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(Universidade Federal de Minas Gerais - Belo Horizonte, MG, Brazil) Orcid: https://orcid.org/0000-0003-2454-681X, Lattes: http://lattes.cnpq.br/8970630770140697

Marcos Raimundo Gomes de Freitas
(Universidade Federal Fluminense - Niterói, RJ, Brazil) Orcid: https://orcid.org/0000-0001-7747-0287, Lattes: http://lattes.cnpq.br/8573989558170120

Maria José Sá
(Universidade do Porto - Porto, Portugal) Orcid: https://orcid.org/0000-0003-0825-2977

Maria Lucia Brito Ferreira
(Hospital da Restauração - Recife, PE, Brazil) Orcid: https://orcid.org/0000-0002-6136-4612, Lattes: http://lattes.cnpq.br/2363390050117897

Marilisa Mantovani Guerreiro
(Universidade Estadual de Campinas - Campinas, SP, Brazil) Orcid: https://orcid.org/0000-0003-1676-1827, Lattes: http://lattes.cnpq.br/4603444396612055

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Oscar Gershanik
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Osvaldo José Moreira do Nascimento
(Universidade Federal Fluminense - Niterói, RJ, Brazil) Orcid: https://orcid.org/0000-0003-3516-485X, Lattes: http://lattes.cnpq.br/2844917988504720

Osvaldo Massaiti Takayanagui
(Universidade de São Paulo - Ribeirão Preto, SP, Brazil) Orcid: https://orcid.org/0000-0002-8190-0275, Lattes: http://lattes.cnpq.br/4345871330138033

Paulo Caramelli
(Universidade Federal de MG - Belo Horizonte, MG, Brazil) ORCID: https://orcid.org/0000-0002-4786-6990, Lattes: http://lattes.cnpq.br/0807993828734491

Pedro Chaná-Cuevas
(Universidad de Santiago de Chile - Santiago, Chile) Orcid: https://orcid.org/0000-0001-9946-3299

Raimundo Pereira da Silva Neto
(Universidade Federal do Piauí - Teresina, PI, Brazil) Orcid: https://orcid.org/0000-0002-2343-9679, Lattes: http://lattes.cnpq.br/4263340418056110

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(Universidade Federal do Estado do Rio de Janeiro - Rio de Janeiro, RJ, Brazil) Orcid: https://orcid.org/0000-0002-1141-8619, Lattes: http://lattes.cnpq.br/4575927827118893

Ricardo Allegri
(Instituto de Investigaciones Neurológicas Dr. Raúl Carrea - Buenos Aires, Argentina) Orcid: https://orcid.org/0000-0001-7166-1234

Ricardo Nitrini
(Universidade de São Paulo - São Paulo, SP, Brazil) Orcid: https://orcid.org/0000-0002-5721-1525, Lattes: http://lattes.cnpq.br/4627329453693645

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(Universidade Federal de Santa Catarina - Florianópolis, SC, Brazil) Orcid: https://orcid.org/0000-0002-9875-6687, Lattes: http://lattes.cnpq.br/4621534549550979

Rubens José Gagliardi
(Santa Casa de São Paulo - São Paulo, SP, Brazil) Orcid: https://orcid.org/0000-0002-3497-0174, Lattes: http://lattes.cnpq.br/1644588288990633

Sérgio Teixeira Ferreira
(Universidade Federal do Rio de Janeiro - Rio de Janeiro, RJ, Brazil) Orcid: https://orcid.org/0000-0001-7160-9866, Lattes: http://lattes.cnpq.br/6999749984515755

Simona Sacco
(University of L'Aquila - L'Aquila AQ, Italy) ORCID: https://orcid.org/0000-0003-0651-1939

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Umbertina Conti Reed
(Universidade de São Paulo - São Paulo, SP, Brazil) Orcid: https://orcid.org/0000-0001-7014-0349, Lattes: http://lattes.cnpq.br/5851667118797836

Vladimir Hachinski
(University of Western Ontario - London, Canada) Orcid: https://orcid.org/0000-0002-0487-734X

Walter Rocca
(Mayo Clinic - Rochester, United States) Orcid: https://orcid.org/0000-0002-1832-7664

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ABSTRACTS

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"De novo" absence status epilepticus: case report with 8-year follow-up

Luma Hussein Salem[1], Felipe Guimarães Kanda[1], Raquel Guimarães Kanda[2], Rafael Guimarães Kanda[3], Paulo Afonso Medeiros Kanda[4]

[1]Universidade de Taubaté, Taubaté SP, Brazil.
[2]Universidade Federal do Cariri, Barbalha CE, Brazil.
[3]Universidade Estadual de Campinas, Campinas SP, Brazil.
[4]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Luma Hussein Salem; Email: lumasalem2345@gmail.com.

Case presentation: A 53-year-old woman went to the emergency room (ER) confused for four days. Lab tests and a CT scan were normal, so, she was referred to a routine psychiatric evaluation. However, her family went to a neurologist, who found a disoriented patient unable to count to 10, perform calculations, remember date or own address. She didn't show focal signs. History of rare seizures in adolescence. The electroencephalogram (EEG) showed continuous, generalized, 3-5 Hz spike-and-wave discharges (SWD). The diagnosis was late-onset absence status epilepticus. Two hours after 5 drops of clonazepam 2.5 mg/ml, she woke up with normal cognition but no recollection of the past week. EEG recorded the next day showed posterior dominant rhythm (PDR) at 7.5 Hz, with brief bursts of fragmented 3 Hz SWD. Upon questioning, she reported having "startles" since adolescence, suggesting idiopathic generalized epilepsy (IGE). Subsequent EEGs on 10/05/2017, 11/21/2018, 02/12/2020, 12/21/2022, and 12/21/2023 had normal background activity with PDR at 10 Hz. All of them showing brief bursts of fragmented, generalized 3-5 Hz SWD. She occasionally has myoclonic seizures but has not had any new absence seizures or convulsions. **Discussion:** Late-onset absence status epilepticus (LO-ASE) is rare and can occur in adults and the elderly as a late relapse of IGE or in de novo patients without a seizure history. De novo LO-ASE is frequently described in women, often triggered by benzodiazepine withdrawal, alcohol or psychotropic drug intoxication, none of which in this history. LO-ASE is characterized by a prolonged and fluctuating confusional state, with or without minor abnormal motor components. The EEG shows various ictal patterns, from typical or atypical generalized spike-and-wave discharges to complex asymmetric irregular sharp and slow waves with anterior predominance. The evolution of our case revealed the persistence of mild and rare morning myoclonic seizures. This LO-ASE likely emerged on a substrate of juvenile myoclonic epilepsy. The eight-year follow-up evidenced the chronic nature of this IGE with a good response to anti-seizure medication. This report provides a detailed view of the clinical and EEG evolution of a rare case of LO-ASE in a middle-aged woman over 8 years period. It emphasizes the need to consider this condition in the differential diagnosis of confusional states in ERs, especially in patients with history of seizures to prevent complications and improve clinical outcomes in such cases. **Final comments:** This case of absence status epilepticus in a middle-aged patient, highlighs the importance of clinical suspicion of ASE and IGE in ER, and the use of EEG in such diagnosis.

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"De novo" mutations in cadasil: a systematic review

Karoline de Alvarenga Leandro Lobo[1], Saulo Machado Moreira Sousa[2], Ralph Lettieri Manhães[1], Vanderson Carvalho Neri[1]

[1]Faculdade de Medicina de Campos, Campos dos Goytacazes RJ, Brazil.
[2]Universidade Estadual do Norte Fluminense Darcy Ribeiro, Campos dos Goytacazes RJ, Brazil.

Correspondence: Karoline de Alvarenga Leandro Lobo; Email: alvarengalobo@gmail.com.

Background: CADASIL is a genetic disease caused by mutations in the NOTCH3 gene. While it is traditionally considered hereditary, recent research indicates the presence of "de novo," mutations. **Objective:** To investigate the latest discoveries regarding "de novo" mutations in CADASIL. **Methods:** A comprehensive systematic review was conducted using: PubMed, Google Scholar, BVS, Jstage, KoreaMed, IMSEAR, MJM, Mycite, WPRIM, LILACS, SciELO, Cochrane Library, and MDPI. Only indexed journals published between 2000 and 2022 were analyzed, resulting in a selection of 41 articles. Duplicated papers (12), non-original articles (3), non-English publications (2), studies not focused on CADASIL and "de novo" mutations (15), and those lacking confirmed mutations (1) were excluded. **Results:** This systematic review presents a collection of case studies concerning non-hereditary NOTCH3 cysteine-altering variants. The described mutations included a heterozygous missense variant NOTCH3 c.1320C> G p.(Cys440Trp) in exon 8, and two heterozygous missense mutations in exon 4 (cys128 to gly and a C-to-T mutation at nucleotide 622). No significant differences in gender prevalence were observed. The mean age of symptom onset was 30.6 ± 9.8 years. Although CADASIL typically manifests between the ages of 10 and 50, the mean age of symptom onset for common hereditary NOTCH3 mutations was 36.7±12.9 years, suggesting a possible anticipation of symptoms in these variant cases. The youngest patient was 15 years old, while the oldest was 40 years old. Migraine was present in 89% of the patients at symptom onset, with 66% experiencing speech changes. However, only 33% reported ocular symptoms, and 22% reported fatigue/asthenia and/or facial asymmetry. In terms of psychomotor and cognitive domains, 33% exhibited psychomotor slowness, and 22% showed changes in concentration, attention, memory, mood, and executive functions, with or without mental confusion. Additionally, 33% presented hemiplegia/paresis. Imaging investigations revealed bilateral white matter lesions in all patients. Parietal lobe lesions were observed in 89% of cases, while 56% exhibited lesions in the temporal lobe and/or basal ganglia, and 33% had periventricular lesions. No significant changes were observed in other regions. No explicit correlations were found between CADASIL and pre-existing comorbidities unless a SARS-CoV-2 infection. **Conclusion:** Although CADASIL is recognized as an hereditary disease caused by mutations in the NOTCH3 gene, "de novo" mutations variants have increasingly been identified as responsible for CADASIL. This review provides insights into "de novo" mutations in CADASIL. These findings emphasize the importance of investigating non-inherited factors and triggering events in CADASIL, because they may contribute to reducing the risk factors associated with this condition.

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Effect of the anti-inflammatory action of β-carbolines on SH-SY5Y cell line (human neuroblastoma) and modulation of TMPRSS11 and Furin receptors

Laura Seixas Bianco[1], Alessandra Linardi[1], Roberta Sessa Stilhano[1]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.

Correspondence: Laura Seixas Bianco; Email: laura.bianco@aluno.fcmsantacasasp.edu.br.

Background: Harmine and harmaline are β-carbolines found in the Banisteriopsis caapi, a vine found in the Amazon rainforest and used with Psychotria viridis to prepare Ayahuasca. β-carbolines are potent MAO-A enzyme inhibitors. Literature has suggested their therapeutic potential due to their immunomodulatory, anti-inflammatory, antidepressant and anxiolytics action. Harmine and harmaline promote neurogenesis in neurospheres and can stimulate the proliferation, migration and differentiation of neural stem cells to adult neurons, indicating possible therapeutic benefits. The SH-SY5Y cell line (human neuroblastoma) is commonly used in neuronal, biochemical and functional properties and as a model for neuroinflammation. High levels of pro-inflammatory cytokines, such as TNF-α and IL-6, are found in COVID-19 patients. Despite the blood-brain barrier, SARS-CoV-2 can cross it and trigger an excessive immune response, releasing cytokines in the CNS, potentially worsening the neurological symptoms of COVID-19, referred to as neuro-COVID. **Objective:** To evaluate the potential anti-inflammatory effects of β-carbolines on the SH-SY5Y cell line and their modulation of TMPRSS11 and Furin receptors, which facilitate viral entry into neuronal cells. **Methods:** β-carbolines (Sigma-Aldrich) were prepared with DMSO and stored at -20°C. SH-SY5Y were cultured in DMEN/F12 supplemented with 10% fetal bovine serum and 1% penicillin and streptomycin. Neuroinflammation was induced by treating cells with LPS (130 ug/mL) for 48 hours. Harmine and harmaline (3-30 μM) were added 24 hours before LPS. **Results:** Harmine (30 μM) reduced basal IL-6 (25 pg/mL) and LPS-induced IL-6 (54.7 pg/mL) to zero. Harmaline (3 μM) significantly reduced basal IL-6 to 2.7 pg/mL and LPS-induced IL-6 to 26.7 pg/mL. Harmine also reduced basal TNF-α (43.1 pg/mL) and LPS-induced TNF-α (103 pg/mL) to practically zero, while harmaline (3 μM) significantly reduced basal TNF-α to zero and LPS-induced TNF-α to 56.6 pg/mL. However, there was no change in the expression of TMPRSS11 and Furin receptors, both in cells induced by LPS or basal and in cells treated by β-carbolines. **Conclusion:** β-carbolines reduced the production of both basal and LPS-induced inflammatory cytokines in the SH-SY5Y cell line, suggesting a potential neuroprotective role in neuroinflammation-induced disorders. Although there were no significant effects on the expression of SARS-CoV-2 receptors, the compounds still show promise in preventing neuro-COVID symptoms caused by neuroinflammation.

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Eyes: a window to the cerebellum

Emanuelly Victória Andrade Fonseca[1], Luma Santos de Oliveira[1], Fabio Victor Vieira Rocha[1], Daniel Mariani Favalessa[1], Igor Flavio Gonçalves do Vale[1], Melissa dos Santos Matos[1], Renann Nunes Pirola[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Emanuelly Victória Andrade Fonseca; Email: mn.andradeduarte@hotmail.com.

Case presentation: A 28-year-old male patient presented with ataxia at the age 1 year and 2 months and never acquired unsupported gait. He currently only gait with bilateral support. He denied other comorbidities and a similar family history. He had suffered a fall from his own height, associated with head trauma, with no warning signs. On examination, he had ocular telangiectasias, atrophy of the interosseous muscles, more prominent on the left, abolished osteotendinous reflexes, grade IV global muscle strength, bilateral facial choreo-dystonia, more pronounced on the left, severe bilateral ataxia, and was in a wheelchair. Facial mimicry and extrinsic ocular muscles were preserved. Evoked nystagmus, slowed saccades and slurred speech were observed. Computed tomography of the skull revealed marked cerebellar atrophy, most evident in the cerebellar vermis, and ectasia of the IV ventricle. After genetic testing, the diagnosis of Ataxia-telangiectasia was confirmed. Laboratory tests showed leukocytosis of 16,230/mm3 and platelet count of 502,000/mm3. **Discussion:** Louis Barr Syndrome or

Ataxia-telangiectasia (A-T) is a rare autosomal recessive disorder, with an incidence of 1:100,000, which usually begins in the first 5 years of life. Its etiology is related to mutations in the ATM gene, whose encoded protein plays a crucial role in cell signaling in response to DNA damage, oxidative stress and genotoxic agents. Patients present with cerebellar degeneration, telangiectasia, immunodeficiency, sensitivity to radiation and a tendency to neoplasms, especially lymphoid ones. In the case in question, TA was suspected due to the presence of neurological signs, such as cerebellar ataxia since childhood, postural instability, fine motor incoordination, abnormal eye movements, involuntary movements, loss of osteotendinous reflexes, ocular telangiectasia and motor neuropathy. In addition, AT can also manifest with sensory neuropathy, cognitive disorders, cutaneous telangiectasia, repetitive sinopulmonary infections, immunoglobulin deficiency, lymphopenia or leukocytosis, and increased alpha-fetoprotein. Nuclear Magnetic Resonance often reveals cerebellar degeneration with atrophy of the vermis and its hemispheres. It is therefore essential to consider AT in any ataxic child when other diagnoses are unlikely. Diagnosis is based on clinical assessment, laboratory tests and genetic testing. Treatment is symptomatic, as there are still no therapies that prevent or slow down neurodegeneration. **Final comments:** The diagnosis of A-T involves clinical data, laboratory tests and genetic testing. Continuous surveillance and early interventions are essential due to immunodepression, marked by low levels of IgA, IgM and IgG production in response to vaccines and infections, generating increased susceptibility to diseases such as recurrent pneumonia, bronchiectasis, interstitial lung disease and cancer. Multidisciplinary care is essential to improve patients' life qualities.

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Management of uncontrolled epileptic seizures in the emergency setting in the southeast region from 2013 to 2023

Letícia Dardes Barbosa[1],Ana Percya Sais da Rocha[2],Jamile Rodrigues Cosme de Holanda[3],Lara Schiavinato Merloto[4],Maiana Lourenço Benvindo da Rocha Martins[5]

[1]Centro Universitário Barão de Mauá, Ribeirão Preto SP, Brazil.
[2]Universidad Central del Paraguay, Ciudad del Este, Praguay.
[3]Universidade Federal de Sergipe, São Cristóvão SE, Brazil.
[4]Universidade Anhembi Morumbi, São Paulo SP, Brazil.
[5]Centro Universitário Novafapi; Teresina PI, Brazil.

Correspondence: Letícia Dardes Barbosa; Email: leticiadardes@hotmail.com.

Background: The rate of untreated epileptic seizures in the Southeast region has increased in recent years, posing a threat to the population, as death can be a consequence. Given that epileptic seizures are sudden and temporary events caused by abnormal electrical activity in the brain, resulting in convulsions, changes in behavior, or unusual sensations, there is a need for prompt emergency team control to avoid gaps, which were not observed between 2013 and 2023. **Objective:** To conduct an epidemiological analysis of the treatment of uncontrolled epileptic seizures as urgent care for the inhabitants of the Southeast region. **Methods:** It consists of an ecological study involving data collection through the Hospital Information System (SIH/SUS). The study population consisted of individuals from the Southeast Region who received treatment for uncontrolled epileptic seizures urgently during the period from 2013 to 2023. The variables analyzed included the number of hospitalizations, average length of stay, and deaths. Data were collected using descriptive statistical analysis techniques. **Results:** During the period from 2013 to 2023, a total of 195,911 cases were reported in the Southeast region. The year 2022 recorded the highest number of hospitalized patients, with 20,902 cases (10.66%), followed by the year 2023 with 19,915 (10.16%), while the year 2020 had the lowest, with 16,475 cases (8.40%). Regarding the average length of stay, it was 5.5 days over the decade. In 2022, the longest stay was recorded, with an average of 5.9 days, while in 2014, the shortest was registered, with an average of 5.0 days. During this period, a total of 6,306 deaths were reported, accounting for 3.21% of the affected population, with the year 2023 recording the highest number of notifications, with 714 episodes (11.32%), and the year 2022 with 707 occurrences (11.21%), while in 2014, there was the lowest number, with 425 episodes (6.73%). **Conclusion:** The prevalence of untreated epileptic seizures is evident, underscoring the need to understand the etiology and classification for better management. It is crucial to control them to improve the patient's routine and habits, in order to address the increasing number of hospitalizations and deaths observed during the studied years (2013/2023). Finally, an initiative from the Ministry of Health (MS) to constantly monitor the patient, while noting the particularities observed in each case, is necessary to achieve a satisfactory holistic response to the patient's health.

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"Split-hand like" syndrom in X-linked form of Charcot-Marie-Tooth disease (CMTX1) caused by GJB1 mutation: a comparative study of eletrophisiologic features

Ricardo Vergara Gianvechio[1], Wilson Marques Junior[1], Pedro José Tomaselli[1], Lucas Gondim Briand Vieira[1], Brenda Cristina Nava[2], Gabriela Rodrigues Ramos[2]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Ricardo Vergara Gianvechio; Email: ricardo.gianvechio@alumni.usp.br.

Background: The X-linked form of Charcot-Marie-Tooth disease (CMTX1) is a common form of inherited neuropathy. It's caused by mutations in the GJB1 gene. Some reports in the literature suggest that a "split hand syndrome-like" condition can be found in patients with CMTX1, characterized by more prominent atrophy in the abductor pollicis brevis (APB) than in the abductor digiti minimi (ADM). Nonetheless, we found no studies numerically describing this possible predilection by means of neurophysiologic parameters. **Objective:** This retrospective study was conducted to objectively evaluate the possible predilection of involvement of the APB (in detriment of ADM) in CMTX1, comparing them to CMT1A patients and to control patients without evidence of peripheral neuropathy and with normal nerve conduction studies (NCS). **Methods:** We analyzed the data from electroneuromyography of 59 patients (21 patients with GJB1 mutations, 26 patients with PMP22 duplication - CMT1A group, and 12 patients without peripheral neuropathy and with normal values of NCS). The epidemiological and clinical baseline characteristics were also collected for comparison. The mean age of the patients undergoing electroneuromyography was 36.1 years in the CMT1X group and 35.5 years in the CMT1A group. The ratio APB/ADM (the ratio between the CMAPs in the APB and in the ADM, respectively) was calculated in each patient. In patients where it was possible to calculate the ratio from both hands, we conventionally used the smaller ratio. We then compared the results from the three groups using a two-tailed Student's t-test. **Results:** The mean APB/ADM in the CMT1X group was 0.646 with a standard deviation of 0.444, while in the CMT1A group, the mean APB/ADM was 1.286 with a standard deviation of 0.578. The mean APB/ADM ratio was significantly lower in patients in the CMTX1 group compared to patients in the CMT1A group (p-value=0.0001) and to patients in the control group (p-value=0.04). There was no statistical difference between the CMT1A group and the control group (p=0.06). **Conclusion:** Although this is a preliminary study, the results suggest a significant predilection for more pronounced atrophy in the APB compared to the abductor digiti minimi ADM in patients with CMTX1. This finding was quantitatively supported by the significantly lower APB/ADM ratios in the CMT1X group compared to both the CMT1A and control groups. The absence of a significant difference between the CMT1A and control groups suggests that this phenomenon is specific to CMTX1. This could be useful to generate hypotheses surrounding the pathophysiology of the disease. Further prospective and/or longitudinal studies should be conducted to evaluate the reproducibility of these results in larger samples and to determine the predictive values of the clinical and neurophysiological findings of "split hand-like syndrome," and the APB/ADM ratio in patients with suspected inherited neuropathies.

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Ataxic sensory ganglionopathy secondary to Sjogren's: a case report

Ana Clara Rodrigues Santos[1], Lucas Porto Ferreira[1], Luiza Serafini Balestrassi[1],Joselene Sales Costa[1], Tamiris Araújo Silva[1], Augusto Cesar Marrafon[1], Franciane Couto Soares[1], João Batista Macedo Vianna[1]

[1]Hospital de Clínicas de Itajubá, Itajubá MG, Brazil.

Correspondence: Ana Clara Rodrigues Santos; Email: anaclaraa.14@hotmail.com.

Case presentation: Patient JRAS, at the age of 45, began suffering from paresthesia in the hands bilaterally, which progressed to the lower limbs, and in three weeks he developed difficulty in walking and tetraparesis. She sought care at the emergency room in her city of origin, with a clinical hypothesis of Guillain Barret Syndrome, where an outpatient electroneuromyography was requested. The examination, carried out in March 2019, showed purely sensory neurogenic impairment and an axonal pattern (lack of response in all nerves studied) in the upper and lower limbs, without changes in the motor nerves, nor signs of conduction block, thus suggesting, ganglionopathy or marked sensory axonal polyneuropathy. Subsequently, during the etiological investigation, the Schirmer test was moderate and severe, and the Rose Bengal Test showed impregnation on the eyelid margin. With the diagnosis of Sjogren's syndrome, therapy was instituted with 3 cycles of pulse therapy during a quarter and symptomatic therapy, such as Gabapentin, with a favorable response. **Discussion:** Sjogren's syndrome is a chronic autoimmune disease that results in inflammation of the exocrine glands, with keratoconjunctivitis sicca and xerostomia being the common manifestations. The extra glandular presentation is multisystemic and may present neurological manifestations, with involvement of the central and peripheral nervous system. Among neurological manifestations, peripheral neuropathy is the most common complication and can be found in 25% of patients. Symptoms may be associated with small fiber neuropathy, mononeuropathy multiplex, cranial neuropathy and, more recently, sensory ataxic neuronopathy associated with immunological attack on the dorsal root ganglion. Sensory ganglionopathies are a subgroup of peripheral neuropathies whose common feature is primary damage to the cells of the bodies of neurons located in the dorsal root ganglia. Among the complications associated with the peripheral nervous system, sensory ataxic ganglionopathy is a rare and severe form that affects less than 5% of patients with Sjogren's. **Final comments:** This case report aims to explore the

characteristics of ataxic sensory neuronopathy, which presents clinically with changes in proprioception, ataxic gait, pseudoathetosis, positive Romberg test and changes in thermal and pain sensitivity. Ganglionopathies are considered rare diseases, whose error and delayed diagnosis are commonly relevant problems, which impacts patient care and can cause irreversible damage with severe functional impact.

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"My brain Robbie": translation to Portuguese to promote a healthy brain

Vivian Proença Lara[1], Glaucia Muniz Proença Lara[2], Eléonore Bayen[3]

[1]Faculdade de Ciências Médicas de Minas Gerais, Belo Horizonte MG, Brazil.
[2]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[3]Global Brain Health Institute/Sorbonne Université, Hôpital de La Pitié-Salpêtrière, Paris, France.

Correspondence: Vivian Proença Lara; Email: vi.prlara@gmail.com.

Background: Scientific research in Neurology and in Epidemiology has shown that early prevention and a healthy lifestyle can mitigate the risk that chronic diseases, including dementia, will develop. A character called "Robbie" was created and a video based on scientific knowledge about brain health prevention was produced as a fun and engaging narrative. Robbie, the brain, explains to us what we can do every day to keep our brain in good shape. This French initiative is a creative way to raise awareness and to promote a better understanding of brain health in children aged 6 to 12 and more broadly to increase awareness among the general public about the importance of brain health for all age groups and in different languages. **Objective:** The goal of this project is to empower children (as well as teenagers, adults and seniors) to keep their brains healthy throughout their lives by providing them with simple public health messages. **Methods:** The video was released in 01/10/18 and was later translated from French into Brazilian Portuguese as "Um cérebro saudável!". The translation was performed during the Alzheimer's Association International Conference on 12/04/2019, in São Paulo. The text was revised by a portuguese teacher. Robbie's voice was interpreted in a simple recorder. The final portuguese video lasts approximately 7 minutes and can be broadcast in different environments, including schools. To support public health interventions on brain health in primary and secondary schools, an educational kit that complements the project was created and translated. It contains activity sheets that can be used by teachers, students and volunteers. The video and the educational material are free to access and cannot be used for commercial purposes. The video is distributed via the YouTube platform (channel @mybrainrobbie1810) and the website https://mybrain-robbie.org. The video is also avaliable in English, Spanish, Mandarin, Greek, Turkish, Slovenian and Lithuanian. On 15/01/2024 a second video was released: "An inclusive world", now available in English and French with a planned translation into Portuguese and other languages soon. **Results:** The eight healthy habits presented in the video that should be observed every day throughout life are: 1. Learning; 2. Exercise; 3. Avoid blows to the head; 4. Have a good diet; 5. Avoid dangerous substances; 6. Sleep well; 7. Taking care of your health; 8. Having time for friends and family. Each one of them is shown and explained by Robbie in the video. Until 26/05/2024 the video was seen 186.518 times in YouTube, in different languages (including Portuguese), in many countries. **Conclusion:** This is as a fun and simple effort to promote global health and also to reduce inequalities all over the world. The eight good brain habits mentioned by Robbie correspond to risk factors for cognitive decline and dementia that can be modified.

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"Rede ELA": a Brazilian initiative on transdisciplinary and interinstitutional research on amyotrophic lateral sclerosis (ALS)

Luna Lila Fernandes de Faria[1], Julia Costa Nacif Santos[1], Maria Goreti de Almeida Oliveira[1], Tiago Antônio de Oliveira Mendes[1], Ricardo Luis de Sousa[1], Clodoaldo Irineu Levartoski de Araújo[1], Lucas Vilas Boas Magalhães[1]

[1]Universidade Federal de Viçosa, Viçosa MG, Brazil.

Correspondence: Lucas Vilas Bôas Magalhães; Email: lvbmagalhaes@ufv.br.

Background: Amyotrophic Lateral Sclerosis (ALS) is a heterogeneous group of fatal neurodegenerative diseases, characterized by involvement of the upper and lower motor neurons in the spinal and bulbar pathways. About 90% of ALS cases are sporadic, with a polygenic pattern, while the remaining 10% comprise monogenic forms, implicated in the development of familial ALS (fALS). The mutation of VAPB gene is associated with the majority of fALS cases in Brazil, but it has an almost insignificant prevalence in developed countries, leading to limited research on the disease caused by this variant. This fact reinforces the need for studies that explore the particularities of ALS in Brazil. **Objective:** To establish a transdisciplinary and interinstitutional research network, in order to expand and integrate knowledge regarding ALS in its multiple dimensions: clinical, diagnostic, therapeutic, epidemiologic, genetic, biochemical and cellular. **Methods:** To establish our network, we initially counted on virtual platforms to recruit professionals from different areas and institutions. In sequence, a questionnaire was used to select and better access each participant's areas of research and potential contributions to the net. Based on this information, 9 working groups were established with defined lines of study, including epidemiology, diagnosis, treatment, alternative therapies and administrative support, each with it's own coordinator. In order to compile knowledge on the topic, define gaps in scientific literature and develop original research projects, weekly meetings took place in each group. Meanwhile, monthly reunions were organized to update the network on the projects. **Results:** The network got its accreditation on 05/10/2024, and has been able to gather 24 researchers from different Brazilian and international institutions, from fields that include medicine, physics and biochemistry. To this moment, there are 12 research projects ideas in process of being developed/registered on cellular and animal ALS models, epidemiology, diagnostic tools and interventions, including stem-cells studies, immunotherapy, and alternative medicine. Furthermore, from the net's foundation, there are ALS patients actively participating in its administrative aspects, which provides the organization a unique perception of the disease's impacts. **Conclusion:** ALS is a complex multifactorial disease, which results in a need for an interdisciplinary approach, both on patient care and on the research perspective of the disease. Our project has given the first steps towards systematically expanding and integrating knowledge on the disease. Despite its advances, "Rede ELA" is still in its early stages and it's our belief that the continuous expansion of contributors from different areas and institutions is essential to its progress and could lead to significant impact on knowledge about ALS.

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"Rete collaterals" at callosomarginal artery: a rare case report

Angelo Ponte de Freitas Campos[1], Rodrigo de Paiva Bezerra[1], Maurício Elias Nunes da Silva[1], Marcel Ken Uehara[1], Lívia Almeida Dutra[1], Gisele Sampaio Silva[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Angelo Ponte de Freitas Campos; Email: campos_angelo@hotmail.com.

Case presentation: We describe the case of a 32-year-old female patient with a history of episodic migraine without aura. She reported a new headache episode that began in the morning, strong intensity, pulsating type, associated with dizziness, nausea, photophobia, and phonophobia. The pain was localized in the right frontal region with progressive evolution, without thunderclap characteristics, but worsened with postural changes. Neurologic examination was unremarkable. She was admitted for diagnostic elucidation and underwent a brain MRI, which showed a nodular cortical/juxtacortical formation in the right frontal region with peripheral blood residues, closely related to the right callosomarginal artery, of vascular nature, suggesting differential diagnoses of pseudoaneurysm or arteriovenous malformation/fistula. Further investigation with cerebral angiography revealed a nodular formation originating from the left callosomarginal artery in the projection of the superior frontal gyrus, characterized by a network of tortuous arterial vessels from which the anterior and middle internal frontal arteries originate. This is likely a remnant of an embryological artery called "Rete mirabile." She didn't undergo a neurosurgery procedure at that time, due to lack of hemorrhage or aneurysm. She was discharged with guidance for analgesia for episodic migraines and scheduled a follow-up and radiological monitoring in three months. **Discussion:** Anatomical variations called "rete-like or rete mirabile" are a rare group of vascular abnormalities. They consist of a complex arterial network primarily found in some mammals such as cows, goats, pigs, and cats. "Rete-like" variations are rare in humans, with an estimated prevalence of 0.01%, and the Asian population seems to be more affected. When present, they are most frequently found in the carotid vascular territory, the middle cerebral arteries, and less commonly described in the posterior circulation. Subarachnoid hemorrhage is the most common clinical presentation (34.2%), usually secondary to aneurysms formed by vessel wall fragility, followed by incidental diagnoses (20%). When indicated, treating aneurysms secondary to rete-like formations can be difficult. Expectant management and serial radiological follow-up are considered in patients with incidental diagnoses. **Final comments:** Despite being an extremely rare variation, they can cause intracranial hemorrhages secondary to aneurysm rupture. High clinical suspicion coupled with selective cerebral angiography is necessary for a correct diagnosis since even MRI can sometimes make a diagnostic mistake. Furthermore, further studies are needed to define the best therapeutic option considering the topography, dimensions, and other epidemiological characteristics of patients.

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18q21,2q23 deletion syndrome associated with agenesis of the corpus callosum

Jessica Vargas Lopes[1], Caroline Dresch Sabadin[2], Laís Machado de Oliveira[3], Rhanna Carolina de Oliveira[3], Vitor Zubeldia[3], Júlia Mascarello[3], Ana Paula Schuur[3], Juliana Dourado Grzesiuk[3], Thyara Boechat[4]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade Estadual do Oeste do Paraná, Cascavel PR, Brazil.
[3]Centro Universitário Assis Gurgacz, Cascavel PR, Brazil.
[4]Dra. Thyara Boechat Neuropeditaria, Cascavel PR, Brazil.

Correspondence: Jessica Vargas Lopes; Email: jessicavargasl@outlook.com.

Case presentation: A female patient, aged 1 year and 1 month, born by cesarean section with a birth weight of 2905 gr. and an Apgar score of 8. She was admitted to the Neonatal Intensive Care Unit (NICU) following birth due to cardiorespiratory arrest, seizures and neonatal sepsis, with a diagnosis of congenital cytomegalovirus infection and epilepsy. At physical examination she presented with axial hypotonia, asymmetrical spontaneous movements, appendicular hypertonia, inability to follow visual objects and bilateral horizontal nystagmus. Electroencephalography (EEG) indicated disorganized brain activity with hemispheric asymmetries, in addition to a fast of medium to high voltage in the central and anterior temporal regions. The speech therapy evaluation revealed hypotonia of speech articulatory structures, and lack of coordination between the tongue, sucking, breathing and swallowing. Cranial magnetic resonance imaging (MRI) showed agenesis of the corpus callosum. A G-band karyotype revealed a partial deletion of the long arm of chromosome 18 present in all the 20 metaphases analyzed. The genomic microarray test identified the position of the terminal chromosomal alteration as 18q21,2q23. The deletion involved a 24.4 Mb segment and was present in most cells (mosaic). An intervention plan was established with a multidisciplinary team, aiming to prevent and mitigate the signs and symptoms of global developmental delay presented by the patient. **Discussion:** Chromosome 18q deletion syndrome (OMIM 601808) is a rare chromosomal disease and most case reports present common findings as impaired intellectual disability, microcephaly, short stature, congenital aural atresia, hearing loss, ophthalmological abnormalities, palatal defects, short frenulum, foot deformities, hypotonia and delayed myelination, in addition to cardiac anomalies. There are a few studies reporting individuals affected with chromosome 18q deletion having neuroradiological findings as smaller corpus callosum, therefore, the present case is one of the rare cases associated with agenesis of the corpus callosum, a congenital malformation which could also contribute to the developmental delay found in the patient. The deletion detected in the reported patient involves 70 coding genes, and two of them are possibly involved with corpus callosum malformations: RTTN and NEDD4L. As alterations in NEDD4L gene have a dominant inheritance it may play an important role in corpus callosum malformations. **Final Comments:** Chromosome 18q deletion syndrome has a low incidence and a high phenotype variability. Axial hypotonia and bilateral horizontal nystagmus were the main findings in the reported patient that are commonly associated with 18q deletions. However, her agenesis of the corpus callosum is a highly rare pathology. Few reports discuss this association, therefore, it is encouraged that more research be done, considering the possible correlation of both disorders, such as observed in this case.

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2 siblings with autosomal dominant Alzheimer's disease secondary to PSEN1 gene mutation: a case report

João Vítor Moreira Nogueira[1], Oliver Reiks Miyajima[2], Alessandra Marangoni Fante[1], Bruna Barbosa Nobre[1], Enzo Lima Alcântara Parente[1], Daniela Vieira Cavalcante[1], Marcelo de Almeida Pinheiro[1]

[1]Unichristus, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: João Vítor Moreira Nogueira; Email: jvmnogueira10@gmail.com.

Case presentation: This study is a report of 2 individuals from the same family who were being followed up as outpatients in a neurology department of a tertiary hospital. A 45-year-old woman presented with amnestic symptoms that began at the age of 36 and progressed to functional impairment, aphasia, social isolation, sphincter dysfunction, visual hallucinations, and significant irritability. Cranial magnetic resonance imaging (MRI) showed signs of diffuse cortico-subcortical atrophy, in addition to discrete hippocampal atrophy, findings that were not expected for her age. Her brother, a 51-year-old man, had amnestic symptoms onset at the age of 45, progressing to irritability, mood swings, and loss of functionality. Cranial MRI showed findings similar to those of his sister. In the family history, similar symptoms were observed in other members, including the grandmother, father, and two sisters, all of whom are now deceased. Sequential genetic analysis of both individuals revealed the presence of a pathogenic mutation in the PSEN1 gene (c552A> C [p.Glu184Asp]), confirming the diagnosis of Autosomal Dominant Alzheimer's Disease (ADAD). **Discussion:** The PSEN1 gene, located on chromosome 14, encodes a protein with the same name that acts as a transmembrane molecule, performing several functions of neurological importance. Its mutation can result in several alterations, including modifications in amyloid metabolism and neurogenesis. Mutations in this gene are the main cause of ADAD, accounting for approximately 50% of cases. In the case report in question, considering that the sister's exome confirmed the existence of the aforementioned mutation, there was an indication to perform genetic screening on all of the patient's first-degree relatives, where her brother would be diagnosed early, increasing the chance of trying to reduce the progression of the disease. Comparative studies between ADAD and late-onset sporadic Alzheimer's disease (DAEIT) indicate that ADAD tends to have presenile onset of symptoms (before age 65), with a more rapid and severe clinical progression, in addition to the presence of cortical symptoms, such as aphasia, and greater loss of hippocampal volume, when compared to DAEIT, which tends to have a more indolent course and without the evident presence of cortical symptoms. Early diagnosis of ADAD in older family members can facilitate management and improve long term outcome and quality of life in younger family members that have ADAD without symptom onset. **Final comments:** It is concluded that these cases highlight the importance of early genetic investigation in cases of presenile onset Alzheimer's disease, especially when associated with a family history of first-degree relatives with similar symptoms. This leads to an accurate and early diagnosis of the disease, which can improve management of the pathology in other family members, in addition to improving quality of life and allowing appropriate genetic counseling.

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80 years of Leão's cortical spreading depression: a historical overview of its impact on neurology

José Antonio Vieira da Silva Calsoni[1], Sofia Gameiro de Souza[1], Raimundo Marcial de Brito Neto[1], Ana Luiza Morgado Costa[1], Rodrigo Amorim Jorge[2], Cristiana Pessoa de Queiroz Faria Goes[3], Jano Alves de Souza[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.
[2]Unigranrio, Duque de Caxias RJ, Brazil.
[3]Universidade Estadual do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: José Antonio Vieira da Silva Calsoni; Email: calsoni.jose@gmail.com.

Background: Aristides Azevedo Pacheco Leão (1914-1993) was born in Rio de Janeiro and entered the medical school at the University of São Paulo. When he was in his 2nd year, he contracted tuberculosis and had to interrupt his course. Although cured he never went back to medical school. Determined to be a scientist, he was accepted at Harvard University (1941), earning his master's degree (1942) and PhD (1943). Then, he became an assistant researcher at the Department of Anatomy at Harvard, where he was able to identify the phenomenon of Cortical Spreading Depression (CSD), which this year marks the 80th anniversary of its publication in the Journal of Neurophysiology. Its discovery by Aristides Leão ushered in a new era of understanding in neurology. This phenomenon, characterized by a wave of neuronal depolarization followed by prolonged suppression of cortical activity, revolutionized the comprehension of migraine pathophysiology, and other neurological disorders. **Objective:** Provide a comprehensive exploration of the historical significance and enduring impacts of Aristides Leão's discovery of cortical spreading depression over the past 80 years. **Methods:** A critical and objective narrative review of literature, including Leão's original research publications, historical archives, and contemporary bibliography. Through a analysis of these sources, we aim to construct a coherent narrative elucidating the evolution of the knowledge surrounding CSD. **Results:** Leão's discovery of CSD in 1944 fundamentally transformed the landscape of neurology. Initially observed in experimental rabbit models, the phenomenon was first described as being related to epilepsy and then there were several studies on this matter. Soon CSD's implications extended to other human conditions like migraine. In 2001, Hadjikhani et al. used fMRI to demonstrate that migraine aura is associated with the propagation of CSD in the visual cortex. Recent studies evaluate the phenomenon of CSD in many aspects such as: spreading depression and cortical hemodynamics; how estrogen and stimulation of the vagus nerve modulate CSD; and the characteristics of CSD in sleep and wake states. Now, non-invasive methods to detect spreading depression is awaited in clinical research. From elucidating the pathophysiology of migraine to informing approaches for other neurological disorders, the impact of Leão's discovery echoes across neurology and his work laid the foundation for future research, inspiring methodological innovations and theories that still influence neuroscience. **Conclusion:** The description of CSD stands as a testament to the transformative power of scientific inquiry. Over the past 80 years, Leão's work has catalyzed paradigm shifts in our understanding of neurological phenomena, paving the way for innovative diagnostic and therapeutic strategies. As we continue to unravel the intricacies of CSD, Leão's legacy endures as an inspiration for future generations of neuroscientists.

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A brain connectivity study associated with peritraumatic dissociation (PTD) in women with posttraumatic stress disorder (PTSD)

Giovana Fontanella Pileggi[1], Raymundo Machado de Azevedo Neto[1], Andrea de Abreu Feijó de Mello[2], Andrea Parolin Jackowski[2], Ana Carolina Milani[2], Bruno Messina Coimbra[2], Marcelo Feijó de Mello[1]

[1]Hospital Israelita Albert Einstein São Paulo, São Paulo SP, Brazil.
[2]Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Giovana Fontanella Pileggi; Email: giovana.pileggi@gmail.com.

Sexual violence against women, including sexual assault, is endemic, and more than 50% of the victims will develop PTSD. A PTD could happen when the violence is happening, being a marker of a worse outcome and worse response to treatment. Functional magnetic resonance (fMRI) is a tool to study brain activity and connectivity using the resting state. However, studies of PTD were scarce. To compare the connectivity between brain areas using the resting state in women with diagnostic PTSD after sexual assault, that present or not PTD, and health controls. We executed seed-to-voxel analysis in target brain areas related to PTSD studies in literature: hippocampus, amygdala, para-hippocampus, insula, and Default Mode Network (DMN) areas, using the CONN software. We found significant differences between PTSD patients with or without PTD. The women with PTD showed increased connectivity between the DMN average and the right and

left frontal cortex, right areas of the cerebellum, right precentral gyrus, lateral areas of the occipital lobe, superior frontal gyrus, precuneus, and supramarginal gyrus. The patients with PTD also showed increased connectivity between the insulae and angular and supramarginal gyrus, lateral occipital cortex and right parietal lobule. Furthermore, increased connectivity was found between the parahippocampus and the supramarginal and angular gyrus, cerebellum, and occipital cortex. On the other hand, women with PTD showed decreased connectivity between the DMN (RLP) and the precuneus, lateral occipital and cuneal cortex. Also, decreased connectivity was found between the insulae and Heschl's, supramarginal, and postcentral gyrus. Finally, decreased connectivity was found between the anterior parahippocampal gyrus and the cerebellum, angular and supramarginal gyrus, and the lateral occipital cortex. These findings suggest that patients with PTD exhibit distinct patterns of brain connectivity, which can be related with their symptoms and experiences. The increased connectivity in the right parietal lobe and lateral occipital cortex suggests heightened sensitivity and intensified interpretation of visual stimuli, potentially linked to re-experiencing the traumatic event. Also, an increased activity in the precuneus and left cuneus indicates intensified retrieval of these traumatic visual perceptions. In addition, reduced activity in the prefrontal cortex and superior frontal gyrus suggests decreased self-reflection and emotional regulation, possibly due to emotional disconnection. The reduced activity in the right precentral gyrus implies a decrease in motor action preparation. These results contribute to a better understanding of the physiological mechanisms associated with PTD on PTSD, which makes it relevant in the development of possible new treatments and further investigations into the implications of PTD in PTSD.

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A Brazilian neuromuscular center experience in diagnosis of rare diseases patients

Erica Nogueira Coelho[1], Laura Alonso Matheus Montouro[1], Eduardo de Paula Estephan[1], João Aris Kouyoumdjian[1], Charles Marques Lourenço[1], Amadeu José Rorigues Queiróz[1], Maria da Penha Ananias Morita[1]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Laura Alonso Matheus Montouro; Email: lauramontouro@hotmail.com.

Background: Rare diseases patients management has been a challenge to Brazilian public healthy providers, especially with regards to etiological diagnosis of genetic disorders in gene therapy era. Difficulties relate mainly to cost and availability of molecular genetic techniques. **Objective:** This study main goal is to share our experience in diagnosis of rare genetic diseases in Neuromuscular Diseases Service of São José do Rio Preto State Medicine School. **Methods:** The 10th edition of International Diseases Classification (CID10) was used to search for patients matching rare diseases codes in the medical data records from 01-Jan-2018 to 15-Apr-2024. The following codes were selected: G11.0, G11.1, G11.2, G11.3, G11.4, G11.8, G11.9, G12.0, G12.1, G12.2, G12.8, G12.9, G23.8, G23.9, G24.1, G24.8, G24.9, G60.0, G60.1, G60.2, G60.8, G60.9, G71.0, G71.1, G71.2 e G71.8. They represent five major groups of rare neurological diseases: Inherited Ataxias/Spastic Hereditary Paraplegia, Motor Neuron Diseases, Dystonic Disorders, Inherited Neuropathies and Primary Inherited Muscular Diseases. Data about those patients and final diagnosis is presented bellow. **Results:** In the Ataxia/Paraplegia group, 179 patients went through evaluation at the Medicine School public outpatient clinic, 68 (38%) of then in our referral Neuromuscular Service, 56 (82,4%) were suspected to have a genetic cause, 25 (36,8%) had access to molecular genetic test and 15 (22,1%) reached the final etiological diagnosis. The genes found were: FXN, ATP1A3, ATXN1, ATXN3, COQ7, SPG11, CACNA1A, ALDH18A1 and SLC16A2. For Motor Neuron Diseases: 197 patients evaluated, 154 (78,2%) by the neuromuscular team, 38 (24,7%) with suspected genetic etiology, 29 (18,8%) tested and 22 (14,3%) with positive results. Described genes: SMN1, SPG11, COQ7, VWA1, TFG and POLG. 13 (9,5%) of a total of 137 patients with dystonic features had appointment in Neuromuscular Diseases Service, 12 (92,3%) with suspected inherited disease, 7 (53,8%) had genetic tests performed and 6 (46,2%) had final diagnosis defined: GCH1, SPR and WDR45. In Inherited Neuropathies group, 339 total patients were evaluated, 251 (74%) in Neuromuscular Service, 55 (21.9%) had the previous diagnosis suspected, 41 (16.3%) tested and 24 (9.6%) confirmed. Following genes: PMP22, FXN, SLC12A6, COQ7, AARS1, GAN, HINT1, LMNA, PRX, SPG11, TFG, TTR, VWA1, DNMT1 and DYNC1H1. Primary Inherited Muscular diseases were suspected in 102 (87.2%) of the 117 patients evaluated, 56 (47.9%) had access to genetic test and 37 (31.00006%) positive results. The genes were: DMD, CAPN3, DYSF, TTN, DMPK, CLCN1, D3Z4, GAA, GMPPB, LAMA2, LMNA, PYGM, RYR1, SGCG, TCAP and VCP. **Conclusion:** Despite genetic disorders may represent an important part of neuromuscular diseases, the access to molecular genetic test is a problem to public healthy services in Brazil, making the final etiological diagnosis a distant reality for the majority of these patients.

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A case of congenital myasthenic syndrome-0,22 responsive to pyridostigmine (so far...)

Wesley Alfredo Gawlinski de Arruda[1], Naiana Posenato[1], Querem Hapuque Zeferini Neves[2], Gabriella Frattari de Araujo Rondon Borges[2], Amanda Martinez Lafetá[2]

[1]Hospital São Vicente de Paulo, Passo Fundo RS, Brazil.
[2]Universidade de Cuiabá, Cuiabá MT, Brazil.

Correspondence: Wesley Alfredo Gawlinski de Arruda; Email: wesley.alfredo@hotmail.com.

Case presentation: A 2-year and 6-month-old girl was referred from her pediatrician due to neurodevelopmental delay. Born by vaginal birth, full-term delivery, with normal metrics at birth and neonatal screening tests within normal limits, she did not require intensive care or resuscitation maneuvers. On physical examination, she was alert, active, and responsive to the examiner, presenting normal growth parameters. However, she had difficulty remaining in the supine position (tended to lateralize), generalized hypotonia (especially cervical), loss of proximal strength, weak palmar and plantar grasp reflexes. Due to presentation as a hypotonic infant, laboratory screening was requested to rule out possible myopathies, brain MRI, and genetic testing for Spinal Muscular Atrophy (SMA), in addition to initiating motor physiotherapy. Initial complementary tests were negative, so an expanded genetic panel was requested, revealing a pathogenic variant chr2:44,328,938 G> A (p.Arg421*) in homozygosis in the PREPL gene (OMIM #609557; 2p21), compatible with a diagnosis of Congenital Myasthenic Syndrome type 22 (CMS22). Subsequently, treatment with pyridostigmine was initiated, resulting in notable clinical improvement and increased speed in developmental milestones acquisition. **Discussion:** Congenital Myasthenic Syndrome (CMS) corresponds to a rare and heterogeneous group of hereditary neuromuscular disorders characterized by involvement of the neuromuscular junction due to autoantibodies acting against Acetylcholine (ACh). It is characterized by muscle weakness, hypotonia, dysphagia, ptosis, facial weakness, and delayed neuropsychomotor development. Among these disorders, CMS-22 is a rare recessive genetic disease caused by mutations in the PREPL gene, which encodes the protein Prolyl Endopeptidase-Like (PREPL), involved in vesicular transport and filling synaptic vesicles with acetylcholine and other neurotransmitters. Symptoms are generally moderate and often respond to pyridostigmine, particularly during childhood. **Final comments:** Although rare, congenital myasthenic syndromes should be suspected in children with hypotonia suggestive of peripheral nervous system disorders, with genetic testing being essential for diagnosis.

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A case of refractory epileptic seizures induced by glufosinate ammonium poisoning

Michelle Zonkowski Ribas[1], Gustavo Zonkowski Ribas[1], Rodrigo Fadel Simão[1], Ricardo Ribeiro Homem Landi[1], Sofia Santos Lima Figueiredo[1], Felipe Cardoso Garcia[1], Theodora Turkot Antunes Pereira[1], Izabella Zgoda[1], Diego Schuster Paes[1]

[1]Hospital Universitário Cajuru, Curitiba PR, Brazil.

Correspondence: Michelle Zonkowski Ribas; Email: michelle.z.ribas@gmail.com.

Case presentation: A 50-year-old man with a history of chronic alcoholism reported ingesting glufosinate ammonium (GLA), an herbicide, associated with alcohol intake on May 16th. Upon admission, he presented with nausea, dizziness, and hallucinations. He experienced an episode of greenish vomiting and subsequently became diaphoretic, tachycardic, and cyanotic, with desaturation and a decreased level of consciousness, requiring orotracheal intubation. During transportation to a tertiary hospital, he developed supraventricular tachycardia, which spontaneously reverted, and had three episodes of epileptic seizures, which only ceased after the administration of 2 vials of diazepam. Upon admission to the hospital, the patient continued to experience epileptic seizures despite sedation with midazolam and fentanyl, leading to the initiation of phenobarbital and continuous sedation with propofol. Following these measures, a video electroencephalogram was performed, which did not identify any seizures. The patient subsequently developed rhabdomyolysis and acute kidney injury, with persistent anuria and metabolic acidosis, requiring hemodialysis sessions. After being off sedation since May 20th, the patient was successfully extubated on May 26th. **Discussion:** The initial symptoms of GLA intoxication are gastrointestinal, primarily due to its irritative effect on mucous membranes. Neurological effects include mental confusion, seizures, memory loss, and coma. Most epileptic seizures are generalized, and more than 50% of them are not controlled solely with benzodiazepines, as demonstrated in our case, where phenobarbital and propofol were necessary. The mechanism underlying these seizures is still poorly understood, but studies indicate hyperstimulation of N-methyl-D-aspartate (NMDA) receptors by glufosinate and its metabolites, such as ammonia. However, it is unknown whether hyperammonemia is a cause or consequence of neurotoxicity. While some studies suggest alcohol may offer protection when combined with GLA ingestion, our case demonstrates severe symptoms despite alcohol consumption. To this day, there is no antidote for this intoxication, and the mechanisms of toxicity remain unclear. While hemodialysis can assist in removing glufosinate from the body, there is no evidence of it preventing neurotoxicity. Additionally, renal excretion proves to be 1.6 to 1.8 times more effective than hemodialysis. Consequently, hemodialysis is reserved for cases of acute renal failure, such as in our situation, and most cases are managed solely with supportive care. **Final comments:** GLA intoxication is rare but presents significant neurological symptoms, necessitating prompt identification for adequate support. Regarding seizures, it is important to note that they are difficult to control and often require additional measures, such as phenobarbital and anesthetics.

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A case of spinal dural arteriovenous fistula

José Víctor Filho de Oliveira[1], Luiz Felipe Mota Freitas[1], Leopoldo Antonio Pires[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Juiz de Fora MG, Brazil.

Correspondence: José Víctor Filho de Oliveira; Email: victorr_oliveira@hotmail.com.

Case presentation: Type 2 diabetic patient, 65 years old, presents with progressive worsening of neuropathic burning pain in the soles of the feet over the last 2 years. Within 2 months it progressed to ascending pain in the posterolateral region of the lower limbs. Additionally, he reported numbness in the anogenital region. Initially diagnosed with musculoskeletal disease, motor physiotherapy was recommended for 6 months. During this period, symptoms worsened during exertion and reports of erectile dysfunction. Proximal weakness in the lower limbs progressed to relatively symmetric crural paraparesis over 8 months, leading to paraplegia and urinary incontinence after intense exertion. In the emergency room, prolonged corticosteroids were prescribed, worsening the condition with notable sphincter dysfunction. Upon admission to our service, cauda equina syndrome was diagnosed. Signs of upper motor neuron involvement were observed asymmetrically in the left lower limb, suggesting conus medullaris syndrome, with sensory dysfunction more pronounced at the L1 level. Symptoms worsened with the Valsalva maneuver. Cerebrospinal fluid analysis and laboratory tests for inflammatory, infectious and autoimmune etiologies were negative. Dural arteriovenous fistula (DAVF) was suspected. Magnetic resonance imaging of the spine revealed extensive signal changes in the spinal cord, from the lower thoracic segments to the conus medullaris, with contrast enhancement and swelling effect. Numerous intradural and extramedullary flow voids were noted, centered at the T11 level. Subsequent spinal angiography demonstrated DAVF at the T12 level, fed by dural branches of the T12 left radicular artery originating from the T12 left intercostal artery, causing extensive venous reflux to L1 with tortuosity of the posterior spinal vein to high medullary levels. **Discussion:** Spinal DAVFs are characterized by an abnormal connection between a radiculomeningeal artery and a radicular vein. This anomaly results in venous congestion and spinal cord ischemia. The vague symptoms associated with DAVFs often lead to diagnostic delay. The slow progression of symptoms can result in extensive myelopathy and irreversible symptoms despite adequate therapeutic intervention. DAVFs should be considered among the differential diagnoses in middle-aged men who present with pain, numbness or weakness in the lower limbs with gradual progression of symptoms associated with bladder, sexual and intestinal dysfunction. Asymmetric onset of symptoms and worsening with exertion and Valsalva maneuver help in the diagnosis. Worsening with the use of corticosteroids is described. MRI typically shows T2 hyperintense lesions and intramedullary flow voids. Catheter angiography is the gold standard for confirmation. **Final comments:** The patient was treated with endovascular therapy, showing notable improvement despite the delay in diagnosis and the use of corticosteroids.

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A case report of complex neurological manifestations associated with polycythemia vera

João Victor Meneses de Aguiar[1], Eduardo Rosa da Silva[1], Bruna Macena Cutti[1], Isadora Oliveira Côrrea[1], Laís Bissoli Perini[2], Edson Pilotto Duarte[1], Bruna Garcia da Silveira[3], Adaucto Wanderley da Nóbrega Junior[1]

[1]Hospital Homero de Miranda Gomes, São José SC, Brazil.
[2]Instituto de Assistência Médica ao Servidor Público Estadual de São Paulo, São Paulo SP, Brazil.
[3]Universidade do Sul de Santa Catarina, Palhoça SC, Brazil.

Correspondence: Laís Bissoli Perini; Email: laisbperini@hotmail.com.

Polycythemia vera is a myeloproliferative disorder of hematopoietic cells characterized by increased blood viscosity due to abnormal and excessive production of erythrocytes, leukocytes, and platelets. It is a rare disease with an annual incidence of 3/100,000 people. Erythrocytosis is the most clinically relevant manifestation, contributing significantly to thrombotic and hemorrhagic events. Neurological complications may occur, primarily due to its association with cerebral ischemic events. **Case presentation:** A 67-year-old male presented with sudden onset paresis in the left upper limb. He reported previous similar episodes, associated with cyanosis in the left hand and bilaterally in the feet, as well as a sensation of "pins and needles". His medical history included a previous ischemic stroke and partial gastrectomy for gastric adenocarcinoma, without requiring chemotherapy or radiotherapy. The patient was also under investigation in the Neurology outpatient clinic for asymmetric tremors in the upper limbs. Neurological examination revealed grade III paresis in the left upper limb with dystonic posture of the 2nd to 4th fingers, hyperreflexia of the left brachioradialis, and a positive Hoffman sign. He exhibited rapid frequency tremors in the hands, bilateral blepharospasm, and adductor-type laryngeal dystonia. Abdominal examination noted a palpable spleen 4 cm below the left costal margin. Admission cranial computed tomography (CT) showed a new ischemic lesion in the right superior frontal gyrus, consistent with recent stroke, and an old ischemic lesion in the right parietal lobe. Additional tests revealed polycythemia (Hb 17.1 g/dL), thrombocytosis (608,000 x 10^9/L), negative serologies, atrial fibrillation on electrocardiogram, and abdominal ultrasound confirming splenomegaly. A JAK2 mutation test confirmed polycythemia vera, supporting our clinical suspicion. Treatment included hydroxyurea to reduce hemoglobin levels, aspirin due to high risk of new ischemic events, and anticoagulation with a factor Xa inhibitor. Following therapy, there was clinical and laboratory improvement. **Final comments:** Increased blood viscosity due to elevated hematocrit is considered the primary factor predisposing polycythemia vera patients to thrombotic events. This case report highlights concurrent neurological symptoms associated with the disease, emphasizing the risk of cerebral infarcts and subsequent neurological manifestations. We highlight in this report the occurrence of dystonic phenomena, previously reported in other polycythemia vera cases. Our primary hypothesis revolves around cortical injury contralateral to the patient's phenomenology, given recent studies emphasizing specific central nervous system activation patterns in dystonia patients. This case underscores the importance of early diagnosis and appropriate therapeutic intervention to mitigate neurological risks associated with polycythemia vera.

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A case report of erdheim-chester disease with neurological manifestations: the relevance of PET-CT scan

Hedra Marques Santos[1], João Henrique Fregadolli Ferreira[2], Lívia Almeida Dutra[2]

[1]Faculdade Israelita de Ciências da Saúde Albert Einstein, São Paulo SP, Brazil.
[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Hedra Marques Santos; Email: hedra.marques@gmail.com.

Case presentation: A 77-year-old male presented with a subacute onset of dizziness, imbalance, intentional tremor of the left arm, dysarthria, and diplopia. He denied any sleep disorders or dysphagia. Additionally, he reported weight loss and was a former heavy smoker. Physical examination revealed ataxic gait, ocular ataxia, normal extrinsic eye movements, normal muscle strength, brisk reflexes, appendicular ataxia, and left hypoesthesia. Brain MRI showed T2 FLAIR lesion at the dorsal pontomesencephalic junction and the left superior cerebellar peduncle, with contrast enhancement, along with small enhancing foci at the right pons, as well as in the deep white matter of the cerebellar hemispheres, and right lateral wall of the fourth ventricle. Perfusion MRI was normal, and spectroscopy revealed an increase in the choline/creatine ratio, and reduction in the N-acetyl-aspartate/creatine ratio. Laboratory results showed normal renal and hepatic function, CRP 1.3, ESR 19mm, serum neurofilament light chain (NfL) of 22.60 pg/mL, normal immunofixation of serum proteins. CSF analysis revealed < 1 cell/mm^3, protein 36 mg/dL, OBC negative, CSF Nfl 1304 pg/mL, IgG index 0.48, negative metagenomic analysis and negative CSF immunophenotyping. Additionally, high-risk paraneoplastic antibodies panel was negative. PET-CT showed an increased in fluorodeoxyglucose-18F (FDG) metabolism in several bilateral bone lesions, and in the pituitary gland. Bone lesions were suggestive of multiple infarcts. A bone biopsy was performed and showed intertrabecular spaces containing fibrous connective tissue, surrounded by rare aggregates of histiocytes with large and foamy cytoplasm. There was no evidence of granulomas or necrosis in the evaluated sections. Immunohistochemistry was positive for CD138 in rare plasma cells, CD20 in rare lymphocytes, CD3 and CD43 in some lymphocytes, and CD68 and CD163 in foamy histiocytes. The patient was diagnosed with ERD. **Discussion:** ERD is a rare histiocytosis characterized by uncontrolled proliferation of histiocytes due to mutations in the MAPK pathway. The presence of CD68+ CD1a- foamy histiocytes is essential for the diagnosis. ECD usually occurs in males between 40 and 70 years of age. Central nervous system involvement may be the only manifestation of ECD and occurs in 40% of patients. The PET-CT in this context enables the detection of metabolic abnormalities throughout the body, including lesions in bones and other organs. This is particularly relevant in cases of ERD, where manifestations can be multifaceted and involve multiple organ systems. Moreover, PET-CT played a crucial role in guiding the bone biopsy, providing information on the location and extent of lesions. **Final comments:** This case report underscores the importance of PET-CT in the evaluation of patients with granulomatous involvement of the central nervous system, providing critical insights for diagnosis and management.

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A case report of super-refractory status epilepticus with a good response to intravenous human immunoglobulin

Rayanne Pereira Mendes[1], Guilherme Bitencourt Barbosa[1], Renato Morais Bueno[1], Victor Ezaki Alves Silva[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Rayanne Pereira Mendes; Email: rayannepmendes@gmail.com.

Case presentation: A 23-year-old female with irregularly treated epilepsy presented to the emergency room complaining of headaches, muscle twitching in limbs, and cognitive changes over the past three days. On admission, she appeared drowsy with no physical abnormalities noted. She developed intense psychomotor agitation, cognitive impairment, opsoclonus, myoclonus, and worsening consciousness, progressing to refractory status epilepticus as confirmed by electroencephalogram showing epileptiform

activity in the left medial temporal region. Cranial CT scan and initial cerebrospinal fluid analysis showed no abnormalities. Serological tests for HIV, syphilis, and viral hepatitis were negative, and rheumatological tests were normal. Subsequent cerebrospinal fluid analysis revealed a cell count of 55 (45% lymphocytes, 55% neutrophils) with normal protein and glucose levels. Despite seven days of anti-seizure medication adjustments and deep sedation, she remained in super-refractory status epilepticus as confirmed by subsequent electroencephalogram. Due to suspicion of autoimmune encephalitis, intravenous human immunoglobulin therapy was initiated, resulting in resolution of status epilepticus; however, she continued to experience a slow awakening process. A repeat cycle of intravenous human immunoglobulin significantly improved her level of consciousness. **Discussion:** Status epilepticus is a condition where mechanisms responsible for terminating seizures fail, potentially leading to neuronal injury or death. Super-Refractory Status Epilepticus persists for 24 hours or more despite anesthesia, including cases where seizures recur after anesthesia reduction or withdrawal. Clear treatment protocols for Super-Refractory Status Epilepticus are lacking, with intravenous human immunoglobulin proposed as a therapeutic option. It is thought to flood the system with non-reactive antibodies, attenuating inflammation through direct interactions with leukocytes and catabolism of pathological antibodies. A systematic review of 33 global Super-Refractory Status Epilepticus cases, including unspecified encephalitis and antibody-defined etiology, found that 42.4% responded to intravenous human immunoglobulin with complete seizure resolution. **Final comments:** While evidence of intravenous human immunoglobulin's superiority over conventional therapies for Super-Refractory Status Epilepticus is lacking, cases like this one, showing significant response to treatment, underscore the need for randomized clinical trials to better assess its efficacy.

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A challenge: anti-GAD-related limbic encephalitis

Ana Letícia Manso Assakawa[1], Isabela Kirach Gaio[1], Maria Júlia Costa Rocha[1], Luís Fernando Fernandes Miranda[1], Pamella Francisquini Gardin[1], Yves Henrique Ramos Mansano[1], Antonio Marcos Reissureição Galindo[1], Matheus da Silva de Oliveira[1], Mateus Mistieri Simabukuro[2], Gabriel Bortoli Ramos[1,2]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.
[2]Universidade de São Paulo SP, Brazil.

Correspondence: Ana Letícia Manso Assakawa; Email: ra128765@uem.br.

Case presentation: A 25-year-old female patient presented with hyperthyroidism, confirmed by elevated TRAb (TSH Receptor Antibody) levels and TSH< 0.01, along with a diffuse goiter on thyroid scintigraphy characteristic of Graves' disease. Following iodine therapy, six months post-diagnosis, she developed hypothyroidism with TSH levels elevated, requiring levothyroxine treatment. During this period, the patient experienced epileptic seizures with mesial temporal semiology and secondary generalization and anterograde amnesia. MRI which revealed T2 and FLAIR hyperintensity affecting the amygdaloid bodies and bilateral hippocampal formations. A SPECT scan showed reduced uptake in the temporal and parietal lobes, anterior and posterior cingulate regions. EEG demonstrated focal epileptiform paroxysms in the bilateral temporal region. Despite treatment with lamotrigine, the patient continued with seizures. Serum anti-GAD levels were markedly elevated (> 2000 IU/mL), suggesting a diagnosis of anti-GAD autoimmune encephalitis. Confirmation was achieved through positive results of cerebrospinal fluid analysis using both cell-based assay and immunohistochemistry. Paraneoplastic investigations were negative. Treatment was a six-month course of immunoglobulin therapy combined with levetiracetam and lacosamide. While there was a slight improvement in seizure frequency with immunotherapy, episodes of secondary generalization ceased. Immunoglobulin therapy discontinued in favor of Rituximab as ongoing immunotherapy. **Discussion:** Limbic encephalitis include autoimmune, infectious, paraneoplastic and systemic etiologies. Among the autoimmune causes, anti-GAD antibodies are notable for their impact on glutamic acid decarboxylase, disrupting GABAergic transmission and causing neuronal hyperexcitability. It is associated with autoimmune diseases such as type 1 diabetes, Graves' disease, and Hashimotos thyroiditis. Neurological manifestations typically present as limbic encephalitis, cerebellar ataxia or stiff person syndrome. Diagnosis relies on clinical suspicion supported by inflammatory findings in cerebrospinal fluid, elevated anti-GAD titers in serum or CSF and characteristic temporal lobe hyperintensity on MRI. PET-FDG and EEG contribute to diagnosis. Treatment challenges arise due to the condition's refractory nature; 80% of patients do not respond adequately to first-line therapies like immunoglobulin. In this case, despite initial treatment, the patient's seizures persisted until anti-GAD autoimmune encephalitis was identified in conjunction with Graves' disease and subsequent hypothyroidism. **Final comments:** This case highlights the importance of considering anti-GAD autoimmune encephalitis in patients presenting with refractory epilepsy. Early diagnosis facilitated prompt initiation of immunotherapy, critical for improving prognosis and minimizing long-term complications. Enhancing access to specific laboratory tests is essential to reduce diagnostic delays.

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A challenger case report: septic-embolic stroke or ischemic stroke due to aortitis syphilitic?

Igor Pereira Matos de Oliveira[1], Patrícia Marques Mendes[2], Yan da Silva Raposo[2], Matheus de Paula Batista[2], Caio Augusto de Martha Santos[2], Enzo Barreto Viana[2], Maycon Melo Lopes[2], Vitor Hugo Machado Silva[2], Eduardo Mendonça Werneck da Silva[2], Laís Aparecida Machado de Siqueira[2]

[1]Santa Casa da Misericórdia de Belo Horizonte, Belo Horizonte MG, Brazil.
[2]Hospital das Clínicas Samuel Libânio, Pouso Alegre MG, Brazil.

Correspondence: Igor Pereira Matos de Oliveira; Email: igorpmoliveira@outlook.com.

Case presentation: A 43-year-old woman attended emergency service with respiratory symptoms and a recent diagnosis of tuberculosis. She presented also aphasia and right brachial mono paresis. She was evaluated for ischemic stroke, and computed tomography (CT) of the brain evidenced bilateral hypodensity (left temporoparietal cortico-subcortical and right temporal subcortical). Serum FTA-ABS test was positive, and VDRL 1:8. In Cerebrospinal fluid analysis, VDRL was negative, bacterial and fungus culture without growth, and cryptococcal antigen test was negative. Transesophageal echocardiogram showed 0.5x0.6 cm thrombus vegetation in the pulmonary valve. Blood culture in three pairs with the finding of Bacillus altudinis/pumilus. CT angiography of the abdominal and thoracic Aorta reported thrombosis in the subclavian artery and aortic arch. Transesophageal Doppler Echocardiography added a filamentary hyperechogenic image in the right ventricular outflow tract, adhered to the pulmonary valve, with random movement. Carotid Doppler ultrasound, and transthoracic echocardiography without abnormalities. Treatment consisted of anticoagulants, antibiotic therapy for endocarditis, COXCIP (tablets containing isoniazid, pyrazinamide, ethambutol, and rifampicin), Crystalline Penicillin 4,000,000 IU, Benzathine Penicillin 2,400,000 IU. He evolved with complete improvement in neurological, cardiac, and respiratory symptoms. **Discussion:** Syphilitic aortitis is an inflammatory condition of the aortic wall, resulting from the hematogenous dissemination of the bacterium Treponema pallidum. Central nervous system involvement may also occur in tertiary syphilis. The presence of nonspecific systemic symptoms, such as fever, asthenia, and weight loss, initially obscured the diagnosis and led to an extensive investigation to determine the underlying cause. A history of smoking and use of illicit substances, including crack and alcohol, may have contributed to the progression of vascular disease and worsening of neurological symptoms. The probable TOAST of ischemic stroke was due to large vessel and cardioembolic involvement, with partial thrombosis of the aortic arch and the formation of septic emboli resulting in bilateral cortico-subcortical cerebral infarctions, evidenced by magnetic resonance imaging. The presence of positive VDRL in serum and hyperproteinorrhaquia in the cerebrospinal fluid also point to the diagnosis of neurosyphilis. Treatment of syphilitic aortitis and its complications involves the administration of antibiotics combined with anticoagulant therapy in cases of arterial thrombosis. Surgical intervention may be necessary in cases of serious vascular complications, such as aneurysms or aortic dissection. **Final comments:** It is important to carry out continuous surveillance and public education about syphilis and its complications and the management of the disease is multidisciplinary.

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A clinical alert on the evolution of a patient with Guillain-Barré syndrome and dengue treated with therapeutic plasma exchange: case report

Eduarda Leitzke[1], Marco Antônio da Croce[1], Júlia Hasegawa Rodrigues[1], Ketelin Roos Tirloni[1], Bianca Tessele[1], Jaqueline Solda Palaoro[2], Bruna Accorsi Machado[2], Gisele de Oliveira Doring[2], Aline Ferreira de Miranda[2], Cristiane da Silva Rodrigues de Araújo[1]

[1]Universidade de Passo Fundo, Passo Fundo RS, Brazil.
[2]Hospital São Vicente de Paulo, Passo Fundo RS, Brazil.

Correspondence: Eduarda Leitzke; Email: leitzkeeduarda@gmail.com.

Case presentation: 23-year-old patient, treated in the emergency room of a hospital in the north of Rio Grande do Sul in November 2013, reporting pain and paresthesia in the lower limbs for 30 days, after a viral illness and a recent trip to Bahia. Physical examination indicated Guillain-Barré syndrome (GBS), confirmed by lumbar puncture. Requested evaluation of the Hemotherapy Service for the therapeutic plasmapheresis (TPE) procedure. After starting the procedure, within three hours the patient had urinary difficulty, requiring a bladder catheter and with greenish urine; six hours later, he developed respiratory failure, hypoxemia, sweating and hypotension, requiring mechanical ventilation. Patient was sent to the Intensive Care Unit with suspected septic shock. Laboratory tests revealed an increase in hematocrit from 43% to 60%. The patient progressed to acute renal failure and underwent renal replacement therapy, but could not tolerate hemodialysis due to clinical instability, progressing to multiple organ failure and death 4 days after the procedure. Cause of death: hemoconcentration shock after TPE. Multidisciplinary review discussed possible causes. Pre-procedure sample confirmed positive IgM for Dengue. **Discussion:** GBS is an autoimmune inflammatory demyelinating polyradiculoneuropathy, which leads to loss of the myelin sheath and tendon reflexes. Symptoms are sensory loss, hypo- or areflexia and limb paresis and/or plegia, mostly symmetrical. Most patients have an infectious disease prior to the onset of symptoms.

Treatment of GBS includes plasmapheresis and intravenous immunoglobulin (IVIg) performed within four weeks of the onset of neurological symptoms, which can slow the progression of the disease by removing harmful antibodies. Dengue is an arboviral infection, the symptoms of which are fever, joint pain, headache, flushed skin and morbilliform rashes. The incidence of neurological symptoms and complications in dengue varies from 1 to 25%, including GBS. In the current dengue scenario, with 4.7 million probable cases by May 2024, it is necessary to evaluate a possible association of dengue with GBS. **Final comments:** Understanding the immunological mechanisms triggering GBS following dengue infection and the increasing incidence of dengue cases in various regions of Brazil, along with the association of these two pathologies, highlights the importance of continuous epidemiological surveillance and the implementation of vector control measures to prevent outbreaks of these diseases and its neurological complications.

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A clinical case of migraine and probable cluster headache in a patient with tuberous sclerosis

Giuseppe Dick Bonato[1], Marco Antônio Delazeri[1], Vanise Grassi[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Giuseppe Dick Bonato; Email: g.d.bonato@hotmail.com.

Case presentation: We present a 35 year-old male with tuberous sclerosis following referred to headache-specialized evaluation. He has been treated for focal epilepsy since age of 4, which was well controlled with lamotrigine 300 mg/day. Diagnosis of tuberous sclerosis was made after skin biopsy a few years before, and brain MRI confirmed generalized dispersed cortical and subcortical tuberous lesions, and small subependymal nodules in left lateral ventricle wall. Skin lesions included hypercromic right forehead plaque and left hallux ungual fibroma. He denied further health issues and previous neurologist had started verapamil 240 mg/day and amitriptyline 75 mg/day for headache. He described two types of headache. Migrainous episodes first presented at 10 year-old, as a moderate bilateral frontal 1-2 hours-long pulsating pain with associated nausea, vomiting, photo- and phonophobia. In such episodes, he needs to stop what he is doing and quietly lie down. The second type of headache started at age of 20 and was a stabbing, acute, severe, left orbital pain. These episodes were associated with marked ipsilateral autonomous symptoms: lacrimation, conjunctival injection, sweating, ptosis and rhinorrhoea. While presenting these second-type episodes, the patient also has photophobia and prefers to lie down, though remains unrestful in bed. They tend to occur in colder months and take up to 8 hours when severe. He had no relevant headache episodes in the previous 10 months under current therapies. Reduction and discontinuation of verapamil was discussed, but patient was very resistant due to good response. As needed subcutaneous sumatriptan was introduced and patient was taught to how to use it. MRI focused on pituitary gland showed possible hypocaptant microadenoma. The patient was then referred to a neuroendocrine team and waits for endocrine screening results. **Discussion:** We present a case with two very distinct headache types: migraine without aura and probable cluster headache - as duration feature of ICHD-3 cluster headache criteria is not fulfilled. Headache associated with tuberous sclerosis can have various causes. In the presented case, however, cluster headache qualities are more closed related to the possible adenoma. Association between both conditions have been described, and it is hypothesized that both hypothalamic activation and local structural changes play a role in developing autonomic features. Cutaneous lesions alone would not fulfill clinical diagnostic criteria for tuberous sclerosis complex updated in 2012, bur radiologic neurologic features do in fact fulfills two major features in said criteria and consequently closes the diagnosis. **Final comments:** Cluster headache is a debilitating diagnosis that can be related to adenomas and pituitary lesions. There is however a question whether subependymal periventricular nodes can play a role in the presented case trigeminal autonomic headache.

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A cluster of Creutzfeld-Jakob disease VV2 phenotype in 2024 in São Paulo

Fernanda Lustosa Cabral Gomez[1], Larissa Braga Mendes[1], Leonardo Villaverde Buback Ferreira[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Luiz José Malta Gaia Ferreira Júnior[1], Daniel Delgado Seneor[1], Márcio Pinheiro Lima[1], Maria Elisabeth Matta de Rezende Ferraz[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Fernanda Lustosa Cabral Gomez; Email: fernandalustosacg@gmail.com.

Case presentation: Case 1 A 58-year-old man was admitted with a one-month history of progressive dizziness and nausea. Neurological exam revealed signs of cerebellar ataxia. Brain MRI and cerebrospinal fluid analysis were unremarkable. He underwent empirical treatment with intravenous immunoglobulin but showed no signs of improvement. Two months later he maintained progressive worsening of cerebellar symptoms along with visual hallucinations and involuntary movements of the head and upper limbs. At this time, his exam revealed worsening of cerebellar signs, global hyperreflexia and choreic movements. A new brain MRI was performed and revealed bilateral T2-hyperintensity in caudate nucleus and putamen with restricted diffusion. 14-3-3 CSF assay was positive and established criteria for probable sporadic Creutzfeldt-Jakob disease (sCJD). The patient died one month later at the palliative care facility. Case 2 A 53-year-old man with a history of chronic alcoholism was admitted with one month of progressive dizziness and headache. Neurological exam revealed central nystagmus and dysmetria. Laboratory and brain image were unremarkable. He was discharged on Thiamine under the hypothesis of Wernicke's encephalopathy. One month later his symptoms worsened, along with visual hallucinations and global rigidity. His exam revealed akinetic mutism and hyperekplexia. Brain MRI showed restricted diffusion of right parietooccipital cortex and protein 14-3-3 from CSF was positive, establishing probable sCJD. The patient died three weeks later at the palliative care facility. **Discussion:** CJD is a rare, fatal and rapidly progressive neurological disease, caused by misfolded prion protein in the brain. The reported prevalence in Brazil per year is 1 to 2 cases per million. CJD is classified in four subtypes: sporadic (sCJD), variant, iatrogenic and familial. Sporadic form is the most common (85%) and is further classified into four subtypes according to prion protein features and genotype: MM1/(MV)1, MM2, VV1, VV2 and MV2. Both our patients had clinical features consistent with the VV2 subtype that most commonly presents prominent progressive ataxia. Patients lived 30 kilometers from each other and were admitted to the same facility within two months. Both cases were reported to the Ministry of Health. **Final comments:** Our article addresses a temporo-spatial cluster of sCJD with the VV2 phenotype in São Paulo. Similar clusters of sCJD have been reported in the past. One possible explanation for this phenomenon is the exposure to a common external factor or, less likely, direct transmission between cases. Our work highlights the need for future research on transmission events and environmental factors that may increase the occurrence of CJD.

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A comprehensive analysis of ATRX and IDH1 expression in gliomas: associations with tumor grade and prognostic implications

Iasmin Maria Rodrigues Saldanha[1], Thiago André Gomes Costa Pereira[1], Guilherme Nobre Nogueira[1], Osvaldo Mariano Viana Neto[1], Cauan Miranda Mateus[1], Leslie Mateus de Sousa Nunes[1], Maria Jania Teixeira[1], Gunter Gerson[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Leslie Mateus de Sousa Nunes; Email: leslienunes@alu.ufc.br.

Background: Management of brain tumors has evolved due to the identification of molecular changes, allowing greater diagnostic accuracy, understanding of the biological behavior of tumors, definition of prognostic factors, and identification of therapeutic targets. IDH mutation is a common change in primary and recurrent gliomas, but the role of ATRX is not yet fully understood. Studies highlight the association between IDH mutations and ATRX inactivation, attributed to deletions, gene fusion, or a combination of these causes. **Objective:** To analyze the expression of wild ATRX and IDH1 mutant in gliomas, looking for possible associations with tumor grade. Understanding these relationships can contribute to more accurate management strategies. **Methods:** An epidemiological, descriptive, transversal, and retrospective study was carried out by reviewing the records of patients diagnosed with gliomas in a tertiary hospital over a period of 10 years. The analysis included 97 patients with different histological types of gliomas, classifying them according to the expression of ATRX and IDH1, in addition to tumor grade. **Results:** The analysis of the records showed a varied distribution of gliomas: 43 cases of astrocytomas (44.3%), 1 case of ganglioglioma (1%), 41 cases of glioblastoma (42.2%), 11 cases of oligodendroglioma (11.3%), and 1 case of pleomorphic xanthoastrocytoma (1%). As for tumor grade, 2 cases of grade I (2%), 31 cases of grade II (31.9%), 9 cases of grade III (9.2%), and 54 cases of grade IV (55%) were identified. Evaluation of ATRX expression revealed preservation of this protein in 79 cases (81.4%): 26 cases of grade II (26.8%), 5 cases of grade III (5%), and 48 cases of grade IV (49.4%). In 18 cases (18.5%), silenced ATRX was observed: 5 grade II cases (5.1%), 4 grade III cases (4.1%), and 9 grade IV cases (9.2%). The analysis of the IDH1 marker showed positivity in 74 cases (76.2%): 1 case of grade I (1%), 33 cases of grade II (34%), 12 cases of grade III (12.3%), and 28 cases of grade IV (28.8%). Negativity for IDH1 was identified in 23 cases (23.7%): 3 cases of grade II (3%), 1 case of grade III (1%), and 19 cases of grade IV (19.5%). In the 74 cases with positive IDH1, ATRX was preserved in 61 cases (82.4%) and silenced in 13 cases (17.5%). In the 23 cases with negative IDH1, the distribution was equal: 11 cases of preserved ATRX (47.8%) and 12 cases of silenced ATRX (52.1%). **Conclusion:** The immunohistochemical analysis of the expression of wild ATRX and mutant IDH1 in diffuse gliomas showed association with tumor grade, corroborating the profile described in the medical literature. When IDH1 is positive, ATRX is often preserved; in cases of negative IDH1, there is equal distribution between preserved and silenced ATRX. The most commonly observed histological types were astrocytomas and glioblastomas, with a prevalence of grade IV tumors. These correlations help in understanding tumor biology and its prognostic value.

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A congenital slow channel myasthenic syndrome in two sisters: case report

Laura Altomare Fonseca Campos[1], Jéssica Cristina Silveira Damasceno[1], Marcelo Sobrinho Mendonça[1], Thiago Cardoso Vale[1]

[1]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.

Correspondence: Laura Altomare Fonseca Campos; Email: lauraaltomare8@gmail.com.

Case presentation: Female siblings, aged 31 and 16, with early childhood onset of eyelid ptosis that worsened throughout the day, associated with falls and weakness, with impairment in school and basic activities of daily living were evaluated. Pregnancy and childbirth were uneventful, and there were no cognitive or neurological deficits or other comorbidities. One of them had dysphagia for liquids during adolescence, which improved after being monitored by a speech therapist. Physical examination showed proximal weakness in the upper and lower limbs, as well as bilateral eyelid ptosis, with no other alterations. Family history included a father and paternal grandfather affected by congenital myasthenia, without consanguinity. The hypothesis of congenital muscle disease was raised, and investigations were carried out with imaging, laboratory and electroneuromyography (ENMG) tests, which showed postsynaptic neuromuscular junction dysfunction, with the occurrence of double action potentials in the motor conduction study in all assessed nerves, bilaterally, suggesting a slow channel mechanism. A genetic test (neuromuscular panel) showed a mutation in the cholinergic receptor nicotinic alpha 1 subunit (CHRNA1) gene in heterozygosity, confirming the diagnosis of Congenital Myasthenic Syndrome 1A (OMIM #601462). Brain MRI was normal. Ophthalmologic and cardiologic evaluations were unrevealing. Spirometry has also been carried out periodically and, to date, mild restrictive disorder has been monitored. Fluoxetine and salbutamol were tried but discontinued due to side effects. **Discussion:** Slow-channel congenital myasthenic syndrome (CMS) consists of a dynamic alteration of the acetylcholine ion channel of the neuromuscular junction, with an autosomal dominant inheritance pattern and a high degree of penetrance. Symptoms vary from mild weakness, which worsens throughout the day, intolerance to physical exercise, to incapacitating weakness, respiratory failure and bulbar symptoms. Unlike Myasthenia Gravis, an important differential diagnosis, slow channel CMS does not respond well to corticotherapy and acetylcholinesterase inhibitors and may worsen with their use. However, it does respond to drugs such as fluoxetine and quinidine, which act on the affected sodium channels. **Final comments:** This report exposes an important differential diagnosis of neurological diseases that present with ptosis and fatigable muscle weakness.

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A "de novo" KIF1A mutation in a patient with spastic paraplegia, peripheral neuropathy and intellectual disability: case report

Aline Yumi Sato Yamazaki[1], Fernanda Nunes Maria[1], Aline Geisler[1], Juliana Cardoso Fiquene[1]

[1]Hospital Santa Isabel, Blumenau SC, Brazil.

Correspondence: Aline Yumi Sato Yamazaki; Email: aline.yumi-andra@hotmail.com.

The KIF1A gene encodes a Kinesin-related protein involved in the anterograde transport of synaptic-vesicle precursors along the axon. Since 2011 several KIF1A-related disorders have been described such as hereditary spastic paraparesis, hereditary sensory and autonomic neuropathy, severe neurodegenerative condition, progressive encephalopathy with edema, hypsarrhythmia, and optic atrophy (PEHO) and NESCAV syndrome. We report a patient presenting spastic paraplegia, peripheral neuropathy and intellectual disability with gene KIF1A mutation. A 58-year-old man presented to our hospital with a progressive gait disorder and peripheral neuropathy. According to his sister, he was born by home birth with no medical assistance. During his childhood, the family observed that he had intellectual delay, as he did not learn to read or to write and dropped out of school after the second grade. He was able to do daily living activities (eg. take a shower, eat by himself), but he did not have a social life, nor a job. Until 15 years old, he had no difficulty walking, then he began to experience gait changes and frequent falls. Currently, he has been walking with the aid of a walker for about 5 years. He had no seizures, dysautonomia, dysphagia, or dysarthria. A neurological examination revealed coherent speech, although intellectual disability. He also presents spastic paraparesis, muscle weakness in the lower limbs, Medical Research Council (MRC) sum score 49/60, deep tendon reflexes (biceps 0, brachioradialis 1, patellar 2, Achilles 0), bilateral Babinski sign. In the investigation, brain MRI was normal, electroneuromyography showed axonal sensitive and motor polyneuropathy with a predominance of the lower limbs, and the genetic panel found a pathogenic heterozygous mutation in the KIF1A gene (p.Arg11Trp). Despite this wide phenotypic spectrum, KIF1A-related disorders as a whole remain extremely rare with many patients receiving a misdiagnosis. In this case reported, the disorder was probably attributed to poor assistance during birth, retarding the diagnosis. Several types of mutations in the same protein have been observed, which leads to the possibility of new variants in these KIF1A-related disorders.

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A diagnostic challenge: is mixed connective tissue disease enough to explain difficult-to-control epilepsy?

Leticia Machado Dumont, Renan Brandão Rambaldi Cavalheiro[1], Paulo Sgobbi[2], Wladimir Bocca Vieira de Rezende Pinto[2], Cristine Mella Cukiert[1], Rafael Chaves Claudino de Queiroga[1]

[1]Irmandade Santa Casa de Misericordia de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Leticia Machado Dumont; Email: leticiamachadodumont@gmail.com.

Case presentation: 27 year-old female patient with a complex clinical presentation, recently diagnosed with GLUT1 deficiency and scleroderma type mixed connective tissue disease (DMTC). The patient had treatment-resistant seizures since childhood, in addition to rheumatological symptoms that began at the age of 16. Since DMTC alone could not explain the occurrence of seizures, the patient was referred from rheumatology to neurology, leading to a investigation and subsequent diagnosis of overlapping GLUT1 deficiency. At 1 year of age, the patient presented with her first seizures, exhibiting different phenomenologies, including myoclonic seizures (the most frequent type). At the age of 3 started treatment with valproic acid, without seizure control. During childhood, the patient presented with learning difficulties, repeating some school years. At the age of 16, she began to experience significant weight loss and muscle weakness in her upper limbs. On neurological examination, she presented with grade IV motor strength in the upper limbs. On ectoscopy, she presented with stigmas of scleroderma. At rheumatological investigation with laboratory tests was confirmed the diagnosis of scleroderma type DMTC (ANA speckled nuclear >1/654, Anti RNP antibody reactive). Treatment with Leflunomide was introduced, with control of the rheumatological condition. Despite DMTC control, the seizures persisted, even with the use of 2500mg/day of valproic acid. That way, the patient was referred to neurology for further investigation. EEGs and cranial MR were performed, without identifying the cause of the seizures. Therefore, it was decided to perform an epilepsy genetic panel, which did not find pathogenic variants, but identified a variant of uncertain significance (VUS) in the SLC2A1 gene, responsible for encoding the GLUT1 protein. Literature review indicated that the VUS in question may be related to GLUT1 deficiency. GLUT1 deficiency is a rare genetic disorder that affects glucose transport to the brain and can lead to a range of neurological problems, including seizures, developmental delay and intellectual disability. DMTC, on the other hand, is a group of autoimmune diseases that affect connective tissue. The association between GLUT1 deficiency and DMTC is rare but has been described in the literature. The VUS identified in the patient's SLC2A1 gene has also been associated with GLUT1 deficiency in other studies. Thus, the combination of GLUT1 deficiency and DMTC in this patient is even rarer than the isolated occurrence of these diseases and represents a diagnostic and therapeutic challenge. **Final comments:** This case illustrates the importance of a comprehensive investigation in patients with refractory epilepsy. The diagnosis of GLUT1 deficiency in adults can be challenging due to the variety of symptoms, but genetic testing has contributed to the elucidation of such cases, allowing for appropriate treatment and patient follow-up.

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A dysarthria, dysphagia, and sialorrhea: impacts on the quality of life in patients with Parkinson's disease

Arthur Jun Deguti Hata[1], Vivian Kaori Segawa[1], Ana Luiza Fabris Moraes[1], Emerson Gisoldi[1], Alice Estevo Dias, Margarete de Jesus Carvalho[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Arthur Jun Deguti Hata; Email: arthur.deguti@gmail.com.

Background: Idiopathic Parkinson's disease (IPD) is a chronic neurodegenerative disease with slow progression and motor and non-motor manifestations, it affects between 1-2% of the population over 65 years old. Dysarthria, dysphagia, and sialorrhea can compromise patients' quality of life and lead to clinical complications. **Objective:** To evaluate the perception of a group of IPD patients regarding speech, swallowing, and saliva control alterations, as well as their impact on quality of life. **Methods:** Observational cross-sectional study with the application of the Radboud Oral Motor Inventory (ROMP) questionnaire in IPD patients. The questionnaire consists of 23 questions, seven related to speech, seven to swallowing, and nine to salivation. Each question contains 5 response (options from 1 to 5), with 1 being normal and 5 being severe alteration. The results were obtained by summing the points of each question, with total scores ranging from 23-115 for speech and swallowing, and 9-45 for salivation. All patients were evaluated using the Hoehn & Yahr (H&Y) and Unified Parkinson's Disease Rating Scale (UPDRS I, II, III, IV, and V) scales for Spearman's correlation analysis with ROMP, using a significance level of ($p \leq 0.05$). **Results:** 25 IPD patients aged 40 to 80 years, including 11 men (44%) and 14 women (56%), H&Y 1-5 and UPDRS 9-96 were evaluated. The mean total points across all patients were 44.08 (SD 16.4), with mean scores for speech was 17.16 (SD 7.5) , 13.56 (SD 6.7) for saliva, and 13.36 (SD 7.7) for swallowing. When separated by sex, men had a total mean of 47.36 (SD 17.3) and women 41.50 (SD 15.8). Spearman correlation of ROMP with UPDRS showed a coefficient of 0.43 ($p=0.03$) and ROMP with H&Y scale was 0.25 (p 0.11). **Conclusion:** This study showed that speech, swallowing, and saliva disorders seriously affect quality of life in IPD, particularly in interpersonal relationships and patients' food safety. Thus, the application of ROMP in IPD patients was useful both for quickly identifying alterations and for guiding appropriate assessment and timely multidisciplinary rehabilitation, aiming to alleviate disease complications and improve well-being.

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A epidemiological analysis of the hospitalization of meningococcal infection in Brazil from 2013-2023

Paulo Wendel Moura de Souza[1], Júlia Viana de Castro[1], Matheus Henrique Brustolim Marcucci[1], Lucas Frigo[1], Cássia Caroline Aguiar da Ponte[1], Yasmin Dantas Babadopulos[1], Amanda Mendes das Neves[1], Sofia Santana de Figueirêdo[1], Rodrigo Guimarães Bandeira[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Júlia Viana de Castro; Email: juliaviana2020@alu.ufc.br.

Background: Meningococci (Neisseria meningitidis) are gram-negative diplococci that cause meningitis and meningococcemia. Symptoms include headache, nausea, vomiting, photophobia, lethargy, rash, multiple organ failure, shock, and disseminated intravascular coagulation. Carriers (and infected patients) may transmit the organism to people who have direct contact with respiratory secretions or who inhale large-droplet nuclei from a carrier or patient. Diagnosis is clinical, confirmed by culture. **Objective:** To analyze the epidemiological profile of confirmed cases of meningococcal infection in Brazil between the years 2013 and 2023. **Methods:** This study is an epidemiological investigation of the hospitalization of meningococcal infections cases between 2013 and 2023 in Brazil. Data were collected from the Sistema de Informação de Agravos de Notificação (Sinan Net) do Departamento de Informática do Sistema Único de Saúde (DATASUS), and data analysis was performed using Jamovi® software. **Results:** From 2013 to 2023, approximately 13,008 cases of meningococcal infection were recorded in Brazil. Of these, approximately 42.91% (≅ 5,582) were in the Southeast region and only 7.92% (≅ 1,031) in the North region. During the pre-pandemic period of SARS-CoV-2 (2013 to 2019), the mean number of cases was 1,464 per year, while during the pandemic (2020 to 2022), it was 602 cases per year, a reduction of 58.86%. From 2019 to 2020, there was a reduction of 44.24%, and from 2020 to 2021, a reduction of 14.59%. The year 2022 had an increase of 59.38% compared to 2021 in terms of the number of cases, and from 2022 to 2023 a small increase of 6.4%. The age group that suffered the most reduction in the mean number of cases in the pandemic period compared to the pre-pandemic period was 10 to 14 years old (0.76%), and the one that was least affected were those from 70 to 79 years old and 80 years old or older (14% and 12%, respectively). **Conclusion:** According to the data analysis, there has been a considerable reduction in the number of cases during the pandemic period (58%) compared to the pre-pandemic period. This is due to social isolation, as well as the restrictive and preventive hygiene measures imposed during the pandemic period to control COVID-19, which may have contributed to lowering the rates of bacterial infections, directly influencing the drop in the number of cases of meningococcal infections. In addition, this difference can also be explained by a reduction in the notification of cases, as well as in hospital demand during the pandemic period, a setting also seen in other diseases, such as encephalitis. This phenomenon can be evidenced by the sharp reduction between 2019 and 2020 and an increase that was also exacerbated between 2021 and 2022, as well as a only slight change between 2022 and 2023, the first post-pandemic year.

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A family with ALS and simultaneous mutations in C9ORF72 and OPTN genes

Francisco de Assis Aquino Gondim[1], Rodrigo Fagundes da Rosa[1], José Marcelino Aragão Fernandes[1], Nayana Freire de Almeida Fontes[1], João Marcelo Albuquerque Beserra de Sousa[1], Wilson Marques Júnior[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Rodrigo Fagundes da Rosa; Email: rodrigofdarosa@gmail.com.

C9ORF72 mutations are the most common identified genetic cause of ALS. The role of OPTN mutations in ALS is less certain. To our knowledge, there is only one report of simultaneous mutations in C9ORF72 and OPTN genes leading to ALS. Here, we report a family with 3 ALS patients, 2 of them with simultaneous C9ORF72 and OPTN mutations. **Case presentation:** Case 1: 62 year-old, male, with history of hypertension and Restless Legs Syndrome, began experiencing dysphonia and dysarthria in January 2020, followed by weakness in the left arm. After four months, he developed weakness in the right arm and fasciculations. EMG performed revealed lower motor neuron disease. In January 2021, he developed leg weakness and falls and in April 2021, riluzole was started. Neurological examination revealed preserved cognitive functions, predominantly left-sided tetraparesis, diffuse hyperreflexia, and proximal and distal arm atrophy. He died on late 2022. Case 2: 56 year-old, female began experiencing dysphonia and dysarthria in September 2019, followed by arm weakness. She underwent gastrostomy in March 2021. She was treated with riluzole and Revised El Escorial, Awaji and Gold Coast criteria. She died in January 2023. Their father had died with a disease pattern consistent with ALS and similar to their children (Cases 1 and 2). Genetic testing revealed that both patients in Case 1 and 2 had simultaneous mutations in the C9ORF72 and OPTN genes (C.1691A> G; p.aSP564Gly, variant of uncertain significance. **Discussion:** Familial Amyotrophic Lateral Sclerosis (fALS) may comprise 10-20% of ALS cases. More than 40 genes have been associated with ALS. The most common genetic cause is a hexanucleotide repeat expansion in the C9orf72 gene, responsible for 30%-50% of familial ALS cases. Optineurin (OPTN) is a cytosolic protein ubiquitously expressed, primarily located in the perinuclear region of the cell, where it associates with the Golgi apparatus and interacts with various molecules. Mutations in OPTN are also known to cause primary open-angle glaucoma (POAG) and Paget's disease. **Final comments:** To our knowledge, our present report with simultaneous mutations in OPTN and C9orf72 genes in ALS patients was previously evidenced only in a single family from England (Bury et al. 2016).

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A gabapentin-induced edema in a patient with mesial temporal lobe epilepsy: a case report

Vanessa Conceição Saar Mautone Lima[1], Ricardo Cezare Araujo[1], Victória Nunes Alvares[1], Lucas Gonçalves Coelho[1], Clara Ribeiro Rizzo[1], Bernardo Leal Cupello[1], Melina da Silva Bernardes[1], Glenda Corrêa Borges de Lacerda[1]

[1]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gaffrée e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: Victória Nunes Alvares; Email: victoriaalvares@edu.unirio.br.

Case presentation: A 50 year-old female with a previous diagnosis of Mesial Temporal Lobe Epilepsy and on active treatment with lamotrigine, gabapentin, topiramate, phenobarbital, valproate and fluoxetine, was admitted with generalized edema and arthralgia in our hospital. On physical examination, the patient presented with hands, heels and facial edema, as well as warmth in the joints of her hands, claudication and a distented abdomen with referred pain on deeper palpation of the right hypochondriac region. Laboratory exams were within the normal limts and the abdominal computed tomography (CT) scan did not show any significant alterations. An in-depth cardiologic investigation was performed in the patient to analyze a possible cardiac-induced edema. However, the Electrocardiogram (EKG), the Transesophageal Echocardiogram (ECOTT), Ambulatory Blood Pressure Monitoring (ABPM) and Lower Limbs Venous Doppler US exams did not show any pathological changes. Therefore, the main hypothesis raised was an antiseizure drug-induced edema, specifically a gabapentin major side effect. **Discussion:** Gabapentin is an antiseizure medication (ASM) mainly used as an adjunct to more potent ASM for managing focal seizures, as it binds to presynaptic voltage-dependent calcium channels, stabilizing the neuronal excitability. Among its various aderse effects, there is weight gain associated with peripheral edema, Although the exact mechanism of its inducing edema remains unknown, some studies suggest that gabapentin might induce peripheral edema in a similar way as calcium channel blockers, such as Amlodipne, do: stimulating arterial dilation without venous compensation. In the presented case, as most frequent causes of edema, such as cardiac, nephrologic and hepatic originated edema, were ruled out, gabapentin was considered as a culprit. A dosis reduction resultued in the edema resolution on our patient. **Final comments:** In conclusion, we emphasize that, among the vairous causes that can possibly induce a peripheral edema, medical providers should always look for pharmacological causes, specially in those patientes on polypharmacy. Even though it isn't a common side effect, neurologists and other healthcare workers should be fully aware of the association between gabapentin and edema so that unecessary interventions can be avoided as the medication can be easily adjusted or even discontued on our patients.

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A huge bilateral paraphalccine cerebral meningioma case report treated in a multimodal way: endovascular and microsurgery

João Lazaro da Silva Rodriguez Oquendo[1], Maria Cândida Barros Arantes Romano[1], Júlia Oliveira e Freitas[1], Fábio Serra Barbosa da Silva[2]

[1]Centro Universitário Tocantinense Presidente Antônio Carlos, Araguaína TO, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Cândida Barros Arantes Romano; Email: candinha_barros-ar@hotmail.com.

This article presents the case of a bilateral, median, solid and volume (5x8x7cm), causing sinus thrombosis at this level, with symptoms non-specific, as the patient only had a headache, with no improvement following conservative treatment, and was submitted to a computerized tomography that showed the lesion. Due to the large extracranial vascular and bone afferent, prior embolization was performed of its main afferent right middle meningeal artery, and discusses its pathophysiology, its image findings, treatment and evolution. Parafalcine cerebral meningiomas represent 7.7% of all neoplasms meningoepithelial. They are currently divided according to their parallel relationship to the superior sagittal sinus into anterior, median (between coronal and lambdoid suture) and posterior. They may appear with non-specific symptoms, and may have differential diagnoses, migraine, tension headache, and psychiatric disorders, according to their location, due to slow growth and brain accommodation mechanisms, respecting the monro-kelly doctrine. The severity of the expansive lesion presented is evident, thanks to the technological advancement, hemodynamic techniques, intraoperative management of hemostasis of the lesion occurs in a more succinct and abbreviated manner, due to the embolization of the tumor previously at first, reducing the morbidity and

mortality of surgery, which in other times was extremely high. Currently, there are still more studies on the topic required in order making the more precise and cost-effective technique, as the technology is new and very costly for the public health system.

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A mechanistic analysis of the neural modulation of the inflammatory system through vagus nerve stimulation: a systematic review and meta-analysis

Paulo Sampaio de Melo[1], Anna C. Gianlorenco[2], Anna Marduy[3], Chi K. Kim[4], Hyuk Choi[5], Jae-jun Song[5], Felipe Fregni[6]

[1]Hospital Santa Izabel, Salvador BA, Brazil.
[2]Universidade Federal de São Carlos, São Carlos SP, Brazil.
[3]União Metropolitana de Ensino e Cultura, Lauro Freitas BA, Brazil.
[4]Korea University, Guro Hospital, Seoul, South Korea.
[5]Korea University, Seoul, South Korea.
[6]Harvard Medical School, Massachusetts, United States of America.

Correspondence: Paulo Ricardo Sampaio de Melo; Email: melosrpaulo@gmail.com.

Background: Current preliminary evidence suggests that VNS modulates the inflammatory system through intricate routes, including neuroendocrine mediated and immunologic communication. Numerous studies have delved into the anti-inflammatory effects of VNS, particularly with cervical and auricular stimulation. Moreover, to the best of our knowledge, despite various studies on the antiinflammatory effects of VNS, a comprehensive review and analysis pooling the results of these studies is currently lacking. **Objective:** To conduct a systematic review and meta-analysis assessing the antiinflammatory effects of various VNS methods while exploring multiple antiinflammatory pathways. **Methods:** We included clinical trials that used electrical stimulation of the vagus nerve and assessed inflammatory markers up to October 2022. We excluded studies lacking control groups, those with combined interventions, or abstracts without full text. We adhered to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses guidelines and the Cochrane Handbook for Systematic Reviews. For each inflammatory marker, a random-effects meta-analysis using the inverse variance method was performed. Methods used include transcutaneous auricular VNS (taVNS), transcutaneous cervical VNS (tcVNS), invasive cervical VNS (iVNS), and electroacupuncture VNS (eaVNS). Main reported outcomes included tumor necrosis factor (TNF)-α, interleukin (IL)-6, IL-1ß, C-reactive protein (CRP), and IL-10. Risk of bias was evaluated using the Cochrane Collaboration Tool (RoB 2.0). **Results:** This review included 15 studies, involving 597 patients. No statistically significant general VNS effect was observed on TNF-α, IL-6, and IL-1ß. However, CRP, IL-10, and interferon (IFN)-γ were significantly modulated by VNS across all methods. Subgroup analysis revealed specific stimulation techniques producing significant results, such as taVNS effects in IL-1ß and IL-10, and iVNS in IL-6, whereas tcVNS and eaVNS did not convey significant pooled results individually. Cumulative exposure to VNS, higher risk of bias, study design, and pulse width were identified as effect size predictors in our meta-regression models. **Conclusion:** Pooling all VNS techniques indicated the ability of VNS to modulate inflammatory markers such as CRP, IL-10, and IFN-γ. Individually, methods such as taVNS were effective in modulating IL-1ß and IL-10, whereas iVNS modulated IL-6. However, different VNS techniques should be separately analyzed in larger, homogeneous, and powerful studies to achieve a clearer and more consistent understanding of the effect of each VNS method on the inflammatory system.

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A non-internuclear cause of walled-eye ophthalmoplegia

João Vitor Gerdulli Tamanini[1], Sofia Monaco Gama[1], Gabriel Casella Speltri[1], Fernanda Abreu[1], Clara Moreira[1], Guilherme Ribeiro[1], Jackeline Neves Pereira[1], Luccas Almeida[1], Larissa Mendes[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: João Vitor Gerdulli Tamanini; Email: jvgtamanini@yahoo.com.br.

Case presentation: A 60-year-old patient presented with a history of two days of facial palsy and binocular diplopia. Past medical history included an upper respiratory infection 1 week before. He presented right-sided hemifacial paralysis involving the superior and inferior quadrants, bilateral exotropia on primary gaze position, adduction restriction of the left eye when looking to the right, and of the right eye when looking to the left. Due to these findings, we made a hypothesis of wall-eyed bilateral internuclear ophthalmoplegia (WEBINO). However, on further examination it was noticed that the patient did not have any form of gaze-evoked nystagmus and presented with up-gaze and convergence palsy. Furthermore, the patient also presented with ataxic gait, absent osteotendinous reflexes and was unable to perform a tandem walk. Considering laboratory assessment, serum examinations were unremarkable, whereas spinal fluid analysis presented 0 cells and 57mg/dL of protein. Motor conduction studies showed reduced amplitude in the right facial nerve potential in the platism and the eye orbicularis muscles. There were no sensitive alterations. The F wave latency was increased in the tibial and normal in the ulnar nerves. The H reflex was absent bilaterally. Regarding the blink reflex, the R1 latency was increased on the right, whereas latency of the left R1 and bilateral R2 and R2c were within normal ranges. Given the clinical, laboratory and nerve conduction study findings we put forward a hypothesis of Miller Fisher syndrome (MFS). **Discussion:** MFS is a rare form of acute-onset immune-mediated neuropathy and is characterised by the triad: ophthalmoparesis, ataxia and areflexia. Specifically, MFS is highly associated with anti-GQ1b antibody. In that sense, both anti-GQ1b antibody and MFS have been associated with internuclear ophthalmoplegia and WEBINO. Our patient presented with several findings suggestive of MFS. Particularly, WEBINO consists of a form of bilateral internuclear ophthalmoplegia with large, wide-angle, exotropia in the primary position, bilateral adduction impairment and nystagmus of the abducting eye. Moreover, WEBINO typically presents with a wide range of vertical gaze alterations such as vertical gaze-evoked nystagmus, skew deviation and vestibular ocular reflex alterations. Lesions that cause WEBINO are frequently located in the pons and/or midbrain and are usually of inflammatory or ischemic aetiology. Particularly, the damaged structure is classically described as the middle longitudinal fasciculus. In a first assessment our patient seemed to present with WEBINO. However, the adduction paralysis,, as well as lack of abduction and gaze evoked nystagmus made the diagnosis of true WEBINO less likely. Normal MRI and alteration in nerve conduction corroborated the pseudo-WEBINO of peripheral nature. **Final comments:** Our patient presented with a pseudo-WEBINO of non-internuclear cause.

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A novel pattern of voxel-based morphometry is associated with focal cortical dysplasia: the "boomerang sign"

William Alves Martins[1], Eliseu Paglioli[1], Ricardo Paganin[1], Thomas More Frigeri[1], Rafael Paglioli[1], Ricardo Bernardi Soder[1], Natalie da Silveira Donida[1], Bibiana Liberman Thomé[1], Tais Michele Werle[1], Andre Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: William Alves Martins; Email: walvesm.br@gmail.com.

Background: More than 30% of patients with epilepsy have seizures refractory to medication and epilepsy surgery remains the only hope of sustained seizure-freedom. It has been shown that complete resection of an epileptogenic structural abnormality seen on MRI is the best predictor of surgical success3, yet a visible MRI lesion is not found in a significant number of patients. Post-processing MRI techniques may harbor a way to correctly identify previously undetected lesions. In this study, we report a relatively specific pattern of VBM abnormalities using the MAP program. **Objective:** To describe a new pattern of MAP positivity associated with focal cortical dysplasia (FCD) **Methods:** We studied all patients in the Porto Alegre Surgery Program (PAESP) who underwent surgery from 2006-2020, selecting patients with MRI negative or FCD positive MRI. All patients had a T1 volumetric sequence analysed using the MAP program, providing statistical maps of cortical-subcortical junction. A cut-off of > 4 SD with exclusion of artefacts upon MRI review was applied. Of 141 patients, MAP was positive in at least one region in 93 patients (66%) (83% in those with lesional MRI and 43% in nonlesional patients. A peculiar, previously undescribed pattern was observed in 29 (31%) of the MAP+ patients, consisting of a Z score > 4 in the "blurring map" at the depth of a sulcus, assuming the shape of a boomerang, its convex portion turned towards the subcortical white matter. Pathology was available in 22 of these patients and the "boomerang pattern" was significantly associated with FCD II (16/22 [73%], p=0.018;), with a trend both for better surgical outcome (55% x 42%, p =0.2) and to lesional MRI (34.8% x 22.2%, respectively; p=0.2). The "boomerang sign" was also linked to total resection of MAP+ region (69% x 42.2%, p=0.017; OR: 2.2 [1.1- 4.3]) and better surgical outcome when completely resected (70% x 22%, p=0.041). **Results:** We have described a new pattern of MAP positivity at the bottom of a sulcus in the format of a "boomerang". Huppertz el al had briefly mentioned a half ring- or horseshoe-shaped pattern in the extension map, but no further investigations were reported. In a study of 5 patients, Stecher et al suggested that a U-shaped asymmetry may underlie FCD in very young children. Here we further characterized this pattern of MAP positivity, showing that the "boomerang sign" is associated with FCD II in 73% of cases and its presence associates with higher rates of complete resection of MAP+ regions. **Conclusion:** The boomerang sign pattern may be a reliable visual clue of MAP positivity, perhaps increasing confidence in lesion localization. Furthermore, it may be a marker of bottom of sulcus dysplasia (BOSD), a peculiar type of FCD. Crucially, because subtle blurring is often seen at the bottom of sulci in patients with otherwise normal MRI, MAP may resolve the issue of whether those represent true abnormality.

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A novel variant of GJB1 related to demyelinating CMTX1 with tremor

Ana Beatriz Bataglia Sperandio[1], Laura Alonso Matheus Montouro[1]

[1]Centro Universitário Padre Albino, Faculdade de Medicina de Catanduva, Catanduva SP, Brazil.

Correspondence: Ana Beatriz Bataglia Sperandio; Email: anabsperandio@gmail.com.

Case presentation: Male, 55 years-old, of consanguineous parents (second cousins). After a fracture in his right ankle at the age of 34, he presented progressive foot deformities associated with paresthesias and weakness. After a few years, the condition progressed to the distal upper limbs associated with intense hand tremors. At

fifties, he needs a walking stick support. The patient has also morbid obesity, systemic arterial hypertension, diabetes mellitus type 2. He denies similar cases in family. On physical examination: atrophy of the thenar and predominantly hypothenar region, claw posture of fingers IV and V on the right and distal interphalangeal joint in swan head on fingers II, III, IV. Atrophy and weakness of legs and feet. High-frequency, medium-amplitude postural tremor in the upper limbs. Global areflexia. Severe sensory ataxia. Distal anormal proprioception. Electroneuromyography: no sensory action potential obtained in the four limbs, motor neuropathy with a primarily demyelinating pattern in the upper limbs (prolongation of distal motor latency and velocity of the compound muscle action potential < 20m/s) and severe neuropathy by non obtained motor potentials in lower limbs. Commercial panel of hereditary neuropathies reveals a hemizygous mutation in the GJB1, variant NM_000166 .6:c.120dup:p.(Glu41Argfs*6). **Discussion:** A large cohort with 160 cases of CMTX1, motor nerve conduction velocity (MNCV) of ulnar and median ranged to 35.0±5.7 and 33.7±5.0, respectively. Therefore, the present case has MNCV far below compared to the majority, reiterating the electrophysiological heterogeneity present in CMTX1. Patients with classic CMT disease phenotype plus tremor were known as Rossy-Lévy Syndrome, usually associated with CMT1A. Tremor is unusual in CMTX1 and has been described in some diseases variants. The c.-17G> A variant, referred by Tomaselli et al, was described in 5 male patients who had postural tremor. Other variants in literature associated with tremor: c.-170T> G, c.-17+1G> T, p.Ala39Val, c.278T> A, p.Arg-164Trp, p.Ile202Asp, p.Trp132Valfs*14, p.A271Lfs*121, p.R183S, c.303dup. By contrast, the Korean cohort by Hong et al, points out that 35% have tremor (21 different variants, some of which are Korean-specific). In Brazil, tremor in the lower limbs in two siblings with p.Trip132Arg was described by Nishiyama. Therefore, this present case has high-frequency postural tremor as one of his symptoms. This variant leads to a change in the reading frame by replacing a glutamic acid with an arginine and consequently early arrest of protein translation, resulting in a truncated protein. This variant is absent in gnomAD, scientific literature and ClinVar. By correlating the phenotype with the type of variant, the potential of its pathogenicity is very high. **Final comments:** This case contributes with a new variant of GJB1, corroborates the electrophysiological heterogeneity and reinforces the phenotypical association of CMTX1 with tremor.

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A portable device for assessing ankle angles during gait in post-stroke individuals: a prototyping study

Tatiana Souza Ribeiro[1], Raiff Simplício da Silva[1], Daiane Carla Rodrigues Cardoso[1], Vitor Vale Pereira[1], Lígia Pablícia Lopes Dantas[1], Maria Amanda Ferreira Quirino[1], Larissa Araujo Gomes[1], Aline Braga Galvão Silveira Fernandes[1], Jefferson Doolan Fernandes[2]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[2]Instituto Federal do Rio Grande do Norte, Parnamirim RN, Brazil.

Correspondence: Tatiana Souza Ribeiro; Email: tatiana.ribeiro@ufrn.br.

Background: Individuals after a stroke commonly experience gait alterations. Limitations in ankle range of motion are among the most common, particularly reduced dorsiflexion during the pre-swing phase and initial contact. Assessing ankle joint angles during gait can assist rehabilitation professionals in planning therapies to improve gait. Portable devices using inertial sensors offer excellent cost-effectiveness for this assessment. However, to the best of our knowledge, no studies have used this technology for this purpose in this population. **Objective:** This study aims to develop and preliminarily test the "Portable device for kinematic gait assessment of individuals post-stroke: DAMA" - a portable device using inertial sensors to obtain ankle angle measurements during the gait of post-stroke individuals. **Methods:** This is a cross-sectional prototyping study. It preliminarily included 3 healthy individuals aged between 20 and 70 years. For the test, the participants were equipped with the DAMA device and Qualisys system® markers for simultaneous data collection by both systems. They walked 4 meters at their usual speed 10 times to collect data on the maximum dorsiflexion angle (MD), maximum plantar flexion angle (MPF), and ankle angles at initial contact (IC) and toe-off (TO). The averages of these measurements were recorded. In the statistical analysis, we compared the data obtained by the two systems using the Wilcoxon test. We also examined the correlations between the averages obtained by the device and those obtained by the Qualisys system® using Spearman's correlation coefficient. **Results:** The means obtained with Qualisys were: IC = -1.93(±0.39); TO= -8.11 (±1.57); MPF= -15.75 (±1.77); MD= 10.90 (±0.55). The means obtained with Qualisys were: IC =5.87 (±4.60); TO= 2,83(±1,96); MPF= -23.56 (±2.78); MD= 12.73 (±2.88). There was no statistical difference between the outcomes collected by the device and those collected by Qualisys: IC (p=0.109); TO (p=0.109); MPF (p=0.109); MD (p=0.285). There was a negative correlation for IC (r=-1.000; p< 0.01) and a positive correlation for MPF (r=1.000; p< 0.01), and a not statistically significant correlation for TO and MD (r=0.5; p=0.667). **Conclusion:** Comparing the two analysis methods revealed no statistical difference between them. However, the limited sample size hinders making precise inferences about their accuracy. The correlation found may suggest good performance of the device for MPF. A larger sample size will be needed for further validation of DAMA in measuring ankle joint angles.

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A pure dysautonomic variant case of Guillain-Barré syndrome

Olga Cassol Silva[1], Joana Laurindo da Silva[1], André Luiz Pereira Martins[1], Carolina Aparecida Donofrio Santos Costa[1], Maria Eduarda Andreatta[1], Mariana Costa Borges[1], Cristine Koehler[1], Gabriela da Silva Cremonese[1], Rafael Marques Mendes[1], Ana Magda da Silva Bruscato[1]

[1]Hospital Governador Celso Ramos, Florianópolis SC, Brazil.

Correspondence: Olga Cassol Silva; Email: olgacassol@hotmail.com.

Case presentation: A 17 year-old female was transferred to a tertiary hospital due to orthostatic intolerance. She was previously healthy. 20 days before the presentation, she had an episode of abdominal pain with diarrhea, was evaluated in a local hospital, diagnosed with urinary infection and discharged with a 3 day course of Ciprofloxacin. After 2 days, she progressed with intense myalgia, fever, nausea and retro orbital pain and a day later with cough and odynophagia. She also had an episode of loss of consciousness that made her come to the ER and be admitted due to hyponatremia and leukocytosis. After laboratory analysis and thorax and abdomen TC exams, she was diagnosed with Chikungunya and atypical pneumonia, treated with Azithromycin and Ceftriaxone and discharged with only residual cough and mild myalgia. After 2 days, she was again transferred to the hospital with episodes of loss of consciousness related to orthostatic position. Her physical exam was normal (including sensitivity, strength and reflexes), except for orthostatic hypotension with no compensation of heart rate. Her blood pressure in lying position was 100x70mmHg and in sitting position after 1 and 3 minutes was 60x40mmHg. She didn't tolerate stand up position and had syncope episodes when tried. Laboratory analysis, head CT, electrocardiogram and echocardiography did not show abnormalities. Cerebrospinal fluid (CSF) analysis showed proteino-cytologic dissociation (1 cell to 90 proteins), glucose of 56 and 10 red blood cells. Electroneuromyography (EMG) revealed slowing of late responses (F waves) associated with incipient signs of sensory, axonal and asymmetric peripheral neuropathy. She was diagnosed with an atypical Guillain-Barré Syndrome (GBS) secondary to Chikungunya infection and treated with a 5 day course of intravenous Immunoglobulin with little improvement. She was discharged with Duloxetin and Fludrocortison for orthostatic hypotension, regarding a mild anxiety underlying condition, and oriented to wear compression socks. At her one-month revaluation consult, she had improved and was well tolerating sitting position and progressing with attempts of upright position during physiotherapy. **Discussion:** This case presents an atypical presentation of GBS, confirmed by typical CSF and EMG findings. Literature data estimates an autonomic dysfunction in ⅔ of all SGB, but the prevalence of a 'pure' dysautonomic syndrome is unknown. Regarding these patients, mortality rate is up to 7%, which reinforces the importance of early monitoring for respiratory or cardiovascular collapse. This patient had had 2 episodes of loss of consciousness at home and a few during medical evaluation, with no evidence of arrhythmias or respiratory dysfunction. **Final comments:** This case report reinforces the importance to keep a close eye on patients with atypical presentations after an infection condition in order to make a correct diagnosis to provide appropriate management and treatment.

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A rare adult-onset leukoencephalopathy as a likely etiology of neurocognitive decline: a case report

Allefy Beltrão Albano[1], Alvaro José Porto Moreira[1], Ana Dolores Firmino Santos do Nascimento[2], Luziany Carvalho Araújo[1], Breno José Alencar Pires Barbosa[1], Fabrício Martins Veras[1], Letícia Klabinske Marques Monteiro[1], Clélia Maria Ribeiro Franco[1], Mariana de Almeida Cruz Silva[1], Matheus Goiana Novaes Correia Carvalho[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Allefy Beltrão Albano; Email: allefymed@gmail.com.

Case presentation: 28-year-old female patient presented to our service, in 2024, for investigation of a cognitive condition characterized by progressive memory loss and difficulty of planning and executing routine activities that has evolved over the last year, accompanied by depersonalization, infantilism, loss of self-care, minimal social provision and urinary and fecal incontinence. Family members reporting two short-term paroxysmal events in the last three months, characterized by sudden loss of consciousness accompanied by hypertonia, with no report of limb shocks. Companion reports the absence of similar cases in the family and the patient was previously functional for daily and instrumental activities of living, with no previous reports of cognitive dysfunction. On physical examination, she presented preserved strength and trophism, with hyperreflexia in the lower limbs, non-straight tandem gait, unmotivated and constant laughter (pseudobulbar affect), distal stereotypical movements with the hands and teeth grinding, with previous self-mutilation in the dental arches. Cognitive examination has made difficult by lack of cooperation, showing temporospatial disorientation, telegraphic speech, significant executive dysfunction, without aphasia and apraxia. Magnetic resonance imaging of the brain was performed, which showed involvement of the periventricular white matter with anterior predominance, symmetrical in sagittal T2/FLAIR sequences.

Complementary tomographic study demonstrated small calcifications adjacent to the anterior horns of the lateral ventricles. **Discussion:** Adult-onset leukoencephalopathy with axonal spheroids and pigmented glia (ALSP) is a rare degenerative disease (prevalence less than 1/1.000.000) of the white matter, with onset in the third decade, characterized by axonal and glial damage due to mutations in the CSF1R gene. Age at onset of less than 60 years, associated with at least two of the following findings: cognitive decline, pyramidal syndrome, extrapyramidal signs and epilepsy, associated with typical findings on brain neuroimaging demonstrating bilateral white matter lesions, periventricular, with frontal predominance associated to specific calcifications, make the diagnosis probable even in the absence of a confirmatory genetic test for the specific mutation. However, such diagnostic criteria for ALSP have limited overall sensitivity along with modest specificity, requiring a comprehensive approach based on Magnetic Ressonance patterns, as classic patterns such as the case in question. **Final comments:** ALSP can represent a unique diagnostic challenge, given its particular adult-onset presentation and the low number of cases described in the literature, with early recognition being essential given the poor prognosis and the potential for familial genetic testing with goal of genetic counseling.

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A rare case of anti-MOG antibody positive encephalomyelitis following dengue infection

Raquel Maria Ayres Monteiro[1], Luis Otavio Caboclo[1]

[1]Faculdade Israelita de Ciências da Saúde Albert Einstein, São Paulo SP, Brazil.

Correspondence: Raquel Maria Ayres Monteiro; Email: raquelmamonteiro@gmail.com.

Case presentation: A 42-year-old female with history of breast cancer and currently on tamoxifen, was admitted for investigation of neurological complications following a recent symptomatic dengue infection. Seven days after onset of dengue, she began to exhibit bladder hesitation followed by urinary retention, abdominal pain, and absence of bowel movement. As her condition progressed, she developed lower limbs and hand grip weakness, and diplopia when looking to the left. Neurological examination indicated tetraparesis with muscle strength grade 4+, hyperesthesia and allodynia in T6 level, diplopia, horizontal conjugate gaze paresis, brisk reflexes, and no cognitive or language alterations. Brain MRI revealed high T2/FLAIR signal in the right cerebral peduncle and thalamus. Spinal MRI revealed extensive lesions with discrete high T2 signal along the cervical and thoracic cord, suggesting infectious, immune, or demyelinating etiology. Cerebrospinal fluid (CSF) showed lymphocytic pleocytosis and elevated protein. Viral panel in CSF was negative. Serum analysis revealed presence of anti-myelin oligodendrocyte glycoprotein (MOG) antibodies. Patient received high-dose methylprednisolone therapy for five days and underwent four sessions of plasmapheresis. After these interventions, the patient fully recovered, showing improved strength, normal balance and gait, and complete recovery of bladder function. **Discussion:** Dengue has been associated with neurological complications, directly through viral infection or indirectly through dysregulated immune responses. This is a rare of a patient with myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) following dengue infection. MOGAD symptoms include optic neuritis, extensive transverse myelitis, acute disseminated encephalomyelitis, brainstem or cerebellar features, or cerebral cortical encephalitis. MOGAD can present as either a monophasic or relapsing disease course, and MOG-IgG cell-based assays are important for diagnostic accuracy. Most patients experience good to excellent visual and motor recovery following corticosteroid and plasmapheresis therapy. However, permanent bladder, bowel, or sexual dysfunction can occur. The effective management of this patient underscores the potential for immune-mediated processes in post-dengue neurological disorders and suggests that early intervention can lead to significant recovery. **Final comments:** Autoimmune encephalomyelitis following dengue infection, particularly when associated with anti-MOG positivity, is rare and underscores the complexity of neurological manifestations in dengue. Early recognition and appropriate treatment are essential for a good prognosis.

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A rare case of cerebellar progressive multifocal leukoencephalopathy in a patient with HIV and subacute ataxia

Pedro Schmidt dos Reis Matos Figueiredo[1], Johnatan Felipe Ferreira da Conceição[1], Flávia de Aguiar Machado[1], Lucas Ladeira de Lucas[1], Helena do Amaral Barreto[1], Caroline Bittar Braune[1]

[1]Universidade Federal Fluminense, Hospital Universátio Antonio Pedro, Departamento de Neurologia, Niterói RJ, Brazil.

Correspondence: Pedro Schmidt dos Reis Matos Figueiredo; Email: pedrosch2010@gmail.com.

Case presentation: A 40-year-old female presented with a one-week history of uncoordinated movements and difficulty walking, accompanied by brief episodes of vertigo. She was diagnosed with HIV approximately 3 months before and since then was regularly on antiretroviral therapy (TDF + 3TC + DTG) and sulfamethoxazole-trimethoprim for neurotoxoplasmosis treatment. On neurological examination, she exhibited debilitating imbalance while standing, significant ataxia on finger-to-nose and heel-to-shin tests bilaterally and bilateral horizontal nystagmus. Laboratory tests showed a viral load of 600 copies/ml and a CD4 cell count of 113 cells/ml. Brain magnetic resonance imaging (MRI) revealed a lesion hyperintense on T2 and FLAIR sequences in the left cerebellar hemisphere, sparing the dentate nucleus, suggestive of the "shrimp sign" described in cases of progressive multifocal leukoencephalopathy (PML) (Figure 1A). Initial cerebrospinal fluid (CSF) analysis was normal, including PCR for John Cunningham (JC) virus and, therefore, no specific treatment was started, and the patient remained hospitalized for follow-up. One month after admission, a new brain MRI was performed, now showing bilateral cerebellar lesions with the "shrimp sign" appearance (Figure 1B), and a new CSF PCR for JC virus was requested and returned positive, thus establishing the diagnosis of PML. **Discussion:** Cerebellar ataxia in HIV patients can have metabolic, infectious, vascular, or autoimmune etiologies, and may or may not be related to the presence of the retrovirus itself. PML is an opportunistic demyelinating disease of the central nervous system, associated with the presence of the JC polyomavirus typically in patients with CD4 counts < 200 cells/ml, and can manifest as an ataxic syndrome due to asymmetric involvement of the cerebellum. The shrimp sign on brain MRI has high sensitivity and specificity for the diagnosis of cerebellar PML. The diagnosis of PML without histopathological findings is based on a combination of typical clinical and neuroimaging findings associated with JC virus positivity in the CSF. When these three criteria are met, the diagnosis is definitive, according to AAN criteria. The treatment of HIV-associated PLM typically involves improving the patients immune status with the use of antiretroviral drugs, although the neurological deficits observed are usually irreversible. **Final comments:** New-onset cerebellar ataxia in HIV patients can be an uncommon clinical manifestation of PML and should be considered among the differential diagnoses, especially in patients with low CD4 cell counts. Despite the improved survival of these patients with the initiation of effective antiretroviral therapy, this condition remains serious and usually has a poor neurological prognosis.

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A rare case of cryoglobulinemia associated with hepatitis B virus infection

Carolina Braga Moura[1], Ana Maria Porto Cortines Laxe Martins da Silva[1], Ana Luiza Morgado[1], Camila Castelo Branco Pupe[1], Osvaldo J. M. Nascimento[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Ana Maria Porto Cortines Laxe Martins da Silva; Email: analaxe@id.uff.br.

Case presentation: A 48-year-old Brazilian woman reported progressive numbness along the lateral aspect of her right leg, evolving into constant numbness in her left foot and intermittent hand numbness, accompanied by weakness in writing. Her medical history included a neonatal blood transfusion, HBV diagnosis at age 20, and recent entecavir initiation. She also had skin lesions diagnosed as urticaria, a thyroid nodule, and a family history of Charcot-Marie-Tooth disease type 1A (CMT1A). Upon neurological examination, the patient exhibited normal gait and muscle strength but presented with distal hypoesthesia, particularly in the lower limbs, asymmetry, and hyperchromic skin lesions. Nerve conduction studies indicated asymmetric sensory axonal polyneuropathy, predominantly in the lower limbs. Laboratory tests confirmed active HBV replication (reactive Anti-HBe, HBV DNA: 759 IU/mL) and cryoglobulinemia. Complement levels were notably reduced (C3: 83 mg/dL, CH50: < 12.8 IU/mL). Genetic testing for CMT1A was negative. Initial treatment with oral prednisone (1 g/kg) and entecavir was ineffective, leading to the initiation of plasma exchange. After one month, the patient experienced persistent numbness in her left foot, but her overall neurological condition improved. Sensory responses, including vibration, tactile, and pain sensation, showed slight improvement. Post-treatment, cryoglobulins were undetectable, and complement levels normalized. **Discussion:** Cryoglobulins are often associated with underlying conditions such as chronic viral infections, chronic liver or kidney disease, autoimmune diseases, lymphoproliferative disorders and genetic factors. Hepatitis C virus (HCV) infection has been the most common etiology associated with cryoglobulinemia, with over 90% of mixed cryoglobulinemias (MCs) linked to HCV. This case underscores the rare occurrence of cryoglobulinemia due to HBV. The presentation with axonal polyneuropathy and skin lesions mimicked other neuropathies, complicating diagnosis. The persistence of symptoms despite initial treatment highlights the necessity for alternative therapeutic strategies such as plasma exchange. **Final comments:** This case contributes to the understanding of cryoglobulinemia associated with HBV, emphasizing the importance of considering HBV in differential diagnoses of cryoglobulinemia. It also underscores the need for vigilant monitoring and flexible treatment approaches to manage this rare but significant complication. The findings highlight the necessity for further research to better understand the pathophysiology and optimal management of HBV-related cryoglobulinemia, potentially improving outcomes for similar patients in the future.

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A rare case of hemichorea secondary to giant perivascular space: a case report

Rafael Saliba Helmer[1], Mayko Nascimento Merscher[1], Camila Caroso Lobo[1], Verena Miranda de Souza[1], Maria Paula Oiticica de Jesus[1], Thiago Ganho[1], Luiz Fernando de Oliveira Urzeda[1], Laura Araujo Paulino[1], Walderico Silva Machado Filho[1], Raissa Gomes Castro e Souza[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Rafael Saliba Helmer; Email: rsalibahelmer@gmail.com.

Case presentation: A 67-year-old male patient was brought to the emergency department due to his daughter's observation of involuntary movements in his right hemibody. The onset of symptoms remained uncertain as the patient had not initially perceived the movements. His medical history included multiple myeloma, which had shown a complete response to chemotherapy and autologous bone marrow transplant in 2022, as well as chronic obstructive pulmonary disease and hypertension. Neurological examination revealed choreiform movements predominantly affecting the distal aspect of the right hemibody, including the ipsilateral hemiface. Brain CT and angio-CT revealed no significant findings apart from an oval hypodensity in the left nucleocapsular region. Subsequent 3T brain MRI revealed an enlarged perivascular space in the left nucleocapsular and sublenticular regions, measuring 1.6x1.6 centimeters. All laboratory tests, including metabolic, hematological, and rheumatological profiles, showed no alterations. Further investigation failed to identify additional conditions justifying the observed symptoms. **Discussion:** Perivascular spaces, typically asymptomatic, may lead to neurological symptoms when extremely enlarged, termed tumefactive Virchow-Robin spaces. Despite the limited literature discussing the consequences of such enlargement, rare symptomatic cases, including chorea, have been documented. The mechanism linking perivascular space enlargement to movement disorders remains unclear, potentially involving proximity to the basal ganglia or disruption of adjacent white matter tracts. This case underscores the importance of considering enlarged perivascular spaces in the differential diagnosis of unilateral movement disorders, urging further research into their pathophysiology for improved diagnostic and management strategies. **Final comments:** Our case highlights that giant perivascular spaces, particularly near the basal ganglia, can present as unilateral movement disorders, such as chorea, despite typically being asymptomatic. This underscores the importance of considering this rare etiology in the differential diagnosis of such conditions, emphasizing the need for comprehensive evaluation and awareness among clinicians.

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A rare case of hypnic headache after meningitis in a patient with HIV, primary or secondary? Case report

Juan Sebastián Sánchez León[1,2], Marcela Nataly Parra Alvarez[1,2], Renato Dumba Monteiro de Castro[1,2], Tais Luise Denicol[1,2], Julian Gomez Gonzalez[1,2], Carolina Matte Dagostini[1,2], Karla Sarai Sandoval[1,2], Pedro Hall Ruschel[1,2], Liselotte Menke Barea[1,2]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Juan Sebastián Sánchez León; Email: juansebastian1516@hotmail.com.

Case presentation: A 76 year old female started 5 years ago with holocranial headache during sleep that cause awakening between 3 and 4 hours in the morning with duration of 20 to 30 minutes without response to treatment with simple analgesic, significantly affecting her quality life. Patient didn't have previous history of headache. One year before starting headache, she had an bacterial meningitis and was diagnosed with HIV, after treatment of meningitis she started antiretroviral therapy. First she started with intermittent bilateral temple headache from mild to moderate intensity, but after few months, headache change presentation to current characteristics. For these symptoms was decided to make a cerebrospinal fluid analysis that was normal and an brain MRI that showed ventricular enlargement disproportionate to cerebral atrophy, being suspected of normal pressure hydrocephalus; however, she didn't have compatible clinical characteristics, so that diagnosis was ruled out. Due to the characteristics of the headache we diagnosed of hypnic headache and started treatment with Indomethacin 25 milligrams once at night. After treatment the patient improved significantly and she didn't have more episodes of headache. **Discussion:** Hypnic headache is a rare primary headache that is characterized by equal or more than 10 headache episodes for month during sleep that cause awakening, with duration from 15 minutes to 4 hours without cranial autonomic symptoms and without better accounted for other ICHD-3 diagnosis. It generally occurs after the age of 50, in the medical literature there are some reports of secondary hypnic headache cases being associated with brain tumors, vascular pathologies, obstructive sleep apnea, low glucose, nocturnal hypertension or associated with medications which in this case were discarded. Headache associated with meningoencephalitis according to ICHD-3 need a temporal association, parallel or on the onset of disease which contrasts with our case, reducing the probability of being secondary, even though there is no greater evidence of this probable association because hypnic headache is a rare type of headache. **Final comments:** Although hypnic headache is rare, it must be suspected in cases of headache during sleep that causes awakening in order to initiate treatment that improves the quality of life of the patients.

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A rare case of spontaneous hypertensive pneumocephalus due to neurotuberculosis

Mayara Romero Canal[1], Mariana Tonon Giovannone[1], Natália Furlani Kassouf[1], Beatriz Gadel Medeiros[1], Natasha Soares Cutolo[1], Ana Luisa Vilela Sá Fortes[1], Fernando Pierini Costa[1], William Thiago Almeida Leite[1], Mauro Gomes Araujo[1], Eduardo de Almeida Guimarães[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Mayara Romero Canal; Email: academico.mayara.canal@gmail.com.

Case presentation: A 78-year-old patient seeks emergency care due to a severe, pulsating headache associated with rhinorrhea that started 14 days ago and worsened over the last 2 days. He had hypertension, DM II, obesity and treatment for neurotuberculosis in childhood. On physical examination, translucent liquid was noted flowing through the nasal cavity associated with signs of meningeal irritation without other changes. initially a head tomography and CSF collection were performed which showed pneumocephalus with frontal air level and bone discontinuation in the E. sphenoid bone (Figure 1 and 2) in the presence of meningoencephalitis (More than 3600 cells with neutrophilic predominance). **Discussion:** Intracerebral aerocele is defined by the presence of gases in regions of the central nervous system and is most commonly observed after trauma, surgery and neoplasms, but rarely spontaneously in infections caused by gas-producing bacteria. Its evolution can be good (asymptomatic) or dramatic to the most aggressive form (hypertensive pneumocephalus) representing a medical emergency. Symptoms are varied (depending on intensity and location) with non-specific neurological deficits, convulsions and changes in the level of consciousness. When located in the posterior fossa, it can displace important structures and result in neurological shock, coma and brain death. For proper diagnosis, a cranial tomography is considered, the change is revealed by the Mount Fuji sign (pathognomonic). The mechanisms of the disease are explained by two main theories: Dandy ("ball valve") which highlights the compaction of gas due to an imbalance of external and intracranial pressure associated with the rupture of dural fissures at the base of the skull that are obstructed by brain structures once opened. And the second Horowitz ("inverted bottle effect") which points to the increase in negative pressure due to the reduction of CSF by external mechanisms with compensation of air entry to balance the pressure differential. **Final comments:** In the case described above, we hypothesized that cerebrospinal fluid rhinorrhea was one of the causes of the pneumatocele, which occurred due to the destruction of brain space connections and also to cerebrospinal fluid loss, involving both mechanisms described. These events can occur in diseases such as tuberculosis and histoplasmosis. The Nasal/sinusoidal infectious manifestation of Mycobacterium Tuberculosis causing the above findings is poorly documented in the medical literature. In general, we observe masses or granulomas in lesion sites associated with B symptoms. In the present case, the pulmonary form was previously treated in childhood, with reactivation occurring in adulthood as an extrapulmonary form, later progressing to sphenoid bone erosion, meningoencephalitis and spontaneous hypertensive pneumocephalus.

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A rare case report: central venous thrombosis associated with a thoracic spontaneous cerebrospinal fluid fistula

Angelo Ponte de Freitas Campos[1], Rodrigo de Paiva Bezerra[1], Marcel Ken Uehara[1], Gisele Sampaio Silva[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Gisele Sampaio Silva; Email: giselesampaio@hotmail.com.

Case presentation: We describe the case of a 35-year-old female patient with a history of spontaneous lower back pain for three weeks, accompanied by nausea, vomiting, and one episode of diarrhea. She denied any history of trauma. The day after the onset of symptoms, she developed an intense, oppressive headache, associated with photophobia and blurred vision. The headache worsened when upright and improved when lying down. Cerebral spinal fluid demonstrated only slight hyperproteinorachia, without pleocytosis and no increase in lactate. Cisternoscintigraphy demonstrated marked delay in cerebrospinal fluid circulation through the usual pathways. She underwent MRI and cranial angio-MRI, which, in addition to the signs suggestive intracranial hypotension, also demonstrated venous thrombosis in subacute evolution, partially recanalized, in the superior sagittal sinus, transverse sinuses, and more recent thrombosis in the cortical veins of the parietal convexities. Further investigation with myelotomography confirmed a ventral median cerebrospinal fluid fistula at the T1-T2 level. The patient underwent a blood-patch and fibrin glue procedure via bilateral foraminal T2-T3 and interlaminar blood-patch at L4-L5. For treatment of CVT she was anticoagulated with apixaban every 12 hours. **Discussion:** Cerebrospinal fluid hypotension headache is a rare and currently underdiagnosed condition. It is generally a cause of chronic headache, which can be spontaneous, more rarely, or secondary to other causes such as trauma, surgeries, cancers, or even congenital diseases. The diagnosis is challenging, as neuroimaging findings can be subtle. Therefore, a high level of suspicion is required, along with image evaluation by experienced radiologists. Magnetic resonance imaging (MRI) of the brain is preferable and could show some indirect and nonspecific signs. Myelotomography is considered the gold standard, although very limited. This exam can identify the exact site of epidural CSF leakage. The timing of diagnosis is crucial for the occurrence of complications, most of which are severe. Treatment involves clinical measures such as hydration and increased caffeine intake, with or

without the use of corticosteroids (conflicting data in the literature). Epidural Blood Patch (EBP) should be considered in patients refractory to clinical measures. **Final comments:** Cerebrospinal fluid hypotension is a rare and still underdiagnosed entity, but it is responsible for high morbidity associated with potentially severe complications. High clinical suspicion combined with insightful radiological interpretation is crucial for diagnosing this disease. Early diagnosis and therapeutic intervention can alter the natural history of the disease and reduce unfavorable outcomes.

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A rare cause of clival mass: giant cell tumor case report and review of literature

Ligia Henriques Coronatto[1], Hennan Salzedas Teixeira[1], Roberto Sobocinski Castro[1], Thaís Rodrigues Barbosa Geraldo[1], Bruna Gutierres Gambirasio[1], Adrialdo José Santos[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Ligia Henriques Coronatto; Email: liscorona@hotmail.com.

Case presentation: A 16-year-old male referred horizontal binocular diplopia two months after a new-onset occipital headache started. Neurological examination revealed right eye abduction paresis. CT scan and later MRI evidenced an expansive lesion that seemed to originate from the clivus, partially calcified in the periphery, that occupied the suprasellar, sellar, and chiasmatic region, also affecting the nasal fossa and paranasal sinuses. MRI confirmed a non-enhancing heterogeneous lesion at this site. He was submitted to endoscopic transsphenoidal partial resection of the clival mass. Pathology discloses giant cells marked for CD68 at immunochemistry (osteoclasts). Histiocytosis markers such as S100 were negative; desmin (a myogenic marker present in myofibroblastic tumors and some sarcomas) was also negative. Ki67 was present in less than 1% of cells, indicating indolent growth. The tissue pattern and immunochemistry were compatible with Giant Cell Tumor of the Clivus. Two months after surgery, the patient's double vision resolved. Adjuvant radiotherapy was performed. In the following months, brain imaging showed tumor shrinkage. Eight months after the end of radiotherapy, the patient remains asymptomatic and MRI remains stable. **Discussion:** Giant Cell Tumor (GCT) is a benign osteolytic neoplasm, most common in young adults between 20 and 40 years old, mainly affecting the metaphyseal or epiphyseal region of the tibia and femur. It represents 4-10% of all primary bone tumors and presents locally aggressive behavior with, rarely, metastases. The pathophysiology is uncertain. Involvement of the skull is rare, accounting for less than 1% of all GCT cases. The sphenoid and temporal bones are more commonly affected. Clivus involvement is rare with very few reports in the literature. The most frequently reported symptoms of GCT of the clivus are headache and diplopia, often associated with increased intracranial pressure and sixth cranial nerve palsy. Total resection of the lesion is generally unfeasible requiring adjuvant therapy. Radiotherapy is most often used for residual tumors and local control. Despite the known efficacy of Denosumab (anti-RANKL monoclonal antibody) for the treatment of extracranial GCTs, little is known about the effectiveness of chemotherapy treatment for giant cell tumors of the clivus. Lines of adjuvant chemotherapy have been described using Doxorubicin, Cisplatin, Methotrexate, Zolendronate, and Raloxefin. **Final comments:** GCT of the clivus remains a diagnostic and therapeutic challenge due to its rarity and locally aggressive behavior. The difficulty in achieving total resection of the lesion requires adjuvant therapies for treatment optimization. Due to the rarity of this disease in the clivus, we describe this interesting case report with excellent response to resection and radiotherapy.

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A rare cause of fecal and urinary incontinence after cerebrospinal fistula surgery: lumbosacral radiculopathy and cauda equina syndrome caused by intrathecal fluorescein - a case report

Amanda Miranda de Mendonça[1], Matheus Alves da Silva[1], Ana Luisa Vieira de Faria[1], Eduardo Mesquita de Souza[1], Rafael Bragança Rodrigues Matias[1], Alex Araujo Fernandes[1], Matheus Luís Braga Munareto[1], Christiane Monteiro de Siqueira Campos[1], Alex Machado Baeta[1], Marcelo Freitas Schmid[1]

[1]Hospital Beneficência Portuguesa, São Paulo SP, Brazil.

Correspondence: Amanda Miranda de Mendonça; Email: amanda.miranda.dv@gmail.com.

Case presentation: A 23 years-old female patient presented with hyaline rhinorrhea, compatible with spontaneous cerebrospinal fluid fistula, which was confirmed through a cisternography, and the patient underwent microsurgical correction of skull base fistula via endoscopic endonasal approach which required a lumbar puncture and intrathecal fluorescein injection . The patient presented epileptic seizures during weaning from sedation, controlled with intravenous lacosamide. During the postoperative care, the patient presented urinary and fecal retention, as well as hypoesthesia in the gluteal region. Her neurological exam showed no muscle weakness, and the only alteration was a hypoesthesia on the gluteal region. Abdomen and pelvis CT showed no alterations. Cerebrospinal fluid (CSF) analysis revealed 12 cells with lymphocytic predominance, 52mg/dL of proteins and 64mg/dL of glucose. Pelvic and lumbosacral plexus MRI revealed hypersignal in the left sciatic nerve, compatible with acute denervation. Electroneuromyography revealed acute motor axonal impairment of the pudendal nerves bilaterally, favoring the hypothesis of radiculopathy from S2 to S4. Infection and rheumatologic diseases were ruled out. Due to the CSF with an inflammatory pattern and the electroneuromyography and MRI, corticosteroid therapy was started with intravenous methylprednisolone 500mg for 3 days associated with urologic physiotherapy, with clinical improvement. **Discussion:** Lumbosacral radiculopathy is most commonly caused by nerve root compression. Noncompressive mechanisms, such as infection, trauma, inflammation, neoplasm and vascular disease are rare. Signs and symptoms vary with which nerve root is affected. The cauda equina syndrome is caused by an injury of two or more nerve roots constituting the cauda equina. Its typically associated with marked neurologic disability, presenting with bilateral leg weakness in multiple root distributions and perineal sensory symptoms, as well as bowel, bladder and sexual dysfunction due to involvement of the S2-S4 spinal nerve roots. Fluorescein is used intrathecally to localize with more precision cerebrospinal fluid fistulas on skull base. Fluorescein can cause weakness in the extremities, epileptic seizures, numbness and cranial nerve deficits, but these are rare and reversible. Studies show that intrathecal fluorescein can cause discrete meningeal reactions, such as an increase in cells between 24 and 48 hours after the procedure and an increase in neutrophils within 24 hours, with a subsequent decrease within 48 hours, usually asymptomatic. **Final comments:** We present a case report of a lumbosacral radiculopathy and cauda equina syndrome caused by the use of fluorescein, a rare case not found by our group in the literature.The use of fluorescein is usually safe, but unusual reactions may be present.

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A rare differential diagnosis of bilateral optic neuropathy

Matheus Hisamitsu Facine[1], Humberto Ramos Crispim[1], Lucas Do Rego Barros Correa Martins[1], Caique Alberto Dosualdo[1], Gabriel Pina Paiva[1], Andressa Regina de Mello Galego[1], Marina Gaglione Teodoro da Silva[1], Murilo Wendeborn Rodrigues[1], Ricardo Kendy Nishiuchi Paiva[1], Renato Arantes Lima Simoes[1]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Humberto Ramos Crispim; Email: hrcrispim.uni@gmail.com.

Case presentation: A forty years old woman, started within 04 days before admission a new onset headache, pulsatile, high intensity, associated to conjunctival hyperemia and lacrimation. Evolving in 02 days, with bilateral blurred vision acuity, with central scotoma, which was progressively getting worse and affiliated to pain when moving her eyes. She denied any other symptom. Her background history was unremarkable as well as her family history.The general and neurological examination was normal, except by the central scotoma and conjunctival hyperemia. She could count fingers at 10 inches bilateral, pupillary reflex was normal and she has no Relative Afferent Pupillary Defect (RAPD) or dyschromatopsia. The intraocular pressure(IOP) and extrinsic eye movement was normal as well. There was no apparent corneal deposit. Fundus examination (FE) noticed diffuse bilateral serous retinal detachment and pockets of fluid in the outer retinal on the left eye. No dermatologic or hearing alteration was seen. Her blood exams, serology and rheumatological tests were normal. Sedimentation velocity was slightly high (49mm/h).Cerebrospinal fluid (CSF) shows pleocytosis and proteinorrachia with normal opening pressure. Confirmed intrathecal production of Immunoglobulin G (IgG). Serum Antibody Anti aquaporin-4 receptor (AQP-4) negative and Oligoclonal Band (BOC) CSF was negative? The Brain Magnetic Resonance Imagem (MRI) showed diffuse parietal thickening with gadolinium enhancement and deposit on eyeball, as well as, posterior wall vitreous chamber thickening. Optical Coherence Tomography (OCT) showed retinal detachment in a bullous serous pattern. Fundus Flurouscein Angiography (FFA) showed multiple bacilar detachment and typical pinpoint leaks - presuming Vogt-Koyanagi-Harada (VKH) syndrome. The patient filled criteria for VHK incomplete syndrome. The treatment was promptly started using Methylprednisolone 1 gram/day for 5 days. She had an excellent improvement in headache and visual acuity: 20/60 left eye and 20/80 right eye was discharged using prednisone 1mg/kg/day. On the 14 days follow up the FE showed resolution of serous detachment and the control OCT showed presence of pigmentary detachment in the right eye, and improvement of retinal detachment. **Discussion:** VKH is rare systemic disease involving various melanocyte-containing organs. This exact cause remains unknown, but there are evidences of an autoimmune process against an antigenic component on the melanocyte configuring an inflammatory T-cell-mediated immune response. The early treatment is essential to avoid complications and permanent vision loss. In this case the patient filled criteria for Incomplete VHK, she had a good improvement on visual acuity after using corticosteroid, confirmed on OCT returned of foveal depression and restored retinal parallelism layer. **Final comments:** This case report highlighted the importance of fast diagnosis VHK to a better prognosis and recovery.

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A rare form of genetic X-linked intellectual disability: case report

Ana Beatriz Landim Ribeiro[1], Luciana Amorim de Matos[1], Giovana Rodrigues França[1], Lisandra Mesquita Batista[2], Carlos Alexandre Gomes Farias[1], Paulo Victor Zattar Ribeiro[2], Lucca Biolcati Palavani[3]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.
[3]Max Planck University Center, Berlim, Germany.

Correspondence: Ana Beatriz Landim Ribeiro; Email: ablr2016@icloud.com.

Case presentation: We presented a 12-year-old boy who started a genetic investigation because of speech and facial dysmorphisms. He was born at 40 weeks of gestation without any complications via Cesarean operation. His birth length was 49cm, and his birth weight was 3.100. During his childhood he was presented with development delay, and still does not speak sentences, he only says the word "no". Today he remains with mild intellectual disabilities. As for other important manifestations, he presented with supratentorial ventriculomegaly, short stature, and behavior alterations such as aggressivity. He also presented with normal initial investigation for FMR1 disorders and karyotype revealing 46XY as result. As dysmorphic findings, we could notice short stature, malar flattening, thick vermillion of the lips, large mouth, sparse eyebrows, and micrognathia. He has no history of genetic diseases in her family, but his mom and his two brothers also had mild intellectual disabilities. He was submitted to a CGH array analysis, and it was found a pathogenic variant XQ25(123832142_124348427)X2 compatible with the diagnosis of Xq25 duplication syndrome. **Discussion:** It's known that Xq25 duplication syndrome is an X-linked neurodevelopmental disorder, characterized by delayed development and intellectual disability associated with abnormal behavior and dysmorphic facial features. To date, there are only thirty-two documented cases in scientific literature. We have described the clinical case of a patient presenting with many of the common features of the Xq25 duplication syndrome. A comparison of our patients phenotype with those previously reported showed similarities, including severe speech delay, facial features and intellectual disabilities. With that being said, it's known that the Xq25 duplication syndrome seems to be a complex condition mainly characterized by neuro psychomotor delay, intellectual disabilities, speech disturbances, epilepsy, brain abnormalities, and organ defects. Multiple X-linked intellectual disabilities are described every year and this reminds us of the importance of differential diagnosis when encountering a boy with intellectual disabilities. **Final comments:** Further detailed reports are certainly needed to better characterize the spectrum associated with this condition and, ultimately, help physicians to assist these patients comprehensively.

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A relapse of coccidioidomycosis meningitis without pulmonary involvement associated with obstructive hydrocephalus and ischemic stroke successfully treated with voriconazole and amphotericin B: a case report

Ana Luisa Vieira de Faria[1], Matheus Alves da Silva[1], Amanda Miranda de Mendonça[1], Eduardo Mesquita de Souza[1], Rafael Bragança Rodrigues Matias[1], Alex Araujo Fernandes[1], Matheus Luís Braga Munareto[1], Christiane Monteiro de Siqueira Campos[1], João Antonio Garreta Prats[1], Alex Machado Baeta[1]

[1]Hospital Beneficência Portuguesa, São Paulo SP, Brazil.

Correspondence: Ana Luisa Vieira de Faria; Email: alfaria96@gmail.com.

Case presentation: A 17-year-old male patient presented with a history of persistent headache. He was previously diagnosed and treated in the USA for coccidioidomycosis meningitis. His infection was complicated with obstructive hydrocephalus that demanded a ventriculoperitoneal shunt and also he developed an ischemic stroke. He was discharged back to Brazil with Fluconazol 1000 mg/day as a maintenance treatment presenting to our hospital after the new headache. His initial neurological examination was unremarkable. The initial CT scan revealed recurrence of the hydrocephalus, and the valve was regulated by the neurosurgery team. Suspecting of relapse of meningitis causing the valve dysfunction, an MRI was made and revealed dilation of the supratentorial ventricular system, diffuse leptomeningeal thickening and impregnation of the basal cisterns, including mesial temporal structures, third ventricle, midbrain, pons, trigeminal, abducens, facial and vestibulocochlear nerves. Also, a cerebrospinal fluid analysis revealed 308 cells with lymphocytic predominance, 188 mg/dL of proteins and 5 mg/dL of glucose. His initial opening pressure was 27 cmH2O. An initial infectious and immunodeficiency screening was negative. We associated Amphotericin B 5 mg/kg, with a subsequent switch from Fluconazole to Voriconazole. He evolved with clinical, radiological and cerebrospinal fluid improvement, being discharged with Voriconazole as a maintenance treatment. **Discussion:** Coccidioidomycosis meningitis is caused by a dimorphic fungus. They are endemic in regions of the southwestern United States, some areas of Central and South America. Spread occurs hematogenously after initial pulmonary infection and can cause destructive meningeal lesions. Meningitis is the most lethal complication of coccidioidomycosis infection. The most common clinical manifestation is persistent headache, present in 75% of patients. Signs of meningeal irritation on physical examination are usually absent; neck stiffness is rarely present. The cerebrospinal fluid presents an increase in cellularity, with a lymphocytic predominance. Glucose is usually reduced and is associated with a significant increase in proteins. We rarely find the organisms in direct microscopic examination of the CSF, if present it is an indication of a high fungal disease burden. On imaging examination we can find leptomeningeal enhancement, cerebral infarction resulting from vasculitis of small and medium vessels, in addition to hydrocephalus, which is the most common complication of coccidioidomycosis meningitis. **Final comments:** We present a case report of a rare infection in Brazil, to highlight the importance of correct follow up of these patients to manage the complications and infectious relapses.

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A slow and complex neurodegenerative disorder with predominant primary lateral sclerosis features in a patient with a erbb4 variant

Leonardo Elias Araujo dos Santos[1], Lívio Leite Barros[2] , Mariana Alcântara Tavares[1], Leonardo José Rodrigues de Araújo Melo[1], Francisco Luciano Honório Barreto Cavalcante[1] , Francisco Duque de Paiva Giudice Junior[1], Mariana Alcântara Tavares[1], Rodrigo Fagundes da Rosa[1], Emanuela Freire Caetano Davi[1], Francisco de Assis Aquino Gondim[3]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Hospital Universitário Walter Cantídio, Fortaleza CE, Brazil.
[3]Universidade Federal do Ceará, Departamento de Medicina Clínica, Fortaleza CE, Brazil.

Correspondence: Leonardo Elias Araujo dos Santos; Email: leonardoaraaujoo0@gmail.com.

Case presentation: A 69-year-old woman began experiencing symptoms in 1998 with rigidity in her toes, hands, and legs, as well as unspecified headaches. Her condition progressed to a gradual loss of movement, resulting in occasional falls and domestic accidents. She was diagnosed with Primary Lateral Sclerosis (PLS) in 2005 and started the use of riluzole. By 2006, she started to need a cane, progressed to a walker in 2012, and has been wheelchair-dependent since 2017. Current complaints include occasional paresthesias, pulsating headaches and persistent sharp pains in the lower limbs (predominantly left), starting in 2024. She performs basic daily activities with grab bars. She also developed nighttime hallucinations, consisting of hands entering through the window and rats on the ceiling, nausea before and during lunch, and urinary urge incontinence for 16 years. Her appetite and sleep are preserved. Past medical history includes congestive heart failure (CHF), arrhythmia and generalized anxiety disorder (GAD). She reports an extensive family history of dementia and psychiatric diseases and denies cases of cancer in relatives. Riluzole was discontinued in 2013 after repeated Electromyography (EMG) did not reveal involvement of lower motor neurons. The neurological examination revealed normal Mini-Mental State Examination (MMSE), mild hypomimia, mixed dysarthria (cerebellar and pyramidal) with voice and head tremor and hypophonia, slight saccadic dysmetria with ocular instability, spastic tetraparaparesis with action tremor predominantly on the left, cerebellar symptoms (index-finger to nose test dysmetria, dysdiadochokinesia), postural arm tremor, mild parkinsonism (bradykinesia, intention, and postural tremor) and rare fasciculations, global hyperreflexia (grade 4) with Babinski and Hoffman signs. She does meet criteria for essential tremor and with abnormal bilateral tremor on Archimedes spiral test. Genetic testing (June 2022) reveals the c.3182G> A variant in the ERBB4 gene, in heterozygosity, with uncertain significance. Neuromagnetic resonance (11/03/22) showed Fazekas score 2 for leukoaraiosis, MTA score 1 for hippocampal atrophy, and GCA score 1 for global cortical atrophy, with no hot bun cross sign in the brainstem. **Discussion:** Current knowledge about the ERBB4 gene indicates that it encodes a cell surface tyrosine kinase protein for neuregulins and EGF family members, regulating the development of the Central Nervous System. Although its significance is uncertain, the ERBB4 variant found suggests it may cause complex pathologies, including pyramidal involvement, parkinsonism, and cerebellar findings **Final comments:** This related case seems to expand the spectrum of ERBB4 related neurodegenerative disease, whose gene is currently associated with HER4 and ALS19. The case report was obtained with the patients understanding and written consent, agreeing to the publication of their clinical information

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A strengthening protocol based on core tasks in Parkinson's disease: a pilot study

Clara Rezende Rocha[1], Maria Eduarda Salum Aveiro[1], Caroline Rodrigues Osawa[1], Pedro Henrique Souza de Andrade[1], Artur Souza dos Santos[1], Daniel Alves de Menêzes[1], Thiago de Andrade[1], Gustavo José Luvizutto[1], Luciane Aparecida Pascucci Sande[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Clara Rezende Rocha; Email: d202010477@uftm.edu.br.

Background: Parkinson's disease (PD) is a neurodegenerative disease whose progression impairs the performance of activities of daily living (ADLs). Its main symptoms are tremor, bradykinesia, muscle rigidity and postural instability, making neurofunctional rehabilitation important for these individuals. Physical exercises are essential for reducing the PD progression and optimize the social participation of individuals. Despite numerous studies on the dose and type of exercise to be performed in PD, no strengthening protocol was found aimed the rehabilitation of core tasks. **Objective:** This is a pilot study that proposes a strengthening protocol aimed at maintaining the independence of PD patients in performing ADLs, with a focus on training core tasks (standing, walking, stepping up/down, and reaching/grasping/manipulating objects). **Methods:** This is a longitudinal study and case report with a 70-year-old man with PD for 12 years and stage 3 on the Hoehn and Yard scale. The protocol contains 9 exercises to be performed in ambulatorial environment and is divided into five weeks with the possibility of progression and regression in terms of difficulty and intensity. Fifteen sessions were realized and the volunteer was assessed using the Unified Parkinson's Disease Rating

Scale adapted by the Movement Disorder Society (MDS-UPDRS), the Dynamic Gait Index (DGI), the Montreal Cognitive Assessment (MoCA) and the Parkinson's Disease Questionnaire (PDQ - 39). **Results:** The protocol included exercises for reaching (anterior, superior and diagonal), step up and down (with and without resistance), free and resisted lower limb exercises aimed at postural transfers, trunk strengthening, gait and balance. The sessions lasted 40 minutes and for each exercise was performed 3 series of 8 to 12 repetitions, which were associated with cognitive double tasks. After five weeks, improvements were seen in the UPDRS (12 points) and DGI (1 point) scores, as well as in the Mobility (12.5%), ADLs (2.5%), cognition (16.67%) and communication (6.25%) domains of the PDQ-39. There was a worsening in the scores of the other domains of the PDQ-39 (emotional well-being, stigma, social support and bodily discomfort) and the MoCA. **Conclusion:** This study developed a strengthening protocol for PD rehabilitation. The protocol was applied for five weeks, three times a week, for 40 minutes to an individual with PD. Improvements were seen in dynamic balance, motor symptoms, mobility, performance of ADLs, cognition and communication, as recorded by the DGI, UPDRS and PDQ-39 scales. No improvement was observed in executive functions and memory (MoCA), as well as in emotional well-being, stigma, social support and bodily discomfort. The protocol can be applied in an outpatient setting and needs to be applied to more volunteers to assess its effectiveness and feasibility.

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A telehealth approach to developmental delays: assessing outpatient children in São Carlos, São Paulo

Guillermo Andrey Ariza Traslaviña[1], Melissa Giannina Sueros Torres[2] , Giovanni Scuracchio Baccala[2], Amanda Dourado Souza Akahosi Fernandes[2]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade Federal de São Carlos, São Carlos SP, Brazil.

Correspondence: Guillermo Andrey Ariza Traslaviña; Email: gtraslavina@fmrp.usp.br.

Background: On March 11, 2020, the World Health Organization officially declared COVID-19 a global pandemic. This announcement precipitated both individual and substantial collective challenges. In response, governments across the globe implemented specific regulations, which included strengthening regulatory frameworks and expanding remote healthcare services. Concurrently, there was a markedly high suppressed demand for outpatient services, particularly for children experiencing developmental delays. Consequently, in acknowledgment of the potential advantages of telehealth for multidisciplinary evaluations, a hybrid approach-integrating both remote and face-to-face methods-was instituted to assess children with suspected neurodevelopmental disorders. **Objective:** describe the clinical and demographic profiles of children evaluated through a multidisciplinary protocol, including neuropediatric, occupational therapy and psychology, between March 2021 and February 2023 at the Federal University of São Carlos. **Methods:** A general online invitation was sent to public and private health clinics, as well as schools. Each participant underwent at least three independent remote consultations, conducted by a team consisting of specialists in each specialty and undergraduate students. Each interview was scheduled according to the availability of the family members and followed a pre-established guide. After completing the initial interview process, a subsequent meeting involving only specialists was scheduled to elucidate the primary diagnostic hypotheses. Following this, a direct consultation with all the specialists was arranged, during which a physical and behavioral assessment was conducted in a controlled environment. At the end of the process, a final document was produced summarizing the consultation, primary diagnosis, and recommendations for treatment or follow-up. The activity concluded in February 2023. **Results:** A total of 94 children were evaluated, including 66 males (70.2%) and 28 females (29.8%). The majority (63%) were of White race, with an average age of 35.7 months. A high percentage had prenatal follow-up (88%), born at term (63%), and had a normal birth weight (63%). Approximately 75.8% were attending school. The evaluation revealed that 39.1% of the children showed delays in motor milestones, 75.8% in language development, and 65.2% in social skills. Subsequent follow-ups diagnosed 64% of the children with autism spectrum disorder. Other diagnoses included developmental delays due to lack of stimulation, epilepsy, reactive disorders, and other developmental and language conditions. **Conclusion:** This research highlights the effectiveness of a hybrid telehealth model, as a strategy to mitigate the high demand for specialized pediatric evaluations. The observed high frequency of autism among the participants may be attributed to preliminary screening by pediatricians or other health professionals prior to their inclusion in the activity.

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A US vs Brazil comparison of mild cognitive impairment conversion to Alzheimer's disease

Gabriella Chinaglia Zanin[1], Caio Hussid de Góes[1], Diego Olympio Peixoto Lopes[2], Maria Tereza Martinez[3], Fernanda Herculano[4], Diogo Haddad Santos[4]

[1]Faculdade de Ciências Médicas da Santa Casa de São Paulo, São Paulo SP, Brazil.
[2]Universidade Santo Amaro, São Paulo SP, Brazil.
[3]CPQuali Pesquisa Clínica, São Paulo SP, Brazil.
[4]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Gabriella Chinaglia Zanin; Email: gabriella.zanin@aluno.fcmsantacasasp.edu.br.

Background: Mild Cognitive Impairment (MCI) is considered a transitional stage between the expected effects of aging in the patient's cognition, and the pathological changes of Alzheimer's disease. The early detection of this transitional stage has grown in importance recently, as new therapies, mainly monoclonal antibody, have been approved by the FDA for Alzheimer treatment, and have shown better efficacy when introduced in early stages of the disease progression. Furthermore, several modifiable quality-of-life related factors influence the conversion rate. **Objective:** The present study's aim was to compare the conversion rate of MCI to Alzheimer in the last 5 years between a US and a Brazilian cohort of patients originally diagnosed with MCI. **Methods:** An electronic medical record aggregator called TriNetX was used to anonymously obtain the patient data used in the study, using exclusively hospital related records. The US cohort was built using a US-only database via selecting patient above 50 years old, who received the MCI diagnosis in the last 5 years time frame and had never received an MCI or Alzheimer diagnosis before. The Brazilian cohort was built using the same criteria as the US but pulled data from the database of a reference neurology center in São Paulo. The MCI diagnosis was considered the index event, while the outcome was an Alzheimer's disease diagnosis within 5 years of the index event. Risk was obtained by calculating the fraction of patients with the selected outcome from the cohort's total. **Results:** Test results were a US cohort of N=204,396, of whom 18,624 patients had the selected outcome, with a risk of conversion of 9,112% and a mean follow-up of 1,5 years. Mean age of cohort was 76 with standard deviation of 10. The Brazilian cohort had N=260 patients, whom 20 had the selected outcome, with a risk of conversion of 7,692% with a mean follow-up of 1 year. Mean age of cohort was 71 with a standard deviation of 10. **Conclusion:** A lot of the variability in risk of conversion result from demographic factors, mainly age, and health factors, such as cardio-embolic events. While the Brazilian cohort showed a lower risk than its US counterpart, it had both a shorter follow-up and had a cohort mean age lower by 5 years. This possibly indicates a higher MCI to Alzheimer conversion rate in Brazilian cohort, a finding that would be congruent with the overall lower educational attainment compared to the US.

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Abetalipoproteinemia: a case report of a diagnostic challenge

Yasmin Ramalho Mandarino[1], Nicole D'Orazio Schilipacke[1], Éder Rodrigues Pereira[1], Victoria Heloina Machado[1], Raul Rodrigues Vilar[1], Laíse Koenig de Lima[2], Maria Eduarda Angelo de Mendonça Fileti[2], Thábata Emanuelle Martins Nunes[2], Carla Heloísa Cabral Moro[2], Alexandre Luiz Longo[2]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Yasmin Ramalho Mandarino; Email: yasminramalhoo@gmail.com.

Case presentation: A 45-year-old female patient, previously diagnosed with acanthocytosis, consulted a neurologist due to difficulty walking and frequent falls since the age of 15. She also reported decreased visual acuity for 20 years and childhood steatorrhea, which improved with a low-fat diet. As for the familial history, the pacient reported that her brother had similar symptoms without a defined diagnosis and that her parents were consanguineous. On examination, the patient presented with right-sided dysmetria, gait ataxia, global areflexia, grade IV+ muscle strength in the limbs, a positive Romberg sign, impaired proprioception, and vibration sense deficit. Fundus examination revealed optic atrophy and retinitis pigmentosa. Additional tests showed acanthocytosis and extremely low levels of cholesterol and triglycerides, with no other significant abnormalities. Cranial MRI results were within normal limits. Given the family history, the patient's symptoms, and laboratory findings, treatment for abetalipoproteinemia was initiated, including supplementation with vitamins A, D, and E, medium-chain triglycerides, and dietary adjustments. Following the commencement of treatment, the progression of the patient's disease stabilized. **Discussion:** Abetalipoproteinemia is an extremely rare condition, occurring in fewer than 1 in 1 million individuals. For the disease to manifest, an individual must possess two defective alleles, as abetalipoproteinemia is a recessive and hereditary disorder (1). Children presenting with steatorrhea, vomiting, and growth retardation are suspected of having abetalipoproteinemia, which may also be incidentally detected in adulthood through health screenings due to extremely low plasma cholesterol levels (3).The diagnosis of abetalipoproteinemia is established in a proband with absent or extremely low levels of LDL cholesterol, triglycerides, and apolipoprotein B (apo B), alongside biallelic pathogenic variants in the microsomal triglyceride transfer protein gene identified through molecular genetic testing (2). Supplementing high doses of fat-soluble vitamins, such as vitamins E and A, can prevent, delay, or alleviate disease complications and improve prognosis, potentially allowing survival into the eighth decade of life. Without treatment, systemic complications due to fat-soluble vitamin deficiencies may occur within the first decade of life (3). **Final comments:** Abetalipoproteinemia, a rare genetic disease, is a complex disorder resulting in the malformation and inadequate secretion of lipoproteins. While there is no cure, treatment can mitigate complications and improve the quality of life for those affected. Early diagnosis is crucial for prompt intervention. Although access to genetic testing can be limited, diagnosis should not be delayed. It is primarily clinical and may not require genetic testing if the characteristic symptoms are clearly present.

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Access to rehabilitation for people with spinal muscular atrophy undergoing treatment with disease-modifying drugs

Eduardo Vital de Carvalho[1], Graziela Jorge Polido[1], Rodrigo de Holanda Mendonça[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.

Correspondence: Eduardo Vital de Carvalho; Email: duvital1983@gmail.com.

Background: Spinal muscular atrophy (SMA) is a neurodegenerative disease caused by mutations in SMN1 (which encodes the survival motor neuron protein). Reduced expression of SMN leads to loss of motor neurons that manifests clinically with severe muscle weakness. The severity of the disease varies from early onset, with respiratory failure during the first months of life, to a mild type, with onset in adulthood, with slow progression. In the last decade, new drug treatment options have been developed, such as modulating the splicing of the SMN2 and SMN1 gene replacement by gene therapy. The first medications were approved for the treatment of patients with SMA and, if started early, can significantly modify the natural course of the disease. Faced with this new era of medicine, new phenotypes emerged and functional demands were reestablished. Since 2007, multidisciplinary care standards have been established, ensuring increased survival and improved quality of life for patients with SMA. New perspectives for early diagnosis, such as neonatal screening, have been recommended, but in Brazil it is not yet well established, so many patients are diagnosed with some degree of active neuronal degeneration and clinical manifestations of the disease. Rehabilitation is essential and plays an extremely important role associated with available treatments. **Objective:** Describe the access that patients with SMA have to rehabilitation with a multidisciplinary team. **Methods:** Retrospective and descriptive study with analysis of medical records. Demographic characteristics such as age, sex, weight and height were collected from patient records. Clinical characteristics such as type of SMA and medication treatment used. Characteristics about rehabilitation such as access, specialties and frequency. **Results:** 104 medical records of patients with SMA were reviewed, 14 (13.4%) of type 1, 56 (53.8%) of type 2 and 34 (32.6%) of type 3. The average age was 12. 97±9.97 years old, average height of 1.02±0.58 and average weight of 35±23. 71 (68.2%) patients have access to some type of drug treatment, 54% to Spinraza, 0.07% to Risdiplan and 0.05% to combined therapy. 38 (36.5%) of patients do not have access to rehabilitation. Of the patients who have access to rehabilitation, 85% undergo motor physiotherapy, 48% respiratory physiotherapy, 30% aquatic physiotherapy, 33% speech therapy, 0.06% psychology, 19% occupational therapy and 0.06% nutritional monitoring. **Conclusion:** We conclude that some patients who receive some drug treatment do not have access to rehabilitation. There is no standard prescription of a rehabilitation program for patients with SMA undergoing drug treatment. Access to some specialties of the multidisciplinary team is still restricted. The multidisciplinary team is extremely important in the follow-up of patients with SMA and aims to minimize complications resulting from muscle weakness.

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Acetazolamide in the treatment of idiopathic intracranial hypertension: a real-world study

Ana Carolina Montouro Storarri[1], Annelise Akemi Higa Lee[1], Lorrane Campidelli Arthuzo[1], Luis Armando Sawada[1], Beatriz Marques dos Santos[1], Thassia Cavalcante de Oliveira Moraes Marques[1], Bruno Pissolati Mattos Gonzaga[1], Renan Barros Domingues[1]

[1]Santa Casa de Misericórdia de São Paulo. São Paulo SP, Brazil.

Correspondence: Ana Carolina Montouro Storarri; Email: anastorarri@gmail.com.

Background: Acetazolamide is a carbonic anhydrase inhibitor widely used in the treatment of idiopathic intracranial hypertension (IIH), a condition characterized by increased intracranial pressure without detectable apparent cause, often resulting in symptoms such as headache, papilledema, and transient visual loss. The efficacy of acetazolamide in managing IIH has been demonstrated in various clinical studies, notably improving patients' symptoms and playing a crucial role in preventing long-term visual complications. The therapeutic approach with acetazolamide is generally well-tolerated, although side effects such as paresthesia, fatigue, and changes in taste may occur. **Objective:** In the present study we aimed to assess the results with Acetazolamide in a real-life cohort of Brazilian patients. **Methods:** The data of IIH patients treated at Santa Casa since 2019 were evaluated, with information obtained during consultations and from medical records. Sociodemographic and clinical data were collected. The outcomes assessed were headache attributed to HII, pulsatile tinnitus, reduced visual acuity, diplopia, and papilledema before and after treatment with acetazolamide. Comparisons were made using the chi-square test. **Results:** Forty patients were included, all of them were female. The mean age was 33.9±11.5 years. Twenty-two patients were obese, 10 were overweight, and in 8 this information was not obtained. The mean BMI was 33,5±4,3. The mean follow-up time was 55.4±48.4 days. The mean Acetazolamide dose was 972.2±523.6. Six patients reported side effects (paresthesias=6 and dizziness and tinnitus=1) with four of them discontinuing acetazolamide, which was replaced by topiramate. The frequencies of observed evaluated outcomes were: headache (90%x0%, P< 0.0001); low visual acuity (75% x 50%, P=0.02); papilledema (75.7% x 54.5%, P=0.001); diplopia (33.3% x 25%, P=0.006); and tinnitus (27.8% x 0%, P=0.0007).

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Acquired hepatocerebral degeneration accompanied by recurrent episodes of hepatic encephalopathy secondary to chronic hepatitis c virus (HCV) infection, presenting as the initial manifestation of hepatitis C in a young patient

Tainá Fátima Ramos Gonçalves[1], Ingrid Lorena da Silva Gomes[1], Guilherme Lago Miranda[1], Pedro Almeida Martins Pontes[1], Giovanni Caniçali Simões[1], Henrique de Moraes Bernal[1], Sephora Sabrina Candido de Almeida[1], Marcele Schettini de Almeida[1], Herval Ribeiro Soares Neto[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assitência Médica ao Servidor Público Estadual de São Paulo (IAMSPE), São Paulo SP, Brazil.

Correspondence: Tainá Fátima Ramos Gonçalves; Email: taina_goncalves1@hotmail.com.

Case presentation: A 36-year-old previously healthy woman presented with a seven-month history of progressively worsening cognitive and behavioral fluctuations, subsequently developing headaches, mental confusion, psychomotor agitation, and speech and language alterations. Neurological examination revealed fluctuating levels of consciousness, difficulty understanding commands, incoherent speech, dysarthria, and extrapyramidal symptoms. Her family history included a mother who died from liver cirrhosis secondary to hepatitis C infection. An extensive evaluation began with neuroimaging. Brain MRI showed spontaneous hyperintensity on T1-weighted sequences in the basal ganglia, consistent with hypermanganesemia. Cerebrospinal fluid analysis was unremarkable. Serum tests, including serologies for HIV, hepatitis B, and syphilis, were negative. Renal and thyroid functions, rheumatologic markers, and serum manganese levels were normal. Positive serology for hepatitis C (anti-HCV reactive with a viral load of 339,000) and laboratory tests revealed altered liver function markers and hyperammonemia (187 µmol/L, reference range: 11-51 µmol/L). Further investigation with abdominal CT and ultrasound indicated chronic liver disease with portal hypertension. A diagnosis of liver cirrhosis (Child-Pugh A6, MELD: 24), hepatic encephalopathy (EH), and hepatocerebral degeneration (AHD) secondary to hepatitis C virus infection was established. Treatment for hepatitis C with velpatasvir/sofosbuvir and management of cirrhosis symptoms with lactulose and ornithine aspartate were initiated and maintained for 12 weeks. The patient continued follow-up with neurology and gastroenterology after hospital discharge, showing improved consciousness and resuming daily activities. **Discussion:** HCV is a leading cause of cirrhosis in Brazil. Acquired hepatocerebral degeneration is one of the neurological conditions linked to chronic liver dysfunction. While hepatic encephalopathy is more common and reversible, AHD is rare and irreversible. HE results from the livers inability to detoxify neurotoxic substances like ammonia, and evidence suggests that multiple HE episodes increase the risk of AHD. The primary cause of AHD is manganese deposition in the basal ganglia, though serum manganese levels can be normal, and other pathophysiological mechanisms may be involved. **Final comments:** Multiple episodes of hepatic encephalopathy in the context of hepatitis C infection are mostly a consequence of cirrhosis and liver dysfunction, as well as the potential neurotoxic effects of HCV. These events are associated with hepatocerebral degeneration, characterized by irreversible basal ganglia manganese deposition. This chronic condition necessitates symptomatic clinical management and control of complications. Additionally, patients with hepatic encephalopathy require careful treatment to reduce the risk of acquired hepatocerebral degeneration and to mitigate symptoms associated with hyperammonemia.

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Active workspace volume combined with trunk inclination upon standing predicts fshd clinical severity

Eduardo Inacio Nascimento Andrade[1], Lawrence Hayward[1], Ben Ridout[2], Jeff Penka[2], Jeff Stauffer[2], Kirsten Tulchin-Francis[3] , Linda Lowes[3]

[1]University of Massachusetts Chan Medical School, Worcester-MA, United States.
[2]LittleSeed, Inc - Powell, OH, United States.
[3]Nationwide Children's Hospital - Columbus, OH, United States.

Correspondence: Eduardo Inacio Nascimento Andrade; Email: neduardoinacio@gmail.com.

Background: Facioscapulohumeral muscular dystrophy (FSHD) is one of the most common muscular dystrophies, and the validation of longitudinal clinical assessments that reflect disease progression are needed for future clinical trials. ACTIVE Workspace Volume (WSV) and OpenCap provide complementary measures to capture 3D motion of the joints and perform kinematic analyses. WSV uses a game to engage patients in a series of tasks using the arms and trunk while measuring the upper limb accessible space. OpenCap uses two iPhones to capture and analyze movements of the upper and lower extremities in space while the patient performs a task. **Objective:** To explore the ability of WSV and OpenCap to predict clinical severity in patients with FSHD. To develop a model using different parameters obtained from ACTIVE and OpenCap that could be used as reliable biomarkers for FSHD therapeutic trials. **Methods:** As part of an ongoing pilot study, we aim to recruit a total of 10 FSHD and

5 control subjects. Here we present the WSV results of 5 initial subjects with FSHD as summarized by the percentage of predicted reachable workspace for each patient's age and height. In addition, the maximum degree of trunk bending when standing from a chair was measured using OpenCap for each patient. FSHD Clinical Severity Scale (Ricci score) was obtained from each patient by a neuromuscular trained neurologist. A multiple linear regression model was performed using the Ricci score as the dependent variable and the first two measures as covariants. **Results:** The mean degree of lumbar bending when standing was 16.4 degrees (SD=10.3). The mean % of predicted reachable workspace achieved by patients was 116% (SD=68%). The mean Ricci score was 2.3 (SD=1.35). The normality of the variables was confirmed using the "Shapiro-Wilk" test. The multiple linear regression model was statistically significant, ($F2,2 = 10337$, $p<0.001$). The model R^2 was 1.00. The percentage of the predicted workspace achieved by each patient was inversely correlated and statistically significant predictor ($b=-0.1$, $SE=0.01$, $t=-7.28$, $p<0.01$) of the dependent variable. The maximum degree of lumbar bending when standing had a positive correlation and was also a significant predictor ($b=0.13$, $SE= 9.078\times10\text{-}4$, $t=143.78$, $p<0.001$) of the Ricci score. **Conclusion:** As muscle weakness progresses, patients with FSHD experience compromise of upper extremity reachable volume. Patients may also need to lean more forward to stand from a chair. ACTIVE Workspace and OpenCap can capture these changes and when combined they may predict the overall severity of disease in patients with FSHD. A larger patient cohort needs to be tested to confirm the use of this model as a clinical biomarker of disease progression.

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Acute bilateral hippocampal swelling in transient global amnesia triggered by sexual orgasm

Gustavo Henrique Tomasi[1], Paula Gasperin[1], Carolina Heck Haubrich[1], Ricardo Bernardi Soder[1], Lucas Porcello Schilling[1], William Alves Martins[1], André Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Carolina Heck Haubrich; Email: carolhhaubrich@gmail.com.

Case presentation: A 50-year-old woman presented with acute disorientation, disordered speech, and repetitive questioning, alongside retrograde and anterograde amnesia. The retrograde amnesia spanned 15 hours before presentation. Her symptoms began seconds into an orgasm during sexual intercourse. Despite attempting to continue her routine, including going to work, her memory dysfunction worsened, leading to multiple timecard punches within a short span (at 11:31h, 11:48h, 11:56h, 11:59h, 12:06h, 12:40h, and 12:41h). She had a history of controlled hypothyroidism and episodic migraine with aura. Neurological exam was normal except for a significant deficit in newly acquired information. Her symptoms improved over the next 10 hours, leading to a complete recovery. Brain CT scan and general laboratory tests were normal. Eight years earlier, she had experienced a similar event triggered by orgasm, with full recovery in approximately six hours, requiring no further investigation. Due to the recurrence and the Valsalva-related nature of her transient global amnesia (TGA), further work-up included electroencephalogram and CTA, both of which were unremarkable. However, brain MRI displayed bilateral hippocampal swelling without diffusion-weighted-imaging restriction. She was discharged two days later, completely asymptomatic and has had no other episodes since. **Discussion:** TGA is characterized by a sudden inability to retain new information and recall recent events, accompanied by repetitive, anxious questioning. Anterograde and retrograde amnesia can persist for variable durations but typically last a few hours. Underlying pathophysiology of TGA remains a mystery, especially the convergence of diverse triggering phenomena into acute memory dysfunction. Investigations are usually non-revealing. Previous studies show involvement of mesial temporal structures and/or thalami, with focal hyperintensity and restricted diffusion on diffusion-weighted MRI. In our case, acute imaging immediately following a recurrent episode of TGA showed bilateral hippocampal swelling without restricted diffusion, suggesting a non-ischemic mechanism of hippocampal insult. These findings support the hypothesis that TGA may result from acute venous congestion of the mesial temporal lobe, which requires a combination of longstanding insufficiency of the internal jugular vein (IJV) and acute, intense Valsalva maneuvers associated with emotionally or physically charged events. The IJV, which continues from the lateral sinus and drains the mesial temporal lobe, has a single valve proximal to its termination in the subclavian vein. Insufficiency of this valve can lead to retrograde venous congestion. In our patient, the swelling was restricted to the hippocampus and disappeared promptly upon recovery. We propose that the Valsalva maneuver during sexual intercourse and orgasm triggered venous congestion with possible IJV insufficiency, leading to bilateral hippocampal edema and amnesia.

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Acute carotid web thrombosis and early reocclusion after successful thrombectomy: a case report

João Paulo Bello dos Santos[1], Letícia Escorse Requião[1], Vinícius Paulo Lima de Menezes[1], Amanda Sanae Esaki[1], Luis Henrique de Castro Afonso[1], Maria Clara Zanon Zotin[1], Giulia Maria Ximenes Verdi[1], Frederico Fernandes Alessio Alves[1], Millene Rodrigues Camilo[1], Octávio Marques Pontes Neto[1]

[1]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Letícia Escorse Requião; Email: leticiaescorse@gmail.com.

Case presentation: A 58-year-old Black woman presented with sudden onset aphasia, right-sided weakness, and numbness 11 hours prior to admission at the Emergency Unit. On examination, her NIHSS score was 13, indicating a left middle cerebral artery (MCA) stroke. She had a history of smoking but no other cardiovascular diseases. CT revealed loss of gray-white differentiation in the left insular and frontal regions, with an Alberta Stroke Program Early CT Score (ASPECTS) of 8. CT angiogram of the head showed a left MCA occlusion and a small wedge-shaped hypodense filling defect along the posterior margin of the left internal carotid artery (ICA), suggestive of a carotid web (CW). Thrombolytic therapy was not administered. Endovascular mechanical thrombectomy (MT) using combined use of contact aspiration and stent-retriever technique was performed, achieving successful recanalization (TICI score 3) within 124 minutes Door-to-Recanalization time. The NIHSS score after the procedure was 17. Blood tests, transthoracic echocardiography, continuous ECG monitoring and evaluation for hypercoagulability testing and vasculitis was performed to investigate the etiolgy. Subsequent CT at 24 hours showed hypodensity in the left deep nuclei and left frontoparietal areas and left hyperdense MCA sign. Carotid doppler ultrasound (CDUS) revealed a hypoechoic mass, suggestive of acute thrombus formation on the CW. Cerebral angiography performed prior to carotid angioplasty, identified reocclusion of the ipsilateral MCA. Due to the risk of hemorrhagic complications, a second MT was not performed. The patient was treated with aspirin 300 mg and clopidogrel 75 mg, followed by angioplasty with stent placement 10 days post-stroke. She was subsequently transferred to a stroke rehabilitation center, the NIHSS score was 14 and the RANKIN score was 4. **Discussion:** Carotid web is a recognized cause of cryptogenic stroke. Our findings are consistent with previous studies describing a higher prevalence of CW in young Black female patients, often ipsilateral to ischemic strokes of undetermined cause. Symptomatic CW is associated with a high rate of recurrent stroke (17% to 40%), particularly in cases involving morphological and hemodynamic changes such as local flow turbulence or in situ thrombus formation. Acute CW thrombosis and early reocclusion following successful MT are rarely reported. Early neurological deterioration (within 3 days) and hemodynamic disturbances related to CW are important red flags in these cases. Follow-up with CDUS may be useful for monitoring thrombus formation. Carotid stenting immediately after MT could be considered to prevent recurrent strokes given the elevated short-term risk post-MT. **Final comments:** The optimal management to prevent recurrent stroke in patients with carotid web remains uncertain. Observational studies generally favor interventional therapy over medical management, but randomized trials are needed to provide more definitive guidance.

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Acute denervation findings on electromyography in patient with central lesion: case report

Eduardo Barbosa de Albuquerque Maranhão[1], Alexandre Medeiros Sampaio Januario[2]

[1]Faculdade de Ciências Médicas de Jaboatão, Jaboatão dos Guararapes PE, Brazil.

[2]SOS NEURO.

Correspondence: Eduardo Barbosa de Albuquerque Maranhão; Email: maranhao.eduardo@hotmail.com.

Background: A subacute isolated deficit in the upper limb can be challenging to characterize in some cases, particularly regarding central versus peripheral nervous system localization. Here, we present the case of a 70-year-old male patient with prostate cancer under investigation for bone metastasis, who developed a subacute isolated weakness in the right upper limb. The symptom onset occurred during a magnetic resonance imaging (MRI), which was complicated by contrast extravasation and difficulty in peripheral venous access. Subsequent electrodiagnostic evaluation performed 45 days after symptom onset revealed symmetric sensory nerve action potentials in the median and ulnar nerves. Motor nerve action potentials of the median, ulnar, and axillary nerves showed significant reduction only in the axillary nerve innervated muscles of the affected limb. During needle electromyography, fibrillations and positive sharp waves were observed in nearly all muscles of the right upper limb, except for rare occurrences in the deltoid and biceps, and frequent findings in muscles such as the flexor carpi radialis, pronator teres, and extensor digitorum communis. During voluntary muscle contraction, muscles including the dorsal interosseous, flexor carpi radialis, pronator teres, and extensor digitorum communis showed markedly reduced motor unit potentials, while muscles like deltoid, biceps, triceps, brachioradialis, and flexor pollicis longus exhibited relatively normal motor unit potentials. Subsequent contrast-enhanced cranial MRI revealed an expansive metastatic lesion involving the lateral aspect of the left precentral gyrus. **Discussion:** The presence of acute denervation findings (such as fibrillations and positive sharp waves) on electromyography in patients with central lesions is a well-documented phenomenon in the literature. One of the prevailing theories for these findings implicates disruption in transsynaptic transmission between the first and second motor neurons, leading to anterior horn degeneration. Typically, abnormal spontaneous activity begins approximately one month after injury onset, becomes less frequent by six months, and is rare after one year, contrasting with upper motor neuron signs. Regarding the predominantly affected muscle groups, as observed in this case, distal muscles in the affected limb often exhibit a higher proportion of acute denervation

compared to proximal muscles. **Conclusion:** Therefore, vigilance is warranted when electromyography reveals signs of denervation in patients presenting with subacute flaccid monoparesis. As exemplified in this case, it still may indicate ongoing pathology involving the first motor neuron, the critical importance of meticulous evaluation for guiding accurate diagnosis and clinical management.

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Acute disseminated encephalomyelitis (ADEM) associated with dengue infection: a case report

Beatriz Pires Paes[1], Bruna da Silva Lima[1], Poliana Lima Campos[1], Beatriz Ballarin Costa[1], Anthony Magalhães Morais Santiago[1], Camilo Carvalho[1], Larissa Lucas Schloicka[1], Andre Gustavo Fonseca Ferreira[1]

[1]Hospital de Base do Distrito Federal, Brasília DF, Brazil.

Correspondence: Beatriz Pires Paes; Email: beatrizpirespaes@gmail.com.

Case presentation: A 48-year-old man was diagnosed with dengue fever confirmed by a positive serology, and after fifteen days, developed paraparesis and a tingling sensation in both legs associated with urinary retention. Ten days after the onset of the condition, he was transferred to the neurological department. At admission, there was asymmetric sensorimotor paraparesis, worse on the left, with sensory level at T10, with tactile, thermal, and palesthetic hypoesthesia. Cerebrospinal fluid analysis showed normal protein levels and a slight increase in opening pressure (27 cmH2O); polymerase chain reaction did not detect dengue virus, and the research for oligoclonal bands was negative. Neuroimaging (MRI) revealed supratentorial and infratentorial intraparenchymal lesions of probable inflammatory/demyelinating etiology, T2 hyperintensity, and gadolinium enhancement in a lesion in the cervical cord (C4-C5), in addition to multiple small peripheral oval lesions, also with T2 hyperintensity and gadolinium impregnation, from T1 to T9. He underwent pulse therapy with methylprednisolone for five days, and his condition improved significantly. Considering the extent of his lesions, we prescribed a four-week-long course of oral corticosteroids afterward. **Discussion:** Acute disseminated encephalomyelitis (ADEM) is a rare immune-mediated central nervous system disorder that determines a rapidly demyelinating process. It can occur during the acute or post-infectious phase of dengue, usually affects young children and has an identifiable trigger. Characterized by acute-onset polyfocal neurological deficits, its symptoms generally appear within 2-21 days after an infection. Encephalopathy is an obligatory feature, and it includes behavioral changes and/or alterations in consciousness, including irritability, not explained by systemic febrile illness or posticta symptoms. Our patient presented with significant irritability. There are no consensus diagnosis criteria for adults but it is usually based on the clinical manifestations described above, along with evidence of demyelination on MRI, which shows hyperintense abnormalities in T2-weighted/FLAIR, with typically bilateral, asymmetrical, large (> 2 cm) and poorly demarcated lesions. Gadolinium enhancement is reported in up to 30% of cases. Clinical manifestations may vary widely but usually include motor and sensory deficits, pyramidal signs, ataxia, brainstem symptoms, and transverse myelitis. CSF pleocytosis and elevated protein levels are typically seen, yet oligoclonal bands are found in less than 10%. Treatment usually consists of IV methylprednisolone for 3-5 days, followed by oral prednisone for 4-6 weeks. **Final comments:** In conclusion, we presented a rare case of ADEM associated with dengue fever in an older male. The diagnosis was based on the clinical manifestations, imaging findings, and laboratory tests. ADEM is a challenging diagnosis, and early treatment is crucial to a better outcome.

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Acute disseminated encephalomyelitis (adem) following recent rabies vaccine

Isabella Nascimento Guedes[1], Matheus Pinto Munoz[1], Lilian Eduarda Bastos da Silva[1], Karoline da Silva Guimarães[1], Gilvan Pires de Castro Junior[1], William Corá[1], Darah Fontes da Silva Assunção, Natasha Pryanka de Araújo Bessa, Roger Santana de Araújo[2], Marina Buldrini Filogonio Seraidarian Seraidarian[2]

[1]Universidade Estácio de Sá, Rio de Janeiro RJ, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Isabella Nascimento Guedes; Email: isabellaguedes21@icloud.com.

Case presentation: Acute disseminated encephalomyelitis (ADEM) following recent rabies vaccine. Clinical case: A 23-years-old female veterinary student was admitted with a history of 3 days weakness, dyspnea, diarrhea and vomiting, and more recently speech impairment and decreased level of consciousness. Emergency department CT showed important cerebellum edema and a decompressive surgery was performed to reduce intracranial pressure. Further head MRI demonstrated abnormalities in cortical and subcortical regions and tumefactive demyelinating lesions diffusely affecting cerebellar and cerebral hemispheres. Patient medical history reveled thar she was submitted to a prophylactic immunization process and the second dose of antirabic vaccine was taken a few days before the illness. As acute disseminated encephalomyelitis (ADEM) can be triggered by preceding infection or immunization, the patient was managed with corticosteroids and immunoglobulin therapy, which led to overall improvement in both radiological and clinical findings. **Discussion:** Rabies is a viral disease transmitted by infected mammals with high neurotropism. Although there is no specific treatment, rabies pre-exposure vaccination is recommended for veterinary staff. The vaccine has inactivated virus and the immunization schedule consists of 3 doses given on days 0, 7 and 28. The hallmarks of rabies encephalitis are primary cortex and basal ganglia lesions, while this case reports affections in white matter and periventricular areas that could be explained by an immune-mediated process. Neurologic impairment is a serious post-vaccination adverse event and its incidence is 1 in 8000 people. The main neurological conditions related to the immunization are: encephalomyelitis, transverse myelitis, cranial nerve neuropathy, peripheral mononeuropathy and Guillain-Barré syndrome. **Final comments:** This case reports acute and severe neurological impairment, probably related to an autoinflammatory response triggered by rabies vaccine. Differential diagnosis could be other autoimmune encephalitis, infectious meningoencephalitis, neurovascular 2 / 2 syndromes or neoplastic lesions. However, taking into account clinical findings, radiological images and patient's evolution those hypotheses were considered less probable.

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Acute disseminated encephalomyelitis as a concomitant presentation of anti-mog, covid and hiv: case report

Juliana Magalhães Leite[1], Thaís Magalhães Lima Leite[2], Lucas Silva Barbosa Bezerra[2], Lívia Gonçalves Santos Costa[2], Ana Lívia de Andrade Rangel[2], Matheus Saraiva Gurgel[2], Rafael de Souza Andrade[1], Assis Martins Maia[1]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[2]Centro Universitário UNIPE, João Pessoa PB, Brazil.

Correspondence: Thaís Magalhães Lima Leite; Email: thaismlleite@gmail.com.

Case presentation: A 21-year-old male patient, a student from Itabaiana-PB, presented with thoracic/lumbar dorsalgia that began 5 days before admission. His condition progressively worsened to paraparesis, and subsequently paraplegia with urinary and fecal retention, with a sensory level at T10, along with drowsiness. He denied any trauma, infectious symptoms, or exposure to river or lake water. Cranial MRI revealed subtle ovoid lesions affecting the deep periventricular white matter of the cerebral hemispheres at the callosal-septal interface of the frontoparietal regions. Cervical spine MRI showed extensive signal alteration affecting the cervical and thoracic spinal cord, associated with mild medullary expansion. Cerebrospinal fluid (CSF) culture was negative, with 14.3 leukocytes (10% polymorphonuclear, 90% mononuclear), glucose 82, protein 68.8, and a negative viral panel. HIV and COVID tests were not performed in the CSF due to lack of availability. Anti-aquaporin-4 antibody was negative, Herpes IgG/IgM was reactive. HIV was reactive with CD4 298 / CD8 692 / VL 102,000 copies/ml, COVID positive. He received 1g of methylprednisolone for 7 days and immunoglobulin for 5 days. The patient showed improved upper limb strength, sensory level improvement, remaining hypoesthesia in the thigh root, and significant radiological improvement. He was discharged on oral corticosteroids and regular antiretroviral therapy (ART). One month after discharge, anti-MOG results were positive. A new cycle of immunoglobulin was administered, and anti-MOG was repeated 3 months post-discharge, remaining positive. **Discussion:** When anti-MOG antibody disease involves the brain, the phenotype resembles acute disseminated encephalomyelitis (ADEM), with mental status changes and good outcomes following treatment with corticosteroids and intravenous immunoglobulins. Unlike ADEM, which is classically monophasic, MOG antibody seropositivity indicates a high probability of relapse. Transverse myelitis is a known manifestation of primary HIV infection. Speculated factors in the formation and infiltration of post-infectious MOG antibodies into the CNS include molecular mimicry and the breakdown of the blood-brain barrier due to prior viral inflammation, although the precise mechanisms of post-infectious demyelinating disease remain to be elucidated. **Final comments:** The presentation of anti-MOG-associated encephalomyelitis in a patient with asymptomatic COVID co-infection, along with a recent HIV diagnosis, sparked significant discussion regarding the possibility of a false-positive anti-MOG result and the appropriate long-term therapeutic approach. Given the severity of the clinical condition, cycles of immunoglobulin were chosen, and anti-MOG antibody levels are being monitored. This case report is highly relevant for academic discussion due to its uniqueness and therapeutic evolution.

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Acute disseminated encephalomyelitis secondary to tuberculosis in a pediatric patient: a case report

Júlio César Soares Barros[1], Antonia Cardoso Silva[1], Guilherme Carvalho de Souza[1], Larissa da Silva Almeida[1], Adriano Cardozo Silva[2], Glicia Florêncio Medeiros[3], João Eduardo Gondim Mendes Leite[3], Bárbara Letícia Figueiredo Fonseca[3]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.
[2]UniFTC, Feira de Santana BA, Brazil.
[3]Hospital Geral do Estado Professor Osvaldo Brandão Vilela, Maceió AL, Brazil.

Correspondence: Júlio César Soares Barros; Email: julio.barros@famed.ufal.br.

Case presentation: A female patient, 11 years old, with a history of dysuria and a diagnosis of hydronephrosis for 2 months, was under follow-up with a pediatric nephrologist. The mother initiated treatment with sulfamethoxazole + trimethoprim on her own, but the patient developed drowsiness associated with paresis in the right

lower limb. She was admitted to the hospital with paresis in the left lower limb and transferred to another service. Her condition worsened, with paresis in the upper limbs and desaturation, leading to transfer to the pediatric ICU. MRI revealed transverse myelitis at various levels, and brain MRI showed acute disseminated encephalomyelitis (ADEM). Infectious etiology investigation revealed a positive rapid molecular test for tuberculosis (TB) in the cerebrospinal fluid, confirming the diagnosis of ADEM caused by TB. The patient underwent pulse therapy with methylprednisolone (later transitioned to prednisolone), followed by human immunoglobulin (4 days), and started TB treatment with rifampicin, isoniazid, pyrazinamide, and ethambutol (RHZE). Azathioprine was also initiated later. The patient received intensive follow-up with motor physiotherapy and occupational therapy, showing satisfactory progress. She improved neurologically and had a favorable clinical evolution, being discharged with general care instructions and outpatient and multidisciplinary follow-up. **Discussion:** ADEM is an inflammatory demyelinating disease of the central nervous system that usually occurs after infections or, less commonly, after vaccination. It is most commonly associated with viral infections, but can also occur following bacterial infections, although these are rare. This case is notable for the association of ADEM with TB, a bacterial infection caused by Mycobacterium tuberculosis. Clinically, ADEM typically presents with an acute onset of multifocal neurological symptoms, including encephalopathy, ataxia, and motor or sensory deficits. In more severe cases, there may be impaired consciousness. In this case, the patient presented with progressive neurological symptoms requiring intensive care. The diagnosis of ADEM was confirmed by MRI, and the infectious etiology was confirmed with a rapid test in the cerebrospinal fluid. The management of ADEM includes immunosuppression with corticosteroids and, in some cases, intravenous immunoglobulin, as performed in this patient. Specific treatment for tuberculosis with the RHZE regimen is crucial to control the underlying infection. The inclusion of azathioprine aimed to prevent relapses and promote long-term remission. **Final comments:** ADEM is a rare condition, especially when associated with TB infection. This highlights the importance of differential diagnosis in patients with acute and progressive neurological symptoms. Rapid and accurate identification of the etiology is crucial for appropriate management and a positive prognosis, as demonstrated by the patients progressive recovery.

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Acute hyponatremia secondary to polydipsia provoking seizure after physical exercise: a case report

Mário Vicente Guimarães[1], Josué Andrade Martins[2], Lawani Rigopoulos[3], Guilherme Requião Radel Neto[2], Julia Souza e Costa[2], Raquel Maria Ayres Monteiro[4], Ana Beatriz Lima Pedroza[2], Tatiana Quintanilha Soares da Silva[2], Bruany Antoniolli Bianchi[2], Natália Moreno Coelho[2]

[1]Universidade de São Paulo, São Paulo SP, Brazil.
[2]Universidade Anhembi Morumbi, São Paulo SP, Brazil.
[3]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.
[4]Faculdade Israelita de Ciências da Saúde Albert Einstein, São Paulo SP, Brazil.

Correspondence: Raquel Maria Ayres Monteiro; Email: raquelmamonteiro@gmail.com.

Case presentation: A 50-year-old man presented at the emergency center with malaise, sweating, abdominal and thoracic pain, followed by a generalized tonic-clonic seizure. He had played an exhaustive tennis game and consumed 4 liters of water. His medical history includes recent gastroplasty and depression, currently treated with quetiapine, trazodone, and melatonin. Post-seizure, he was disoriented with a Glasgow Coma Scale score of 9 and bilateral mydriasis, prompting orotracheal intubation. At the emergency center, he experienced another seizure, treated with intravenous phenytoin, and showed signs of sinus bradycardia, managed with atropine. Imaging tests, including CT and MRI, revealed no abnormalities. His sodium level was critically low at 117 mEq/L, and D-Dimer was elevated at 2400 ng/mL. Upon hospital admission, he was hypertensive and agitated despite sedation. AngioCT found calcified plaques in several arteries but no stenosis. AngioMR and transcranial Doppler results were unremarkable, and EEG indicated disorganized brain activity. The echocardiogram showed no abnormalities. The primary diagnosis is a seizure resulting from hyponatremia due to excessive water intake. **Discussion:** Acute hyponatremia occurs when symptoms develop within 48 hours, often leading to severe cerebral edema and poor prognosis, while chronic hyponatremia develops over a longer period. This patients condition likely resulted from dilutional hyponatremia due to excessive low-sodium fluid intake and increased ADH during intense exercise, leading to symptoms such as cellular edema and increased intracranial pressure. The case underscores the importance of carefully correcting acute hyponatremia to avoid osmotic demyelination syndrome, with a recommended increase in serum sodium of no more than 8 to 10 mEq/L per day. Additionally, differential diagnoses like hyperthermia or hypoglycemia should be considered in athletes presenting with seizures after strenuous activities. **Final comments:** This case highlights the importance of promptly recognizing and understanding the variations of exercise-associated hyponatremia (EAH) to manage it appropriately based on its specific cause. Hyponatremia should always be considered in patients presenting with seizures following intense activity and excessive fluid intake.

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Acute intermittent porphyria in siblings: a case report

Lucas Gonçalves Coelho[1], Victoria Nunes Alvares[1], Clara Ribeiro Rizzo[1], Isabella Navarro Diaz Horta[1], Maria Eduarda Braz Ferreira Leão Caldas dos Santos[1], Bernardo Leal Cupello[1], Isadora Gomes Mesquita[2], Patricia Gomes Pinheiro[2], Clarissa de Araujo Davico[2], Karina Lebeis Pires[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gafrée e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: Lucas Gonçalves Coelho; Email: lucasgcoelho@edu.unirio.br.

Case presentation: Two sisters, 49 and 54 years old, are followed up at the neurology outpatient clinic due to the diagnosis of Acute Intermittent Porphyria since 2001, detected by family screening. The younger sister reports periods of abdominal pain associated with menstruation, which awaken her and radiate to the left lower limb. In addition, the patient refers to anxiety, chronic fatigue, frequent cramps and a "strange sensation" in her hands and feet, which is relieved by biting her nails or wearing socks. Genetic molecular testing was performed and confirmed a mutation of uncertain significance in the HMBS gene (c.1004G> T). The older sister has a similar clinical presentation, characterized by intermittent abdominal cramps since 2014, associated with sporadic numbness in the fingers and depression. On physical examination, distal hypopalesthesia is observed in the four limbs. Her genetic molecular test also demonstrated a mutation of uncertain significance in the HMBS gene (c.100G> T). Both patients reported improvement in pain over the use of 5% glucose solution, increased intake of carbohydrates in the diet or use of ibuprofen. **Discussion:** Acute Intermittent Porphyria (AIP) is a rare disease caused by a deficiency of the enzyme Porfobilinogen deaminase, leading to accumulation of neurotoxic precursors of the heme group (porphyrins). The clinical manifestations of AIP are diverse and nonspecific, including abdominal, neuropsychiatric and autonomic symptoms. In the cases presented, additionally to the similarity of the symptoms presented by the sisters, there is a family history of the condition, emphasizing the importance of genetic screening for the early diagnosis of the disease, ensuring fulfilled monitoring and treatment, especially in courses of crisis which come from the disease. **Final comments:** The incorporation of Acute Intermittent Porphyria in the differential acute neurovisceral diagnosis disease is crucial, especially in patients with family history. Besides, it is necessary to emphasize the importance of genetic counseling in the family of those patients, aiding early detection and treatment. Therefore, implementing strategies that prevent acute thus is important to improve the quality of life of patients with AIP.

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Acute intermittent porphyria mimicking guillain-barré syndrome: a case report

Iasmim Portela Maifrede[1], José Víctor Filho de Oliveira[1], Luiz Felipe Mota Freitas[1], Stephania Neves Scapim[1], Leopoldo Antonio Pires[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário (EBSERH), Juiz de Fora MG, Brazil.

Correspondence: Iasmim Portela Maifrede; Email: iasmimportela@hotmail.com.

Case presentation: Woman, 18-year-old, previously healthy, presented with a history of mild to moderate recurrent colic abdominal pain associated with pollakiuria, diarrhea and depressed humor since november 2023. After a month, the pain became more intense and was associated with constipation, palpitations, diaphoresis, and dark urine. The symptoms worsened after episodes of alcohol intake, prompting visits to the emergency room. In early March 2024, she was hospitalized in her hometown due to intestinal subocclusion, followed 4 days later by dysautonomia, proximal predominant crural paraparesis, episodes of intermittent mental confusion and urinary incontinence. Laboratory tests revealed electrolyte disturbances (hypocalcemia, hypomagnesemia, and hypokalemia) associated with leukocytosis. The condition rapidly progressed to tetraparesis and peripheral facial diparesis with dysphagia and hypophonia, requiring orotracheal intubation. The diagnosis of Guillain-Barré syndrome was considered, despite normal cerebrospinal fluid findings, and immunoglobulin was initiated without improvement. Due to the presence of abdominal pain associated with acute flaccid paralysis with dysautonomia, the possibility of polyneuropathy secondary to acute intermittent porphyria (AIP) was raised and confirmed by 24-hour urine delta-aminolevulinic acid (ALA) and quantitative porphobilinogen levels, with results exceeding 12.0 mg/L and 3.1 mg/24h, respectively. Intravenous glucose support was initiated, and the patient progressed with partial improvement of symptoms, but with persistent bulbar muscle weakness and symmetrical distal paresis, currently showing progressive gains with rehabilitation. **Discussion:** AIP is a rare metabolic disorder with an estimated prevalence of 0.5 to 10 per 100,000 individuals worldwide. Up to 23% of carriers have polymorphic neurological manifestations. This case report describes an acute presentation of abdominal pain with neurological manifestations, such as proximal-predominant paresis, associated with dysesthesias, areflexia, autonomic manifestations, and facial paralysis. The dark urine color present in the picture is one of the most characteristic aspects of AIP. Among the laboratory alterations, hyponatremia, hypocalcemia and leukocytosis are possible findings in patients with AIP. The neurological manifestations of AIP attacks can assume a character similar to Guillain-Barré Syndrome,

however, central nervous system manifestations (such as in the case, confusional syndrome), in addition to recurrent abdominal pain and association of symptom onset with medication and/or alcohol intake, can help differentiate the two pathologies. **Final comments:** AIP is a rare disease that is difficult to diagnose, but it should be considered in patients with diverse neurological manifestations, especially those with recurrent abdominal pain, as an alternative diagnosis.

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Acute intermittent porphyria presenting with encephalopathy and osmotic demyelination syndrome

Rafael Bragança Rodrigues Matias[1], Eduardo Mesquita[1], Ana Luisa Vieira de Faria Donadon[1], Amanda Miranda de Mendonça[1], Alex Araujo Fernandes[1], Matheus Luís Braga Munareto[1], Alex Machado Baeta[1], Hennan Salzedas Teixeira[1], Karlla Danielle Ferreira Lima[1]

[1]Beneficência Portuguesa de São Paulo, São Paulo SP, Brazil.

Correspondence: Rafael Braganca Rodrigues Matias; Email: rafaelwaa@gmail.com.

Case presentation: A 28-year-old female, previously healthy with an unclear early paternal death, presented with abdominal and right flank pain, constipation, and was treated for urinary infection. She was hospitalized for 11 days, during which hyponatremia and hypokalemia were observed, and discharged with a serum sodium level of 110. She returned with seizures and rhabdomyolysis, sodium at 104, and underwent a rapid correction of 30 mEq in 24 hours. Admitted to our service with suspected encephalopathy and new abdominal pain, examination revealed diffuse hypotonia and areflexia, with proximal strength graded III. Acute intermittent porphyria was suspected; hematin was initiated. MRI showed central pontine myelinolysis signs, hyperintensity in bilateral striatum, cerebral cortex, cerebellum, dorsolateral thalamus aspects, especially in the pons. Laboratory tests indicated porphobilinogen in urine, isolated ALA of 6.2, ALA/creatinine ratio of 32.5 mg (limit 4.5). Genetic studies found no abnormalities in mother and sisters. Gastrostomy and tracheostomy were performed; the patient improved with hematin but remains bedridden. Early identification of porphyria is vital for treatment and outcome; initial misidentification and erroneous sodium correction led to encephalopathy, complicating the diagnosis with overlapping osmotic demyelination syndrome during an acute porphyria attack. **Discussion:** Porphyrias are a group of rare metabolic disorders affecting the heme biosynthetic pathway. Depending on the defective enzyme, these disorders can manifest with neurovisceral and/or cutaneous symptoms. Neurovisceral porphyrias are characterized by acute attacks triggered by excessive heme production. During an acute attack, patients typically experience severe abdominal pain, vomiting, and tachycardia. Other possible symptoms include hypertension, hyponatremia, peripheral neuropathy, and mild mental symptoms. In severe cases, symptoms may escalate to seizures and psychosis. If left untreated, the attack can become very severe, affecting the peripheral, central, and autonomic nervous systems, potentially leading to paralysis, respiratory failure, coma, and even death. **Final comments:** In this case the failure to identify classic signs of porphyria led to misdiagnosis and a delay in the correct treatment. Additionally, the mechanism of hyponatremia was not considered, leading to an incorrect correction and complications such as osmotic demyelination syndrome. This case report highlights the importance of disseminating information about the classic clinical presentation and the factors clinicians should consider when including porphyria in their diagnostic hypotheses. Early treatment has the potential to interrupt the damage process.

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Acute intermittent porphyria: identification of novel variant in the HMBS gene

Camila Rodrigues Nepomuceno[1], Nayara de Paula Guerreiro[1], Maria Fernanda Hamamoto Leati[1], Andre Vinicio Cruz Mesias[1], Gabriella Corrêa Dousseau[1], Daniel Thomas Pereira Lopes[1], Cristiane de Araújo Martins Moreno[1], Arthur de Medeiros Dias[2], Ana Claudia Piccolo[2], Maria Sheila Guimarães Rocha[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil. [2]Hospital Dr Alipio Corrêa Neto, São Paulo SP, Brazil.

Correspondence: Camila Rodrigues Nepomuceno; Email: camilarnepomuceno@gmail.com.

Case presentation: A 19-year-old woman has made multiple visits to the emergency room this year due to recurring abdominal pain. In April 2024, she was admitted to another hospital due to ascending areflex flaccid paralysis, where she was initially diagnosed and treated for Guillain-Barré syndrome (GBS). However, cerebrospinal fluid showed no cytoprotein dissociation. Her upper limbs were more severely affected than her lower limbs, and she also experienced seizures, confusion, and reduced level of consciousness. Electrolyte disturbances, particularly hyponatremia, were consistently observed during hospitalization. Given this context, porphyria was suspected, and a 24-hour urine analysis was conducted, revealing: Uroporphyrins 3078 mcg, Heptaporphyrins 183 mcg, Hexaporphyrins 48 mcg, Pentaporphyrins 98 mcg, Coproporphyrins I 153 mcg, and Coproporphyrins III 261 mcgall exceeding the laboratory reference values. She was then transferred to our service and qualitative analysis of ALA and PBG in urine was also positive. Subsequently, genetic sequencing revealed the variant c.751del: p. (Arg251Glyfs*4) in heterozygosity in exon 11 of the HMBS gene. This variant was classified as probably pathogenic (ACMG evidence: PM2, PVS1) and has not been previously described; it is absent in ClinVar and Gnomad databases. The patient received targeted treatment with hematin and glucose, which effectively controlled the crisis. **Discussion:** Acute intermittent porphyria (AIP), the most prevalent and severe form of hepatic porphyrias, is an autosomal dominant disorder caused by mutations in the HMBS gene. These mutations lead to a deficiency in hydroxymethylbilane synthase (HMBS), the third enzyme in heme biosynthesis. Common disease-causing mechanisms associated with the HMBS gene include missense, frameshift, splice site, and nonsense mutations. Symptoms of AIP include severe diffuse abdominal pain, headache, muscle weakness (often presenting as areflex flaccid paralysis, which can clinically mimic GBS), autonomic neuropathy, neuropsychiatric symptoms, hyponatremia, myoglobinuria, seizures, and coma. The penetrance rate of AIP is low, and the condition exhibits high clinical heterogeneity. Some mutations may influence the penetrance or severity of clinical manifestations, and several modifying genes have been identified over the years, including the PEPT2 gene, PPARA gene, cytochrome P450 gene, and ABCB6 gene. As mentioned above, we identified a novel frameshift variant in the HMBS gene. This variant has not been previously described and is absent from both the ClinVar and Gnomad databases. **Final comments:** Early diagnosis of AIP is crucial. This can be achieved through quantitative and qualitative porphyrin tests. Prompt diagnosis can prevent the progression to severe acute attacks, as well as chronic manifestations. Advances in genetic sequencing have led to the identification of numerous variants, aiding in genetic counseling and crisis prevention.

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Acute myelorradiculitis after dengue infection

Bárbara Luisa Coraspe Gonçalves[1], Luiz Guilherme Gerhardt Fernandes[1], Maria Luiza Levindo Coelho Martinis[1], Ana Carolina Andorinho de Freitas Ferreira[1], Bárbara França Amorim[1], Cristiane Borges Patroclo[1], Marina Brasileiro Cesar Leitao[1], Marcelo Cagy[1], Osvaldo Pantoja de Oliveira[1]

[1]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.

Correspondence: Barbara Luísa Coraspe Gonçalves; Email: barbaracoraspeg@gmail.com.

Case presentation: A 52-year-old man, living in Rio de Janeiro, presented with fever, headache, myalgia, retro orbital pain, nausea, vomiting and fatigue. He sought emergency care and received symptomatic medication. After 5 days, he evolved with severe sensory ataxia, strap-like pain in the flanks level with irradiation to lower limbs. There was mild proximal paraparesis with a sensory loss level at T10, in addition to urinary and fecal retention, leading to hospitalization. Blood analysis disclosed hyponatremia (118mmol/L), positive NS1 and leukopenia (3,310/mm^3), negative C-reactive protein. CSF analysis pointed to 6 leukocytes/mm^3 (100% mononuclear), glucose 62mg/dL and protein 240 mg/dL, no growth of bacteria or fungi. Electromyography and MR were unremarkable. The neurological examination was compatible with myeloradiculitis, which was regarded as an acute arboviral complication. The patient received treatment with methylprednisolone for 5 days, physiotherapy and motor rehabilitation. He was discharged after partial clinical improvement. After 2 months, patient had recovered completely from ataxic gait, however, it remained light neuropathic pain in a waistcoat pattern. **Discussion:** In the first half of 2024, Brazil surpassed the mark of 2 million dengue (DENV) cases. According to data from the Ministry of Health, the country registered 2,010,896 cases in the first 11 weeks of this year, and more than 500 deaths. Symptoms of DENV fever include headache, retro-ocular pain, severe myalgia, arthralgia, rash, hemorrhagic manifestations and leukopenia. Although neurological manifestations might be rare, they reach up to 21% in Brazil. They encompass examples of direct invasion, immunemediated response and metabolic disturbance with neurological repercussion, such as: encephalopathy in parallel to cerebral edema and secondary to hyponatremia, fulminant hepatic failure or hypoxia; Guillain-Barré syndrome; encephalitis and myelitis. Myelitis is a rare neurological complication. It involves mostly males and presents usually with lower limbs sensorimotor impairment and sphincter disturbances within 2 weeks of dengue onset. The outcome is favorable in general, and the recovery may be full. It is caused by autoimmune reaction or direct invasion. The direct infection of the nervous system may stem from sequential infection, notably considering the hyperendemicity (the cocirculation of multiple dengue serotypes) and the neurotropic properties. The DENV2 and DENV3 viruses have been proved to cross the blood-brain barrier and invade the CNS. Our case fits in the most prevalent profile and seemingly derives from direct invasion. However, it was associated to normal MRI. This condition has been punctually reported, especially in MOGrelated disorders. **Final comments:** This report depicts a case of acute myeloradiculitis sparked during DENV infection presenting with normal MRI. The outcome resulted in full recovery of gait and residual thoracic radicular pain.

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Acute spinal cord compression due to extramedullary hematopoiesis

João Victor Cabral Correia Férrer[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Ruan Gambardella Rosalina de Azevedo[1], Victor Cardoso de Faria[1], Rafael Tuzino Leite Neves Maffei[1], Adriel Rego Barbosa[1], Andre Felipe Candêas Amorim[1], Marianna Pinheiro Moraes de Moraes[1], José Luiz Pedroso[1], Orlando Graziani Povoas Barsottini[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Eduarda Slhessarenko Fraife Barreto; Email: madusfbarreto@gmail.com.

Case presentation: A 26-year-old man with beta-thalassemia major presented with paresthesia in lower limbs, genital numbness, urinary retention, constipation and erectile dysfunction, without low back pain, one day after falling from his own height. Physical examination disclosed tactile hypoesthesia on the posterior territory of the thigh and leg on the left and absent left Achilles reflex. Spinal cord magnetic resonance imaging (MRI) revealed T2 hyperintense and T1 heterogeneous contrast- enhancing soft tissue mass in the presacral region and in the lumbosacral epidural space, compressing the spinal cord and its roots. Electroneuromyography showed signs of acute denervation in myotomes S1-S4 on the left. He underwent decompressive laminectomy L5-S2 and radiotherapy sessions. Biopsy confirmed extramedullary hematopoiesis (EMH). **Discussion:** This condition occurs as a compensatory and physiological reaction in hemoglobinopathies and myeloproliferative disorders, such as thalassemia, sickle cell anemia, polycythemia vera and myelofibrosis. The most common sites are the liver, spleen and lymph nodes. EMH-induced spinal cord compression is rare and usually presents a subacute progression. Acute course is uncommon, and trauma can be a trigger. Low signal intensity on both T1- and T2-weighted MR images is characteristic of iron deposition and strongly suggests EMH. Treatment options are variable including blood transfusions, cytostatic agents, radiotherapy and decompressive surgery, this last option allowing an immediate decompression and an accurate histological diagnosis. **Final comments:** It is important to know about uncommon neurological complications of systemic diseases. The early diagnosis and correct treatment are essential to reduce clinical sequelae and to improve quality of life.

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Acute vestibular syndrome in the emergency department: be alert!

Letícia Tozzini Tavares da Silva[1], Patryck Garcia do Prado[2], Giovanna Reis Coelho[1], César Minoru Toita Koga[3], Marcos Christiano Lange[3], Valéria Cristina Scavasine[3], Caio César Diniz Disserol[3,4,5]

[1]Universidade Positiva, Curitiba PR, Brazil.
[2]Fundação Escola do Ministério Público do Estado do Paraná, Curitiba PR, Brazil.
[3]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[4]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[5] Hospital Israelita Albert Einstein, Curitiba PR, Brazil.

Correspondence: César Minoru Toita Koga; Email: cesarmtkoga@gmail.com.

Case presentation: A 76-year-old male with hypertension, dyslipidemia, and atrial fibrillation on edoxaban presented to the emergency department with sudden, complete right-sided sensorineural hearing loss, followed by vertigo, imbalance, nausea, and vomiting. He was treated symptomatically and discharged with oral prednisone. While vestibular symptoms improved, hearing loss remained. A neurology clinic evaluation after a month revealed an altered right head impulse test and ipsilateral anacusis, with normal strength, reflexes, coordination and sensory examination. Brain magnetic resonance imaging (MRI) taken 2 weeks after symptom onset revealed right labyrinth hemorrhage. **Discussion:** Labyrinthine hemorrhage (LH) is a rare condition involving bleeding within the intralabyrinthine compartment, presenting as sudden sensorineural hearing loss (SNHL) and any other dysfunction of the compartments of the labyrinth, such as vertigo, nausea, vomiting, nystagmus, and imbalance. While most LH cases are idiopathic, potential causes include hematological malignancies, autoimmune diseases, cerebral venous thrombosis, and anticoagulant therapy. However, LH associated with direct oral anticoagulants has not been reported. It is often characterized by profound hearing loss and resemble acute vestibular neuritis or labyrinthine ischemia, making the diagnosis challenging. The HINTS battery differentiates acute vestibular neuritis from stroke but may not adequately distinguish from LH or labyrinthine infarction. Furthermore, the HINTS plus battery, which includes acute hearing loss, could be used to differentiate LH from acute vestibular neuritis, but is limited in distinguishing LH from anterior inferior cerebellar artery (AICA) ischemic infarction, posing a risk of thrombolysis in patients with ongoing labyrinthine bleeding. Computed tomography (CT) alone lacks sensitivity in identifying intralabyrinthine disorders. Only brain MRI can detect labyrinthine hemorrhage in the acute phase, revealing hyperintensities in the labyrinth on T1 and FLAIR sequences. Treatment data for LH are limited. Common approaches include oral or systemic corticosteroids and intratympanic corticosteroid therapy. Discontinuing anticoagulants is recommended, but the risk-benefit balance is debated. In cases associated with anticoagulants, resuming therapy after 30 days and switching to a lower-risk agent like apixaban can be considered. Prognosis for hearing impairment in LH is generally poor, with most patients experiencing permanent hearing loss, while vestibular symptoms typically resolve within weeks due to central compensation. **Final comments:** LH is a rare cause of sudden SNHL and vestibular syndrome, often resulting in permanent ipsilateral deafness. Due to the scarcity of high-quality management data, treatment strategies should be individualized. Case reports such as this are important for raising awareness and acknowledgement of LH, since it may mimic acute vestibular neuritis.

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Acute weight loss and alexythymia after stroke in the right caudate nucleus

Janaina Mariana de Araujo Miranda Brito Marques[1], Paulo Roberto de Brito Marques[1]

[1]Universidade de Pernambuco, Recife PE, Brazil.

Correspondence: Janaina Mariana de Araujo Miranda Brito Marques; Email: marianaaraujo6@gmail.com.

Case presentation: 78-year-old woman, admitted to a neurology outpatient clinic complaining of weight loss of 30 kg after an ischemic stroke 6 months ago. Refers to a patient who had chest pain 6 months ago and underwent cardiac catheterization in the emergency room with normal results. Immediately after the procedure, she presented left hemiparesis, followed by hyporexia and adipsia. She reported not feeling hungry or thirsty and when she ate felt disgust and a sensation of change in the texture and flavor of the food (as if she were drinking oil). Her behavior from extroverted and communicative changed to reclusive with a feeling of social inadequacy and difficulty identifying and expressing what she felt, her mood became depressed. The neurological examination showed left hemiparesis with hyperreflexia and the presence of striatal hand (Fig.1). Mini-Mental State Examination1 23/30, Montreal Cognitive Assessment2 13/30, Depression Scale3 (Zung) 46/80, Toronto Alexithymia Scale4 73/100. Brain magnetic resonance imaging (MRI) showed hyperacute ischemic insult in the right caudate nucleus (Fig. 2). **Discussion:** The Caudate Nucleus (CN) contains several neuronal clusters that are functionally connected to cortical areas that are part of networks involved in the cognitive and emotional process, whose lesions compromise multiple cognitive and behavioral domains. Personality change, psychiatric disorders, and cognition symptoms can do clinical syndrome. Lesions located in the right CN through their interconnections, with the orbitofrontal, medial and anterior prefrontal, and cingulate cortical areas, are involved in these behaviors. We report the case of a 78-year-old woman, who, in addition to the motor deficit, developed acute weight loss, mood changes, hyporexia, adipsia, bizarre taste perceptions, and alexithymia, which we infer as possible disconnection of networks with the right insula and hypothalamus. The cognitive and emotional consequences of CN lesions, as illustrated in the patient, emphasize the concept of network disturbance as a cause of cognition and behavioral manifestations of vascular disorders. **Final comments:** There are few studies on cognition and behavioral disorders caused by unilateral CN injury. Although these syndromes present strong similarities, there are subtle differences, which likely reflect the specific contribution of neuronal processing within each cognition and emotional network.

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ADEM as the first manifestation of HIV in a young adult presented at a Brazilian tertiary hospital

Bruno Barros de Padua Andrade[1], Rodrigo Ferreira Faria[1], Giovanna Santos Rodrigues[1], Luiz Fernando Mohallem Reynaldo[1], Victoria Lot Avezum Mestriner[1], Elizardo Nogueira Junior[1], Marilia Mamprim de Moraes Perin[1], Matheus Spilari[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Bruno Barros de Padua Andrade; Email: brndepadua@gmail.com.

Case presentation: A 28-year-old previously healthy male was admitted following a motorcycle crash. He reported asymmetric paraparesis and hypoesthesia with a sensory level at T6, both worse on the left side, since eight days prior to admission. Additionally, he described urinary incontinence and constipation during the initial assessment and had difficulty describing the chronological order of events, presenting as slightly drowsy. Initially, he underwent an MRI study based on the hypothesis of spinal cord syndrome and encephalopathy, which revealed tumefactive lesions with a pseudotumorous aspect in the right frontal (2), temporo-parietal (1) cortex, at the supratentorial level, and multiple coalescing lesions at the level of the cervico-thoracic transition. Due to the characteristics and topography of the lesions, serologies and CNS inflammatory markers were requested, revealing HIV+ (ELISA and Western blot), a pathology the patient was previously unaware of. Until the CD4 count and further serologies were realized, IVIG (0.4g/kg/d) was initiated. After subsequent exams came back negative (CD4 749 cells/mm^3), treatment was transitioned to Methylprednisolone 1g/d. Upon discharge, the patient had improved motor and sensory capacity in the upper limbs but retained motor and sensory deficits in the lower limbs. As treatment progressed, the patient also showed improvement in consciousness, achieved sphincter competency, and no longer required intermittent catheterization. **Discussion:** Acute disseminated encephalomyelitis (ADEM) is a rare inflammatory and demyelinating disorder of the central nervous system. Distinctively, ADEMs pathological changes tend to have a perivenous localization and commonly affect children following exposure to infections and vaccines. It has been associated with various viral infections including EBV, CMV, and less commonly HIV. There are few descriptions in the literature of patients with atypical manifestations of ADEM and HIV, generally presented in immunocompetent individuals or during seroconversion (CD4 counts > 500 cells/μL), due to the necessity of "autoreactive" circulating T-cells that predispose the development of an autoimmune response to HIV. **Final comments:** Although the initial approach to multifocal deficits of subacute onset should consider demyelinating disorders, particularly in HIV-infected patients, it must be distinguished from a spectrum of other opportunistic diseases that may present similarly, especially neuroinfections. Recognition and diagnosis must be prompt and should not delay treatment.

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ADEM refractory to immunoglobulins as a presentation of mogad in a young woman

Adria Rodrigues da Silva[1], Bruna Beppler[1], Carol Zapelini[1], Lia Maki Hatisuka Imai[1], Luis Sidônio Teixeira da Silva[1], Lúcio Baena de Melo[1], Mônica Marcos de Souza[1], Gabriel Rodrigues Espelho Rossi[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Ádria Rodrigues da Silva; Email: adriarodriguess@hotmail.com.

Case presentation: Female patient, 43 years old, admitted to a tertiary hospital with a history of neck pain and sporadic holocranial headache for 2 years, worsening over the past 2 weeks. Symptoms included febrile episodes, paresthesia, and paresis in the left upper limb (LUL), followed by other limbs asymmetrically, confining her to a wheelchair. She also experienced tremors in the upper limbs, mental confusion, and urinary retention. Previously healthy, she had nodular skin lesions (neurofibromatosis type I). Neurological examination revealed inattentiveness, restlessness, aphasia, mild left facial paralysis, asymmetric tetraparesis, brisk reflexes, Hoffmann and Babinski signs present, bilateral exhaustible clonus, and predominant ataxia in the LUL. Cranial CT scan was normal. Cerebrospinal fluid (CSF) analysis showed 59 cells (88% lymphocytes) and 60 proteins. Serum serology for cytomegalovirus (CMV) IgM and IgG was positive, but negative in CSF with undetectable viral load. Oligoclonal bands were exclusively found in CSF. Neuroaxis MRI revealed T2/FLAIR signal abnormalities in the white matter adjacent to the left lateral ventricle body, left frontal lobe, and left amygdala, oval-shaped with contrast enhancement, and from C4 to T4 and lower dorsal levels/spinal cord cone, associated with pial surface enhancement. Despite switching from acyclovir to ganciclovir without improvement after 7 days of admission, intravenous human immunoglobulin 2g/kg was infused. The patient deteriorated, requiring therapeutic plasma exchange, and eventually discharged with improved consciousness, motor function, and ataxia, on Azathioprine 50 mg/day and Prednisone 10 mg/day. Subsequently, serum MOG-IgG test was positive (1:100), initiating periodic immunoglobulin therapy, weekly physiotherapy, and showing neurological improvement. **Discussion:** MOG antibody-associated disease (MOGAD) is an inflammatory demyelinating disease of the central nervous system (CNS) with heterogeneous clinical manifestations, ranging from isolated optic neuritis to multifocal CNS demyelination. Detection of MOG-IgG in serum confirms the diagnosis in patients with compatible clinical phenotypes and MRI findings. Concurrent infections such as rubella, CMV, and Epstein-Barr are reported, suggesting compromised blood-brain barrier during these exposures, leading to peripheral blood leakage of MOG antigens and substantial MOG-IgG production. In this case, however, the timing of CMV infection couldnt be determined. Crises often respond to high-dose corticosteroids, with plasma exchange reserved for refractory cases. Maintenance therapy relies on empirical immunosuppressants, though further studies are needed for treatment guidance. **Final comments:** MOGAD is a recently discovered, rare disease with variable phenotypes, challenging diagnosis, and uncertain treatment.

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ADEM-like presentation in an immunocompetent host after disseminated tuberculosis: TB or not TB?

Jackeline Neves Pereira[1], Pedro Henrique Fraiman[1], Raphael Gusmao Barreto[1], Fernanda Ferreira Abreu[1], Gabriel Casela Speltri[1], Paula Massarani Peçanha-pietrobon[1], Marianna M. Moraes[1], Paulo Roberto Abrão Ferreira[1], Orlando Graziani Povoas Barsottini[1], Jose Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Jackeline Neves Pereira; Email: jac.nevesp@gmail.com.

Case presentation: A 23-year-old woman in the late stages of pregnancy presented to the Hospital with fever and abdominal pain. She was diagnosed with pyelonephritis and underwent an emergency cesarean section due to fetal bradycardia. During the days following the surgery, she maintained abdominal pain and an abdomen computerized tomography (CT) scan revealed a pancreatic solid-cystic mass, signs of pyelonephritis, colonic swelling, and peritoneal implants. She developed pneumoperitoneum due to a cecum perforation, the histopathological analysis confirmed disseminated tuberculosis (TB), in addition to an incidental pancreatic tumor suggestive of pseudopapillary pancreatic neoplasm (later confirmed through biopsy). She tested negative for HIV and infectious hepatitis. After two weeks of standard TB treatment, she developed optic neuritis in the right eye, rapidly deteriorating to a hand movement perception. The first hypothesis was an effect of Ethambutol, which was then suspended. Nevertheless, one week later, the contralateral eye was also affected. The brain Magnetic Resonance (MR) revealed signs of bilateral optic neuritis. Cerebrospinal fluid (CSF) study was negative for TB and neoplasm cells, besides a normal glucose, protein, and cell count. Despite Methylprednisolone pulse therapy and plasmapheresis, she maintained bilateral visual loss and developed weakness in the right arm. A new brain MR revealed hyper-intensities in the frontal lobe, corpus callosum, and brain stem, compatible with demyelinating lesions. The brain biopsy was negative for TB and compatible with demyelination. The auto-immunity panel was negative for either surface or intranuclear antibodies, including anti-MOG antibodies. After hospital discharge, the patient presented a progressive partial recovery from her neurological deficits until the day of this report and could see the face of her child for the first time after 5 months of having given birth. **Discussion:** The presented case had a series of challenges to the correct diagnosis of her neurological symptoms: an immunocompetent patient, a pancreatic neoplasm, repetitive negative molecular tests for TB in the CSF, Ethambutol side effects similar to the initial visual deficit, and progression of neurological symptoms during treatment despite corticosteroids use. Indirect effects of TB in the central nervous system are not the most common presentation of the disease, especially of demyelinating nature, presenting as Acute Demyelinating Encephalomyelitis (ADEM) like syndrome, as we describe here. **Final comments:** ADEM-like syndrome related to TB has been reported before and should be kept in mind by neurologists, especially in countries with a high prevalence of the disease. The differential diagnoses are mostly Ethambutol side effects or direct TB effects in the CNS. The rapid progression and characteristics of lesions monitored by MRI should help to differentiate, in addition to the usual benign course with symptoms and image regression.

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Adherence to AHA life's essential 8 criteria in stroke patients: an observational study

Catarina Dantas Corrêa[1], Eduarda de Almeida Joly[1], Gabriel de Camargo de Brito[1], Gabrielle Beatriz da Cruz Ferreira[1], Pedro Paulo Daniel Rocha[1], Renata Dal-prá Ducci[1], Valéria Cristina Scavasine[1], Juliano André Muzzio[1], Viviane de Hiroki Flumignan Zétola[1], Marcos Christiano Lange[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Catarina Dantas Correa; Email: catarinadcorrea@gmail.com.

Background: Cardiovascular diseases are a major cause of mortality and morbidity worldwide. In Brazil, cardiovascular diseases are the leading cause of non-traumatic death. The American Heart Association (AHA) Lifes Essentials 8 framework (comprising Diet, Physical Activity, Nicotine Exposure, Sleep Health, Body Mass Index, Blood Lipids, Blood Glucose, and Blood Pressure) provides a holistic approach to health, shifting the focus from merely treating cardiovascular diseases to promoting and preserving overall health. **Objective:** To evaluate the adherence of stroke patients to the American Heart Association (AHA) Lifes Essential 8 criteria. **Methods:** This observational study analyzed data from 41 patients admitted to a stroke unit over a four-month period. Data were collected on diet, physical activity, smoking, sleep, BMI, non-HDL levels, blood glucose, and blood pressure, with scores ranging from 0 to 100 according to the Lifes Essential 8 criteria. **Results:** The median overall Lifes Essential 8 score for the study population was 58 (IQR 50-65). Median scores for lifestyle habits were as follows: adherence to diet, 50 (IQR 25-50); physical activity, 0 (IQR 0-40); smoking, 75 (IQR 0-100); and sleep quality, 90 (IQR 40-100). For risk factors, the median scores were: BMI, 70 (IQR 70-100); non-HDL levels, 80 (IQR 60-100); blood glucose, 100 (IQR 60-100); and blood pressure, 30 (IQR 5-100). **Conclusion:** This study demonstrated low adherence scores for blood pressure, physical activity, and diet according to the Lifes Essential 8 criteria. These findings suggest that health intervention strategies and policies should focus on improving physical activity and diet among stroke patients.

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Admission trends for epilepsy in the Northeast region over the last 10 years and the impact of the COVID-19 pandemic

Willian Alves Costa[1], Danilo Silva Nogueira[1], Juliana Alencar Isacksson Vieira[1], Vinícius Gabriel Nunes de Freitas[1], Pedro Antônio Tavares Pinto[1], Agnus Gabriel Bitencourt Magalhães[1], Francisco Edvan Batista Filho[1], Thiago da Costa Brito[2]

[1]Universidade Federal do Amapá, Macapá AP, Brazil. [2]Universidade Estadual do Pará, Belém PA, Brazil.

Correspondence: Danilo Silva Nogueira; Email: danilo-nogg@hotmail.com.

Background: Epilepsy is one of the most prevalent chronic neurological conditions in Brazil. It is characterized by recurrent epileptic seizures, related to abnormal electrical activity in the brain, without a readily defined or identified causal factor. **Objective:** this study aims to outline an epidemiological overview of hospital admissions due to epilepsy in the Northeast region over the past decade, as well as to evaluate the implications of the COVID-19 pandemic. This works aims to make. **Methods:** This is a descriptive, retrospective, and cross-sectional study involving the collection and analysis of data from the Hospital Information System (SIH/SUS), extracted from the Department of Health Informatics of the Unified Health System (DATASUS). The category "Epilepsy" was analyzed in the Northeast region from 2014 to 2023, considering variables such as Region, Federative Unit, year, race, age group, sex, and hospital admissions. The collected data were organized into tables using EXCEL 2016 software. **Results:** The Northeast region constitutes the second highest region in terms of epilepsy hospital admissions reported over the past 10 years (128,605 cases). The majority of cases were reported in the states of Pernambuco and Bahia (27.2% and 27.08% respectively). Conversely, Sergipe had the lowest number of recorded cases (2.31%). Over these 10 years, there was a progressive increase in cases, with 2022 and 2023 showing the highest prevalence (12.6% and 11.9% respectively). A sharp decline was observed in 2020, with a prevalence of 9.2%. In Pernambuco, there were variations in this

pattern regarding the peak years; instead of 2022 and 2023 , 2021 and 2022 showed the highest prevalence (3.3% and 3.4% respectively). Regarding demographic profile, males (58.6%) and individuals of mixed race (67.2%) constituted the majority of cases. In terms of age group, prevalence was highest among children aged 0 to 9 years (38.9%), particularly in the state of Ceará (25.12%). Conversely, individuals aged 15 to 19 years and those aged 80 years and above were the least affected (5.4% and 4% respectively). **Conclusion:** Therefore, based on the results obtained, it was observed that epilepsy has been on the rise in the Northeast region over the past 10 years, particularly in the state of Pernambuco and among males and individuals of mixed race. Regarding age groups, there was notable prevalence among children aged 0 to 9 years; however, these findings are closely linked to the high incidence rates affecting this age group in the state of Ceará. Additionally, it was noted that the Covid-19 pandemic had a significant impact on the number of hospital admissions, as evidenced by the significant decrease reported in 2020.

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Advances in modulation of the microbiota-gut-brain axis: treatment of neuropsychological disorders and functional gastrointestinal disorders

Jocsã David Gomes Santos[1], Luan Kelves Miranda e Souza[1]

[1]Instituto de Educação Superior do Vale do Parnaíba, Parnaíba PI, Brazil.

Correspondence: Jocsã David Gomes Santos; Email: jocsadavid.jd@gmail.com.

Background: The bidirectional communication between the gastrointestinal system and the central nervous system, known as the microbiota-gut-brain axis, is important for the regulation of physiological and psychological functions. In this context, contemporary science has revealed the role of the gut microbiota in modulating this axis, offering new therapeutic perspectives for neuropsychological and gastrointestinal disorders. The influence of the microbiota on mental health and gastrointestinal function has attracted significant scientific interest, highlighting the need to explore interventions based on axis modulation. **Objective:** To review recent advances in the modulation of the microbiota-gut-brain axis, focusing on the application of microbiota-based interventions for the treatment of neuropsychological and functional gastrointestinal disorders. **Methods:** This is an integrative literature review with a cross-sectional, retrospective design, using the descriptors "Microbiome-Brain-Gut Axis", "Neurocognitive Disorders", and "Gastrointestinal Microbiome" from the Health Sciences Descriptors (DeCS), combined with the Boolean operator AND. The databases used were PubMed, Scielo, and Dynamed. The research included articles in Portuguese, English, and Spanish, within the time frame of 2020 to 2024. Original articles, systematic reviews, and meta-analyses were included, while articles unrelated to the topic, letters to the editor, editorials, and opinion articles were excluded. **Results:** The modulation of the microbiota-gut-brain axis through probiotics involving Bifidobacterium and Lactobacillus reduces cortisol levels and has shown efficacy in reducing symptoms of neuropsychological disorders, such as depression and anxiety, as well as functional gastrointestinal disorders, such as irritable bowel syndrome. Additionally, the administration of serotonin receptor agonists, such as psilocybin or 2,5-dimethoxy-methylamphetamine, has shown significant therapeutic potential, as elevated serotonin levels in the intestinal lumen promote the proliferation of bacteria like Turicibacter sanguinis, which can regulate genetic pathways in the gastrointestinal system and be used as treatments for depression, anxiety, and inflammatory bowel diseases. Studies also highlight the importance of personalizing interventions based on the individual microbiota profile of patients to increase efficacy. **Conclusion:** Microbiota-based therapies represent a significant advancement in the treatment of neuropsychological and gastrointestinal disorders, with a reduced side effect profile compared to traditional treatments. The incorporation of these interventions into clinical practice could revolutionize the management of various conditions, significantly improving patients' quality of life.

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Advances in the treatment of glioblastoma: traditional therapeutic strategies vs. modern therapies - systematic review

Graziele Gleice da Silva[1], Ana Silvia Suassuna Carneiro Lúcio[1], Alexandre Jorge de Andrade Negri Júnior[1]

[1]Faculdade de Ciências Médica da Paraíba, João Pessoa PB, Brazil.

Correspondence: Graziele Gleice da Silva; Email: grazielle_gleice@hotmail.com.

Background: Glioblastoma (GBM) is a grade 4 astrocytoma (WHO), the most common primary malignant tumour of the central nervous system (CNS). It is responsible for about half of the malignant CNS tumours, with a 5-year survival rate of 7.2%. The standard GBM therapy includes neurosurgery, radiotherapy (radiation therapy), and chemotherapy. This type of therapy has not been very successful due to recurring side effects. There is however a promising, modern form of therapy consisting of immunotherapy and virotherapy, combining oncolytic viruses [OVs] and cancer vaccines. Additionally, significant progress is notable in tomo-, NanoTherm-, or individualized multimodal immunotherapy (IMI). Ongoing challenges in GBM treatment include insufficient resection, a high degree of heterogeneity, and an immunosuppressive microenvironment. **Objective:** To highlight the most modern therapies in the treatment of glioblastoma and their effectiveness compared to traditional therapies. **Methods:** This is a systematic review of the literature guided by the question: "What are the new therapies for glioblastoma?" The article selection took place between February and May 2024 carried out through the U.S. National Library of Medicine (PUBMED) database. The inclusion criteria were: primary original articles addressing new therapies in GBM treatment, available in Portuguese and in English. There were no restrictions on the study design, however, only articles from 2021 to 2023 were selected. Exclusion criteria included articles that did not address the theme. The following indexed descriptors from Health Sciences Descriptors (DeCS) were used: "Glioblastoma", "Immunotherapy", and "Virotherapy". **Results:** Immunotherapy combined with virotherapy and cancer vaccines is an innovative therapeutic regimen. Personalized medicine encompasses the patient as a whole in the context of immunotherapy, such as individualized multimodal immunotherapy (IMI). This is based on individual tumour-host immune interactions and the concept of OV-induced immunogenic tumour cell death (ICD). Ongoing challenges in GBM treatment include insufficient resection, a high degree of heterogeneity, and an immunosuppressive microenvironment. Treatment with OVs is more beneficial and safer than regular treatment, with fewer side effects, making it a promising course of treatment for GBM. **Conclusion:** Given that there are modern treatments available, a transformation in the clinical trials is needed, both to their specific approach and to individual patient approach. This will lead to more targeted guidelines and better humanization of GBM patients' treatment.

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Advantage of FilmArray® over conventional methods in the diagnosis of acute bacterial infections of the central nervous system, including meningitis caused by listeria monocytogenes

Renan Domingues[1], Irineu Massaia[1], Camila Spinelli[1], Myrna Monteiro[1], Márcio Vega[1], Lais Guerra[1], Daiane Salomão[1], Carlos Senne[1]

[1]Senne Liquor Diagnóstico, São Paulo SP, Brazil.

Correspondence: Renan Barros Domingues; Email: contato@renandomingues.med.br.

Background: The etiological diagnosis of bacterial meningitis is fundamental for supporting accurate therapeutic decisions, making the early initiation of antimicrobials crucial. **Objective:** In this study, we evaluated the performance and utility of FilmArray® compared to conventional microbiological methods in the diagnosis of bacterial meningitis. **Methods:** We retrospectively evaluated CSF data from 2502 samples of patients suspected of having CNS infection, focusing on the CSF data from cases where a bacterial agent was identified. **Results:** Ninety cases were positive for bacteria, with the following agents identified: E. coli K1 = 3, H. influenzae = 17, L. monocytogenes = 8, N. meningitidis = 16, S. pneumoniae = 35. Of these, only 21 (23.3%) were positive with conventional microbiological methods, including culture and/or bacterioscopy, with the following distribution: E. coli K1 = 0/3 (0%), H. influenzae = 4/17 (23.5%), L. monocytogenes = 2/8 (25%), N. meningitidis = 5/16 (31.2%), S. pneumoniae = 8/35 (22.8%). For the 8 samples positive for L. monocytogenes, the cell count was 853.4 cells/mm3 (90-2560 cells/mm3), with neutrophils in CSF ranging from 17% to 78%. The protein level was 146.5 mg/dl (69-263 mg/dl), glucose was 30.5 mg/dl (12-238 mg/dl), and lactate was 82.1 mg/dl (44.9-160.3 mg/dl). Only two cases had a positive culture. **Conclusion** The study confirms previous data indicating that FilmArray® significantly increases the sensitivity of the etiological diagnosis of bacterial meningitis for all agents included in the panel, including L. monocytogenes. The results of this study corroborate the low sensitivity of conventional microbiological methods and suggest that molecular methods constitute the best tool for the etiological diagnosis of these cases.

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Aerobic physical exercise improves quality of life in temporal lobe epilepsy

Shai Oisiovici Laks[1], Nathalia Volpato[1], Nikolas Coelho[1], Mateus Henrique Nogueira[1], Clarissa Lin Yasuda[1], Luciana Ramalho Pimentel da Silva[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Shai Oisiovici Laks; Email: s243177@dac.unicamp.br.

Background: Aerobic physical exercise (APE) has been showing improvement of quality of life (QoL) and mood disorders in a series of chronic brain diseases [1]. A prior study from our group showed that people with temporal lobe epilepsy (TLE) with an active lifestyle present better QoL, but the influence of APE in the QoL of TLE is still unclear[2]. As drug-resistance is commonly associated with TLE, the possibility of seizures happening during the activities might prevent patients from benefiting from APE. **Objective:** We aimed to analyze the influence of APE in the QoL of TLE patients, also focusing on the seizure worry subitem of the QoL in epilepsy 31 (QOLIE-31) questionnaire. **Methods:** We selected and reviewed data from

51 individuals with TLE who participated in a 6-month APE program under the supervision of a board-certified physical trainer. TLE patients were divided into training (TLE-training) and control (TLE-control) groups. The TLE-training group went through an APE program of 6 months, while the TLE-control group was oriented to keep their routine. Both groups were assessed by the QOLIE-31 before and after the intervention (APE-time). A mixed-between subjects ANOVA was conducted to assess the impact of the APE intervention compared to TLE-control pre- and post-intervention on the QoL. We set p< .05 as statistically significant. **Results:** There was a significant interaction between intervention-groups and APE-time (p=.0005), showing that only patients on the TLE-training group presented with higher QoL after the intervention (p=0.001). As for seizure worry, we have not found any significant differences between groups (p=0.50). **Conclusion:** The APE improved QoL and did not increase the SW, which might be a feature of concern for both patients and healthcare practitioners. Further studies should focus on long term interventions to evaluate the long-term impact of APE in the QoL of TLE patients.

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Age of onset and clinical characteristics of neuromyelitis optica spectrum disorder: a cross-sectional study in Espírito Santo

Vinícius Andreata Brandão[1], Igor Flavio Gonçalves do Vale[1] , Mackeyle Siqueira de Souza[1], Sávio Cesar Rodrigues Pestana[1], Victória Pagani Samora Sousa[1], Paula Zago Melo Dias[2], Bruno Batitucci Castrillo[2], Carlos Alberto Magirius Peixoto[2]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.
[2]Hospital Universitário Cassiano Antônio Moraes, Vitória ES, Brazil.

Correspondence: Vinícius Andreata Brandão; Email: viniciusandreatab@gmail.com.

Background: Neuromyelitis optica spectrum disease (NMOSD) is a rare autoimmune condition, usually associated with antibodies against aquaporin-4 (anti-AQP4), which leads to demyelination of the central nervous system, mainly affecting the optic nerve and spinal cord. Typically, the onset of these symptoms occurs between the third and fourth decades of life, although, less frequently, they can appear in any age group. **Objective:** To assess the age of onset of symptoms in patients diagnosed with NMOSD with positive IgGaquaporin-4. **Methods:** This is a cross-sectional study carried out at a neurology outpatient clinic in a public health service in Espírito Santo, Brazil. The electronic medical records of patients diagnosed with NMOSD were reviewed to assess the age of patients at the first disease attack. Data was categorized into late onset NMOSD (LONMOSD), after the age of 50; and very-late onset NMOSD (VLONMOSD), after the age of 70. **Results:** The average age of onset of symptoms among the 31 patients studied (28 women and 3 men) was 38.5 years, ranging from 13 to 79 years. Among these patients, six (19.3%) started their symptoms before the age of 20, while eight (25.8%) had their first symptoms after the age of 50. Of those who had their first symptoms after 50, 16.19% were classified as LONMOSD and 9.67% as VLONMOSD. Patients who experienced their first attack after the age of 50 had a lower incidence of optic neuritis, with an average of 0.6 attacks per person (standard deviation of 0.7) over two years of disease. In contrast, patients below the age of 50 had an average of 1 attack per person (standard deviation of 0.6). Regarding longitudinally extensive transverse myelitis (LETM), both age groups had an average of 1 attack over two years. **Conclusion:** The average age of onset found in this study aligns with global literature, which indicates a peak prevalence of NMOSD around the age of 40. However, the prevalence of NMOSD after the age of 50 is notably higher than the global average. The clinical characteristics observed in patients with LONMOSD demonstrates higher incidence of spinal cord lesions compared to optic neuritis (ON), often resulting in a poorer prognosis compared to patients with onset before age 50. Furthermore, older patients are more likely to have comorbidities and reduced immune function, which contribute to worse clinical outcomes and a lower life expectancy free from disability, despite aggressive immunosuppressive treatment.

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Age profile of mortality in viral and bacterial meningitis cases: an epidemiological study (2012-2022)

Paulo Victor de Santiago Gonçalves[1], Leonardo José Rodrigues de Araújo Melo[1], Leslie Mateus de Sousa Nunes[1], Victor Domingueti Vallim Fonseca[1], Francisco Luciano Honório Barreto Cavalcante[1], Cássia Caroline Aguiar da Ponte[1], Cynthia Paz Rodrigues[1], Leonardo Elias Araujo[1], Lucas Soares Radtke[1], Heitor Rios de Medeiros Virginio[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Paulo Victor de Santiago Gonçalves; Email: paulo.santiago6571@gmail.com.

Background: Infections that affect the meninges, causing inflammation known as meningitis, can result from various bacterial and viral pathogens. These agents typically exhibit higher incidence rates across different age groups, capitalizing on the varying immunological characteristics of individuals at different life stages. **Objective:** This study aims to delineate the age profile of patients with confirmed deaths due to viral and bacterial meningitis between 2012 and 2022. **Methods:** We conducted an epidemiological investigation of confirmed fatal cases of viral and bacterial meningitis spanning the years 2012 to 2022. Data were collected from the DATASUS Notifiable Diseases Information System, and statistical analysis was performed using Jamovi software. **Results:** Based on epidemiological data from DATASUS for the period 2012 to 2022, deaths attributed to clinical complications of bacterial meningitis (6,613 deaths) significantly outnumbered those caused by viral meningitis (683 deaths). When examining patient age groups, we observed that 11.72% of deaths occurred in children under 1 year old due to bacterial infections, and 8.5% were related to viral infections. These findings indicate a declining trend up to age 19, followed by an increase with advancing age. The highest mortality rates occurred in the 50 to 69 years age group for bacterial meningitis (30.6% of deaths) and the 40 to 59 years age group for viral meningitis (26.2% of deaths). Similar to the first two decades of life, there is a decline in mortality beyond age 70. **Conclusion:** The collected data allows us to construct an age profile of meningitis victims. Notably, very young children face a higher risk of severe infection and mortality. As physiological development progresses (up to the end of adolescence), the death rate tends to decrease, only to rise again, peaking around age 50 for bacterial variants and age 40 for viral variants. However, after this peak, cases decline beyond age 70 for both studied variants.

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Albendazole plus praziquantel versus albendazole alone in the treatment of neurocysticercosis: a systematic review and meta-analysis

Luísa Menegaz Veronese[1], Gabriela Oliveira Caetano[1], Jade Lingiardi Altoé[2], Laíse Lênin Leodoro Silva[1], Maria Eduarda Silva[1], Artur Menegaz de Almeida[1], Ítalo Barros Andrade[1], Michele Kreuz, Davi Tanajura Costa[3]

[1]Universidade Federal de Mato Grosso, Sinop MT, Brazil.
[2]Universidade Cesumar, Maringá PR, Brazil.
[3]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Artur Menegaz de Almeida; Email: arturomenegaz@gmail.com.

Background: Neurocysticercosis is a parasitic neurological disease, considered by the World Health Organization (WHO) as one of the most neglected tropical diseases in the world, also being an important public health concern in Brazil. The eggs of the Taenia solium hatch and the larvae spread through highly vascularized tissues, including the brain. Most patients are asymptomatic, but in some cases, cysts can cause inflammation, which can lead to headaches, seizures, and intracranial hypertension. The preferred treatment for neurocysticercosis remains unclear: the combination therapy of albendazole plus praziquantel has been associated with greater benefits than albendazole alone. **Objective:** To assess the safety and efficacy of albendazole plus praziquantel therapy for neurocysticercosis in comparison to albendazole alone. **Methods:** MEDLINE, Embase, and Cochrane databases were systematically searched until May 11, 2024. Data were examined using the Mantel-Haenszel method and 95% confidence intervals (Cls). Heterogeneity was assessed using I^2 statistics. Primary outcomes were the reduction of cysts, seizure recurrences, and possible adverse events. R, version 4.3.2, was used for statistical analysis. **Results:** A total of 4 randomized controlled trials and 313 patients, in which 129 (41%) were submitted to combined therapy. Compared to albendazole monotherapy, the combination of albendazole plus praziquantel regimen achieved better rates of cyst resolution (68% vs 42%; odds ratio [OR] 2.87; CI 95% 1.74-4.72; P = 0.00003; I^2 = 1%). Furthermore, no difference was seen regarding the occurrence of adverse events (18% vs 23%; risk ratio [RR] 1.17; CI 95% 0.76-1.79; P=0.481345; I^2= 12%). Seizure relapse endpoint did not reach a statistically significant difference between groups (10% vs 9%; risk ratio [RR] 1.36; CI 95% 0.68-2.72; P= 0.383208; I^2= 0%). **Conclusion:** In this meta-analysis, consistent results suggest that albendazole plus praziquantel combined therapy is associated with more beneficial outcomes than albendazole alone in treating neurocysticercosis.

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Alien hand syndrome associated with lesion in the corpus callosum and frontal lobe

Sávio Roberto Silva Costa[1], Fernando Gabriel dos Santos Santiago[1], Gustavo Celeira de Sousa[1], Danusa Neves Somensi[1], Emanuel de Jesus Soares de Sousa[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Gustavo Celeira de Sousa; Email: gustavo.c.sousa0@gmail.com.

Case presentation: Male, 59 years old, describes complaints of involuntary movements in the right hand that began 1 year after a stroke. These movements have characteristics of grasping, strangling, or pulling. However, the movements can also precede the motor act, causing the limb to make gestural movements in relation to speech or perform movements that the patient intended to do with the contralateral limb. Additionally, it presents motor incoordination in the left upper and lower limbs after the stroke. MRI shows gliosis in the left frontal lobe from the superior frontal gyrus to the precentral gyrus, as well as signs of ischemia in the corpus callosum and cingulate gyrus. On the physical examination, he is in good general condition, scoring 20/30 on the mini-mental state examination. Neurological examination reveals pathological findings such as absence of movement in the right foot, hyperreflexia, and increased reflexogenic area in the right patellar reflex. Motor coordination is

altered in the index-nose test with the left hand, and cranial nerve evaluation shows right nasal hemianopsia. **Discussion:** This case represents a rare condition known as alien hand syndrome, defined as involuntary and seemingly purposeful autonomous activity, combined with the patients erratic perception, usually associated with difficulty recognizing the limb in the absence of visual cues. It is worth noting that the hand is the most commonly affected limb, but the alteration can be found in other limbs as well. The pathophysiology describes lesions in the frontal lobe or the anterior portion of the corpus callosum, and, more rarely, in the parietal region. The clinical presentation varies according to the affected location. Thus, lesions in the frontal lobe, such as the supplementary motor area, cingulate gyrus, and medial prefrontal cortex, mainly affect the dominant hand and are characterized by frontal release signs - grasping and groping. In lesions of the corpus callosum, intermanual conflict is presented, while parietal lesions provide a sensation of limb estrangement and the appearance, mainly, of the levitation phenomenon. Therefore, the similarity between the imaging findings of the patient in question and the characteristic clinical manifestations, mainly of the frontal lobe and corpus callosum lesion variants, is evident. **Final comments:** The onset of this syndrome is generally due to acute focal lesions, and in this condition, recovery occurs within 1 year. However, in chronic and degenerative brain lesions, the clinical symptoms may persist for life. Currently, there is no specific treatment or rehabilitation for the condition, but it does not cause any harm to the patient's physical integrity.

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Alterations in the motor development of children with autism spectrum disorder level 2, according to the portage inventory: a cross-sectional study

Larissa da Fonseca Guimarães[1], Laércio da Silva Paiva[1], Débora Bachin Carvalho[2,3], Noelia Barbosa de Oliveira[3], Lívia Joelma Almeida de Lima e Silva[3], Clara Criez de Lima Santos[3], Lucas Tomita Sodré[3], Marina Ortega Golin[1]

[1]Centro Universitário Faculdade de Medicina do ABC, Santo André SP, Brazil.
[2]Universidade Nove de Julho, São Paulo SP, Brazil.
[3]FisioPETI, São Bernardo do Campo, SP, Brazil.

Correspondence: Débora Bachin Carvalho; Email: dbachinc@gmail.com.

Background: Autism Spectrum Disorder (ASD) is classified into three levels of severity. At level 2, the child needs substantial support and is subject to motor skills delays. A frequently used instrument to assess these children is the Operationalized Portage Inventory (OPI). Although the OPI is widely used, studies describing the identified motor difficulties are still rare. **Objective:** To analyse the overall motor performance of children with ASD level 2, according to the OPI, and to check the correlation to their performance in language, cognition, self-care and socialization. **Methods:** This is a retrospective observational cross-sectional study, analyzing 47 medical records of children aged between three and six years old, seen at FisioPETI Rehabilitation Clinic, São Bernardo do Campo / SP. The project was approved by the Research Ethics Committee of the Faculdade de Medicina do ABC University Center, protocol no. 70421023.0.0000.0082. The following data was collected: age, gender, Childhood Autism Rating Scale (CARS) score, developmental classification (in years) and quantification of developmental delay (in months) in each dimension of the OPI. Pearson's correlation test was used to relate the delay in overall motor performance to the other dimensions. The sample power calculation to quantify the delay in global motor performance was 100%. **Results:** In the sample of 47 patients (CARS median: 33), 74.47% were male, 91.49% showed delay in global motor performance, 97.87% in self-care and 100% in language, cognition and socialization. The median age was 4.0 years old, the global motor development, 2.7 (years); self-care, 2.5; cognition, 1.7; socialization, 1.6 and language, 0.9. The quantification of developmental delay in months (average) was 19.93 in global motor, 21.06 in self-care, 29.76 in cognition, 31.52 in socialization and 36.87 in language. There was a significant correlation between developmental delay and all other dimensions: language ($p< 0.001$), cognition ($p< 0.001$), socialization ($p< 0.001$) and self-care ($p< 0.001$). **Conclusion:** The largest proportion of children with ASD level 2 showed delays in all neurodevelopmental dimensions assessed by the OPI, however, motor development was less severe. A correlation was found between the severity of the motor alteration and the impairment in all other developmental dimensions.

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Altered cerebral vasoreactivity on transcranial color-coded sonography related to akinetic-rigid phenotype of Parkinson's disease and individuals with Parkinson's disease and risk factors for cerebrovascular disease

Rodrigo Tavares Brisson[1], Rita de Cássia Leite Fernandes[1], Josevânia Fulgêncio de Lima[1], Liene Duarte Silva[1], Marco Antônio Sales Dantas de Lima[1], Ana Lucia Zuma de Rosso[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Rodrigo Tavares Brisson; Email: quimicarod@hotmail.com.

Background: A correlation between worse functional outcomes in Parkinson's disease (PD) patients with cerebrovascular disease (CVD) or the Akinetic-rigid phenotype has been argued in recent studies. **Objective:** To evaluate the association of cerebral hemodynamics impairments, assessed by Transcranial Color-coded Doppler sonography (TCCS), on PD patients with different phenotypes of the disease and with risk factors for CVD. **Methods:** Idiopathic PD patients (n = 64) were divided into motor subtypes: Akinetic-rigid (AR) (n = 31) and Tremor-dominant (TD) (n = 33) and into two groups regarding vascular risk factors: when ≥2 were present (PDvasc) (n = 20) and < 2 (PDnvasc) (n = 44). In a parallel analysis, the Fazekas scale on brain magnetic resonance imaging (MRI) was applied to a sample to assess the degree of leukoaraiosis. TCCS examinations were prospectively performed obtaining middle cerebral artery Mean Flow Velocities (Vm), Resistance Index (RI), and Pulsatility Index (PI). The Breath-Holding Index (BHI) was calculated to assess cerebrovascular reactivity (cVR). Standardized functional scales were administered (UPDRS III and Hoehn&Yahr). **Results:** The phenotype groups were similar in age, disease duration and demographic parameters, but there were significantly higher H&Y scores on RA group (2,66±1,6 vs 2,21±0,78; p=0.05). cVR was impaired in 64% of AR vs. 36% of TD (odds ratio: 3,18). AR group exhibited lower BHI (0.56 ± 0.32 vs. 0.97 ± 0.60; $p = 0.001$), lower Vm after apnea (45.8 ± 10.9 cm/s vs. 53.18 ± 10.4 cm/s; $p = 0.008$), higher PI (0.91 ± 0.24 vs. 0.8 ± 0.15; $p = 0.029$). PDvasc group showed higher PI (0.97 ± 0.22 vs. 0.80 ± 0.18; $p = 0.001$), higher IR ($0.59 \pm 0,09$ vs. 0.54 ± 0.08 ; $p = 0.042$) and higher frequency of altered cVR (75% vs. 38%; $p = 0.007$ - odds ratio: 4,7). There was a significant predominance of higher values on Fazekas scale in the PDvasc group (p=0.004). We found no difference between the Fazekas scale when comparing motor subtypes groups but there was a trend toward higher scores in the AR phenotype. **Conclusion:** TCCS, a cost-effective method, displayed impaired cVR in Parkinsonian patients with risk factors for CVD with higher degree of MRI leukoaraiosis. PD patients with the AR disease phenotype also presented higher frequency of impaired cVR on TCCS and greater functional impairment, although with just a trend to higher scores on MRI Fazekas.

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Alternating hemiplegia in childhood associated with epilepsy, a rare disease related to ATP1A3 gene mutation: case report

Vanessa Lopes Senssuliin[1], Andrea Senna Pessoto Serafin Gallina[1]

[1]Universidade Anhembi Morumbi, Piracicaba SP, Brazil.

Correspondence: Vanessa Lopes Senssulini; Email: vsenssulini@gmail.com.

Case presentation: A 3-year-old female patient, only child of non-consanguineous parents, began experiencing bilateral muscle spasms and rotary eye movements at three months of age. During that period, transfontanelle ultrasound and electroencephalogram (EEG) were both normal. Despite this, she was diagnosed with epilepsy and started treatment with Phenobarbital combined with oxcarbazepine. Over a year, attempts to taper medication resulted in worsened symptoms, including tonic-clonic seizures, persistent eye movements, and right-sided hemiplegia. Due to refractory symptoms and clinical deterioration, Levetiracetam was added, followed by Clobazam, yet seizures persisted. Various diagnostic tests were conducted, including cranial MRI and EEG, all yielding no significant abnormalities. A geneticist recommended genetic testing for ataxias and epilepsy, identifying a heterozygous pathogenic variant in the ATP1A3 gene (Gly947Arg). Flunarizine was subsequently added to the treatment plan based on genetic findings, alongside ongoing multidisciplinary management. **Discussion:** The patient was ultimately diagnosed with Alternating Hemiplegia of Childhood (AHC) associated with the ATP1A3 gene mutation, despite initial treatment for epilepsy. The link between AHC and this mutation is recent, with most cases identified post-2012. Diagnostic criteria for AHC, established since 1993, include onset before 18 months, autonomic phenomena, cognitive impairment, recurring episodes of hemiplegia involving both sides of the body, neurological abnormalities like choreoathetosis, symptom disappearance during sleep, and reoccurrence upon awakening. The patient exhibits five of these criteria. However, definitive diagnosis is hindered by the disease's rarity (annual incidence < 1 per 100,000 newborns), phenotypic variability, and limited genetic data in epidemiological studies. The significance of a definitive diagnosis resides in its capacity to guide optimal clinical management, even in the absence of a specific treatment, with current management focusing on symptomatic measures and multidisciplinary care to improve patient quality of life. **Final comments:** Prospective, randomized, controlled studies are needed to develop more effective treatments that not only alleviate symptoms but comprehensively address the disease. Increased research publication will deepen understanding of disease mechanisms, diagnosis, and clinical aspects, benefiting clinical practice and a broader patient population with this disease. Etics committee approval : 79893624.0.0000.5492

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Alzheimer's disease and antipsychotics, a troublesome match

Gabriella Chinaglia Zanin[1], Caio Hussid de Góes[1], Diego Olympio Peixoto Lopes[2], Maria Tereza Martinez[3], Fernanda Herculano[4], Diogo Haddad Santos[4]

[1]Faculdade de Ciências Médicas da Santa Casa de São Paulo, São Paulo SP, Brazil.
[2]Universidade Santo Amaro, São Paulo SP, Brazil.
[3]CPQuali Pesquisa Clínica, São Paulo SP, Brazil.
[4]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Gabriella Chinaglia Zanin; Email: gabriella.zanin@aluno.fcmsantacasasp.edu.br.

In the context of Alzheimer's disease, antipsychotic medications are used to treat behavioral and psychological symptoms of dementia, such as agitation, aggression, psychosis and delirium, with a prevalence between 40 to 50%. The choice of antipsychotic is particularly delicate, as they have important side effects, and some have been associated with higher mortality. While between 20 to 50% of patients with Alzheimer receive antipsychotic prescriptions, there are comparatively few studies with Brazilian data related to their usage. The present study's aim was to analyze antipsychotic drugs prescribed to patients with Alzheimer in the last 5 years in reference neurology center in São Paulo, while comparing it to the general Brazilian usage of antipsychotic in Alzheimer patients. An electronic medical record aggregator called TriNetX was used to anonymously obtain the patient data used in the study, using exclusively hospital related records. The Brazilian cohort was built using a database that coalesces patient records of several institutions throughout national territory, via selecting patient with Alzheimer diagnosis who were at least 50 years old when they received the diagnosis. The reference center cohort was built using the same criteria as the Brazilian but pulled data only from the database of a reference neurology center in São Paulo. Antipsychotic use event was defined as the first prescription of the drug, regardless of time of use or subsequent administrations. Test results were a Brazilian cohort of N=5,944, of whom 33% had been prescribed any type of antipsychotic, with 27% having received Quetiapine, 16% Haloperidol, 6% Risperidone and 4% Olanzapine. The reference center cohort had N=250 patients, whom 28% had been prescribed any type of antipsychotic, with 24% having received Quetiapine, 16% Haloperidol, 8% Risperidone and 4% Olanzapine. The main points of antipsychotics prescription in the context of any dementia are the increased mortality risk and side effects. While the prescription pattern is similar, the reference center administered antipsychotics to less patients. The most prescribed drug, Quetiapine, is an off-label prescription, as only Risperidone and Haloperidol are validated for usage in behavioral and psychological symptoms of dementia. This is possibly related to its less severe side effects compared to other antipsychotics, but it could also be a result from earlier evince of antipsychotics use where it appeared as the only drug to not increase mortality, a find contested by more recent review. An important find is the high usage of Haloperidol compared to other antipsychotics, as it is the drug most associated with increased mortality, especially when administered as injection, due to its neurotoxicity. As there is important evidence against its use in the context of dementia, and it should be viewed as a very restricted drug.

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Alzheimer's disease and Parkinson's disease: prolonged work absence in Brazilian public university employees in ten years

David Greco Varela[1,2], Antonio de Souza Andrade Filho[1,2], Frederico Luiz da Silva Figueroa[1,3], Isabel Fernandes Pereira[1], Ana Carolina Campos Carvalho[1,3], Camila Gomes de Souza Andrade[1,4], Vitoria de Araujo Carneiro Santos[1,5], Adonay Ferreira Lisboa[1,3], Vitor Raphael Heliodoro Barreto[1,3], Natalia Lino Dorea da Silva[1,3]

[1]Fundação Baiana de Neurologia e Neurocirurgia, Salvador BA, Brazil.
[2]Universidade Federal da Bahia, Salvador BA, Brazil.
[3]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[4]Universidade Salvador, Salvador BA, Brazil.
[5]União Metropolitana para o Desenvolvimento da Educação e Cultura, Lauro Freitas BA, Brazil.

Correspondence: David Greco Varela; Email: dgvarelabr@yahoo.com.br.

Background: In Brazil, around one hundred thousand new cases of Alzheimer's Disease are diagnosed each year. For Parkinson's disease, it is estimated that two hundred housand people are affected. The progressive aging of the population of public employees, associated with the change in legislation that allowed them to remain in active service until 74 years old, makes it important to know what the frequency of sickness is associated with the two most prevalent degenerative neurological diseases worldwide. **Objective:** Verify the frequency of Alzheimer's disease and Parkinson's disease associated with prolonged work absence in Brazilian public university employees in the last ten years. **Methods:** It was a cross-sectional study in which a retrospective survey was carried out, including work absences lasting fifteen days or more associated with Alzheimer's disease and Parkinson's disease, between 01/01/2014 and 12/31/2023. Gender, occupation and type of neurological disease were analyzed. The population was 5,564 public university employees, with a predominance of women (53.8%). The staff with age between 66 and 74 years old was 8.1%. **Results:** Alzheimer's diseases accounted for 0.12% of the total of 10,027 absences due to illness over the ten years. Parkinson's disease accounted for 0.18%. The frequency of absence from Alzheimer's disease in women when compared to the total number of absence by gender was 0.12% (12/7498) and 0.11 (3/2529) for men. The frequency of absence of Parkinson's disease in women when compared to the total number of absence by gender was 0.12% (12/7498) and 0.35 % (9/2529) for men. The workers most frequently affected by Parkinson's disease were teachers 55.5% (10/18) and administrative assistants 33.3% (6/18). For Alzheimer's disease, medical doctors were most affected 41.6% (5/12) and laboratory assistants 16.6% (2/12), professionals of the health sector were 66,6% (8/12). **Conclusion:** The present study found no difference in the frequency of sick leave between men and women for Alzheimer's disease. For Parkinson's disease, the frequency of absenteeism was higher in men compared to women. For Parkinson's disease, there was a predominance of sick leave for education professionals, while Alzheimer's disease was more common among health professionals.

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Amyloid angiopathy-related inflammation: more than amyloid spells

Giuseppe Dick Bonato[1], Marciéli Gerhardt[1], Luis Del Carmen Vega Gutierrez[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Giuseppe Dick Bonato; Email: g.d.bonato@hotmail.com.

Case presentation: An 83 year-old woman is brought to a tertiary care hospital after being found with sensory impairment. The previous day she showed behavior change and had inespecific complaints. She also presented an episode of possible focal seizure. At first evaluation she was brad-psychic and presented mixed aphasia. Her blood pressure was elevated, being treated with parenteral antihypertensive drugs. Family denied previous history of hypertension, and she lived alone without need for help in daily activities. She presented major depression, being treated with citalopram. Moreover, she frequently took oral analgesics due to chronic back pain. First impressions of vascular event or hypertension encephalopathy were not confirmed. Brain MRI showed signs of amyloid angiopathy-related inflammation features, such as corticosubcortical assymetrical bleeds, white matter hyperintensities and leptomeningeal enhancement. Electroencephalogram showed asymmetric frontal epileptogenic activity. Lumbar puncture revealed 106mg/dl with no pleocytosis. Extensive infectious screening showed no other plausible cause for clinical presentation other than amyloid angiopathy-related inflammation, and we proceeded to administer methylprednisolone 1g per day during 5 days followed by oral corticoids with progressive dose tapering. Before being discharged, she had considerable improvement, presenting fully oriented and alert. In the next two months, she was admitted twice to the same hospital. At first, she presented urinary tract infection complicated by delirium, being discharged at basal cognitive level after treatment. A second admission was due to right femur fracture after falling, with surgical correction. She presented vomits at 7th postoperative day with possible bronchoaspiration and consequent cardiorespiratory arrest. She did not respond to intensive care measures and passed away the following day. **Discussion:** Amyloid angiopathy-related inflammation is an important differential of several more frequent medical conditions. Its potential treatability deem importance of identifying and diagnosing this disease. The two major proposed pathophysiological mechanisms involve primary angiitis-like features, and even if they are pathologically distinct, no clinical nor radiological characteristics seems to be able to differentiate them. Treatment basis involve immunomodulation, first line being high dose IV corticosteroids with progressive oral tapering. Response to treatment may be effective in more acute symptoms, while chronic impairments may be permanent. **Final comments:** The presented case conveys an older patient presenting inflammatory features associated with vascular changes compatible with the discussed disease. Our patient presented a good outcome after first hospitalization. Complications related to advanced age and worse prognosis-associated incidents (i.e. delirium and femur fracture) lead to patient's disease.

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Amyotrophic lateral sclerosis and multiple sclerosis: a uncommon association

Jéssica Cristina Silveira Damasceno[1], Laura Altomare Fonseca Campos[1], Júlia Lima Vilela[1], Marcelo Sobrinho Mendonça[1], Thiago Cardoso Vale[1]

[1]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.

Correspondence: Jéssica Cristina Silveira Damasceno; Email: drajessicadamasceno@gmail.com.

Case presentation: A 51-year-old woman, previously hypertensive, diabetic, and with a history of depressive disorder, began experiencing progressive weakness in her limbs, predominantly in her lower limbs, in 2019, leading to loss of ambulation and wheelchair dependence in 2023. She had a family history of amyotrophic lateral sclerosis (ALS) in her father and cousins, without genetic confirmation. In June 2022, she underwent electromyography of all four limbs and thoracic paravertebral muscles, revealing active denervation in all studied segments, consistent with motor neuron disease. Genetic testing revealed a mutation in the vesicle associated membrane protein B (VAPB) gene at position CHR20:58,418,318, confirming ALS diagnosis due to VAPB gene mutation. During diagnostic investigation, brain MRI made in 2021 showed hyperintense ovoid lesions on T2/FLAIR and hypointense ones on T1, suggestive of demyelinating lesions without contrast enhancement. She had no history suggestive of clinical relapses. Cervical and thoracic spinal cord MRI also revealed demyelinating lesions without contrast enhancement. She was diagnosed with radiologically isolated syndrome (RIS) and began undergoing periodic neuro-axis MRI scans. In September 2023, she experienced subacute worsening of strength, primarily in the upper limbs, and a repeat cervical spinal cord MRI showed a lesion in the right lateral C7-T1 with hyperintensity on T2 and STIR sequences, with contrast enhancement, suggestive of inflammatory activity. Cerebrospinal fluid showed oligoclonal bands. She underwent a 5-day course of methylprednisolone pulse therapy without clinical improvement. She was subsequently diagnosed with multiple sclerosis (MS) and started treatment with glatiramer acetate. **Discussion:** Familial

forms of ALS are associated with mutations in various genes. Mutations in the VAPB gene follow an autosomal dominant inheritance pattern and typically manifest as slowly progressive weakness, fasciculations, cramps, and preferential involvement of the lower motor neurons. Despite clinical, electrophysiological, and confirmatory genetic diagnosis of ALS due to VAPB gene mutation, the patient exhibited a faster progression rate since 2019 than typically observed for the disease. However, given the subacute worsening of strength, including in the upper limbs in September 2023, corresponding to the cervical spinal cord lesion, there was a possibility of a bout of MS rather than just ALS progression. There are no reports in the literature suggesting a causal association between ALS and MS, being the occurrence of both diseases in this patient likely coincidental. **Final comments:** This case highlights a rare association between familial ALS and MS and underscores the diagnostic challenge in this association, as motor symptoms from one condition can overlap with those of the other, complicating the differentiation between RIS and MS.

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Amyotrophic lateral sclerosis associated with a rare pathogenic VCP gene variant: a case report

Yasmin Luíse Lucena e Silva[1], Samila Marissa Pinheiro Gomes[1], Aglagilson Fernandes Das Chagas[1], Moisés Vasco de Freitas[1], Emanuela Maria de Aguiar Azevedo[1], Gabriel Belo Cavalcante Linhares[1], Jonnathan Camara da Silva Vianna[1], Eduardo Martiniano de Lima[1]

[1]Universidade Federal Rural do Semi-Árido, Mossoró RN, Brazil.

Correspondence: Yasmin Luíse Lucena e Silva; Email: yasmin.silva43227@alunos.ufersa.edu.br.

Case presentation: Amyotrophic lateral sclerosis (ALS) is a motor neuron disease, affecting both upper and lower motor neurons. It is a progressive, paralytic, incurable condition of unknown etiology. Despite that, over 120 genes have been implicated in ALS. The following case report describes a patient who was diagnosed with ALS and was later found to have a rare pathogenic variant of the VCP gene. A 51-year old man, with a past medical history of lumbar hernia, acute myocardial infarction, hypertension and obesity, presented with progressive left lower limb weakness. He had no history of poliomyelitis and was initially evaluated by an orthopedist, who prescribed pregabalin. Unfortunately, the weakness progressed, later affecting the other lower limb and, eventually, both arms. The patient developed dysarthria and dysphonia. Currently bedridden, he is unable to speak and feeds via tube. Electroneuromyography, ordered about 7 months after symptom onset, was consistent with lower motor neuron disease, affecting the cervical, thoracic and lumbar regions. The condition was more evident on the left hemibody, mainly affecting the left lower limb. Split-hand index was decreased bilaterally, measuring 3.4 on the left and 5.0 on the right side. Finally, an oral swab sample was sequenced by next generation sequencing, which detected a heterozygous pathogenic variant of the VCP gene, namely NM_007126.5: c.277C> T: p. (Arg93Cys). The exact same variant was identified in close relatives, including a sibling and a cousin, who were diagnosed with slowly progressive motor neuron disease and inclusion body myositis, respectively. Furthermore, a case of Guillain-Barré syndrome and other neurological diseases with motor impairment in distant family members have been reported. **Discussion:** Pathogenic variants of the VCP gene have been linked to cases of ALS, as well as other conditions, such as inclusion body myopathy with early-onset Paget disease and frontotemporal dementia. This gene encodes the valosin-containing protein, an enzyme that mainly plays a role in protein degradation. Although ALS is a fairly common diagnosis, NM_007126.5: c.277C> T: p. (Arg93Cys) is a very rare variant of the VCP gene. It is considered a missense variant and studies indicate that it is expected to disrupt VCP protein function, but the exact mechanisms behind disease development are not fully understood. This case highlights an uncommon genetic abnormality detected in three related individuals, which is unusual considering that around 90% of ALS cases are sporadic. In addition, since these patients received different diagnoses, this report reinforces that the conditions mentioned here have an overlapping genetic basis. **Final comments:** This report features a rare genetic variant linked to ALS and other conditions. Further investigation is needed to better understand how this variant causes disease, which can aid genetic counseling and clinical management of patients.

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An atypical case of hereditary distal motor neuropathy (HDMN) associated with mutation on DCTN1 gene

Roberto Pereira Santos[1], Rhanice Soares Huguenin Câmara[2], Amanda de Morais Affonso[2], Caio Tardin[1], Matheus Jorand Cecatto[1], Ana Carolina Leite[1], Barbara Pina Aiello[1], Filipi Britto[1], Victor Evangelista Rodrigues Pereira[1], Marcos Raimundo Gomes de Freitas[1]

[1]Universidade Federal do Rio de Janeiro, Hospital Universitário Clementino Fraga Filho, Serviço de Neurologia, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Rio de Janeiro, Faculdade de Medicina, Rio de Janeiro RJ, Brazil.

Correspondence: Roberto Pereira Santos; Email: r.santos.ufrj@gmail.com.

Case presentation: A 20-year-old woman has presented with a scuffing gait since early childhood, bilateral dorsiflexion paresis worse on the left, and distal muscle atrophy in the lower limbs (LL), which has remained non-progressive. She shows no signs or symptoms of sensory, vocal, or parkinsonian issues. There were no similar cases in her family, and her asymptomatic, non-consanguineous parents were unaffected. Exon 29 of the DCTN1 gene revealed a heterozygous variant of uncertain significance (VUS) c.3502G> A (p.Val1168Ile). This gene typically follows an autosomal dominant pattern and is associated with Hereditary Distal Motor Neuropathy type 7B (dHMN7B) and Perry syndrome. Other pathogenic variants have also been linked to Amyotrophic Lateral Sclerosis (ALS). This case stands out due to its childhood onset, lack of progression, absence of vocal involvement, and negative family history of a dominant genetic variant that typically manifests in adulthood with vocal symptoms. Genetic testing of the parents will confirm the presence of this mutation. **Discussion:** Pathogenic variants in the DCTN1 gene typically exhibit an autosomal dominant inheritance pattern and are associated with Hereditary Distal Motor Neuropathy type 7B (OMIM#607641) and Perry syndrome (OMIM #168605). Some variants are also implicated in Amyotrophic Lateral Sclerosis (ALS), with both dominant and recessive inheritance patterns observed. Hereditary Distal Motor Neuropathy (dHMN) primarily affects distal motor function in the lower limbs, progressing to involve proximal limb regions and hands. Some variants may exhibit subclinical sensory and pyramidal signs, leading to differential diagnoses such as ALS, Spinocerebellar Ataxia, Hereditary Spastic Paraplegia, axonal Charcot-Marie-Tooth disease, and distal myopathies. A subgroup known as NMDh-plus may present additional features including bradykinesia, rigidity, retinitis pigmentosa, cognitive delay, hyperCKemia, and joint deformities. While mutations in over 30 genes have been linked to NMDh, identification of the specific pathogenic mutation occurs in fewer than half of cases (WU, 2022). The classification of NMDh variants and their related genes is detailed in Table 1, adapted from Zhang et al. (ZHANG, 2021). The dHMN7 subtype, encompassing types 7A and 7B, typically manifests in adulthood with vocal cord paralysis and follows an autosomal dominant inheritance pattern (ZHANG, 2021). **Final comments:** This case represents an unusual presentation of NMDh associated with a VUS in the DCTN1 gene. Further research is needed to elucidate the functional impact of this variant and its role in the pathogenesis of neurological disorders.

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An atypical presentation of neuroleptospirosis: a case report and literature review

Ana Letícia Amorim de Albuquerque[1], Monique Blauth Tovo[1], Rhaná Carolina Santos[1], Giovanna Polo[1], Lorenna Lima Regueira Costa[1], Matheus Zschornack Strelow[1], Matheus Bernardon Morillos[1], Diógenes Guimarães Zan[1]

[1]Universidade do Vale do Rio dos Sinos, São Leopoldo RS, Brazil.

Correspondence: Ana Letícia Amorim de Albuquerque; Email: analeticia.neurologia@gmail.com.

Case presentation: Male, 43 years old, no known comorbidities. Presented with progressive caudal-cranial paresis in lower limbs. A week prior, he experienced acute urinary retention and episodes of intermittent fever. Neurological exam showed drowsiness, hypophonia, paraplegia associated with reduced strength in upper limb and cervical musculature. Tactile sensitivity was absent up to the T4 level, along with loss of sphincter control. Reflexes were increased in the upper limbs and absent in the lower limbs. Initial investigations revealed elevated hepatic and canalicular enzymes, elevated lactate dehydrogenase, and signs of systemic inflammation. Cerebrospinal fluid (CSF) had a total protein of 109 mg/dL, the ratio of CSF/Serum glucose was 0.86 and 640 cells/mm3, 80% lymphocytes. Magnetic resonance imaging revealed foci of hypersignal on T2 and subcortical FLAIR in the brainstem, middle cerebellar peduncles and at the level of the thalamus and basal nuclei with extension to the corona radiata, bilaterally, with minimal restriction in diffusion. MRI assessment of the cervical spine showed longitudinally extensive myelitis, with an incipient degenerative disc disease at C4-C5, C5-C6 and C6-C7, and T2 hyperintense lesion diffusely compromising the cervical spinal cord. The patient was initially treated with ceftriaxone and ampicillin for suspected CNS bacterial infection, and dexamethasone to reduce cerebral inflammation. A new detail emerged: the patient had contact with a dead rat without protective gear a week before symptom onset. Leptospirosis was confirmed by a positive serum IgM-ELISA. Clinical improvement was noted by day 9 of therapy as the patient was already receiving empirical ceftriaxone. He remained with lower limb plegia. The patient was discharged with physiotherapy and follow-up with neurology. **Discussion:** Leptospirosis is prevalent in tropical countries, such as Brazil, having episodic outbreaks after torrential rains and flooding, especially in precarious sanitary situations. Human contraction typically arises from exposure to contaminated soil, water and infected animals. Rodents are primary carriers. Humans may contract the infection through cuts and mucous membranes. Neurological involvement can lead to vasculitis, microcirculation and endothelial dysfunction, developing patchy demyelination in the CNS. Neurological manifestations occur in 15% of cases and can be manifested as encephalitis. This case underscores the importance of considering leptospirosis in the differential diagnosis of acute neurological syndromes, particularly in regions where the disease is endemic. **Final comments:** The case emphasizes the necessity for prompt recognition and treatment of leptospirosis to mitigate severe complications. Increased awareness and early intervention are paramount in managing such infections, particularly in tropical regions with poor sanitation and frequent rodent exposure.

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An atypical presentation of stiff person spectrum disorder: a case report

Catarina Sodré de Castro Prado[1], Luiza de Aguiar Lima[1], Márcia Cristina Antunes Ribas[1], Gabriel R. de Freitas[1], Camila Pupe[1], Marco Antonio Araujo-leite[1], Caroline Bittar Braune[1]

[1]Universidade Federal Fluminense – Ebserh, Hospital Universitário Antonio Pedro, Departamento de Neurologia, Niterói RJ, Brazil.

Correspondence: Luiza de Aguiar Lima; Email: luizadealima@gmail.com.

Case presentation: A 25-year-old female presented with difficulty walking and dysarthria. After 4 months she developed dysphagia, convergent strabismus, urinary retention and vertigo. She had the previous diagnosis of congenital nephrogenic diabetes insipidus, Hashimotos disease and lichen planus. In the hospital the patient began with painful muscle spasms in the upper and lower limbs, along with tachycardia and profuse sweating, with no loss of consciousness, and a good response to treatment with benzodiazepines, baclofen and opioids. On neurological examination she exhibited involuntary contractions of the orbicularis oris muscle, spasticity and hyperreflexia of the upper and lower limbs, global reduction of strength with sarcopenia, convergent strabismus with bilateral lateral rectus paresis, worse on the right eye, and tongue paralysis. Neuroaxis magnetic resonance imaging showed no alterations that could justify her symptoms. The cerebrospinal fluid analysis was normal, including bacterial, viral and fungal panels and oncotic cytology, as was her serum screening for HIV, syphilis, toxoplasmosis, cytomegalovirus and hepatitis B and C. The antinuclear antibody (1:80) was positive with a fine speckled pattern. She deteriorated with respiratory failure and needed to undergo an orotracheal intubation. Her muscle weakness and ophthalmoplegia also progressed, and the patient could not lift her limbs against gravity or move her eyes. She remained oriented, with no alterations of cognition or behavior. She evolved with anisocoria, with her left pupil not reacting to light. Electromyography showed a sustained contraction of both agonist and antagonist musculature (biceps and triceps) simultaneously. The Anti-GAD-65 Antibody was requested and came back with a high titer of 29.541 UI/ml. The diagnosis of Stiff Person Spectrum Disorder (SPSD) was established and treatment with intravenous immunoglobulin was initiated with 0,4 grams per kilogram of weight during 5 days. After two weeks the patient began to improve, being able to stick out her tongue, move her head and eyes, remaining only with paresis of the right lateral rectus. Her right pupil also started responding to light and she was able to raise her lower limbs against gravity. **Discussion:** The diagnosis of SPSD was suspected based on the clinical scenario of painful muscle spasms, which became increasingly frequent, the good response to benzodiazepines and baclofen, the rigidity of extremities, the involvement of multiple cranial nerves bilaterally, and the personal history of autoimmune disease. It was further supported by electromyography findings, high titer of Anti-GAD-65 and the marked improvement after immunoglobulin. **Final comments:** This case highlights the importance to bear in mind the different clinical presentations of SPSD, and so to maintain a low threshold of suspicion for this treatable condition.

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An epidemiological analysis of Guillain-Barré syndrome in the neurology service of the UNIMED BH hospital

Ana Luiza de Oliveira Batista Passos[1], Andréa Jerusa de Souza Santos[1], Glaucia Lara Resende[1]

[1]UNIMED, Belo Horizonte MG, Brazil.

Correspondence: Ana Luiza de Oliveira Batista Passos; Email: anaobps@hotmail.com.

Guillain-Barré Syndrome (GBS) is a rare immune-mediated condition that affects the peripheral nervous system and is characterized by an ascending flaccid paralysis, however, it sometimes presents in an atypical form with clinical variants. It has a diagnosis based on clinical, CSF and electrophysiological criteria. In this series of cases, we portray patients admitted to the Neurology service of the Unimed Hospital of the Contorno Unit (Belo Horizonte/MG) with a diagnosis of Guillain-Barré Syndrome between April 2022 and June 2024. 29 patients were included, 66% male and 34% female. The average age was 40.3 years (range 15 - 77 years). Of these, 27% had a respiratory or gastrointestinal infection preceding the condition; 38% had been vaccinated in the last 4 weeks and 80% of patients who had an infectious trigger in the gastrointestinal tract required transfer to the ICU. The months with the highest incidence of cases were December 2022 and November 2023. The majority received treatment with human intravenous immunoglobulin (86%) and none received treatment with plasmapheresis. Half had an Erasmus GBS Respiratory Insufficiency Score (EGRIS) ≤ 3 points and the other half EGRIS ≥ 4. Three patients developed respiratory failure in the first week after the onset of symptoms. Of those with EGRIS ≤ 3, 30% went to the ICU and did not require invasive ventilation. In relation to those with EGRIS ≥ 4, 70% went to the ICU, and of these 28% required invasive ventilation. There was 1 death. Our data and results presented are compatible with those described in the literature regarding triggers for autoimmunity and prognoses. We question whether the coronavirus pandemic could be an aggravating factor for the increase in the incidence of Guillain Barré Syndrome.

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An epidemiological analysis of hospitalizations due to epilepsy in Brazil from 2013 to 2023

Matheus Henrique Brustolim Marcucci[1], Guilherme Ângelo Braga de Sousa[1], Leslie Mateus de Sousa Nunes[1], Matheus Sant'anna de Oliveira Gonçalves[1], Hurik Pontes Rocha[1], Paulo Wendel Moura de Souza[1], Leonardo Elias Araujo[1], Francisco Luciano Honório Barreto Cavalcante[1], Letícia da Costa Rios[1], Letícia Gadelha Caldas[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Matheus Henrique Brustolim Marcucci; Email: matheus.marcucci@alu.ufc.br.

Background: A temporary and reversible change in brain function, not caused by fever, drugs, or metabolic disorders, manifested by repeated epileptic seizures. The origins of seizures are often unidentified (idiopathic), but they can be related to various brain problems, such as structural abnormalities, strokes, or tumors. **Objective:** To analyze the epidemiological profile of epilepsy hospitalization cases in Brazil between 2013 and 2023. **Methods:** This study is an epidemiological investigation of confirmed epilepsy hospitalization cases between 2013 and 2023. Data was collected from the Sistema de Informação de Agravos de Notificação (Sinan Net) of the Departamento de Informática do Sistema Único de Saúde (DATASUS), and data analysis was performed using Jamovi® software. **Results:** After data analysis, 592,028 epilepsy cases were found in Brazil between 2013 and 2023. The highest numbers were in the Southeast with 249,189 cases (42.09%) and the Northeast with 138,242 cases (23.35%), followed by the South, Midwest, and North with 20.75%, 8.29%, and 5.52% respectively. Regarding temporal analysis, an average of 53,820.72 (±5196.94) cases per year was found, with a constant growth in diagnoses from 2013 to 2019, a turning point reducing the number of diagnoses in 2020, followed by another growth period until 2023. Of the observed cases, 57.90% of patients were male and 42.10% were female, with the predominant age group being 1-4 years old (16.68%), followed by 40-49 years old (10.14%) and 50-59 years old (10.13%). The most prevalent race was mixed race with 39% of the cases, and the least prevalent was indigenous with 0.16% of diagnoses. **Conclusion:** According to the data analysis, a concentration of cases was identified in the Southeast (42.09%), the Northeast (23.35%), and the South (20.75%), possibly due to the population concentration in these regions of Brazil. A possible underreporting of cases was found in 2020, likely caused by the COVID-19 pandemic, during which people avoided leaving their homes, making diagnoses more difficult. Regarding the patient profile, there was a prevalence of male individuals (57.90%) and patients aged 1-4 years (16.68%). This can be explained by the fact that children's brains are still developing, making them more susceptible to congenital or genetic anomalies. As for race, a low diagnosis rate was found in the indigenous population, which may be caused by both the concentration of this population in the country and the underreporting of cases in this population.

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An epidemiological analysis of hospitalizations due to transient ischemic attack and non-specific strokes (hemorrhagic or ischemic) in Ceará from 2013 to 2023

Matheus Henrique Brustolim Marcucci[1], Guilherme Ângelo Braga de Sousa[1], Hurik Pontes Rocha[1], Paulo Wendel Moura de Souza[1], Matheus Sant'anna de Oliveira Gonçalves[1], Leonardo José Rodrigues de Araújo Melo[1], Cássia Caroline Aguiar da Ponte[1], Paulo Victor de Santiago Gonçalves[1], Oliver Reiks Miyajima[2], Júlia Viana de Castro[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Matheus Henrique Brustolim Marcucci; Email: matheus.marcucci@alu.ufc.br.

Background: Non-Specific Stroke (NS-Stroke) is brain damage causing a sudden, focal, neurological deficit from a vascular disease, lasting over 24 hours. In contrast, a Transient Ischemic Attack (TIA) is a temporary, reversible neurological deficit, usually lasting less than an hour, caused by a brief vascular blockage in the brain. **Objective:** To analyze the epidemiological profile of hospitalization cases of NS-Stroke and TIA in Ceará between 2013 and 2023. **Methods:** This study is an epidemiological investigation of the hospitalization of NS-Stroke and TIA cases between 2013 and 2023. Data were collected from the Sistema de Informação de Agravos de Notificação (Sinan Net) do Departamento de Informática do Sistema Único de Saúde (DATASUS), and data analysis was performed using Jamovi®. **Results:** According to the data found in DATASUS, between 2013 and 2023, approximately 74,400 cases of NS-Stroke were recorded in Ceará. A temporal analysis of NS-Stroke cases showed an average of 6,763.6 (±471.28) cases/year, with periods of increase (2013-2016, 2017-2019, 2020-2021 and 2022-2023) and years of decline (2017, 2020, 2022). Among the NS-Stroke diagnoses, 53.08% were male and 46.92% were female. Additionally, 60.67% of cases were from the metropolitan regions of Fortaleza and Cariri, while 39.32% were from outside the metropolitan region. The most prevalent race was mixed race at 70.36%, and the second least prevalent was black at 0.96%. Regarding TIA, during the analyzed period (2013-2023), 3,735 cases were recorded, with an average of 339.54 (±59.21) cases/year, with periods of increase (2013-2016, 2018-2019 and 2020-2023) and years of decline (2017, 2018, 2020). Among the TIA diagnoses, 51.91% were male and 48.09% were female. Moreover, 68.30% of cases were from the metropolitan regions of Fortaleza and Cariri, while

31.70% were from outside the metropolitan region. The most prevalent race was mixed race at 76.33%, and the least prevalent was black at 1.74%. **Conclusion:** According to the data analysis, significant differences were found in the reporting of Stroke cases, with NS-Stroke (74.400 cases) having a higher number than TIA (3,737 cases). This difference can possibly be explained by the underreporting of TIA cases, since, despite having symptoms similar to NS-Stroke, the rapid disappearance of symptoms often leads to not seeking medical attention. Additionally, there is the difficulty of recognizing symptoms by untrained individuals. In both cases, no significant gender differences were found. Higher hospitalization were found in metropolitan regions, both in NS-Stroke (60.67%) and TIA (68.30%), likely due to more hospitals and support networks for recognizing and treating these diseases in metropolitan areas. Regarding races, there was severe underreporting of cases in the black population (0.96% in NS-Stroke and 1.74% in TIA). This could be caused either by a lack of case reporting in this population or by a racial identification error.

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An epidemiological analysis of hospitalizations due to viral encephalitis in Ceará from 2013-2023

Matheus Henrique Brustolim Marcucci[1], Oliver Reiks Miyajima[2], Guilherme ângelo Braga de Sousa[1], Matheus Sant'anna de Oliveira Gonçalves Matheus Gonçalves[1], Hurik Pontes Rocha[1], Paulo Wendel Moura de Souza[1], Lucas Soares Radtke[1], Emanuel de Assis Bertulino Martins Gomes[1], Pedro Vitor Ferreira Rodrigues[1], Sarah Braga de Castro[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Matheus Henrique Brustolim Marcucci; Email: matheus.marcucci@alu.ufc.br.

Background: Viral encephalitis is the most common type of encephalitis, characterized by inflammation of the brain caused when viruses like herpes simplex, herpes zoster, cytomegalovirus, or West Nile virus directly infect the brain. Alternatively, inflammation can be triggered by agents such as viruses, vaccines, or other stimuli. This inflammation can manifest with clinical symptoms including fever, headache, personality changes or confusion, seizures, paralysis or numbness, and drowsiness, which may progress to coma or death. **Objective:** To analyze the epidemiological profile of hospitalization cases of viral encephalitis in Ceará between 2013 and 2023. **Methods:** This study is an epidemiological investigation of hospitalization cases of viral encephalitis between 2013 and 2023. Data were collected from the Sistema de Informação de Agravos de Notificação (Sinan Net) of the Departamento de Informática do Sistema Único de Saúde (DATASUS), and data analysis was performed using Jamovi® software. **Results:** According to data analysis from 2013 to 2023, 863 cases of Viral Encephalitis were found in Ceará. In terms of temporal analysis, an average of 78.45 (±14.58) cases per year were identified. Regarding patient demographics, 57.24% were male and 42.76% were female. Moreover, 71.96% of cases were from metropolitan areas of Fortaleza and Cariri, while 28.04% were from non-metropolitan regions, predominantly affecting the age groups of 1-4 years (17.72%) and 5-9 years (12.97%). In terms of racial profile of hospitalizations, individuals of mixed race (parda) accounted for 761 (84.70%) of the hospitalizations, while individuals of black race accounted for only 1 (0.11%) case. **Conclusion:** According to the data analysis from 2013 to 2023 in Ceará, there was little variation in the number of hospitalizations per year, as evidenced by the low standard deviation (14.58), which was not significantly affected by the COVID-19 pandemic period. There was a slight predominance of male individuals (57.24%) and of individuals residing in metropolitan areas (71.96%), possibly influenced by factors such as population density and better access to healthcare systems, facilitating diagnosis. Regarding age groups, a higher number of cases were found among individuals aged 1 to 9 years (30.69%), possibly explained by children's naturally lower immunity and increased exposure to pathogens during activities such as play and school interactions. The racial profile indicates a potential underreporting of hospitalizations among black individuals or a misinterpretation of racial classification.

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An epidemiological analysis of hospitalizations due to viral meningitis in Ceará from 2013-2023

Matheus Henrique Brustolim Marcucci[1], Lucas Frigo[1], Cynthia Paz Rodrigues[1], Ana Cecília Neves Rios Parente[1], Oliver Reiks Miyajima[2], Ana Luiza Souza Santos[1], Paulo Wendel Moura de Souza[1], Hurik Pontes Rocha[1], Matheus Sant'anna de Oliveira Gonçalves[1], Guilherme Ângelo Braga de Sousa[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Matheus Henrique Brustolim Marcucci; Email: matheus.marcucci@alu.ufc.br.

Background: Viral meningitis is an inflammation caused by a virus in the protective layers of the brain and spinal cord, the meninges, and the subarachnoid space (the space between the pia mater and arachnoid mater filled with cerebrospinal fluid). The condition includes symptoms similar to bacterial meningitis but milder, such as fever, headache, neck stiffness, nausea, vomiting, loss of appetite, irritability, drowsiness or difficulty waking up, lethargy, and photophobia. **Objective:** To analyze the epidemiological profile of hospitalziation cases due to viral meningitis between the years 2013 and 2023. **Methods:** This work is an epidemiological investigation study of confirmed hospitalization cases of viral meningitis between 2013 and 2023. Data were collected from the Sistema de Informação de Agravos de Notificação (Sinan Net) of the Departamento de Informática do Sistema Único de Saúde (DATASUS), and data analysis was performed using Jamovi® software. **Results:** The following data were found in DATASUS between 2013 and 2023: 2,776 diagnoses of meningitis in the state of Ceará, 909 of which were viral meningitis (32.74% of cases). The temporal analysis found an average of 82.63 (±26.79) annual cases of viral meningitis, maintaining an average of 90.42 cases/year from 2013-2019, which drastically reduced to 54 cases/year between 2020-2022, and increased again to 104 cases in 2023. Among the diagnoses, approximately 58.86% were male and 41.14% were female, 77.88% were from the metropolitan regions of Fortaleza or Cariri, and 22.12% were from non-metropolitan regions. The most affected age group was children aged 1-4 years, with 15.40% of the cases. The most prevalent race was mixed race with 801 cases (88.11%), and the least prevalent was black with 4 cases (0.41%). **Conclusion:** According to the analysis of the aforementioned data, a reduction in the number of cases was identified during the period from 2020-2022. This underreporting is possibly a result of the COVID-19 pandemic, during which people, fearing the virus, stayed indoors and away from hospitals, thus hindering diagnosis. A higher prevalence was found in males (58.86%) and in people from metropolitan regions (77.88%), which can possibly be explained by the greater number of hospitals and support networks for diagnosis. The age group most affected by viral meningitis is around 1-4 years, which may be due to individuals of this age having naturally weaker immune systems than adults and being more exposed to pathogens through play. Regarding races, the black race is drastically less reported than others, which may indicate underreporting or even a misidentification error.

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An epidemiological analysis of incidence of brain cancer in brazil from 2013 to 2023

Paulo Victor de Santiago Gonçalves[1], Leonardo José Rodrigues de Araújo Melo[1], Pedro Vitor Ferreira Rodrigues[1], Victor Domingueti Vallim Fonseca[1], Cynthia Paz Rodrigues[1], Francisco Luciano Honório Barreto Cavalcante[1], Cássia Caroline Aguiar da Ponte[1], Paulo Wendel Moura de Souza[1], Letícia da Costa Rios[1], Leonardo Elias Araújo[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Paulo Victor de Santiago Gonçalves; Email: paulo.santiago6571@gmail.com.

Background: Brain neoplasms are serious clinical conditions with a high mortality rate. Tumors affecting the central nervous system are often of metastatic origin, meaning they initially arise in other parts of the body and later spread to the brain. **Objective:** To analyze the epidemiological profile of confirmed cases of brain neoplasms between the years 2013 and 2023. **Methods:** This study is an epidemiological investigation of confirmed brain tumor cases between 2013 and 2023. Data were collected from the DATASUS Notification of Diseases Information System, and data analysis was performed using Jamovi software. **Results:** According to the data from DATASUS, between 2013 and 2023, approximately 158.313 cases of malignant brain neoplasms were registered. Of these, 43.5% were recorded in the southeast region, 23.23% in the south, 20.8% in the northeast, 8.35% in the central-west, and 4.08% in the northern region of the country. Among the total cases, 47.75% of patients were female, and 52.24% were male. Regarding patients' race, approximately 3.15% of cases were in Black individuals, while 78.79% were in White and mixed-race individuals. The annual average of cases during the analyzed period was 14.392 (± 1019.492). **Conclusion:** Based on the collected data, a prevalence of cases was identified in the southeast region (43.5%), which can be attributed to the higher population density in that area. No significant difference in the number of cases was observed between genders. However, a limitation of this study was the low incidence observed in Black individuals (3.15%), possibly due to severe underreporting of neoplasm cases in this population or errors in racial identification.

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An epidemiological analysis of viral encephalitis in children in Northeast Brazil between 2018 and 2023

Maria Cecília Leal Menezes[1], Luana Cristina de Oliveira Lima[1], Arthur Vinícius Feitosa Santos[1], Sabrina Sthéphanie Ataíde Martins[1], Vagner Costa da Cunha Júnior[1], Thiago Pariz da Silva Dumas[1], Arthur Aragão Nunes[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Maria Cecília Leal Menezes; Email:

Background: Viral Encephalitis (VE) is an inflammatory process that occurs in the brain parenchyma. It can result in permanent neurological sequelae and high morbidity rates, especially in children, although it can affect people of all ages. Therefore, a better understanding of its epidemiology can contribute to the construction of prevention and control strategies. **Objective:** To characterize the profile, according to epidemiological indicators, of hospitalizations due to VE in the Northeast of Brazil in the period from 2018 to 2023. **Methods:** This is a retrospective study with a quantitative approach, conducted based on an analysis of the secondary database from the SUS Hospital Information System (SIH/SUS) and the

Brazilian Institute of Geography and Statistics (IBGE), in the period between January 2018 and December 2023, regarding admissions due to VE in the Northeast region. Among the risk factors analyzed are age group, federative unit and color/ethnicity. As for the research numbers, they were organized in a spreadsheet in the Microsoft Excel program. **Results:** The sample studied consisted of 4,813 hospitalizations for viral encephalitis. The prevalence was in children aged 1 to 4 years, representing 15.41% of the total, followed by those aged 5 to 9 years (11.13%). In contrast, the lowest occurrence was among elderly people over 80 years old, corresponding to 2.28%. Among Brazilian states, Pernambuco recorded the highest number of cases (1,894 patients) and the highest prevalence per 100,000 inhabitants (20.9), with emphasis on hospitalizations among children aged 1 to 4 years. On the other hand, Alagoas had the lowest number of admissions (101), as well as the lowest rate per 100,000 inhabitants (3.2). Furthermore, viral encephalitis was more common among mixed race people, with 3,462 cases (71.93%), and less common among indigenous people, with only 4 cases (0.08%). **Conclusion:** The present study showed a higher occurrence of hospitalizations for VE in children aged 1 to 9 years in Northeast Brazil, possibly due to the immature immune system at this age, making them more susceptible to the disease. Moreover, there was an increase in admissions in the brown population and a drop in the indigenous population, suggesting a possible relationship with the majority number of brown people in the Northeast and the smaller number of indigenous people. Thus, racial, geographic and age factors were suggested in order to justify the distribution of viral encephalitis in the Northeast so that effective public policies can be created aimed at preventing the pathology, ensuring better control of it.

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An extradural hematoma case report of the posterior fossa treated conservatively

Maria Cândida Barros Arantes Romano[1], João Lazaro da Silva Rodriguez Oquendo[1], Júlia Oliveira e Freitas[1], Fábio Serra Barbosa da Silva[2]

[1]Centro Universitário Tocantinense Presidente Antônio Carlos, Araguaína TO, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Cândida Barros Arantes Romano; Email: candinha_barros-ar@hotmail.com.

This article presents a case of posterior fossa epidural hematoma, of considerable volume, with non-specific symptoms and no signs of brain herniation or meningeal irritation, treated conservatively, without surgery, where there was good outcome, and discusses its pathophysiology, radiological findings, alternative treatments and their indications as well as the evolution of this patient. Epidural hematomas after trauma to the posterior fossa are very rare, representing only 0.3% of head injuries. The majority of extradural hematomas in the fossa posterior are venous, occurring based on fractures of the occipital bone with following injury to the sigmoid bone or transverse sinus. Venous bleeding may also occur as a result of arachnoid granulation or emissary veins rupturing. Due to its mild symptoms, and it being essential to perform a computed tomography in any suspected case. Early recognition is important so that you can reduce the time of brain herniation and thus reduce the morbidity and sequelae that the injury can cause.

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An ominous complication: explosive mucormycosis initially mimicking optic neuritis

Eduarda Rosa Fernandes[1], Victor Garcia Ortiz[1], Giussepe Dick Bonato[1], Marco Antonio Delazeri[1], Carolina Heck Haubrich[1], Guilherme Vanik[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Victor Alfonso Garcia Ortiz; Email: victorgarcia.or@gmail.com.

Case presentation: A 36 year-old male was transferred to our tertiary care hospital due to an extensive brain tumor. Two weeks prior to admission, he presented headache with right ocular pain and progressive vision loss. He was admitted to a secondary care facility with presumed optic neuritis. As previous medical history, he presented type 1 diabetes, hypertension and terminal chronic kidney disease. At the time, brain CT showed no lesions although angiography suggested a possible vasculitic process in the anterior circulation. He then received daily pulsetherapy with IV methylprednisolone 1g. On the sixth day of therapy, he progressed with consciousness impairment with a new brain CT showing extensive tumoral lesion at the right frontoparietal region and deep nuclei, as well as transtentorial and subfalcine herniations with diffuse cranial sinuses thickening. MRI further charactherized the lesions with extensive supra and infratentorial edema and water diffusion restriction in the anterior cerebral artery on the right. The patient underwent urgent decompression craniotomy but suffered progressive clinical deterioration. Rhinoscopy showed extensive inflammation of facial sinuses with infiltrative lesions. He receive empirically ceftriaxone, meropenem, vancomycin, anfotericin B, dexamethasone and acyclovir. Brain biopsy suggested infection by Rhyzopus spp. and extensive necrosis and inflammation, confirming mucormycosis. Despite all treatment efforts, he evolved with untreatable intracranial hypertension, evolving to brain death days after the procedure. **Discussion:** Mucormycosis is a rare fungal infection with high mortality and morbidity. It affects immunocompromised patients, but presents a high predilection for those with poorly-controled diabetes. Clinical syndromes englobe pulmonar and rino-orbit-cerebral mucormycosis. **Final comments:** We present a diabetic dialysis patient with invasive mucomycoses who underwent immunossupressant therapy due to presumed optic neuritis with ominous results. We highlight the imperative exclusion of infectious etiologies prior to starting immunossupressant therapy, particularly with possible previous immunosupression, and the need for close monitoring for neurological worsening during treatment. Mucormycosis is a serious and often unrecognized disease that demands prompt multidisciplinary treatment.

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An overview of multiple sclerosis associated comorbidities

Caio Hussid de Góes[1], Gabriella Chinaglia Zanin[1], Diego Olympio Peixoto Lopes[2], Maria Tereza Martinez[3], Fernanda Herculano[4], Diogo Haddad Santos[4]

[1]Faculdade de Ciências Médicas da Santa Casa de São Paulo, São Paulo SP, Brazil.
[2]Universidade Santo Amaro, São Paulo SP, Brazil.
[3]CPQuali Pesquisa Clínica, São Paulo SP, Brazil.
[4]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Caio Hussid de Góes; Email: caio.goes@aluno.fcmsantacasasp.edu.br.

Multiple sclerosis is an inflammatory, autoimmune demyelinating disease of the central nervous system. Much like most chronic diseases, patients living with Multiple Sclerosis tend to develop associated comorbidities, resulting in decreased quality of life and increased economic burden of disease. The study's objective is to analyze the most common diagnosis received by Multiple Sclerosis patients before the initial Multiple Sclerosis diagnosis, as well as within 10 years of the diagnosis, without actively seeking the comorbidities. An electronic medical record aggregator called TriNetX was used to anonymously obtain the patient data used in the study, using exclusively hospital related records. The cohort was built using a Brazilian-only database via selecting patients who received the Multiple Sclerosis diagnosis in the last 10 years time frame, had never received a Multiple Sclerosis diagnosis before, and were at least 18 years at the time of the diagnosis. The associated diagnosis were gathered as the first instance per patient, regardless of subsequent frequency. Test results were a cohort of N=3,093 patients, with age at index of 40.2 ± 13.7 years and current age of 43.8 ± 13.8 years. Pre-Multiple Sclerosis most common comorbidity reported was headache at 8% of patients, followed by anxiety at 2%, fatigue at 2%, visual disturbances at 1%, and diabetes mellitus and hypothyroidism at less than 1%. Post Multiple Sclerosis diagnosis common comorbidity reported was headache at 10% of patients, followed by anxiety at 3%, fatigue at 2%, visual disturbances at 1%, and diabetes mellitus and hypothyroidism at 1%. Coexisting diseases increase the complexity of patient management, particularly with the increasing number of immunosuppressive disease-modifying therapies and the subsequent overlapping of their side-effects and comorbidity symptoms. The results obtained related to anxiety and depression post Multiple Sclerosis diagnosis were lower than the expected prevalence between 20 to 40% of patients. As most studies investigating these comorbidities actively seek them in patients via questionnaires, it is likely that spontaneous referral from the patient of symptoms typical of them occurs at a lower rate. Also, the prevalence of co-autoimmune diseases post Multiple Sclerosis diagnosis, hypothyroidism and diabetes mellitus, were lower than the expected. Hypothyroidism prevalence is reported as close to 20%, though it typically is subclinical in Multiple Sclerosis context, being diagnosed based on serum antibodies, which were not investigated in this research. Expected diabetes mellitus prevalence is between 5 and 7%, a discrepancy that may be a result from the age bias of the cohort, as most studies focusing on Multiple Sclerosis and Diabetes explore populations above 45 to 50 years. Thus, it can be concluded that comorbidities diagnosis in Multiple Sclerosis require patient referred symptoms, but also active seeking from the attending clinician.

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An uncommon neurological presentation of dengue fever: spinal subdural hematoma

Maria Paula Oiticica de Jesus[1], Giovana Barros e Silva Ribeiro[1], Alberto Rolim Muro Martinez[1], João Luiz Mourão[1], Nivaldo Adolfo da Silva Junior[1], Marcondes Cavalcante França Junior[1], Raissa Gomes Castro e Souza[1], Rafael Saliba Helmer[1], Verena Miranda de Souza[1], Kaio Cezar Ferreira[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Maria Paula Oiticica de Jesus; Email: mpaulaoiticicaa@hotmail.com.

Case presentation: A 36-year-old woman with a previous history of cirrhosis due to an autoimmune hepatitis presented with insidious onset of back pain associated with limitating headache and nausea. A rapidly progressive symmetric paraparesis with lower limb areflexia and urinary retention took place two weeks later leading the patient to seek for medical attention. Laboratory workup disclosed thrombocytopenia (38,000) and mildly elevated INR (1.4). Initial cranial and thoracolumbar spine CT scans were unremarkable. Lumbar puncture revealed markedly elevated protein levels (> 1000 mg/dL) in the absence of pleocytosis. Initially suspected as Guillain-Barré syndrome, the patient did not respond to immunoglobulin therapy. Subsequent serological testing revealed positive IgM and IgG for dengue fever. The

patient's condition deteriorated with headache worsening and onset of tonic-clonic seizures. A new cranial CT scan revealed non-communicating hydrocephalus, necessitating external lumbar drainage (ELD). A subsequent MRI of the brain revealed leptomeningitis and arachnoiditis affecting basal cisterns, posterior fossa, and cervical spine segments. The patient progressed to flaccid tetraparesis and areflexia. Neuroaxis MRI was performed, revealing extensive heterogeneous collections occupying a significant portion of the intradural cerebrospinal fluid spaces within the vertebral canal, more pronounced at thoracic and lumbosacral levels. Signal characteristics suggest a likely subacute hemorrhagic nature. Notably, there was compressive effect on the spinal cord, most pronounced between T1 and T4 levels, with moderate narrowing of the cord caliber and extensive signal alteration spanning virtually the entire cervical cord and extending into the medulla, consistent with compressive myelopathy. **Discussion:** The patient, who was thrombocytopenic due to an underlying disease, along with dengue fever, experienced spontaneous spinal subdural hematoma complicated by hydrocephalus leading to severe hyperproteinorrachia due to the obstruction of CSF flow. The clinical course was further complicated by leptomeningitis and arachnoiditis, which may have arisen from local irritation secondary to hemorrhage or, considering the patient's immunosuppressed state, an infectious context (although a specific pathogen was not identified despite an extensive pathogens screening and lack of response to empirical treatment employed). **Final comments:** This case exemplifies a complex diagnostic challenge in a patient with autoimmune hepatitis-related cirrhosis and concurrent dengue infection, complicated by severe neurological manifestations. The diagnostic journey was marked by extensive imaging and laboratory investigations. This underscores the need for heightened vigilance and tailored therapeutic strategies in patients with overlapping immunosuppressive conditions and infectious complications.

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An under-recognized case of mioclonic epilepsy: NUS1

Ruan Gambardella Rosalina de Azevedo[1], Adriel Rêgo Barbosa[1], Andre Felipe Candêas Amorim[1], Gabriel Pinheiro Martins de Almeida e Souza[1], João Victor Cabral Correia Férrer[1], Márcio Pinheiro Lima[1], Pedro Henrique Almeida Fraiman[1], Rafael Tuzino Leite Neves Maffei[1], Victor Cardoso de Faria[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo/SP;

Correspondence: Ruan Gambardella Rosalina de Azevedo; Email: ruangambardella@gmail.com.

This case reports a 13-year-old female of Japanese descent with distal tremors since she was seven months old. Her mother experienced an uneventful pregnancy, with eutocic vaginal labor. The patient's milestones of motor development were appropriate for the age, but there was speech delay and worsening of tremors with aging that made it difficult to perform some manual tasks. The parents informed learning disability since kindergarten, without social impairment. Physical examination was marked by: preserved language, slowed speech, dysmetria worse in the upper limbs, appendicular myoclonus that hindered walking, normoactive reflexes. There were no signs of: ocular motricity impairment, dystonia, parkinsonism. The neuropsychological test performed at the age of eight described mild intellectual disability and impairment of all skills assessed, including motor and visual skills. Laboratory tests resulted normal for renal, thyroid and liver function. There were normal blood levels of ammonia, lactate, total cholesterol, acid-base balance, and inflammatory markers. Brain magnetic resonance imaging was negative for malformations, anomalous enhancements and atrophies. Electroencephalogram revealed rare generalized interictal epileptiform activity of anterior predominance. Exome performed at nine years-old resulted on heterozygous NUS1 mutation, with 1.5mb deletion on the long arm of chromosome 6. This genetic testing was compatible with the hypothesis of Myoclonus, epilepsy, ataxia, intellectual disability syndrome (MEAID). This is a rare disease, often with de-novo occurrence, and early age onset (from a few months to 13 years). The phenotype is characterized by various combinations of epilepsy, intellectual disability, cerebellar ataxia, and cortical myoclonus. The patient is using valproic acid 30mg/kg/day and experiencing regular psychotherapy, with behavior and myoclonus control.

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An unusual case of variegate porphyria with nerve enlargement, conduction block and idiopathic intracranial hypertension

Michelle Abdo Paiva[1], Andre Macedo Serafim Silva[1], Rodrigo de Holanda Mendonça[1], Ana Lucila Moreira[1], Nícolas Ruan dos Santos Cavalcante[1], José Pedro Soares Baima[1], Carlos Otto Heise[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Michelle Abdo Paiva; Email: michelle.abdop@gmail.com.

Acute hepatic porphyrias are a group of metabolic diseases where the main targets of neurotoxins ALA and PBG are the central and peripheral nervous system. Typical acute attack is characterized by an initial autonomic neuropathy, usually abdominal pain, followed by central nervous system symptoms or sensorimotor neuropathy. Chronic features have been suggested to be related to a small fiber neuropathy, but further investigation is still due. Here we report a patient with variegate porphyria with unusual findings during follow up, related to central and peripheral nervous systems. **Case presentation:** Female patient with previous history of acute attack (abdominal pain, urinary retention and tetraplegia) at age 27. She was evaluated at age 42 and as sequelae she had mild distal weakness and neuropathic pain in both hands and feet. She was diagnosed after genetic panel showed a pathogenic variant on PPOX - c.503G> A. Electrodiagnostic studies (EDX) were performed and revealed reduced compound motor action potential (CMAP) amplitudes in nerves of the lower limbs and both median nerves. Also, conduction block in both median nerves suggested acquired demyelinating neuropathy. Late responses showed absence of F waves and H reflexes in the tibial nerves. Sensory studies were normal. Neuromuscular ultrasound showed enlarged cross-sectional area in both median nerves in the forearm, consistent with neural thickening outside typical compression site. Six months later, she presented with psychosis and reported migrainous headache. Brain MRI suggested idiopathic brain hypertension. Lumbar puncture demonstrated increased opening pressure of 32 cmH2O. A new attack was discarded with biochemical exams. **Discussion:** Acute porphyric neuropathy is described as an axonal predominantly motor neuropathy. Secondary demyelinating findings have been described. A primary demyelinating neuropathy and partial conduction block, as seen in our case, have been reported in the acute phase and related to more severe acute attacks. A chronic distal axonal sensory-motor polyneuropathy can persist. Specific ultrasonography features of porphyric neuropathy have not been reported. **Final commentws:** This unpredicted finding can be a source of misleading diagnosis as inflammatory neuropathy. Finally, intracranial hypertension is not usually seen and further studies are needed to establish a relationship.

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An unusual presentation of small vessel disease: NIT1 gene mutation

Camila Rodrigues Nepomuceno[1], Nayara de Paula Guerreiro[1], Maria Fernanda Hamamoto Leati[1], Felipe Cenicio Juliatti[1], Joao Roberto Ribeiro Pimenta[1], Larissa Neves de Lucena[1], Cristiane de Araujo Martins Moreno[1], Julian Leticia de Freitas[1], Maria Sheila Guimaraes Rocha[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Camila Rodrigues Nepomuceno; Email: camilarnepomuceno@gmail.com.

Case presentation: A 39-year-old woman presented at our hospital with a sudden neurological deficit characterized by disproportionate right hemiparesis and severe motor aphasia. When asked about past medical history, relatives reported that during the immediate postpartum period seven years ago, the patient had a similar condition, resulting in non-limiting motor and language sequelae. In childhood, the patient had a cognitive delay, which evolved over the years into cognitive dysfunction with amnestic and executive predominance. The family history included information about consanguinity (her parents were first-degree cousins); she has eight siblings without comorbidities. Brain MRI showed T2/FLAIR hyperintensity in periventricular white matter and bilateral middle cerebellar peduncle, also demonstrated diffusely widened perivascular spaces in the striatum and acute stroke in the left middle cerebral arterial territory. CSF, echocardiogram, and MR angiography showed no abnormalities. Whole exome sequencing revealed a homozygous pathogenic variant c.883C> T (p.Arg295*) in the NIT1 gene, consistent with small vessel disease. **Discussion:** Given the cognitive changes associated with white matter disease and stroke, leukoencephalopathy and its differential diagnoses were considered. Whole exome sequencing showed a mutation in the NIT1 gene responsible for small vessel disease. The Nitrilase1 (NIT1) protein belongs to the nitrilase protein family. Its role in human health and disease is not yet fully understood. Studies in mice have indicated that NIT1 functions as a metabolite repair enzyme. Additionally, NIT1 is believed to act as a tumor suppressor by activating the TGFβ-Smad and WNT signaling pathways. Rutten et al. demonstrated that biallelic loss-of-function variants in NIT1 cause genetic cerebral small vessel disease. In this study, all patients exhibited a distinct neuroimaging phenotype, with the extent of white matter hyperintensities varying among patients, even those of the same age. Our patient showed an image pattern similar to leukoencephalopathy on MRI. This condition brings to a newly identified cerebral small vessel disease inherited in an autosomal recessive manner. This type of inheritance can obscure the genetic nature of the disorder, as patients often appear to be sporadic cases. **Final comments:** We describe an uncommon cause of cerebral small vessel disease with a distinct neuroimaging phenotype consistent with diagnoses of leukoencephalopathy caused by biallelic NIT1 loss-of-function variants.

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Anabolic androgenic steroids triggering reversible cerebral vasoconstriction syndrome

Cássia Marin Dapper[1], Natália Contini[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Natália Contini; Email: nataliacontini4@gmail.com.

Case presentation: A healthy 24-year-old male developed spontaneous and severe thunderclap headache (TCH), during low impact physical exercise. Neurological examination and vital signs were normal. Noncontrast brain computed tomography (CT), CT arteriography, laboratory tests and cerebrospinal (CSF) analysis displayed no

abnormal findings. The headache responded satisfactorily to intravenous analgesia. At the same day, after total pain improvement, patient requested to be discharged. One day after, he returned to the hospital after a new episode of TCH and severe nausea, not associated with physical effort. Once again, vital signs, neurological examination, and CT study were normal. Laboratory tests including blood count, lipid profile, prothrombin time, partial thromboplastin time, anti-nuclear antibody, anti-HBs, HBsAg, anti-HCV, anti-HIV and VDRL were unremarkable. Liver and renal function tests were above the limit of normality. Patient later revealed using an anabolic androgenic steroid (AAS) called oxandrolone in the past 6 weeks during gym time. At the same day of admission, a Brain MRI angiography revealed narrowing segments of P2 and P3 of posterior cerebral arteries, especially on the right side. There was no signs of bleeding or ischemia. Headache and nausea had gradual improvement after symptomatic medications. Liver and renal function tests came to normality and, after seven days, patient did not present any more episodes of headache. A MR angiography after four weeks demonstrated improvement in alternating stenotic areas in vessels. Thus, diagnosis of reversible cerebral vasoconstriction syndrome (RCVS) was established. **Discussion:** The features of RCVS are thunderclap headache, lack of subarachnoid hemorrhage by CSF and CT analysis, exclusion of cerebral vasculitis by CSF, and angiographic demonstration of multifocal segmental cerebral artery vasoconstriction that resolves with time. It is associated with some potential triggers. The most common are postpartum period, oral contraceptive medication, immunosuppressants drugs and vasoactive drugs, as antidepressants, nasal decongestants and illicit drugs. In the presented case, none of them was identified. The temporal relationship between the AAS initiation use and syndrome development can implicate the supplement as a contributing cause. Supraphysiologic doses of AAS have been associated with dyslipidemia, arrhythmias, cardiovascular remodeling and hypertension. Moreover, there is some evidence that androgen use is associated with thrombosis, which also increase the risk of cerebral venous or arteries events. The association between RCVS and AAS use has not previously been made. Indirect prothrombotic and proatherogenic mechanisms and direct cardiac effects can explain this possible correlation. **Final comments:** Thus, it is advisable to consider the potential roles of anabolic androgenic steroids in cases of otherwise unexplained cerebrovascular disease.

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Analgesics overuse in patients with headache in the neurology outpatient clinic of a university center: clinical-epidemiological profile

Ana Beatriz Rezende dos Santos Correa[1], Bruna Verdelho Leite[1], Daniele Azevedo Valente[1], Celice Cordeiro de Souza Bergh Pereira[1], Walther Augusto de Carvalho[1]

[1]Centro Universitário do Pará, Belém PA, Brazil.

Correspondence: Daniele Azevedo Valente; Email: danieleazevedos84@gmail.com.

Background: Medication-overuse headache is a secondary headache that usually occurs in patients suffering from primary headaches, worsening the intensity and frequency of attacks and is often underdiagnosed and treated inappropriately because of the population's lack of knowledge about it and high frequency of self-medication with over-the-counter analgesic agents. **Objective:** To identify the prevalence of analgesics overuse by patients diagnosed with headache treated from June 2021 to June 2023 in a neurology outpatient clinic of a medical specialty center in the city of Belém/PA. **Methods:** A cross-sectional, observational, analytical-descriptive study was carried. **Results:** 166 medical records of patients with headache were included, in which there was a higher prevalence of females (85.5%), aged between 41-60 years (45.2%) and the main type being migraine with or without aura (61.4%). A prevalence of 28.3% of analgesic overuse was found, in wich dipyrone was the most associated acute analgesic (44.7%) and the prophylactic medciation most associated was amitriptyline (34%). It is noteworthy that in 51.2% of the medical records there was no adequate recording of information about the use of analgesics, which could lead to a possible underdiagnosis. **Conclusion:** It is concluded that a considerable portion of patients treated for headaches was doing analgesics overuse, predominantly females, the majority using dipyrone and amitriptyline. It was also revealed that there is a need for greater vigilance regarding the excessive use of analgesics in this population, either through greater encouragement for patients to use a headache diary or through the incorporation of specific forms for the care of patients with headache in the electronic medical records system.

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Analysis of admissions by stroke in Southeast region since 2020 to 2023

Marcela Cristina de Souza Lemus[1], Anna Beatriz Dias Bertozzi[2], Ana Lígia Tiemi Soares Nishimura[3], Gabriela Silva Toro[4], Lívia Maria Oliveira Franco Vieira[5], Lucas Fernando Silva Moreira[6], Vitória Stefanny Souza Barbosa[7], Pedro Lucas Gomes Rodrigues[8], Yasmim Melo Costa de Azevedo[9], Cayara Mattos Costa[10]

[1]Universidade Federal do Rio de Janeiro, Macaé RJ, Brazil.
[2]Faculdade de Minas BH, Belo Horizonte MG, Brazil.
[3]Universidade de Gurupi, Gurupi TO, Brazil.
[4]União Metropolitana de Educação e Cultura, Lauro de Freitas BA, Brazil.
[5]Centro Universitário Inta, Sobral CE, Brazil.
[6]Centro Universitário FIPMoc, Montes Claros MG, Brazil.
[7]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil
[8]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[9]Universidade Nove de Julho, São Paulo SP, Brazil.
[10]Universidade Federal do Maranhão, São Luís MA, Brazil.

Correspondence: Marcela Cristina de Souza Lemus; Email: marcelalemus47190@gmail.com.

Background: Cerebrovascular diseases are one of the principal causes of death in Brazil, in the last four years more than 106 thousand obits were registered by stroke (AVC), according to DATASUS. Recent studies have shown a relation between the grave acute respiratory syndrome caused by SARS-CoV-2 and the development of grave neurological manifestations, as the AVC. Due to the increase of cases of this disease after the pandemic, this study is important to understand the data of AVC admissions. **Objective:** Analyze the admissions by AVC in the southeast region between 2020 and 2023. **Methods:** Ecological study realized with Hospitals System Informations of SUS data, vinculated to DATASUS. It was evaluated the number of admissions of ischemic AVC or acute hemorrhagic AVC treatment (procedure: 0303040149) in the period of 2020 to 2023 in southeast region, including the variables: total of admissions, average cost, average time of hospitalization and mortality tax per year of service. As this was a study that uses secondary databases, it wasnt necessary the approval of the Ethics Committee. **Results:** In the period analyzed, the southeast region presents a total of 356,513 admissions to AVC treatment. During this period, there was a progressive growth of admissions, from 80,326 to 95,888 hospitalizations, which represents an increase of 19.37% between 2020 and 2023. The average cost per hospitalization increased from R$1,529.13 in 2020 to R$1,934.84 in 2023, without significant alterations between 2020-2021 and 2022-2023. The average time of hospitalization remained constant between the years analyzed, approximately 7.6 days. In relation of the mortality tax, in the first two years of COVID-19 pandemic, there was an increase of 16.68/100,000 habitants in 2020 to 17.61/100,000 habitants in 2021, while between 2022 and 2023 there was a reduction of mortality tax of 15.97 to 14.93 per 100,000 habitants, respectively. It is possible that the data presented here are underestimated by possible sub notifications. **Conclusion:** It was observed an increase in the total number of internations in the period analyzed, in addition to an increase of the average cost of hospitalizations. The tax values of mortality are consistent with the most critical phase of the first years of pandemic, being higher by 2021 and decreasing in the last two years.

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Analysis of contraindications to chemical thrombolysis in patients within the therapeutic window for stroke in a public tertiary neurology service in the state of Pernambuco

Ana Caroline Paiva Simeao[1], Ana Caroline Paiva Simeao[1], Mayhara Rosany da Silva Santiago[1], Heloisy Maria Nunes Galvao[1], Nereu Alves Lacerda[1], Raissa Josefa Pereira de Moura[1], Luis Felipe Barros e Silva Pereira Ribeiro[1], Fernando Tenorio Travassos[1]

[1]Hospital Pelopidas Silveira, Recife PE, Brazil.

Correspondence: Ana Caroline Paiva Simeao; Email: carolpaivasimeao@gmail.com.

Background: Stroke is a significant cause of morbidity and mortality worldwide, being the second leading cause of death. Chemical thrombolysis is currently a major milestone for the acute treatment of ischemic stroke, with the greatest effectiveness within the first 3 hours, extending up to 4.5 hours. Currently, alteplase is the only approved chemical thrombolytic, but other thrombolytics like tenecteplase and other tissue reperfusion techniques are being studied to improve treatment response. **Objective:** To conduct an epidemiological profile of patients within the therapeutic window at a tertiary neurology hospital between February and May 2024, evaluating contraindications to the use of thrombolytics. **Methods:** A cross-sectional, observational study was conducted, including all patients within the therapeutic window for chemical thrombolysis at a public tertiary neurology service in the State of Pernambuco, observing the contraindications to thrombolysis. **Results:** A total of 71 thrombolysis protocol activations were evaluated at the institution, where 6 were patients outside the window or with an indeterminate onset, 27 underwent chemical thrombolysis (18 female and 9 male), and 44 did not undergo thrombolysis (22 female and 22 male). The reasons for not performing thrombolysis included: 11 cases of hemorrhagic stroke, 11 cases with a low NIHSS (≤4), 6 cases of extensive ischemic stroke with ASPECTS ≤8 or high NIHSS (> 23), 2 cases missed the window due to uncontrolled blood pressure for thrombolytic infusion, and 6 cases had other reasons, including patient refusal, issues with cranial CT, and recent stroke or traumatic brain injury. **Conclusion:** The majority of patients had some contraindication to chemical thrombolysis with alteplase, with hemorrhagic stroke and low NIHSS being the main reasons for not performing thrombolysis. Training the team on the importance and speed of indicating and contraindicating chemical thrombolysis is crucial. It is also important to evaluate that, in addition to chemical thrombolysis, there is the possibility of thrombectomy, which is not always available in public tertiary neurology services and could have been an alternative for patients with contraindications to chemical thrombolysis.

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Analysis of deaths due to cerebrovascular diseases in the state of São Paulo between 2010-2022: an ecological study

Cristina de Souza Lemus[1], Maria Clara Missio dos Santos[2], Nivea de Mattos Amarante Rodrigues[1], Tainara Maria de Souza Abrão[3], Marjorie Correia de Andrade[4]

[1]Universidade Federal do Rio de Janeiro, Macaé RJ, Brazil.
[2]Universidade Nove de Julho, São Bernardo do Campo SP, Brazil.
[3]Pontifícia Universidade Católica de Minas Gerais, Poços de Caldas MG, Brazil.
[4]Universidade Potiguar, Natal RN, Brazil.

Correspondence: Marcela Cristina de Souza Lemus; Email: marcelalemus47190@gmail.com.

Background: Cerebrovascular diseases are among the main conditions responsible for deaths, physical disability, and impaired quality of life globally. According to data from the World Health Organization, six million individuals succumb annually to these diseases, making them the second leading cause of annual mortality. **Objective:** This study aims to analyze deaths from cerebrovascular diseases in São Paulo from 2010 to 2022. **Methods:** An ecological, observational study, using data from the Mortality Information System available on the website of the Department of Informatics of the Unified Health System (DATASUS). The information collected pertained to death notifications by residence with cause coded as "CID-BR-10: 070 - cerebrovascular diseases". Deaths were evaluated by age group, sex, year of occurrence, and color/race in the state of São Paulo between 2010 and 2022. Approval by the Ethics Committee was not required. A limitation of the study is the possible underreporting of cases. **Results:** In the analyzed period, 284,188 deaths from cerebrovascular diseases were recorded in the state of São Paulo. It was observed that between 2010 and 2020, the number of annual records did not exceed 22,308 deaths, in 2016. However, from the year 2021, a growing increase in these values was observed, reaching 23,058 deaths in 2022. The elderly, aged 60 years and older, represent 80.6% of the deaths during this period, with those over 80 years old being the most affected, accounting for 100,711 deaths (43.98%). Regarding sex, although male cases were predominant in the analyzed interval, deaths among men and women are close, totaling 144,064 and 140,110, respectively. In the analysis by color/race, a predominance of white color was observed, with 194,186 cases, representing 68.33% of the total deaths. The capital of São Paulo recorded 125,550 occurrences, corresponding to approximately 44% of the total of all analyzed regions, compared to 30% of the total cases registered outside the metropolitan region. **Conclusion:** The observed data indicated an increase in the number of deaths between 2021 and 2022, with the elderly being the majority of victims of cerebrovascular accidents. This phenomenon can be explained by the higher prevalence of comorbidities in this age group. Additionally, studies in the literature highlight the various disease mechanisms of SARS-CoV-2, such as vascular and nervous system involvement, which may be related to the increase in deaths from cerebrovascular diseases during the pandemic periods.

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Analysis of efficiency between the cost, length of stay and clinical outcomes for stroke in the states of the Northern region of Brazil

Sávio Roberto Silva Costa[1], Emilly Alessandra Cruz dos Reis[1], Luan Nascimento Pereira de Amorim[1], Danusa Neves Somensi[1], Mariângela Moreno Domingues[1], Emanuel de Jesus Soares de Sousa[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Sávio Roberto Silva Costa; Email: saviorsc20@gmail.com.

Background: Stroke is one of the leading causes of morbidity and mortality across the globe, posing a significant challenge to healthcare systems. Characterized by the interruption of blood flow to the brain, stroke can result in severe and permanent neurological damage. Treating stroke patients involves high costs, including hospital expenses, medication, rehabilitation and long-term care. In addition, length of stay is a critical factor that influences both costs and patients' clinical outcomes. Thus, understanding these dynamics can provide valuable insight into improving treatment processes, reducing expenses and maximizing the quality of patient care. **Objective:** To evaluate the efficiency of stroke management in the northern region based on financial variables and the clinical outcome. **Methods:** This is a cross-sectional, observational study with a quantitative approach. The data was collected from the Department of Information and Informatics of the Unified Health System (DATASUS), in the hospital morbidity section (SIH/SUS). Variables related to the number of hospitalizations, total expenditure, average expenditure per hospitalization, average length of stay and mortality rate were analyzed; data was collected from 2014 to 2023. Bioestat 5.3 software was used for the statistical analysis. A p-value of less than 0.05 was considered statistically significant for this study. **Results:** In the period from 2014 to 2023, 17,018 hospitalizations for stroke were recorded in the Northern region, with 84.1% of the records occurring in Amazonas and Pará, where 11,839 and 2,479 hospitalizations were reported, respectively. Amapá had the lowest number of notifications in the period (73). The average regional cost per hospitalization was R$905.88; the states with the highest costs were Roraima (R$1,270.68) and Rondônia (R$1,227.31), while Acre had the lowest average cost per hospitalization (R$715.98). With regard to hospital stays, the North region had an average of 6.7 days, which was exceeded by four states, Amapá (12.1), Roraima (10.2), Acre (8.4) and Amazonas (6.9), while the lowest average was in Rondônia (4.9). The regional mortality rate during the period was 11.06, and the state with the highest death rate was Amapá (16.44), followed by Roraima (12.24) and Amazonas (11.84). The states with the lowest death rates were Rondônia (7.63%) and Tocantins (6.4%). **Conclusion:** Based on the data collected, there was a concentration of hospitalizations in Amazonas and Pará, which also had the highest total expenditure on hospitalizations for stroke in the North. Most of the costs were related to hospital services. The states of Amazonas, Acre, Amapá and Roraima stood out with average hospital stays and mortality rates higher than the regional average. In terms of efficiency, the most efficient state in the North was Tocantins, while Amapá was the least efficient in terms of expenditure and clinical outcomes for stroke.

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Analysis of epilepsy cases in Brazil in the last five years

Caroline Patrícia Amaral Costa[1], Manuel Edmundo Teles Neto[2], Chan David Ribeiro[3], Rafael Reis de Oliveira[1], Moisés Cardoso Portilho[2], Laura Lino Gonçalves[3], Luiz Carlos Figueiredo Filho[3], Luan Nascimento Pereira de Amorim[3]

[1]Universidade Federal do Pará, Belém PA, Brazil.
[2]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.
[3]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Caroline Patrícia Amaral Costa; Email: caroline.costa@ics.ufpa.br.

Background: Epilepsy is a serious syndrome in which the patient is in an imbalance of various modulators of the synaptic action potential, such as receptors, neurotransmitters, enzymes or the neurons themselves. These alterations result in episodes of focal or generalized cortical hyperexcitation. Acute symptomatic seizures affect around 29 to 39 people per 100,000 inhabitants, mainly children under the age of 1 and the elderly, who are predisposed to cardiovascular complications. Due to the complexity of epilepsy, it is crucial to analyze patient data in Brazil in order to identify risk groups, protective factors and strategies to reduce morbidity and mortality. **Objective:** To analyze the epidemiological profile of epilepsy cases over the last 5 years in Brazil. **Methods:** This is an epidemiological, ecological and temporal study, which used data collected from 2019 to 2023, available in the SUS Hospital Information System (SIH/SUS), provided by the SUS Information Technology Department (DATASUS). The following variables were selected: number of procedures, expenditure per hospital admission authorization (AIH), length of stay, average length of stay, deaths and mortality rate by Region/Federation Unit and Year of Processing. The data was tabulated using the Microsoft Excel 2021 program. **Results:** With regard to hospital admission authorizations (AIH), 228,181 cases were approved, with the Southeast region predominating with 41.54% (n=94777, p< 0.0001), while the North region had only 6.46%. In terms of average length of stay, the regions had an average of 5.8 days, with the Northeast having the longest with 6.1 days, while the Midwest had 5.1 days. In addition, with regard to the number of deaths, 6,901 deaths were recorded during this period, with the Southeast having 48.37% (n=3338, p< 0.0001), followed by the Northeast with 28.73%, while the total mortality rate for the period was 3.02, with the average rate by region predominating in the Southeast with 3.52. Finally, the average value per total hospitalization was R$876.32, and there was significant variation between the regions, with the Southeast region having an average of R$988.05 (p<0.0001) and the North having the lowest average with R$565.75. **Conclusion:** During the period analyzed, it should be noted that there was a significant predominance of the Southeast region in the number of hospitalizations and deaths, which may be associated not only with the demographic density of the region, but also with the high demand and complexity of the patients. Therefore, it is understood that the high cost per hospitalization associated with this region. Furthermore, based on the data obtained from the Northeast, it can be concluded that there is a greater need for prolonged treatment and intensive measures in this region, which may also be related to the considerable number of deaths.

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Analysis of four attention tests and the influence of education and intelligent coefficients

Hellen Rose Maia Salazar[1], Ana Luiza Caldas Coutinho[2], Sergio Luis Schmidt[1]

[1]Universidade Federal do Estado do Rio de Janeiro, Programa de Pós Graduação em Neurologia, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Ana Luiza Caldas Coutinho; Email: analu.coutinho@edu.unirio.br.

The neurological circuit that involves attention includes prefrontal areas, thalamus, anterior cingulum, and brainstem. Other cognitive domains, such as memory, use more complex neocortical networks. When combined, all those networks form an individual's general intelligence. Although these cognitive domains can be affected even without attention deficits, the opposite is not true as attention is a prerequisite for them. Therefore, properly excluding attention deficits is a core process for the correct analysis of other cognitive processes. In this context, an ideal attention test would be one that can measure attention performance with minimal interference from

general intelligence. Currently, most attentional tests are anchored in years of formal education. However, to our knowledge, there aren't attentional tests standardized based on the Intelligent Quotient (IQ). Our main objective is to analyze if there was influence of intelligence scores on attentional performance through different neuropsychological tests. We also aimed to identify whether these attention tests were affected by different educational levels. This study age range was of 18 to 65 years. Education levels were divided as: elementary (0 to 8 years of instruction), high school (9 to 12) and higher education (13+). All participants were submitted to the following attention tests: Continuous Visual Attention Test (CVAT), Diffuse Attention Test for Drivers (TADIM), Discriminative Attention Test for Drivers (TADIS), and Pictorial Visual Memory Test (TEPIC). Intellectual performance was measured through the R-1 Non-Verbal Intelligence Test. R-1 raw points were divided into 3 clusters based on the percentile as: ≤ 25%; > 25% and < 75%; and ≥ 75%. For each attention test we analyzed the correlations between the R-1 raw scores and attention performance. A multivariate analysis of covariance (MANCOVA) was used to test the effect of R-1 clusters in the attention performance. Educational level was used as a covariate. The sample included 1036 participants (75.8% males). The mean age was 31.8 years. Education levels were represented as 17% elementary, 48% high school, 29% higher education and 5% not informed. The correlations indicated significant coefficients for TADIM (r = 0.486, P = 0.000), TADIS (r = 0.502, P = 0.000), and TEPIC (r = 0.311, P = 0.000). The CVAT variable that reached the highest correlation coefficient was the intrinsic alertness subdomain (r = 0.143, P = 0.000). A MANCOVA indicated a significant effect of the R-1 cluster for all the tests, excepted for the CVAT. The educational levels were also found equally significant for the TADIM, TADIS, and TEPIC except the CVAT. We concluded that TADIM, TADIS, and TEPIC test results were significantly affected by IQ and educational levels. Then, standardized data of these tests need to be corrected not only for the educational level, but also for the individual's IQ.

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Analysis of functional/dissociative seizures (FDS) semiology based on video-electroencephalography (VEEG)

Luiza Barbosa de Oliveira[1], Gláucia Monteiro Oliveira[1], Vivian Proença Lara[1], Caio Cobucci Esteves[1], Igor Pereira Matos de Oliveira[1], Gustavo Barros Loureiro[1], Vanessa Alves Lobato[1], Aline Curcio de Moraes[1], Helena Barbosa Horta Santos[1], Maria do Carmo de Vasconcellos Santos[1]

[1]Santa Casa de Belo Horizonte, Belo Horizonte MG, Brazil.

Correspondence: Luiza Barbosa de Oliveira; Email: luiza.neurologia@gmail.com.

Background: Functional/dissociative Seizures (FDS) are episodes that may resemble epileptic seizures, but lack abnormal brain electrical activity. The gold standard for diagnosis is videoelectroencephalography (VEEG), however, diagnosis is often delayed due to limited centers offering VEEG facilities. Therefore, estimating the most frequent semiology in FDS can be an useful tool to help physicians in order to diagnose and manage these disorders. **Objective:** Detailed semiological analysis may assist in diagnosis. Then, the research focus is to analyze the semiology of documented FDS from our VEEG unit from 2020 to 2023. **Methods:** We retrospectively analyzed medical and VEEG records of patients with FDS with at least one typical event documented. Records were then reviewed for the presence or absence of semiological signs considered as potential discriminators between FDS and Epileptics Events according to the pertinent literature. Prevalence in our study allowed us to select 7 practical semiological features with potential to distinguish FDS from Epileptics Events. These items were: eyes closed, body position (opisthotonus), asynchronous limbs movement, head movements side-to-side, duration > 2 minutes, complete unresponsiveness and rapid postictal reorientation. Comparative analyses were carried out to identify significant differences between patients diagnosed with FDS and concomitant epilepsy from those with FDS and no concomitant epilepsy. For this, the Chi-square test (via simulation of Monte Carlo) was performed. In all analyzes carried out, the differences obtained were considered statistically significant when the p value< 0.05. Statistical analyzes were performed using the Stata® program. **Results:** In the period from 2020 to 2023, 828 patients were evaluated at the VEEG unit. Of these, 11.35% (n=94) documented FDS. Concomitant epilepsy was detected in 48.94% of patients positive for FDS. Considering the semiology of FDS the presence of asynchronous limb movements (23.89%), forced eye closure and duration greater than 2 minutes (15.79% each) were the most frequent signs. Data on semiology indicated a significant difference only in relation to the rate of forced eye closure. According to the results presented, this type of signal was observed at statistically lower rates in FDS patients and concomitant epilepsy, when compared to patients without epileptic events. **Conclusion:** The diagnosis of FDS can be challenging, therefore, an evaluation of semiology can help. When the patient has a normal interictal EEG, it may be possible to make a probable diagnosis of FDS, even without VEEG. For that, review of the clinical event suggestive of FDS by an experienced physician is needed.

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Analysis of hospitalizations for carpal tunnel syndrome surgery in Brazil from 2014 to 2023

Laisa de Paula da Costa Lobato[1], Bárbara Alessandra Solano Silva[2] , José Antônio Solano Silva[1], Luan Victor Silva Ribeiro[1], Hálisson Ferreira Freitas Filho[3], Leandro Gabriel Costa Macedo[1], Ricardo Ferreira Silva[1], Dirceu Jose de Freitas Cardoso[1], Bárbara Coutinho Morgado[2]

[1]AFYA Faculdade de Ciências Médicas , Abaetetuba PA, Brazil.
[2]Centro Universitário do Pará, Belém PA, Brazil.
[3]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Laisa de Paula da Costa Lobato; Email: laisaikilman@gmail.com.

Background: Carpal tunnel syndrome is a neuropathy characterized by compression of the median nerve at the wrist level, capable of affecting sensation in the region. It is one of the upper extremity disorders that generates substantial costs and disabilities, with an estimated prevalence between 4% and 5% of the population. In many cases, conservative treatment is ineffective, necessitating surgical intervention. Potential complications include suture dehiscence, infection, and tendon injuries, contributing to increased hospitalizations of these patients. **Objective:** To analyze the epidemiological profile of hospitalizations for carpal tunnel syndrome surgery in Brazil from 2014 to 2023. **Methods:** An ecological, retrospective, quantitative study using data collected from the Sistema de Informações Hospitalares (SIH/SUS) via Departamento de Informática do Sistema Único de Saúde (DATASUS). Variables were analyzed using descriptive statistics, focusing on surgical procedures for carpal tunnel release in Brazil during the study period, including annual hospitalization numbers by state, deaths, and average length of stay. **Results:** A total of 126,335 hospitalizations were reported, with the majority in 2023 (18,934 or 15%) and the fewest in 2020 (7,714 or 6.1%), potentially reflecting increased "home office" employment post-pandemic leading to repetitive strain injuries, such as typing. The Southeast region had the highest number of hospitalizations (60,745 or 48%), whereas the North had the lowest (1,802 or 1.42%), possibly due to higher rates of repetitive strain injuries in jobs involving finger and hand movements, such as typing, compared to predominantly manual labor in the North. Regarding deaths, 3 were recorded, which may be explained by the low complexity of carpal tunnel syndrome surgery. 2 deaths occurred in the South region and 1 in the Northeast. The average length of hospital stay in Brazil was 0.6 days, likely due to the straightforward postoperative course and low complication rates. The Central-West region had the highest average stay (1.3 days), while the Southeast had the lowest (0.4 days). **Conclusion:** Analysis of hospitalizations for carpal tunnel syndrome surgery revealed significant variations across Brazil. The peak in 2023 may be attributed to increased "home office" practices leading to repetitive strain injuries. The Southeast region experienced the highest number of hospitalizations, likely due to a higher prevalence of jobs involving repetitive finger and hand movements. The low mortality rate and short hospital stays highlight the safety and low complexity of this surgery. These findings underscore the importance of developing public policies aimed at treating and preventing this condition.

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Analysis of hospitalizations for stroke before and during the COVID-19 pandemic in Bahia: an epidemiological approach

Victor Vale da Silva[1], Júlia Ribeiro Oliveira Campos[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Afya Faculdade de Ciências Médicas de Itabuna, Itabuna BA, Brazil.

Correspondence: Victor Vale da Silva; Email: vitor.futebolclube@hotmail.com.

Background: Cerebrovascular accident (CVA) is one of the main causes of mortality and disability globally. In Bahia, the COVID-19 pandemic brought significant changes to the healthcare system, impacting the management of several medical conditions, including stroke. This objective study analyzes the profile of stroke hospitalizations in Bahia, comparing the pre-pandemic and during the pandemic periods, with the aim of identifying changes in hospitalization patterns and obtaining information to improve stroke management during periods of health emergencies. **Objective:** To evaluate the epidemiological profile of hospitalizations for stroke in Bahia between the pre-pandemic (2017-2019) and pandemic (2020-2022) periods. **Methods:** This is an epidemiological, retrospective and descriptive study that collected secondary data from the Hospital Morbidity System (SIH/SUS) to analyze the epidemiology of hospitalizations for stroke in the state of Bahia. The pandemic period analyzed was from January 2020 to December 2022, being compared to the pre-pandemic period, from January 2017 to December 2019. The variables studied included: number of hospitalizations, age group, sex, year and color /race. Data analysis was carried out in the Microsoft Excel program, using graphs to generate descriptive statistics. **Results:** In the period 2017-2020, there were 37,045 hospitalizations for stroke, rising to 41,842 during the pandemic (increase of 12.95%). Average hospital admissions decreased before the pandemic (7.6 days in 2017-2018, 7.3 days in 2019), but increased during the pandemic (7.3 days in 2020, 7.4 days in 2021, 7.7 days in 2022). The death rate, which was falling before the pandemic, increased during the pandemic. The number of hospitalizations for men increased from 18,673 to 21,061 (an increase of 12.8%) and for women from 18,372 to 20,781 (an increase of 13.1%). The male mortality rate reduced slightly (17.8 to 17.6), while the female mortality rate increased (17.91 to 18.18). The number of hospitalizations of black and brown people increased from 18,695 to 26,227 (an increase of 40.3%). In the elderly, hospitalizations increased by 10.9%, in adults aged 20 to 59, by 17.9%, and in young people under 20,

by 61.1%. **Conclusion:** The results indicate an increase in hospitalizations for stroke during the pandemic, with longer hospital stays and a change in the downward trend in the mortality rate. There were increases for both men and women, with variations in the death rate by sex. Analysis by race showed an increase in hospitalizations among black and mixed-race patients, possibly associated with the fact that these groups constitute the majority of those facing conditions of socioeconomic vulnerability. All age groups recorded an increase in hospitalizations, especially younger people. Therefore, these results highlight the need for prevention and treatment strategies adapted to each group in order to improve stroke management in challenging contexts.

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Analysis of hospitalizations for the treatment of polyneuropathies in Brazil between 2019 and 2023

Ana Clara Lacerda Freitas[1], Aline Teixeira[1], Laysa de Souza Maia[2], Andressa Gonçalves Vicente[3], Ana Alice Lemos Lima[4], Gabriela Hernandez Dumani[5], Ana Cristina Carvalho e Silva[3], Estefany Moreno de Souza[6], Geovana Franklim Gomes e Silva[7], Juliana Braga Rodrigues de Castro[8]

[1]UniFunorte, Montes Claros MG, Brazil.
[2]Faculdades Integradas Aparício Carvalho, Jaru RO, Brazil.
[3]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[5]Universidade Franciscana, Santa Maria RS, Brazil.
[6]Universidad de Aquino Bolivia, Santa Cruz de La Sierra, Bolivia.
[7]Centro Universitário de Patos de Minas, Pato de Minas MG, Brazil.
[8]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Ana Clara Lacerda Freitas; Email: anaclf15@yahoo.com.br.

Background: Polyneuropathies are conditions that affect multiple peripheral nerves, causing a wide range of symptoms that vary depending on the nerves involved. These conditions can result from various causes, including metabolic diseases, nutritional deficiencies, toxins, infections, and autoimmune disorders. In the Brazilian context, the impact of polyneuropathies on public health is significant, with an increasing number of cases diagnosed each year. **Objective:** To analyze hospitalizations and healthcare costs related to the treatment of polyneuropathies in the Brazilian Unified Health System (SUS) between 2019 and 2023. **Methods:** This ecological study utilized data extracted by the Department of Informatics of the Brazilian Unified Health System (DATASUS). Data analysis was performed using Google Sheets, covering the number of cases and duration of hospitalizations for the treatment of polyneuropathies in Brazil, average cost per hospitalization, total cost, and mortality rate from 2019 to 2023. **Results:** Between 2019 and 2023, there were 37,810 hospitalizations for the treatment of polyneuropathies in Brazil, with 2022 having the highest number of hospitalizations (8,402), followed by 2019 (8,167), 2021 (7,509), 2023 (7,100), and 2020 with the lowest number of hospitalizations (6,632) in the period analyzed. There was a reduction in 2020, likely due to the impact of the COVID-19 pandemic, followed by a gradual recovery in subsequent years, until a decline again in 2023. The Southeast Region had the highest number of hospitalizations, with 13,412, followed by the Northeast (9,752), South (9,098), North (3,317), and Central-West with 2,231. The total expenditure for these hospitalizations was R$ 50,262,017.26, with the highest in 2022 (R$ 13,091,569.51) and the lowest in 2020 (R$ 7,554,768.12). The average cost per hospitalization increased from R$ 1,029.95 in 2019 to R$ 1,602.97 in 2023, reflecting an increase in costs over time. Additionally, the average hospital stay increased from 8.7 days in 2019 to 9.3 days in 2023, as well as the mortality rate, from 2.15% in 2019 to 2.66% in 2023. **Conclusion:** The year with the most hospitalizations was 2022, which consequently had the highest total expenditure. Additionally, the data indicate an increase in costs related to hospital resources and a rising mortality rate, highlighting significant challenges in the treatment of polyneuropathies. Through this analysis, it is possible to identify potential contributing factors, evaluate the effectiveness of existing interventions, and guide public health policies to improve the prevention, diagnosis, and management of these conditions.

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Analysis of melatonin as a therapeutic tool in the treatment of neurologically-originated insomnia: a systematic review

Hálisson Ferreira Freitas Filho[1], João Victor Souza de Barros[1], Larissa Suzan Basilio e Silva[1], Felipe José Silva e Silva[1], Luan Víctor Silva Ribeiro[2] , Izabella Carolline Menescal Ferreira[1], Igor Moreira Miguez Godoy[3] , Kaliana Kennedy Oliveira Calixto[1], Antônio Marcos da Silva Henriques[1], Nathália Coimbra Coêlho[1]

[1]Universidade Federal do Pará, Belém PA, Brazil.
[2]Afya Faculdade de Ciências Médicas
[3]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.

Correspondence: Hálisson Ferreira Freitas Filho; Email: halisson.filho@ics.ufpa.br.

Background: Insomnia, a common symptom among sleep disorders, is primarily characterized by difficulty sleeping due to mental or physical conditions. To determine the pathogenesis, a thorough analysis of the patients history is necessary to identify any underlying cause. The diagnosis refers to a symptom of insomnia that requires specific attention. The therapeutic approach can be cognitive-behavioral, which is the most commonly used, or pharmaceutical, which is also widely utilized and is the focus of this study. **Objective:** Explore the efficacy of using melatonin as a treatment for neurologically-originated insomnia. **Methods:** This involves conducting a systematic literature review of the past 10 years on the "PubMed" platform using the descriptors: "insomnia", "melatonin", and "neurological. Initially, 30 articles were selected, and of these, only 3 were included based on the following criteria: english language, systematic review, clinical trial, meta-analysis, and relevance to the topic. Twenty-six articles were excluded based on the exclusion criteria: not being in english, studies not aligned with the proposed theme, and incomplete articles. **Results:** Most studies assessed the effects of a specific dosage of slow-release melatonin on quantitative variables such as total sleep time, sleep latency, and wakefulness after sleep onset. In studies with elderly patients, in which insomnia is highly prevalent, as in other special populations like children, the results of melatonin use have been inconsistent. Although subjective improvement in sleep quality has been observed with the slow-release form, no benefit on total sleep time has been demonstrated using **Objective:** parameters. In a randomized, double-blind, placebo-controlled, crossover study with children affected by epilepsy and sleep problems, it was observed that slow-release melatonin improves sleep latency and wakefulness after sleep onset, a similar outcome of a systematic review with adults patients with neuropsychiatric disorders. However, these were small studies, so the results may not be generalizable to an open scope. **Conclusion:** Therefore, there is not enough evidence to recommend the use of melatonin for the treatment of insomnia, and a larger scale study is necessary to analyze the efficacy for secondary outcomes, safety, and tolerability in children with epilepsy.

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Analysis of morbidity due to dementia in elderly individuals in Southeast Brazil (2008-2024): an ecological study

Anna Gabriella Venâncio Neves de Moraes Tosta[1], David Cohen[2], Larissa Farah de Oliveira[3] , Hadassa Lucena Sales Santos[4] , Paulo Fernando Katsuo Ogatha Ito[5]

[1]São Leopoldo Mandic, Campinas SP, Brazil.
[2]Universidade Luterana do Brasil, Canoas RS, Brazil.
[3]Universidade Municipal de São Caetano do Sul, São Caetano do Sul SP, Brazil.
[4]Universidade Ceuma, Imperatriz MA, Brazil.
[5]Universidade para o Desenvolvimento do Estado e da Região do Pantanal, Campo Grande MS, Brazil.

Correspondence: Anna Gabriella Venâncio Neves de Moraes Tosta; Email: Annagabriellatosta@gmail.com.

Background: Dementia is a syndrome of degeneration of higher cortical functions, involving memory, thinking, comprehension, language, and judgment. It has become increasingly important as a public health problem due to the growth of the elderly population, projecting that these individuals may reach 23.8% by 2040, thus nearly a quarter of the Brazilian population. **Objective:** To analyze the morbidity and mortality due to dementia in the elderly population in the Southeast Region from January 2008 to March 2024. **Methods:** This is an ecological, cross-sectional, descriptive study with a quantitative approach, conducted in May 2024, using data collected from the Hospital Information System of SUS (SIH/SUS - Sistema de Informações Hospitalares do Sistema Único de Saúde), available from the Department of Informatics of the Unified Health System (DATASUS - Departamento de Informática do Sistema Único de Saúde). The variables used were hospitalizations, total diagnostic value, deaths, and mortality rate. The collected data were organized in Microsoft Excel spreadsheets and analyzed using descriptive statistics. **Results:** Mortality rates, deaths, hospitalizations, and total diagnostic value were observed, respectively, for each state in the Southeast Region. In Minas Gerais, it was 19.04% (626 deaths and 3,288 hospitalizations) totaling R$15,725,524.48; in Espírito Santo, it was 7.38% (9 deaths and 122 hospitalizations) totaling R$323,552.44; in Rio de Janeiro, it was 15.11% (950 deaths and 6,289 hospitalizations) totaling R$38,420,414.65; in São Paulo, it was 7.18% (511 deaths and 7,114 hospitalizations) totaling R$40,252,064.51. Hence, it was possible to verify that the mortality rate in the state of Rio de Janeiro is lower than that of Minas Gerais; additionally, it has an approximately 50% higher total value allocated compared to the number of hospitalizations that occurred in the state of Minas Gerais. Thus, there is an unequal distribution of funds in Brazil, resulting in avoidable deaths. **Conclusion:** The data examined highlight the discrepancy in resources allocated to Minas Gerais compared to Rio de Janeiro. Therefore, the lack of support professionals such as nurses, psychologists, and physiotherapists for this population exacerbates this situation, consequently requiring more investment in dementia prevention measures and care for diagnosed patients with comprehensive services. For example, early recognition of the disease and differential diagnosis to mitigate the impacts of this disease, improve the patients quality of life, and increase their survival.

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Analysis of mortality due to intracranial trauma in São Paulo from 2020 to 2023: an ecological study

Maria Clara Calixto Caetano[1], Giulia Chiavegato Locatelli[2] , Nicolas Inkei Chiba Frazao[3], Bruna Ravany Farias Marques[4] , Beatriz Moraes Florenzano[5], Juliana Braga Rodrigues de Castro[6]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade Nove de Julho
[3]Universidade Federal do Pará, Belém PA, Brazil.
[4]UNIFG, Guanambi BA, Brazil.
[5]Faculdade Santa Marcelina São Paulo, São Paulo SP, Brazil.
[6]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Maria Clara Calixto Caetano; Email: maria.caetano@sou.unaerp.edu.br.

Background: Intracranial Trauma is recognized as one of the leading causes of death and disability, constituting a significant public health issue. This trauma can impair consciousness and, depending on its severity, can affect physical, cognitive, emotional, or behavioral functioning. The Glasgow Coma Scale (GCS) and the pupillary light response are used to assess the severity of brain injury. According to the GCS score, trauma can be classified as mild, moderate, or severe. Falls are the most common cause of such injury, with motor vehicle incidents being the second leading cause. Several complications can arise, including intracranial hemorrhage, cerebral edema, elevated intracranial pressure, vasospasm, seizures, infection, hydrocephalus, memory loss, and hyponatremia. Early patient management and rapid intervention are crucial for a favorable prognosis. **Objective:** This study aims to analyze and present the epidemiology of intracranial trauma in the state of São Paulo from 2020 to 2023, identifying the factors and epidemiological characteristics of the affected patients. **Methods:** This is an ecological, retrospective, and descriptive study, with data collected from the Department of Informatics of the Unified Health System (DATASUS). The variables analyzed included mortality rate, race/ethnicity, sex, age group, and municipalities, focusing on intracranial trauma in the state of São Paulo from 2020 to 2023. **Results:** The mortality rate due to intracranial trauma in the state of São Paulo from 2020 to 2023 was 11.00 per 100,000 population. For males, the rate was 11.55 per 100,000, while for females it was 9.36 per 100,000. The age group with the highest rate was 80 years or older, with 21.45 per 100,000. Regarding race/ethnicity, the Indigenous population had the highest rate at 25.00 per 100,000, followed by Asians at 15.52 per 100,000. The municipality with the highest rate was Tapiratiba, with 100 per 100,000, followed by Queluz, with 50 per 100,000. **Conclusion:** Mortality due to intracranial trauma in the state of São Paulo during the evaluated period was higher among males, likely related to biological factors. The age group 80 years and older had the highest rates, suggesting the influence of factors associated with aging. Among municipalities, Tapiratiba had the highest rate at 100 per 100,000. Regarding race/ethnicity, Indigenous people were the most affected, indicating a possible relationship with socioeconomic and cultural factors. Understanding these data is crucial for developing strategies, such as public policies and programs, aimed at reducing the observed rates.

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Analysis of prevalence and vaccination coverage against bacterial meningitis in Brazil from 2013 to 2022

Monyque Pinheiro Charchar[1], Rafaela Gruppi Rodrigues[1], Eloah Mourão Corrêa[1], Fabíola de Carvalho Chaves de Siqueira Mendes[1]

[1]Centro Universitário do Pará, Belém PA, Brazil.

Correspondence: Monyque Pinheiro Charchar; Email: mcharchar10@gmail.com.

Background: Meningitis is an inflammatory process of the meninges that can be infectious or non-infectious. Vaccination against its etiological agents is therefore an effective strategy for prevention and better prognosis. **Objective:** To analyze the prevalence of bacterial meningitis and its relationship with vaccination coverage in Brazil between 2013 and 2022. **Methods:** An ecological study was carried out using information available on the Notifiable Diseases Information System (SINAN) of the SUS Information Technology Department (DATASUS). We analyzed data on the number of cases of bacterial meningitis caused by Neisseria meningitidis, Streptococcus pneumoniae, and Haemophilus influenzae, and vaccination coverage for Pentavalent (VPenta), Pneumococcal (VPC), and Meningococcal C (VMC) by region of Brazil from 2013 to 2022. **Results:** The average vaccination coverage from 2013 to 2022 was 88.9%, 87.4%, and 84.3% for PCV, CMV, and VPenta, respectively. When analyzing coverage by year, in 2020 and 2021, the PCV and CMV vaccines showed a drop in coverage (below 80%) in most regions of Brazil, except for the South and Central-West regions only in 2020. As for VPenta, there was a drop in 2019 and 2021 in all regions, with average coverage of 71%, improving only in the Southern region in 2021 (80.8%). Regarding the prevalence of meningitis, there was a decrease in all regions when comparing the years 2019 (total of 16,684 cases), 2020 (7,076), and 2021 (6,867), with an increase in 2022 (12,238). Thus, considering the coincidence of low vaccination coverage and prevalence of bacterial meningitis with the most critical period of the SARS-CoV-2 pandemic, it is hypothesized that this may justify the low demand for health services, underreporting, as well as social isolation contributing to the reduction in contagion. It should also be noted that, as there was an increase in the number of cases in all regions in 2022, the hypothesis of the effect of social isolation during the pandemic is reinforced, as this year the distancing rules were less strict. On the other hand, vaccination coverage did not increase to the same extent in some regions, such as in the North of the country, where average coverage remained below 80% for all three vaccines, and in the Northeast and Southeast regarding CMV. **Conclusion:** The years 2020 and 2021 recorded low vaccination coverage and a reduction in the prevalence of bacterial meningitis, findings that may be related to the COVID-19 pandemic. However, in 2022, there was no significant resumption of coverage in some regions, suggesting that other factors interfered with vaccination. This highlights the need for the government to address issues negatively impacting immunization rates, such as flaws in production and distribution processes, unequal access between Brazilian regions, social inequality, and increasing misinformation in society.

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Analysis of spina bifida hospitalizations in the northeast region of Brazil over the last ten years

Hálisson Ferreira Freitas Filho[1], Kaliana Kennedy Oliveira Calixto[1], Larissa Suzan Basilio e Silva[1], Felipe José Silva e Silva[1], Luan Víctor Silva Ribeiro[2], João Victor Souza de Barros[1], Igor Moreira Miguez Godoy[3], Izabella Carolline Menescal Ferreira[1], Antônio Marcos da Silva Henriques[1], Nathália Coimbra Coêlho[1]

[1]Universidade Federal do Pará, Belém PA, Brazil.
[2]Afya Faculdade de Ciências Médicas
[3]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.

Correspondence: Hálisson Ferreira Freitas Filho; Email: halisson.filho@ics.ufpa.br.

Background: Spina bifida is a congenital anomaly caused by failures in the neural tube closure mechanism during the fourth week of gestation. This condition can affect the spinal cord and meninges, leading to a range of neurological and orthopedic complications. The Northeast is the Brazilian region with the highest prevalence of hospitalizations due to this congenital malformation in the past ten years. **Objective:** To delineate the epidemiological profile of hospitalizations of patients with spina bifida in Northeast Brazil over the last ten years. **Methods:** This is a retrospective study using data obtained through the Department of Informatics of the Unified Health System (DATASUS), regarding hospitalizations for spina bifida in Northeast Brazil, between January 2014 and December 2023. **Results:** From the analysis of this period, a total of 4285 hospitalizations and 108 deaths were recorded in the Northeast region, representing 40.4% of hospitalizations and 44.8% of deaths due to spina bifida in Brazil. Among the states in the region, Bahia accounted for 29.1% of hospitalizations, followed by Maranhão with 20.8%. The highest number of hospitalizations occurred in 2015, with 15% of the total, showing a gradual decrease in the following years. Regarding age groups, individuals under 1 year old represented the age group with the highest number of hospitalizations (62.5%) and deaths (74.0%). Concerning sex, males predominated with 50.8% of hospitalizations and 55.5% of deaths. In terms of mortality rate, Alagoas had the highest rate (6.03), followed by Maranhão (4.36). The white race/color group was the most affected, with a mortality rate of 4.17. **Conclusion:** The data on hospitalizations and deaths reveal regional inequalities and vulnerabilities in the country. It is necessary to implement public policies with investments focused on improving health infrastructure and technologies for early detection and treatment of spina bifida, as well as on training health professionals for the proper management of patients from birth, in order to reduce the impact of hospitalizations and mortality in individuals under 1 year old.

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Analysis of the correlation between trunk control and quality of life in individuals with post-stroke hemiplegia: a cross-sectional study

Marina Rigobello Muraro[1], Samuel Lidione da Silva[1], Wellington Felipe Conceição[1], Heloísa Freiria Tsukamoto[1]

[1]Centro Universitário Filadélfia, Londrina PR, Brazil.

Correspondence: Samuel Lidione da Silva; Email: lidione67@gmail.com.

Background: A stroke commonly leads to permanent dysfunctions such as hemiplegia/hemiparesis. These functional alterations significantly compromise the quality of life (QoL), potentially causing social and emotional impairment for affected patients. With the lesion, the motor incapacity of synergistic trunk muscles interferes with the execution of activities of daily living (ADLs), weakening the patient's subjective well-being. Literature results have shown that interventions aimed at improving trunk movement functionality favor an improvement in QoL, but it is still unclear whether trunk control is directly correlated with QoL in post-stroke individuals. **Objective:** To analyze the correlation between trunk control and aspects of QoL in individuals with post-stroke hemiplegia. **Methods:** This is a cross-sectional study with a quantitative approach, with a convenience and intentional sample composed of 16 individuals. The evaluation of trunk control and QoL variables was performed using the following questionnaires: Trunk Impairment Scale (TIS) and Stroke Impact Scale (SIS). Pearson and Spearman correlation analyses were used, according to data normality, considering a significance level of 5%. This study was approved by the Research Ethics Committee (CAAE: 67926323.2.0000.5217). **Results:** Participants had a mean age of 57±12.7 years. The final scores of the questionnaires were: TIS total 16.5±2.9 and SIS total 64.2±8.9. Statistically significant correlations were found in the following variables: with a moderate effect between the total scores of SIS and TIS (p=0.018 and r=0.582), in the ADLs domain and physical problems (p=0.004; r=0.674), and total TIS with the ADLs domain (p=0.003; r=0.685). For the SIS total and the ADLs domain variables, there was a strong correlation (p=0.00001; r=0.867). **Conclusion:** The lack of motor control in trunk movement adversely impacts the quality of life (QoL) in post-stroke individuals, with

a statistically significant correlation observed between these factors in this study's sample. Understanding these results underscores the importance of early clinical focus on addressing trunk control issues to enhance the subjective well-being prognosis for these individuals. Further research with larger sample sizes and objective measures of trunk control outcomes is necessary to validate the data presented.

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Analysis of the effects of melatonin in astrocytes in a mouse model of Parkinson's disease

José Victor Ribeiro Silva Gomes[1], Gustavo Amorim Alves[1], Ana Paula Martins Inácio[1], Kananda Oliveira Garcia Ruiz[1], Miguel Henrique dos Reis[2], Mônia Léia Martins[1], Ana Lívia Teixeira[2] , Gustavo Schiavoni de Almeida[1], Liliane Vanessa Costa Pereira Mendes[1], Laila Cristina Moreira Damazio[1]

[1]Universidade Federal de São João del-Rei, São João del Rei MG, Brazil.
[2]Centro Universitário Presidente Tancredo de Almeida Neves, São João del Rei MG, Brazil.

Correspondence: José Victor Ribeiro Silva Gomes; Email: josevrsgomes@gmail.com.

Background: Parkinson's disease (PD) is the second most common neurodegenerative disease in the world, only behind Alzheimer's. The etiology of PD is unclear, and the major pathological feature found in the brain is the loss of dopaminergic neurons in the substantia nigra (SN), part of the midbrain. The causes of those deaths are related to oxidative stress, mitochondrial dysfunction and protein abnormalities. Besides the neuronal cells, recent findings show an important role of glial cells in the development of neurodegenerative diseases. Among them, astrocytes are the most abundant and act, primarily, through a process called reactive astrogliosis in reaction to an abnormal or pathological state. In this process, astrocytes can change the brain environment, releasing cytokines and inflammatory mediators which, in dysfunctional situations, can damage and even kill neurons and other glial cells. In this context, melatonin can be used to attenuate this process, once its role in reducing oxidative stress and diminishing neuronal deaths has been shown. In addition, recent findings suggest that melatonin may play a role in astrogliosis. **Objective:** Evaluate the effects of melatonin in the population of astrocytes in a mouse model of Parkinson's disease. **Methods:** 32 male Wistar mice were used in the research, divided in 4 groups: submitted to an induction surgery of PD through the application of a toxin (6-OHDA) in the striatum or a sham surgery (control) and intraperitoneal injections of melatonin (20 mg/kg) or saline (0.5 mL/100 g, control) 5 days a week during 5 weeks before the surgery. One week after the surgery, the animals were euthanized and samples of their brains were used to make microscope slides for immunohistochemical analysis. Statistical analysis was performed using ANOVA one-way, p< 0.05. **Results:** We analyzed the process of astrogliosis comparing the quantity of astrocytes in the samples of each group. The count was made in two areas of the slide at 40x magnification. Two proteins were targeted in immunohistochemistry: GFAP (Glial Fibrilar Acid Protein) and vimentin. We evaluated astrogliosis in two regions of the mouse brain: SN and striatum. Although the astrogliosis was higher in the intervention group in all the analyses made, there was no statistically significant difference among the groups. The analysis made by GFAP in SN showed less astrogliosis in the control group (p=0.035). A larger number of astrocytes were identified using GFAP rather than using vimentin. **Conclusion:** Astrogliosis is an important process in the healthy brain. However, changes in it can contribute to the development of neurodegenerative diseases. Melatonin, as an antioxidant agent, may prevent the emergence of such damage. However, in this research, we did not find enough evidence to support the idea of a protective role of melatonin in PD. Further studies are necessary to elucidate this issue fully.

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Analysis of the epidemiological profile of epilepsy in the Southeastern region of Brazil over the last five years

Marcelo Pereira de Souza[1], Bianca Amado Cardozo[1]

[1]Universidade Nove de Julho, Osasco SP, Brazil.

Correspondence: Marcelo Pereira de Souza; Email: marsouzz@hotmail.com.

Background: Epilepsy is a functional condition of the brain that results in epileptic seizures caused by an imbalance between the concentrations of the neurotransmitters GABA and glutamate. In the last five years, this condition was responsible for 28.54% of all hospitalizations for diseases of the nervous system in Brazil. Based on this result, it is important for a neurologist to know how to recognize epileptic events and treat refractory ones. **Objective:** To analyze the Brazilian epidemiological profile in the Southeast Region in relation to the prevalence of epilepsy cases associated with epileptic seizures. **Methods:** This is an ecological, retrospective, quantitative and descriptive study, whose data was obtained from consultations carried out on the Sistema de Informações Hospitalares (SIH/SUS), through the DATASUS platform, for the period from January 2019 to December 2023. The Southeast Region and its respective states were analyzed. **Results:** In relation to the Southeast region, this condition is present at all ages, with a distribution of 5,921 cases in Espírito Santo, representing 28.7% of total hospitalizations for nervous system diseases in the region (20,622); 33,885 in Minas Gerais, totaling 29.4% of hospitalizations (115,186); 14,492 in Rio de Janeiro, making up 26.1% of the total (55,484 hospitalizations) and 60,350 in São Paulo, corresponding to 27.6% (219,001 hospitalizations). In terms of age group, the prevalence is highest between 1 and 9 years old, which represents a total of 21.1% of hospitalizations in the region (24,196). Furthermore, it is clear that there is a significant decrease from the age of 10 in all the states in the region. **Conclusion:** Based on the analysis of this study, we can see the significant number of neurological hospitalizations associated with epilepsy, especially in children aged between 1 and 9 years, in Brazil over the last five years, with the southeast region standing out.

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Analysis of the epidemiological profile of hospitalizations for epilepsy in Brazil between 2019 and 2023

Thairon Guilherme de Santana[1], Victória Maria Rabelo Lopes[1], Rayane Eunice Brandão de Lima[1], Gabriella de Oliveira Barboza[1], Maria Eduarda Garcia Marvulle[1]

[1]Universidade de Marília, Marília SP, Brazil.

Correspondence: Thairon Guilherme de Santana; Email: thaironguilherme@hotmail.com.

Background: Epilepsy is characterized as a chronic neurological disease associated with episodes of seizures resulting from spontaneous and abnormal electrical discharges of neurons. In addition, psychiatric comorbidities, such as depression and attention deficit hyperactivity disorder, are often associated with this pathology, which can have a significant impact on the quality of life of these individuals. **Objective:** This study aims to analyze the epidemiological profile of hospitalizations for epilepsy in Brazil between 2019 and 2023. **Methods:** This is an observational, descriptive, cross-sectional and retrospective study. We used data from the Department of Informatics of the Unified Health System (DATASUS) on hospitalizations for epilepsy in Brazil between 2019 and 2023. The variables used to classify the epidemiological profiles were: gender, age group and color/race. Cases that did not fit these variables were excluded. **Results:** Between 2019 and 2023 there were 239,393 cases of hospitalization due to epilepsy in Brazil, of which 42.18% (100,980) were female and 57.82% (138,413) male. Among male hospitalizations, there were more cases among brown men under the age of 20 with 34,693 (25.06%) cases, followed by brown men aged between 20 and 59 with 28,728 (20.75%) hospitalizations, and the third most frequent epidemiological profile among males was white men aged between 20 and 59 with 19,865 (14.35%). Among female hospitalizations, there were more brown women under the age of 20 with 27,400 (27.13%) cases, the second most common profile was white women under the age of 20 with 17,320 (17.15%), followed by brown women aged between 20 and 59 with 16,214 (16.05%) hospitalizations. **Conclusion:** Based on the analysis of the data, it can be concluded that the three main epidemiological profiles with the most hospitalizations due to epilepsy are brown men under 20 years of age and between 20 and 59 years of age, and brown women under 20 years of age, showing that with advancing age hospitalizations for this reason are decreasing.

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Analysis of the epidemiological profile of migraine cases in Brazil between 2018 and 2023

Gabriella de Oliveira Barboza[1], Maria Eduarda Garcia Marvulle[1], Victória Maria Rabelo Lopes[1], Thairon Guilherme de Santana[1]

[1]Universidade de Marília, Marília SP, Brazil.

Correspondence: Gabriella de Oliveira Barboza; Email: gabriellaobz@gmail.com.

Background: Migraine is a type of primary headache, characterized by intense and pulsating pain, which may also be accompanied by photophobia, nausea, vomiting and other symptoms. The main triggers of this headache are stress, exposure to screens, sudden changes in temperature, poor quality of food and sleep, as well as anxiety and depression. Furthermore, it is a highly disabling and very prevalent condition worldwide, especially among women in the job market, and is an important cause of work absenteeism. Furthermore, because migraines are poorly understood and accepted in the workplace, many do not notify employers about this issue, resulting in the individual remaining unproductive in the workplace, a fact that generates important financial losses. **Objective:** This work aims to analyze the epidemiological profile of hospitalizations for migraine in Brazil, among individuals in the labor market, between the years 2018 and 2023. **Methods:** This is an epidemiological, retrospective, observational and descriptive study, using data from the Department of Informatics of the Unified Health System (DATASUS) from January 2018 to December 2023. The variables used were gender, age range from 20 to 69 years old, year of care and region. **Results:** The region with the highest prevalence of migraine cases was the northeast, followed by the southeast region, which together correspond to approximately 59.14% of the total. Furthermore, women experience more than double the rate of cases compared to men. Furthermore, there is also a higher frequency between the ages of 30 and 39 and a lower frequency between 60 and 69 years old. **Conclusion:** Based on the data presented, it is observed that, in fact, the prevalence of migraine is higher among women of reproductive age, a fact explained mainly by female neuroendocrine factors. Furthermore, the high frequency of cases in the northeast of Brazil may be related to the greater incidence of sunlight

in the region, the population's exposure to more intense temperatures and less access to specialized treatment health services. Finally, migraine is a health problem that generates both increased costs in the health sector and losses in the quality of life and work productivity of the Brazilian population, demonstrating the need for interventions mainly related to worker health.

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Analysis of the incidence of hospitalizations and deaths due to cerebral palsy and other paralytic syndromes in Brazil from 2019 to 2023

Lucas Charuri de Andrade Castello Branco[1], Barbara Zorzi Sanfins[1], Francisca Isabelly Leite de Almeida[2], Moisés Cardoso Portilho[3], Fernando Gabriel Rodrigues Baia[4], Karolliny Amador Kzam[3], Vinícius Adriano Coelho Amaral[4], Bruno Kauê Rodrigues Vilhena[4], Paola Carolina Santos Foinkinos[4], Luan Nascimento Pereira de Amorim[4]

[1]Faculdade São Leopoldo Mandic, Campinas SP, Brazil.
[2]Centro Universitário Unifacisa, Campina Grande PB, Brazil.
[3]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.
[4]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Lucas Charuri de Andrade Castello Branco; Email: julucascastellobranco@gmail.com.

Background: The Chronic Non-Progressive Encephalopathy, better known as cerebral palsy, is the most common disability in childhood and comprises a group of motor disorders due to permanent and non-progressive injuries to the fetal or childhood nervous system. In Brazil, a country with great heterogeneity and inequality in access to healthcare, the distribution of cerebral palsy is not homogeneous throughout the territory, which justifies carrying out epidemiological studies to analyze this disease in the country. **Objective:** Analyze the epidemiological profile according to hospitalizations and deaths from cerebral palsy and other paralytic syndromes in Brazil from 2019 to 2023. **Methods:** This is a quantitative, retrospective and cross-sectional study developed using data obtained from the Ministry of Healths Hospital Information System at DATASUS/TABNET, considering the variables: region, age group, gender and character of care of hospitalizations and deaths from cerebral palsy and other paralytic diseases in the period from 2019 to 2023. **Results:** A total of 39,118 hospitalizations for cerebral palsy and other paralytic syndromes were observed during the period, with the Southeast region having the highest percentage (45.24%), specifically the state of São Paulo (7,718), followed by the Northeast (24.2%) and Midwest (21.33%) regions, with a predominance of male patients aged between 20-29 years, with elective care, a profile similar to that of the other regions. Of all the deaths, there were 704 from the disease in the period, with the Southeast region accounting for 59.51% of cases, with a profile of male patients, but in the 50-59 age group, although in all states the most affected age group is the 60-69 age group, due to senility, predisposition to atherosclerotic events and hormonal factors. With regard to the mortality rate, the state of Mato Grosso do Sul has the highest rate for cerebral palsy and other paralytic syndromes (8.43), although the southern region leads the way (3.32). **Conclusion:** The study reveals that the majority of hospitalizations for cerebral palsy and other paralytic syndromes occurred in the Southeast, particularly in the state of São Paulo (7,718). Predominantly, the patients were young men (20-29 years old) and the care was elective, i.e. non-emergency, indicating that the hospitalizations were planned or programmed, which suggests a significant need for continuous and specialized care for this age group and demographic profile. As far as deaths are concerned, the Southeast again leads the prevalence (59.51%), although there is no relationship with the number of hospitalizations, since the age group of deaths is 50-59 years. These findings highlight the need for public health strategies focused on prevention, early diagnosis and appropriate management of the complications associated with these conditions, especially in the regions and age groups most affected.

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Analysis of the incidence of Parkinson's disease inpatients and deaths in brazil during the years 2019 to 2023

Paola Carolina Santos Foinkinos[1], Karolliny Amador Kzam[2] , Matheus Prado dos Santos[1], Fernando Gabriel Rodrigues Baia[1], Vinicius Adriano Coelho Amaral[1], Bruno Kauê Rodrigues Vilhena[1], Luan Nascimento Pereira de Amorim[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.
[2]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.

Correspondence: Paola Carolina Santos Foinkinos; Email: paola.csfoinkinos@aluno.uepa.br.

Background: Parkinson's disease is a degenerative disease of the central nervous system, characterized by motor, cognitive and autonomic alterations, which starts insidiously and causes socio-functional impairments, especially in the elderly population. Research shows that there is currently an increase in the mortality rate of the disease and that it could become a public health problem, due to the trend towards an ageing population in Brazil. Studies into the dynamics, course and impact of Parkinson's disease in the country are needed. **Objective:** To analyze the epidemiological profile of hospitalization and death rates due to Parkinson's disease in Brazil between 2019 and 2023. **Methods:** This is a cross-sectional, retrospective and quantitative study, in which secondary data were collected from the Department of Informatics of the Unified Health System (DATASUS), from the years 2019 to 2023. The following variables were analyzed: authorizations for hospital admissions, mortality rate, number of cases and average length of stay in hospital institutions. **Results:** There were a total of 4401 hospitalizations for Parkinson's disease in Brazil, from 2019 to 2023, with 46.76% of hospitalizations in the Southeast region, with emphasis on the state of São Paulo (22.47%). The average length of stay was 13.4 days. The longest average length of stay (33.4 days) was recorded in the state of Rio de Janeiro. The mortality rate was 7.84%, being more significant in the North region (10.29%), with emphasis on the state of Roraima (33.33%). **Conclusion:** Analysis of the study showed that the Southeast region accounts for almost half of all hospitalizations for Parkinson's disease in Brazil and that hospital stays require considerable maintenance costs, given the average length of stay of 13 days, which can reach a month in certain states. In addition, it is clear that less developed regions face obstacles in the management of Parkinson's disease, since it has a considerably higher mortality rate than the national average. It is therefore necessary to develop local measures to mitigate regional disparities with regard to the appropriate treatment of Parkinson's, with the aim of reducing the length of hospitalization and the number of deaths.

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Analysis of the morbidity and mortality of muscular dystrophies in Brazil: 2019 to 2023

Lucas Charuri de Andrade Castello Branco[1], Barbara Zorzi Sanfins[1], Francisca Isabelly Leite de Almeida[2], Moisés Cardoso Portilho[3], Luan Daher Fernandes[4], Luiza da Paz de Souza[4], Suzana Gomes de Oliveira[4], Arley Nikson Oliveira Barreto[4], Luiz Carlos Figueiredo Filho[4], Luan Nascimento Pereira de Amorim[4]

[1]Faculdade São Leopoldo Mandic, Campinas SP, Brazil.
[2]Centro Universitário Unifacisa, Campina Grande PB, Brazil.
[3]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.
[4]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Lucas Charuri de Andrade Castello Branco; Email: julucascastellobranco@gmail.com.

Background: Muscular dystrophies are neuromuscular diseases characterized by structural alterations or functional deficiency of the muscles. They fall into the group of myopathies, which are degenerative and hereditary diseases of genetic cause, transmitted as autosomal dominant, recessive or X-linked traits. The main clinical manifestations are: weakness, progressive degeneration of skeletal muscles, muscle fatigue and tiredness, atrophy, myotonia and cardiac and respiratory impairment, with fatigue being the symptom that most generates functional incapacity in the patient. Despite the fact that there is no cure for this condition, treatments are available from the health system to improve the patients quality of life. In this sense, it is important to evaluate the variables related to the treatment of this pathology. **Objective:** To analyze the morbidity and mortality of muscular dystrophies in the last 5 years (2019-2023) in Brazil. **Methods:** This is a cross-sectional, retrospective and quantitative study, in which data was collected from the Department of Informatics of the Unified Health System (DATASUS), referring to the years 2019 to 2023. Secondary data was collected from the SUS Hospital Morbidity database (SIH/SUS). **Results:** In the period considered, a total of 2,903 hospitalizations authorized for the treatment of muscular dystrophies were found in Brazil, all of which were of medium complexity. Among the regions, the Northeast predominated (38.04%), followed by the Southeast (37.13%), which also had a substantial number of hospitalizations. Of the total number of hospitalizations, the majority (76.6%) are urgent, while the remainder (23.3%) are elective. Finally, of the 2,903 hospitalizations, there were 68 deaths, which corresponds to a total mortality rate of only 2.34% associated with the clinical condition described. **Conclusion:** Therefore, based on the data analyzed about hospitalizations authorized for the treatment of muscular dystrophy, it not only affects secondary care, but also primary care, given the counter-referral that occurs in PHC. In addition, these hospitalizations occur differently throughout Brazil, highlighting the country's continental characteristics. For example, the Northeast Region concentrates the cases of hospitalizations, and the classification of these hospitalizations differs between elective and emergency cases. There was a recovery rate of 97.66% of the people who were hospitalized, meaning that the treatment of this problem is being efficient and promoting the reduction of these cases, which denotes a total of more than 2,500 people who suffered from these hospitalizations. It is important to note that the medium-complexity infrastructure, as well as the working conditions of health professionals, interfere with the outcome of hospitalizations of patients with muscular dystrophy.

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Analysis of the prevalence of diagnoses and age group impact of brain cancer in Brazil over the last 5 years

Bernardo Caetano Novaes[1], Guilherme Galardino Santos[1], Samuel José Feres Ozava[1], Victor Bruno dos Santos Teixeira[1], Heloisa Magda Resende[1]

[1]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.

Correspondence: Bernardo Caetano Novaes; Email: bernardo16.novaes@gmail.com.

Background: Brain tumors include various malignant neoplasms that can originate from different types of cells in the central nervous system. They can be primary, developing within the brain tissue itself, or secondary, resulting from metastases of other neoplasms. The global incidence of the primary tumors is low, with approximately 3 cases per 100,000 people. Although rare, the prevalence of these tumors has been increasing in recent decades. They are highly incapacitating and have a high morbidity and mortality, with a 5-year relative survival rate of only 37%. **Objective:** To calculate the prevalence of brain cancer diagnoses over the last 5 years in the federative regions of Brazil and to analyze which age group is most affected. **Methods:** The methodology of this study involved data collecting from the Oncology Panel of DATASUS (Department of Information and IT of the Unified Health System) from 2019 to 2023 and population data from the 2022 IBGE (Brazilian Institute of Geography and Statistics) census to calculate prevalence. The age group analysis was conducted based on data from the Oncology Panel of DATASUS, defining the age groups as: 0-19 years; 20-39 years; 40-59 years; 60-79 years; and over 80 years. **Results:** This data collecting resulted in 29,958 cases of brain cancer diagnosed in Brazil. The federative region with the highest prevalence (per 100,000 inhabitants) was the South (21.50), followed by the Southeast (13.07), Central-West (13.03), Northeast (8.85), and North (6.49) regions. Regarding the most affected age groups, in the North region over the last 5 years: 0-19 years (330); 20-39 years (304); 40-59 years (519); 60-79 years (310); over 80 (16). In the Northeast: 0-19 years (946); 20-39 years (1,131); 40-59 years (2,172); 60-79 years (1,425); over 80 (48). Southeast: 0-19 years (1,758); 20-39 years (1,813); 40-59 years (4,386); 60-79 years (4,359); over 80 (169). South: 0-19 years (787); 20-39 years (1,071); 40-59 years (2,802); 60-79 years (2,703); over 80 (119). Central-West: 0-19 years (423); 20-39 years (465); 40-59 years (1,042); 60-79 years (830); over 80 (30). Thus, it can be observed that in Brazil, the most affected age group is 40-59 years. **Conclusion:** Based on the data collected over the last five years, the highest prevalence was observed in the South region (21.50/100,000 inhabitants) and the lowest in the North region (6.49/100,000 inhabitants). The most affected age group was 40-59 years in all regions, with 4,386 cases in the Southeast, 2,802 in the South, 2,172 in the Northeast, 1,042 in the Central-West, and 519 in the North. These results indicate a higher prevalence of cases in middle-aged individuals (40-59 years) and significant regional variation, suggesting the need for analysis of factors contributing to the development of brain cancer. Prevention and early diagnosis strategies are necessary nationwide, aiming to ensure equitable access to quality healthcare across all regions.

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Analysis of the prevalence of tuberculous meningitis in the state of São Paulo between 2012 and 2022

Gabriel Pereira Silva Rocha[1], Letícia César de Oliveira[2] , Sofia Pereira Figueiredo[3], Mirella Simini Santolim[4], Michely Laiany Vieira Moura[5]

[1]Universidade de Mogi das Cruzes, Mogi das Cruzes SP, Brazil.
[2]AFYA Faculdade de Ciências Médicas, Palmas TO, Brazil.
[3]Centro Universitário Mauricio de Nassau de Barreiras, Barrareias BA, Brazil.
[4]Universidade Nove de Julho, São Paulo SP, Brazil.
[5]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Gabriel Pereira Silva Rocha; Email: irineu33rocha@gmail.com.

Background: Meningitis is a morbidity that can lead to serious consequences for the infected person. Among the various etiologies, tuberculous meningitis stands out. It occurs when the Mycobacterium tuberculosis bacillus spreads from its primary site of infection to the Central Nervous System which can cause severe neurological damage, with a mortality rate of approximately 50%. It is therefore highly relevant to learn about the prevalence of such a serious disease, which is caused by the complication of a public health issue in Brazil and worldwide, such as tuberculosis. **Objective:** To describe the prevalence of tuberculous meningitis cases in the state of São Paulo, Brazil, from 2012 to 2022. **Methods:** It is characterized as an ecological, cross-sectional, descriptive and retrospective study of an epidemiological nature, supported by the database of the Department of Informatics of the Unified Health System (DATASUS), with information provided by the Notifiable Diseases Information System (SINAN). An analysis was conducted of confirmed cases of meningitis caused by Mycobacterium tuberculosis in the state of São Paulo between 2012 and 2022. The variables were reported using descriptive statistics. The electronic search was carried out employing the term "Tuberculous Meningitis" and "Brazil". **Results:** Between 2012 and 2022, 546 cases of meningitis caused by Mycobacterium tuberculosis were reported in the state of São Paulo. The highest prevalence among the cities in the state was recorded in Pariquera-Açu, with a rate of 0.0253%, 5 confirmed cases and a population of 19,723 inhabitants in the time period evaluated. The second highest incidence was detected in Louveira, with a rate of 0.0020%, followed by Ribeirão Preto, which recorded a rate of 0.0087% between 2012 and 2022. The low prevalence rates in the analyzed cities are justifiable by the fact that tuberculous meningitis is the most severe complication of all forms of tuberculosis, as well as by the decline in its lethality since 1945 with the discovery of chemotherapy drugs specific for tuberculosis. However, it is necessary to carry out a prevalence analysis with the aim of promoting the monitoring of notified cases, especially in the state of São Paulo, which has a population of 44.04 million during the period of analysis of the study, and is thus classified as a large center susceptible to the spread of the disease. **Conclusion:** Tuberculous meningitis has presented a reduced prevalence rate in the cities of the state of São Paulo, although tuberculosis continues to be a public health problem in Brazil and to affect patients who, once they are carriers of the Mycobacterium tuberculosis bacillus, are susceptible to developing a severe form of tuberculous meningitis. It is necessary to constantly monitor the prevalence of this comorbidity in the state of São Paulo in order to keep reported cases of the disease under control.

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Analysis of the profile and therapeutic response of Parkinson's disease patients undergoing deep brain stimulation

Rhaymysom Jasmy Gomes Abreu[1], Rafaella Resplande Xavier[1], Beatriz Fulador[1], Helen Luiza Silvestrini[1], Letícia Lage Silva[1], Hingrid Cristine Arruda de Oliveira[1], Albérico Rocha Lima Neto[1], Victor Hugo de Souza Silva Gomes[1], Heloise Helena Siqueira Borge[1]

[1]Hospital Geral, Cuiabá MT, Brazil.

Correspondence: Beatriz Fulador; Email: beatriz_fulador@hotmail.com.

Background: Parkinson's disease (PD) is a neurodegenerative disorder characterized by cardinal motor symptoms such as bradykinesia, resting tremor, rigidity, and postural instability. Its pathophysiology results from degeneration of the nigrostriatal system, leading to decreased dopamine production. Deep Brain Stimulation (DBS) offers an innovative approach to managing the debilitating symptoms of PD. DBS involves implanting electrodes in specific areas of the brain, which receive stimulation through electrical currents, modulating neurotransmitters involved in the disease's etiopathogenesis. Proper patient selection for DBS must follow precise indications. **Objective:** To evaluate the effectiveness of deep brain stimulation indications in a reference service based on the response of motor symptoms in assisted patients. **Methods:** This is a retrospective study with data from patients with Idiopathic PD who underwent DBS from 2022 to 2024. Data were obtained from medical records, including variables such as sex, age, Mini-Mental State Examination (MMSE), Geriatric Depression Scale (GDS), Unified Parkinson's Disease Rating Scale (UPDRS) score, and levodopa (Prolopa) dosage. **Results:** The sample consisted of 7 patients, with an M:F ratio of 1:1.3, and a mean age of 55 years (50-61). Regarding the disease duration and DBS implantation, 57% of patients underwent surgery between the 4th and 6th year of the disease, and the rest between 7 to 10 years. Through the analysis of the MMSE, a screening and follow-up tool for cognitive impairment, it was found that 4 patients (57.14%) had preserved cognitive function, i.e., MMSE score ≥ 27, 2 patients (28.57%) had mild cognitive impairment (21-24), and 1 (14.28%) was classified with moderate cognitive impairment (10-20). Regarding the Geriatric Depression Scale, only 2 showed mild depression, while the others had no psychiatric abnormalities. All individuals in the analysis showed a reduction in the UPDRS percentage when compared before and after DBS implantation, with a variation between 45.45% and 88.88%, and an average reduction of 68.1%. Regarding the use of Prolopa, a significant reduction in dosage was observed. Before DBS implantation, patients used an average of 528 mg/day of Prolopa. Post-DBS, an average reduction of 32.38% in dosage was observed. **Conclusion:** The application of scales prior to DBS implantation aims to guide the indication. These yielded positive results for individuals with PD. Consequently, it is concluded that to achieve the expected outcomes, it is essential to establish a routine of continuous patient evaluation for correct and precise indications, thereby obtaining better results for the patients.

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Analysis of the profile of demyelinating and autoimmune diseases at the first specialized referral center in the state of Alagoas

Túlio Marlus Castro Lucena[1], Assis Porfirio Furtado Nogueira[1], Vitor Gustavo Leão Souto[2], Samyra Melo Vital[3], Patrícia Pereira Nunes Ribeiro[1], Maria Júlia Tabosa de Carvalho[2], Geovane Brito Affonso[2], Érika Santos Machado[1], Darlan de Araujo Benevides[1], Nayra Roberta Sales Salvador[1]

[1]Hospital Geral do Estado, Maceió AL, Brazil.
[2]Hospital Universitário Professor Alberto Antunes, Maceió AL, Brazil.
[3]Hospital Metropolitano de Alagoas, Maceió AL, Brazil.

Correspondence: Túlio Marlus Castro Lucena; Email: tuliomarlus.neuro@gmail.com.

Background: In Maceió, the first specialized service for demyelinating and autoimmune diseases in the state of Alagoas was implemented for the screening, accurate diagnosis, appropriate treatment, and follow-up of affected individuals. **Objective:** To evaluate the epidemiological profile of patients treated at the referral outpatient clinic for demyelinating and autoimmune diseases in the state of Alagoas. **Methods:** A review of the medical records of patients seen at the service from its inauguration in August 2022 until June 2024 was conducted. Patients under investigation without a confirmed diagnosis were excluded from the analysis. **Results:** Currently, 65 patients with confirmed diagnoses are being followed at the demyelinating and autoimmune diseases outpatient clinic. Of these, 33 patients have been diagnosed with Multiple Sclerosis, including 24 women and 9 men, representing more than 50% of the clinic's patients, which aligns with global epidemiology as it is the most common demyelinating disease. Additionally, the proportion of approximately 70% women among those affected also

corroborates the literature on the disease. Among other demyelinating and autoimmune diseases, 13 patients are followed for Neuromyelitis Optica, with 8 women and 5 men. Regarding inflammatory myelitis of different causes, 13 patients are being followed, including 8 women and 5 men. Four patients are being followed for inflammatory optic neuritis, with 2 women and 2 men. One male patient is being followed for autoimmune encephalitis, and one male patient is being followed for MOG antibody-associated disease. **Conclusion:** Demyelinating and autoimmune diseases have always represented a diagnostic challenge within neurology. Therefore, the presence of specialized centers for accurate diagnosis and appropriate follow-up of patients is of utmost importance, providing earlier diagnosis, better care, and improved quality of life for patients affected by these diseases.

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Analysis of the profile of hospitalizations due to multiple sclerosis in the sus in 2023

Vitória Felice Camargos Muller[1], Vinícius Felice Camargos[2] , Thamyrez Quézia de Assis[1], Pedro Bruno Paixão Ribeiro[1], Ana Carolina Nascimento de Sousa[1], Thiago Cardoso Vale[1], Rodrigo Lima Vieira[3]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[2]Centro Universitário IMEPAC, Araguari MG, Brazil.
[3]Universidade do Estado do Pará, Santarém PA, Brazil.

Correspondence: Vitória Felice Camargos Muller; Email: vitoriafelicec@gmail.com.

Background: Multiple sclerosis (MS) is an immune-mediated inflammatory demyelinating disease of the central nervous system that mainly affects women aged between 28 and 31 years. The progression of neurological disability in MS is highly variable. Its impact on individuals varies according to the severity of signs and symptoms, relapse frequency, rate of worsening, and residual disability. **Objective:** To assess the profile of hospitalizations for MS in the SUS (Brazilian Unified Health System) in the year 2023. **Methods:** The study consists of a retrospective and descriptive analysis of epidemiological data of patients hospitalized for MS in the SUS in 2023. The database used was the TABNET - DATASUS platform. The eligibility criteria included patients who were in SUS healthcare services, in the hospital admission modality, with ICD-10 (International Classification of Diseases) specified as MS. Regarding data representation, they were tabulated in Microsoft Excel, and inferential statistics were performed using BioEstat Software (Version 5.3). Due to the qualitative nature of the correlated data (gender, age group, type of hospitalization, and race), the Chi-Square test, with a significance level of 5%, was used. **Results:** During the year 2023, there were 1,369 hospitalizations for MS in the SUS. Of these, 938 (68.5%) were female. Regarding the type of hospitalization (TH), 688 (50.3%) patients were treated as emergencies. In terms of color/race, 700 (51.1%) were white, while 601 (43.9%) were brown. The most prevalent age group (AG) for hospitalizations was between 30 to 39 years (29.1%), with the majority (54.1%) of these hospitalizations being elective. On the other hand, in the AG between 10 to 19 years, emergency treatments (56.6%) prevailed. Concerning in-hospital mortality, the AG between 60 to 69 years old was the most prevalent (36.3%). Regarding statistical inference, significant correlations were found between being brown and emergency hospitalization ($p < 0.0001$), being white and elective hospitalization ($p < 0.0001$), AG (over 60 years) and mortality ($p < 0.0001$), and AG (between 30 to 59 years) and emergency hospitalization ($p = 0.0199$). Meanwhile, there were no correlations between AG and emergency hospitalization in those under 30 years ($p = 0.604$) and over 60 years ($p = 0.3222$). **Conclusion:** After a detailed analysis of this study, it is observed that women between 30 to 39 years old were more prevalent, as described in the literature. Despite recent advances in therapeutic options, MS is still a disease that can cause severe conditions requiring emergency hospitalization, especially in younger individuals. In addition, the mortality rate from MS remains significant, particularly among the elderly. Therefore, this research aims to encourage future studies and provide direction both for early diagnosis and treatment of MS, to optimize the quality of care and improve the prognosis for these patients in the SUS.

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Analysis of the profile of patients who died due to post-stroke infection in a reference hospital in Salvador-BA

Ana Beatriz Brazileiro de Sant'anna[1], Joanna Sousa da Fonseca Santana[1], Amanda Hellen Sousa Leal[1], Ana Clara de Lemos Guimarães[1], Maria Clara Bastos de Sá[1], Giovanna Souza da Conceição Brito da Silva[1], Fabiana Oliveira de Araujo[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Ana Clara de Lemos Guimarães; Email: anaguimaraes21.1@bahiana.edu.br.

Background: Stroke affects approximately 16 million patients annually, leading to sequelae and complications. Bacterial infections are the most common consequences of this condition and can exacerbate patient recovery post-stroke. Therefore, identifying high-risk groups for infections can expand knowledge about the topic and promote specialized care for this group. **Objective:** To analyze the patients profile due to post-stroke infection among patients admitted to the emergency department **Methods:** This is a cohort study, observational and retrospective, as a preliminary result from a major stroke study with 6,993 patients. The partial results are based on secondary data collected from electronic medical records of patients admitted to a reference hospital in Salvador, Bahia. The study population comprises patients diagnosed with stroke, aged over 18 years, admitted between 2020 and 2022. The study was analyzed using Excel and SPSS and was approved by the Ethics and Research Committee (CEP). Variables: age, sex, diagnosis [Ischemic Stroke (IS), Hemorrhagic Stroke (HS), Subarachnoid Hemorrhage (SAH), and Central Venous Thrombosis (CVT)], death, length of hospital stay, site of infection and admission window. **Results:** 62 out of 330 patients contracted a post-stroke infection (18.7%). Of those infected, 24 died (35.8%). The average age of the group with infection was 65 years (ranging from 19 to 94 years) with a slightly higher number of cases in women (59.7%). The distribution of stroke types among patients with infection, in ascending order, were: IS (65.7%), HS (22.4%), and SAH (4.5%). However, the highest incidence of infection was in the SAH group (37.5%). The most common comorbidities in patients with infection were hypertension (62.6%), diabetes mellitus (35.8%), and previous stroke (22.3%). The infections most associated with these patients were lower respiratory tract infections (53.7%), upper respiratory tract infections (43.2%), and urinary tract infections (25.3%). The average length of hospital stay was 29 days, and the admission window (time between the onset of symptoms and arrival at the hospital) had an average of 71 hours. **Conclusion:** The profile of patients is balanced between the sexes, with an average age of 65 years. It was more pronounced in cases of IS but prevalent in SAH. The most frequent infection sites (respiratory infection and urinary tract infections) align with what the literature reviews already identify as expected. The wide variation in the admission window and a longer length of hospital stay compared to the average of the sample and represented about 1/5 of the deaths in this study. Thus, there is a need for intensive clinical surveillance, preventive strategies for patients with comorbidities in order to improve clinical practices in these cases.

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Analysis of the strategies involved in the management of epilepsy in the hospital emergency environment

Rafael Alvarenga de Oliveira Pereira[1], Pedro Battaglia Bonetti[1], Rafael Gonçalves Brandi[1], Vinicius Moreira Martins Couto[1], Gabriel Lucas da Costa Oliveira[1], Bruno Vasconcelos Stefanini[1], João Pedro Costa Romão[1], Vitor Fernando Bordin Miola[2], Luís Henrique dos Santos Sousa[3], Nicolas Minavia Delgadillo[1]

[1]Universidade Metropolitana de Santos, Santos SP, Brazil.
[2]Universidade de Marília, Marília SP, Brazil.
[3]Universidade de Pernambuco, Serra Talhada PE, Brazil.

Correspondence: Rafael Alvarenga de Oliveira Pereira; Email: rafael.oliva192@gmail.com.

Background: Epilepsy is a chronic neurological disorder characterized by recurrent seizures, which are episodes of abnormal electrical activity in the brain. This activity can lead to a variety of symptoms, ranging from brief lapses in consciousness or involuntary movements to generalized seizures involving loss of consciousness and violent muscle contractions. The causes of epilepsy can vary, including brain injuries, congenital anomalies, genetic disorders, and underlying medical conditions. Treatment typically involves the use of antiepileptic medications to control seizures, although in some cases, surgery or other therapies may be recommended. This disorder is recurrent in emergencies, making it important to understand the management of epilepsy in this scenario. **Objective:** To carry out a systematic review of the literature on epilepsy and its management in emergency settings, with the purpose of analyzing the strategies involved in treating this condition in emergency hospital settings. **Methods:** Thus, managing epilepsy in an emergency requires swift and effective actions to control seizures and ensure patient safety. Key points include safeguarding the patient from injury during the seizure, ensuring an unobstructed airway, and administering antiepileptic medication, such as benzodiazepines, intravenously or intramuscularly, to halt seizures. Additionally, it is essential to assess and treat any underlying medical conditions that may be triggering seizures. Continuous monitoring of vital signs and maintaining a calm and safe environment are crucial during epilepsy management in the emergency setting. Following seizure control, investigating underlying causes and adjusting antiepileptic treatment are necessary to prevent recurrences. **Conclusion:** Hence, appropriate management of epilepsy in emergency situations is crucial to prevent severe complications, such as brain injuries, respiratory difficulties, and other medical emergencies associated with seizures. Moreover, effective management can help reduce the stigma surrounding epilepsy and enhance the long-term quality of life for the patient by ensuring adequate seizure control and minimizing negative impacts on physical and emotional health.

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Analysis of the use of benzodiazepines and cognitive changes

Tatiana Pizzolotto Bruch[1] , Camila Pezente Dal Toe[1], Cecília Juliani Felippe[1], Ingridy da Paz Bauer[1], Débora Dagostin Casagrande[1], Mirela Comin Salvaro[1], Gaia Batista[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Tatiana Pizzolotto Bruch; Email: tatibruch@yahoo.com.br.

To evaluate the correlation between the use of BZD and the presence of cognitive changes among university employees in Southern Santa Catarina, data were collected from a sample of 122 individuals. Information was gathered through the application of the Montreal Cognitive Assessment (MoCA), a screening tool for detecting Mild Cognitive Impairment (MCI). Variables such as age, sex, race, education, marital status, BZD use, medication dosage, duration of use, and reason for prescription were also collected. The study found no statistically significant relationship between BZD consumption and cognitive decline (p=0.077). However, the executive function domain of the MoCA test showed a statistically significant difference between BZD users and non-users (p=0.010). A high prevalence of medication use was reported, at 21.3%. The general profile of users included women, Caucasians, in stable relationships or married, primarily prescribed for anxiety, with a prevalent duration of use ranging from 0 to 12 months.

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Anatomopathological features of MRI negative epilepsies in a cohort from Southern Brazil

Fernanda Schuh Martins[1], William Alves Martins[1], André Luis Fernandes Palmini[1], Ricardo Paganin[1], Eliseu Paglioli[1], Rafael Paglioli[1], Thomas Frigeri[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Fernanda Schuh Martins; Email: fschuhmartins@gmail.com.

Background: Pharmacoresistant MRI-negative focal epilepsies present significant challenges for neurologists. While surgical intervention can potentially render patients' seizure-free, outcomes often vary significantly. The underlying pathology, aside from the localization of the epileptogenic zone, is a crucial factor influencing these outcomes. Certain etiologies are known to correlate with lower rates of post-surgery seizure freedom. **Objective:** This study evaluated the anatomopathological findings in a cohort of MRI-negative patients who underwent epilepsy surgery at the Porto Alegre Epilepsy Surgery Program (PAESP) from 2012 to 2019. **Methods:** The study included all MRI-negative patients who underwent surgery and had anatomopathological exams reviewed. Reports were available for 57 patients. Results showed Focal Cortical Dysplasia (FCD) I was the most common etiology, found in 18 (31.6%) patients. Mild Malformation of Cortical Development type II (MOGHE) was seen in 14 (21.1%), normal pathology in 10 (17.5%), FCD IIA in 9 (15.8%), mMCD I in 4 (6.5%), FCD IIB in 2 (3.2%), and FCD IIIA in 2 (3.5%). Rates of seizure-freedom were: 38.9% for FCD I, 22% for FCD IIA, 100% for FCD IIB, 33.3% for MOGHE, 20% for normal pathology, 25% for mMCD I, and 50% for FCD IIIA, with significant differences between etiologies (p=0.464). Median follow-up was 44 months (12-146). **Results:** In this cohort of MRI-negative patients, the pathology was consistent with previous studies, with FCD type I being the most common pathological substrate. Our 12-month seizure-free rate was 47.4%, with late recurrence in 30% of initially seizure-free patients. This could be due to more widespread underlying diseases, such as FCD type I, which is prevalent in nonlesional epilepsies. In a large cohort of MRI-negative patients, Wang et al. found FCD I in 40% of cases, corroborating our findings. Additionally, mild malformations of cortical development (mMCD) were a common finding in 27.6% of nonlesional cases. mMCD, as described in Palminis classification, includes heterotopic neurons in subcortical white matter (mMCD 1) or oligodendrocyte hyperplasia with blurring of the gray matter-white matter junction (mMCD II or MOGHE). Our study corroborates that mMCD II may be more common in nonlesional epilepsies than previously shown. **Conclusion:** In this southern Brazil cohort, FCD type I and MOGHE were the most common pathological substrates. These conditions may present a less distinct transition to normal cortex compared to other etiologies (e.g., FCD IIB), potentially leading to incomplete surgical resection of the epileptogenic zone and explaining late surgical recurrences in these patients.

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Anatomy of vertical hemispherotomy: a study in cadaveric brains

Maíra Piani Couto[1], Marcus Vinícius Giglio[1], Bianca Gomes Wanderley[1], Leticia Mansano de Souza[1], Enrico Ghizoni[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Leticia Mansano de Souza; Email: dra.leticiamansano@gmail.com.

Clinically intractable epileptic encephalopathies caused by hemispheric pathology can currently be treated by two surgical techniques, peri insular lateral hemispherotomy and vertical parasagittal hemispherotomy. Peri Insular hemispherotomy involves steps such as supra-insular and infra-insular windows and resection of the insula. The vertical parasagittal approach disconnects through a posterior frontal cortical window or the corpus callosum, continuing through the internal capsule and resecting the mesial temporal region. The vertical approach has advantages, such as complete disconnection at the level of the thalamus and lower risk of injury to the contralateral hemisphere. May result in seizure relief in patients with unilateral foci, although incomplete disconnections may occur, especially in extensive cortical malformations. Precise anatomical knowledge is crucial to the success of the procedure. It is essential to develop a universally acceptable hemispherotomy technique with consistent anatomical standards. In this sense, we carried out an anatomical study with photographic documentation of three adult cadaver heads in the Microneuroanatomy Laboratory of a University. Step-by-step disconnection of the hemisphere involves the steps: (1) Patient Position, Skin Incision, Craniotomy; (2) Resection (superior frontal rotation); step (3) Posterior callosotomy of the body; (4) approach to the Inferior Horn of the Lateral Ventricle; (5) disconnection of Projected Fibers from the anterior part of the Frontal Lobe; stage (6) the final disconnection of the Limbic System. It is essential to know the sequence of steps, know the anatomical landmarks and observe them all from different angles of vision, such as the steps (1): corpus callosum, foramen of monro, choroid plexus, collateral triangle, bulb callosum, falcotentorial junction and internal veins; stage (2): stria terminalis, fornix, choroid plexus, choroidal fissure and inferior choroidal point; stage (3): corpus callosum, septum pellucidum; stage (4): anterior cerebral artery (ACA), carotid bifurcation, olfactory triangle; stage (5): middle cerebral artery (MCA), corpus striatum, optic tract, posterior cerebral artery (PCA), and inferior choroidal point. The most challenging part is the final disconnection of the limbic system, requiring identification of the arteries through the arachnoid membrane, mainly the ACA, MCA and PCA, to avoid postoperative ischemic complications in the brainstem or thalamus. Vertical hemispherotomy surgery is established for intractable epilepsy secondary to diffuse, usually unilateral, hemispheric disease resistant to medical therapy. A solid anatomical understanding is a challenge even for neurosurgeons. Thus, a step-by-step understanding of the surgical procedure using cadaveric brains can serve as an intraoperative guide in disconnecting an entire cerebral hemisphere, beneficial for beginners in epilepsy surgery.

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Angioedema induced by thrombolytic therapy in stroke: a case series from Brazilian Northeast

Raphael Pinheiro Camurugy da Hora[1], Manuela Magalhães Dardenne Tenório[1], Ana Dolores Fimino Santos so Nascimento[1]

[1]Hospital da Restauração, Secretaria de Saúde de Pernambuco, Recife PE, Brazil.

Correspondence: Raphael Pinheiro Camurugy da Hora; Email: raphaeldahora@hotmail.com.

Background: The advent of intravenous thrombolysis with rtPA has drastically changed the landscape of cerebrovascular diseases. However, this therapy is not without potential complications, one of which is orolingual angioedema, a local inflammatory reaction that can be life-threatening. **Objective:** To report and analyze cases of orolingual angioedema occurring during rtPA thrombolysis in patients with ischemic stroke, highlighting the clinical presentation, management, and outcomes. **Methods:** Four cases of orolingual angioedema were identified during a one-year period among patients receiving rtPA thrombolysis for ischemic stroke at a tertiary hospital in Northeast Brazil. Clinical data, including patient demographics, comorbidities, time to onset of symptoms, treatment administered, and outcomes, were collected and reviewed. **Results:** Case 1: A 66-year-old hypertensive and diabetic male presented with sudden hemiparesis and dysarthria. Edema of the lips and left hemi-tongue was noted 55 minutes into the infusion. The infusion was stopped, and hydrocortisone and adrenaline were administered. Orotracheal intubation was performed due to worsening edema. Case 2: A 65-year-old hypertensive and diabetic male developed right hemiparesis and dysarthria. Facial angioedema, particularly of the left lower lip, was observed 53 minutes into the infusion. The infusion was stopped, and hydrocortisone and diphenhydramine were administered, leading to improvement of the edema. Case 3: A 63-year-old obese and hypertensive female developed anterior and right tongue edema at the end of the alteplase infusion. Hydrocortisone, promethazine, and adrenaline were administered, resolving the condition after 24 hours. Case 4: A 58-year-old male with no comorbidities developed tongue edema and respiratory discomfort 58 minutes into the thrombolytic infusion. The infusion was stopped, and dexamethasone and supplemental oxygen were administered. **Conclusion:** Our study found a prevalence of angioedema similar to that reported in the literature (1.2% of thrombolysed cases over one year). There was a predominance of initial patterns involving half of the lips or tongue, and ACE inhibitor use in half of the cases. Notably, all events occurred near the end of the infusion. We recommend periodic oroscopy during the infusion to enable early detection of this adverse effect, thereby ensuring better care for patients.

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Anterior temporal lobectomy vs amygdalohippocampectomy in the treatment of mesial temporal pole epilepsy: a functional connectivity analysis

João Fernando Cloclet Pio da Silva[1], Maíra Piani Couto[1], Letícia Mansano de Souza[1], João Vitor Gerdulli Tamanini[1], Brunno Machado Campos[1], Enrico Ghizoni[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Letícia Mansano de Souza; Email: dra.leticiamansano@gmail.com.

The objective of this study was to differentiate the two most accepted approaches to surgically treat temporal lobe epilepsy, the Anterior Temporal Lobectomy (TPAH) and Selective Amygdalohippocampectomy (TSAH), from the perspective of functional connectivity of different areas of the brain with the healthy temporal lobe pre and postoperatively. The main hypothesis was that, from the

point of view of postoperative functional connectivity, Selective Amygdalohippocampectomy would behave like Anterior Temporal Lobectomy, which presumably has better seizure control. This is a retrospective study of 101 refractory mesial Temporal Lobe Epilepsy patients treated surgically at the Epilepsy Service of a University, 57 submitted to temporo-polar amygdalohippocampectomy (TPAH) and 44 submitted to transsylvian selective amygdalohippocampectomy (TSAH). We sought to investigate the consequences to global and contralateral temporal pole connectivity of each approach and compare both groups. In the analysis of patients undergoing TPAH, it was found that the preoperative healthy (right) temporal lobe was more connected to the regions of the left medial temporal gyrus, orbital part of the left inferior frontal gyrus, left superior frontal gyrus (medial segment), left frontal pole, left and right anterior cingulate gyrus, brainstem and right subcallosal area, when compared to the postoperative healthy temporal lobe. When the TSAH group was compared to the TPAH, no increase in connectivity was detected. When doing the inverse comparison, TPAH vs TSAH, the connectivity was only increased in the lingual gyrus. This study's findings showed an almost insignificant difference in how the neuroconnectivity of the brain was altered after surgery between the two distinct approaches. Thus, no benefit was shown for choosing one over the order, from a functional connectivity perspective. When comparing the selective access group to the post operative TPAH, the healthy temporal poles (right) in patients submitted to selective access surgery did not present any increase in connectivity when compared to the healthy temporal poles (right) of patients submitted to TPAH. This study's findings showed an almost insignificant difference in how the neuroconnectivity of the brain was altered after surgery between the two distinct approaches. Thus, no benefit was shown for choosing one over the order, from a functional connectivity perspective.

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Anti-AMPAR encephalitis in a patient with rapidly progressive dementia and ataxia: a case report

Pedro Cougo Samueli[1], Isabela Wandarti Ribas[2], Gustavo Machado Pereira[3], Izadora Celant Miranda da Silva[1], Elora Sampaio Lourenço[1], Emanuel Cassou dos Santos[1], João Henrique Fregadolli Ferreira[4], Caio César Diniz Disserol[1,5,6], Livia Almeida Dutra[4]

[1]Instituto de Neurologia de Curitiba-INC, Curitiba PR, Brazil.
[2]Faculdade Evangélica Mackenzie do Paraná, Curitiba PR, Brazil.
[3]Univerisdade Positivo, Curitiba PR, Brazil.
[4]Hospital Israelita Albert Einstein, Instituto do Cérebro, São Paulo SP, Brazil.
[5]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[6]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Pedro Cougo Samueli; Email: drpedrocougosamueli@gmail.com.

Case presentation: ISW, male, 75 years old, presenting complaints of 3 weeks onset of progressive cognitive decline. He showed a pattern of a multidomain impairment characterized by anterograde amnesia, visual spatial impairment, semantic paraphasia, visual and auditory hallucinations, irritability and compulsive smoking. No episodes of seizures or transient loss of consciousness reported. The patient had a previous history of postural tremor, long-term alcohol and tobacco consumption and colon cancer resection 30 years ago. On physical examination, He showed moderate disorientation regarding space and time and no language impairment, no signs of nystagmus or ophtalmoparesis. He had bilateral postural tremor, paratonic rigidity of the right upper limb and mild tandem gait ataxia, but no dysmetria or dysdiadokokinesia. He also had palpable right axillary lymph nodes. MRI showed only diffuse unspecific T2/FLAIR hyperintensities and no signs of cerebellar atrophy. 2h EEG recording was normal. CSF showed mild pleocytosis (6,5 WBC) 64% lymphomonocytic, normal glucose and negative oligoclonal bands. Also, tests for viral infection (CSF PCR panel for HHV-1, HHV-2, VZV and Enterovirus) resulted negative. B1 levels were normal (85 mcg/dL), as well as B12 and homocysteine (321 and 12mcg/dL, respectively). ANA titers were 1:320 nuclear fine speckled pattern, but no other autoimmune, rheumatological or infectious serology was positive. Chest, abdomen and pelvis CT resulted negative for malignancy screening, except for the right axillary nodes enlargement. CSF encephalitis antibody panel resulted positive for anti-AMPAR encephalitis. The patient evolved with bronchoaspiration, refractory respiratory arrest and death before biopsy and other diagnostic tests and therapeutics could be performed. **Discussion:** Anti-AMPAR encephalitis is an autoimmune encephalitis that was first described in 2009. Patients typically present features compatible with limbic encephalitis (altered mental status, cognitive impairment, psychiatric symptoms) with or without seizures. There is a moderate risk of association to neoplasm (breast, thymoma and small cell lung cancer). Although there is a description of one patient with anti-AMPA encephalitis who presented parkinsonism responsive to immunotherapy, other movement disorders, such as ataxia, are not characteristic of the disease. Although this case describes a patient with characteristics of limbic encephalitis, mild CSF monomorphic pleocytosis with a diagnosis of anti-AMPA-R encephalitis, the unfortunate outcome prevented a full assessment of paraneoplastic causes as well as the gait ataxia itself. **Final comments:** This case highlights the need for high diagnosis suspicion for autoimmune encephalitis in patients presenting rapid onset progressive cognitive symptoms, altered mental status, behavioral changes, seizures, or movement disorders, associated with CSF abnormalities, after the exclusion of other possible causes.

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Anticholinisterasic, antidepressant, antipsychotic e hypnotic use in a sample of individuals with dementia: comparison between patient and caregiver report and plasma levels

André Luiz Rodrigues Palmeira[1], Mayra Pacheco Pachado[2], Tiago Franco de Oliveira[3], Bruno Pereira dos Santos[3], Sarah Eller[3], João Otávio Ancelmo Barankevicz[3], Carolina Fontela[3], Leonardo Martins de Paula[3], Liana Lisboa Fernandez[3]

[1]Santa Casa de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade de Maastritch, Maastricht, Netherlands.
[3]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: André Luiz Rodrigues Palmeira; Email: andreluizrp05@gmail.com.

Background: Several psychotropic drugs play an importante role on dementia treatment, and polypharmacy is frequently observed among this population, as well as drug inadequate use **Objective:** To estimate anticholinesterasic drugs and memantine use in this population; to compare data about drug use obtained through medical history with those collected by plasma measure of antidepressants, antipsychotics, and hypnothics. **Methods:** This present sample is part of a study regarding plasma biomarkers through mass spectroscopy method and its relationship with oxidative stress markers. In this very sample, several drugs and its metabolites were studied and data was compared with clinical history. **Results:** A hundred and seven individuals were recruited to this study, all of them with some sort of cognitive impairment (CDR 0.5= 5.61%; CDR 1.0= 47.66%, CDR 2.0= 40.19%, CDR 3,0= 6,54%). About 46.73% of the sample did not use anticholinesterasic drugs; between the users, 9.35% used galantamine, 16.82% rivastigmine and 27.10% donepezil. Memantine use was observed in 12.15% of the sample. Antidepressant use was encountered in 44,86%, being the most common escitalopram (16.82%), duloxetine (5.61%), fluoxetine and bupropion (4.67% each). When compared to mass spectroscopy, only 36.45% did use antidepressants, being escitalopram found in 11.21%, fluoxetine in 7.48%, buproprione in 2.8% and duloxetine in none of the participants. 23.37% of the sample used antipsychotics, being quetiapine the most representative (18.69%); nevertheless, by mass spectroscopy method, only 4.67% showed detectable seric levels. Hypnothic use (benzodiazepines and zolpidem) were found in 1.87% of the sample. Inconsistencies between what was related by the participants and their caregivers, and mass spectroscopy were found in 36.45% of the sample. **Conclusion:** In this sample, inconsistencies were found between clinical history and mass spectroscopy on psychotropic drugs use. In this kind of patient, to which polypharmacy is of common occurrence, attention to correct drug use is imperative, in order to avoid adverse events related to drug use and thus morbidity increase, to assure correct adherence, and to be aware of non-prescribed drug use.

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Anticoagulation strategy for patients presenting with ischemic strokes while using a direct oral anticoagulant: a systematic review and meta-analysis

João Paulo Mota Telles[1], Giulia Isadora Cenci[1], Gabriel Marinheiro[2], Gabriela Borges Nager[3], Rebeka Bustamante Rocha[4], Fernanda Ferreira Bomtempo[1], Eberval Gadelha Figueiredo[1], Gisele Sampaio Silva[5]

[1]Universidade de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[3]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[4]Universidade Federal do Amazonas, Manaus AM, Brazil.
[5] Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: João Paulo Mota Telles; Email: joao.telles@fm.usp.br.

Background: This meta-analysis examines four therapeutic approaches following a stroke in patients already on DOACs: continuing with the same DOAC, changing to a different DOAC, increasing the current DOAC dosage, or switching to a vitamin K antagonist (VKA), such as warfarin. **Methods:** Systematic review of literature from the Medline, Embase, and Cochrane databases, conducted in line with PRISMA guidelines. The analysis focused on six studies with varied patient demographics, examining as outcomes as recurrent ischemic stroke, intracranial hemorrhage, other bleeding events, and mortality. **Results:** Four studies compared the same regimen of DOAC and the change to warfarin. The group that maintained the same DOAC regimen had a significantly decreased risk of ischemic stroke (RR, 0.55; 95% CI 0.43, 0.70; p< 0.001; I2= 0%), intracranial hemorrhage (RR, 0.37; 95% CI 0.25, 0.55; p< 0.001; I2= 0%), and hemorrhagic events (RR, 0.44; 95% CI 0.30, 0.63; p< 0.001; I2= 6%). Warfarin increased the risk of mortality (HR, 1.85; 95% CI 1.06, 3.24; p= 0.03; I2= 84%) compared to the same DOAC. Changing the DOAC was not superior to maintaining the same DOAC regarding ischemic stroke (RR, 0.97; 0.66,1.42; p= 0.88; I2= 74%), intracranial hemorrhage (RR, 1.10; 95% CI 0.76, 1.60; p= 0.62; I2= 0%), hemorrhagic events (RR, 1.03; 95% CI 0.78, 1.37; p= 0.82; I2= 0%), or mortality (HR, 0.98, 95% CI 0.85, 1.13; p= 0.78; I2= 0%). Switching DOAC dose was not superior compared to maintaining DOAC dose regarding ischemic stroke (RR, 0.88; 95% CI 0.66, 1.18; p= 0.39; I2= 0%), intracranial hemorrhage (RR, 1.04; 95% CI 0.64, 1.67; p= 0.88; I2= 0%), and hemorrhagic events (RR, 1.10; 95% CI 0.69, 1.74; p= 0.69; I2= 0%). DOAC change

was superior to warfarin concerning ischemic stroke (RR, 0.57; 95% CI 0.34, 0.95; p= 0.03; I2= 67%), intracranial hemorrhage (RR, 0.33; 95% CI 0.21, 0.52; p< 0.001; I2= 0), and hemorrhagic events (RR, 0.46; 95% CI 0.27, 0.81; p< 0.01; I2= 40%). Three studies compared the switch of DOAC dose and the switch to warfarin. Ischemic stroke (RR, 0.58; 95% CI 0.41, 0.83; p< 0.01; I2= 0%) and intracranial hemorrhage (RR, 0.36; 95% CI 0.21, 0.61; p< 0.001; I2= 0%) were significantly decreased in DOAC dose change group compared to the warfarin group. Regarding hemorrhagic events, DOAC dose change was also superior (RR, 0.37; 95% CI 0.22, 0.62; p< 0.001; I2= 0%) between these treatment groups. There was no difference between switching DOAC dose and changing DOAC agent regarding ischemic stroke (RR, 1.14; 95% CI 0.90, 1.44; p= 0.28; I2= 0%). Intracranial hemorrhage (RR, 1.05; 95% CI 0.63, 1.77; p= 0.84; I2= 0%) and hemorrhagic events (RR, 0.99; 95 % CI 0.60, 1.64; p= 0.98; I2= 0%) **Conclusion:** Post-stroke adjustments to anticoagulation therapy do not yield additional benefits. Additionally, the results suggest that warfarin may be less effective than DOACs for preventing stroke recurrence, bleeding complications, and death in this patient population.

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Anticonvulsant potential of phenolic compounds in animal models of epilepsy: a systematic review

Rodrigo Esteves Lagares[1], Marcos Otávio Bueno[1], Mariana Siqueira Leite[1], Lucas Volpi Candido[1], Paola Ribas Gonçalves dos Santos[1], Igor Rodrigues de São Tiago[1], Alberto José Rodríguez Renfigo Valmorbida[1], Gustavo Colombo Dal-Pont[1]

[1]Universidade Alto Vale do Rio do Peixe, Caçador SC, Brazil.

Correspondence: Rodrigo Esteves Lagares; Email: rodrigolagares15@gmail.com.

Background: Epilepsy is a neurological condition of debilitating character marked by recurrent and spontaneous convulsions, affecting millions of people on a global scale. Despite the evolution of antiepileptic treatment, many patients continue to suffer from refractory seizures and the adverse effects of available drugs. Consequently, there is a continuous necessity to discover new treatment alternatives that may offer superior efficacy and less toxicity. In this context, phenolic compounds, widely found in plants, food and fruits, have stirred interest due to their antiepileptic properties. **Objective:** To investigate the pharmacological potential of the phenolic compounds in animal models of epilepsy. **Methods:** This is a systematic review carried out using PRISMA methodology. The database PubMed was used to search articles in the last 5 years (2020-2024), in English and Portuguese. As inclusion criteria, preclinical trials of rodents submitted to the induction protocol of convulsion/status epilepticus by pentylenetetrazol (PTZ) were defined. The articles were analyzed blindly by the authors, through the Rayyan® software. **Results:** Though the search strategy, 166 articles were found, and after careful filtration and analysis, 17 articles were included in this review. The experimental tests in mice demonstrated a significant reduction not only in the frequency of acute epileptic crisis, but also in the symptoms, after administration of the phenolic compounds. Research with resveratrol, ferulic acid, salvianolic acid B, quercetin, daidzin and genistein, among others, have highlighted their capacity to reduce convulsions induced by PTZ, attenuate cognitive and inflammatory deficits associated with epilepsy and protect against neuronal damage in animal models. These effects were attributed to the modulation of many signaling pathways, including AKT/CREB/BDNF, TLR4/NF- κB, JAK2/STAT3 and antioxidant pathways. Another relevant matter was the neuroprotective potential of some phenolic compounds, such as daidzin and quercetin, the latter also acting through the suppression of inflammatory cytokines like tumor necrosis factor alpha (TNF-α) and interleukin-1β (IL-1β), as well as the activity of the enzyme COX-2 (cyclooxygenase-2). **Conclusion:** Although promising, the results of these studies are mainly based on animal models, requiring more studies for better comprehension of these mechanisms. However, natural compounds present a significant therapeutic potential in epilepsy, offering an alternative and complementary approach to the conventional treatments.

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Antiganglioside antibody screening accuracy and sensitivity in a tertiary neuromuscular outpatient clinic

Ana Flávia Andrade Lemos[1], Amanda Vitória Freire de Oliveira[1], Lucas Gondim Briand Vieira[1], Mariana Ribeiro Pereira[1], Mateus Gustavo Favaro[1], Rodrigo Siqueira Soares Frezatti[1], André Cleriston José dos Santos[1], Giuliana França Teixeira Mandia Grossi[1], Pedro José Tomaselli[1], Wilson Marques Junior[1]

[1]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Ana Flávia Andrade Lemos; Email: anaflaviaandradelemos@gmail.com.

Background: Gangliosides are glycosphingolipids highly abundant in the nervous system. They are composed of a ceramide lipid tail attached through glycosidic linkage to a glycan headgroup containing one or more sialic acid residues. They play important roles in the modulation of membrane proteins and ion channels, in cell signalling, and in communication among cells. Antibodies against ganglioside subtypes, such as GM1, GD1a, GT1a, and GQ1b gangliosides, have been associated with various neurological disorders. **Objective:** To analyse the association of anti-ganglioside antibodies (AGAs) and their subtypes with neuromuscular diseases in a tertiary neuromuscular outpatient clinic. **Methods:** Clinical and serological data were retrospectively gathered from patient records using a standardized proforma. We included patients evaluated from 2015 to 2024. Sera from 197 patients were tested using enzyme-linked immunosorbent assay (ELISA) for AGA. Patients were divided according to predefined clinical categories based on a review of the current literature on AGA. Final diagnoses were combined with the AGA results for statistical analysis. **Results:** From 197 patients included, 75 (38.26%) were female and 122 (61.92%) male. The average age at the onset of symptoms was 47.5 years for females and 45.4 years for males. Fifty-seven cases (28.93%) tested positive for one of the seven AGA subtypes (GQ1b, GT1b, GD1b, GD1a, GM3, GM2, GM1). A positivity rate of 20% for AGA GM1 and GM2 was observed in patients with multifocal motor neuropathy (MMN). Other correlations found were: 57% of patients diagnosed with Miller-Fisher Syndrome (MFS) tested positive for AGA, with 28% being GQ1b. Acute motor axonal neuropathy (AMAN) exhibited the highest positivity rate at 58%, with 23% for GM1. Chronic inflammatory demyelinating polyneuropathy (CIDP) showed a 29% positivity rate, with 57% being GQ1b. **Conclusion:** Antibodies against ganglioside complexes, or gangliosides and other glycolipid complexes, are emerging as useful markers of various clinical features and pathological subtypes; however, they lack specificity and accuracy.

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Anti- IgLON5 disease presenting with bulbar dysfunction, ocular paralysis and facial myokymias

Carolina Heck Haubrich[1], Leticia Bassani Devens[1], Carollina Danezi Felin[1], Eduarda Rosa Fernandes[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Carolina Heck Haubrich; Email: carolhhaubrich@gmail.com.

Case presentation: A 65-year-old woman presented with acute severe dysphagia and stridor. The previous month, she had two aspiration pneumonias treated with antibiotics. Over the past year, she experienced unexplained falls and intermittent diplopia. Her medical history was unremarkable except for a thyroidectomy for stage I thyroid papillary carcinoma two years earlier. Neurological examination revealed supranuclear vertical palsy, slight bilateral facial weakness, facial myokymia, fasciculations, bilateral dysmetria, and an unsteady gait. She reported minor sleep problems, controlled with Zolpidem. Initially suspected of having myasthenia gravis, she showed subtle improvement with Pyridostigmine, but tests for MG were negative. She then developed acute respiratory distress, requiring mechanical ventilation. CSF revealed lymphocytic pleocytosis and increased protein. IVIG treatment stabilized her symptoms, but further tests for autoimmune and neoplastic causes were negative. A few months later, she experienced another episode of acute respiratory failure. Suspecting IgLON5, a Video-PSG demonstrated severe central and obstructive apneas, periodic complex movements, and undifferentiated NREM sleep. CSF tests (CBA) confirmed anti-IgLON5 antibodies. She was on a regimen of monthly IVIG and Rituximab, when she passed away 20 months after diagnosis. **Discussion:** Anti-IgLON5 is an auto-immune tauopathy recently identified as characterized by both REM and non-REM parasomnias, sleep apnea, and stridor. Its clinical features can resemble Parkinson's disease, MSA or Supranuclear Palsy, delaying the diagnosis. Initial presentations include sleep disorder, bulbar dysfunction, PSP-like syndrome, and cognitive impairment. Symptoms such as central hypoventilation and aspiration pneumonia due to dysphagia can be life-threatening. The presence of antibodies against extracellular IgLON5 epitopes is a hallmark of the disease. IgLON5 is a neuronal adhesion molecule whose function is not fully understood, yet it is widely expressed in the central nervous system. Neuropathology also reveals hyperphosphorylated tau protein accumulation in the hypothalamus, hippocampus and brainstem tegmentum, correlating with memory deficits, sleep disorders, and movement disorders. Associations with HLADRB1*10:01 and HLADQB1*05:01 alleles have been noted. Patients typically show a mild or no response to immunotherapy, likely due to irreversible IgLON5 antibodies. In a case series, half of the patients had a mean survival of 35 months post-diagnosis, with respiratory disturbances being the leading cause of death. **Final comments:** This case is notable due to the rarity of IgLON5 disease and its first reports in Latin America. The patient's acute respiratory failure and severe bulbar dysfunction prompted early gastrostomy and a broad differential diagnosis, initially considering myasthenia gravis. However, a sustained lack of improvement and unrevealing electroneuromyography led to the diagnosis of IgLON5.

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Anti-MAG: an important differential diagnosis of CIDP

Adriana Alves Rapp[1], Filipe Nolasco de Souza e Silva[1], Marcel Leal Ribeiro[1], Rayanderson Nunes da Gama[1], João Gabriel Teixeira Mota[1], Julia Cordeiro Braga[1], Lucas Vergne Cheade Lins[1], Brenda Francisca Cardoso Brito[1], Victor Xavier da Silva[1], Diego Sant'ana Sodré[2]

[1]Hospital Santa Izabel, Salvador BA, Brazil.
[2]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Adriana Alves Rapp; Email: adrianarapp15.2@bahiana.edu.br.

Case presentation: Patient, 77 years old, hypertensive with a history of prostate cancer, began experiencing weakness in the lower limbs in 2014 with progressive difficulty in walking, which worsened in 2016, requiring support for ambulation and the use of a cane since then. On physical examination, there was noticeable muscle weakness affecting the lower limbs with proximal strength graded as 4/5 and distal strength as 3/5, reduced vibratory sensation, and sensory ataxia. Osteotendinous reflexes were +/IV in the upper limbs and absent in the lower limbs. Other aspects of the neurological physical examination were unremarkable. During the investigation, in 2021, a electroneuromyography showed signs of demyelination such as reduced conduction velocity, temporal dispersion with conduction block, suggestive of chronic demyelinating sensory-motor polyradiculopathy. Concurrently, immunofixation of serum proteins revealed the presence of IgM. Due to the clinical presentation and complementary exams, AntiMAG antibody testing in 2022 was positive for IgM (titer 1/10). Initially, the patient was treated with prednisone but showed little response, also complaining of muscle pains during the treatment period. Currently, the patient is stable on Azathioprine, awaiting approval for Rituximab therapy with neurological stability. **Discussion:** AntiMAG neuropathy is predominantly a demyelinating disease characterized by a distally developed sensory or sensory-motor polyneuropathy in the context of IgM monoclonal gammopathy (sometimes of undetermined significance or secondary to lymphoplasmacytic lymphoma). The most common clinical manifestation involves weakness and altered sensation distally, primarily in the lower limbs, although some patients may experience gait instability and tremors. AntiMAG neuropathy is an important differential diagnosis of CIDP, particularly the distal variant (which accounts for 2/3 of patients with this phenotype), hence the mandatory need for adequate further investigation, including immunofixation of serum and urine proteins (often repeated if IgM is not initially detected). The confirmatory test is serological testing for AntiMAG antibodies. Traditionally, AntiMAG neuropathy does not respond to the usual treatments used for CIDP (such as corticosteroids or immunoglobulin), emphasizing the importance of correct differential diagnosis. The current optimal therapeutic option is sequential Rituximab infusion (anti-CD20 monoclonal antibody), with improvement seen in approximately 30-50% of cases. **Final comments:** Reinforcing that CIDP is a diagnosis of exclusion is essential for appropriately addressing possible differential diagnoses, among which is Anti-MAG neuropathy.

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Anti- MDA5 dermatomyositis with severe myopathy: an unusual presentation

Elisa Gabriela Barros Cunha[1], Márcio Victor de Souza Silva[1], José Orlando Ferreira Tomé Filho[1], Kevellyn Cruz Aguilera[1], Hermany Capistrano Freitas[1], Lara Albuquerque de Brito[1], Natália Merten Athayde[1], Eduardo Braga de Oliveira[1], Paula Camila Alves de Assis Pereira Matos[1], Cleonisio Leite Rodrigues[1]

[1]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Elisa Gabriela Barros Cunha; Email: elisacunha4@gmail.com.

Case presentation: A 71-year-old woman with a history of hypertension,dyslipidemia, and hypothyroidism presented to our service with a 4-month history of dysphagia, dysphonia, muscle weakness, and spots on her face and hands. One month before the onset of symptoms, she had experienced a self-limited flu-like illness. She was taking losartan, hydrochlorothiazide, levothyroxine, and simvastatin. On general physical examination, hyperchromic lesions were observed on her lower eyelids and palms. Neurologically, she exhibited predominantly proximal muscle weakness in all four limbs (MRC 45), amyopathic gait, and required support from another person. Additional tests revealed CPK levels of 4705 U/L and ANA 1:5000 with a membrane-type nuclear pattern. Other rheumatological markers, including anti-SCL70, JO1, RO, LA, mitochondria, and RNP antibodies, were negative. Electroneuromyography indicated severe subacute myopathy with proximal predominance, fibrillations, and electrical myotonia. A muscle biopsy of the biceps brachii showed focal myositis with mild atrophy, and immunohistochemistry was positive for anti-CD68, CD3, CD4, CD8, and MHC-I antibodies, consistent with inflammatory myopathy. A serum antibody panel for myositis was positive for anti-MDA5 and anti-RO52. Neoplastic screening, including chest CT, upper digestive endoscopy, and thyroid ultrasound, showed no malignancy. Chest CT revealed thickening of the bronchial walls and posterior basal reticular opacities. During hospitalization, the patient received pulse therapy with methylprednisolone 1g for 5 days, and azathioprine was initiated. After discharge, based on the antibody panel results, monthly pulse therapy with gradual tapering was continued. At the last outpatient follow-up, 10 months post-discharge, the patient showed improvement in hyperchromic lesions and muscle strength (MRC 56), and was walking with a walker. The dose of azathioprine (2.1 mg/kg/day) was maintained, and methylprednisolone pulse therapy was gradually tapered. **Discussion:** Dermatomyositis (DM) has a global incidence of 5-10 per million per year and a prevalence of 6-7 per 100,000 per year. The most commonly associated antibodies are anti-Mi2, MDA5, NXP2, TIF1, and SAE. In DM associated with anti-MDA5, muscle involvement is generally less common than in other DM presentations, with skin rash, polyarthralgia, and interstitial lung disease being more frequent. This case is unusual in that the primary manifestation was muscle involvement. Anti-Ro52 is a non-specific antibody that can be present in various rheumatological diseases, occurring in 27-62% of anti-MDA5 DM cases and is associated with a worse prognosis due to increased lung involvement. **Final comments:** This case describes anti-MDA5 dermatomyositis with atypical severe muscle impairment. Early suspicion and therapeutic intervention in the context of inflammatory myopathy allowed for a relatively favorable outcome.

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Anti-MDA5 dermatomyositis: two phenotypic extremes

Pedro Nogueira Fontana[1], Caio César Alves Lins de Oliveira[1], Carolina da Cunha Correia[1], Lara Candido Pena[1], Alex de Novais Batista[1], Ana Valeska Lisboa Carvalho[2], Ana Maria Dutra[3], Igor Dias Brockhausen[3], Roseli Corazzini[3], Alzira Alves Siqueira Carvalho[3]

[1]Hospital Universitário Oswaldo Cruz, Recife PE, Brazil.
[2]Hospital Alberto Urquiza Wanderley, UNIMED, João Pessoa PB, Brazil.
[3]Centro Universitário Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Caio César Alves Lins de Oliveira; Email: caiolins97@gmail.com.

Background: Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by muscle weakness and typical skin lesions, with subgroups delimited by specific autoantibodies. Anti-MDA5 is associated to severe skin changes, including necrotic lesions, interstitial lung disease (ILD), normal CK and little or no weakness. In these cases, the muscle biopsy is usually normal or presents mild and nonspecific changes. **Methods**: We report a case of anti-MDA5+ DM (DM-MDA5) with atypical clinical and laboratory presentation, comparing it with a typical case. **Results:** Case 1: 40-year-old man came to medical attention due to skin lesions on his face 10 months ago. He then began to experience weakness in the lower limbs, progressing to the upper limbs, in addition to fever, dysphagia and dyspnea. On examination, he presented Gottron's papules, malar rash, shawl sign and proximal muscle strength grade 3 and distal grade 5. Laboratory tests revealed leukocytes of 2100/mm3, platelets 91,000/mm3, CK 77IU/L (< 171UI/L), aldolase: 8.7U/L (< 7.6U/L), ferritin 2855ng/mL (< 400ng/mL), ANA 1:80 (dotted cytoplasmic pattern). The following were negative: rheumatoid factor, anti-DNAds, anti-RNP, anti-Sm, anti-SSA/SSB, anti-Scl70, anti-centromere, anti-LKM1, anti-AMA, anti-TPO, anti-GAD, lupus anticoagulant and anti-cardiolipin (IgG/IgM). Serum immunofixationsuggested IgG/kappa monoclonal protein. Myelogram revealed granulocytic and megakaryocytic hyperplasia. Imaging evaluation showed splenomegaly and muscular edema with gadolinium enhancement in the right thigh and left arm. In neoplastic screening, thyroid infiltrative follicular papillary carcinoma was found. Electromyography showed a myopathic pattern with spontaneous activity. Muscle biopsy revealed necrotic and regenerating muscle fibers, perifascicular atrophy, perivascular inflammatory infiltrate, sarcolemmal C5b9 and sarcoplasmic immunoexpression for MXA. DM-specific autoantibody panel was positive for anti-MDA5. Case 2: 50-year-old man sought medical attention due to skin lesions. He presented Gottron's sign, palmar erythema, necrotic and scaly skin lesions on the back and shoulders and calcinosis cutis. On examination, he presented grade 4+ proximal weakness and dyspnea. Chest CT showed ILD. Muscle biopsy showed mild and nonspecific changes, without MHC-I immunoexpression. Myositis autoantibodies panel was positive por MDA5. **Conclusion:** we illustrate two extremes of the DM-MDA5 presentation spectrum: a hypomyopathic form and another with muscular involvement even with normal CK. Furthermore, case 1 presented characteristics of macrophage activation syndrome secondary to DM, and concomitant thyroid carcinoma. We highlight 1) wide phenotypic variability in DM-MDA5, 2) DM with severe weakness and normal CK and 3) DM-MDA5 with severe muscle involvement and associated with thyroid neoplasia.

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Antiseizure drugs impair fine motor dexterity in people with epilepsy without lesions in the motor areas

Lucas Scárdua-Silva[1], Marco Tulio Medina[1], Gabriela Ribeiro Prada[1], Marina Koutsodontis Machado Alvim[1], Rafael Batista João[1], Thais Maria Bezerra[1], Fernando Cendes[1], Mateus Henrique Nogueira[1], Clarissa Lin Yasuda[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Lucas Scárdua-Silva; Email: scardua@unicamp.br.

Background: Several neuroimaging studies have demonstrated gray matter atrophy in the precentral gyri, basal ganglia and alterations of corticospinal tracts in focal and generalized epilepsies; however, little is known about the fine motor alterations in people with epilepsy (PWE). Besides, fewer studies have investigated the negative impact of adverse effects of drugs, symptoms of anxiety and depression on motor performance. **Objective:** Here, we aimed to analyze the impact of adverse effects of antiseizure medication on gross and fine motor dexterity in PWE and the relationship with associated factors such as depression and anxiety. **Methods:** We interviewed and tested 195 people with epilepsy (PWE, median of 35 years of age and 11 years of education) at a Tertiary Centre (before their outpatient appointment) and 252 healthy volunteers (HV, median of 35 years of age and 15 years of education) at the University Campus. All individuals performed the 9-hole peg test (9-HOLE), Box and Block Test (BBT), Tower of Hanoi (TH), the Block design subtest of the Wechsler Adults Intelligence Scale (BD-WAIS), Generalized Anxiety Disorder Questionnaire (GAD-7) and Neurological Disorders Depression Inventory for Epilepsy (NDDI-E). Patients also answered the Liverpool Adverse Events Profile (LAEP; 80% with adverse events and 20% without adverse events). PWE was separated into seizure-free (76 subjects), lower-frequency (13 subjects)

and pharmacoresistant (85 subjects). We used SPSS23 for statistical analyses with non-parametric tests for group comparisons (Mann-Whitney and Spearman correlation). We excluded subjects with intellectual or gross motor impairment. **Results:** Overall, the PWE performance was 15-22% worse than their age-paired controls ($p< 0.05$). Group with adverse events (AE_pos) was balanced to the group without adverse events (AE_neg) in terms of age ($p=0.8$) and education ($p=0.6$). However, the AE_pos presented higher proportion of patients with pharmacoresistant seizures ($p=0.03$), depression (NDDI, $p<0.001$) and anxiety (GAD-7, $p<0.001$). Interestingly, the two groups performed similarly for fine motor and complex tasks [9-HOLE (bimanual, $p>0.05$); tower of Hanoi ($p=0.5$) and BD-WAIS ($p=0.3$)]. On contrary, the AE_pos presented a poor bilateral performance on the gross motor test of Box and Block Test (average of 10% worse; $p<0.05$, bilaterally). We did not observe strong correlations between performance and intensity of anxiety or depression, regardless the higher scores. **Conclusion:** The impact of adverse events was mainly observed as higher intensity of anxiety and depression symptoms, and specifically on Box and Block Test. We speculate there is a 'floor effect" present in these groups of patients, associated with long duration of epilepsy, explaining similar performance on other tests. The treatment of anxiety and depression may improve motor performance. Future analyses with subtypes of epilepsies and the different medications are necessary.

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Anti-seizure prophylaxis and electroencephalographic changes in bulsufan therapy: a case report

Luiza Gonçalves Fraga[1], Luís Guilherme Pacheco de Almeida[1], Mariane Panka[1], Patrícia Coral[1], Carlos Eduardo Soares Silvado[1], Marlon Wycliff Caeira[1], Luciano de Paola[1], Mayara de Rezende Machado[1]

[1]Universidade Federal do Paraná, Complexo Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Luiza Gonçalves Fraga; Email: luizaag.fraga@gmail.com.

Case presentation: We present a case of a female patient aged 5 years old diagnosed at 16 months with Chediaki-Higashi syndrome (CHS), a rare primary immunodeficiency syndrome of autosomal recessive inheritance characterized by recurring infections and oculocutaneous albinism. During her disease course, she presented a first episode of bilateral tonic-clonic seizure (BTCS). Three years later, meanwhile a febrile illness, she presented a new seizure, and started on levetiracetam 30mg/kg/day. Due to disease's progression, she underwent bone marrow transplantation (BMT) receiving conditioning with bulsufan (BU) 19.6mg/kg, alongside fludarabine and antithymocyte globulin. Due to the neurotoxicity of BU and a history of epilepsy, levetiracetam dose was increased to 40 mg/kg/day. On the fourth day of infusion, the patient developed two episodes of seizures, and started phenytoin and phenobarbital as an add-on therapy to levetiracetam. Ancillary testing with a CT scan and electroencephalogram (EEG) showed only brain atrophy and diffuse and persistent slowing of background activity, predominantly in the delta range. Patient evolved with clinical improvement. **Discussion:** BU has been used since 1959 and is commonly used in the conditioning regime for BMT. It easily crosses the blood-brain barrier, reaching levels similar to those in blood or even higher. Neurotoxicity figures among its side effects, manifesting as seizures in around 7.5-10% of patients and due to this high incidence, antiseizure prophylaxis with phenytoin, phenobarbital, benzodiazepines or levetiracetam is reasonable. The first dose is given 12 hours before the first dose of BU and continues until 48 hours after the last dose. BTCS generally occurs during therapy, mainly between the third and fourth day, with myoclonus and muscle spasms also being reported. The ictal/interictal EEG may show generalized spike/polyspike-wave discharges, while follow up examinations show a persistent slowing of background with intermittent rhythmic delta activity, which may persist for 20-30 days. Even without an unequivocal seizure, encephalographic abnormalities can occur in up to 60% of patients. **Final comments:** Though seizures are rare in CHS patients, those on BU show a seizures' incidence of approximately 1.3%, being more common in patients with predisposing factors such as previous seizures/epilepsy or previous brain injury. Electroencephalographic changes can occur at any time during BU therapy and may persist for weeks, as the delta diffuse slowing presented by the patient in this case after treatment with 3 antiseizure medications. This report reinforces the importance of knowing potential side effects of drugs used in current medical practice and how antiseizure prophylaxis in selected cases can impact patients' prognosis.

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Anti-SOX1 antibody-positive caused by small-cell lung cancer presenting with chorea: a case report

Victoria Lin[1], Victor Rebelo Procaci[2], João Henrique Fregadolli Ferreira[1], Livia Almeida Dutra[1]

[1]Instituto do Cérebro, Hospital Israelita Albert Einstein, São Paulo SP, Brazil.
[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Victoria Lin; Email: vickylin1001@gmail.com.

Case presentation: A 60-year-old woman with liver metastasis and prior diagnosis of small-cell lung cancer, treated with radiotherapy, prophylactic cranial irradiation, and chemotherapy until 2022, was admitted in November 2023 to investigate involuntary movements she had experienced over the last year. In March 2023 she developed involuntary movements of the lips and, after three months, involuntary movements of the feet and legs as well. Neurological exam revealed chorea involving the face, legs, and feet. Hung-up knee jerk and motor impersistence were also present. Brain magnetic resonance imaging showed subtle brain volume reduction, subcortical white matter hyperintensities, and hemosiderin foci in the right thalamus. Upper abdominal magnetic resonance imaging revealed a hypovascular nodule in liver segment VII. Positron emission tomography-computed tomography indicated glycolytic hypermetabolism in a right peri-hilar lung mass and the hepatic nodule. She was treated with haloperidol 5 mg daily and intravenous immunoglobulin for five days, resulting in partial improvement. However, nine days later, she relapsed and the chorea progressively worsened. Cerebrospinal fluid and serum testing with a tissue-based assay and immunoblot revealed strong reactivity to anti-SOX1 antibodies. The haloperidol dosage was increased to 5 mg twice a day and oncology started durvalumab for the treatment of the small-cell lung cancer, which resulted in an almost complete resolution of the chorea. **Discussion:** Anti-SOX1 antibodies are partly characterized as onconeural due to their correlation with neoplastic diseases, particularly small-cell lung cancer. These antibodies are detected in up to 36% of patients with this neoplasm and are considered serological markers for this condition, an association reinforced by the present case. Chorea is not reported as a neurological symptom in anti-SOX1-positive patients with cancer, despite the common association of the antibody with intracellular antigens. Regarding treatment, the low prevalence of anti-SOX1-related paraneoplastic syndrome has precluded trials, resulting in a lack of guidelines. Syndromes associated with intracellularly targeted antibodies tend to be refractory to treatment. Moreover, given the probability of intracellular antigen, immune checkpoint inhibitor therapy was approached with caution, as it could increase the risk of aberrant T cell activation and immune-mediated adverse events, potentially aggravating the chorea, which did not occur in our case. **Final comments:** Anti-SOX1 antibodies are linked to small-cell lung cancer and chorea is associated with paraneoplastic syndromes, although a direct connection between them has rarely been documented. Further research is needed to recognize and treat these presentations.

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Anxiety, depression, and excessive daytime sleepiness in patients with parkinson's disease: a possible association?

Mariana Carvalho[1], Diego Monteiro de Carvalho[2], Marcus Vinicius Della Coletta[3], Alvaro Siqueira da Silva[2], Marcello Facundo do Valle Filho[2], Raissa Costa Said[1], Emillaine Alves Noronha[1], Agatha Lopes Azedo[1], Yenly Gonzalez Perez[2], Carlos Mauricio Oliveira de Almeida[2]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade Federal do Paraná, Curitba PR, Brazil.

Correspondence: Agatha Lopes Azedo; Email: agathalopesazedo@gmail.com.

Background: Parkinson's disease is a neurodegenerative disorder associated with the loss of nigrostriatal dopaminergic neurons, affecting 1% to 2% of the population over the age of 60. In addition to motor symptoms such as rigidity and postural instability, it manifests through non-motor symptoms including sleep disorders, anxiety, and depression, which can negatively impact the clinical course and quality of life of these patients. **Objective:** To analyse the association between anxiety, depression, excessive daytime sleepiness, and the stages of Parkinson's disease as determined by the Hoehn-Yahr scale. **Methods:** This was an observational, cross-sectional study conducted over 12 months at a tertiary referral hospital in Manaus, AM. The study was approved by the local ethics committee under CAAE n° 58684622.2.0000.0007. Inclusion criteria were patients with Parkinson's disease aged >18 years, diagnosed according to the London Brain Bank criteria, with staging of 1-3 on the Hoehn-Yahr scale. For the diagnosis of depression and anxiety, a single question was used, and for sleepiness, the Epworth Sleepiness Scale was used, with a score greater than 10 indicating excessive daytime sleepiness. **Results:** Among the 52 individuals, 33 (63.46%) were depressed and 19 (36.54%) were not; 42 (80.77%) were anxious and 10 (19.23%) were not; 34 (65.38%) were classified with excessive daytime sleepiness, and 18 (34.62%) were not. No significant association was observed between the stages of Parkinson's disease as per the Hoehn-Yahr scale and depression (X^2= 3.27; $p=0.65$), nor between Parkinson's disease and anxiety (X^2=3.54; $p=0.61$). No significant association was found between anxiety and sleepiness (X^2=4.28; $p=0.11$) or between depression and sleepiness (X^2=3.29; $p=0.19$). **Conclusion:** Our results support the literature, demonstrating that a large proportion of patients with Parkinson's disease exhibit anxiety, depression, and excessive daytime sleepiness. However, no significant association was found between the stages of Parkinson's disease and anxiety or depression, nor between anxiety and depression with excessive daytime sleepiness.

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Aphasic status in a patient with serous ovarian carcinoma and CNS recurrences: case report and narrative literature review

Valdecir Boeno Spenazato Júnior[1], Thales Pardini Fagundes[1], Mayara Apolinário Januzzi[1], Isabela Rios Felix[2], Antonio Torrecilha Manfrinato[2]

[1]Fundação Pio XII, Barretos SP, Brazil.
[2]Faculdade de Ciências da Saúde de Barretos Dr. Paulo Prata, Barretos SP, Brazil.

Correspondence: Isabela Rios Felix; Email: isabelar.felix13@gmail.com.

A 51-year-old woman with a history of high-grade serous ovarian carcinoma stage IIIC had a CNS recurrence in 2019. She underwent resection and radiotherapy for lesions in the left frontal, temporal, and parietal lobes, followed by radiosurgery in July 2021 for a right frontal motor area lesion. The following year, a new nodule appeared in the right cerebellar hemisphere. By 2023, the cerebellar nodule increased in size, leading to multidisciplinary discussions about its nature, confirmed as radionecrosis in February 2024, with bevacizumab treatment initiated. In April 2024, the patient presented with acute onset speech difficulty, unable to pronounce several words, and was disoriented in time, person, and space. She had trouble understanding complex commands, could not repeat phrases, and had naming difficulties, along with ideomotor and ideational apraxia, partial alexia without agraphia, and echolalia. Cranial CT and lab tests showed no significant changes. A new MRI showed few evolutionary changes, with a slight increase in the edema of the left parietal lesion. The CSF analysis revealed no cells, and there were no signs of leptomeningeal disease. EEG findings were compatible with focal epilepsy and electrographic seizures, confirming Focal Non-Convulsive Status Epilepticus - Aphasic Status. The patient received levetiracetam, leading to complete improvement. Aphasic status does not present symptoms other than aphasia and is classified as an atypical seizure condition. Although rare, aphasia can occur in both focal seizures and status epilepticus, and it may be caused by multiple etiologies such as structural lesions, metabolic disturbances, and neurological diseases. In this case, despite the patient having a parietal lesion, it had no modifications and no disease progression. Moreover, laboratory abnormalities were absent. The diagnosis can be confirmed through EEG, however, a prolonged tracing is often necessary. Despite being challenging, early diagnosis is essential for the initiation of anticonvulsant treatment, as delays can lead to increased mortality. In this case, the patient began treatment with Levetiracetam and showed complete improvement, with no new seizures after one month of follow-up, reassuring the diagnosis. Treatment and changes in EEG patterns are important tools for retrospective diagnosis in such cases. Aphasic status is rare, likely due to underdiagnosis, and requires a high level of clinical suspicion. It should be considered in any patient with sudden aphasia or prolonged changes in fluency and comprehension. A simple EEG may not be suggestive, but it is an essential tool. Treatment should be initiated early with anticonvulsants to improve the status.

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Application of artificial intelligence algorithms in the diagnosis of temporal lobe epilepsy compared to conventional methods: a systematic review

Sayros Akyro Soares Martins[1], Raphael Fernandes Casseb[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Sayros Akyro Soares Martins; Email: sayrosakyro@gmail.com.

With the aim of evaluating the impact of artificial intelligence (AI) algorithms on the diagnosis of temporal lobe epilepsy (TLE), we conducted a systematic literature review of the past 10 years, assessing the performance of AI compared to conventional methods, the main data used (clinical or neuroimaging), and the algorithms employed. Studies included met the following criteria: published in English, included patients with TLE, evaluated the use of AI in diagnosing TLE in comparison to conventional methods, and reported performance metrics such as accuracy, sensitivity, and specificity. Studies focused on animal models or in vitro, not written in English, lacking comparison with a conventional diagnostic method, not published in peer-reviewed journals, and lacking performance metrics were excluded. The risk of bias in the studies was assessed using the QUADAS-2 bias scale. Statistical evaluation and meta-analysis of the data were performed using SPSS software. Literature searches were conducted using the databases PubMED, Scopus, Web of Sciences, Cochrane, and Embase. Search terms included "Epilepsy, Temporal Lobe/diagnostic imaging," "Artificial Intelligence," "Machine Learning," "Convolutional Neural Network," and "Magnetic Resonance Imaging." The initial search yielded 165 studies. Of these, 34 duplicate versions and 109 studies that did not meet the inclusion and exclusion criteria were removed, resulting in 22 studies being included. The study involved a total of 4,326 patients, of whom 2,114 had TLE and 2,212 were part of the control group (healthy individuals, patients with epilepsy other than TLE, or patients with neurodegenerative diseases). The median accuracy of the AI algorithms was 0.878. 63.64% of the studies used machine learning algorithms, 27.27% used deep learning algorithms, and 9.09% used both. The results showed the great potential of AI algorithms in assisting the diagnosis of temporal lobe epilepsy, with performance comparable to conventional methods. Deep learning algorithms have increasingly gained relevance in this issue, but the machine learning algorithm Support Vector Machine is still the most used (59.09% of the included studies). The main limitation of the study was the heterogeneity of the available performance metrics, with some studies not reporting relevant statistical information such as standard deviation or confidence interval, preventing the inclusion of half of the pre-selected studies in the initial search. When appropriate, future studies should report more robust statistical metrics, such as the sensitivity, specificity, and F1-score. Systematic review registration: https://www.crd.york.ac.uk/prospero/. Registration number: CRD42024545284

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Application of artificial intelligence and mass spectrometry for biomarker discovery in plasma and CSF of patients with cognitive decline

Leandro Cesar Monteiro[1], Isadora Cristina Ribeiro[2], Liara Rizzi[2], Ítalo Karmann Aventurato[2], Marjorie Cristina da Rocha Silva[2], Marcio Luiz Figueredo Balthazar[2]

[1]Fundação Oswaldo Cruz, Instituto Carlos Chagas, Paraná PR, Brazil.
[2]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Leandro Cesar Monteiro; Email: lcesarmonteiro@gmail.com.

Background: The early diagnosis of cognitive decline, including Alzheimer's disease (AD), remains a significant challenge in neurology. Identifying reliable biomarkers in plasma and cerebrospinal fluid (CSF) can revolutionize diagnostic approaches and patient management. The integration of artificial intelligence (AI) with mass spectrometry (MS) offers a promising avenue for biomarker discovery due to its high sensitivity and specificity. **Objective:** This study aims to employ AI-driven analysis and MS to identify potential biomarkers in plasma and CSF of patients exhibiting cognitive decline. **Methods:** Plasma and CSF samples were collected from patients diagnosed with cognitive decline and matched controls. Mass spectrometry (LC-MS) was utilized to perform a comprehensive proteomic analysis of the samples. The resulting datasets were then processed and analyzed using AI algorithms tailored toward shortlisting patterns and identifying biomarkers correlated with cognitive decline. **Results:** Preliminary analysis has identified a set of proteins in both plasma and CSF that show significant differences between patients with cognitive decline and controls. The AI models have successfully highlighted potential biomarkers that may be involved in the pathophysiology of cognitive decline, offering promising targets for further validation. For example, in samples from AD patients, Beta Amyloid proteins (P05067) were found in quantities of around 270 spectral counts. A group spectral count can account for protein variants. In contrast, the analysis of samples from patients with cognitive decline (CD without AD identified) the same Beta Amyloid protein with a lower number of spectral counts, around 60. This preliminary finding suggests the potential of this differential biomarker in the analyzed samples. **Conclusion:** The integration of AI with MS in the analysis of plasma and CSF samples presents a robust methodology for biomarker discovery in cognitive decline. This approach not only enhances our understanding of the underlying mechanisms but also paves the way for the development of early diagnostic tools and personalized therapeutic strategies.

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Application of the new criteria for optic neuritis in a patient seronegative for anti-AQP4 and anti-mog antibodies

Francyane Peixoto Ramos de Abreu[1], Luana Gomes Dias Pimentel[1], Marina Papais Alvarenga[1], Regina Maria Papais Alvarenga[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.

Correspondence: Francyane Peixoto Ramos de Abreu; Email: francypra@gmail.com.

Case presentation: In 2022, a 46-year-old white male sought ophthalmological care complaining of visual alterations, mentioning a blurring of the central visual field in the right eye. Numerous tests were carried out and found to have mild dysfunction in the right eye. However, at the end of 2023, the patient reported a worsening of the condition, which led him back to the ophthalmologist, who ordered angiofluorescein, retinal mapping, MRI of the skull/orbits and visual field, in which loss of the temporal visual field in the right eye was detected. In April 2024, the patient noticed bilateral blurriness and sought medical assistance. On examination of the fundus of the eye, bilateral papillary oedema was identified, and corticosteroids were prescribed for a month, during which 50% of the condition improved. In May 2024, the patient went to the neurologist with a report specifying the alterations in the fundus of the eye and the presence of visual deficit bilaterally, with greater involvement of the right side. The suspicion of optic neuritis arises considering the papilla oedema. The neurologist requests a skull CT scan, optical coherence tomography, and visual evoked potential and asks the ophthalmologist for a new opinion to rule out ophthalmological causes. The skull CT was normal, and the ophthalmological causes were ruled out, but the visual evoked potential confirmed the suspicion of optic neuritis, concluding that there was a conduction defect in the visual pathways on both sides, with the right side being the most affected and compatible with demyelinating and axonal involvement of the optic nerve. The patient was informed of the disease and expressed the wish to be investigated, so he was admitted to Rio de Janeiro to have his cerebrospinal fluid examined and the markers anti-aquaporin 4 (AQP4) and autoantibody against myelin oligodendrocyte glycoprotein (Anti MOG) measured. After the tests were carried out, the cerebrospinal fluid was found to be normal, with no oligoclonal bands and no AQP4 or MOG markers. The patient is currently waiting to complete one month of physiotherapy before undergoing a new MRI. **Discussion:** The absence of lesions in the brain, spinal cord and AQP4 and MOG antibodies speaks in favour of isolated optic neuritis. In this report, the patient initially presented with a visual field deficit on the right, and within more than a year had a worsening episode in which the visual loss became bilateral, as well as papilla oedema assessed bilaterally by fundoscopy. The

change in evoked potential showed demyelinating and axonal involvement of the optic nerve, confirming the diagnostic suspicion of optic neuritis. **Final comments:** It is therefore believed that this is relapsing isolated NO (RION), due to the recurrence of symptoms, involvement restricted to the optic nerve and the absence of autoantibodies typically related to the disease.

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Application of the proposed criteria for the diagnosis of mog- associated disease (MOGAD) in a population from Rio de Janeiro

Gabriela Barroso Girafa[1], Kelma Macedo Pohlmann Simoes[1], Vanderson Carvalho Neri[1], Marina Papais Alvarenga[1], Marcos Papais Alvarenga[1], Claudia Cristina Ferreira Vasconcelos[1], Marcell Pourbaix Morrisson Guimarães[1], Cleonice Alves de Melo Bento[1], Priscila Mendonça do Sacramento[1], Regina Maria Papais-Alvarenga[1]

[1]Universidade Federal do Estado do Rio de Janeiro, Pós-Graduação da Neurologia, Rio de Janeiro RJ, Brazil.

Correspondence: Gabriela Barroso Girafa; Email: gabrielagirafa@hotmail.com.

Background: In 2023, an international panel proposed new criteria for MOG-associated disease (MOGAD) based on clinical and imaging data and titers of MOG-IgG antibodies. **Objective:** To apply the MOGAD criteria in a series of patients with positive serum antibodies against the myelin oligodendrocyte glycoprotein (MOG-IgG) from Rio de Janeiro. **Methods:** Between 2016 and 2023, Neuromielitis optica (NMO), high risk NMO and acute disseminated encephalomyelitis (ADEM) patients from Rio de Janeiro were tested for anti aquaporin 4 antibody (AQP4-IgG) and MOG-IgG by the cell-based assay (CBA) method. The new MOGAD criteria were applied in the cases positive to MOG-IgG. **Results:** From 516 patients tested, 94 were positive for AQP4-IgG. Among the 422 negative cases for AQP4-IgG, MOG-IgG was present in 28 patients, five under 18 years old and 23 adults. Sixty percent of the patients were female, 70% caucasians and 30% afro-descendants. The age ranged between eight and 47 years (median 31). Two patients were seropositive for both antibodies. Optic neuritis (ON) ocurred in twenty patients, myelitis in 11 and brainstem syndrome in two patients . Two pediatric patients presented with ADEM. We didnt have titration in our test, making it necessary to have additional supporting clinical or MRI features, such as bilateral simultaneous clinical ON (12 patients), longitudinal optic nerve involvement (six patients), optic disc oedema (four patients), perineural optic sheath enhancement (three patients), longitudinally extensive transverse myelitis (four patients), conus lesion (1 patient), central cord lesion (two patients), Ill-defined T2-hyperintensity involving pons, middle cerebellar peduncle or medulla (four patients), multiple ill-defined T2 hyperintense lesions in supratentorial and often infratentorial white matter (four patients), cortical lesions (two patients). In total 23 of 28 patients (82%) fullfield the 2023 MOGAD criteria. Didn´t met the criteria one case of isolated unilateral ON, one case of transerse myelites no-LETM, two case of double seropositivity and one case of optic neurits and transverse myelitis without additional supporting criteria. **Conclusion:** Eighty- two percent of the patients MOG-IgG positive met the new criteria for MOGAD, inspite of not having MOG-IgG titration.

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AQP4-IGG-seropositive neuromyelitis optica spectrum disorder (NMOSD) associated with teratoma and parotid tumor: a case report

Gabriel Isaac Correia[1], Elisa Gouvea Gutman[2], Caio Faria Tardim[1], Anna Beatriz Scherer Vanzan[2], Ingrid Pereira Marques[1], Carlos Eduardo Ferreira Ramos[2], Larissa Araujo Duarte[1], Marcos Martins da Silva[1], Roberto Pereira Santos[1], Soniza Vieira Alves-Leon[2]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Elisa Gouvea Gutman; Email: gutman811@gmail.com.

57-year-old female patient presented paresis and paresthesia in the right lower limb, accompanied by gait disturbance and urinary incontinence. Upon examination, she exhibited proximal predominance paresis in the right lower limb, hyperreflexia in the lower limbs predominantly on the left side, and diminished vibratory, tactile, and thermal-analgesic sensations with a T6 spinal level. During the investigation of myelitis, right ovarian teratoma and lymphoepithelial cyst in the right parotid were identified. Magnetic resonance imaging (MRI) revealed an extensive spinal cord lesion with longitudinal hypersignal from T3 to T7, predominantly central, without contrast enhancement. Brain imaging identified nodular lesions in the periventricular, subcortical, and medulla white matter, with hypersignal in T2 and FLAIR, without contrast enhancement or diffusion restriction. The hypotheses of Neuromyelitis Optica Spectrum Disorder (NMOSD) or paraneoplastic syndrome were considered. General biochemistry and rheumatological panels were normal. Cerebrospinal fluid analysis had normal findings and excluded infectious diseases. Antibody testing for anti-aquaporin 4 (AQP4) and anti-MOG IgG by ELISA method was negative. She evolved with improvement of the neurologic symptoms during the investigation and was discharged with outpatient follow-up. Six months later she evolved with similar symptoms. She tested negative for a new anti-MOG test, but positive for anti-AQP4, using the CBA method, establishing the diagnosis of NMOSD. She received pulse therapy with 1g/day of methylprednisolone for 5 days followed by 5 sessions of plasmapheresis on alternate days that did not yield significant improvement in symptoms. Treatment with azathioprine 100mg (2mg/kg/day) + prednisone 40mg/day was initiated, considering access to disease-modifying therapies in the Unified Health System. The relationship between teratomas, parotid tumor and NMOSD has been previously reported in literature, although the mechanism is still unclear. AQP4 has been detected in human parotid excretory duct epithelia, and a tumor with overexpression of excretory duct epithelial cells may lead to upregulation of AQP4 production. Additionally, histopathologic analyses of the teratomas revealed AQP4 immunoreactivity and infiltration of lymphocytes in the neural tissue component. In genetic susceptible patients, this can be the trigger to produce anti-AQP4 antibodies that induce the acute NMOSD attack. The coexistence of autoimmune disease and tumor may be explained by the immunotolerant role of HLA-G expression, inhibiting the immune responses of MHC-unrestricted NK and MHC restricted T cells. Presence of anti-AQP4-IgG does not exclude a paraneoplastic nature of NMOSD; however, paraneoplastic cases are rare and most of them are secondary to CNS lymphoma and intramedullary tumors. This case illustrates one more association between these conditions and reinforces the demand for further translational searches.

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Aquaporin-4 positive NMOSD patients have worse quality of life

Karina Nonato Pingituro Domingues[1], Samira Luísa dos Apóstolos Pereira[2], Raquel Vassão Araujo[3], Dagoberto Callegaro[2]

[1]NMO Brasil, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[3]Cronicos do Dia a Dia, Garulhos SP, Brazil

Correspondence: Karina Nonato Pingituro Domingues; Email: karina.npd@gmail.com.

Objective: To report quality of life in a cohort of people with Neuromyelitis Optica Spectrum Disorder (NMOSD), comparing patients based on the results of the Aquaporin-4 test. **Methods:** We evaluated 68 patients in six reference centers over a period of 18 months. Inclusion criteria was diagnosis of NMOSD according to IPND 2015. Demograhic and clinical data were evaluated. To evaluate life quality we use the Brazilian version of life quality SF-36. The research is in progress and will be applied in several regions of Brazil. **Results:** 68 NMOSD patients in the states of São Paulo, Paraíba, Goiás, Ceará, Bahia and Rio Grande do Sul, with 82% of female preponderance were evaluated. Median age was 41 and disease duration was 4.63 years. 33% are Aquaporin-4 positive, 31% Aquaporin-4 negative and 36% do not know or did not want to inform. According to the instrument, results are seen on a scale ranging from 0 to 100, where zero is the worst state and 100 is the best. Results obtained: regarding the physical functioning domain, the mean value obtained was 34 in Aquaporin-4 positive patients, 52 in Aquaporin-4 negative patients and 46 in patients who did not know/want to inform; regarding the role limitations due to physical health domain, the mean value obtained was 5 in Aquaporin-4 positive patients, 38 in Aquaporin-4 negative patients and 31 in patients who did not know/want to inform; regarding the social functioning domain, the mean value obtained was 45 in Aquaporin-4 positive patients, 59 in Aquaporin-4 negative patients and 63 in patients who did not know/want to inform; regarding the role limitations due to emotional problems domain, the mean value obtained was 9 in Aquaporin-4 positive patients, 36 in Aquaporin-4 negative patients and 50 in patients who did not know/want to inform. **Conclusion:** In this research, it was demonstrated that all domains had a worse result in Aquaporin-4 positive NMOSD patients, in relation to Aquaporin-4 negative patients or patients who were unable to inform the result, with greater emphasis on the domains limitation due to physical aspects and limitation due to emotional aspects, demonstrating a reduction in the Quality of Life of people with Aquaporin-4 positive NMOSD.

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Arboviruses evolving with Guillain-Barré syndrome and impact on immunoglobulin stock: a case report

Isadora Ribeiro Doria[1], Larissa da Cruz Cardoso[1], Sandy Karoline Carnaúba dos Santos[2], Augusto Costa Ataides[1], Beatriz Palladino Ferreira[1], Matheus Henrique Fonseca Melo[3], Agnaldo Queiroz Plaster[2], Maria Luísa Ribeiro Lima[4], Mireli Martins Montijo[1]

[1]Universidade do Estado de Mato Grosso, Cáceres MT, Brazil.
[2]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.
[3]AFYA, Itabuna BA, Brazil.
[4]Centro Universitário Governador Ozanam Coelho, Ubá MG, Brazil.

Correspondence: Isadora Ribeiro Doria; Email: isardoria@gmail.com.

Case presentation: There was a high incidence of arboviruses in this endemic region, with a significant number of cases progressing to a neurological condition, acute polyradiculoneuritis, also known as Guillain-Barré syndrome, with five cases in a one-month period. One patient remains hospitalized at this time, while the others have been discharged. EER, a 31-year-old female, reported 30 days of diffuse abdominal pain, high fever,

headache, tachycardia, dysphagia, right-sided hemiparesthesia, and subsequently bilateral hemiparesthesia. She arrived at the hospital in a wheelchair with the above-mentioned symptoms Chikungunya infection was identified in her history, as with most patients who developed this syndromic condition. They also denied drug allergies, comorbidities, alcohol use, and drug use, and all tested negative for Human Immunodeficiency Virus. On physical examination, she presented with tetraparesis predominantly on the left side, muscle strength grade II, with preserved tone and trophism. The patient developed proteinuria and leukopenia associated with polyneuropathy, with flaccid paraplegia, but remains hospitalized due to difficulties with treatment with IV Human Immunoglobulin. **Discussion:** Guillain-Barré syndrome is a rare but serious complication of arbovirus infections, and the recent surge in cases has overwhelmed the healthcare system. Specific treatment for this syndrome aims primarily to accelerate the recovery process, reduce complications associated with the acute phase, and involves the use of IV Human Immunoglobulin, compounded by the emergency situation of its supply at the state and national levels. Many patients in need of this treatment face challenges and clinical complications. **Final comments:** Therefore, it is crucial to study the etiology and pathophysiology of arbovirus progression to acute polyradiculoneuritis, along with developing alternative management protocols and treatments due to the persistent seasonal characteristics of this public health problem. This can directly impact the incidence of Guillain-Barré syndrome and its associated neurological syndromes, with socioepidemiological impacts that are still immeasurable.

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Area postrema syndrome as an initial manifestation of AQP4 -AB negative NMO spectrum disease treated with rituximab: a case report

Renata de Andrada Santana[1], Marina Papais Alvarenga[1], Regina Maria Papais Alvarenga[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.

Correspondence: Renata de Andrada Santana; Email: renata.andradaa@gmail.com.

Case presentation: A brown woman, currently 41 years old, reports that in 12/2012, at the age of 29, she suddenly felt intensely nauseous and developed incoherent vomiting. A few days later, she started having recurrent hiccups interspersed with vomiting and dyspnea. She was hospitalized for almost a month investigating gastric causes, and underwent endoscopy and radiological exams, being discharged without a diagnosis after improving. On January 30, 2013, she suddenly presented with urinary retention weakness of the lower limbs and imbalance, evolving within 72 hours with tetraplegia and numbness from the neck down. When she returned to the hospital, she underwent imaging tests. The MRI showed an extensive longitudinal lesion extending from the bulb to the lumbar spinal cord, with edema and contrast uptake. She was started on IV methylprednisolone (1 gram for five days) followed by human immunoglobulin and showed little improvement in her right upper limb deficit. In May 2013, she underwent four sessions of plasmapheresis, followed by exponential improvement in the paralysis of the lower limbs over the following months. The AQP4 antibody test was negative. The patient never showed any visual impairment. Preventive treatment was started with a monoclonal antibody, Rituximab, in 05/2013 at a dose of 500mg 1x a week for 4 weeks, followed by a dose of 500mg 1x in 6-6 months, accompanied by physiotherapy. Over the last 10 years, she has progressed with complete recovery of motor dysfunction and normalization of the MRI image of the spine. At the last assessment on 03/2024, she walked freely and the MRI study showed no lesions. The sequelae are neurogenic bladder and fecal urgency. **Discussion:** The area postrema is located around the IV ventricle in the medulla oblongata. Physiologically, it works as a chemosensitive vomiting/hiccupping center and an epicenter for monitoring and integrating systemic autonomic signals, together with the nucleus of the solitary tract and the dorsal motor nucleus of the vagus. Demyelination in this territory leads to episodes of nausea, vomiting, and incoercible hiccups lasting longer than two days and is known as Area Postrema Syndrome (APS). APS, optic neuritis, and longitudinally extensive myelitis constitute the three central signs of "Neuromyelitis Optica Spectrum Disorders" (NMOSD). These diseases are currently classified by serological status into AQP4 positive and negative. Treatment of these groups differs because the new FDA-approved drugs for NMOSD are only indicated for AQP4-positive cases. **Final comments:** In describing this rare case of AQP4-AB antibody-negative NMOSD initiated by APS, we emphasize the efficacy and safety of the off-label drug Rituximab. This CD 20 cell immunosuppressant shows excellent results for both AQP4-AB positive and negative NMOSD cases and is indicated by expert consensus as a first-line drug for these diseases.

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Arnold-Chiari malformation type I and chronic vocal cord paralysis: case report

Kelly Rejane Leite Figueiredo de Oliveira[1], Moacir Pereira Leite Neto[2], Ricardo Queiroz Gurgel[3], Pedro Garcia Dias de Barros[2], Isabel Monique Leite Romualdo[4], Cláudio Gleidiston Lima da Silva[2]

[1]AFYA, Fortaleza CE, Brazil.
[2]Universidade Federal do Cariri, Barbalha CE, Brazil.
[3]Universidade Federal de Sergipe, São Cristóvão SE, Brazil.
[4]Escola Estadual de Educação Profissional Presidente Médici, Campos Sales CE, Brazil.

Correspondence: Kelly Rejane Leite Figueiredo de Oliveira; Email: krlfigueiredo@gmail.com.

Case presentation: MCOLS, 53 years old, born in Saboeiro, Ceará. 28 years ago, she gradually developed dysphonia. Eight years later, she developed symmetrical paresis in her hands and paresthesia and pain in all four limbs, asymmetrical, particularly on the left. From then on, she developed difficulty walking and recurrent attacks of neck pain and left-sided hemicranial headache, the latter appearing in stitches, with strong intensity for up to 10 minutes, triggered by coughing or throbbing, with at least five episodes per month. On examination: dysphonic speech, symmetric palate elevation and reduced amplitude. Muscle strength grade 4: deltoid, rhomboid, biceps brachii, triceps, pronator teres, extensor carpi radialis, flexor carpi radialis, interossei and abductor pollicis brevis bilaterally. Grade 1 reflexes: styloradial, bicipital and triceps bilaterally. In sensitivity assessment, there is "shawl" hypoesthesia of C4-C6. Complementary exams: 10/21/2011 Laryngoscopy: nodular formation in the middle third of both vocal folds compatible with an intrachordal cyst; presence of irregular glottal cleft with double spindle predominance. 04/03/2017 Magnetic resonance imaging of the cervical spine: posterolateral disc protrusions at levels C5-C7, determining an impression on the ventral surface of the dural sac, causing a reduction in the amplitude of the left intervertebral foramen at C6-C7; herniation of the cerebellar tonsils through the foramen magnum by approximately 0.9 cm; extensive syringomyelic cavity from C2 to the thoracic segment. 03/13/2018 Electroneuromyography of the upper limbs: acute and chronic radiculopathy of the C7 root on the left side, with signs of active and chronic denervation in the flexor carpi radialis, extensor carpi radialis and left triceps muscles; sensory and motor neuropathy of the median nerve at the level of the wrist, bilaterally, compatible with severe carpal tunnel syndrome bilaterally. **Discussion:** Arnold-Chiari malformations are defined as a spectrum of abnormalities of the posterior part of the brain, involving the cerebellum, brain stem, skull base and spinal cord. Type I is the most common and is characterized by the caudal displacement of the cerebellar tonsils and the medial and inferior part of the posterior lobe of the cerebellum through the spinal canal, above 3 to 5 mm from the foramen magnum. The patient presented multiradiculopathy from C5 to T1, bilateral preganglionic hypoesthesia, C4-C6 hypoesthesia, motor deficit of the upper limbs caused by type I malformation. It was associated with extensive syringohydromyelia, dysphonia related to bulbar paresis, cervicogenic paroxysmal headache and cyst on the vocal folds. After a surgical approach, the headache, dysphonia, sensory and motor deficits gradually resolved after surgical treatment of the Chiari malformation, as well as neurophysiological recovery from the assessed radiculopathy.

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Arquivos de Neuro-Psiquiatria: a bibliographic study of the first 20 years (1943-1963)

Rodrigo Fagundes da Rosa[1], Francisco Duque de Paiva Giudice Junior[1], Francisco Luciano Honório Barreto Cavalcante[1], Leonardo Elias Araujo dos Santos[1], Emanuela Freire Caetano Davi[1], João Marcelo Albuquerque Beserra de Sousa[1], Mariana Alcântara Tavares[1], Wagner Patrik Sousa Silva[1], Caio Levi Mendes Costa[1], Francisco de Assis Aquino Gondim[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Rodrigo Fagundes da Rosa; Email: rodrigofdarosa@gmail.com.

Background: In 2023, the Arquivos de Neuro-Psiquiatria celebrated 80 years. Founded in June 1943 by Oswaldo Lange, Adherbal Tolosa, and Paulino Longo, it has been the official journal of the Brazilian Academy of Neurology (ABN) since 1970. Over the decades, it has undergone several upgrades, including indexing in international databases, peer review, and transitioning to open access, reaffirming its commitment to academic excellence and the internationalization of Brazilian research. This article, the first in a series of four, analyzes the first 20 years of Arquivos de Neuro-Psiquiatria. **Objective:** To analyze publication trends, authorship patterns, and editorial changes over the first 20 years, highlighting its historical significance. **Methods:** All editions published by the journal in its digital repository from 1943 to 1963 were analyzed, with each article being examined individually. Additionally, all physical editions available in the Professor Jurandir Marães Picanço Health Sciences Library were also scrutinized. Data from both sources were tabulated independently by two blinded, independent researchers. Subsequently, those two individuals cross-verified the collected data. **Results:** Between 1943 and 1963, the journal published 83 issues and 2 supplements in 21 volumes, totaling 923 articles. The annual publication rate was 4 issues, except in 1943, with 3 issues, and in 1954, with 6 publications due to two extra supplements. The study analyzed 479 articles classified as article, case report, update, practical note, and technical note, excluding book reviews, tributes, and non-original articles. A total of 893 authors participated, averaging 1.91 authors per article. The articles were categorized as follows: 325 articles, 109 case reports, 19 practical notes, 13 preliminary notes, 9 updates and 9 in other categories. Horacio Martins Canelas was the most prolific author, contributing with 35 articles, followed by Rolando ngelo Tenuto with 29. Only 3.92% were women, notably Maria Elisa Bierrenbach Khoury, the first woman to co-author an article, Iracy Doyle with 3 main authorships, and Maria I. Valente with 1 authorship and 6 co-authorships. Geographically, 418 articles came from Brazil,

notably São Paulo (352), Rio de Janeiro (21), and Bahia (21), as well as 61 studies from other countries. Oswaldo Lange was the main Editor throughout the entire study period. **Conclusion:** The first 20 years of the Arquivos de Neuro-Psiquiatria established it as a vital platform for disseminating knowledge, with significant contributions from Brazilian researchers. Although female participation was low, the journal took steps toward inclusion. The stability of the editorial board and the involvement of influential authors were crucial for the journal's growth and quality, laying a solid foundation for its continued trajectory of academic excellence and internationalization.

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Artificial intelligence algorithms to detect cerebral small vessel disease tomography markers and their association with clinical outcomes in patients with acute ischemic stroke undergone thrombectomy

Thiago Oscar Goulart[1], Rui Kléber Do Vale Martins-Filho[2], Millene Rodrigues Camilo[2], Octávio Marques Pontes-Neto[2]

[1]Harvard, Boston MA, USA.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.

Correspondence: Thiago Oscar Goulart; Email: tgoulart@hsph.harvard.edu.

Background: Although mechanical thrombectomy (MT) has expanded its indications over the last years, establishing a robust benefit even for the elderly, some trials have shown neutral effects regarding this subgroup. One possible explanation for these findings is the vulnerability of the patients treated under these trials, which means these populations were frailer and, accordingly, portended a significant burden of cerebral small vessel disease (cSVD), which may work as a surrogate marker for brain frailty. **Objective:** To study the influence of the cSVD markers on the baseline CT scan in the functional outcome and develop Artificial Intelligence (AI) algorythms to detect them. **Methods:** A trained vascular neurologist blinded to clinical details evaluated the CT scans of 351 patients. We focused on markers of cSVD (leukoaraiosis, old lacunes, atrophy), and calculated the Brain Frailty Score (BFS) and modified Small Vessel Disease score (mSVD) (Figures 1 and 2). Then we carried out a multivariable back-stepwise logistic regression and random forest to identify the variables related to disability at 90 days, and other model to analyze the association with Hemorrhagic Transformation. We segmented the images with 3DSlicer and develop an U-Net model to detect leukoaraosis. **Results:** The group with disability at 90 days was older, had higher NIHSS at admission, more prevalence of diabetes mellitus (DM), higher levels of systolic blood pressure and glucose levels at admission, poorer collaterals, lower rate of MT, and higher rates of severe cSVD markers at baseline CT scan. Figure 3 shows the variables associated with poor outcomes in a back-stepwise logistic regression. The model performance was excellent: AUC=0.962 (Figure 3), with no signs of heteroskedasticity (Figure 4). In other models with HT and Symptomatic HT as dependent variables, cSVD markers also were associated with these outcomes. The random forest model determined the variables importance in these models (Figure5). The U-net model had great performance to detect severe leukoariosis (accuracy=100%, macro-precision=100%, AUC of ROC curve=1.00) **Conclusion:** Markers of cSVD on baseline CT scans are related to poorer outcomes, such as disability, mortality and symptomatic Hemorrhagic Transformation in patients with LVO and might be one of the main causes of the lack of benefit of MT in elderly patients, highlighting its importance even in acute LVO context. We also developed a AI software with great performance to detect severe leukoaraiosis.

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Artificial intelligence algorithms to differentiate between contrast extravasation and hemorrhagic transformation in tomography of acute ischemic stroke after thrombectomy

Thiago Oscar Goulart[1], Júlio César Nather Júnior[2], Renato Okabayashi Miyaji[3], Danilo Pinheiro Fonseca[4], Octávio Marques Pontes-Neto[4], Millene Rodrigues Camilo[4]

[1]Harvard, Boston MA, USA.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.
[3]Universidade de São Paulo, São Paulo SP, Brazil.
[4]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Thiago Oscar Goulart; Email: tgoulart@hsph.harvard.edu.

Background: Mechanical Thrombectomy (MT) treats Acute Ischemic Stroke (AIS) removing the thrombus, and improves clinical outcome. However, it uses high quantities of contrast, with risk of Contrast Extravasation (CE), but also Hemorrhagic Transformation (HT), both detected by cranial Computed Tomography (CT). However, to differentiate between them, it is necessary to perform a control CT 72 hours after MT (Figure 1). **Objective:** To develop a tool capable of differentiating HT from EC in cranial CT. **Methods:** 398 CTs were classified into four classes: HT, CE, HT with CE, and without HT nor CE. We selected 111 CT performed until 72 hours after angiotomography+MT, segmented the images with 3DSlicer (Figure 2). Then, we had 2 approaches: 1) Three classification models were developed based on phenotypic characteristics: Support Vector Machines (SVM), Random Forest (RF) and Logistic Regression (LR); 2) The U-Net, a Convolutional Neural Networks (CNN) model for biomedical image segmentation. **Results:** SVM and RF had ROC AUC above 92% for the three classes. Cross-validation demonstrated good accuracy (SVM=0.947±0.064; RF=0.893±0.076; LR=0.956±0.048) and macro-precision (SVM=0.947 ± 0.066. RF=0.901±0.075, LR=0.956±0.049). The U-Net also presented good predictive performance after cross-validation: accuracy=0.956±0.072 and macro-precision = 0.965±0.055. Multiclass ROC Curves exhibited AUC ≥ 0.98 for the three classes (Figure 3). **Conclusion:** In this study, we developed a tool with high performance to distinguish between CE and HT with two different approaches, which could help in clinical decision-making, such as introduction of anticoagulants and antiaggregants

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Aseptic meningoventriculitis after spinal anesthesia and Epstein Barr finding as a differential diagnosis of rapidly progressive dementia: case report

Luciano Gonçalves do Nascimento Júnior[1], Gilmar Leite Pessoa Filho[1], Felipe Nunes de Araújo[1], Ana Luisa Castelo Branco Gomes[1], Rafael de Souza Andrade[1], Paulo Antonio Farias Lucena[1], Luíza Alves Monteiro Torreão Villarim[1], Alex Tiburtino Meira[1], Matheus Gurgel Saraiva[1], Juliana Magalhães Leite[1]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.

Correspondence: Gilmar Leite Pessoa Filho; Email: gilmarpessoa@gmail.com.

Case presentation: A 61-year-old female patient from João Pessoa (Paraíba), with a medical history of hypertension and diabetes, underwent uterine polypectomy under spinal anesthesia. Four days post-procedure, she developed an intense headache associated with nausea and vomiting, progressing subacutely to disorientation, recent memory difficulty, disinhibition, incoherent speech, and gait disturbance, and initiated investigation for rapidly progressive dementia. After 40 days, with progressive worsening, she was admitted in a somnolent state, with neck stiffness, bradykinesia in the upper limbs (worse on the right), and left VI paresis. Initial computed tomography revealed cortical sulci effacement with marked supratentorial ventricular dilatation. Contrast-enhanced magnetic resonance imaging demonstrated signs of inflammatory/infectious leptomeningitis affecting cortical gyri and basal cisterns, changes suggesting ventriculitis with moderate supratentorial ventricular dilatation; FIESTA sequence revealed aqueductal stenosis with thin septations. Cerebrospinal fluid analysis revealed limpid fluid with glucose of 69; proteins 111; leukocytes 20 (80% mononuclear); non-reactive VDRL and negative bacterial, fungal cultures, and malignant cells. Viral panel by RT-PCR detected Epstein-Barr Virus (EBV). Serologies for hepatitis C and B, HIV, and VDRL were negative. Initial empiric treatment included vancomycin 2g/day, meropenem 6g/day, and dexamethasone 16 mg/day for 21 days. After aseptic ventriculitis was confirmed, with serial negative CSF cultures, a ventriculoperitoneal shunt was performed. The patient improved cognitively and in his level of consciousness. At 8-month follow-up, she returned to the neurology outpatient clinic with daily activities maintenance and remission of previous symptoms. **Discussion:** Post-spinal anesthesia meningitis is extremely rare (< 0.01%) with potentially severe complications. The reported case suggests DIAM, diagnosed based on a high suspicion level. Aseptic cerebrospinal fluid confirms the diagnosis, facilitating antibiotic withdrawal, shorter hospital stay, and reduced costs. **Final comments:** Serial investigations ruled out infectious causes for the clinical-radiological presentation, suggesting inflammatory process as a cause for aqueductal septations. The presence of EBV in the case is uncertain as to whether it is a concomitant finding or a causative and aggravating agent. The patient had a favorable clinical outcome.

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Assesment of the quality of life of elderly individuals attending a senior community center

Natalli Studnicka[1], Tatiana Pizzolotto Bruch[1], Camila Pezente Dal Toe[1], Pedro Mendes Maciel[1], Virgílio Fernandes Bronzatti[1], Gustavo Beltrame[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Natalli Studnicka; Email: natalli.stu@gmail.com.

A study was conducted to assess the quality of life of elderly attendees of a senior community center in Criciúma. During the first semester of 2023, 286 elderly individuals were interviewed. The questionnaire covered personal data such as gender, age, race/ethnicity, marital status, education level, and socioeconomic status, and included an evaluation of quality of life using the WHOQOL-OLD scale. Results showed an Overall Score of 4.16 for the elderly individuals' quality of life. The highest scoring domain was "Autonomy" (4.24), while the lowest was "Death and Dying" (4.08). Statistical analyses revealed significant differences between genders and between married and separated/divorced individuals in the "Past, Present, and Future Activities" domain. Differences were also found between married and single individuals in the "Social Participation" and "Intimacy" domains. However, no statistically significant differences were found between the variables and the other domains of the WHOQOL-OLD. In conclusion, elderly participants in the senior

community center demonstrated overall good quality of life, particularly in autonomy. Married individuals exhibited better quality of life than singles in domains related to activities, social participation, and intimacy as assessed by the WHOQOL-OLD.

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Assessing dysphagia and ventilatory dysfunction in inclusion body myositis: clinical insights and muscle ultrasound findings

Ana Lucila Moreira[1], Bruna Moreira de Souza Proença[1], André Macedo Serafim da Silva[1], Cristiane de Araújo Martins Moreno[1], Eliene Dutra Campos[1], Mariana Cunha Artilheiro[1], Tatiana Ribeiro Fernandes[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas,São Paulo SP, Brazil.

Correspondence: Bruna Moreira de Souza Proença; Email: brunamsproenca@gmail.com.

Background: Inclusion Body Myositis (IBM) is characterized by progressive muscle weakness, with dysphagia affecting 40-80% of patients. Weakness in the pharynx and tongue significantly contributes to dysphagia, which can occur at any disease stage. Patients often adapt to minor swallowing difficulties over time, while asymptomatic respiratory function impairment is also common. Its impact on patients' quality of life is profound, while aspiration pneumonia and respiratory complications are the most common causes of death. Therefore, monitoring for swallowing and respiratory complications is crucial in managing IBM. Imaging techniques, like ultrasound (US), offer potential as non-invasive tools for monitoring muscle changes. The US morphologic study of the tongue and respiratory muscles may provide valuable insights into disease processes. **Objective:** To evaluate dysphagia and respiratory impairment using US in conjunction with clinical analyses. **Methods:** We conducted a cross-sectional study involving ten IBM patients. We analyzed clinical and epidemiological data descriptively to characterize the patient population. Dysphagia assessment used the IBM Functional Rating Scale (IBM-FRS), and we categorized ventilatory restriction based on FVC measurements by spirometry. We examined the patients with B-mode ultrasound, evaluating the diaphragm, intercostals, and tongue muscles (digastrics, geniohyoid, and genioglossus) in the transverse plane. All examinations took place on the right side using a fixed preset. Muscle classification followed the Modified Heckmatt Scale (1=normal, 2=probably abnormal, 3=abnormal). **Results:** The study included ten IBM patients, with 4 males and 6 females, with a median age of 61.5 years and median duration of disease of 9 years. Dysphagia was present in 60% of the patients, who reported occasional choking. US findings of the tongue muscles showed that the genioglossus muscle was altered in all patients and the least altered muscle in the tongue was the geniohyoid. In terms of respiratory function, the FVC of the patients ranged from 25% to 93% of the predicted values, with a median FVC of 67.0%. Ventilatory restriction was observed in the following proportions: 33.3% normal, 11.1% mild, 33.3% moderate, 11.1% severe, and 11.1% very severe. US evaluation of the respiratory muscles revealed that the intercostal muscles and the diaphragm were normal in only 20% of the patients. **Conclusion:** Abnormalities in the genioglossus muscle were frequently observed in patients with dysphagia. Ventilatory restriction was commonly seen in patients with abnormalities in the respiratory muscles. US evaluation of the tongue and respiratory muscles can provide insight into disease processes and guide therapeutic decisions even in patients without complaints of dysphagia or difficulty breathing. This evaluation may serve as an objective assessment of treatment response in the future.

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Assessing stroke awareness in Aparecida-SP city: a cross-sectional study from 2023 and 2024

Beatriz Barbosa Nishigima[1], Mariana Rachas Reis[1], Anna Luísa Mennitti[1], Fernanda Yumi Abe[1], Rubens José Gagliardi[1]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.

Correspondence: Beatriz Barbosa Nishigima; Email: beatriz.nishigima@gmail.com.

Background: Stroke is a leading cause of mortality and disability in Brazil. Despite its severity, it often goes unrecognized, leading to delayed treatment and unfavorable outcomes. This research, conducted as part of a community outreach program, aims to assess the level of stroke awareness among residents of Aparecida-SP city, from 2023 to 2024. By identifying knowledge gaps, it is possible to develop targeted interventions to improve stroke prevention and management, thereby mitigating its impact on communities. **Objective:** To conduct a descriptive analysis of the overall knowledge concerning stroke recognition and prevention in a low-income community in Aparecida (SP), between 2023 and 2024. **Methods:** This is a descriptive cross-sectional study on data from a structured questionnaire available on the REDCAP platform applied in Aparecida-SP city in January 2023 and 2024, during a community outreach program organized by medical, nursing and speech therapy students. The study population was composed of people aged 18 years or older, who were registered for an outpatient appointment and signed the term of consent. In 2024, previous respondents were excluded. They were analyzed demographically and the questions about stroke included definition, symptoms, prevention methods, what to do in case of suspicion and if they knew someone who had a stroke. **Results:** Collecting data in both years resulted in a sample of 534 answers, 258 in 2023 and 276 in 2024. Among all interviewees, 63.8% were women, the most prevalent age group was between 50-59 years (28,3%) and 31.6% of them had incomplete primary education. In the evaluated sample, 71.9% knew someone who had a stroke and 60.7% couldn't define the disease. Considering the total participants, 304 people (57%) know how to identify at least one of the stroke symptoms out of a list. This list included, from the most to least cited: paresthesia in the face, arm or leg (52%), weakness in the face, arm or leg (42.4%), mental confusion, difficulty speaking or understanding (38.5%), severe headache of sudden onset and unknown cause (35.5%), difficulty with movement balance and coordination (34.2%), hemilateral symptoms (30.6%) and vision impairment (15.2%). When asked about preventive measures, 272 (52.8%) knew at list one, being physical activity the most mentioned (69.5%) and dyslipidemia control (17.8%) the least commented in both years. On the occasion of witnessing a stroke, 58.6% would call an emergency service, 0.74% would check the timing and 40.6% wouldn't know what to do. **Conclusion:** Despite the prevalence and mortality of stroke, the population knows little essential information about it. This lack of knowledge contributes to the high incidence of the disease. Therefore, it is important to promote campaigns and healthcare policies to raise awareness of stroke and its prevention.

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Assessing the efficacy of donanemab in reducing amyloid plaque levels in Alzheimer's disease: a systematic review and meta-analysis of randomized clinical trials

Victor Hugo Palhares Flávio-Reis[1], Yago Marcos Pessoa-Gonçalves[1], Marlos Aureliano Dias-Sousa[2], Gustavo de Oliveira Almeida[1], Chamberttan Souza Desidério[1], Carlo José Freire Oliveira[1], Wellington Francisco Rodrigues[1]

[1]Universidade Federal do Triângulo Mineiro, Instituto de Ciências Naturais e Biológicas, Laboratório de Imunologia e Bioinformática, Uberaba MG, Brazil. [2]Universidade Federal do Triângulo Mineiro, Hospital de Clínicas, Departamento de Neurologia, Uberaba MG, Brazil.

Correspondence: Yago Marcos Pessoa-Gonçalves; Email: yagompg98@gmail.com.

Background: Alzheimer's disease (AD) is characterized by the accumulation of beta-amyloid (Aβ) peptides plaques in the brain. Donanemab was developed to bind to amyloid plaques and operate through microglial-mediated phagocytosis. In animal studies, this drug has demonstrated efficacy in reducing the number of plaques without causing microhemorrhages. **Objective:** To evaluate the efficacy in humans of Donanemab for the reduction of amyloid plaque levels in AD. **Methods:** This study is a meta-analysis of randomized controlled trials that evaluated the efficacy of Donanemab in reducing amyloid plaque levels of patients with AD. Embase, Pubmed and Cochrane were searched for trials published up to may 2024. Data were extracted from published trials and Cochrane tool for assessing risk of bias was employed. The extracted data were input into Review Manager (RevMan) 5.3 (Nordic Cochrane Centre, Denmark) to calculate the effect size. RevMan calculated as effect measure the mean difference with a 95% confidence interval (CI) using the inverse variance statistical method. **Results:** Regarding the risk of bias in each RCT, three were assessed to have an overall low risk of bias. Only one RCT was found to have some concerns regarding the risk of bias. Out of 528 database results, 146 duplicate articles were excluded. Of the remaining 382 studies, only 4 studies met the inclusion criteria. Four randomized controlled trials comprising 2142 patients were included. Of the participants studied, 1060 (51%) received Donanemab and 1072 (49%) received placebo. Donanemab was associated with a significant reduction of 84.68 amyloid plaque levels (95% CI, -89.77 to -79.58, P<0.00001) when compared to placebo. When conducting subgroup analyses and assessing secondary outcomes, it was observed that Donanemab (95% CI, 1.68 to 4.27, P<0.00001) presented a Integrated Alzheimer's Disease Rating Scale (iADRS - range from 0 to 144, with lower scores indicating greater cognitive deficits) value of 2.97, meaning a significant improvement in patients performing activities of daily living. Regarding the Clinical Dementia Rating Scale-Sum of Boxes (CDR-SB; scores range from 0 to 18, with higher scores indicating greater impairment), no statistically significant difference was observed, with a combined value of -0.29 (95% CI, -1.81 to 1.23, P=0.71). **Conclusion:** This meta-analysis suggests that Donanemab is capable of reducing amyloid plaque levels in patients with AD. Additionally, improvements in the iADRS scale were also noted, although the CDR-SB scale did not show statistical significance. Randomized trials are warranted to further investigate this question.

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Assessing the impact of serological profiles on clinical outcomes in suspected paraneoplastic neurological syndromes

Mariana Ribeiro Pereira[1], Ana Flávia Andrade Lemos[1], Amanda Vitória Freire de Oliveira[1], Lucas Gondim Briand Vieira[1], André Cleriston José dos Santos[1], Rodrigo S. S. Frezatti[1], Giuliana França Teixeira Mandia Grossi[1], Vanessa Daccach[1], Pedro José Tomaselli[1], Wilson Marques Junior[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.

Correspondence: Mariana Ribeiro Pereira; Email: pribeiromariana@gmail.com.

Background: Paraneoplastic neurological syndromes are a rare group of disorders caused by autoimmune responses that are often, but not always, associated with known cancers. Antibodies directed to intracellular neuronal antigens and neuronal cell-surface or synaptic antigens have been described as causative. Among paraneoplastic autoantibodies, anti-Hu, anti-Ma2/Ta, anti-Ma, anti-Ri, and anti-Yo antibodies are known for their association with various neurological manifestations. **Objective:** To analyze patients serological profile and clinical evolution with suspected paraneoplastic neurological syndromes. **Methods:** Clinical and serological data were retrospectively gathered from patient records using a standardized proforma. Data collection included key parameters such as sex, age at disease onset, type of neurological syndrome, cancer type (if present), and clinical outcomes. 274 patients with neurological disorders suspected to be paraneoplastic were included. **Results:** Neuronal autoantibodies were detected in 10/274 (3.78%) of patients. A broad range of neurological symptoms was recorded in patients who tested positive, including cerebellar and sensory ataxia (5), rapidly progressive dementia (2), multiple mononeuritis (1), lower motor neuron syndrome (1), and unresponsible bilateral optic neuritis (1). The most frequently detected autoantibodies were Yo (4/10, 40%), followed by Hu (2/10, 20%) and Ma2/Ta (2/10, 20%). Anti-Ma antibodies were identified in one patient, and anti-Ri antibodies were found in another. Among the 264 patients who tested negative for antibodies, a wide range of diagnoses were observed, with no clear association with specific types of cancer. This group generally exhibited a better prognosis and more favorable responses to symptomatic treatment. Patients positive for paraneoplastic autoantibodies exhibited severe and more rapidly progressive neurological manifestations, regardless of the associated cancer. These findings suggest that the presence of these autoantibodies is a critical factor in managing these patients. However, our cohort shows reduced sensitivity and specificity in detecting a positive diagnosis, possibly suggesting an elevated testing rate. **Conclusion:** The presence of neuronal autoantibodies is associated with more severe neurological manifestations irrespective of association with cancer. In contrast, patients negative for these autoantibodies presented with more varied neurological symptoms and generally better prognosis. Therefore, early identification of autoantibodies remains crucial for patient care, even in the absence of detectable tumors.

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Assessment and management of patients with syndromic craniosynostosis associated with hydrocephalus

Letícia Mansano de Souza[1], Sérgio Lourenço Polo Filho[1], João Pedro Leite Pereira[1], Fernando Cendes[1], Enrico Ghizoni[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Sérgio Lourenço Polo Filho; Email: s205772@dac.unicamp.br.

Craniosynostosis is the premature closure of cranial sutures, causing deformities and cranioencephalic disproportion. It can be diagnosed before birth with imaging methods. There are primary types, simple or complex, and secondary types, with complexes being syndromic and non-syndromic. Syndromic forms are linked to genetic conditions such as Apert syndrome. The main treatment is surgery to mitigate complications such as intracranial hypertension and hydrocephalus, focusing on reducing intracranial pressure to preserve cognitive and visual functions. The present study, with a retrospective and non-randomized cohort design, aimed to establish correlations between the presence of hydrocephalus in patients with syndromic craniosynostosis, whether or not undergoing surgical procedures for treatment in a tertiary hospital. The computed tomography images were analyzed in the hospital database using the Horos 4.0.1 for Apple software to determine the size of the anatomical structures of the project patients. A total of 21 patients with syndromic craniosynostosis were included in the research, with a mean age of 5 years (standard deviation of 2.2). In the sample, 43.2% of patients were female. Of the patients evaluated, 71.6% of patients had Apert syndrome, 14.92% had Crouzon, 10.44% had Pfeiffer and 2% had Chotzen. In the neuroimaging exams of the research patients, 61.9% showed early closure of the bilateral coronal sutures, with a brachycephalic skull shape. Anomalous emissary veins were observed in 57% of patients, 76% of patients had hypoplastic jugular foramen. In most of the analyzed cases of Apert, Pfeiffer and Crouzon syndromes, a higher incidence of development of hydrocephalus was observed compared to ventriculomegaly. Hydrocephalus is more common in these patients due to compression of intracranial structures and disturbances in CSF absorption, which can result in changes in cranial anatomy. Surgical approaches were used in the research patients, including posterior decompression (40.4% of the sample), posterior distraction (33.3%) and fronto-orbital advancement (21.4%), also known as FOA, and posterior decompression or distraction plus FOA (4.8%). In post-surgical neuroimaging studies, encephalomalacia was observed, frequently associated with focal epilepsy, suggesting pathophysiology similar to traumatic epilepsy, non-genetic or resulting from brain malformations. Among patients with epilepsy, 61.1% developed the disease after surgery, while 38.8% already had it before the surgical procedure. Craniosynostosis presents a complex challenge that involves early diagnosis, multidisciplinary management and specialized surgical treatment. This study provides valuable information about the association between different genetic syndromes and the development of hydrocephalus. However, there are challenges such as the continued need for research to better understand and personalized treatment of these complex conditions.

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Assessment of changes in the sleep/wake cycle of medical students at a private university

Clara Alves Caram[1], Lucas Seri Iijima[1], Mércia Tancredo[1]

[1]Pontifícia Universidade Católica de São Paulo, Sorocaba SP, Brazil.

Correspondence: Clara Alves Caram; Email: claracaram@outlook.com.

Background: Sleep is an essential biological process for humans and has an important role in maintaining homeostasis in the body's environment. It is a complex state of physiological processes with its own behavioral characteristics. Studies show that anxiety disorders and depression affect a high proportion of university students. Among Medicine students, this rate is even higher, due to the high workload and emotional stress faced by students on this course. For this reason, many students use psychotropic drugs, such as antidepressants and anxiolytics, in order to reduce the symptoms of these diseases. Discovered in the 1950s, these medications act on the central nervous system and have a direct impact on the sleep/wake cycle. This, in turn, is essential for the proper functioning of higher brain functions such as memory and learning. **Objective:** To understand changes in the sleep/wake cycle of medical students at a private university. **Methods:** After approval by the Ethics and Research Committee and acceptance of the Free and Informed Consent Form, medical students at a private university responded to a digital form (Google Forms®) developed for this research, on sleep/wake rhythm and the use of psychotropic drugs, with questions based on the Pittsburgh Sleep Quality Index, adapted for Brazil. **Results:** Among 102 students, the majority had good or very good sleep (56.9% and 11.7% respectively), but 24.5% had poor sleep and 6.8% had very poor sleep. In relation to students with very poor or poor sleep, 21.6% of students use antidepressants/anxiolytics and 6.8% do not. Most (68%) have been using psychotropic drugs for more than 2 years. Furthermore, 59.6% use it before starting graduation. When segmenting for clinical indication, the majority (90%) of participants with poor or very poor sleep experience anxiety and/or depression. Furthermore, 68% resorted to therapy and only 3% use sleeping pills. **Conclusion:** The predominant profile of medical students with changes in the sleep/wake cycle is mainly characterized by the diagnosis of anxiety and/or depression, use of antidepressants and/or anxiolytics prior to entering college and for more than 2 years and positively the search for complementary treatment with individual psychotherapy and the non-use of specific sleep medications.

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Assessment of disability in patients with progressive multiple sclerosis in a Brazilian hospital

Marcelle Sanjuan Ganem Prado[1], Amanda Selvatici dos Santos Dias[1], Camila Lopes Figueiredo[1], Darah Fontes da Silva Assunção[2], Eliana Tomomi Shimabukuro da Cunha[1], Tatiane Souza Gutierrez[1], Guilherme Sciascia Do Olival[1], Maria Fernanda Mendes[3]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.
[3]Santa Casa de Misericórdia de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.

Correspondence: Marcelle Sanjuan Ganem Prado; Email: cellesanjuan@yahoo.com.br.

Background: Progressive multiple sclerosis (PMS) is a type of MS that leads to a gradual increase in disability without relapses. **Objective:** To evaluate changes in EDSS functions in patients with Progressive Multiple Sclerosis (pwMSP), aiming to define rehabilitation strategies to improve quality of life (QoL). **Methods:** It is a retrospective observational study involving 33 patients diagnosed with pwMSP who were treated at Santa Casa de Misericórdia hospital in São Paulo, Brazil. Medical records were analyzed between the months of December/23 to March/24. The EDSS scale and functional systems analysis were used to collect data. Person's Chi-square test was used for statistical analysis. **Results:** Of the 33 patients evaluated with pwMSP, 6.1% (2) had EDSS less than or equal to 5.0; 54.5% (18) had EDSS between 5.5 and 6.5 and 39.4% (13) had EDSS greater than 7.0. Younger ages were observed in the group with EDSS greater than 7 (48.5 years) compared to those with EDSS < 5.5 and EDSS between 5.5 and 6.5 (56 yo and 58.9 yo respectively). Among the most altered parameters in EDSS have brain stem function (p=0.05), mainly dysarthria, in patients with EDSS greater than 7. As for the complaint of bladder changes, this was more present in EDSS greater than 7.0 (p=0.03), the same did not occur with intestinal functions. Cognitive and coordination changes in the upper limbs were the most frequently observed sequelae in patients with minor EDSS, although without statistical significance. **Conclusion:** Although the sample is small, the findings provide us with information about the main characteristics of pwMSP in the studied group, indicating the needs of pwMS regarding possible interventions to improve QoL.

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Assessment of respiratory function in Friedreich's ataxia

Celiana Figueiredo Viana[1], Cristina Saade Jaques[1], Flavio Moura Rezende[1], Orlando Graziani Povoas Barsottini[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Celiana Figueiredo Viana; Email: celianafigueiredo@hotmail.com.

Background: Friedreich's ataxia (FRDA) is the most common cause of autosomal recessive ataxia. It is caused by expanded guanine-adenine-adenine (GAA) triplet repeats in the first intron of the frataxin gene (FXN), resulting in reduction of mRNA and protein levels of frataxin in different tissues. FRDA can lead to sleep disorders, respiratory complications and dysphagia. The purpose of this study is to evaluate the respiratory function in Friedreich ataxia. **Objective:** To evaluate the lung function of individuals diagnosed with Friedreich's ataxia through respiratory function tests and phrenic nerve conduction when compared to the healthy controls. Furthermore, to correlate respiratory assessment data with demographic data (age, duration of the disease), anthropometric data (weight, height, BMI, cervical and abdominal circumference) SARA and ICARS scales on the Friedreich's group. **Methods:** A total of 15 individuals with FRDA and 20 healthy controls were recruited. The patients and controls underwent, spirometry assessment, inspiratory and expiratory pression test, sniff test and Peak cough flow test. **Results:** Thirty-five participants, with an age of 37.1 years (standard deviation 11.0) were included in the study. Among them, 42.9% (15 participants) were from the Friedreich's group, while 57.1% (20 participants) were healthy controls. There were no differences in sex or age between the groups, but Friedreich's group had significantly lower weight, height, and BMI ($p=0.04$, $p<0.01$ and $p<0.01$, respectively). Friedreich's group had significantly lower values of respiratory parameters (forced vital capacity (FVC), forced expiratory volume in first second (FEV1), PEF, oxygen saturation, respiratory and cardiac frequencies, maximal inspiratory effort in the supine and seated positions, and sniff nasal inspiratory pressure, (all parameters with $p<0.01$), when compared to the control group. There were no differences among the mean and maximum phrenic amplitude's ($p=0.42$ and $p=0.37$, respectively) between groups. For both SARA ($rho=0.70$, $p<0.01$) and ICARS (0.59; $p=0.03$), the Friedreich's group shows a moderate correlation in the duration of disease. There was no statistically significant correlation among SARA or ICARS scale and anthropometric data. **Conclusion:** Evaluating respiratory function in patients with FRDA is crucial for guiding respiratory physiotherapy and preventing life-threatening complications. More studies are necessary to identify respiratory complications in Friedreich ataxia and these findings could be useful when approaching progressive neurological disorders.

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Assessment of the accuracy of stroke protocol screening by the emergency medical services

Victoria Cavalcanti de Souza[1], Ester Namie Hanai[1], Alice Volpato Rocha[1], Laura Louise Marques Nobrega Vieira[1], Gabriel Volpato Rocha[1], Viviane de Hiroki Flumignan Zétola[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Victoria Cavalcanti de Souza; Email: victoriacavalcantisouza@gmail.com.

Background: Early recognition of Stroke (Cerebrovascular Accident, CVA) within the treatment window directly influences patient outcomes and prognosis. Most patients will access qualified healthcare centers through the ambulance service. However, we have limited data on the challenges in the prehospital period and the screening performed. **Objective:** To evaluate the accuracy of the Stroke Protocol of the Emergency Medical Service (SAMU) in identifying and referring stroke cases. **Methods:** This is an observational retrospective study in which patients directed by the Stroke Protocol via SAMU to a designated hospital in Curitiba from January 2023 to May 2024 were included in the analysis. The evaluated outcome was the final diagnosis upon patient admission, based on medical record review. In cases where the diagnosis was not specified in the patient charts, complementary imaging exams were reviewed with the assistance of a specialist neurologist. Cases that remained without a specific diagnosis were grouped separately. **Results:** Medical records of 297 patients were reviewed. 206 cases (69%) were confirmed with a diagnosis of Stroke (ischemic [176; 85.43%] and hemorrhagic [30; 14.56%]), thus accurately diagnosed by SAMU (accuracy of 69%). Among the main differential diagnoses were seizure (29; 9.76%), stroke mimics (17; 5.72%), syncope (6; 2.02%), headache (6; 2.02%), and subarachnoid hemorrhage (4; 1.35%). 18 patients (6.06%) were grouped into other diagnoses, and 11 (3.70%) did not have a specified diagnosis at the time of hospital discharge. **Conclusion:** The prehospital diagnostic accuracy of the SAMU Stroke Protocol was 69%. Therefore, there is plenty of room for improvement in diagnostic precision. It is important to continue conducting training sessions with the National Institutes of Health Stroke Scale (NIHSS) and to make adjustments and enhancements to ensure the advancement of the Stroke Protocol and provide a better prognosis for those affected by stroke within the appropriate time frame.

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Assessment of the pain perception threshold by electrical algometry in migraine and healthy individuals: preliminary study

Renata Michelini Guidi[1], Felipe Franco de Graça[2], Antônio Augusto Fasolo Quevedo[1]

[1]Universidade Estadual de Campinas, Faculdade de Engenharia Elétrica e de Computação, Campinas SP, Brazil.
[2]Universidade Estadual de Campinas, Departamento de Neurologia, Campinas SP, Brazil.

Correspondence: Renata Michelini Guidi; Email: guidirm@gmail.com.

Background: Pain is the brain's interpretation of stimuli captured by nerve structures in the skin, viscera and muscles. Migraine is a type of headache with severe pain symptoms and has a major impact on daily occupational activities and quality of life. Measuring the pain perception threshold plays an important role in evaluating pain improvement throughout treatment. Electrical algometry quantifies the threshold of pain perception and pain tolerance through electrical stimulation of nociceptors present in the skin. **Objective:** To evaluate the pain perception threshold by electrical algometry in migraine and healthy individuals and to establish a protocol for the use of electrical algometry. **Methods:** Observational study was conducted with 14 female individuals, 7 with chronic migraine and 7 healthy. The anatomical points were assessed bilaterally as the temporal, frontal and masseter muscles and the C5/C6 zygapophyseal joint. Measurements were carried out using electrical algometer with two different protocols, the ramp mode and random mode. The difference between the algometry protocols on the sides of the body was evaluated for each anatomical point. Predominant pain side and the contralateral side for migraines, and the dominant and non-dominant side for healthy patients. **Results:** The migraine group (41.4±7 years) had 71% of participants with predominant pain on the right side and 29% with predominant pain on the left side. There was no statistical difference between algometry protocols in the anatomical points evaluated. The only exception was the temporal muscle on the predominant pain side with a significant difference between the algometry protocols, which suggests that there was accommodation of the nociceptor fibers in ramp mode (pain threshold = 7.5 ±1.2 mA), presenting lower pain threshold value (6.4 ±1.2 mA) in random mode. This may be related to greater local sensitivity, which corroborates the participants' reports that the headache is more intense and more present in the temporal region compared to other areas. However, for the healthy group (39±3.5 years), there was significant difference between the algometry protocols on both sides and in all anatomical points, suggesting a probable accommodation of peripheral nociceptor fibers. When comparing groups, the pain thresholds showed significant differences in all anatomical points. The migraine group had lower pain thresholds compared to the healthy group. **Conclusion:** Our study is the first to evaluate pain thresholds in migraine with electrical algometry, showing that pain thresholds are lower in migraine compared to healthy individuals. This may be related to the presence of dysfunctions in pain modulation mechanisms or greater local sensitivity due to the chronic disease. The most appropriate evaluation protocol for electrical algometry seems to be random mode, which is capable of reducing interference from the accommodation of nociceptor fibers in measurements.

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Assessment of trigeminal nerve integrity in young adults

Eduardo Rocha Arruda[1], Carlos Ernesto Garrido Salmon[2], Antonio Carlos dos Santos[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Filosofia, Ciências e Letras de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Eduardo Rocha Arruda; Email: duarruda@alumni.usp.br.

Background: Some chronic orofacial pain diseases have a well-defined etiology, such as classic trigeminal nerve (TN) neuralgia. However, in idiopathic trigeminal neuralgia, the individual presents the same symptoms as classic neuralgia, but not the same cause, which suggests a problem directly in the nerve. Diffusion Tensor Imaging (DTI) is the MRI technique that allows the assessment of white matter integrity using diffusion metrics such as fractional anisotropy (FA). Studies on the integrity of the trigeminal nerve in young, healthy adults contribute to defining a standard of normality, but are scarce in the current literature and have a small number of individuals. **Objective:** To evaluate the integrity of the NT of healthy young adults, using the DTI and verify the behavior of the AF values, as well as the bilateral asymmetry of this metric considering the effect of the individuals' age and sex. **Methods:** Structural and diffusion MRI data from 312 healthy individuals, 155 men and 157 women, aged between 22 and 40 years, were obtained from the Human Connectome Project, HCP database and separated into 8 groups considering age and sex. T1w images were used for bilateral marking of the regions of interest (ROIs): NT, root entry zone, middle portion of the cistern and Merckel's cave. The marking of ROIs, the generation of FA maps and the calculation of the average of this metric in the aforementioned regions were done using the MRTrix3 software. With the FA values, bilateral asymmetry was determined in all ROIs according to the expression: A = (FAd - FAe)/(FAd + FAe)*100. The results were analyzed by applying the ANOVA test with a significance threshold of $p<0.05$. **Results:** When comparing the FA values and their asymmetry in the different ROIs, there were no differences between any of the 8 groups when considering the age effect. FA values are higher bilaterally in women in the regions of the NT, root entry zone and in the middle portion of the right cistern (FA women (0.32±0.12); FA men (0.28±0.11); $F(1.898) = 19.78$; $p<0.001$). In the middle portion of the left cistern and Marckel's cave bilaterally, there was no significant difference between the sexes. **Conclusion:** Age does not influence FA values

or laterality asymmetry in any portion of the NT in young adults. However, sex should be considered a confounding factor when evaluating the integrity of the TN, the root entry zone and the middle portion of the right cistern.
Keywords: Trigeminal nerve, DTI, young adult.

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Assistive technology for communication accessibility in Machado Joseph disease

Bernardo Caetano Novaes[1], Lara Valentim Vale[1], Igor Dutra Braz[1], Claudia Yamada Utagawa[1]

[1]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.

Correspondence: Bernardo Caetano Novaes; Email: bernardo16.novaes@gmail.com.

Background: Machado Joseph disease (MJD), or spinocerebellar ataxia type 3 (SCA3), is an autosomal dominant neurodegenerative disorder, affecting 1 per 50,000-100,000 individuals. It is the most common hereditary ataxia. The manifestations include dysarthria, lack of motor control, and ophthalmoplegia, creating communication barriers that require assistive technologies to maintain patients´s autonomy, independence, quality of life. **Objective:** This study aims to understand the needs of a 53-year-old patient with MJD through reports from the patient, her family members, and caregiver, and to promote communication accessibility interventions using assistive technologies to optimize her quality of life. **Methods:** Approved by the Ethics Committee (CAAE 163692622.7.0000.5237), a clinical follow-up was performed for one year. Quality of life assessments and semi-structured interviews were conducted with the patient who had been living with the disease for eight years, as well as with family members and the caregiver regarding communication difficulties (verbal, visual/reading, writing, and gestural) and the strategies used to overcome them. Six months after the proposal of assistive technologies, new interviews were conducted, including their perceptions of using these technologies. **Results:** The analysis of the interviews led to the implementation of several assistive technologies, considering the patient's difficulties, disease stage, smartphone access as her preferred communication device. The implemented tools were screen zoom and character enlargement, replacement of typed texts with audio conversations, use of Google Assistant, and Voice Access. The enlargement of characters and screen zoom facilitated the reading of texts and images, mitigating diplopia. Audio communication, despite the patients' slowed speech, made social communication easier. Google Assistant, designed for speech-to-text conversion, had limited use due to slowed speech but it helped convert speech into written messages. Although Voice Access also converted speech to text, the patient preferred not to use it continuously due to its impact on smartphone usability. Other technologies such as SymboTalk, Head Mouse, and TalkBack were proposed but not evaluated due to the current stage of the patient. However, we believe they could be useful as the disease progresses. **Conclusion:** This study proposes interventions with communicational assistive technologies, recommending the use of audio in social networks, screen and character enlargement, and Google Assistant for voice typing in the early stages of the disease. Voice Access did not have good adherence from the patient. The interventions showed a significant positive impact, suggesting that these strategies may benefit other patients with MJD in similar disease stages.

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Association between age and time from symptom onset to admission to a stroke unit in Northeastern Brazil

Marivaldo Yslan Silveira Leal[1], Luisa Dias Moreno Aravena[1], João Pedro Fernandes Gonçalves[1], Gabriel Souza Suzart[1], Lucca Borges Ornelas[1], Vinicius Bessa Mendez[1], Pedro Antonio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Luísa Dias Moreno Aravena; Email: luisaaravena@ufba.br.

Background: Stroke is still socially considered as a disease that affects only the elderly. There is a lack of awareness about the frequency of stroke in the younger population and less perception of stroke-like symptoms as an emergency. Moreover, premature stroke may represent an evaluation challenge for healthcare professionals, resulting in misdiagnosis and diagnosis delay. Despite lower mortality, younger patients present impaired quality of life, social and economical burden as part of the labour force population. Therefore, early identification is needed to implement correct management and etiology investigation, in order to improve functional outcomes. **Objective:** To evaluate the association between age and ictus-admission time in a stroke unit (SU). **Methods:** This is a cross-sectional study that included patients with confirmed acute stroke in a SU in northeastern Brazil between July 2023 and April 2024. Confirmed acute stroke, age over 18 years and time interval between ictus and study inclusion below 72 hours were the inclusion criteria. Demographic and clinical data were analyzed. Age was dichotomized into elderly (≥60 years) and non-elderly (<60 years), based on the Brazilian Statute of the Elderly. We subtracted the time of admission to the SU from the time of symptom onset to find the ictus-admission time. The Mann-Whitney test was used to evaluate the association between age and time between ictus and admission. **Results:** Two hundred eight patients were enrolled. The majority of patients were female, 108 (51.9%), and the mean age was 65.7 years old (SD=±12.5). The median time between ictus and admission in SU was 7.9 hours (IQR=2.5-8.5). In the dichotomous analysis, <60 years of age were 28.4% of the patients and yours median ictus-admission time was 5 (IQR=3-15), while ≥ 60 years accounted for 71.6% and had a median ictus-admission of 4 (IQR=2.5-6) hours. The Mann-Whitney test showed that age has an effect on the time from ictus to admission (U=3,526; p=0.026). 40.2% of non-elderly patients and 64.4% of those elderly were admitted within the thrombolysis time. The median admission NIHSS for <60 years was 8 (IQR=4-14) and for ≥60 years was 7 (IQR=4-12), but there was no effect of age on patient admission NIHSS (U=3,886; p=0.620). **Conclusion:** Our study indicated that non-elderly patients had a lower ictus-admission gap and thus arrived faster at the healthcare service compared to those elderly. However more studies are needed to understand the factors that influence this time difference.

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Association between alpha-1-antitrypsin levels and intracranial aneurysms: a case-control study

João Paulo Mota Telles[1], Jefferson Rosi Junior[1], Julia Leal Bassan[1], Eberval Gadelha Figueiredo[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: João Paulo Mota Telles; Email: joao.telles@fm.usp.br.

Background: Comprehending the mechanisms involved in the formation of intracranial aneurysms can contribute in predicting the risk of its development and provide different types of treatment. **Objective:** This case-control study aimed to compare alpha-1-antitrypsin levels between patients with ruptured and unruptured intracranial aneurysms. **Methods:** We retrospectively analyzed medical records of patients that had collected alpha-1-antitrypsin levels from 2018-2022. A total of 233 patients were identified as having unruptured intracranial aneurysms and 114 were identified as having ruptured intracranial aneurysms. The control sample was formed by 55 patients that had intracranial arteriovenous malformation without aneurysms. **Results:** Alpha-1-antitrypsin mean levels in the unruptured group were 134.9 (±23.3), compared with 132.1 (±20.3) in the ruptured group, which was similar to 132 (±19), the alpha-1-antitrypsin mean level in the control group. Alpha-1-antitrypsin level was not associated with the presence (compared to controls, OR 1.07, 95% CI 0.93-1.25) or rupture (compared to unruptured, OR 0.94, 95% CI 0.84-1.04) of intracranial aneurysms. Alpha-1-antitrypsin levels were not associated with vasospasm (OR 0.94, 95% CI 0.93-1.25) or functional outcomes (mRS ≤ 2, OR 0.77, 95% CI 0.51-1.27). **Conclusion:** alpha-1-antitrypsin levels did not differ between patients with ruptured and unruptured aneurysms. Alpha-1-antitrypsin levels were also not associated with vasospasm or functional outcomes.

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Association between cannabis exposure and stroke: a systematic review and meta-analysis

Isabella Christina Amaral de Lara[1], Ísis Larissa de Brito Dichtl[2], Larissa Emi Tanimoto[3], Hugo de Souza Barreto[4], Artur Menegaz de Almeida[4], Ítalo Barros Andrade[5], Lilianne Rodrigues Fernandes[6]

[1]Universidade Cesumar, Maringá PR, Brazil.
[2]Universidade Estadual do Piauí, Teresina PI, Brazil.
[3]University of Buenos Aires, Buenos Aires, Argentina.
[4]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[5]Faculdade Santo Agostinho de Vitória da Conquista, Vitória da Conquista BA, Brazil.
[6]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.

Correspondence: Isabella Christina Amaral de Lara; Email: isabellaadelara@gmail.com.

Background: As the most widely used drug globally, cannabis has been the subject of numerous studies investigating its health effects. Recent studies have explored the potential link between cannabis use and stroke events, revealing conflicting findings. **Objective:** To determine the real association between cannabis and stroke episodes, contributing to the existing evidence on this topic. **Methods:** We searched PubMed, Scopus, and Web of Science databases for studies investigating the association between cannabis exposure and stroke occurrences. Outcomes of interest included overall stroke incidence and specifically ischemic stroke rates. We computed odds ratios (ORs) for binary endpoints, with 95% confidence intervals (CIs). Heterogeneity was assessed using I^2 statistics. For this meta-analysis, statistical analysis was performed using Review Manager 5.4.5 software. **Results:** A total of 21 studies and 146,873,842 patients were included. Most patients were women (71.60%). The meta-analysis revealed that the ischemic stroke rate was homogeneous between cannabis users and non-users (OR 1.22; 95% CI 1.10-1.35; p = 0.0001; I^2=41%). In contrast, the overall stroke rate was significantly higher in the non-users group (OR 1.85; 95% CI 1.52-2.27; p<0.00001; I^2= 62%). The results showed a significant difference in favor of the control group in terms of increased stroke risk (0.446370% vs 0.273191%; OR 0.72; 95% CI 0.70-0.74; p<0.00001; I^2=99%) or ischemic strokes (0.290268% vs 0.244143%; OR 1.08; 95% CI 1.04-1.12; p<0.0001; I^2=95%) compared with the non-cannabis exposed group. **Conclusion:** While previous studies have produced inconsistent results, this meta-analysis indicates that cannabis use is not significantly associated with higher rates of stroke or ischemic stroke compared to the control group. These findings highlight the importance of continued research to further understand how cannabis use can affect health outcomes.

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Association between diffuse glioma and ischemic stroke in a pediatric patient: a case report

Angelo Ponte de Freitas Campos[1], Rodrigo de Paiva Bezerra[1], Marcel Ken Uehara[1], Beatriz Casella[1], Alessandra Milani Prandini de Azambuja[1], Vicente Odone Filho[1], Gisele Sampaio Silva[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Angelo Ponte de Freitas Campos; Email: campos_angelo@hotmail.com.

Case presentation: We describe this case of a female patient, 2 years old, previously healthy. At three months she was diagnosed with a diffuse BRAF-KIAA1549 glioma, due to an episode of bilateral nystagmus and drowsiness. MRI revealed a contrast-enhancing expansive lesion in the hypothalamic-chiasmatic region associated with multiple leptomeningeal implants in the cervicodorsal transition without signs of myelopathy. After the introduction of chemotherapy, she was readmitted at 2 years of age due to acute worsening of strabismus with reports of abnormal hand and head movements. A new Cranial MRI showed an extensive area of diffusion restriction and signal alteration in T2/FLAIR in the left frontoinsular cortical area, consistent with a recent ischemic event in the territory of the left superior M2 division of the middle cerebral artery. Increased T2/FLAIR signal in the left cerebellar hemisphere, suggestive of subacute recent ischemic foci. Cerebral angiography revealed moderate stenosis at the origin of the superior division of the M2 segment of the left middle cerebral artery. Further investigation of a cerebrovascular event in a young patient was carried out with TTE showing no microbubble passage and TCD showing no active embolization. Regarding thrombophilia screening, there were no abnormalities justifying the event. **Discussion:** Few studies correlating gliomas and cerebrovascular events are currently available in the literature. Observational studies that have identified the tumor's topography with the highest risk of ischemic events. Cancer represents one of the major causes of ischemic stroke classified as ESUS (Embolic Stroke of Undetermined Source) with an estimated subgroup incidence of 10% of cases. Several mechanisms described in the literature explain the occurrence of ESUS in these patients, among which are paradoxical embolism, non-bacterial thrombotic endocarditis, aortic atheroma, and more rarely, tumor embolism. Regarding treatment, the current literature still diverges on the best therapy to adopt. There is a lack of data on the best treatment regimen for this subgroup of patients with ischemic stroke. In summary, there has been no superiority among anticoagulants as well as an increased risk of bleeding if antithrombotics are chosen. **Final comments:** Oncological patients with acute ischemic stroke should be regarded as a heterogeneous subgroup, thus requiring extensive investigation of the underlying cause of the cerebrovascular event. Patients with the investigation ends in an unknown cause or ESUS, anticoagulation or even antithrombotic therapy still represents a frontier of knowledge available in the literature. In these cases, the decision should be individualized, weighing the risks and benefits of each proposed therapy. Serum D-dimer levels appear to be a useful tool in aiding the early initiation of secondary prophylaxis as well as in monitoring these patients, given its association with increased risk of ischemic stroke.

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Association between early ictus-admission time in a stroke unit and functional outcome in hyperacute stroke patients not eligible for reperfusion therapies

Caio Fontes do Nascimento[1], Marcos Paulo Rodrigues de Oliveira[1], Alex de Cerqueira Silveira Figueiredo[1], Nara Samara Silva dos Santos[1], Luana Araújo Passos[1], Pedro Antonio Pereira Jesus[2], Bruno Bacellar Pedreira[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil. [2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Caio Fontes do Nascimento; Email: caiofontesdn@hotmail.com.

Background: Stroke is currently one of the leading causes of morbimortality in Brazil and worldwide. Unfortunately, the availability of reperfusion therapies (RT) in Brazil's public health system is limited, and the time window for the effective use of these therapies is relatively short. The development of stroke units (SU) has provided a new perspective for patients admitted to hospitals who are not eligible for RT, showing evidence of better prognoses compared to conventional hospital care settings. **Objective:** To analyze the correlation between the interval from ictus to admission in a stroke unit and functional outcome among patients who did not meet the criteria for RT. **Methods:** This study was a prospective cohort that followed patients who suffered a stroke with a time from ictus to admission to a SU of less than 72 hours between July 2023 and May 2024. All described information was collected from medical records. The functional outcome variable at hospital discharge, at 30 days, and at 90 days was measured using the modified Rankin Scale (mRS). The Mann-Whitney test was used to assess the difference in average time from ictus to admission between the groups, and the Spearman test was utilized to measure the correlation between the variables. **Results:** A total of 114 patients met the inclusion criteria. The mean age was 63.23±12.97, with 21% of all patients showing delirium during hospitalization and a median ictus-admission time of 360 minutes (range: 30 - 3270). When comparing patients with good outcomes (mRS<3) in hospital discharge, 30 days and 90 days and those with bad ones (mRS≥ 3), no difference was found in ictus-admission time ($p<0.05$). Furthermore, there was no statistical significance in correlations between outcomes in hospital discharge (rho = -0.188, p=0.054), 30 days (rho = -0.119, p=0.223) or 90 days (rho = -0.169, p=0.133), and the ictus-admission time, even when analyzing patients with mRS below (rho = -0.153, p=0.216) or above 1 (rho = -0.453, p=0.188), younger (rho = -0.351, p=0.168) or older (rho = -0.050, p = 0.695) than 55 years, and with (rho = -0.004, p=0.989) or without (rho = -0.156, p=0.215) delirium. **Conclusion:** The sample studied exhibited epidemiological data and delirium occurrence frequencies consistent with those reported in current literature. However, no statistically significant correlation was found between the time from ictus to admission in SU and functional outcomes. We attribute this, in part, to the exclusion criterion of a time from ictus to admission greater than 72 hours, which significantly restricted the sample size. Well-established evidence indicates that admission to SU can enhance patients prognosis compared to other hospitalization settings. The results of the study reinforce the importance of RT in improving patient outcomes, however further studies are necessary to determine if early admission can also contribute to better results.

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Association between microembolic signals on transcranial doppler and brain atrophy in patients with Chagas disease

Ayla Morais Vitório Guerra[1], Cárita Victória Carvalho de Santana[1], Renan Carvalho Castello Branco[2], Pedro Fernandes Abbade[1], Isabella Reis Vieira[1], Beatriz Malaquias Neves[1], Israela Souza Brito Santos[1], Bruno Felipe Santos de Oliveira[1], Thiago Cerqueira Silva[1], Jamary Oliveira Filho[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil. [2]Universidade Federal do Recôncavo da Bahia, Cruz das Almas BA, Brazil.

Correspondence: Ayla Morais Vitório Guerra; Email: aylaguerra1@hotmail.com.

Background: Chagas disease (CD) is a neglected chronic disease associated with central nervous system impairment, such as stroke, cognitive dysfunction, microembolic signals (MES) and brain atrophy. However, the relationship between MES and brain atrophy in patients with CD has not been studied. **Objective:** To investigate the association between MES and central nervous system volume in patients with heart failure (HF) of chagasic and non-CD origin. **Methods:** Cross-sectional study conducted between 2002 and 2024 with participants from a cardiology outpatient clinic in Salvador-BA, aged > 18 years, diagnosed with HF by Framingham criteria; CD was confirmed by ELISA or immunofluorescence test, in duplicate. We excluded those with a history of previous stroke, dialysis, chronic parenchymal liver disease, history of malignancy, use of anticoagulants, or metallic prostheses. Eligible participants underwent bilateral transcranial Doppler (TCD) examination of the middle, anterior (ACA), and posterior (ACP) cerebral arteries for one hour to detect signs of MES. Additionally, they underwent cranial magnetic resonance imaging (MRI) to evaluate volume of several brain structures after automated volume assessment using FreeSurfer version 7.4.1 software. We performed univariate tests to assess associations between MES and brain volumes. **Results:** The sample consisted of 336 participants with HF, 45% male, 51% with CD. The mean age among CD participants was 55.5±10.6 years, and the mean ejection fraction was 50.4±17.7%. The presence of MES in TCD was observed in 19 (11%) of CD participants and 11 (7%) non-CD participants. Univariate tests in participants with CD showed that the MES group had a lower mean volume of the ventral diencephalon (3.3±0.7 ml non-MES vs 3.1±0.8 ml MES; p = 0.047). In non-CD participants, the MES group had a lower mean volume of the isthmus of the cingulate gyrus (2.6±0.4 ml non-MES vs 2.4±0.3 ml MES; p = 0.008) and a lower mean volume of the paracentral cortex (3.7±0.5 ml non-MES vs 3.2±0.6 ml MES). No significant differences in the presence of territorial and lacunar infarcts were observed between the groups with and without MES. **Conclusion:** Presence of MES may influence cerebral volume in several anterior circulation structures, highlighting the need for further investigation into its role in the progression of cerebral atrophy in patients with HF, both CD and non-CD patients.

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Association between migraine and autism spectrum disorder: a case series of 10 patients

Eduardo Souza Cardoso[1,2,3], Gabriel Eduardo Pereira Cardoso[4], Adileia Santos da Silva[3], Carla Soares Souza[3], Maria da Gloria Canto de Sousa[2], Ana Patricia Pascoal Queiroz de Araujo[2], Grace Patricia Santana Pereira Cardoso[5], Jéssica de Andrade Ribeiro Lima[2], Maiêva Pereira Ribeiro[2]

[1]Universidade Estadual de Feira de Santana, Feira de Santana BA, Brazil.
[2]Universidade do Estado da Bahia, Salvador BA, Brazil.
[3]Clinica IBIS, Salvador BA, Brazil.
[4]Faculdade Zarns, Salvador BA, Brazil.
[5]Hospital Portugues, Salvador BA, Brazil.

Correspondence: Gabriel Eduardo Pereira Cardoso; Email: Gabrielwer204@gmail.com.

Background: Migraine is a common neurological disorder marked by severe headaches, nausea, and sensitivity to light and sound. Autism Spectrum Disorder (ASD) Level 1 support is characterized by difficulties in social interaction, communication, and restricted behaviors,

often accompanied by sensory hypersensitivity and emotional dysregulation. This case series explores the overlap between migraine and ASD Level 1 support in 10 patients, emphasizing sensory hypersensitivity and emotional dysregulation. **Objective:** This study aims to explore the association between migraine and subsequent diagnosis of ASD Level 1 support, with a focus on sensory hypersensitivity and emotional dysregulation observed during clinical follow-ups. **Methods:** We included 10 patients with a confirmed diagnosis of migraine. These patients were followed up regularly and assessed for symptoms indicative of ASD Level 1 support. The focus was on identifying signs of sensory hypersensitivity and emotional dysregulation, which are common in both migraine and ASD. **Results:** The study included 10 patients, with a mean age of 32 years, comprising 7 females and 3 males. Migraine with aura was present in 60% of cases, with an average duration of migraine diagnosis of 5 years. The average time to ASD diagnosis was 2 years after the migraine diagnosis. All patients exhibited heightened sensitivity to light and sound during migraine episodes, and these sensory sensitivities persisted as characteristics of ASD. Additionally, 100% of the patients showed aversion to certain textures, sounds, and lights. Emotional dysregulation, including difficulties in managing emotions, frequent mood swings, and episodes of anxiety and irritability, was reported by 90% of the patients. This emotional instability significantly affected their social interactions and daily functioning. The coexistence of migraine and ASD Level 1 support, with pronounced sensory hypersensitivity and emotional dysregulation, suggests a potential interrelation between these conditions. Understanding the overlap in symptoms can facilitate early identification and comprehensive management, improving patient outcomes. The findings underscore the importance of recognizing sensory and emotional symptoms in patients with migraine, which may indicate an underlying ASD diagnosis. **Conclusion:** Identifying ASD in patients with migraine, particularly considering sensory hypersensitivity and emotional dysregulation, can significantly influence treatment strategies and improve quality of life. Further research is essential to elucidate the mechanisms linking these conditions and develop integrated therapeutic approaches. Early intervention and multidisciplinary care are crucial for managing these overlapping conditions effectively.

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Association between risk of obstructive sleep apnea and stroke recurrence: a cohort study

Aurea Maria Lago Novais[1], Juliana Cerqueira Gomes[1], Marina Soledad Benitez Bitencourt[1], Thaise Braga de Oliveira[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Aurea Maria Lago Novais; Email: aureanovais20.1@bahiana.edu.br.

Background: Obstructive sleep apnea (OSA) is a disease of upper airways, diagnosed with polisomnography, that is well associated with stroke through diverse mechanisms. Despite that, it still lacks studies about its role in prognosis of stroke, especially in stroke recurrence. **Objective:** This study aims to elucidate the possible association between stroke recurrence and risk of OSA. **Methods:** We performed a cohort. Patients were evaluated of risk of OSA with STOP-Bang and SOS scores and remotely evaluated with Rankin, Barthel, questioned about stroke recurrence and another hospitalization during 3.6 and 12 months. We performed univariate analysis (T test, Mann-Whitney or chi-square) and a multivariate analysis with logistic regression (considering P < 0.05). **Results:** We obtained 430 patients, 69.1% classified as high risk with STOP-Bang and 50,2% with SOS. 271 finished 3 months of follow-up, 180 finished 6 months and 112 finished 12 months. There was significant difference between groups in hypertension (p<0.001), sex (p<0.001), alcoholism (p=0.01), diabetes (p=0.002), obesity (p=0.01) and previous regular use of medication (p<0.001). In high risk group, Rankin 3 months median 2 (1-4; p=0.03), Rankin 6 months median 1 (1-4), Rankin 12 months median 1(1-4). In low risk group, Rankin 3 months median 2 (1-4), Rankin 6 months median 1 (1-3), Rankin 12 months median 1 (1-3). 15 patients recurred with stroke (71% of high risk; OR 1.2; CI 95% 1-1.4; rs=-0.2; p=0.05). 29 evolved to death (14 of high risk; OR=1.03; CI95% 0.55-1.9; p=0.92). This is the first study to describe association between OSA and stroke recurrence, with a relevant higher rate of recurrence in patients with high risk, denoting the role of OSA in occurrence and prognosis of cerebrovascular disease. Also, patients with high risk tend to have a worst recovery, especially in thirst 3 months,, in comparison to high risk. Higher rates of regular use of medications in high risk appears to reveal the presence of multiple comorbidities, with coexisting mechanisms of impairment. **Conclusion:** In face of this, it is important now to investigate the importance of, not only use risk scores in cerebrovascular disease, but treat obstructive sleep apnea as part of secondary prophylaxis.

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Association between stroke etiology and functional outcomes

João Pedro Fernandes Gonçalves[1], Marivaldo Yslan Silveira Leal[1], Luísa Dias Moreno Aravena[1], Gabriel Souza Suzart[1], Lucca Borges Ornelas[1], Vinicius Bessa Mendez[1], Pedro Antonio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador, BA, Brazil.

Correspondence: Luísa Dias Moreno Aravena; Email: luisaaravena@ufba.br.

Background: The TOAST classification is a widely used and well-described diagnostic approach regarding ischemic stroke etiology. However, the association of different stroke etiologies and functional outcomes is not well established. **Objective:** To assess the effect of different etiologies on functional outcomes of ischemic strokes. **Methods:** We conducted a prospective cohort of patients with a confirmed acute ischemic stroke in a single stroke unit in Brazil. The inclusion criteria were: confirmed acute ischemic stroke, the time interval between ictus and study inclusion below 72 hours and age over 18 years. Exclusion criteria included: RASS scale -4 or -5 at admission, diagnosis of severe bilateral vision or hearing loss and severe sensory aphasia. Stroke etiologies were divided following the Trial of ORG 10172 in Acute Stroke Treatment (TOAST) Classification. Functional outcomes were assessed by the modified Rankin scale (mRs) 90 days post-stroke by telephone interview. The mRs was dichotomized into good functional outcomes (mRs = 0-2) and poor functional outcomes (mRs = 3-6). The chi-square test evaluated the association between stroke etiology and functional outcomes. Anova, Kruskal-Wallis and chi-square tests assessed differences between possible confounding variables. **Results:** In total, 514 patients were included in the final analysis with a mean age of 63.1 (±13.3) years and 54.5% were men. The etiologies investigated and the frequency of poor functional outcomes were: large-artery atherosclerosis (48.6%), cardioembolism (38.9%), small-vessel occlusion (36.7%), incomplete investigation (33.8%), multiple causes (72%) and other defined causes (55.6%). Incomplete investigation was associated with good functional outcomes at 90 days (p<0.05) and multiple causes were related to poor functional outcomes (p<0.05). Relevant differences in age were found: incomplete investigation (61.1) showed a lower mean age when compared to large-artery atherosclerosis (67.6) and multiple causes (70.1), also multiple causes had a higher mean age compared to other causes (50.5). Incomplete investigation had a lower NIHSS median (7.0) compared to cardioembolism (9.0), while multiple causes had a higher NIHSS median (12.0) than small-vessel occlusion (6.0). No differences were found in the thrombolysis frequencies. **Conclusion:** Patients with an incomplete investigation may have better functional outcomes at 90 days, while patients with multiple possible causes may have worse functionality post-stroke. Unfortunately, the associations found may be due to the differences in confounding variables, such as age and NIHSS at admission. Further studies with larger samples and multivariate analysis are needed to help determine the role of stroke etiology in functional outcomes.

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Association of neuromyelitis optica and rapidly progressive IgA glomerulonephritis: a case report

João Henrique Reis Duque Estrada[1], Caroline Vianna Costa Pinto[1], Luiza Guimarães Póvoa[1], Victor Waldhelm Cozer[1], Felipe Schmidt[1], Diogo Terrana[1], Sofia Mermelstein[1]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: João Henrique Reis Duque Estrada; Email: joaohestrada2@gmail.com.

Case presentation: Our case involves a 35-year-old patient admitted for the investigation of sequential bilateral visual acuity loss and ascending paresthesia associated with gait disturbance. The patient reported experiencing a probable area postrema syndrome in March 2023, followed by partial cervical myelitis a few months later, and severe optic neuritis affecting both eyes by September 2023, when admitted to the Neurology ward. The preliminary diagnostic workup for infectious and autoimmune causes was negative, except for a positive anti-AQP4 antibodies detected by cell-based assay. She was then, diagnosed with Neuromyelitis Optica (NMO), with a relapsing course with minimal functional improvement between episodes. Additionally, upon admission, she presented with elevated creatinine levels (3.5 mg/dL or 17 mL/min/1.73 m2), urinary protein/creatinine ratio of 0.63 mg/dL and dysmorphic hematuria on sediment microscopy and denied the use of nephrotoxic medications. Previous tests had shown creatinine levels of 0.9 mg/dL. This led to a concurrent diagnosis of rapidly progressive glomerulonephritis due to IgA nephropathy, confirmed by renal biopsy showing crescents and segmental IgA deposition in the glomerular mesangium. Prompt treatment with pulse therapy and plasma exchange resulted in rapid improvement of neurological symptoms and renal dysfunction. The patient's management included ongoing oral corticosteroids and Rituximab infusion as broad immunosuppressive therapy, targeting both NMO and IgA nephropathy. **Discussion:** To our knowledge, this is the first report describing the association between NMO and rapidly progressive glomerulonephritis due to IgA deposits. The simultaneous involvement of the nervous system and kidneys suggests autoimmune hyperactivity of the humoral immune system, although the mechanisms are not fully understood. One potential link between brain and kidney dysfunction could involve aquaporins. There are at least 10 different isoforms of aquaporins found throughout the human body. AQP4 is located in astrocytic foot processes, in the periventricular regions, the medullary floor of the fourth ventricle, and the area postrema. AQP4 antibodies, known as a diagnostic marker for NMO, can cause significant damage to astrocytes and are considered pathogenic. Interestingly, AQP4 is also present in the medullary collecting tubules of the kidney. Therefore, it's reasonable to hypothesize that antibodies against AQP4 in the serum of an NMO patient could potentially attack the kidney and lead to renal disorders. However, it's important to note that our patient has IgA mediated nephropathy, and the anti-AQ4 is a IgG antibody. **Final comments:** It is of utmost importance to discuss the autoimmune comorbidities of patients with NMO, as they are increasingly being described and contribute to unraveling the pathophysiology of autoimmune mechanisms related to the disease, thereby guiding more effective and eficient treatments.

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Ataxia and Parkinsonism related with CAG expansion size in a cohort of Huntington's disease in the Brazilian Amazon region

Marcus Vinicius Della Coletta[1], Emanuelle Campos Amaral[2], Maria Esther Lima Rios Evangelista[3], Fernanda Almeida Carvalho[2], Danilo Lemes Reis[2], Manuela Brandao Ferreira Teles[2], David Franca Ferreira Cruz[2], Diana Vieira Brito[2], Helio Afonso Ghizoni Teive[1], Carlos Henrique Ferreira Camargo[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[3]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Emanuelle Campos Amaral; Email: eca.med22@uea.edu.br.

Background: Huntingtons disease is caused by the expansion of CAG repeats in the HTT gene and exhibits wide variability in age of onset, symptomatology, and disease progression. There is no published data about genetic and clinical aspects of HD in the Brazilian Amazon population. **Objective:** We conducted genetic and clinical assessments in 22 families, with genetic diagnosis in 51 patients with HD in the state of Amazon, Brazil. The main objectives are evaluating demographics, genetic characteristics, and clinical presentations of HD in the Amazonian population. **Methods:** Genetic testing was performed on 51 patients exhibiting HD symptoms. Clinical assessments, including the Unified Huntingtons Disease Rating Scale (UHDRS), were conducted to evaluate motor and cognitive functions. Data on age of onset, illness duration, CAG repeat sizes, and parental transmission were collected and analyzed. **Results:** The study analyzed a cohort of patients with a mean age of 44.9 years, ranging from 11 to 71 years. The average age of onset of the first symptoms was 33.1 years, with a range from 6 to 62 years. The average illness duration was 12.8 years. The average size of CAG expansions was 47.4, with a range between 39 and 84. Paternal transmission was confirmed in 27 patients (52.9%), while maternal transmission was confirmed in 20 patients (39.2%). Patients exhibited a variety of clinical manifestations, including chorea in 37 patients (72.5%), parkinsonism in 36 patients (70.5%), ataxia in 31 patients (60.7%), and dystonia in 30 patients (58.8%). Tics were observed in 8 patients (15.6%).The average score on the Unified Huntington's Disease Rating Scale (UHDRS) was 59.8, assessed in 38 patients, with a range from 7 to 120. These data highlight the diversity of clinical manifestations and genetic variability among patients, underscoring the complexity of diagnosing and managing the disease. **Conclusion:** The study results indicate that HD in the Amazon region manifests predominantly with chorea, parkinsonism, and ataxia, aligning with previously reported clinical profiles in other populations. The observed high prevalence of chorea suggests a significant impact on motor functions, leading to a decreased quality of life for affected individuals. Additionally, the average age of onset in this population is consistent with global trends, although the relatively large CAG repeat sizes may contribute to the severity of symptoms. The more severe forms of ataxia and parkinsonism were significantly related to the size of CAG expansions.

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ATP1A3 related disorders: a case series

Victor Rebelo Procaci[1], Raphael Pinheiro Camurugy da Hora[1], Pedro Lucas Grangeiro de Sá Barreto de Lima[2], João Victor Cabral Correia Ferrer[1], Débora Palma Maia[3], Francisco Eduardo Costa Cardoso[3], Paulo Ribeiro Nóbrega[2], Thiago Yoshinaga Tonholo Silva[1], Orlando Graziani Povoas Barsottini[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[3]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Victor Rebelo Procaci; Email: vrebelop@gmail.com.

Background: ATP1A3 related disorders have a complex clinical presentation and can manifest from childhood to adulthood, presenting as differential diagnosis to several diseases with ataxia, hemiparesis, parkinsonism, dystonia, fever, encephalopathy, epileptic seizures, psychosis, among other features. The description of a clinical spectrum in a cohort of patients is essential for better disease characterization and development of new therapeutic methods. **Objective:** To present a case series of patients diagnosed with mutations in the ATP1A3 gene, through the analysis of medical records from the neurology outpatient clinic of the Universidade Federal de São Paulo and multiple Brazilian universities. **Methods**: This is a retrospective case series that we analyzed medical records from the neurology outpatient clinic of the Universidade Federal de São Paulo, Universidade Federal do Ceará and Universidade Federal de Minas Gerais and included patients with an established diagnosis of ATP1A3 mutation, independent of the phenotype presented. Clinical and genetic data was acquired through medical records analysis. **Results:** We identified 6 cases with a known mutation in ATP1A3, four male and two female. Age of onset varied from less than a year to 17-year-old. We found three main phenotypes: rapid-onset dystonia parkinsonism (2), CAPOS syndrome (2) and alternating hemiplegia of childhood (2). Besides the main phenotype, we identified other neurological features in our cases. Dystonia was present in one patient with AHC and one with CAPOS syndrome. Cognitive impairment was present in four patients. Epilepsy was present only in patients with AHC. AHC patients had positive response to flunarizine therapy. Spasticity, dysarthria, bradykinesia, REM sleep behavior disorder, neuropathy, developmental delay, autism, hypotonia were some of the neurological features that we also identified in our cohort. **Conclusion:** Our cohort highlights the phenotypic heterogeneity of ATP1A3 related disorders, emphasizing the need to consider a combination of neurological symptoms instead of classifying phenotypes in classic separate syndromes. More research is needed to clarify the spectrum of this disorder.

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Atrophy of the frontal lobe after white matter ischemia due to occlusion of the cervical portion of the internal carotid artery

Karina Carvalho Higa[1], Carlos Cesar Conrado Caggiano[1], Leandra Schneider[1], Pedro Arthur Possan[1], Mateus Pinto Marchetti[1], Isabela Louise Weber[1], Lucas Shigueo Hiromoto[1]

[1]Hospital Nossa Senhora das Graças, Curitiba PR, Brazil.

Correspondence: Karina Carvalho Higa; Email: karinacarvalho.fdc@gmail.com.

Case presentation: Patient K.L.L.F, a 37-year-old female, previously healthy presented with tonic-clonic movements followed by a decreased level of consciousness. Brought to the emergency department more than 12 hours after the ictus. On admission, neurological examination revealed a GCS score of 8 (Eye Opening 2; Verbal Response 2; Motor Response 4) and a NIHSS score of 31. MRI and cranial MR angiography showed hyperintense signal on FLAIR with diffusion restriction in the head of the caudate nucleus, lentiform nucleus, anterior limb of the internal capsule, and mesial portions of the left temporal lobe, consistent with an area of acute ischemia; the rest of the brain parenchyma had normal signal intensity of white and gray matter; and thrombosis of the entire length of the left internal carotid artery in its cervical and intracranial segments. The patient underwent mechanical thrombectomy with recanalization of the left carotid artery and left decompressive craniectomy within the first 24 hours of hospitalization. Patient showed good postoperative progress, with a slight left-sided facial droop, but no other abnormalities. In subsequent outpatient consultations, patient began to experience mood disturbances. Initially, with anxiety symptoms, progressing to major depressive disorder and semantic memory impairment on neuropsychological evaluation. Patient continued to have significant mood impairment, with poor response to instituted measures, as well as cognitive impairment. An MRI, two years later, showed changes compatible with atrophy of the left temporal and frontal lobes - a retractable lesion, hyperintense on T2/FLAIR, affecting the left lentiform nucleus and caudate nucleus, causing focal dilation of the left lateral ventricle, and prominence of the cerebral sulci in the left temporal and frontal lobes. **Discussion:** The loss of white matter volume after ischemic events reflects the sensory input from the brainstem as well as the cortical projections of the affected major nuclei. Changes in cortical geometry do not seem to reflect gray matter atrophy but rather the remodeling of the cortical surface due to underlying white matter atrophy. Over time, the reduction in white matter volume appears to result in cortical deformation, but not in gray matter volume loss. Acute unilateral lesions induce central compensation processes by reorganizing sensorimotor networks within the ipsilateral hemisphere and the contralateral hemisphere. The lack of changes in gray matter may reflect the heterogeneous interindividual projection pattern of the affected regions and their connections with various areas of the cerebral cortex. The clinical presentation depends on the affected area and the type of brain tissue involvement. **Final comments:** Strokes lead to a reduction in white matter volume in both the ipsilesional and contralesional hemispheres over time. This reduction in white matter volume is a consequence of cortical remodeling following a stroke.

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Attention deficit hyperactivity disorder and autism spectrum disorder in adults

Tatiana Pizzolotto Bruch[1], Camila Pezente Dal Toe[1], Amanda Cardoso da Silva[1], Suélen Galuppo[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Tatiana Pizzolotto Bruch; Email: tatibruch@yahoo.com.br.

To investigate the presence of symptoms of Attention Deficit Hyperactivity Disorder (ADHD) and Autism Spectrum Disorder (ASD) in university students using validated and translated psychological assessment tools in Brazil. A cross-sectional study was conducted, evaluating 433 students through online questionnaires: the Adult Self-Report Scale (ASRS-18) for ADHD and the Autism Spectrum Quotient - 10 items (AQ-10) for ASD. Analysis of 433 questionnaires identified ADHD symptoms in 62.4% and ASD symptoms in 15.7% of the students. Common ASD symptoms included difficulties in attention to details and communication, while ADHD symptoms primarily involved inattention. Significant associations were also found between ADHD and ASD symptoms and treatment for depression and/or anxiety. Therefore, it was concluded that symptoms of ADHD and ASD significantly impact personal relationships and academic profiles of students, as these psychological disorders are strongly linked to human performance in both social and professional contexts.

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Atypical case of HaNDL syndrome: an unusual presentation in a man

Nereu Alves Lacerda[1], Luis Felippe Barros[1], Mayhara Rosany da Silva Santiago[1], Ana Caroline Paiva Simeão[1], Raissa Josefa Pereira de Moura[1], Heloisy Maria Nunes Galvão[1], Felipe Franco da Graça[2], Natália Nunes Tenório[1], Heron Alves Vale[1], Ana Beatriz Roberto Lima Borba[1]

[1]Hospital Pelópidas Silveira, Recife PE, Brazil. [2]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Nereu Alves Lacerda; Email: nereulacerda@gmail.com.

Case presentation: A 23 year-old man, without any comorbidities aside from obesity and history of pain, was hospitalized due to a mild headache in the occipital region, which was not associated to photophobia or phonophobia, did not have a progressive characteristic and did not have improving or worsening factors. Afterwards the pain evolved with a change of pattern, now being a severe holocranial headache associated with an ictal difficult of speech and right hemiplegia. Neurological clinical exam showed motor aphasia, hemiparesis on the right (strength grade 2), ipsilateral hypoesthesia and bilateral papilledema. Notably, soon after admission the patient had complete improvement of the symptoms, but during hospitalization he persisted with sporadic episodes of headache with the mentioned characteristics. Laboratory exams and head CT without contrast had unremarkable results. However, on CSF analysis it was noted increased intracranial pressure (ICP = 52 cmH2o) and a lymphocytic (96%) pleocytosis, but no alterations regarding protein or glucose levels. Serologies with a viral panel were performed for infectious investigation, with all results being negative. We proceeded the investigation with angiotomography, magnetic resonance imaging and arteriography of cervical and intracranial vessels that were within the patterns of normality. Thus the diagnosis of headache with neurological deficits and cerebrospinal fluid lymphocytosis (HaNDL syndrome) associated with idiopathic intracranial hypertension was established. We initiated treatment with acetazolamide, after which a significant clinical improvement along with decrease in opening intracranial pressure on CSF was observed. Henceforth, the patient was discharged with orientation of return for outpatient follow-up. **Discussion:** HaNDL syndrome is a rare, benign and self-limited condition, characterized by headache associated with aphasia and other sensitive or motor deficits, along with CSF lymphocytosis. As in the presented case, opening intracranial pressure of the CSF might be increased, and the investigation of viral etiologies have negative results. In general, the syndrome has a great recovery rate even without long-term treatment, considering its self-limited nature. However, it is relevant to manage acute symptoms and give orientations about possible recurrences. It is important to highlight that the syndrome might initiate with a thunderclap headache, which makes it relevant to consider HaNDL syndrome in patients with this pattern of headache, even though it is an exclusion diagnosis. **Final comments:** HaNDL syndrome is a rare and possibly sub diagnosed condition that might be associated with comorbidities like idiopathic intracranial hypertension and have a thunderclap headache pattern, which makes it difficult to diagnose. Therefore, it is important to be aware of this diagnosis in cases of headache with warning signs.

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Atypical cryptococcosis with restricted diffusion mimicking radiological features of ischemic stroke: a case series

Alex Araújo Fernandes[1], Matheus Alves da Silva[1], Matheus Luís Braga Munareto[1], Eduardo Mesquita de Souza[1], Rafael Bragança Rodrigues Matias[1], Ana Luisa Faria Donadon[1], Amanda Miranda de Mendonça[1], Victor Hugo Rocha Marussi[1], Hennan Salzedas Teixeira[1], Alex Machado Baeta[1]

[1]Beneficência Portuguesa de São Paulo, São Paulo SP, Brazil.

Correspondence: Ana Luisa Vieira de Faria; Email: alfaria96@gmail.com.

Case presentation: Case 01 - A 70 year old man with Gilbert Syndrome presented with subacute fever and headache. His neurological examination showed signals of bilateral parkinsonism and appendicular ataxia. The MRI showed multiple foci of restricted diffusion in cerebellum, basal ganglia and cortex associated with leptomeningeal enhancement. The vessel wall imaging sequence showed concentric parietal thickening of middle cerebral arteries, compatible with vasculitis. His lumbar puncture showed normal opening pressure, 15 cells, 31 glucose, 281 proteins, 47 lactate and had a positive testing for Cryptococcus neoformans. He received amphotericin B liposomal and fluconazole. His neurological symptoms completely improved, and he was discharged with Fluconazole treatment. Case 02 - A 66 year old man presented with a one week headache and confusion and new sudden dysarthria onset. He had Budd-Chiari syndrome and cirrhosis. The neurological examination showed mild scandid dysarthria. His initial CT was normal, but MRI showed bilateral cerebellar restricted diffusion. Besides the images of ischemic stroke, his symptoms keep worsening in the hospital, incompatible with a vascular diagnosis. A lumbar puncture was performed, showing a normal opening pressure, 1152 cells with neutrophile predominance, 228 protein, 68 glucose and a positive testing for Cryptococcus neoformans. He received amphotericin B liposomal and fluconazole, with neurological improvement, but evolved with gastrointestinal bleeding and died of hypotensive shock. **Discussion:** Neurocryoptococcosis is an infection of the central nervous system that generally affects immunocompromised patients, but can affect immunocompetent patients and has a high mortality and morbidity. It usually presents with headache and fever; the presence of other symptoms are less common. Central nervous system cryptococcosis produces a wide variety of MRI features. Most common imaging finding in MRI is leptomeningeal and pachymeningeal enhancement, followed by dilatation of perivascular spaces e cryptococcomas. Restricted diffusion is rare, more common as a complication of the disease. While some of the radiographic features may be secondary to vascular injury, like in case one, restricted diffusion may be secondary to intramyelinic edema making the extracellular water restricted in its movement. **Final comments:** These cases presents atypical clinical and radiographic features of neurocryptococcosis. Early diagnosis is critical for successful treatment and avoiding neurological complications from this lethal infection, even in atypical cases.

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Atypical features of epileptic seizures

Mariana Holanda Gameleira[1], Samir Buainain Kassar[2], Mariana Lages Sarmento Barbosa[2], Fernando Tenório Gameleira[3], Felipe Camilo Santiago Veloso[4], José Gois de Almeida Neto[5], Maria Cristina Falcão Raposo[6], Rebeca Teixeira Costa[7], Davysson Castro[5]

[1]Hospital da Restauração, Recife PE, Brazil. [2]Centro Universitário Tiradentes, Recife PE, Brazil. [3]Hospital Universitário Professor Alberto Antunes, Maceió AL, Brazil. [4]Universidade Estadual de Ciências da Saúde de Alagoas, Maceió AL, Brazil. [5]Universidade Federal de Alagoas, Maceió AL, Brazil. [6]Universidade Federal de Pernambuco, Recife PE, Brazil. [7]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Mariana Holanda Gameleira; Email: marianagameleira_@hotmail.com.

Background: Epileptic seizures can reproduce from the known generalized tonic-clonic convulsive seizures, to piloerection, episodes of pure apnea, the most several types of pain, speech block, etc. The atypical manifestations of these seizures have not yet been properly defined. Clinical acumen is essential to improve diagnostic accuracy. **Objective:** To describe the prevalence and the clinical features of atypical epileptic seizures in patients with epilepsy, and to know the epidemiological profile of patients presenting atypical manifestations of epileptic seizures. **Methods:** A cross-sectional study was carried out with 1,244 patients from an epilepsy outpatient clinic of an University Hospital between February 2013 and May 2019. The presence of atypical epileptic seizures, and the epidemiological profile of the patients were analyzed. The prevalence of atypical epileptic seizures was determined in patients with epilepsy. For statistical analysis, we performed a case-control study, in which the control population were the patients who presented only typical seizures. We have excluded 305 patients due to uncertain diagnosis of epilepsy, and/or due to cognitive impairment, enough to compromise the reliability of their provided information. **Results:** A prevalence of 5.79% of patients with atypical epileptic seizures was observed, of which 1.85% had exclusively atypical seizures. In this group, female and patients over 50 years hegemony were observed, both with a prevalence of 60.8%. In addition, in atypical epileptic seizures patients, there was an evident predominance of seizures with a focal component, with indeterminate epileptic syndrome, normal neurological examination and with a frequency of more than one seizure per month. **Conclusion:** The prevalence of atypical epileptic seizures in patients with epilepsy is not negligible. The knowledge of these kind of seizures facilitates the early diagnosis of epilepsy, as well as provides an adequate treatment, and improves the patient's quality of life.

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Atypical initial manifestations of multiple sclerosis: a case report

Jonh Heberthon de Souza Araújo[1], Andréia Maria dos Santos Carvalho[1], Monique Melo Fortaleza[1], Michelly Bruna Soares de Moura[1], Raimundo Feitosa Neto[2] , Aline Gomes da Costa[1], Janine Midori Figueiredo Watanabe[1]

[1]Universidade Estadual do Piauí, Teresina PI, Brazil. [2]Universidade Federal do Piauí, Hospital Universitário, Teresina PI, Brazil.

Correspondence: Jonh Heberthon de Souza Araújo; Email: johnheberthon@hotmail.com.

Case presentation: A 35-year-old female was admitted to the neurology service on May 14th, 2024 presenting hemilingual ageusia, diplopia, and tinnitus. A month earlier, the patient reported having symptoms including increased fatigue and memory loss, as well as the onset of ageusia. The progression of the clinical condition led to diplopia, tinnitus, vertigo, and a differently patterned headache, which persisted up to the time of consultation. She denied motor symptoms or any other sensory manifestations. Her past medical history included chronic headaches since the age of 15, causing recurrent episodes of dizziness and fibromyalgia. A brain magnetic resonance imaging (MRI) performed on May 21st revealed more than nine lesions including: rounded lesions in the periventricular white matter and bilateral centrum semiovale; a lesion in the right forceps major of the corpus callosum and in the infratentorial compartment of the right posterior portion of the pons; and in the left paramedian region of the pontomedullary junction. Oligoclonal bands were present in the cerebrospinal fluid, confirming the diagnosis of multiple sclerosis. The patient was hospitalized for treatment with methylprednisolone pulse therapy for five

days, leading to clinical recovery. **Discussion:** Multiple sclerosis (MS) is a chronic inflammatory and neurodegenerative disease that affects the central nervous system and results from a demyelination process. It typically manifests between the third and fourth decades of life and can affect various areas, with notable otolaryngological symptoms, especially olfactory and audiovestibular. The tinnitus and vertigo reported in this case are part of the common audiovestibular alterations seen in MS patients due to the central nervous system demyelination. The diplopia reported by the patient results from lesions in the cranial nerves or in the brainstem, and it is among the most common visual complaints. Gustatory alterations, on the other hand, are extremely rare, especially when they occur early. However, they should not be underestimated and need careful investigation as potential indicators of multiple sclerosis, especially in young women. Clinically, it presents as transient ageusia, and on MRI, as demyelinating lesions in the cerebral taste area and pontine lesions, consistent with the patient's examination findings. The combination of symptoms and MRI findings points to brainstem involvement as the initial manifestation of the case, which is atypical and could potentially lead to a delayed diagnosis. **Final comments:** Few reported cases show ageusia, especially associated with diplopia, tinnitus, and vertigo, as initial symptoms of multiple sclerosis, making this an atypical manifestation of the disease. Therefore, a thorough anamnesis and a high degree of clinical suspicion are required for diagnosis in such situations.

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Atypical presentation of hypertensive cerebral lobar hemorrhage in a puerperal woman

Thayná Tavares Cutrim Everton[1], Vinicius Gabriel Peneluppi dos Santos[1], Bianca de Araujo Sobral[1], Gustavo Yukihito Anzou de Souza[1], Bruno Barros de Padua Andrade[1], Guilherme Previtale Vitorino[1], Luis Eduadro Belini Soares[1], Rodrigo Ferreira Faria[1], Giovana Santos Rodrigues[1], Luiz Fernando Mohallen Reynaldo[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Bianca de Araújo Sobral; Email: biancadeasobral@gmail.com.

Case presentation: MB, 41 year old, on the 7th postpartum day, was admitted to the emergency with a sudden onset of thunderclap headache upon waking, along with a sudden focal deficit of right-sided hemiparesis associated with motor aphasia. In the initial evaluation, she presented with hypertension (210/120 mmHg) and a cranial tomography revealed bifrontal hemorrhage with extravasation into the subarachnoid space and ventricles. She was referred to a tertiary care service, where arterial and venous angiographic tomography were performed, excluding cerebral venous thrombosis. Subsequently, she underwent cranial resonance and angiography, also without abnormalities, excluding ruptured aneurysms, arteriovenous malformations, angiomas, and cavernomas. During hospitalization, she experienced hypertension refractory to intravenous drugs, seizure crises, and a decreased level of consciousness, initiating measures for intracranial hypertension, invasive monitoring (ICP), and treatment for Status Epilepticus. After blood pressure control and electroencephalography, she showed awakening with partial remission of motor deficits and aphasia. After discharge, she underwent a post-hemorrhage follow-up exam to exclude structural causes and continues to have motor sequelae, for which she is undergoing rehabilitation. **Discussion:** Non-traumatic intracerebral hemorrhage (ICH) is a public health issue with an annual incidence of 30/100000 people, a mortality rate of 40%, and accounts for 20% of cerebrovascular events. This report describes a case of hypertensive ICH with bifrontal bleeding in a postpartum woman, a rare condition without precedent in the literature. Its most common topographies are basal nuclei (31%), putamen (30%), thalamus (25%), lobes (25%), ventricles (11%), pons (8%), and cerebellum (8%). In the lobar topography, 24% are unilateral and 26% multilobar, with the parietal being the most frequent (31%). The bifrontal finding is rare and generally associated with central thrombosis, which was ruled out by angiography, angiotomography, or angioresonance. The clinical presentations of ICH vary depending on its topography. In this case, there was a sudden focal deficit on the right side that became bilateral, indicating bilateral injury. The incidence of postpartum ICH is 0.05%, with maternal mortality between 60% (12% of maternal deaths). 50% of postpartum ICHs result from the rupture of an aneurysm or arteriovenous malformation, and rarely from the rupture of a carotid-cavernous fistula or cavernous angioma, Moyamoya disease, sickle cell anemia, and disseminated intravascular coagulation. In this case, the hypertensive peak on the postpartum was considered the cause of the bleeding. **Final comments:** This report highlights a rare presentation of hypertensive intracerebral hemorrhage due to its atypical topography and postpartum incidence. Despite its rarity, the diagnosis of such an entity must be prompt due to the high mortality involved with therapeutic delays.

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Atypical presentation of Rasmussen's encephalitis as progressive hemispheric atrophy syndrome without epilepsy: an unusual case report and the diagnostic challenge of a rare condition

Lucas Wilson Matos Gomes[1], Victor Hugo de Souza Silva Gomes[1], Gabriel Carvalho Alves[1], Gabriel Santos de Castro[1], Danyelle Rute Francisco e Silva[1], Letícia Leão Alvarenga[1], Guilhermes Henrique Cavalcante[1], Luany Patrícia Liberato de Oliveira[1], Denise Sisterolli Diniz[1], Delson José da Silva[1]

[1]Universidade Federal de Goiás, Goiânia GO, Brazil.

Correspondence: Lucas Wilson Matos Gomes; Email: lucaswilson95@hotmail.com.

Case presentation: A 22-year-old female patient, mixed race, presented five years ago with incomplete and disproportionate spastic hemiparesis predominantly in the right lower limb. She developed complete facial paralysis on the left side and, two years ago, began to experience decreased visual acuity on the left side, executive memory changes, and a decline in phonemic verbal fluency. She reports a febrile epileptic seizure at age 2 during an episode of acute pharyngitis but denies other paroxysmal events. Physical examination revealed complete facial paralysis on the left, lateral rectus muscle ophthalmoplegia with visual acuity of 20/70 (Snellen) in the left eye, and complete hemiparesis on the right with pyramidal release signs. Cranial MRI showed hyperintense areas on T2/FLAIR in the left temporal pole and cingulate gyrus, associated with hemiatrophy of the same hemisphere, accentuation of cerebral sulci and fissures, and hyperintense signals on T1/MTC in the left corticospinal tract from the precentral gyrus to the corona radiata. A biopsy of the left cingulate region showed histomorphological findings of a perivascular inflammatory infiltrate predominantly of CD4 and CD8 T lymphocytes, without identifiable cytotoxic cells, microglial activation with rare microglial nodules, and diffuse gliosis. Testing for Epstein-Barr virus (EBV) antigens was inconclusive, and tests for cytomegalovirus (CMV) and herpes simplex types 1 and 2 were negative. Cerebrospinal fluid (CSF) analysis showed oligoclonal bands with an intrathecal production pattern type 2. Infectious serologies for other agents and tests for autoantibodies anti-AQP4, anti-MOG, anti-NMDA, and anti-GAD were negative. The electroencephalogram was within normal limits and showed no evidence of new clinical or electrographic seizures since the patient's follow-up began. Based on the imaging and histopathological findings, the patient meets the diagnostic criteria for Rasmussen's encephalitis (RE) according to the Bien et al. (2005) criteria. **Discussion:** RE is an immune-mediated syndrome characterized by progressive chronic encephalitis affecting one cerebral hemisphere. It typically results in intractable focal epilepsy, progressive focal neurological deficits, and intellectual decline related to the affected hemisphere, with neuroimaging revealing progressive unilateral cerebral hemiatrophy. This case presents an atypical evolution by not showing one of the main characteristics of the disease: refractory epilepsy. Therefore, it was necessary to proceed with a brain biopsy, which resulted in findings compatible with RE, although not specific. **Final comments:** This is a difficult diagnosis due to the rarity and atypical presentation of the syndrome. Thus, there is a need to review the diagnostic criteria to increase the sensitivity and specificity of diagnoses, as in the case reported above.

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Atypical presentation of rhabdomyolysis due to epilepsy in a patient with McArdle disease

Raíssa Josefa Pereira de Moura[1], Cleberson Alberceli Vieira de França[1], Flávio Laurentino de Sousa Júnior[2], Mayhara Rosany da Silva Santiago[1], Ana Caroline Paiva Simeão[1], Nereu Alves Lacerda[1], Renata Amaral Andrade de Mendonça[1], Heron Alves Vale[1], Natália Nunes Tenório[1], Ana Beatriz Roberto Lima Borba[1]

[1]Hospital Pelópidas da Silveira, Recife PE, Brazil. [2]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Raíssa Josefa Pereira de Moura; Email: rayssamoura16@gmail.com.

Case presentation: A 45 year-old woman, previously diagnosed with Glycogen Storage Disease Type V (McArdle disease), was hospitalized due to bilateral intense muscle pain in gastrocnemius, knees and triceps that did not respond to medications. Neurological examination revealed proximal tetraparesis, predominantly on lower limbs, and global hyperreflexia. There was no history of physical exertion prior to this clinical condition and no signs or symptoms of infectious diseases were present. Initial laboratory exams showed blood urea nitrogen (BUN) of 28 mg/dL and creatinine 0.84 mg/dL, but had ascending levels of CPK (2289-10.005 U/L). During hospitalization, we also noted mild contractures of short duration on quadriceps, without any facial involvement or impairment of consciousness. However, after one of those contractures, the patient had a tonic-clonic seizure, which was followed by an acute kidney injury (Creatinine 3.1 mg/dL; BUN 57 mg/dL) and progressive increase on CPK levels (CPK = 169.926 U/L). Interictal electroencephalogram showed frequent bilateral frontal-polar epileptiform activity. The patient is still on hemodialysis due to rhabdomyolysis, but after administration of anti-seizure medications the CPK levels started to decrease. **Discussion:** McArdle disease is a genetic metabolic myopathy that occurs due to deficiency of the enzyme myophosphorylase, which causes intolerance to physical exercise. The disorder is mainly characterized by muscle pain, weakness and fatigue after periods of physical activity. Severe muscular damage can occur following intense physical exertion, resulting in acute kidney injury and rhabdomyolysis. Although physical exertion is a known trigger for muscular damage on McArdle disease, the patient in this case had no history of previous physical exertion. Traumas, inflammatory and infectious diseases are also potential etiologies of rhabdomyolysis on those patients, but there was also no clinical or laboratory evidence that pointed to such

causes. Epileptic seizures are not a common occurrence on McArdle disease, but are a possible precipitant factor of rhabdomyolysis that must be actively investigated if clinical and laboratory worsening without a clear factor is identified. There are reports of magnetic resonance spectroscopy questioning a possible accumulation of glycogen on the brain of patients with McArdle disease, which raises the possibility of acute symptomatic seizures of metabolic etiology. **Final comments:** Here we highlight the importance of actively investigating occult epileptic seizures as an etiology for patients with McArdle disease who progress to rhabdomyolysis, especially when there is not a clear cause for the muscular damage.

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Atypical presentation of Turner-type X-linked syndromic intellectual developmental disorder: a pediatric case report

João Vítor Moreira Nogueira[1], Oliver Reiks Miyajima[2], Alessandra Marangoni Fante[1], Bruna Barbosa Nobre[1], Enzo Lima Alcântara Parente[1], Marcelo de Almeida Pinheiro[1]

[1]Unichristus, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: João Vítor Moreira Nogueira; Email: jvmnogueira10@gmail.com.

Case presentation: A 5-year-old female patient, daughter of a young non-consanguineous parents with no family history of neurological impairment, was referred by her pediatrician for early stimulation with occupational therapy (OT), physical therapy, and speech therapy at the age of two months due to global delay in neuropsychomotor development, partially reaching the expected milestones for her age. At the age of three, due to persistent delay in language comprehension, low inhibitory control, visual and auditory hypersensitivity, she was evaluated by a neurogeneticist who, upon physical examination, identified apparent intellectual disability, craniofacial dysmorphisms such as broad forehead, ocular hypertelorism, hypoplasia of the ear lobes, dysmorphic upper labial frenulum, thin upper lip and prominent nasal columella, alongside skeletal anomalies in the toes. Following investigation, complete exome sequencing was requested, which revealed a de novo pathogenic variant in heterozygosity in the HUWE1 gene (chrX:53.647.391 G> A), confirming the diagnosis of Turner-type X-linked syndromic intellectual developmental disorder with OMIM nº 309590. Subsequent multidisciplinary evaluation indicated low performance in expressive language, delay in intellectual acquisition and processes, auditory and visual hypersensitivity, fine motor dyspraxia and reduced sustained attention. Currently, the patient undergoes regular multidisciplinary follow-up with psychology, psychomotricity, speech therapy and OT, showing improvements in motor and cognitive development. **Discussion:** Turner-type X-linked syndromic intellectual developmental disorder is a rare neurodevelopmental disorder with a genetic basis caused by pathogenic variants in the HUWE1 gene. This disorder follows an X-linked inheritance pattern, however, in this case, atypically presented due to the de novo mutation, as there is no family history of the disease. The phenotype of this condition is highly variable. Heterozygous females, as in the case in question, may present mild manifestations or be asymptomatic. Affected individuals usually show global developmental delay since childhood, with intellectual impairment of varying intensity. Common dysmorphic features include small palpebral fissures, low-set dysplastic ears, long face, thin upper lip, and mild distal skeletal anomalies, such as brachydactyly. **Final comments:** It is concluded that Turner-type X-linked syndromic intellectual developmental disorder, caused by pathogenic variants in the HUWE1 gene, presents a wide range of phenotypic manifestations, thus highlighting the importance of genetic evaluation of patients with global delay and dysmorphisms, even if minor, in addition to an early multidisciplinary approach to optimize the development of multiple domains compromised by the underlying disease, including language, motor skills, cognition, among others.

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Atypical presentations of MUSK-congenital myasthenia syndrome

Henrique Alves Bezerra[1], Nicole Lopes Cavalcante Machado[1], Eduardo de Paula Estephan[2], Maria Sheila Rocha[2], Paulo Victor Sgobbi[3], Antonio Edvan Camelo-filho[3], Pedro Lucas Grangeiro de Sá Barreto Lima[4], Paulo Ribeiro Nóbrega[4], Cristiane Araujo Martins Moreno[2]

[1]Faculdade Santa Marcelina, São Paulo SP, Brazil.
[2]Hospital Santa Marcelina, São Paulo SP, Brazil.
[3]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[4]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Henrique Alves Bezerra; Email: riq_alves@hotmail.com.

Background: Congenital myasthenic syndromes (CMS) are a group of disorders caused by mutations in genes that encode proteins at the neuromuscular junction. The typical clinical presentation involves congenital or childhood-onset muscle weakness affecting ocular, respiratory, bulbar, and limb muscles, alongside fatigability. However, adult-onset and atypical clinical features have also been reported, depending on the specific gene involved. The MUSK gene, which encodes the muscle-specific kinase (MuSK) protein crucial for neuromuscular synapse formation, is rarely implicated in CMS. The clinical phenotype associated with MUSK mutations is highly variable, ranging from severe neonatal presentations to mild early-childhood limb-girdle weakness often accompanied by early ventilatory insufficiency. **Methods:** Six patients with CMS caused by biallelic variants in the MUSK gene were evaluated at three tertiary centers. They provided informed consent for inclusion of their clinical data in this study. **Results:** We presented the clinical details of six patients with MUSK-CMS. Four males and two females aged 19 to 58 years (mean 36.5 +-12). One patient manifested symptom congenitally and the remaining five in adulthood (mean onset age for adult-onset cases was 21.33 +-6 years). Clinical presentation included mild limb-girdle weakness, respiratory dysfunction, and varied responses to treatments such as salbutamol and pyridostigmine. Our findings underscore the diverse clinical presentations of MUSK-CMS. While childhood-onset cases may not always involve significant respiratory issues, adult-onset presentations can be debilitating, particularly with respiratory involvement. The positive response to salbutamol in all patients suggests its efficacy as a potential treatment. Three patients treated with pyridostigmine showed no worsening of symptoms. Adult-onset forms of MUSK-CMS, uncommon but marked by significant respiratory involvement, differ from the typical congenital or childhood-onset phenotype described in the literature. The varying treatment responses, including pyridostigmine's lack of exacerbation of symptoms, highlight the heterogeneity among individuals with MUSK mutations. **Conclusion:** MUSK-CMS is a rare yet clinically diverse condition that may present in adulthood despite the typical congenital or childhood-onset patterns. Our study emphasizes the importance of recognizing and comprehending the diverse clinical presentations and treatment responses in MUSK-CMS. Tailored management strategies are crucial to optimizing outcomes in this patient population, underscoring the need for further research to elucidate underlying mechanisms and refine therapeutic approaches.

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Aura change in migraine: be careful!

Taís Luise Denicol[1,2], André Luiz Girotto[1,2], Carolina Matte Dagostini[1,2], Juan Sebastián Sánchez León[1,2], Pedro Hall Ruschel[1,2], Renato Dumba Monteiro de Castro[1,2], Alexandre Laisson Prado Taschetto[1,2], Fernando Kowacs[1,2], Liselotte Menke Barea[1,2], Paulo Valdeci Worm[1,2]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Taís Luise Denicol; Email: taisldenicol@gmail.com.

Case presentation: Female, 49 years old, previously hypertensive and with migraine with aura. Medications: Losartan 50mg SID and atenolol 25mg SID. Denies smoking and other habits. Patient had infrequent episodic migraines with typical visual aura since youth without prophylactic treatment. Positive family history of migraine with aura. The aura was characterized by a fortification spectrum with gradual onset lasting 15-20 minutes followed by a unilateral, pulsatile and periorbital pain associated with nausea, fono/photophobia. However, six months ago, the attacks increased in frequency to 3-4x per week and started with a chronic cough. Two months ago, there was a change in the pattern of the aura, which sometimes became a figure similar to a disco ball. She had 03 episodes of this aura and in the last one followed by a generalized tonic-clonic seizure in which she was referred to a tertiary health service. On physical examination, breath sounds were abolished at the base and decreased at the apex on the left lung. No changes in neurological examination. A chest x-ray was performed with extensive pleural effusion on the left and a head computed tomography showing an intra-axial occipital lesion on the right. It is complemented by a computed tomography of the chest, which reveals a lesion of probable primary neoplastic nature in the left lower lobe. Resection of the extra-axial right occipital brain lesion and continued with the falx and intra-axial lesion with anatomopathological analysis showed metastasis of adenocarcinoma in connective tissue and brain parenchyma. The immunohistochemical study associated with the histopathological findings and clinical data received are compatible with adenocarcinoma metastasis, with a probable primary site in the lung. **Discussion:** Most headaches are a primary disorder, however it is important that during any medical evaluation, alarm signs have been looked for systematically, like the change of pattern of aura, seizures and frequency increase. Red flags have been promoted in the medical literature and it helps the clinician to initiate a workup plan. SNNOOP10 list is a systematic way to screen patients with a new headache and will help to detect a secondary cause. This patient showed pattern change or recent onset of headache that in SNNOOOP10 suggests neoplasms, headaches attributed to vascular, nonvascular intracranial disorders. **Final comments:** The importance of investigating patients with changes in headache and aura patterns that are listed in SNNOOOP10 is reinforced. It is worth noting that even patients with primary headaches disorder can develop secondary headaches disorder and they deserve attention, as in the case of this patient.

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Autoimmune necrotizing myopathy associated with 3-hydroxy-3-methylglutaryl-coa reductase (HMGCR) antibody after COVID infection: a case report

Samara Andrea da Costa Fonseca[1], Elberth Henrique Miranda Teixeira[1], Caio Eduardo Rodrigues Falcao[1], Luiziane Maria da Silva Alves[1], Pablo Vinicius Silveira Feitoza[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Samara Andrea da Costa Fonseca; Email: samaradacostafonseca@gmail.com.

Case presentation: 64-year-old woman from Manaus, Amazonas. On January 6, 2021, she started suffering from flu-like symptoms and was diagnosed with COVID four days later. Over the following week, she presented with muscle weakness in the lower and upper limbs, with difficulty climbing stairs and walking, including difficulty supporting her neck; associated with dysphagia, myalgia and cramps. On physical examination, the patient was hypotonic, with hypotrophy in the proximal region of the upper and lower limbs; symmetrical tetraparesis with a proximal crural predominance; hyporeflexia in the triceps, biceps and patella bilaterally; preserved superficial and deep sensitivity; eumetrics and eudiadochokinetics; cranial nerves without alterations; myopathic gait requiring bilateral support for walking; absence of skin lesions. Laboratory tests showed hyper-CPKemia of 6,893 U/L. Etiological stratification revealed a histopathological pattern of autoimmune necrotizing myopathy on muscle biopsy and positivity for HMGCR antibody (235, normal <20UA). She is being followed up with methotrexate with a good therapeutic response. **Discussion:** This case of confirmed autoimmune necrotizing inflammatory myopathy associated with HMGCR antibody demonstrates the relevance of SARS-CoV-2 virus infection as a trigger for the development of autoimmune neurological diseases. Autoimmune myopathies represent a set of rare, heterogeneous pathologies and are a major challenge in the medical field when it comes to differential diagnoses. Immune-mediated necrotizing myopathy is a recently described form of idiopathic inflammatory myopathy characterized by a severe clinical picture with rapidly progressive muscle weakness, axial involvement, early muscle atrophy and high CPK titers. Currently, there are two main categories, with anti-signal recognition particle autoantibodies (SRP-IMNM) and anti-3-hydroxy-3-methylglutaryl-coa reductase autoantibodies (HMGCR-IMNM). Serum anti-SRP autoantibodies in IIM were first detected in 1982 and anti-HMGCR autoantibodies were first detected in 2010-2011. **Final comments:** This case study enabled us to make a differential diagnosis between inflammatory myopathies. This report is extremely relevant, as these nosological entities share numerous signs and symptoms, which makes diagnosis challenging, especially considering the correlation with COVID infection.

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Autoimmune optic neuritis in a patient with hiv on irregular treatment: case report

Paulo Henrique Martinelli Oliveira[1], Amanda Michelucci dos Santos[1], Rebeca Aranha Barbosa Sousa[1], Larissa Miyashiro[1], Raderi Luiz Cardoso Do Santos[1], Lucas Silva Dias[1], Laiz Helena de Pádua[1], Gustavo Carvalho Costa[1], Alexandre Coelho Marques[1]

[1]Hospital Municipal Dr. José de Carvalho Florence,

Correspondence: Paulo Henrique Martinelli Oliveira; Email: paulohmoliveira96@gmail.com.

Case presentation: Patient E.E.M.F, female, 35 years old, diagnosed with HIV 13 years ago, with irregular treatment (CD4 count: 315, Viral load: 2830 copies). Discontinued antiretroviral therapy in April/2023. In December/2023, developed pain upon movement of the left eye, associated with loss of visual acuity ("counting fingers at 1 meter"), and left optic disc edema was observed without other additional findings. At the time, due to suspicion of Cytomegalovirus infection, intravenous Ganciclovir was initiated with no improvement in symptoms. On the 7th day of antiviral therapy, she underwent Methylprednisolone 1g/day for 5 consecutive days, showing significant improvement in visual acuity (20/70) by the end of pulse therapy. In March/2024, she again presented a similar condition, experiencing a new worsening of visual acuity (20/400) in the left eye. Again treated with Methylprednisolone 1g/day (5 days), with partial recovery after treatment (20/70). Cerebrospinal fluid evaluation showed mild pleocytosis (05 cells/mm^3) and proteinorrhaquia (59 mg/dL), presence of oligoclonal bands, and positive PCR for Epstein-Barr Virus (EBV). Research was negative for other viruses. Magnetic resonance imaging revealed a slight hypersignal on FLAIR/T2 in the right frontal semi-oval center. **Discussion:** Although HIV infection predominantly affects lymphocytic load (CD4) and Multiple Sclerosis (the most prevalent autoimmune disease of the central nervous system) has its pathophysiology intrinsically associated with lymphocytes, recent studies demonstrate a relationship between such pathologies, with viral infection potentially acting as a trigger for a demyelinating outbreak. Furthermore, the positive PCR for EBV in the cerebrospinal fluid may be related to disease activity. **Final comments:** The associations and neuroimmunological triggers related to autoimmune diseases in HIV patients remain largely unknown. Given the high epidemiological focus of HIV in Brazil and its potential concurrence with such autoimmune phenomena, the study and investigation of these pathologies are essential.

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Automatic prediction of ASPECTS for patients with posterior circulation infarcts

Aditya Santosh Kondepudi[1], Manuela Yphyn Faria Sung[2], Noam Rotenberg[1], Andreia Vasconcellos Faria[1]

[1]Johns Hopkins University, Baltimore, US. [2]Universidade Estadual do Oeste do Paraná, Cascavel PR, Brazil.

Correspondence: Manuela Yphyn Faria Sung; Email: manuela.yphyn@gmail.com.

Background: The Alberta Stroke Program Early CT Score (ASPECTS) visually assesses the extent and location of ischemic core in Middle Cerebral Artery (MCA) ischemic strokes. Due to its simplicity, ASPECTS gained popularity and was adapted for diffusion-weighted MRIs (DWI). However, variability in human evaluation affects ASPECTS' effectiveness in treatment selection. Automatic classification methods that can replicate expert opinion might add an extra level of reproducibility and aid urgent medical decisions like thrombolysis or prognostic prediction. **Objective:** We previously developed a fully automatic system to calculate MCA-ASPECTS, matching consensus expert readings. This study extends the system to calculate ASPECTS in posterior circulation strokes (PCA-ASPECTS). **Methods:** The study included MRIs of patients admitted to our stroke center with ischemic stroke between 2009 and 2019. This subset of the public "Annotated Clinical MRIs and Linked Metadata of Patients with Acute Stroke" (https://www.icpsr.umich.edu/web/ICPSR/studies/38464). Stroke cores were manually traced. Models identifying ischemic regions were trained on 1,414 MRIs with ischemic evidence in DWI and tested on an independent dataset of 160 patients with posterior circulation infarcts. The models' efficiency was compared to consensus expert evaluations and inter-evaluator comparisons. **Results:** The Random Forest model was the most efficient for classifying stroke locations in the posterior circulation. Balanced accuracy (0-1 scale, with 1 being perfect) was: PCA 0.87; cerebellum 0.92; midbrain 0.84; pons 0.84; thalamus 0.87. This performance rivaled inter-evaluator classification. The model's accuracy in predicting unilateral or bilateral lesions in the PCA, cerebellum, and midbrain averaged 0.9. Accuracy for predicting PCA-ASPECTS was 0.65, and 0.85 for "flexible" ASPECTS (±1 point tolerance). We developed a fully automated system for calculating PCA-ASPECTS with accuracy comparable to inter-evaluator agreement. While accuracy for total score calculation was slightly lower due to multiclass issues, accuracy for predicting individual affected regions and flexible ASPECTS rivaled or exceeded inter-evaluator agreement. This system is part of our Acute Stroke Detection and Segmentation tool, ADS (https://www.nitrc.org/projects/ads), which outputs digital infarct masks, proportions of brain regions injured, automatic radiology reports, and MCA-ASPECTS, all with explanatory features. ADS is public, free, accessible to non-experts, and runs in real time on local CPUs with minimal computational requirements. **Conclusion:** We developed an automated and accessible system to accurately predict PCA-ASPECTS, potentially enhancing acute stroke assessment and supporting urgent medical decisions and reproducible research.

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Autosomal recessive spinocerebellar ataxia type 8 and associated non-cerebellar features

Bruno Wendorff Lucci[1], João Carlos Cervelin[1], Fernanda Coan Antunes[1], Pricila Bernardi[1], André Dias de Oliveira[1], Gisele Espíndola[1]

[1]Universidade Federal de Santa Catarina, Florianópolis SC, Brazil.

Correspondence: Bruno Wendorff Lucci; Email: brunowlucci@hotmail.com.

Case presentation: A 58-year-old man presented with lifelong difficulties in running and climbing stairs. He maintained functionality until age 30, when symptoms gradually escalated, including incoordination, imbalance, slurred speech, double vision, hearing loss, and urinary incontinence. Family history revealed consanguineous parents. Neurologic exam findings included an ataxic gait requiring unilateral support, thoracic kyphosis, slurred and nasal speech, hypermetria on saccadic movements with torsional nystagmus on lateral gaze, diffuse hypotonia, normal strength and reflexes without pyramidal signs, bilateral reduction in distal vibratory sensation up to the malleoli, dysmetria, and dysdiadochokinesia. MRI revealed significant volumetric reduction in cerebellar hemispheres, vermis, and cerebellar peduncles. Electroneuromyography indicated moderate to severe sensory-motor peripheral axonal polyneuropathy, predominantly sensory, affecting the distal lower limbs. Audiometry showed bilateral sensorineural hearing loss. Genetic testing identified a homozygous mutation in the SYNE1 gene. **Discussion:** Autosomal recessive spinocerebellar ataxia type 8 (SCAR8), caused by mutations in the SYNE1 gene, is a rare autosomal recessive disorder. This gene, located on chromosome 6q25.2, is one of the largest in the human genome and comprises 146 exons. Onset typically occurs between ages 6 and 42, with classical symptoms including cerebellar ataxia and motor neuron disease. Patients often experience decreased physical activity performance compared to peers and may initially present with ALS-like symptoms. Other manifestations can include cognitive deficit, brainstem dysfunction with oculomotor abnormalities, skeletal anomalies and urinary incontinence. In South America, SYNE1 mutations account for 1.89% of autosomal recessive cerebellar ataxia cases with a known genetic cause. Large cohort studies indicate that 64-81% of SCAR8 patients exhibit ataxia-plus syndromes, characterized by ataxia with additional features. The SYNE1 gene's size and its role in nuclear stability are linked to degenerative conditions, as mutations affect nucleus stability, size, and shape. Truncating mutations near Nesprin-1's N-terminal are associated with late-onset cerebellar phenotypes, while C-terminal mutations cause early-onset, complex presentations. Imaging studies show reduced cortical thickness, volume reduction in subcortical structures, brainstem, and particularly the cerebellum. White

matter disruption is evident in both the brainstem and cerebellum, while spinal cord is typically spared. **Final comments:** This case illustrates the importance of recognizing extra-cerebellar symptoms in SCAR8, which align with findings that most cases of SCAR8 present with ataxia-plus features. Comprehensive genetic testing is crucial for accurate diagnosis and understanding of SCAR8, highlighting the need for further research to elucidate the pathomechanisms and genetic variations of this disorder.

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B12 vitamin deficiency and dementia in the elderly

Gabriela de Castro Cavalcante Mendonça[1], Antônio Pinheiro Goes Curvello[1], Italo Francisco Valença da Costa[1], Luisa Villanova Valadares[1], Nathalia Spacassassi Rodrigues[1], Sabrina Gomes de Oliveira[1]

[1]Centro Universitário de Maceió, Maceió AL, Brazil.

Correspondence: Gabriela de Castro Cavalcante Mendonça; Email: gabrieladecastrocm@gmail.com.

Background: Alzheimer's disease is the most common form of dementia, with a prevalence of 70% of cases, with risk factors including advanced age, genetics, low educational level and cardiovascular disease. Vitamin B12 deficiency, linked to cognitive decline, can cause neurological degeneration and affects DNA and myelin synthesis. **Objective:** Evaluate the relationship between vitamin B12 deficiency and dementia. **Methods:** A systematic literature review was realized on the correlation between vitamin B12 deficiency and cognitive decline in dementia. The information was found in articles in the PubMed database between 2012 and 2022. Inclusion criteria involved systematic review and meta-analysis studies, written in English and focused on the cognitive function of vitamin B12. The selection of articles was carried out by 5 reviewers, independently, with differences resolved by consensus. 7 articles that met the criteria were included. **Results:** Vitamins from the B complex, especially vitamin B12, have an immunomodulatory and neurotrophic function in the nervous system. In the meantime, its deficiency is associated with reduced myelination of axons, impairing cognition and the integrity of the brain structure. Studies have shown the prevalence of vitamin B12 deficiency, through elevated serum markers, such as methylmalonic acid and homocysteine, in elderly patients (> 60 years old) with dementia. Thus, it was demonstrated that B12 supplementation, associated with adequate nutrition, has a significant role in improving the Mini Mental State Examination (MMSE) score, a test whose function is the cognitive assessment and tracking of dementia, indicating a reduction of cognitive decline in elderly patients diagnosed with dementia. B12 deficiency, prevalent due to reduced gastrointestinal absorption associated with aging and certain health conditions, is not only linked to cognitive decline and dementia, but may also be a reversible cause of mental deterioration. Some studies indicate that supplementation may be more effective in elderly people with confirmed cognitive impairment, with regular monitoring of B12 levels being part of a preventive approach. Vitamin supplementation in elderly people with cognitive impairment and dementia demonstrated significant improvements, benefiting more than half of test participants. It is important to note that vegetarianism was not related to the worsening of cognitive diseases, indicating that other factors may be more determining for the development of these conditions. **Conclusion:** As it has an immunomodulatory and neurotrophic function in the nervous system, vitamin B12, when deficient, becomes an important factor in the investigation of cases of dementia in the elderly. Furthermore, the studies analyzed demonstrated improved performance on the Mini Mental State Examination (MMSE) among participants who received adequate B12 supplementation.

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Bacterial meningitis in the Brazilian public health system: a regional comparison over the last decade

João Lucas Lima Diogo[1], Ana Clara Costa de Siqueira Campos[1], Gabriela Morais de Medeiros Dias Melo[1], Joanna Sousa da Fonsêca Santana[1], Lara Millena Miranda Frota[1], Luísa Lettiery Andrade Santos[1], Maria Antônia Vianna Santos[1], Maria Eduarda Kobayashi Teixeira[1], Ruben Costa Santos Neto[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: João Lucas Lima Diogo; Email: joaodiogo20.2@bahiana.edu.br.

Background: Bacterial meningitis is a global public health issue, with an incidence of 80 cases per 100,000 inhabitants in developing countries. Despite the existence of vaccines against the main pathogens, it remains highly prevalent among children and the elderly. The mortality rate can reach 54%, leading to debilitating neurological sequelae. Therefore, it is crucial to improve healthcare services and implement effective public health policies to reduce the incidence and impact of bacterial meningitis. **Objective:** To analyze the demographic distribution and hospital morbidity of bacterial meningitis in Brazil. **Methods:** This is a descriptive ecological study that analyzes the number of hospitalizations and mortality rates due to bacterial meningitis in the state of Bahia from 2014 to 2023. Data sources included the Hospital Information System (SIH) and Mortality Information System (SIM) provided by DATASUS, as well as data on resident population. Variables considered were number of hospitalizations, deaths, and mortality rate. Mortality rate was calculated as deaths per thousand inhabitants. **Results:** The Southeast region stood out with the highest number of hospitalizations and deaths, totaling 14,572 hospitalizations and 1,590 deaths, respectively. In contrast, the Southern region showed the greatest reduction in the number of deaths, with a decrease of 62 deaths from 2015 to 2016, and the highest percentage reduction in mortality rate, decreasing by 33.23% from 2015 to 2016. The Southeast region had the highest absolute growth in one year, with an increase of 614 hospitalizations from 2014 to 2015, while the North region recorded the highest percentage growth in hospitalizations, increasing by 225.8% from 2014 to 2015. The Northeast region had the highest percentage increase in mortality rate, rising by 58.49% between 2020 and 2021. Regions ranked in descending order of highest mortality rates were: Southeast, North, Northeast, South, and Midwest, with the Southeast region presenting the highest average rate of 10.91 and the Midwest region the lowest at 8.33. **Conclusion:** This study highlights significant regional variations in understanding the epidemiology of bacterial meningitis. The Southeast region had the highest number of hospitalizations and deaths, while the Southern region showed the greatest reduction in mortality. In contrast, the Northeast region experienced a concerning increase in mortality rate between 2020 and 2021, and the North region saw a significant increase in hospitalizations. Study limitations include the inability to establish causality due to the descriptive nature of the data and potential inconsistencies in data quality collected by SIH and SIM. Further studies are recommended to better understand regional factors and develop specific strategies to mitigate the incidence and burden on the healthcare system.

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Balint syndrome in the three-territory sign outbreak

Bárbara Luisa Coraspe Gonçalves[1], Maria Luiza Levindo Coelho Martinis[1], Luiz Guilherme Gerhardt Fernandes[1], Bárbara França Amorim[1], Marina Brasileiro Cesar Leitão[1], Osvaldo Pantoja de Oliveira[1], Ana Carolina Andorinho de Freitas Ferreira[1], Cristiane Borges Patroclo[1], Marcelo Cagy[1]

[1]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.

Correspondence: Bárbara Luisa Coraspe Gonçalves; Email: barbaracoraspeg@gmail.com.

Case presentation: A 71-year-old man, with hypertension, diabetes mellitus, and metastatic prostatic cancer, woke up with left faciobrachial paresis and dysarthria. He arrived at the Hospital Emergency 8 hours after the ictus, when the first head CT showed a right frontal stroke. Just three days later, a neurology consultation was requested. The patient was very inattentive, especially to his left side. Initiation of horizontal saccadic eye movements was impaired bilaterally and it was noticed signs of ocular apraxia and simultagnosia. Although there were superposed findings of left extinction and hemianopsia, there was also a consistent Balint syndrome (BS). A novel CT showed additional biparieto-occipital and right frontal ischemia. **Discussion:** BS is a rare neurologic manifestation of constrained attention and limited spatial orientation in visual processing. It was firstly reported by Reszo Balint in 1909 in a patient also suffering from left neglect. It is described by a triad of simultagnosia, oculomotor apraxia and optic ataxia. This syndrome derives from bilateral parietal lesion, including parietooccipital junction. Etiologies are diverse: cerebrovascular disease (especially watershed infarctions), tumour, trauma, prion disorders, viral infections such as HIV and neurodegenerative conditions, mainly the posterior variant of Alzheimer's disease (Posterior Cortical Atrophy). Simultagnosia is the inability to see multiple stimuli at a time, regardless of visual acuity or field. It can be classified as ventral and dorsal types. In the ventral type, there is involvement of occipitotemporal ("what") pathway. Its dysfunction impairs the visual discrimination of objects without affecting the perception of their position. On the other hand, the "dorsal" or parietal cortical ("where") pathway, participates in motion perception. Its damage compromises spatial-temporal analysis, leads to the inability to judge objects position and size, and flawed visually guided eye and hand control. Hence, this pathway lesion is classically related to the BS, although both pathways might share similar processing mechanisms. Optic ataxia is the difficulty in reaching under visual guidance in the absence of motor or primary visual deficits. Oculomotor apraxia is the inability to voluntarily shift gaze in spite of intact eye rotation. The three-territory sign consists of cerebral infarctions of the bilateral anterior and posterior circulation. It is highly predictive of Trousseau's syndrome, a hypercoagulable state associated with advanced cancer. **Final comments:** BS is the result of higher-level visual processing dysfunction, characterized by the triad of optic ataxia, oculomotor apraxia, and simultagnosia. The current case depicts a rare clinical manifestation caused by multiple vascular territories ischemia, parallel to the condition of metastatic cancer.

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Basal ganglia calcification associated with hypocalcemia and hypomagnesemia: a case report of Fahr's disease

Sarah Cardoso Moreira Fagundes[1], João Victor Vieira Giacometti[1], Rodolfo Souza de Faria[1]

[1]Faculdade de Medicina de Itajubá, Itajubá MG, Brazil.

Correspondence: João Victor Vieira Giacometti; Email: joaovgiacometti@hotmail.com.

Case presentation: A 47-year-old male patient underwent thyroidectomy in 2012 due to seven benign nodules, resulting in the partial removal of the parathyroid glands.

Continuous hormone replacement therapy with levothyroxine sodium and calcium citrate malate associated with vitamin D3 was recommended. However, the patient autonomously discontinued the treatment the same year, staying without following medical recommendations for the next 8 years, leading to progressive myasthenia in the limbs and dysarthria. In 2020, then, he experienced eight seizures and weekly episodes of muscle spasms, attributed to severe hypocalcemia and hypomagnesemia, which cause hyperexcitation of the central nervous system. The following year, bilateral brain calcifications in the basal ganglia, thalami, dentate nuclei, and subcortical regions of uncertain origin were detected through computed tomography scans. In 2023, after six months of diligent home follow-up due to the fact that he still did not recognize the importance of his treatment, the patient finally adhered to a daily medication regimen including calcitriol 0.5mg, magnesium chloride 500mg, calcium carbonate 500mg with vitamin D3 400mg, levothyroxine sodium 150mg, clonazepam 2mg, and phenobarbital 100mg, resulting in the cessation of seizures and clinical improvement. However, the patient's speech rigidity progressively worsened. **Discussion:** Fahr's disease affects less than 2% of the global population and typically manifests from the fourth decade of life. Despite its uncertain pathophysiology and etiologies, it is agreed that hypocalcemia and hypomagnesemia, associated with brain calcifications, are hallmark conditions of this pathology. Alongside a probable genetic predisposition, the factors leading to this disease are linked to hyperexcitation of the nervous system, resulting in seizures, spasms, and muscle rigidity. In the discussed case, chronic serum reduction of calcium and magnesium levels, following thyroidectomy and partial parathyroidectomy, was exacerbated by persistent non-adherence to treatment. Additionally, brain imaging emphasizing cerebral calcinosis supported the suspicion of Fahr's disease. Thus, the proposed treatment aimed to alleviate neuromuscular symptoms by normalizing the serum profile. However, the persistent worsening of speech rigidity suggests a possible irreversible involvement of brain areas due to the chronic inadequately treated condition. **Final comments:** This study details a rare case of Fahr's disease, a condition linked to brain calcifications and severe hypocalcemia. The description of this case is crucial for enriching the limited knowledge about this pathology, contributing to the investigation of etiologies and treatments to improve the quality of life of affected patients. The limitations of this research include the scarce scientific literature and the challenging clinical detailing of the case due to the prolonged progression of the disease.

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Basal ganglia encephalitis with fulminant Parkinsonian features and dysautonomia in a child: look no further than levodopa

Marciéli Gerhardt[1], Andressa Noschang[1], William Alves Martins[1]

[1]Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Marciéli Gerhardt; Email: marcieligerhardt@yahoo.com.br.

Case presentation: A 5-year-old presented to our service with a history of altered mental status, rigidity and tremors. A few days prior, he presented a minor upper respiratory illness, which improved upon receiving azithromycin for 5 days. Six days after, he became confused and developed a slow and monotonous voice tone. At the emergency room, head CT was unremarkable. Lumbar puncture showed 7 cel/mm^3 (100% lymphocytes) with slightly increased protein on CSF, prompting initiation of acyclovir for possible HSV encephalitis. Despite treatment, his condition worsened with supervening spasms, akinetic mutism, worsening conscience and severe dystonic "seizures". MRI showed bilateral striatum and caudate hyperintensities, with minor gadolinium enhancement. At the same time, PCR for HSV and other viruses returned negative, and he was started on methylprednisolone 20mg/kg and IVIG 400mg/kg/day for 5 days each. Acyclovir was weaned off and he was transferred to our hospital. At arrival, he had painful dystonia in which his lower limbs would extend with flexion of his arms, at times with oromandibular spams - so severe he eventually lesioned his lower lips - intermixed with low frequency tremors. After the immunotherapy he could interact, however still exhibited akinetic mutism. Alongside worsening of his parkinsonian features, he developed spiking temperatures (39-40.5°C) and his CPK increased slowly. Due to this, we provided a trial of levodopa/benzerazide up to 20mg/kg/day, which controlled his dystonic seizures and significantly improved his rigidity. A new LP was done and neuronal surface antibodies (CBA method), as well as onconeural antibodies were negative. Subsequently, both IgG and IgM antibodies against Mycoplasma pneumoniae were positive, as well as low serum titers of anti-GAD (1/64). Six weeks after onset he was discharged with a diagnosis of autoimmune/post-infectious basal ganglia encephalitis (BGE), already walking and speaking a few words. A week after discharge, he was completely asymptomatic and began weaning off levodopa. **Discussion:** BGE is a rare autoimmune or post-infectious encephalitis that affects mostly the caudate and putamen. Symptoms include severe Parkinsonism, dystonia and altered consciousness. A few causes have been reported, including direct invasion by M. pneumoniae and other respiratory viruses. This, however, has been challenged for the discovery of anti-DR2 antibodies against the basal ganglia, leading to an autoimmune etiology, perhaps with an infectious trigger. Our case of BGE showed significant improvement after immunotherapy and levodopa. **Final comments:** Due to the rarity of this condition, it is unlikely large studies will give definitive answers over which treatment course to follow. Nonetheless, our case demonstrates high dosages of levodopa could improve parkinsonism and cease dystonic 'seizures' and dysautonomia, while immunotherapy has not fully stopped the autoimmune process.

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Behavioral variant frontotemporal dementia and the DYSF gene: causal relationship or coincidence? A case report

Thamires Marx da Silva Santos[1], Paulo Caramelli[1], Leonardo Cruz de Souza[1], Elisa de Paula França Resende[1]

[1]Universidade Federal de Minas Gerais, Hospital das Clínicas, Belo Horizonte MG, Brazil.

Correspondence: Thamires Marx da Silva Santos; Email: marxthamires@gmail.com.

Case presentation: The patient is a 75-year-old woman, daughter of consanguineous parents, with a family history of muscular dystrophy and neurofibromatosis, but no reported familial cases of dementia. She was diagnosed with behavioral variant (bv) frontotemporal dementia (FTD) at the age of 65, presenting symptoms such as forgetfulness, visual hallucinations, lack of empathy, and unmotivated laughter. She did not present clinical features of motor neuron disease. Magnetic resonance imaging studies revealed significant frontotemporal atrophy, without significant white matter hyperintensities (Fazekas scale = 1). A genetic panel was performed, confirming the presence of the DYSF gene mutation in heterozygosity variant c.5979dup (p.Glu1994Argfs*3). Of note, the patient did not present other pathogenic mutations such as GRN, MAPT or TARDBP. **Discussion:** Current literature does not report an association between FTD and mutations in the dysferlin (DYSF) gene, which are associated with specific myopathies. However, there are reports linking this gene to Alzheimer's disease, indicating a possible role in neurodegeneration. **Final comments:** This clinical case raises the question of whether the presence of the DYSF gene mutation constitutes a mere coincidence or if there is a causal relationship with the manifestation of bvFTD. Previous study showing the accumulation of dysferlin intra-neuronal in the brain may suggest a pathological substrate for neurodegenerative disorders. Further investigation with other samples and additional studies may help clarify this relationship and its relevance in understanding the pathogenesis of FTD.

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Beneficial nucleosides reposition at TK2d: the first Brazilian cohort of treated patients

Cristiane Araujo Martins Moreno[1], Tatiana Ribeiro Fernandes[1], Gabriel Saboia de Araújo Torres[1], André Macedo Serafim da Silva[1], Mariana Hiromi Manoel Oku[1], Gabriella Corrêa Dousseau[1], Michelle Abdo Paiva[1], Clara Gontijo Camelo[1], Mariana Cunha Artilheiro[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Cristiane Araujo Martins Moreno; Email: cristianeammoreno@gmail.com.

Background: Thymidine kinase 2 (TK2) is the enzyme responsible for the phosphorylation of deoxyribonucleosides deoxythymidine and deoxycytidine to deoxythymidine and deoxycytidine monophosphates, respectively. Both are phosphorylated to generate the deoxynucleosides triphosphates needed for the replication of mtDNA. Biallelic pathogenic variants within the TK2 gene result in TK2 deficiency (TK2d) and lead to DNA maintenance defects and depletion of mitochondrial DNA. This condition typically presents with a progressive and threatening myopathy. A supplementation therapy using monophostate nucleosides to bypass the biochemical defect was developed and showed pronounced clinical benefits at open-label trials, enhancing the importance of an early diagnosis and early treatment to change the lethal natural history of the disease. Here we present the clinical findings and follow up of eight Brazilian patients receiving the nucleosides reposition through compassionate program. **Methods:** Eight patients followed at a large Terciary Center in Brazil were receiving supplementation therapy with deoxynucleosides (D'Thd and dCtd) in a compassionate program by the sponsor, offered orally at a dosage of 400mg/kg/day for each compound. **Results:** Patients were 20(±16) years old, two presented infantile-onset and the others childhood-onset. All of them were severely affected and attended to the requirements for a compassionate program. The protocol was approved by ANVISA in addition to the Clinical Hospital at FM/USP. One patient received the treatment for 15 months and presented an increase of 28 points at Chop-Intend, increased weight, descreased time-on ventilation and regained the ability to swallow. Two patients were on treatment for 6 months, both presented a significantly increased at MFM-32. Five adult patients with severe presentation of the disease were receiving the reposition for one month. Despite not presenting improvement at motor scales, all of them referred subjective improvements. They were more active and feeling less fatigued. **Conclusion:** TK2d is a progressive disease with high mortality rates. However, due to a potential treatment capable of changing the natural history of the diseas, the early recognition of this disease is urgent. We demonstrasted positive from the first patients receiving the treatment in Brazil.

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Beyond cataplexy: which parameters could help differentiate narcolepsy type 1 from narcolepsy type 2?

Christianne Martins Correa da Silva Bahia[1,2], Luis Cristovao Porto[1], Victoria Mendes Oliveira[1], Gilson

Costa dos Santos Junior[1], Danielle Angst Secco[1], Romulo Vianna[1], Carlos Otavio Brandão[3], Emmanuel Mignot[4], Bruna de Freitas Dias[4], Soniza Vieira Alves Leon[2,5,6]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ,Brazil.
[2]Universidade Federal Fluminense, Niteroi RJ, Brazil.
[3]Neurolife, Rio de Janeiro RJ, Brazil.
[4]Standford University, California, USA.
[5]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ,Brazil.
[6]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ,Brazil.

Correspondence: Christianne Martins Correa da Silva Bahia; Email: christiannemartins@gmail.com.

Background: The latest edition of the International Classification of Sleep Disorders (ICSD-3) has redefined narcolepsy terminology, replacing the subdivisions " with" or "without" cataplexy to narcolepsy type 1 (NT1) and type 2 (NT2), based on hypocretin-1 levels in cerebrospinal fluid (CSF). However, this biomarker is not readily available in many countries, including Brazil, and the lumbar puncture required for its measurement may deter patients. Also, hypocretin could be low in other neurological disorders with damage to the blood-brain barrier. Cataplexy and a positive multiple sleep latency test (MSLT) are used to diagnose NT1 without hypocretin data. Nevertheless, these criteria have limitations, as both NT1 and NT2 patients may exhibit atypical or absent cataplexy, and most episodes are not witnessed by the specialist, leading to diagnostic challenges. **Objective:** To compare clinical and genetic variables between NT1 and NT2, focusing beyond cataplexy and hypocretin measurement. **Methods:** Narcolepsy was diagnosed according to ICSD-3 criteria. Classification into NT1 and NT2 were based on CSF hypocretin levels measured by radioimmunoassay. Clinical, neurophysiological, and genetic data were obtained. Univariate and multivariate analyses were performed to compare the two groups using Chi-Square or ANOVA with the aid of Epi Info® and Partial Least Square Discriminant Analysis (PLS-DA), Variance Importance of Projection (VIP score), cross-validation with R2 and Q2 values and Area Under the ROC Curve (AUROC) were generated on MetaboAnalyst 6.0. **Results:** A matrix table encompassing 73 variables and 33 patients (17 NT1 and 16 NT2) was generated. NT1 subjects exhibited higher Body Mass Index (BMI), Epworth Sleep Scale (ESS) scores and frequency of cataplexy, along with lower Sleep Latency (SL), Total Sleep Time (TST) and diagnostic delay compared to NT2 ($p<0.05$). Cataplexy was reported by 15 NT1 (88.3%) and three NT2 (18.7%) subjects. Detailed characterization allowed differentiation between typical and atypical cataplexy. No NT2 patients present typical cataplexy, but atypical cataplexy was present in 17.6% of NT1 cases. The most common features of atypical cataplexy included triggers by negative emotions.HLA-DQB1*06:02 had the highest ranking on the VIP Score. Shorter sleep latency, typical cataplexy, and higher BMI were also significant parameters in differentiating NT1 from NT2 in both univariate and multivariate analyses ($p<0.05$). **Conclusion:** No single test alone is perfect for distinguishing NT1 and NT2. While detailed anamnesis can aid in the interpretation of typical and atypical cataplexy, it remains subjective. Therefore, objective parameters beyond cataplexy, such as HLA-DQB1*06:02, shorter sleep latency, and higher BMI, are essential for a more accurate diagnosis of NT1 and NT2. This differentiation can be helpful since new drugs based on hypocretin function have been developed.

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Beyond myelopathy: neuromuscular manifestations of HTLV-1

Andre Vinicio Cruz Mesias[1], Maria Fernanda Hamamoto Leati[1], Camila Rodrigues Nepomuceno[1], Daniel Santos Uchoa[1], Gabriella Corrêa Dousseau[1], Daniel Thomás Pereira Lopes[1], Cristiane de Araújo Martins Moreno[1], Julian Letícia de Freitas[1], Maria Sheila Guimarães Rocha[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Maria Fernanda Hamamoto Leati; Email: mfleati@gmail.com.

Case presentation: Patient 1: A 68-year-old woman presented in 2008 with progressive lower limb weakness that progressed to wheelchair dependence in less than 3 years, accompanied by a 30 kg weight loss over 2 years. Motor strength examination revealed proximal (MRC grade 2) and distal (MRC grade 4) weakness in the lower limbs, along with proximal atrophy. In the upper limbs, proximal weakness (MRC grade 4) was observed. Osteotendinous reflexes were preserved, except for the abolished patellar reflex. Plantar cutaneous reflexes showed flexion. Laboratory tests indicated normal CPK and aldolase levels, but serology was positive for HTLV-1. Cerebrospinal fluid (CSF) analysis confirmed HTLV-1 positivity. Electromyography (EMG) demonstrated asymmetric proximal myopathy, more severe in the lower limbs, with early recruitment and small amplitude potentials. A muscle biopsy indicated inclusion body myositis, showing CD8+ cells and macrophages surrounding and invading non-necrotic fibers, rimmed vacuoles, mitochondrial alterations, and protein aggregates. Patient 2: A 59-year-old man reported reduced strength in his lower limbs, particularly the left leg, for several years without knowing the exact onset. Significant limitations became apparent after 2020, when he required unilateral support to walk. Motor strength examination revealed MRC grade 4 weakness in the right lower limb and MRC grade 3 in the left. Atrophy, fasciculations, spasticity in the lower limbs, and signs of pyramidal release were noted in all four limbs. He walked with a scissor gait, more severe on the left side, using unilateral support. HTLV serology was positive. MRI did not indicate myelopathy. Electroneuromyography (ENMG) met the criteria for diffuse lower motor neuron involvement (four segments) with active denervation and chronic reinnervation, classifying this case within the HTLV-ALS spectrum. **Discussion:** There are several clinical and radiological presentations of neuromuscular diseases related to HTLV-1, as seen in our cases. The most frequent complications are tropical spastic paraparesis/myelopathy (HAM/TSP) and adult T-cell leukemia/lymphoma (ATL). Some cases of HAM/TSP mimic ALS (Amyotrophic Lateral Sclerosis), potentially leading to diagnostic confusion, though differing mainly in autonomic and sensory symptoms in addition to the bulbar segment. Inflammatory myositis associated with HTLV-1 has few reported cases despite in vitro evidence of myotoxicity by the virus. So far, there are several case reports of patients with IBM and HTLV-I infection. In addition, we have polyneuropathy also composing other neuromuscular presentations described with HTLV. **Final comments:** Both patients in these cases exhibit rare complications related to chronic HTLV-1 infection, highlighting the importance of clinical reasoning for accurate investigation given the wide range of differential diagnoses.

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Bilateral asynchronous lateralized periodic discharge in a patient with intraparenchymal hemorrhage

Marcillio Holanda Bezerra[1], Carlos Alexandre Twardowschy[2]

[1]Universidade Federal do Maranhão, Hospital Universitário, São Luís MA, Brazil.
[2]Pontifícia Universidade Católica do Paraná, Curitiba PR, Brazil.

Correspondence: Marcillio Holanda Bezerra; Email: marcillio.holanda@hotmail.com.

Case presentation: A pregnant, 43 years old, 35 weeks of gestation, was admitted with pre-eclampsia and seizure. On physical examination: the patient presented scale coma Glasgow 14 (A4V4M6) and left hemiparesis. Brain MRI showed: Image with low signal on the SWI, T2 and FLAIR sequences with hypersignal foci on T1, without enhancement after the use of venous contrast in the lentiform nucleus up to the corona radiata on the right, measuring 2.2 x 2.1 x 2.0cm, corresponding to intraparenchymal hemorrhage. An EEG was performed, which showed asynchronous lateralized periodic discharges (LPD) starting in the right temporal region, followed a few milliseconds by LPD in the left temporal region (Bilateral Asynchronous LPD). Patient was discharged after the 12th day of hospitalization, GSG 15 and with left brachiofacial paresis. **Discussion:** One of the changes to the ACNS Standardized Critical Care EEG Terminology in the 2021 version was the addition of the term asynchronous bilateral LPD. Bilateral LPDs are less common than other rhythmic and periodic patterns. It is estimated to occur in less than 1% of ICU patients, but the incidence of bilateral asynchronous LPDs is not known. These are defined as a lateralized periodic pattern that consistently begins in one hemisphere and propagates to the other hemisphere more than 80% of the time. Unlike independent bilateral LPDs, which are associated with acute bilateral brain injury in more than 70% of cases and consequently tend to have a poor prognosis, bilateral asynchronous LPD is associated with unilateral brain injury. We highlight in the present case that the right hemisphere involved with the intraparenchymal hemorrhage was where the bilateral asynchronous LPDs consistently began. **Final comments:** Bilateral asynchronous LPD is a rare pattern, and more studies are needed on its prevalence, pathophysiology, etiologies and prognosis.

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Bilateral facial palsy secondary to dengue fever: a case report

Guilherme Rocha Cardoso[1], Gustavo Alves Cangussú[1], Gustavo de Souza Campos[1], Luídi Neves Nascimento[1], Victor Santos Vilares[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1], Emmanuel Santos Trindade[1], Priscila Santos Oliveira[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Érica Otoni Pereira Miranda; Email: ericaotoni15@gmail.com.

Case presentation: A 50 year old female patient was referred to the neurology outpatient clinic with complaints of facial palsy for 10 days. She reported hospitalization due to dengue hemorragic fever 30 days ago. Subsequently, she developed a bilateral facial palsy, with difficulty in facial mimicry movements and blinking. She also presented progressive cephalocaudal paresthesias, worsening in the perioral region right hemiface and limbs. There were no reports of headache, fever, or associated visual and auditory impairments. Upon physical examination, it was noticed a bilateral seventh cranial nerve impairment, with ptosis, generalized facial mimicry limitation, reduced strength and inability to completely close the eyelids. In view of the above, a clinical diagnosis of bilateral facial palsy secondary to dengue was made. The therapeutic approach was corticosteroid therapy with prednisone 40 mg/day, facial physiotherapy and ocular protection with ophtalmic lubricating solution and occlusion at night. **Discussion:** Dengue is an acute febrile illness with a systemic and dynamic nature, and it has a highly variable severity spectrum. It is endemic in Brazil, transmitted by the Aedes aegypti mosquitoes, mainly in hot and rainy months. Besides its classic manifestations such as fever, myalgia and circulatory shock in severe cases, dengue can also present with neurologic involvement. The literature describes cases of encephalitis, meningitis, Reye's syndrome, and polyradiculoneuritis, which includes Guillain-Barré syndrome (GBS) and

its variants. This report presents a patient with a typical pattern of polyradiculopathy, characterized by bilateral facial motor impairment, which may indicate bilateral seventh cranial nerve involvement. Additionally, there was sensory impairment, with diffuse paresthesias throughout the body. These findings point to the diagnosis of a GBS variant, characterized exclusively by bilateral facial paralysis and paresthesias. GBS can occur early or late in the disease course and is likely mediated by an immune mechanism in which antibodies (due to cross-reaction) are directed against myelin and/or axon, resulting in increased latency for the transmission of neuronal signals to the innervated regions and a reduced compound muscle action potential, indicating a decrease and/or blockage in impulse conduction velocity. Furthermore, GBS secondary to arbovirus infection presents a faster clinical course compared to GBS from other causes. This is a rare post-infectious manifestation in dengue cases, with few cases described in the literature. **Final comments:** Therefore, it is suggestive that the bilateral facial palsy in this patient is due to GBS variant secondary to arbovirus infection. Despite being a low-incidence clinical presentation, it presents significant implications that must be carefully analyzed for diagnosis, as dengue has various spectrums of nervous system involvement.

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Bilateral fusiform internal carotid artery aneurysms: case report and challenges in the approach

Maria Clara Motta Barbosa Valente[1], Filipe Brito Ferraz da Silveira[1], Camila Lopes Figueiredo[1], Rafael Chaves Claudino de Queiroga[1], Letícia Machado Dumont[1], Cícero Rodrigues Veloso[1], Lorenna Castro Di Filice[1], Tomaz Lima Moreira[1], Vivian Dias Baptista Gagliardi[1], Rubens José Gagliardi[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Clara Motta Barbosa Valente; Email: mariaclarambvalente@gmail.com.

Case presentation: A 55-year-old male patient with a history of hypertension and smoking presented with an 8-month history of progressive dysphagia and weight loss. He was initially investigated for gastroesophageal reflux disease. Two months prior to hospital admission, he experienced an episode suggestive of a seizure (without further clinical details). A cranial and cervical CT scan revealed bilateral intracranial expansile lesions of undetermined etiology. He was admitted to the emergency department with left hemiparesis and decreased level of consciousness (Glasgow Coma Scale: 9). Neurological examination revealed left hemiparesis, hyperreflexia and bilateral extensor plantar responses. CT angiography showed fusiform dilatation of the right internal carotid artery (26 x 16.8 mm) and irregularities and dilatation of the left internal carotid artery (maximum diameter of 9.3 mm). There was also displacement of the left posterior cerebral artery, superior cerebellar artery, and basilar artery, suggesting an adjacent mass. Angiography confirmed bilateral aneurysms. The patient's level of consciousness deteriorated and he required endotracheal intubation. A repeat CT scan showed brainstem displacement due to aneurysm compression. A joint discussion with neurosurgery considered endovascular or bypass approaches, but the patient developed loss of brainstem reflexes and brain death before the procedure. **Discussion:** Bilateral fusiform internal carotid artery aneurysms (FCIAs) pose a therapeutic challenge due to their extensive size and the inability to perform techniques such as clipping due to the absence of a neck. We present a case of bilateral FCIA with brainstem compression. In this case, the risks of surgical and non-surgical approaches were high, as the latter would lead to an unfavorable outcome due to compression of vital structures. Two options were considered: open bypass or stress test with occlusion of the right internal carotid artery and assessment of collateral circulation, followed by endovascular intervention for aneurysm exclusion. However, there is no gold standard treatment technique due to the complexity and individuality of each case. **Final comments:** Despite the unfavorable outcome, this case highlights the importance of discussing the approach technique and the therapeutic challenges of FCIAs, especially in the presence of adjacent structure compression. The rarity of these cases limits the evidence for the best treatment, and decision-making regarding the timing of intervention is crucial due to the risk of rupture with aneurysm expansion and adjacent structure compression.

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Bilateral optic neuritis and probable MOG-associated demyelinating syndrome in pediatric patient: a case report

Danielli Regina da Silva[1], Renata Cristina Franzon Bonatti[1], Airton Sena Oliveira Sales[1], Laisy Bárbara Souza Silva[1], Guilherme Victor Rocha Pereira[1], Marcos Luiz Belotti de Souza[1], Cairo Cruvinel[1], Lucas Santos Andrade[1], Lorenzo Pereira de Almeida Melo Montes[1], Alex Eduardo da Silva[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Danielli Regina da Silva; Email: d202011051@uftm.edu.br.

Case presentation: Male patient, 11 years old, mixed race, presenting with progressive bilateral decreased visual acuity for three days and weight loss of eight kilograms over one week. Denies fever, headache, nausea, vomiting, weakness, speech changes, confusion, ocular trauma, recent infections, or prior comorbidities. Diagnosed with myopia one year ago, corrected with visual aids. Neurological examination revealed bilateral papilledema, midriasis, and absent pupillary light reflexes, without other abnormalities. Cranial computed tomography showed mild elongation and tortuosity of the optic nerves. However, magnetic resonance imaging demonstrated symmetric T2 and diffusion hyperintense areas in the posterior orbital and anterior intracranial portions of the optic nerves and optic chiasm, with anomalous post-contrast enhancement in the proximal intracranial portions of the optic nerves and faint hyperintensity in the Flair sequence in the right thalamic pulvinar. Serological tests were negative for antinuclear antibodies, rheumatoid factor, HLA-B-27 antigen, as well as Cytomegalovirus, VDRL, Treponema pallidum, toxoplasmosis, HBSAg, anti-HBS, HIV, and Herpes simplex virus. Cerebrospinal fluid analysis was normal, with negative results for Cryptococcus neoformans antigen, IgG antibodies against aquaporin 4 (AQP4-IgG), and other serologies. Provisional diagnosis of bilateral optic neuritis and probable MOG-associated central nervous system demyelinating syndrome (MOGAD). Treated with five days of methylprednisolone pulse therapy, resulting in gradual improvement of bilateral visual acuity. Oral corticosteroid initiated for outpatient follow-up. **Discussion:** Optic neuritis in childhood, a rare and severe autoimmune inflammatory demyelinating disorder of the central nervous system, occurs in only 5 to 10% of cases. In cases negative for AQP4-IgG antibodies, like the one described, the presence of antibodies against myelin oligodendrocyte glycoprotein (MOG-IgG) is the leading etiological hypothesis, particularly with bilateral anterior involvement while sparing the optic chiasm. However, due to limitations in resources within the Unified Health System, specific confirmatory testing could not be conducted. Initial treatment with methylprednisolone pulse therapy followed by oral corticosteroids is recommended, as demonstrated in this case. **Final comments:** This case underscores the importance of a meticulous approach to diagnosing neurological disorders in children, including detailed clinical history, thorough neurological examination, and imaging studies. Diagnostic confirmation through specific tests such as AQP4-IgG and MOG-IgG is crucial for guiding appropriate and safe treatment in this age group, where optic neuritis presents unique challenges compared to adults.

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Bilateral petrosal apex cephalocele and empty sella turcica - case report

Renan Iegoroff[1], Alvaro Marcelo Huchani Huanca[1]

[1]Afya, IPEMED, São Paulo SP, Brazil.

Correspondence: Renan Iegoroff; Email: renan_iego@outlook.com.

Case presentation: Woman, 42, Caucasian, early childhood education teacher. Has been working with neurology for migraines with aura for approximately ten years, with partial pain control and irregular prophylactic treatment, and for ten months resuming treatment with Topiramate 25mg/day. No history of metabolic endocrine changes, absence of galactorrhea and vision changes, presenting grade 1 obesity as a comorbidity and a previous history of two episodes of pulmonary thromboembolism of unclear cause, on neurological examination, without evidence of focal signs and cephaliatric examination only with finding of painful palpation of the greater occipital nerve on the left, a magnetic resonance imaging examination was performed without contrast (23.12.23) which revealed an empty sella turcica, when carrying out additional tests for differential diagnosis. This was evidenced on the Sela Turcica Magnetic Resonance Imaging with Contrasts (21.01.24) enlargement of the Bilateral Cavum of Meckel at the expense of cystic formations with characteristics of bilateral petrous apex cephalocele; in laboratory tests (01/24/24) resting prolactin, IGF-1, cortisol, aldosterone, fasting insulin, TSH, T4L, FSH were within normal limits. **Discussion:** Cephalocele of the petrous apex is a cystic lesion resulting from the herniation of Meckel's Cavum, and the bilateral condition is a rare phenomenon, described in the literature until 2022 around 21 cases, it is believed that this pathology occurs due to increased intracranial pressure with herniation of the meninges, and must be differentiated by its characteristics from cholesteatomas, mucocele and paragangliomas, which in this case were discarded due to the well-defined characteristics of the image. When reviewing the 21 bilateral cases, 80% of the cases were in women, around 55% of the cases bilateral lesions presented with an empty sella turcica, 32% presented with headache and 0.01% (2 cases) underwent surgical interventions, and always without changes in endocrinological functionality. Management, therefore, is mostly conservative, with management of the symptoms associated with the condition, in the cases reported surgically, one of them had surgery due to the initial diagnosis of cholestoma with lateral gaze paralysis, and the other case had surgery due to bilateral cranial changes with cerebrospinal fluid leaks and recurrent meningitis. **Final comments:** The rarity of the described case and the growing clinical association of presentation with empty sella turcica are evident in the literature, making it necessary for the neurologist to understand the differential diagnosis and recognition of the lesional aspect due to the management being mostly conservative.

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Bilateral profound sensorineural hearing loss due to cerebrovascular event

Hélio Aquaroni Farão Gomes[1], Caroline de Barros Viegas[1], Ana Beatriz Marangoni Baston[1], Pedro Machry Pozzobon[1], Gabriel Pinheiro Modolo[1], Renan Macionil Cotrim[1], João Marcio Borgue de Oliveira[1], Luiz Felipe Moraes Schwerz Bonadiman Blanco[1], Itamar Meireles Andrade Santos[1], Dominick Esthephanny Monteiro Silva[1]

[1]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Hélio Aquaroni Farão Gomes; Email: heliofarao@hotmail.com.

Case presentation: A 59-year-old patient with a history of diabetes, hypertension, and smoking presented with a sudden onset of vertigo, nausea, and hearing loss in the left ear. The following day, he noticed facial paralysis on the same side and was referred to the hospital 20 hours after the onset of symptoms. Upon admission, the patient complained of horizontal diplopia and hearing loss. Neurological examination revealed dysarthria, altered balance in orthostasis with a tendency to fall backwards, left lateral rectus paresis, and peripheral facial paresis on the left. Additionally, inexhaustible multidirectional nystagmus, bilateral sensorineural hypoacusis, hypoesthesia on the left side, and dyskinesia between the trunk and limbs were observed. A CT scan of the brain showed hypodensity in the bulbopontine transition. Arterial angiography revealed occlusion of the basilar artery. The patient was admitted to the stroke unit. Further investigation with magnetic resonance angiography showed hypoplasia of the right vertebral artery, hypersignal in the left vertebral artery, and occlusion of the basilar artery with reperfusion by communicating arteries near the intracranial source. Brain MRI revealed hypersignal in the pons, middle cerebellar peduncle, dentate nucleus of the left cerebellum, and the right cerebellar hemisphere. This affected region overlaps with the facial and vestibulocochlear nerve nuclei on the left. A transcranial Doppler was also performed, showing subocclusion of the basilar artery from its origin to the middle third. Following the investigation, prophylaxis was started with 300 mg of aspirin (AAS) and 75 mg of clopidogrel for 3 months. Evaluation by the otorhinolaryngology department showed continued anacusis on the left and hypoacusis on the right. Audiometry revealed bilateral profound sensorineural hearing loss, with a voice detection threshold of 80 decibels on the right and over 100 decibels on the left, indicating greater impairment on the left. Electro-oculography was compatible with mixed cochleovestibulopathy. **Discussion:** Posterior circulation strokes can manifest with non-specific symptoms, causing a delay in diagnosis compared to anterior circulation cases. Sensorineural deafness has been described in stroke cases, especially in those involving occlusion of the anteroinferior cerebellar artery, but it usually occurs unilaterally. **Final comments:** In this case, due to the occlusion at the origin of the basilar artery with collateral filling at its uppermost portion, the anteroinferior cerebellar artery, which originates from the initial part of the basilar artery, was bilaterally impaired. Consequently, the bilateral labyrinthine artery was compromised by the pattern of the lesion. Brainstem lesions with a better prognosis are typically microvascular and usually have more unilateral involvement, unlike this case, which involved a large vessel lesion.

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Bilateral schwannomas: a case of schwannomatosis

Daniel Mariani Favalessa[1], Ivo Gabriel Ribeiro Carvalho[1], Melissa dos Santos Matos[1], Guilherme Valiate Picoli[1], Guilherme Marques Nunes Mamede[1], Renann Nunes Pirola[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Daniel Mariani Favalessa; Email: dmfavalessa@gmail.com.

Case presentation: A 55-year-old male reported that at 27 years old, he began experiencing paresthesia followed by loss of strength in his right hand, progressing from distal to proximal, and subsequently involving the rest of the upper limb. After three months, similar symptoms occurred in the left upper limb. At 52 years old, he noticed the appearance of two bilateral supraclavicular cervical nodules. He has no complaints regarding the lower limbs, bulbar, vestibular, or ocular symptoms. He has a history of epilepsy since age six, with seizures starting with facial paresthesia, progressing to impaired consciousness and disjointed speech, without significant postictal symptoms. He denies a similar family history, skin spots, or other neurological diseases. On examination, bilateral interscalene tumors were observed, with no café-au-lait spots, interosseous muscle atrophy more pronounced on the left side, grade IV strength in the upper limbs, except for left shoulder abduction (grade II). Global areflexia, preserved superficial sensation, and distal hypopallesthesia in the lower limbs and left hand were noted. Electroneuromyography indicated severe demyelinating sensorimotor polyneuropathy with signs of activity. A biopsy of the left tumor indicated schwannoma. **Discussion:** Schwannomatosis is a term used by neurologists to describe conditions involving multiple schwannomas, excluding NF1 and NF2. In 2022, its classification was updated to include a spectrum of schwannoma predisposition syndromes, such as those involving SMARCB1 and LZTR1 genes. Among these, the LZTR1 gene has the most significant diagnostic overlap with NF2, making differentiation challenging. However, the absence of bilateral vestibular schwannomas is particularly reliable in ruling out NF2 in patients over 30 years old. In this case, the patient presents with severe demyelinating sensorimotor polyneuropathy associated with bilateral peripheral schwannomas and a history of epileptic seizures since childhood, without skin, eye, or cranial nerve involvement. The biopsy revealed lesions consistent with non-intradermal schwannomas, confirming unspecified schwannomatosis, with genetic testing necessary for specific diagnosis. However, genetic testing is still difficult to access and does not determine prognosis or inform medical treatment, making clinical diagnosis often sufficient. Treatment is mainly symptomatic, focusing on pain management. **Final comments:** Finally, the diagnostic complexity of schwannomatosis and the difficulty in distinguishing it, especially from NF2, are highlighted. This requires a comprehensive approach combining clinical data, laboratory tests, imaging studies, and, when possible, genetic testing. Management is symptomatic, with the primary focus on pain relief.

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Bilateral third nerve palsy as a manifestation of pituitary apoplexy: a case report

Caroline Zapelini[1], Bruna Beppler[1], Adria Rodrigues da Silva[1], Lia Maki Hatisuka Imai[1], Luis Sidonio Teixeira da Silva[1], Lucio Baena de Melo[1], Gabriel Rodrigues Espelho Rossi[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Caroline Zapelini; Email: zapelinicarol@gmail.com.

Case presentation: A 73-year-old female patient presented with frontal headache, fever, myalgia, and nausea. After six days of progression, the patient was admitted to a tertiary hospital due to bilateral third nerve palsy, complete on the right side and incomplete on the left side. There was no alteration in the level of consciousness or signs of meningeal irritation. Laboratory tests showed mild leukopenia, no thrombocytopenia, normal coagulation profile, and no renal or hepatic dysfunction. Cerebrospinal fluid analysis revealed 432 cells with predominance of lymphocytes, protein level of 141, and glucose level of 81. Bacterial cultures were negative. Empirical acyclovir treatment was initiated and later discontinued after negative herpes PCR. Dengue fever IgM antibodies were detected in the blood. Brain MRI showed an enlarged hypophysis with suprasellar extension, heterogeneous appearance, and contrast enhancement, suggestive of a pituitary macroadenoma with hemorrhagic degeneration. The dimensions were approximately 2.2 x 1.7 x 1.4 cm (estimated volume of 2.74 cm^3). Hormonal tests confirmed hypopituitarism. The patient was diagnosed with pituitary apoplexy and started on dexamethasone, leading to complete resolution of bilateral III cranial nerve palsy in a few days. Follow-up MRI showed reduction of the adenoma, and the patient remained stable without adrenal crisis. She was discharged with outpatient follow-up. **Discussion:** Bilateral third nerve palsy is uncommon and may be associated with lesions in the nucleus or cavernous sinus, such as tumors, hemorrhages, aneurysms, and other etiologies. In the described case, where ophthalmoplegia is due to pituitary apoplexy (PA), there is a sudden increase in content in the sella turcica, raising intrasellar pressure and leading to compression of adjacent structures in the cavernous sinus, such as the oculomotor nerve. Bilateral third nerve palsy in PA is rare. PA is an endocrine emergency resulting from hemorrhage or infarction in a pituitary tumor and must be promptly recognized to avoid possible complications such as acute adrenal insufficiency. In the reported case, besides the atypical presentation, the patient had an ongoing dengue infection, with cerebrospinal fluid (CSF) changes suggesting a differential diagnosis of viral meningoencephalitis. PA usually does not affect CSF, although aseptic meningitis has been described in some cases. Dengue fever has been described as a precipitating factor for PA in patients with pituitary adenoma and has been linked to coagulation abnormalities. Although the patient did not show laboratory abnormalities such as thrombocytopenia, elevated INR, and TTPA, the infectious condition may have contributed to the case's progression. **Final comments:** The case illustrates a rare presentation of AP, with bilateral involvement of the oculomotor nerve, associated with dengue fever

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Bilateral vestibulopathy due to aminoglycoside otoxicity in patients with chronic kidney disease: a series of cases

Thaís Laura Almada da Fonseca[1], Isabelly Arruda Cardoso Slavec[1], Yasmin Coelho Patrial[1], Enã Rezende Bispo Do Nascimento[1], Aline Mizuta Kozoroski Kanashiro[1]

[1]Hospital Regional de Mato Grosso do Sul, Campo Grande MS, Brazil.

Correspondence: Thaís Laura Almada da Fonseca; Email: thaisalmada123@gmail.com.

Bilateral vestibulopathy (BVP) is an uncommon disease, with an incidence of approximately 28/100,000 cases per habitant. In around 50% of cases BPV is idiopathic, however it can be secondary to ototoxicity, bilateral Ménière's disease, bilateral infectious/inflammatory vestibular diseases; autoimmune, neoplastic, genetic and other causes. Despite the well-established diagnostic criteria by the Barany society in 2017, recognition of this condition is still not achieved by many doctors in clinical practice. This is the report of a series of 5 cases of BPV treated in 2023, in an outpatient clinic specialized in vestibular disorders. All patients had a previous diagnosis of chronic kidney disease (CKD), among them, 4 were on renal replacement therapy. There was history of previous use of aminoglycoside (gentamicin) in a dose adjusted for renal function, within one month of the onset of symptoms. The otoneurological examination was altered in all 5 cases, with pathological vestibuloocular reflex (VOR) on both sides and imbalance, with fall during the examination on an unstable surface and substantial worsening when closing the eyes. The main symptoms are those of chronic vestibulopathy and are generally reported by the patient as a non-specific imbalance. However, in most cases the patient is not able to spontaneously report the complaints characteristic of this disease. After a guided anamnesis, it is possible to identify the presence of oscillopsia, dizziness and imbalance during gait and head movements, which typically improve when in a static position with

the head still, and worsen in dark environments or on uneven surfaces. The diagnosis is based on the typical clinical history and the presence of bilateral altered VOR, which can be acessed through complementary exams such as the video head impulse test (v-HIT) and caloric test. This case series is a warning regarding the use of aminoglycosides as the first line of infectious treatment in patients with severe renal dysfunction. Furthermore, it is important to highlight the need to identify BPV, given the functional impact on patients' lives.

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Bilateral wrist drop revealing symmetric polyradiculoneuropathy and optic neuritis secondary to dengue infection successfully treated with human immunoglobulin - a case report

Matheus Luís Braga Munareto[1], Matheus Alves da Silva[1], Alex Araújo Fernandes[1], Amanda Miranda de Mendonça[1], Ana Luísa Faria Donadon[1], Rafael Bragança Rodrigues Matias[1], Eduardo Mesquita de Souza[1], Karlla Danielle Ferreira Lima[1], Alex Machado Baeta[1], Bruno Shigueo Yonekura Inada[1]

[1]Beneficência Portuguesa, São Paulo SP, Brazil.

Correspondence: Matheus Luís Braga Munareto; Email: matheusbragamunareto@gmail.com.

Case presentation: A 67-year-old male patient with prediabetes presented with a history of fatigue and subfebrile state associated with subacute, bilateral and symmetric wrist drop. He also developed blurred vision. He tested positive for dengue with NS1 exam. His examination showed symmetric strength loss, more evident in wrist and finger extension and thumb abduction. There was no synkinetic wrist extension, his reflexes were diminished in the arm and he had no sensory loss. There was proximal loss of strength in the elbow extension bilaterally. He had a decreased visual acuity of 20/160 in his left eye associated with afferent pupillary defect in this side. In view of these findings, magnetic resonance imaging of the limbs and orbits were performed. His arms MRI demonstrated denervation pattern of the left posterior interosseous nerve (posterior branch of the radial nerve), symmetrical thickening and hyperintensities of the roots and trunks of the brachial plexus. His orbital MRI was suggestive of anterior left optic neuritis. His electroneuromyography showed chronic and bilateral (symmetric) preganglionic motor impairment in the cervical region, of a moderate degree at the level of C7 and a marked degree at the level of C8/T1, with signs of acute denervation, compatible with multiradicular involvement. His CSF was unremarkable. The testing for DENV in the cerebrospinal fluid was unavailable. Due to the important strength and visual impairment, we started immunoglobulin for five days, with complete clinical improvement. **Discussion:** Dengue fever is the most prevalent arbovirus globally. It is an arthropod-borne viral disease caused by the four dengue virus serotypes (DENV 1, 2, 3, 4). DENV infection can be categorized into three stages: the febrile phase, critical phase, and recovery phase. Not all infection cases experience critical phases. Although dengue usually presents as a febrile illness, it can present with neurological symptoms. Besides the high prevalence of dengue infection, the neurological manifestations of DENV are rare, but may affect the central and the peripheral nervous system. The pathogenesis of dengue-associated neurological manifestations is thought to be immune-mediated, and there is some evidence in vitro for neurotropism. Encephalitis is one of the most common neurological manifestations of DENV infection. Optic neuritis and polyradiculoneuropathy are rare clinical manifestations. There is no specific antiviral treatment for dengue infection but corticosteroids and immunoglobulin may be an option for severe neurological manifestations. There is a lack of evidence to guide the treatment in these cases, due to the infrequency of neurological manifestations. **Final comments:** We present a case of a rare manifestation of a very common arbovirus disease. The case highlight the importance of correct follow up of these patients to manage the complications, including the unusual ones.

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Biopsy diagnosis of fungal infections affecting the central nervous system

Isabella Queiroz Barbosa[1], Évely Almeida Souza[1], Gabriel Lédo Pereira de Oliveira[1]

[1]Faculdades Santo Agostinho de Vitória da Conquista, Vitória da Conquista BA, Brazil.

Correspondence: Isabella Queiroz Barbosa; Email: qbisabella@hotmail.com.

Background: Fungal infections affecting the central nervous system (CNS) represent a significant challenge in clinical practice due to their complexity and diversity of presentation. These infections generally affect immunocompromised patients. Early and accurate diagnosis is fundamental to defining treatment and improving the patient's prognosis. Analysis of cerebrospinal fluid (CSF) is the method of first choice when there is a suspicion of fungal infection in the CNS, mainly because it is not very invasive and has high sensitivity. However, in more challenging cases, or where CSF analysis is inconclusive or cannot be performed, tissue biopsy is a viable alternative. **Objective:** To analyze data on cases of CNS fungal infection diagnosed by tissue biopsy. The evaluation will be carried out with data from a health service responsible for the pathology service of a local referral hospital, in the years 2022 to 2024. **Methods:** A descriptive analysis of biopsy results was carried out in order to draw up a diagnostic profile for fungal infections that affect the CNS using this method. These tests were provided by a pathology laboratory and comprise all the histological analyses from the neurosurgery department of a local referral hospital. The inclusion criteria were cases in which the patients had a suspected fungal infection of the CNS or in which the infectious diagnosis was incidental. **Results:** Between 2022 and 2024, the laboratory received 139 requests for analysis of neurological biopsies from the hospital. Of this total, around 4% were diagnosed with fungal infections, totaling 5 cases, 2 of which were cryptococci, and of these, 100% had another pathology as their suspected diagnosis. The following prevalence rates were also found: 35% of Gliomas (including glioblastomas), 20% of Meningiomas, 5% of carcinoma metastases, as well as other varied diagnoses such as Medulloblastoma, Hemangioblastoma, Ependymoma and Schwannoma. This distribution corroborates current literature, which indicates neurocryptococcosis as the most common fungal infection in the CNS, with an annual global incidence of 3.9 cases per 100,000 inhabitants. **Conclusion:** Fungal infections in the CNS are rare, but represent a significant diagnostic challenge. Tissue biopsy analysis, although less frequently used than CSF analysis, has proven to be a relevant tool in cases where the diagnosis is unclear or the CSF is inconclusive. This study highlights the need to consider fungal infections, such as neurocryptococcosis, in the differential diagnosis of CNS lesions, especially in immunocompromised patients or when symptoms do not correspond to more common pathologies. Accurate identification of these infections is vital for appropriate treatment and improving the patients prognosis.

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Biopsy-proven myosistis and myocarditis due to immune checkpoint inhibitor treatment

Lucas Bruno Rezende[1], Bruna Moreira de Souza Proença[1], Beatriz Gioppo Betini[1], Gustavo Ramos de Lima[1], Augusto Bragança Reis Rosa[1], Ana Carolina Zetehaku[1], Carolina Novaes Perniquelli[1], Antônio Francisco de Araújo Netto[1], Mariana Cavalcanti Costa[1], Rafael Paternò Castello Dias Carneiro[1]

[1]Hospital Nove de Julho, São Paulo SP, Brazil.

Correspondence: Lucas Bruno Rezende; Email: lucasbrunorezende@hotmail.com.

An 83-year-old woman with locally invasive non-metastatic lung adenocarcinoma was admitted to the hospital due to mialgia, weakness and blurry vision. Two weeks earlier, she had received her second cycle of treatment with nivolumab and ipilimumab, anti-PD1 and anti-CTLA-4 immune checkpoint inhibitors respectively. Bilateral semiptosis, multidirectional ophtalmoplegia and proximal muscle weakness grade III predominantly affecting the limb girdles were noted on neurological examination. Bulbar function, cranial nerves and deep tendon reflexes were normal. CK was elevated with levels of 10.970 U/L. Brain MRI was unremarkable. Thigh MRI suggested myositis and disclosed patchy bilateral areas of edema and gadolinium enhancement affecting the quadriceps femoris, adductors, hamstring and gluteal muscles. Shortly after admission, hemodynamic instability due to a 3rd grade atrioventricular block ensued. Cardiologic work-up showcased high troponin levels, but no structural cardiomyopathy on transthoracic echocardiography. Cardiac catheterization was pursued and an 80% stenosis in the anterior descendant artery was treated with a drug-eluting stent, but bradyarrhytmia did not resolve and a transvenous pacemaker was put in place. Neurophysiological studies were unavailable at the time and serum anti-acethylcoline receptor antibodies were negative. Since a concomitant neuromuscular junction disorder could not be completely ruled out, prompt treatment with both pulse-dose steroids and intravenous immunoglobulin was then instituted whilst preparations for muscle biopsy were being undertaken. Left quadriceps femoris biopsy confirmed the diagnosis of immune checkpoint inhibitor-related myositis with findings of multifocal muscle necrosis and mild inflammatory infiltrate constituted mainly by macrophages and CD8-positive lymphocytes. The patient eventually died due to infectious complications of her critical illness.

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Bisegmental angioplasty of the right vertebral artery: case report

Isabela de Carvalho Florencio[1], Rodrigo Ibañez Tiago[2], Lucas Blanco Insaurralde da Luz Silva[2], Amanda Boutrik[2], Renata Gratão Rezende[1], Guilherme Augusto de Oliveira Soares[1], Carolina Cândida Vaz[1], Rodolfo Antonio Corona[1], Mateus Magalhães Barbosa[1], Felipe Inácio Ferreira da Silva[3]

[1]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.
[2]Universidade Federal de Mato Grosso do Sul, Faculdade de Medicina, Campo Grande MS, Brazil.
[3]Hospital do Coração do Mato Grosso do Sul, Campo Grande MS, Brazil.

Correspondence: Isabela de Carvalho Florencio; Email: isabelacflorencio@hotmail.com.

Case presentation: A 56-year-old male with a history of posterior circulation ischemic stroke one month ago, TOAST atherothrombotic, presented with sequelae of left hemifacial paresis and abducens nerve palsy in the left eye. The patient was on acetylsalicylic acid and clopidogrel. He was admitted to the emergency department due to recurrent left hemibody paresthesia, worsening over the past two days. Upon admission, clopidogrel was replaced with ticagrelor. Angiographic study via catheter of the aortic arch, subclavian, vertebral, and carotid arteries revealed an occluded dominant left vertebral artery and sub occlusion in the distal V4 segment,

affecting the posterior inferior cerebellar artery, as well as sub occlusion at the vertebrobasilar junction. Despite the medication switch and prophylactic heparin, the patient continued to experience recurrent focal symptoms of posterior circulation five days post-switch, leading to the indication for endovascular treatment. The right vertebral artery at the V4 segment was treated with percutaneous transluminal angioplasty using an Appolo stent (2.5 x 8 mm - MICROPORT), and angioplasty with a balloon (Treck 1.5 x 6 mm - ABBOT) was performed at the vertebrobasilar junction. Final control demonstrated total regression of the stenoses with the Appolo stent well-adhered to the vessel lumen and no signs of direct procedure-related complications. The patient was discharged while continuing acetylsalicylic acid and ticagrelor. A follow-up cerebral angiography performed six months after the procedure confirmed the stent was well-adhered with no signs of stenosis. **Discussion:** Vertebral artery stenosis is responsible for approximately 20% of ischemic strokes in the posterior circulation, potentially resulting in devastating neurological deficits. This ischemia is related to hemodynamic changes in many patients, due to proximal vertebral artery obstruction. For symptomatic patients who do not respond to dual antiplatelet therapy, endovascular therapy is beneficial, leading to lower rates of symptom recurrence and improved survival. It is known that morphological factors of vertebral artery lesions can affect the outcome of angioplasty therapy. Therefore, even with a promising result, it is necessary to maintain patient follow-up due to the possibility of restenosis, particularly at the vertebrobasilar junction. **Final comments:** We report a case of a patient with previous complete occlusion of the left vertebral artery who had a stroke and transient ischemic attacks while on antiplatelet agents and atorvastatin due to critical stenoses of the right vertebral artery. Angioplasty with stent placement resulted in clinical stabilization and the absence of new ischemic events. This case underscores the importance of endovascular intervention in patients with complex vertebral stenoses and reinforces the need for multidisciplinary management to optimize outcomes.

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Bladder pattern profile of children with Zika virus congenital syndrome

Larissa Coutinho de Lucena[1], Vitor dos Santos Rodrigues[2] , Jhulia Thaysa Macena da Costa[1], Amanda Georgia Diniz de Campos[3], Marília da Costa Paiva[1], Amanda da Silva Santos[1], Milena Thaisa Silva de Lima[2], Fernanda Gomes Mourão[1], Gabriel Alves Mendes Vasiljevic[1], Lilian Lira Lisboa[1]

[1]Instituto Santos Dumont, Macaíba RN, Brazil.
[2]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[3]Centro Maurício de Nassau, João Pessoa PB, Brazil.

Correspondence: Larissa Coutinho de Lucena; Email: larissa.lucena@isd.org.br.

Background: Congenital Zika virus syndrome (CZS) is characterized by a range of alterations in the anatomy and physiology of the central nervous system (CNS). These changes reverberate through all other body structures as well as neuropsychomotor development. In consensus with the evidence in the literature, a correlation is described between bladder dysfunction and motor functionality, which influences the production capacity in the urinary tract. **Objective:** To characterize the urinary profile of children with urinary sequelae caused by CZS. **Methods:** This is a cross-sectional study, carried out between 2023 and 2024, approved under CAAE 51865521.5.0000.0129 and financed by the CNPq/MCTI/FNDCT-18/202, UNIVERSAL call (403498/2021-0), carried out in patients diagnosed with CZS using the evaluation protocol recommended by the Brazilian Ministry of Health, which included Immunoglobulin G and Immunoglobulin M serological tests. The patients were evaluated using cystometry in the urodynamic study to assess the following variables: detrusor pressure, hyperactivity and maximum cystometric capacity; renal ultrasound examination in search of dimensions, echo textures and contour; and micturition and bowel movement diaries. **Results:** Ten children were enrolled, being eight (80%) female and two (20%) male, with a median age of 7.2 years. Bladder function showed a mean detrusor pressure of 23.3 cmH2O, 95% CI [16.93, 29.67]; mean expected cystometric capacity of 252 ml, 95% CI [240.92, 263.08]; and mean maximum cystometric capacity of 58.7 ml, 95% CI [33.98, 83.42]. It was also noted that 70% of the children had 20% or less of the maximum expected capacity in residual volume. All of the upper urinary tract assessments made by ultrasound showed normal dimensions, echotexture and contour. Urination was not triggered. **Conclusion:** Bladder alterations caused by CZS, over the long term, are indicative of the possible triggering of upper urinary tract lesions as a result of the high pressures generated by these alterations. The findings also suggest the need for interventions that promote an increase in maximum cystometric capacity in accordance with the expected capacity. More extensive and in-depth studies on the subject need to be carried out so that neurogenic bladder and bowel can be monitored and treated, even with low-cost resources such as transcutaneous electrostimulation, with the aim of minimizing future deleterious effects on the upper urinary tract that could limit the rehabilitation and quality of life of these patients.

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Body composition in older adults with mild cognitive impairment: effect of 6 months of resistance training

Isadora Cristina Ribeiro[1], Thiago Mattos Frota de Souza[1], Jose Igor Vasconcelos[1], Gabriel Barzon Silva[1], Marco Carlos Uchida[1], Paula Teixeira Fernandes[1], Italo Karmann Aventurato[1], Marjorie Cristina Rocha da Silva[1], Brenda Costa Gonçalves[1], Marcio Luiz Figueredo Balthazar[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Isadora Cristina Ribeiro; Email: isadora.cr@hotmail.com.

Background: Mild cognitive impairment is the diagnosis applied to older adults who present a cognitive loss unnatural to aging, affecting one or more domains of cognition. These individuals maintain their functional independence and are at greater risk of developing dementia, such as Alzheimer's disease, which is related to obesity and a decrease in muscle strength. Investigating the impact of interventions that promote changes in body composition in this population, such as physical exercise, helps to understand how non-pharmacological tools can be effective against risk factors for dementia. **Objective:** This study investigated the influence of resistance exercises practicing for 6 months on body composition in older adults with mild cognitive impairment. **Methods:** older adults (N=22) diagnosed with mild cognitive impairment following the National Institute on Aging and Alzheimer's Association criteria underwent a resistance training protocol twice a week for 6 months. The protocol consisted of large muscle group exercises (3 sets of 10 repetitions at 80% of 1RM). Before and after the intervention, participants were assessed for body composition on a Tanita® BC-108 bioimpedance scale. For measurement, the individual's sex and the "non-athlete" option were selected. The participant removed the handles electrodes from the equipment, keeping their arms away from the torso. The participants' body composition data to be evaluated were body fat percentage, lean mass, fat mass, body water, and basal metabolic rate. Data normality was assessed using the Shapiro-Wilk test. Due to the normal distribution of the data, the paired T-test was used to compare the initial and final moments of the intervention in the IBM SPSS Statistics 22 software. **Results:** Data are presented as mean and standard deviation, and before and after the intervention, respectively: body fat (%): 32.35 (8.68), 32.87 (9.76); lean mass (kg): 52.84 (12.35), 51.94 (11.10); fat mass (kg): 25.60 (9.69), 26.21 (11.50); body water (kg): 38.38 (8.62), 38.51 (7.96); basal metabolic rate (kcal): 1515.40 (332.97), 1503.95 (299.99). As a result of the comparison between the pre and post-intervention moments, no significant changes were found for any of the measures: body fat percentage: p=0.52, lean mass: p=0.37, fat mass: p=0.41, body water: p=0.77, basal metabolic rate: p=0.55. We point out as a limitation that another factors relevant to body composition changes, such as diet, for example, were not evaluated. We suggest further investigations with larger populations and longer durations to contribute to the knowledge of the body composition changes in mild cognitve impairment. **Conclusion:** We highlight that to our knowledge this is the first study to compare bioimpedance parameters in elderly people with mild cognitive impairment after resistance training. Our findings suggest that 24 weeks of resistance training were not enough to change body composition in older adults with mild cognitive impairment.

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Botulinum toxin as a therapeutic option in Graves' orbitopathy: a case report

Vinícius Andreata Brandão[1], Lavínia Damacena Perin[1], Jovana Gobbi Marchesi Ciríaco[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Vinícius Andreata Brandão; Email: viniciusandreatab@gmail.com.

Case presentation: Female, 62 years old, with a significant past pathological history, including systemic arterial hypertension (SAH), mitral valve disease secondary to rheumatic fever corrected with implantation of a metal prosthesis, iron deficiency anemia, actinic colitis after radiotherapy for neoadjuvant treatment of cervical cancer, as well as Graves' disease treated with total thyroidectomy in 2003, presenting postoperatively with refractory bilateral exophthalmos, worse on the right. A therapeutic attempt was made with botulinum toxin administration, with control of ocular proptosis with injections at 4-month intervals since 2015. **Discussion:** Graves' orbitopathy (OG) is the main extrathyroidal manifestation of Graves' disease. It is characterized by inflammation of the extraocular muscles and orbital connective tissue, which becomes progressively larger, leading to ocular proptosis, which can result in damage to the cornea or conjunctiva. In general, exophthalmos does not regress with the treatment of thyroidopathy and surgical intervention is necessary to decompress the orbit and thus allow for better accommodation of the eyeballs. However, in some cases surgery is not appropriate and may be contraindicated, such as in complex patients with a high surgical risk. Therefore, effective and safer alternatives can be chosen, such as the administration of botulinum toxin type A (BTXA) for controlled induction of eyelid ptosis. Botulinum toxin is a potent neurotoxin from the bacterium Clostridium botulinum that selectively inhibits the secretion of acetylcholine vesicles at the neuromuscular junction, resulting in chemical denervation. Its use in the levator palpebrae muscle has been shown to be effective in reducing the symptoms resulting from Graves' orbitopathy, as it allows for better occlusion of the eye, leading to better aesthetics and a reduction in the risk of complications due to conjunctival and corneal exposure. **Final comments:** The use of botulinum toxin A for Graves' orbitopathy is effective, with the potential to substantially improve symptoms, providing a viable alternative for patients at high surgical risk.

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Botulinum toxin as an ally in the treatment of belly dancer syndrome: a case report

Leticia Machado Dumont[1], Filipe Brito de Ferraz[1], Rafael Chaves Claudino de Queiroga[1], Camila Lopes Figueiredo[1], Cícero Rodrigues Veloso[1], Maria Clara Motta Barbosa Valente[1]

[1]Irmandade Santa Casa de Misericordia de São Paulo, São Paulo SP, Brazil.

Correspondence: Leticia Machado Dumont; Email: leticiamachadodumont@gmail.com.

Case presentation: This case report describes a female patient, 48 years old, diagnosed with neuromyelitis optica (NMO) in 2010 after presenting an episode of optic neuritis in the right eye. In 2020, the patient presented a new bout of NMO, this time characterized by T8 paraparesis and crural paraparesis. After the attack, the patient began to present painful involuntary movements in the abdominal and dorsal region, radiating to the lumbar region, characterized as contraction of bilateral internal and external oblique muscles. An investigation with Electroneuromyography was carried out in September 2020, the report demonstrated spontaneous discharges of action potentials with arrhythmic activity and variable frequency, suggestive of myokymia at rest in the rectus abdominis muscles and more markedly in the left oblique muscle. Based on the diagnostic hypothesis of belly-dancer syndrome (BDS), an attempt was made at pharmacological therapy with levetiracetam, without satisfactory response, nor when combined with carbamazepine and gabapentin. Due to the failure of pharmacological therapy, the patient underwent application of botulinum toxin to the left rectus abdominis and left external oblique muscles, showing partial improvement in symptoms, with a reduction in the intensity and frequency of muscle spasms. **Discussion:** "Belly-dancer" syndrome is a rare syndrome involving repetitive and involuntary movements of the abdominal wall muscles, also known as diaphragmatic myoclonus. Various etiologies have been associated with the phenomenon, from medications, post-operative abdominal surgery, stroke, metabolic changes, trauma and autoimmune disorders, as is the case with NMO. Such changes lead to abnormal excitation of the phrenic nerve, via the central nervous system or along the nerve. Another proposed mechanism is irritation of the diaphragm muscle itself. Regarding treatment, there are reports of relative success with the use of phenytoin, carbamazepine, as well as clonazepam, fluoxetine, gabapentin, pimozide, valproic acid, among others. Thus, this syndrome becomes a therapeutic challenge, as the response to pharmacological therapy is variable. The use of botulinum toxin is one of the most recent therapies and appears as a promising alternative for treatment, especially in cases refractory to medication. The toxin works by blocking the release of acetylcholine at the neuromuscular junction, which results in temporary paralysis of the affected muscles. In the present case, the application of botulinum toxin provided partial improvement in the patient's symptoms, which corroborates the literature that points to the effectiveness of this therapeutic approach in BDS. **Final comments:** BDS can be a debilitating condition, but with proper treatment, patients' quality of life can be significantly improved. The application of botulinum toxin appears to be a promising alternative for treating the syndrome, especially in cases refractory to pharmacological therapy.

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Botulinum toxins on muscular spasticity rehabilitation in a reference center in Northern Brazil

Gustavo Celeira de Sousa[1], Vitor Ferreira Baia[1], Fernando Gabriel dos Santos Santiago[1], Savio Roberto Silva Costa[1], Danusa Neves Somensi[2], Mariângela Moreno Domingues[1], Emanuel de Jesus Soares de Sousa[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Gustavo Celeira de Sousa; Email: gustavo.c.sousa0@gmail.com.

Background: Muscle spasticity is the exacerbation of stretch reflex, resulting in continuous muscle contractions and is a common sequel to various neurological disorders, especially those that affect the first motor neuron. In Brazil, epidemiology of spasticity is directly related to the etiology of this condition, which may be secondary to ischemic insult, as in stroke and cerebral palsy; or traumatic brain and spinal cord injury. One of the main treatments for this condition is intramuscular injection of botulinum toxin A (TBA), a neurotoxin that blocks acetylcholine liberation on neuromuscular junction, causing reduction of muscle tonus. **Objective:** Analyzing and monitoring results of TBA treatment in patients with spasticity in a specialized service. **Methods:** This is a cohort study that followed treatment of spasticity in 43 individuals. The location and amount of TBA applied was defined according to the patient's clinical characteristics and current Ministry of Health guidelines. Patients were evaluated monthly using modified Ashworth scale (MAS). In addition, epidemiological and demographic data were collected. Statistical analysis of collected data was performed using GraphPad Prism 9.0 software; simple statistical calculations and p-value calculation were performed using the chi-square test. For this study, a p-value less than 0.05 was considered statistically significant. **Results:** Regarding etiology, about 29% of the patients had cerebral palsy, followed by patients with stroke sequelae. It was observed that spasticity pattern was different amongst between different etiologies for spasticity, of note patients with cerebral palsy-related spasticity had predominance of affection of lower limbs (LL) muscles bilaterally, while patients with stroke-related spasticity had bigger prevalence of unilateral upper limb (UL) and lower limb commitment. The main muscles of the UL affected were brachial biceps, finger flexors, finger flexors, while in LL the most affected were gastrocnemius, hamstrings, and leg adductor. Regarding the patient's age, it was observed that individuals with cerebral palsy usually were under 20 years old; stroke sequelae patients usually were over 30 years old. From start of treatment to deadline it was evidenced substantial reduction in muscle spasticity, mainly in lower limbs muscles, with statistically significant difference, thus having a positive impact in gait and improvement of quality of life. The results related to muscles of UL showed slight improvement of spasticity, but no statistically significant difference. **Conclusion:** This research reinforced the effectiveness of TBA treatment, remarkably in lower limb spasticity, causing an improvement not only in the kinetics of the limbs, but also functionally, by facilitating movement and enabling advances of physical therapy treatment. Thus, it is possible to conclude that use of TBA is associated with positive outcomes and improvement on individuals' quality of life.

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Bow Hunter's syndrome in a patient with paroxysmal vertigo: case report

Mariana Akemi Takegawa Miyahara[1], Santhiago Calvelo Graça[1], Rachel Leirner Argelazi[1], Mauro Gualano da Costa Abreu[1], Camila Lopes Figueiredo[2], Filipe Brito Ferraz da Silveira[2], Cícero Rodrigues Veloso[2], Maria Clara Motta Barbosa Valente[2], Vívian Dias Baptista Gagliardi[2], Rubens José Gagliardi[2]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Irmandade Santa Casa de Misericordia de São Paulo, São Paulo SP, Brazil.

Correspondence: Mariana Akemi Takegawa Miyahara; Email: akemi.mariana99@yahoo.com.br.

Case presentation: A 51-year-old female patient with a history of systemic hypertension, depression, Systemic Lupus Erythematosus, and a previous stroke ten years before was admitted for a neurology evaluation at the Emergency Room due to significant neck pain and paroxysmal vertigo occurring with head rotation to the left. Additionally, she experienced a sudden event of left hemiparesis, lasting less than 24 hours, attributed to a Transient Ischemic Attack (a head CT at the time only described the previous lesion). During the neurological examination, performing cervical rotation to the left by approximately sixty degrees induced vertigo, oscillopsia, and erratic rapid saccadic eye movements. In a motor assessment, she presented a slight deficit in strength on the left side and hypoesthesia. Brain and cervical MRI and MRA showed no current or previous infarction; MRA findings suggested hypoplasia and dissection of the left vertebral artery. Transcranial Doppler revealed a stenotic process in the left vertebral artery, along with spontaneous brain embolic activity during insonation of the left vertebral artery (V4 segment), proximal basilar artery, and right posterior cerebral artery. Further investigation with arteriography showed irregularities and moderate stenosis (around 40%) in the right vertebral artery; at the transition of its V3/V4 segments, this narrowing became critical during a left head rotation maneuver. Additionally, there was occlusion of the left vertebral artery in its V3 segment. Considering the findings-occlusion of the left vertebral artery and compression of the right vertebral artery during head rotation resulting in vertigo and nystagmus, confirmed by angiography - a diagnosis of Bow Hunter's Syndrome or Rotational Vertebrobasilar Insufficiency was made. In this context, platelet anti-aggregation with Clopidogrel was introduced due to the patient's allergy to aspirin. Cardiological changes and thrombophilia were ruled out, and the patient showed good recovery during follow-up visits. **Discussion:** Vertigo is a common complaint in emergency rooms worldwide, including Brazil. Diagnosing its cause is crucial, with benign paroxysmal positional vertigo (BPPV) being the most frequent diagnosis due to its high prevalence and benign nature. However, considering a stroke is essential, even if no other focal neurological signs are present, as vertigo can be a primary symptom of a vertebrobasilar stroke. Early identification and treatment of a stroke are critical to reduce the risk of permanent damage and improve outcomes. **Final comments:** Managing vertigo in the emergency department remains an important challenge. In cases of acute onset and continuous duration, the hypothesis of ischemic posterior circulation stroke must be considered. Additionally, when transient symptoms are associated with cervical and head movements, vertebral artery compression syndrome should be considered, as seen in Bow Hunter's syndrome.

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Brachial plexitis: an unusual presentation in dengue virus infection

Caio Vinicius Barroso de Lima[1], Marcelo Annes[1], Vinícius Boaratti Ciarlariello[1], Cristina Gonçalves Massant[1], Anna Maria Andrei[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Caio Vinicius Barroso de Lima; Email: caiolimaneurologista@gmail.com.

Case presentation: We report the case of a 61-year-old female patient. She presented in the emergency room with acute weakness of the left upper limb, upon awakening, 3 days before hospitalization. The patient denied pain, fever or other associated symptoms in the last few weeks. She had monoplegia in the left upper limb, hypoesthesia and areflexia with flaccidity of the same limb. Initially

investigated with cranial and cervical angiotomography, excluded cerebral vascular events. In laboratory, only mild thrombocytopenia was relevant. Cranial and cervical MRI were also unremarkable. A brachial plexus MRI demonstrated thickening and enhancement of the left C6, C7 roots, the upper and middle trunks with changes in diffusion. Edema of the left scalene, left teres major and infraspinatus muscles were also seen, related to denervation. Electroneuromyography revealed severe hyperacute involvement of the left brachial plexus. CSF demonstrated slight pleocytosis with lymphomonocyte predominance. Finally, specific research confirmed positive IgG and IgM serology for dengue virus with negative NS1 antigen. **Discussion:** Neurological dengue is classified as a severe form of dengue. Neurological complications are seen in 4.2 - 13% of all DENV. Brachial neuritis is characterized by severe neuropathic pain (95%), sudden weakness (80%), hypoesthesia (78%) and atrophy of the proximal musculature. Most of cases are unilateral. One of the manifestations associated with dengue virus is the immune-mediated neurological complications (IMNC), which include brachial plexus neuritis. Majority of IMNC cases occur in the subacute (7-30 days) latency period. All patients achieved complete clinical recovery after a course of intravenous methylprednisolone (MP) and immunoglobulin (IVIg). Studies hypothesize that post-dengue infection leads to an autoimmune response against the myelin sheath, causing brachial neuritis due to demyelination of the plexus. Unlike most reports, our patient did not demonstrate pain related to brachial plexitis. She also did not present any classic viral symptoms of dengue preceding the neurological condition. Not even fever, which is the symptom most related to the dengue IMNC. Corroborating literature, our case presented in a subacute interval of dengue virus infection (negative NS1 with positive IgM IgG indicates infection between 7 - 29 days). According to recent literature, we treated her as a dengue IMNC case: immunomodulation with 3 days of MP concomitantly with IVIg for 5 days, and a complete response should be assessed within the next 3 months. **Final comments:** Dengue is a viral infection with worldwide distribution, especially in underdeveloped tropical countries. Where diagnostic resources are often limited and access to appropriate treatment is also scarce. Therefore, diagnostic suspicion is crucial for adequate investigation and preservation of resources, as well as aiming the best outcome for the patient.

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Brachial plexus axonotmesis after coronary artery bypass surgery: a case report

Natana Rangel da Silva Ribeiro[1], Natália Curcio Fedrizzi[1], Ana Maria Hoppe[2]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Natana Rangel da Silva Ribeiro; Email: natanarangel@gmail.com.

Case presentation: A 60-year-old man with a history of coronary artery disease, pre-diabetes, hypertension and smoking, was admitted for a coronary artery bypass surgery. After awakening from sedation, he noticed weakness and numbness in his left hand. On examination, sensation was diminished on the left palmar and dorsal surfaces of fourth and fifth fingers, medial edges of the hand and of the ipsilateral forearm. He had weakness of left interossei and thumb abductors, and of extensor digitorum. Triceps, biceps and brachioradialis reflexes were preserved. Electrodiagnostic studies were realized 1 month later. Nerve conduction studies demonstrated decreased left ulnar compound muscle action potential amplitude, recorded from the first dorsal interosseous and the abductor digiti minimi muscles, and reduced medial cutaneous of the forearm and ulnar sensory nerve action potentials amplitudes. On needle electromyography, evidence of active denervation and also early reinnervation in the left first dorsal interosseus, extensor indicis and flexor pollicis longus was seen, suggesting a Sunderland second-degree axonotmesis injury of the lower trunk of left brachial plexus. **Discussion:** The brachial plexus is formed by the C5, C6, C7, C8 and T1 nerve roots, and is divided into roots, trunks, divisions, and cords. It is responsible for the upper limbs innervation and its injuries can be classified as preganglionic, which are not repairable, or postganglionic, which can be spontaneous or surgically repaired. Through electrodiagnostic studies, nerve injuries can also be classified by the Seddon and Sunderland classification systems as neuropraxia, axonotmesis or neurotmesis. Neuropraxia is a demyelinating injury with no axon loss, and complete recovery within days to weeks. Axonotmesis is a partial loss of axons with intact endoneurim, and complete (Sunderland second-degree axonotmesis) or partial (Sunderland third-degree axonotmesis) recovery. Neurotmesis is a severe damage to axons, myelin sheath and connective tissue elements, with surgical repair being essential for recovery. In all injuries described, physiotherapy plays an important role in rehabilitation and is the best course of action before surgery. After surgery, it can also help restore neurological function. **Final comments:** Patients undergoing open heart surgery must be closely monitored for peripheral nerve injury during the postoperative period, as missed injuries may subsequently lead to delay in recovery, morbidity, and poor functional outcome. The investigation can be done with detailed history and examination, complemented with timely electrophysiological studies.

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Brachial plexus injury after breast prosthesis exchange

Sara Pessoa de Oliviera[1], Julia Gasparino de Oliveira[1]

[1]Centro Universitario Integrado, Campo Mourão PR, Brazil.

Correspondence: Julia Gasparino de Oliveira; Email: julia.gasparino@grupointegrado.br.

Female patient who underwent mastopexy surgery and breast implant replacement on Agt 24, 2022. Postoperatively, she developed anesthesia and brachial paraplegia. She was prescribed Etna® and Dexador® and referred for physical therapy. She sought neurological consultation on Sep 1, 2022, reporting hypoesthesia, paresthesia (tingling), and weakness in the upper limbs. Physical examination (upper limbs): arm abduction (strength (S) grade 1 on the right and 3 on the left), flexion and extension of forearms (S grade 2), and hands (S grade 4); upper limb hypoesthesia; globally reduced deep tendon reflexes (DTR) grade 1; absent Hoffman sign. Upper limb nerve conduction studies were requested on Sep 16, 2022, revealing bilateral brachial plexopathy with signs of acute axonal loss. By September 22, 2022, there was partial improvement in upper limb weakness - arm abduction (S 2 on the right and 3 on the left), flexion and extension of forearms (S 4 on the left and 3 on the right), and hands (S 4). Dexador® and Etna® were gradually discontinued, and Millgama® and Pregabalin were prescribed along with continued physical therapy. Cervical spine MRI on Sep 30, 2022, showed a posterior central disc protrusion at C5-C6 level indenting the ventral aspect of the dural sac; brachial plexus MRI showed mild T2 hyperintensity. On Oct 20, 2022, the patient exhibited difficulty maintaining isometry, upper limb hypoesthesia, and paresthesia in the right thumb. Physical examination showed improved upper limb strength (S 4 in hands and forearms; S 3 in arm abduction and shoulder elevation), grade 2 brachioradialis reflex on the right, and grade 1 elsewhere. Medications and physical therapy were continued. At the last visit, the patient reported complete improvement in movements, reducing physical therapy sessions. Nerve conduction studies on Feb 3, 2023, did not reveal significant abnormalities, demonstrating clear improvement in electrophysiological patterns compared to previous studies. Neurological examination showed S 5 in both upper limbs, preserved tone, reflexes grade 2, and intact tactile sensation. The patient was discharged. Disscution Fernandes notes that most peripheral nervous system injuries involve the brachial plexus, mainly from traumas during shoulder manipulation, leading to functional and sensory loss. This case details brachial plexopathy post-breast reconstruction surgery, highlighting varied surgical techniques impacting outcomes. Initial assessment revealed proximal upper limb weakness on the Medical Research Council (MRC) Scale. Electromyography confirmed bilateral plexopathy, aligning with complex diagnostic challenges requiring advanced imaging. Follow-up showed muscle strength improvement and MRI findings suggesting transient plexopathy. Early suspicion and treatment led to full recovery in five months, emphasizing non-surgical intervention benefits. Clinical vigilance and prompt patient response significantly influenced prognosis.

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Brain abnormalities in early-onset spinal muscular atrophy with neurodevelopmental disorders

Rodrigo de Holanda Mendonca[1], Raquel Diógenes Alencar Sindeaux[1], Clara Gontijo Camelo[1], Erasmo Barbante Casella[1], Kellen Faria Silva Chillon[1], Antônio José da Rocha[2], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.
[2]Alta Medicina Diagnóstica, São Paulo SP, Brazil.

Correspondence: Rodrigo de Holanda Mendonca; Email: rodrigo.holanda@hc.fm.usp.br.

Background: Recent literature has reported the occurrence of neurodevelopmental comorbidities in early-onset spinal muscular atrophy linked to 5q (SMA-5q). Furthermore, previous reports before the advent of disease-modifying therapies (DMT) suggested brain involvement in 5q-SMA. **Methods:** This cross-sectional study of a case series included five patients with early-onset and pre-symptomatic SMA-5q under treatment with DMT and who presented with neurodevelopmental disorders, including four cases with autism. These patients underwent brain MRI with volumetric analysis utilizing Neuroquant® software and tractography using diffusion tensor imaging (DTI). **Results:** Patients with type 1 SMA-5q exhibited a consistent pattern of volumetric reduction in the frontal lobes and reduced representation of association fibers corresponding to the superior longitudinal fasciculus. In contrast, pre-symptomatic patients who received treatment did not show any abnormalities on brain imaging. In an older SMA-5q type 1 patient with advanced disease, used as a control, the spectrum of imaging changes was more severe, presenting marked atrophy in both frontal lobes. **Conclusion:** Brain involvement in early-onset SMA-5q may occur even in patients treated with new DMT, and its correlation with neurodevelopmental disorders remains an open question.

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Brain aneurysm neurogenetics: a systematic review

Graziele Gleice da Silva[1], Ana Silvia Suassuna Carneiro Lúcio[2], Alexandre Jorge de Andrade Negri Júnior[2]

[1]Cabedelo.
[2]Faculdade de Ciências Médicas da Paraíba, Cabedelo, PB, Brazil.

Correspondence: Graziele Gleice da Silva; Email: grazielle_gleice@hotmail.com.

Background: Genetic factors are crucial to the formation and clinical evolution of intracranial aneurysms. Several loci have been associated with its pathogenesis, many of which implicate several pathways related to endothelium maintenance and extracellular matrix integrity. Subarachnoid hemorrhage (SAH) and intracranial aneurysm (IA) have a complex origin, entailing the interaction of several genes and environmental factors. The rupture of the extracellular matrix of an arterial wall, which confers strength and elasticity to intracranial arteries, is a probable factor in the pathophysiology of intracranial aneurysms. In this review, the focus lies on genetics association, linkage, and gene expression of SAH and IA. Loci neurogenetics studies include functional structural-protein-codifying genes of extracellular matrix. The elastin and type 1A2 collagen genes are the most promising candidates given their allelic association with IA.**Objective:** To determine whether polymorphism of genes implicated in the maintenance of vascular endothelium may modify the risk of developing IAs. **Methods:** This is a systematic review carried out through the following databases: Medical Literature Analysis and Retrieval System Online (MEDLINE/PubMed), Latin-American and Caribbean Health Sciences Literature (LILACS/Bireme), Science Direct, Web of Science, and Scientific Electronic Library Online (SCIELO). The filters used were title, subject, and type. The terms used in the search for the articles were standardized by the Medical Subject Headings (MeSH) and Health Sciences Descriptors (DeCS). The descriptor "brain aneurysm" was combined using the boolean operator AND with the descriptors: "neurosurgery" and "genetics". The guiding question of the study was: How can the study of the genetic profile of brain aneurysms improve the prevention and diagnosis of this pathology? **Results:** The T allele of the rs1799983 polymorphism of the endothelial nitric oxide synthesis gene (eNOS) is correlated with a decreased risk of developing the disease. Likewise, the G allele of the rs1799983 polymorphism of the eNOS increased the risk of developing intracranial aneurysm. A comprehensive understanding of the genetic basis that governs the risk of developing IA and the resulting aneurysmal subarachnoid hemorrhage may aid in the screening, clinical management, and risk stratification of these patients, and may also allow the identification of putative mechanisms for the further development of medications. **Conclusion:** The association of the rs1799983 polymorphism of the eNOS with an elevated risk of IA reinforces the hypothesis that genetic variants in the eNOS gene may be crucial in its pathogenesis. Future studies need to demonstrate which alleles are related to the origin and incidence of aneurysms, as well as determine the screening for this pathology.

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Brain changes in an elderly woman with Hypomelanosis of Ito

Patricia Soares Rodrigues Mello[1], Igor Castro Tavares[1], Rafael Cavalcanti Fernandes Rocha[2], Massanobu Takatani[2], Nise Alessandra de Carvalho Sousa[2], Satiko Andrezza Ferreira Takano[2]

[1]Universidade Nilton Lins, Manaus AM, Brazil. [2]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Satiko Andrezza Ferreira Takano; Email: satikotakano.neuro@gmail.com.

Case presentation: A 66-year-old woman with amnestic, visuospatial, executive, and attention deficit complaints, also presenting with peripheral vertigo, headache, and depressive and anxiety disorder. Physical examination revealed hypochromic spots in the abdomen, and upper and lower left limbs, present since childhood. Laboratory tests, including complete blood count, and renal, hepatic, and thyroid function tests, were within normal limits. Magnetic resonance imaging (MRI) of the skull revealed multiple enlarged perivascular spaces in the bilateral periventricular white matter and cavernomas. Skin biopsy showed mild lymphohistiocytic infiltrate in superficial vessels, favoring the diagnosis of Hypomelanosis of Ito. **Discussion:** Hypomelanosis of Ito or Incontinentia pigmenti achromians is a rare, multisystem neurocutaneous disease with hypopigmentation of the skin along the Blaschko lines. The cause is genetic, but not fully known, and chromosomal alterations with somatic mosaicism may occur. The clinical presentation is related to dermatological findings, unilateral or bilateral, and 75% have extracutaneous neurological, musculoskeletal, cardiac, and genitourinary manifestations. Neurological manifestations may include cognitive impairment, epilepsy, spasticity, cerebellar ataxia, and microcephaly. Neuroradiological alterations include lissencephaly, polymicrogyria, laminar heterotopias, cerebellar atrophy, atrophy of the posterior third of the corpus callosum, dilation of the Virchow-Robin spaces, and tumors. Subcortical alterations may aggravate the patient's cognitive symptoms. **Final comments:** Recognition of clinical, dermatological, and brain changes in patients with Hypomelanosis of Ito is necessary for early diagnosis of this disease, allowing for appropriate treatment with a multidisciplinary approach

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Brain changes in pediatric multiple sclerosis

Gabriel de Deus Vieira[1], Fernanda Ferrão Antônio[1], Alfredo Damasceno[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Gabriel de Deus Vieira; Email: gabrieldedeusvieira@gmail.com.

Background: Pediatric-onset multiple sclerosis (POMS) comprises about 5% of all multiple sclerosis (MS) patients. It presents similarly to the adult disease, but with a more inflammatory disease. **Objective:** we aimed to evaluate clinical and neuroimaging data in POMS subjects. **Methods:** A total of 10 pediatric MS patients were enrolled (under 18 years of age). For neuroimaging comparisons, 16 controls were selected. Brain volume evaluations were performed on volumetric T1 and 3D-FLAIR images using the Sequence Adaptive Multimodal Segmentation, a FreeSurfer pipeline to segment brain structures. All patients underwent to timed 25-Foot Walk Test, 9-Hole Peg Test, Symbol Digit Modalities Test (SDMT) and Expanded Disability Status Scale. Cortical gray matter volume, deep white and gray matter structures were analyzed. The paramagnetic rim lesions were assessed for each lesion with SWI sequence. Statistical analysis was performed using the SPSS software. To compare data between groups, the General Linear Model and Mann-Whitney U test were used. Correlations were performed using Spearman correlation coefficient. **Results:** The MS group consisted mainly of girls (n=8), with a mean age of 15.3 years old. All patients had a relapsing-remitting clinical, 60% used fingolimod, 20% interferon beta-1a, 10% natalizumab and 10% rituximab. None of them presented severe physical disabilities. There was a higher choroid plexus and lower thalamic volumes in POMS group compared to controls. Regarding PRL, 90% had this type of lesion. The percentage of white matter lesions with a paramagnetic rim lesions was 18.8%. Regarding correlations, the SDMT had a strong correlation with paramagnetic rim lesions percentage ($R=-0.800$, $p<0.01$). **Conclusion:** POMS is characterized by a greater number of relapses compared to adults, having a higher frequency of paramagnetic rim lesions and its correlation with SDMT (a cognitive test, related to information processing speed). In addition, the MS group presented changes in volumetry, with atrophy of the thalamus and enlargement of the choroid plexus.

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Brain damage in carriers of DMD pathogenic variants

Mariana Rabelo de Brito[1], Thiago Junqueira Ribeiro de Rezende[1], Stephany da Silva Passos[1], Anamarli Nucci[1], Fernando Cendes[1], Marcondes Cavalcante França Jr.[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Mariana Rabelo de Brito; Email: mariana_rabelo_@hotmail.com.

Background: Duchenne muscular dystrophy (DMD) is typically a male disease, but mounting evidence indicates that females carrying DMD mutations (cDMD) can also be affected. More recently, cognitive impairment has been noticed in women with cDMD. Despite this, no studies have attempted to evaluate brain damage using imaging tools, particularly MRI. **Methods:** Sixty-six women (33 with cDMD and 33 controls) underwent cognitive assessment using the Addenbrooke's Cognitive Examination Revised (ACE-R) and 3T MRI to evaluate gray matter (GM) and white matter (WM). FreeSurfer was used to assess cerebral GM and DTI multi-atlas for cerebral WM. All analyses were corrected for multiple comparisons. **Results:** The average age of women with cDMD was 38.5 years, and their average educational level was 9.4 years of formal education. The mean score on the ACE-R was 73.4±9.6 points, with almost 50% of the patients (n=16) scoring below the expected range, suggesting some degree of deficit. Cortical thinning was observed in the right inferior parietal, bilateral precuneus, and bilateral superior parietal pole. Additionally, a reduction in gyrification index was seen at the bilateral gyrus and sulcus parietal. However, we did not find a correlation with the patients' cognitive deficit. **Conclusion:** Cognitive impairment has been observed in women with cDMD, who are also characterized by structural damage, especially in the temporal lobe and its gyrus and sulcus. Our results shed some light on the pathophysiology underlying the disease.

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Brain death after a cardiac arrest: main characteristics of a group

Rafaela dos Santos Braga[1], Gisele Sampaio Silva[1], Maramelia Miranda[1], Ingrid Marques[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Rafaela dos Santos Braga; Email: braga.rafaela@gmail.com.

Background: The occurrence of brain death in patients with hypoxic-ischemic brain injury after resuscitation from cardiac arrest creates opportunities for organ donation. About two-thirds of patients admitted to hospital or an intensive care unit after cardiac arrest die before hospital discharge. Most of these deaths are due to hypoxic-ischemic brain injury. Brain death is an unfavorable outcome for the individual patient; however, it creates opportunities for organ donation. **Objective:** Knowing the epidemiology of brain death after cardiac arrest could help physicians organize a timely screening and identification of potential organ donors after resuscitation. **Methods:** We reported the main characteristics of a group of patients that evaluated to brain death after a cardiac arrest. **Results:** On period of six months, 45 patients were notified as potential donor under the Brazilian criteria of brain death and the cause of death was described as hypoxic-ischemic encephalopathy after a cardiac arrest. Patients from 0 to 78 years old were included, with a median age of 42.66 years old, 3 cases under 2 years old, 7 cases between 2-18 y-o, 24 cases between 18-65 y-o and 11 cases above 65 y-o. The eldest patient was 78 years old. There was 27 female (60%) and 18 men (40%). Mean time of cardiac arrest was 11 minutes, with a minimum of 2 minutes and the maximum of 30 minutes. The number of days since the cardiac arrest included from 3 days to 10 days, with a median of 5.5 days. In patients with primary brain

injury as the cause of cardiac arrest a median of 4.6 was the number of days until brain death was confirmed. Cardiac arrest is a catastrophic event with high mortality. Fortunately, survival has improved over the past several decades; however, mortality rates are still high, and wide variability in survival exists, plus some underreported data. Nowadays, there is an urgent need to improve postcardiac arrest care, and there are multiple knowledge gaps related to patient management and neuro prognostication. The postcardiac arrest syndrome includes varying levels of brain injury, circulatory dysfunction, and multiorgan ischemia/reperfusion injury and dysfunction. The number of days from the cardiac arrest until the confirmed brain death varied from 3 10 days, (median 5.5 days). In some cases, the occurrence of brain death has been reported within 1 day after cardiac arrest. This is common when cardiac arrest is due to neurological causes, most commonly a subarachnoid hemorrhage. When a neurological rather than a cardiac cause of arrest is suspected, a brain computerized tomography immediately after the return of spontaneous circulation is recommended. If brain computerized tomography demonstrates a catastrophic brain injury, an early evaluation for brain death may be considered. **Final comments:** This small study had the intention to bring up the importance of studying the population who suffered a cardiac arrest.

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Brain morphometric changes in SAA positive individuals with idiopathic Parkinson's disease

Stella Duarte Pinto[1], Andrei Bieger[2], Daniel Teixeira dos Santos[3], Rafaela Ravazio[1], Christian Mattjie[1], Lara Angi Souza[4], Thomas Hugentobler Schlickmann[4], Rodrigo C. Barros[1], Eduardo R. Zimmer[4], Artur Francisco Schumacher Schuh[3,4]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.
[3]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[4]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Stella Duarte Pinto; Email: stella.pinto@edu.pucrs.br.

Background: Molecular diagnostics are becoming increasingly important for detecting Parkinson's disease (PD) in its early stages. The α-synuclein seed amplification assay (SAA) has shown high sensitivity and specificity in detecting synucleinopathies in the cerebrospinal fluid (CSF). Differences in brain morphometry between PD patients with and without a positive SAA test are still unknown. **Objective:** This study analyzed cortical and subcortical morphometric alterations comparing them with the outcome of the SAA in patients with idiopathic PD. **Methods:** We used T1-weighted 3-dimensional brain MRI images from participants of the Parkinson's Progression Markers Initiative (PPMI) database with idiopathic PD who also underwent CSF collection and SAA analysis. Using Freesurfer's v7.4.1 recon-all pipeline, we extracted volumes of cortical and subcortical regions of interest, according to the Desikan-Killiany-Tourville atlas. Volumes were normalized according to estimated total intracranial volume. Linear models evaluated differences in volumes between positive and negative SAA groups, correcting for age and sex. **Results:** A total of 267 participants (95.5% positive SAA, 63.2% males, mean age 60.6 years, standard deviation 8.1) were included. In patients with idiopathic PD, volume changes in positive SAA were observed in the right parahippocampal (p=0.02), right fusiform (p=0.03), and left posterior cingulate gyri (p=0.04), and in the left frontal pole (p=0.05). Subcortical structures presented changes in volumes of left putamen (p=0.02) and left globus pallidus (p=0.03). Conclusions The results of this study demonstrate a clear association between SAA positivity and specific morphometric changes in the brains of patients with idiopathic PD. These changes include significant reductions in cortical thickness and subcortical volumes in critical regions associated with memory, cognition, and motor control. These findings suggest that SAA may be a valuable biomarker not only for the early detection of PD but also for monitoring disease progression through structural changes in the brain. Identifying specific morphometric changes can help personalize therapeutic strategies and monitor the efficacy of treatments over time. To consolidate these findings, future studies should include larger samples and consider the clinical heterogeneity of PD. Additionally, integrating neuroimaging data with other diagnostic modalities, such as molecular and clinical biomarkers, may provide a more comprehensive view of PD progression and treatment responses. **Conclusion:** This study significantly contributes to understanding the relationship between α-synuclein and structural brain changes in Parkinson's disease. Detecting morphometric changes associated with SAA positivity can enhance the diagnosis and treatment of PD, offering new perspectives for managing this neurodegenerative condition.

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Brainstem encephalitis and subacute sensory neuropathy accompanied by colon cancer: is there a possible paraneoplastic association?

Camila Rodrigues Nepomuceno[1], Gustavo Celeira de Sousa[1], Maria Fernanda Hamamoto Leati[1], Heitor Nunes de Oliveira Sento-sé Neto[1], Flavio Vieira Marques Filho[1], Arthur de Medeiros Dias[1], Graziella Aguiar Santos Faria[1], Maria Sheila Guimaraes Rocha[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Camila Rodrigues Nepomuceno; Email: camilarnepomuceno@gmail.com.

Case presentation: A 51-year-old woman was transferred to our service due to flaccid areflexic tetraparesis and diffuse nociceptive and neuropathic pain which began seven days before hospitalization. The onset of symptoms occurred four days after the resection of a colonic tubular adenoma. Neurological examination showed preserved consciousness, cognition, memory, language and muscle strength. Important impairment on sensory pathways, mainly on profound system (sensory ataxia, pseudoparesis, pseudoathetosis) was observed. Other relevant clinical finding was multidirectional nystagmus. Laboratory tests did not show significant changes, including anti-SSA and anti-SSB antibodies. CSF analysis evidenced elevated protein levels and light pleocytosis. On MRI, signal hyperintensity in pons paramedian region. Her electroneuromyography also showed pure sensitive polyneuropathy non-length-dependent. The anatomopathology of colon biopsy revealed moderately differentiated adenocarcinoma of the colon. Onconeural antibodies were not available for our patient due to the resource-limited setting. Treatment with methyl-prednisolone pulse therapy was instituted, followed by immunosuppressive therapy with azathioprine and prednisone. On follow-up of less than a year patient had significant control of symptoms and improvement on autonomy. **Discussion:** Subacute sensory neuropathy (SNN) and brainstem encephalitis are included in paraneoplastic neurological syndrome (PNS) and are rarely accompanied by colon cancer. Typically, SNN is associated with small-cell lung cancer (SCLC) and involves Hu, amphiphysin, CRMP5, and PCA2 antibodies, which are present in both central and peripheral syndromes. According to a 10-year retrospective study conducted by the Mayo Clinic, out of 115,081 cancer patients, only 58 (0.05%) were diagnosed with a paraneoplastic neurological syndrome, and among these, five patients had colon cancer. These patients presented with peripheral neuropathy and a cerebellar syndrome, similar to the present case. Given the unknown antigen expression status and a follow-up period of less than two years, our patient was classified as having possible PNS according to the PNS-Care score. **Final comments:** PNS treatment targets underlying malignancies. Early neural destruction can limit clinical improvement despite treatment. Consequently, high-risk neurological phenotypes should prompt an intensive search for malignancy. It is also crucial to consider colon adenocarcinoma, although rare, as a potential cause of PNS.

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Brazilian carotid web registry - an ongoing cohort

Alessandra Braga Cruz Guedes de Morais[1], Ana Silvia Sobreira Lima Verde[2], George Nilton Nunes Mendes[3], Vitória Cristina Almeida Flexa Ribeiro[4], Adson Freitas de Lucena[2], Francisco José Arruda Mont'alverne[2], Fernanda Martins Maia Carvalho[2,4], Gisele Sampaio Silva[1,5], Fabricio Oliveira Lima[2]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]Hospital Geral de Fortaleza, Serviço de Neurologia, Fortaleza CE, Brazil.
[3]Centre Hospitalier de l'Universite de Montreal, Neurology Department, Montreal, Canadá.
[4]Universidade de Fortaleza, Fortaleza CE, Brazil.
[5]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Alessandra Braga Cruz Guedes de Morais; Email: alebcgmorais@gmail.com.

Background: Carotid web (Web-Ca) is considered an underrecognized cause of stroke. The lack of prospective studies results in a poor understanding of its clinical characteristics, recurrence rate, treatment strategies, and stroke prevention. **Objective:** We present a prospective ongoing Brazilian cohort to describe the epidemiology, treatment outcomes, and long-term prognosis of patients diagnosed with Web-Ca. **Methods:** Patients admitted to our stroke unit or attended in our stroke clinic were systematically evaluated. This involved an in-depth review of neuroimaging data (CT Angiography and/or MR Angiography) at our center, from 2017 to 2023. Two experienced stroke neurologists confirmed the diagnosis. Data on demographics, risk factors, treatment, and patient outcomes were gathered using a standardized questionnaire. **Results:** A total of 42 cases were identified (57.1% male) with a median age of 50.5 (IQR 42.25-58). Most of the patients were identified after an ischemic stroke, and the initial NIHSS median was 11 (IQR 4-15.5). Twenty-two patients (52.4%) were treated with either endarterectomy or angioplasty, and 32 (78%) were considered symptomatic. There were three cases of colorectal neoplasm associated (7.7%). In addition, Web-Ca was identified in two cases of Intracranial hemorrhage, one case of subdural hematoma, and five cases of subarachnoid hemorrhage. **Conclusion:** The ongoing Brazilian cohort study provides valuable insights into epidemiology, treatment outcomes, and long-term prognosis of patients with Web-ca. This study underscores the importance of recognizing Web-Ca in the differential diagnosis of ischemic stroke etiology and also in non-ischemic stroke patients. Suggesting the necessity for further research on its management and prevention.

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Brazilian medical students' knowledge and interest in headache and other common pain conditions: a comprehensive survey

Caroline Matielo Ricci[1], Fernanda Firmiano Casarotto[1], Isadora Miarelli Fortuna[1], Lucas da Silva Oliveira[1], Hilton Mariano da Silva Júnior[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Caroline Matielo Ricci; Email: carol.iccir@gmail.com.

Background: Pain, a pervasive issue in healthcare, necessitates that future doctors are well-versed in various pain syndromes, their origins, diagnoses, and treatments. It also seeks to evaluate their preparedness to manage patients afflicted by diverse pain types in the emergency room and outpatient settings. However, studies that aimed to estimate the knowledge and interest of medical students in primary pain syndromes, particularly headaches, are scant. **Objective:** To assess the knowledge of medical students about the main pain syndromes, to quantify the frequency of pain-related classes in medical school and identify potential gaps in the medical curriculum related to this topic. **Methods:** This is a cross-sectional and descriptive study involving medical students from various Brazilian universities. An online questionnaire was used as the data collection instrument. It included questions about demographic data (age, gender, university, year of graduation, faculty, city, and state), objective questions to assess knowledge about the main pain syndromes and questions about the frequency of pain-related classes during medical school. The questionnaire link was made available to students through the university communication channels, social media, and promotion by student organizations and responses were collected over seven months. Subsequently, the results were analyzed. **Results:** Five hundred twenty responses were obtained from undergraduate medical students from fifty-four medical universities in various states nationwide. From these responses, it is noted that although most students (90%) recognize how frequent headache complaints are in the emergency room, a large portion (56.9%) have not had classes on how to evaluate these patients initially, and about 70.6% have never participated in conferences on this complaint. In contrast, only 33.7% have not had classes in the emergency department to evaluate patients with chest pain. Similarly, only 26.7% have not learned how to approach abdominal pain in the emergency room. These data highlight the negligence of educational institutions regarding the topic of headaches compared to other types of pain frequently encountered in the hospital setting. **Conclusion:** Our results reveal a substantial knowledge gap concerning headaches and other pain syndromes in the medical education curriculum despite the high prevalence of these conditions in clinical practice. This deficiency in medical training leads to patient dissatisfaction with the healthcare system, frequent cases of malpractice, and undertreatment. Your role as future doctors is crucial in addressing this issue. Therefore, medical students must receive comprehensive headache management education, particularly in emergencies. This can be addressed by integrating dedicated classes and events into the academic curriculum to enhance proficiency in pain management in clinical practice and healthcare services.

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Breakthrough coma: a severe myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) case report

Josenice Gomes Trindade Seidel[1], Thiago Gonçalves Guimarães[1], Ahmad Kassem El Zein[1], Lucas Henrique de Moura Rogerio Garcia[1], Matheus Rodrigues Nogueira[1], Joana Laura Aihara Barbosa[1], Luiz Gustavo Brenneisen Santos[1], Tarso Adoni[1], Julia Martins Brunelli[1], Gabriela Almeida Pimentel[1]

[1]Hospital Sírio Libanês, São Paulo SP, Brazil.

Correspondence: Josenice Gomes Trindade Seidel; Email: josegomes15@hotmail.com.

Case presentation: A 40-year-old black woman was admitted to our emergency department due to a 30-day history of progressive headache, unresponsive to analgesics, associated with fever, bilateral visual deficit, bladder dysfunction, and lower limb paresthesia. She deteriorated with tetraparesis and encephalopathy, despite treatment with acyclovir and antibiotics. No previous infections, vaccinations, or neurologic symptoms were reported. Neurological examination revealed somnolence, mental confusion, and flaccid bilateral hemiparesis, ataxia, and light-near dissociation. The patient evolved with coma and required ventilatory support. Brain magnetic resonance imaging (MRI) showed multiple T2/FLAIR-hyperintense tumefactive lesions affecting the supratentorial white matter, cerebral cortex, basal ganglia, brain pedunculi, the whole brainstem, optic nerves, optic chiasma, and optic tracts, with gadolinium enhancement. Cervical MRI showed a mild non-tumefactive lesion in C2. The cerebrospinal fluid analysis revealed increased protein with lymphocytic pleocytosis and normal glucose levels, negative bacterioscopy and cultures. Oligoclonal bands were negative. No infectious agents were detected. Electroencephalogram did not reveal epileptic discharges. A comprehensive molecular panel for infectious meningitis and encephalitis came out negative. Of all rheumatological markers, only rheumatoid factor was positive. The whole-body 18F-FDG PET/CT scan showed no hypermetabolic lesions. Brain biopsy revealed an active chronic inflammatory process with necrosis, cellular debris, and secondary demyelination (predominantly perivascular lymphomononuclear inflammatory infiltrate). A Cell Based Assay (CBA) for anti-MOG was positive. Anti-aquaporin-4 antibody was negative. Acyclovir and antibiotics were discontinued. Plasma exchange and high dose methylprednisolone were initiated. The patient had a dramatic response and close to full recovery from neurological deficits. Three-month follow-up showed regression of MRI lesions. **Discussion:** MOGAD is a recently identified acquired autoimmune central nervous system demyelinating disease. It is typically associated with acute disseminated encephalomyelitis (ADEM), optic neuritis (ON) or transverse myelitis, and is less commonly associated with cerebral cortical encephalitis, brainstem or cerebellar presentations. The MOGAD spectrum is broad and varies according to age, with ADEM being more common in children, and recurrent ON and longitudinally extensive transverse myelitis (LETM) being more common in adults. MOGAD attacks needing ventilatory support are rare. Most attacks respond well to treatment. We report a case of MOGAD with ADEM phenotype in an adult patient, evolving into coma, with need of ventilatory support, an uncommon, life-threatening, complication. **Final comments:** MOGAD should be considered as a cause of coma in adult patients with inflammatory diseases of the central nervous system suggestive of ADEM.

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Bright light therapy for non-seasonal depressive disorders: a systematic review and meta-analysis

Artur Menegaz de Almeida[1], Francisco Cezar Aquino Moraes[2], Maria Eduarda Cavalcanti[3], Jorge Henrique Cavalcanti Orestes Cardoso[3], Fernanda Tamashiro[4], Celso Miranda[5], Lilianne Fernandes[6], Michele Kreuz[7], Francinny Alves Kelly[8]

[1]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.
[3]Universidade de Pernambuco, Recife PE, Brazil.
[4]Universidad de Buenos Aires, Buenos Aires, Argentina.
[5]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[6]AFM, Abaetuda PA, Brazil.
[7]Universidade Luterana do Brasil, Canoas RS, Brazil.
[8]DPIC, São Paulo SP, Brazil.

Correspondence: Artur Menegaz de Almeida; Email: arturomenegaz@gmail.com.

Background: Seasonal humor disorders are prone to have a link with daylight exposure. However, the effect of external light on non-seasonal disorders remains unclear. Evidence is lacking for the validity of Bright Light Therapy (BLT) as an adjunctive treatment for these patients. **Objective:** To assess BLT effectiveness as an adjunctive treatment for non-seasonal depressive disorders. **Methods:** In March 2024, a comprehensive search of publications in MEDLINE, Embase, and Cochrane databases for randomized clinical trials (RCTs) evaluating BLT effects in non-seasonal depressed patients. Truly randomized clinical trials published since 2000 were eligible. Comparisons between BLT and Dim Red Light (DRL) or antidepressant monotherapy alone were considered for inclusion. Using the systematic review approach on randomized controlled trials published from January 1, 2000, through March 25, 2024, differences between patients treated with and without BLT were estimated using the Mantel-Haenszel method; heterogeneity was assessed using I^2 statistics. R Studio, version 4.3.2, was used for statistical analysis. Remission of symptoms, response to treatment rates, and depression scales were assessed. **Results:** In this systematic review and meta-analysis of 11 unique trials with data from 858 patients, statistically significant better remission and response rates were found in the BLT group (40.72% vs. 23.52%; OR 2.42; 95% CI 1.503.91; p=0.0002; I^2=21%; and 60.36% vs. 38.56%; OR 2.34; 95% CI 1.463.75; p=0.0003; I^2=41%, respectively). Subgroup analysis based on follow-up times (<4 weeks; >4 weeks) also showed better remission and response rates with BLT: Remission (<4 weeks; 27.38% vs. 9.21%; OR 3.59; 95% CI 1.458.88; p=0.005; I^2=0%;) and (>4 weeks; 46.59% vs. 29.08%; OR 2.18; 95% CI 1.194.00; p=0.01; I2=47%). Response (<4 weeks; 55.55% vs. 27.35%; OR 3.65; 95% CI 1.817.33; p=0.0002; I2=35%) and (>4 weeks; 63.03% vs. 44.91%; OR 1.79; 95% CI 1.013,17; p=0.04; I2=32%). **Conclusion:** This meta-analysis established BLT as an effective adjunctive treatment for non-seasonal depressive disorders. Additionally, our results suggest that BLT may improve the time to respond to the initial treatment.

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BRPF1-associated disease mimicking congenital myasthenic syndrome

Mariana Hiromi Manoel Oku[1], Clara Gontijo Camelo[1], Cristiane de Araujo Martins Moreno[1], Raquel Diogenes Alencar Sindeaux[1], Gabriella Corrêa Dousseau[1], Gabriel Saboia de Araujo Torres[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Mariana Hiromi Manoel Oku; Email: mari.hiromi.mo@gmail.com.

Case presentation: We describe a case of a two-year-old Haitian girl who presented with bilateral ptosis since birth. She is the first child of a non-consanguineous Haitian couple, born via vaginal delivery, at term, with no history of obstetric complications. The parents reported bilateral eyelid ptosis with mild fluctuation and weak crying since her first weeks of life. There was no description of feeding difficulties, choking spells, hypotonia, or respiratory distress. She was diagnosed with cystic fibrosis after neonatal screening and a confirmatory chloride sweat test. Neurological exam at 11-month-old showed mild generalized hypotonia and weakness with axial involvement. There was no social/cognitive delay. In addition, there was bilateral partial ptosis without clear fatigability. Brain MRI was unremarkable and electromyography with repetitive nerve stimulation showed no signs of neuromuscular transmission dysfunction. Creatine phosphokinase levels were normal. Pyridostigmine was prescribed with no neurological improvement. Genetic testing revealed a pathogenic mutation at the BRPF1 gene, confirming the diagnosis of BRPF1-associated intellectual development

disorder with dysmorphic facies and ptosis. **Discussion:** BRPF1 gene mutations and consequent BRPF1 haploinsufficiency have been described to result in decreased H3K23 acetylation which leads to epigenetic deregulation and impaired development. The typical phenotype consists of ptosis, hypotonia/weakness associated with dysmorphic facies, intellectual developmental disorder, and epilepsy. Nevertheless, milder phenotypes with congenital ptosis and normal intellectual development have been reported. Bilateral ptosis and fatigable weakness are typical findings in congenital myasthenic syndrome (CMS). There is a wide clinical variability ranging from mild symptoms such as exercise intolerance or fluctuating ptosis to severe phenotypes with disabling weakness, bulbar symptoms, and respiratory insufficiency. The age of presentation (neonatal, childhood, adult-onset), course of the disease, and response to acetylcholinesterase inhibitors are also variable, which leads to a broad differential diagnosis, especially in childhood-onset forms. We present a case of a rare genetic disease that can mimic CMS and therefore should be included as a differential diagnosis in patients presenting with ptosis and weakness. **Final comments:** BRPF1-associated disease can mimic CMS and should be considered as a differential diagnosis even in children with mild or no intellectual development disorder.

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Bulbar hematoma presenting as Wallenberg syndrome due to possible radiotherapy-induced vasculopathy as an atypical etiology: a case report

Alex de Novais Batista[1], João Herculano Lins[1], Maria Clara Arcoverde Santana[1], Virgílio Sátyro da Nóbrega Segundo[1], João Eudes Magalhães[1], Letícia Nunes Campos[1]

[1]Universidade de Pernambuco, Recife PE, Brazil.

Correspondence: Alex de Novais Batista; Email: alexnovaisb@gmail.com.

Case presentation: A 38-year-old woman presented to our outpatient clinic in late 2023 with left-sided limb weakness and numbness. Initially, she presented with hiccups, which lasted for four days and progressed to numbness and weakness in the left upper and lower limbs, visual disturbance in the left eye, and a staggering gait. Because of these symptoms, the patient sought emergency care. However, stroke features were not recognized in the computed tomography scan and no thromboembolic treatment was prescribed. Regarding her medical history, the patient underwent chemotherapy and radiotherapy for nasopharyngeal cancer from 2006 to 2007. As a result, she developed hypothyroidism and hypercholesterolemia, which were adequately treated with levothyroxine and rosuvastatin. The patient also presented a right foot drop due to Hansen's disease, which she was treated for in 1998. Neurological examination revealed left hemifacial hypoesthesia, impaired left corneal reflex, mild left trapezius weakness, and mild tongue deviation to the left. Other findings included incomplete left hemiparesis (grade 4), hemihypoesthesia, decomposing finger-to-nose movement, and hyperreflexia. A brain magnetic resonance imaging (MRI) showed a right-sided, elongated, lobulated hemorrhagic lesion at the bulbomedullary junction extending to the C3 level (2.5 x 1.4 cm). The patient was diagnosed with Wallenberg syndrome due to a bulbar hemorrhagic stroke. In early 2024, the patient demonstrated significant strength improvement (grade 5). Follow-up brain angiography and echocardiogram were unremarkable, while lab tests showed hypercholesterolemia. Although she had atherothrombotic factors, the stroke was attributed to vasculopathy induced by her previous radiotherapy, which probably affected microvessels after a long interval. Six months after the stroke, a new brain MRI showed complete resorption of the hematoma with gliosis. **Discussion:** Wallenberg syndrome results from infarction or hemorrhage in the lateral portion of the medulla oblongata, causing patients to experience vertigo, dysphagia, hoarseness, hiccups, ipsilateral facial hypoesthesia, and contralateral body sensory deficits. Common etiologies for the syndrome include vertebral artery occlusion, posterior inferior cerebellar artery occlusion, and vertebral artery dissection. None of these causes were found in our patient. Radiation-induced vasculopathy is a known but rare complication that can lead to cerebrovascular events years after treatment, often involving large vessels. Furthermore, the presence of a hematoma, as opposed to ischemia, is likely due to vessel wall fragility from an actinic lesion. **Final comments:** In this case, radiation-induced vasculopathy was a possible cause of the bulbar hemorrhagic stroke leading to Wallenberg syndrome. The atypical etiology and stroke nature highlight the importance of considering cerebrovascular disease as a long-term complication of cancer therapy.

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CADASIL presenting with Foix-Chavany-Marie syndrome: a case report

Beatriz Pires Paes[1], Bruna da Silva Lima[1], Poliana Lima Campos[1], Beatriz Ballarin Costa[1], Anthony Magalhães Morais Santiago[1], Camilo Carvalho[1], Larissa Lucas Schloicka[1], Andre Gustavo Fonseca Ferreira[1], Pedro Manzke de Carvalho[1], Plínio Rodrigo Máximo Macêdo[1]

[1]Hospital de Base do Distrito Federal, Brasília DF, Brazil.

Correspondence: Beatriz Pires Paes; Email: beatrizpirespaes@gmail.com.

Case presentation: A 49-year-old man, cocaine user, alcoholic, and smoker, presented initially with dengue fever symptoms, with a positive serology. After three days, these symptoms were improving. However, he progressed within 24 hours to bilateral facial palsy, severe dysphagia, sialorrhea, hypophonia, and significant dysarthria. He also developed psychomotor agitation and emotional lability that did not improve with the use of antipsychotics, and persisted during the entire month of hospitalization. Infectious-metabolic causes were ruled out and lumbar puncture was within normal limits. The initial hypothesis was a post-infectious autoimmune response (rhombencephalitis). Therefore, we performed five days of pulse therapy with methylprednisolone, without much improvement. After two weeks, we carried out five plasmapheresis sessions, but the patient evolved with only partial betterment. Magnetic Resonance Imaging (MRI), only available one month after hospitalization, revealed in the T2 sequence multiple confluent hyperintensity foci with involvement of the white matter of the cerebral hemispheres predominantly in temporal lobes, and foci of heterogeneous contrast enhancement in the posterior topography of the left corona radiata, centrum semiovale and right frontal periventricular centers in a perivenular course. Throughout hospitalization, a family history of frequent neurovascular events was evident, with three brothers having previous histories of ischemic stroke, one of which was diagnosed with CADASIL at 60 years old. **Discussion:** Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is a rare monogenic cerebral small vessel disease (SVD). As with all SVDs, CADASIL manifests on brain MRI with various combinations of white matter hyperintensities, lacunar infarcts, microbleeds, and brain atrophy, predominantly in locations such as anterior temporal poles, the basal ganglia and thalamus, and superior frontal gyri. It usually affects young or middle-aged adults, and it's typically characterized by migraine, subcortical ischaemic events, mood disturbances, and cognitive impairment. This condition can have a myriad of different presentations, and in this case, the anterior operculum was involved bilaterally. The Foix-Chavany-Marie syndrome (FCMS), also known as anterior opercular syndrome, is a rare neurological condition that presents with voluntary facial, pharyngeal, lingual, and masticatory paralysis bilaterally. FCMS can be differentiated from other conditions, such as bulbar paralysis and cranial nerve palsies by intact extraocular movements, preserved brainstem reflexes, and the absence of lower motor neuron signs. **Final comments:** CADASIL remains a diagnostic challenge, especially with atypical manifestations, such as presented in this patient with FCMS. This syndrome was consistent with our patient's presentation since he had central facial palsy, dysphagia, and sialorrhea.

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Calcinosis of the cervical spine: a cause of myelopathy in crest syndrome

Natália Contini[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Natália Contini; Email: nataliacontini4@gmail.com.

Case presentation: A 53-year-old patient with CREST (Calcinosis, Raynaud's phenomenon, Esophagopathy, Sclerodactyly and Telangiectasias) syndrome for the past 30-years presented with slowly progressive weakness in her lower limbs and vesical incontinence. She had been treated with prednisone 20mg/day and azathioprine 150mg/day. Magnetic resonance imaging demonstrated severe compression at cervical spine due to a spinal tumoral calcinosis, with signs of myelopathy. The patient refused surgical intervention due to increased surgical risk. She remains clinically stable two year after diagnosis, without progression of her neurological deficits. **Discussion:** CREST syndrome is an acronym for calcinosis, Raynaud's phenomenon, esophageal dysfunction, sclerodactyly and telangiectasias. Tumoral calcinosis is a classical finding of this syndrome, however, it has only rarely been reported to cause myelopathy. Here, we report a 54-years-old woman with CREST syndrome who presented with slowly progressive spastic paraparesis and urinary incontinence, in whom diagnostic investigation demonstrated spinal tumoral calcinosis of the cervical spine. This diagnosis should be suspected in CREST patients presenting with spinal cord symptoms. Tumoral calcinosis is characterized by focal or multifocal soft tissue calcification, occurring usually in periarticular tissue or justa-articular regions of the extremities. Calcinoses occur in a dystrophic fashion in this disorder, where calcification is promoted in damaged or devitalized tissues in patients with normal calcium levels. It is common in areas that experience recurrent microtrauma. Thus, calcinosis of the spine is being recognized, as the spine is a common place for microtrauma, due to the extensive flexibility and susceptibility of some areas to degenerative disc disease and intervertebral disc herniation. Spinal tumoral calcinosis is defined as tumor-like deposits of calcium usually involving both the vertebral bodies and the posterior elements of the cervical, thoracic, and lumbar spine. It is a rare manifestation of CREST syndrome. Differential diagnosis remains difficult, as it may mimic neoplasms and infections. However, in CREST syndrome presenting with progressive paraparesis and spinal masses, this diagnosis should be primarily considered, especially in the absence of infection. Although patients may be asymptomatic, surgical resection remains the only effective treatment for patients with progressive symptoms. In these cases, surgery can be recommended, as the patient can be symptomatic and/or have extensive destruction of the vertebrae and surrounding structures. **Final comments:** Calcinosis of the spine can cause morbidity in patients with CREST syndrome. When intervention is necessary, surgical resection with spinal cord decompression and fusion can be effective in managing symptoms and preventing further spinal cord injury.

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Cannabidiol for acute stroke treatment: cast study

Millene Rodrigues Camilo[1], Jaime Eduardo Cecílio Hallak[1], Priscilla Queiroz de Lima[1], Alline Cristina de Campos[1], Rodrigo Bazan[2], Maurício Yonamine[3], José Alexandre de Souza Crippa[1], Octávio Marques Pontes-Neto[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade Estadual Paulista, Botucatu SP, Brazil.
[3]Universidade de São Paulo São Paulo SP, Brazil.

Correspondence: Millene Rodrigues Camilo; Email: millene.camilo@gmail.com.

Background: Cannabidiol is a non-psychoactive phytocannabinoid derived from Cannabis sativa. In animal models, it has been demonstrated that cannabidiol is associated with reduced in cerebral infarction volume, increased blood flow, and modulation of neuroinflammatory processes. Although clinical studies have already shown good tolerance and few adverse effects, none have yet been carried out to evaluate the safety and efficacy of cannabidiol in acute stroke patients. Primary **Objective:** To assess the safety of the use of cannabidiol when administered to patients with ischemic stroke within the first 7 days of the onset of symptoms. Secondary objectives (1) evaluate the functionality and severity of the stroke using the modified Rankin scale and the NIHSS 90 days after the stroke; (2) assess anxiety, depression, and drowsiness through specific questionnaires on admission, on the 7th day or hospital discharge and after 90 days; and (3) evaluate the final volume of cerebral ischemia. **Methods:** This is a phase II clinical trial of intervention, double-blind, randomized, multicenter, prospective, placebo-controlled to evaluate the safety and efficacy of oral treatment with cannabidiol in doses of 150mg, 300mg, and 600mg in the first 7 days of evolution of an acute ischemic stroke. Randomization will be stratified by stroke center and severity (NIHSS <10 or >10). A total of 80 participants will be included, 20 in each group: placebo, 150mg, 300mg, and 600mg of cannabidiol. **Results:** The use of cannabidiol in ischemic stroke patients will likely be safe and points to possible neuroprotection, allowing the continuation of a phase III study.

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Cannabidiol in the cognitive neurology of aging: current evidence, challenges and future prospects

Tereza Raquel Xavier Viana[1], José Eduardo Martinelli[1]

[1]Faculdade de Medicina de Jundiaí, Jundiaí SP, Brazil.

Correspondence: Tereza Raquel Xavier Viana; Email: espacotr@hotmail.com.

Background: Population aging represents a growing challenge for global health, with a corresponding increase in neurodegenerative diseases affecting cognition. Cannabidiol (CBD), a non-psychoactive component of Cannabis sativa, has emerged as a potential therapeutic agent in the cognitive neurology of ageing, due to its anti-inflammatory, antioxidant and neuroprotective properties. **Objective:** To investigate the current evidence of CBD in the cognitive neurology of aging, identifying potential benefits, methodological challenges and future prospects as a therapeutic intervention to improve cognitive function in the elderly and mitigate the impacts of age-related neurodegenerative diseases. **Methods:** This is an exploratory and descriptive systematic review conducted according to the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) and PICO (Population, Intervention, Comparison, Outcome) guidelines. Searches were carried out in the PubMed, SciELO, Cochrane Library and BVS databases, using the descriptors: "Cannabidiol" and "Cognitive Impairment" AND "Neurodegenerative Diseases". We included studies published in the last 5 years (2019 - 2024), in English and Portuguese, which investigated the effects of CBD on older people with cognitive impairment, dementia or age-related neurodegenerative diseases. Experimental studies, controlled clinical trials and systematic reviews were considered for inclusion. **Results:** During the initial search, nine articles were identified, of which one was a duplicate. After reviewing the titles, abstracts and conclusions of the remaining eight, five were excluded because they did not meet the inclusion criteria. Three studies were selected for full reading and inclusion in this research. These studies highlight that CBD has the potential to mitigate neuroinflammatory processes, reduce oxidative stress and modulate neuronal plasticity in brain ageing. Pre-clinical evidence indicates benefits in cognitive function and improved quality of life in the elderly. However, variations in methodological quality and the dosage used highlight the need for more robust research and well-controlled clinical trials. **Conclusion:** This study highlights the growing body of evidence supporting the therapeutic potential of CBD in the cognitive neurology of ageing. Although promising, the use of CBD faces significant challenges, including the need for longitudinal studies, standardization of formulations and dosages, and accurate understanding of its mechanisms of action. Future research should focus on filling these knowledge gaps, with a view to translating pre-clinical findings into tangible clinical benefits for an increasingly large and vulnerable ageing population.

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Cannabinoid treatment for autism spectrum disorder (ASD): a systematic review with single-arm meta-analysis

Jonas Leite de Souza Filho[1], Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Maria Antônia Oliveira Pereira[2], Victor Gonçalves Soares[3], Celli Veloso Cavalcanti[1], Dário Correia Negreiros[1], Laura Martins Soares Cortez[1], Gielson de Sousa Silva[1], Lara Beatriz Alves Batista[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Universidade Estadual do Piauí, Teresina PI, Brazil.
[3]Universidade Federal dos Vales do Jequitinhonha e Mucuri, Diamantina MG, Brazil.

Correspondence: Jonas Leite de Souza Filho, Email: jonaslf25@gmail.com.

Background: Autism spectrum disorder (ASD) is a complex neurodevelopmental condition. It can exhibit a range of symptoms, from deficits in communication and social interaction to repetitive disorders and stereotypical behaviors. Currently, despite increased prevalence of this condition, drug therapy is still limited. The use of cannabinoid chemical compounds, derived from the cannabis plant, has shown potential as a therapeutic alternative for individuals with ASD. **Objective:** The present study aims to evaluate the effectiveness of cannabinoid treatment on behavioral symptoms of ASD. **Methods:** We systematically searched PubMed, Embase, and Cochrane for studies reporting. All included studies used plant extracts containing the same concentrations of Cannabidiol and Tetrahydrocannabinol in a 20:1. The outcomes of interest were mean change from baseline for the Autism Parenting Stress Index (APSI), Home Situation Questionnaire-ASD (HSQ), and the prevalence of success regarding anxiety problems and behavioral problems. Mean changes and prevalences were pooled using a random-effects model with 95% confidence intervals (CIs). We used R software version 4.3.2 and the extension package "meta" for all statistical analysis. **Results:** Five studies were included, consisting of one randomized clinical trial, two retrospective, and two prospective, involving a total of 469 patients. The mean change from baseline for the APSI outcome was -0.66 (95%CI -0.85 to -0.47; I^2=0%) and for the HSQ outcome was -1.35 (95% CI -1.78 to -0.92; I^2=0%). The prevalence of success regarding anxiety problems was 37.65% (95%CI 29.73% to 45.56%; I^2=0%). The prevalence of success regarding behavioral problems was 51.13% (95%CI 34.84% to 67.41%; I^2=84%). **Conclusion:** We observed the effects of cannabinoid treatment on behavioral symptoms of ASD, with positive effects on anxiety and parental stress. However, due to the scarcity of blinded clinical trials with control groups, there is still limited evidence to justify its widespread use, making further studies necessary. Furthermore, as the scales used assessed behavioral symptoms, there were no observable positive effects on core ASD symptoms.

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Cardiac calcified amorphous tumor-induced ischemic stroke treated with intravenous thrombolysis: a case report

Pedro Almeida Martins Pontes[1], Igor Izoton[1], Richardson da Silva Nogueira[1], Gunnar Willy Pereira Crepaldi[1], Henrique de Moraes Bernal[1], Ingrid Lorena da Silva Gomes[1], Giovanni Caniçali Simões[1], Werley de Almeida Januzzi[1], Marcele Schettini de Almeida[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Pedro Almeida Martins Pontes; Email: pedroalmeida.pontes@gmail.com.

Case presentation: A 69-year-old woman with a history of smoking, obesity, hypertension, cardiac arrhythmia, and a report of a transient ischemic attack one year ago presented with sudden weakness in her left leg and numbness throughout her entire left side. She was admitted three hours after the onset of symptoms and stroke protocol was initiated. Her neurological examination revealed a National Institutes of Health Stroke Scale (NIHSS) score of five points (4.1, 6A.4), with no movement in the left leg. Brain computed tomography (CT) showed no acute brain lesion, and angio CT did not reveal any major vessel occlusion. The patient underwent intravenous thrombolysis without complications, maintaining the same NIHSS score at its conclusion. Her electrocardiogram revealed a left bundle branch block. A point-of-care ultrasound (POCUS) was also performed. In the apical 4 chamber view, there was an irregular hyperechoic image, on the basal inferolateral wall of the left ventricle, attached to the mitral annulus, adjacent to the posterior mitral leaflet. Due to the suspicion of a cardioembolic stroke mechanism caused by a cardiac mass, further tests were conducted. Transthoracic echocardiogram revealed an amorphous, irregular nodular image, measuring 3.0 x 2.5 cm, suggestive of a calcified tumor with necrotic content inside. Cardiac MRI showed a nodular formation on the basal inferolateral wall, adjacent to the mitral valve, measuring 2.8 x 4.2 x 2.5 cm, showing iso-signal on T1 and T2 and heterogeneous late enhancement. Brain MRI showed diffusion restriction in the right frontal and parietal lobes. At the time of discharge, the patient had grade IV strength in her left leg and no facial paralysis (NIHSS 1 point). In joint management with Cardiology, her surgical risk was considered too high at the moment, therefore a diagnosis of probable Cardiac Calcified Amorphous Tumor (CAT) was made. She was discharged with oral anticoagulation for secondary stroke prophylaxis. **Discussion:** To our knowledge, this is the first case of stroke caused by CAT that was treated with intravenous thrombolysis. Cardiac CAT is a rare type of non-neoplastic cardiac tumor, occasionally associated with cardioembolic stroke. It is suspected that the embolization mechanism of these tumors is related to either a part of the mass itself detaching or a

thrombus attached to the mass causing emboli. Surgical treatment is usually performed, involving excision of the mass. However, there are reports of patients with contraindications to surgery who underwent conservative treatment, with periodic monitoring and anticoagulation, who remained free of recurrence of strokes. **Final comments:** More studies are needed to better establish the risks of stroke in the population with CAT, as well as the appropriate treatment for preventing new cerebrovascular events in this disease.

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Cardioembolic ischemic stroke in a young man with delayed diagnosis of patent foramen ovale: a case report

Nicoli Lindissey Zuim[1], Amanda Stefani Fernandes Donon[1], Carolaine Cristina Quirino[1]

[1]Universidade Nove de Julho, Bauru SP, Brazil.

Correspondence: Nicoli Lindissey Zuim; Email: nicolind14@gmail.com.

Case presentation: GCR, 25 years old, male, presented to the Emergency Department of the Clinical Neurology Department with a complaint of headache in the nape region, which was more intense on the right side, associated with fainting. Unremarkable general physical examination. Neurological examination revealed upper foveal, hemiplegia, temporal hemianopia, and hypoesthesia, all on the left with a National Institutes of Health Stroke Scale (NIHSS) score of 18 on admission. Further tests were ordered and revealed abnormalities: non-contrast brain computed tomography (CT) with loss of cortico-subcortical differentiation in the putamen, M1, and M2 territories, as well as a cerebral/neck angiogram showing dissection of the right internal carotid artery with subocclusion in M1 on the right. The patient reported previous surgery 45 days ago for a left ankle injury and denied any further comorbidities. He reported a family history of cerebrovascular events. With a presumptive diagnosis of ischemic stroke of large vessels with Tandem occlusion, mechanical thrombectomy intervention was initiated for the patient. In 24 hours, the patient developed intracranial hypertension and delineation of the ischemic area in the territory of the right middle cerebral artery. Consequently a right decompressive craniectomy was performed. To investigate the possible causes of the event, a transesophageal echocardiogram was requested. It revealed a patent foramen ovale (PFO), with passage of macrobubbles between the right and left chambers after Valsalva maneuver. In addition, lower limb venous Doppler confirmed deep vein thrombosis of fibular veins in the middle and proximal third of the left lower limb, providing a potential source of emboli. The findings allowed the etiology of the ischemic stroke to be elucidated. **Discussion:** In the presented case, the complexity of cerebrovascular events is evident, as well as the importance of a multifaceted diagnostic and therapeutic approach during follow-up. The suggestive picture of a significant neurological event was corroborated by the findings on neurological examination, as well as by imaging tests and the NIHSS score of 18, indicating a severe stroke due to the prothrombotic state and a genetic predisposition to vascular events. Therefore, the combination of patent foramen ovale and deep vein thrombosis indicates a paradoxical and fortuitous embolic mechanism, where a venous thrombus passes to the arterial system through the patent communication between the chambers of the heart, resulting in a malignant and complexly evolving ischemic stroke. **Final comments:** In summary, it is important to emphasize the need for a comprehensive approach to the diagnosis of stroke in young patients, especially when they have atypical risk factors or a medical history suggestive of prothrombotic states. Prompt intervention combined with a thorough etiological investigation is crucial for effective management and prevention of future events.

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Carotid artery stenosis after 13 years of radiotherapy for head and neck cancer: a case report

João Henrique Reis Duque Estrada[1], Caroline Vianna Costa Pinto[1], Luiza Guimarães Póvoa[1], Victor Waldhelm Cozer[1], Diogo Terrana[1], Felipe Schmidt[1], Sofia Mermelstein[1]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: João Henrique Reis Duque Estrada; Email: joaohestrada2@gmail.com.

Case presentation: A 58-year-old male patient, hypertensive, with a history of laryngeal cancer treated with ionizing radiotherapy in 2010, presented to the emergency department of his municipality with a right brachio-crural hemiplegia, non-fluent aphasia, and general malaise with symptom onset of approximately 1 hour and 30 minutes. Despite no contraindications, reperfusion therapies were not performed. He was then transferred to a tertiary hospital for etiological investigation, where he underwent magnetic resonance imaging of the head and neck, as well as other complementary exams, which showed severe right carotid artery stenosis, suggestive of actinic involvement. He underwent carotid angioplasty with non-pharmacological stent placement, without complications, and was discharged 24 hours later. In addition to the diagnosis of actinic carotid artery stenosis, the patient started treatment for primary hypothyroidism of probable actinic etiology after initial exams. **Discussion:** Carotid artery stenosis is an important late complication in patients who have undergone head and neck radiotherapy. This complication sometimes presents with ischemic stroke, occurring years after radiotherapy, as seen in the present case. The incidence of cerebrovascular events ten years post-irradiation is prevalent in up to 34% in large studies. With increasing survival rates among patients undergoing radiotherapy for head and neck tumors, actinic carotid artery stenosis has become an important topic for discussion. Actinic carotid stenosis is characterized by vascular obstructions usually distal with diverse pathophysiology, which can be explained by endothelial vascular injury, vasa vasorum injury, and accelerated atherosclerosis, with extensive fibrosis of the media and adjacent tissues, sometimes accompanied by atheromatous plaques, which often confuses the etiological diagnosis. According to the literature, the main therapeutic approach for actinic carotid stenosis is angioplasty with stent rather than surgical endarterectomy proposed for most cases, due to the adherence of the actinic lesion to the muscular planes and adjacent fascia, besides the high prevalence of restenosis after endarterectomy in these patients. Even though there is evidence of associated atheromatous plaques, these are usually disproportionately smaller than the fibrotic component. **Final comments:** The high incidence of actinic carotid stenosis after head and neck radiotherapy is a condition often underdiagnosed due to its atheromatous component. Better understanding of these lesions is necessary to assist in prevention, diagnosis, and choice of the most appropriate therapeutic approach.

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Carotid web and ischemic stroke in young adult: a case report

Brenda Leticia Lopes Batista[1], Barbara Borges de Oliveira[1], Daniel Duarte Rolim[1], Victor Souza Torres de Vasconcelos[1], Mariana Holanda Gameleira[1], Igor Santos Sales[1], Ana Dolores Firmino Santos do Nascimento[1], Ubiratan Alves Viturino da Silva[1], Ana Rosa Melo Correa Lima[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Brenda Leticia Lopes Batista; Email: brendalopes.academico@gmail.com.

SLNL, 44 years old, female, massage therapy professional, married, admitted to the neurological emergency department of a tertiary hospital with a history of right-sided hemiparesis, aphasia and combined gaze deviation to the left with a sudden onset lasting 3 hours and 30 minutes. On admission, she underwent a head tomography (CT) scan without acute injuries, with an Alberta Stroke Program Early CT Score (ASPECTS) of 10 points. In the absence of contraindications, as well as within the therapeutic window, chemical thrombolysis was performed with alteplase, with slight gingival bleeding. The patient had an initial NIHSS of 20 points, with a final NIHSS of 9 points, showing partial improvement in strength deficit and aphasia. The patient remained in the stroke unit and underwent control neuroimaging the following day, showing hypodensity in the territory of the left middle cerebral artery. The patient was discharged from the stroke unit for follow-up in the neurology ward, where the etiological investigation continued. Hypertensive patient with recent diagnosis, dyslipidemic, overweight and without other comorbidities. She evolved with significant improvement, presenting some phonemic and semantic paraphasias during hospitalization. During the etiological investigation, she underwent magnetic resonance imaging of the brain that showed diffusion restriction with hypersignal on T2/FLAIR affecting the cingulate gyrus, precuneus, insula and left frontoparietal opercular region without post-contrast enhancement, compatible with recent ischemia. Remaining unchanged. Usg doppler of carotid and vertebral arteries without changes. However, resonance angiography of intracranial vessels showed irregularities in the posterior wall of the origin of the left internal carotid artery, insinuating itself into the lumen, compatible with the same side of the ischemiated territory; suggesting carotid membrane or carotid web. Patient was released home and returned for elective angioplasty with carotid stent to treat carotid web. She was again discharged from hospital with a final NIHSS of 3 points, using dual antiplatelet therapy with acetylsalicylic acid (ASA) and clopidogrel, in addition to a high-potency statin. Carotid web is a rare event, often undiagnosed, representing a risk of recurrence for stroke in the absence of other risk factors. The webs are protrusions into the lumen of the vessel, similar to a shelf, and can be considered a variant of fibromuscular dysplasia. Regarding treatment, there are favorable clinical results with both angioplasty and endartectomy, although there are few reports describing angioplasty with stent. For this reason, there must be a high clinical suspicion in young patients, without defined causes, in cases of cryptogenic stroke, for timely diagnosis and prevention of recurrences and future disabilities.

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Carotid web under an ulcerated carotid atherosclerotic plaque: a case report

Pedro Almeida Martins Pontes[1], Giovanni Caniçali Simões[1], Henrique de Moraes Bernal[1], Ingrid Lorena da Silva Gomes[1], Guilherme Lago Miranda[1], Tainá Fátima Ramos Gonçalves[1], Mateus Paquesse Pellegrino[1], Marcele Schettini de Almeida[1], André Luiz de Rezende[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Pedro Almeida Martins Pontes; Email: pedroalmeida.pontes@gmail.com.

Case presentation: A 49-year-old woman, with a history of hypertension and diabetes, presented with a sudden onset of aphasia. She was admitted to the emergency room with aphasia and dysarthria and a NIHSS score of 9 points. A brain computed tomography (CT) showed a hypodense area in the left frontal region, associated

with regional bleeding. There was a report of a previous event with a similar presentation one month ago, when she was diagnosed with an ischemic stroke of undetermined mechanism, and was prescribed dual antiplatelet therapy (DAPT) and high-potency statins. Angio CT revealed a mild (less than 50%) stenosis in the left carotid bulb region. Digital Angiography confirmed this suspicion, showing an ulcerated plaque in the left carotid bulb with adjacent contrast retention, without significant stenosis. Cardiac and laboratory tests were unremarkable. Based on these findings and CCS stroke analysis, stroke etiology and mechanism were defined as possible large vessel atheroembolism secondary to the aforementioned atherosclerotic plaque. She was again discharged on DAPT and statins. Over the next two months, despite proper adherence to therapies, she experienced three new episodes of sudden neurological deficits, characterized by aphasia and right-sided hemiparesis. All events were confirmed by Brain MRI as new ischemic strokes. A new Angio CT showed an occlusion of the middle cerebral artery in the distal M1 region. Reperfusion therapies were not performed, due to arrival outside of the appropriate time window. Initially, a new angiography was performed with an identical report to the first one. On the third angiography, a carotid web was identified, with an ulcerated atherosclerotic plaque on its surface at the left carotid artery bifurcation. Thus, a diagnosis of ischemic stroke secondary to carotid web associated with an ulcerated atherosclerotic plaque was established. Angioplasty was performed, and DAPT was maintained. The patient has been free from new ischemic events for 16 months, with progressive rehabilitation of previous deficits. **Discussion:** Carotid web is a shelf-like lesion that typically occurs on the inner wall of the carotid bulb and is suspected to be an intimal variant of fibromuscular dysplasia. The protrusion of this lesion into the carotid artery lumen causes blood stasis, increasing the risk of thrombus formation and subsequently the risk of stroke. Retrospective studies show a risk of up to 40% of recurrence of stroke, depending on the choice of drug prophylaxis. The most effective treatment for these lesions is not completely known, but angioplasty appears to be superior to DAPT and to anticoagulation. **Final comments:** Carotid web is a condition that should be investigated as a possible cause of ischemic stroke, especially in young patients without a defined etiology for the event or those who experience recurrence despite optimized clinical treatment.

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Carotid web: a rare etiology of ischemic stroke

Ana Luísa dos Santos Carregosa[1], Eduarda Leitzke[1], Carlos Eduardo Marisco Ceretta[1], Robert Luís Kern[1], Pedro Damassini[1], Gustavo Roberto Minetto Wegner[1], Luciana Estacia Ambros[1]

[1]Universidade de Passo Fundo, Passo Fundo RS, Brazil.

Correspondence: Ana Luísa dos Santos Carregosa; Email: carregosaluisa@gmail.com.

Case presentation: A 62-year-old hypertensive female presented to the emergency department with Broca's aphasia, ataxia, and swallowing difficulty, which began upon waking 3 hours prior. She exhibited difficulty following commands and had an NIH Stroke Scale (NIHSS) score of 6 upon arrival. The patient denied smoking and is allergic to azithromycin. Neurological examination revealed preserved cranial nerves with a slight deviation of the labial commissure, intact sensation, and preserved symmetric muscle strength in all four limbs. Cranial computed tomography (CT) revealed a left frontoparietal hypodense area consistent with stroke, along with a hyperdense middle cerebral artery (MCA) sign. As it was a wake-up stroke, cranial magnetic resonance imaging (MRI) was performed, showing no mismatch, rendering the case ineligible for thrombolysis. Subsequent magnetic resonance angiography revealed proximal occlusion of the left middle cerebral artery at the M1-M2 junction. Percutaneous mechanical thrombectomy was performed, along with full heparinization, followed by prophylactic heparin and dual antiplatelet therapy. Follow-up CT scan showed hemorrhagic transformation in the territory of the left MCA extending into the subarachnoid space, which was later reabsorbed without new lesions. Additional examinations demonstrated a small concentric plaque in the left carotid artery, with no other abnormalities. Cervical and intracranial CT angiography revealed a membrane at the transition from the left common carotid artery to the left internal carotid artery, likely representing fibromuscular dysplasia, suggestive of a "carotid web". The patient was discharged with follow-up by speech therapy and physiotherapy, along with instructions regarding signs of severity. **Discussion:** Carotid web is a rare cause of ischemic stroke, defined as an intimal fibromuscular dysplasia found in the carotid bulb or the beginning of the internal carotid artery. Epidemiologically, its incidence in ischemic stroke is approximately 1.2%, more common in men and individuals of African descent, and typically presents unilaterally. Histologically, it lacks calcification and atherosclerosis, with its pathophysiology stemming from disturbed normal laminar flow and increased platelet aggregation. Diagnosis is made through imaging modalities such as digital subtraction angiography, computed tomography angiography, magnetic resonance imaging, and ultrasound. **Final comments:** Despite its rarity, understanding of carotid web is crucial as it constitutes a part of the etiological differential diagnoses of ischemic stroke, especially in patients without atherosclerotic disease or cardiac arrhythmias. Diagnosis through imaging modalities such as angiography is essential for appropriate management and follow-up.

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Carotid web: an atypical and underrecognized cause of ischemic stroke

Melissa Fernandes Vilela de Freitas[1], Natália Rebeca Karpejany[2], Marx Lima de Barros Araújo[2], Tibério Silva Borges dos Santos[2], Sabrina Borges Batista[1], Danielle Cortellazzi Colonna Romano[1], Taianara Sampaio Reis[2], Gabriela Yumi Nakamura[1], Marta Maria da Silva Lira Batista[2], Amanda Melo Barradas[2]

[1]Universidade Nove de Julho, São Paulo SP, Brazil. [2]Universidade Federal do Piauí, Hospital Universitário, Teresina PI, Brazil.

Correspondence: Melissa Fernandes Vilela de Freitas; Email: melissa.fernandesvf1@gmail.com.

Case presentation: A 37-year-old woman presented with a sudden onset of numbness and weakness affecting her left side. She had no history of hypertension, diabetes, dyslipidemia, smoking, cardiomyopathy, or other conditions. Initial non-contrast brain computed tomography (CT) and magnetic resonance imaging (MRI) revealed a non-lacunar acute ischemic stroke in the right anterior circulation. Cardiac evaluation included an electrocardiogram (ECG) to check for arrhythmias and an echocardiogram to assess cardiac function and detect potential sources of emboli; however, no sources of emboli or significant vascular pathology were identified. Given the negative initial work-up, a digital subtraction angiography (DSA) was performed. This detailed imaging revealed a carotid web (CW) in the ipsilateral carotid bulb associated with the stroke. Other cervical arteries, including the left carotid and both vertebral arteries, appeared normal. The patient was started on dual antiplatelet therapy with acetylsalicylic acid and clopidogrel to prevent further strokes and prepared for a future stenting procedure. **Discussion:** A CW is an intimal variant of fibromuscular dysplasia with recently recognized implications in patients with cryptogenic ischemic stroke. Its hallmark is a focal shelf-like intraluminal protrusion in the posterior aspect of the carotid bulb. CW is considered a rare entity and a significant proportion of cases have been associated with recurrent ischemic strokes, most frequently in younger adults who lack other known risk factors. It is best demonstrated and diagnosed on CT angiography (CTA) of the neck because of its ability to resolve calcium and create multiplanar reconstructions. Although they can be readily visualized on CTA, carotid webs may be missed or misinterpreted because they do not typically cause hemodynamically significant stenosis and can mimic arterial dissection, non-calcified atherosclerotic plaque and intraluminal thrombus. Options for management include antiplatelet therapy, carotid endarterectomy and carotid artery stenting. Modern management strategies for cryptogenic stroke include long-term cardiac monitoring, further investigation for structural cardiac disease and a diagnostic workup for arterial hypercoagulability, however, these strategies are not likely to capture the possibility of a carotid web. **Final comments:** This case underscores the importance of considering a CW as a potential cause of cryptogenic ischemic stroke, especially in younger adults without typical risk factors. It highlights the necessity of thorough vascular imaging and the consideration of CW in stroke patients lacking other risk factors. The patient's management with dual antiplatelet therapy illustrates the critical role of tailored treatment strategies in such cases. Comprehensive diagnostic and therapeutic approaches are essential for improving patient outcomes in rare but impactful conditions like CW.

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Carotid-cavernous fistula with drainage to the cavernous sinus: a case report and literature review

Carlos Eduardo Marisco Ceretta[1], Ana Luísa dos Santos Carregosa[1], Eduarda Leitzke[1], Tami Zang Crestani[1], Werner Petry Silva[2], Luciana Estacia Ambros[1]

[1]Universidade de Passo Fundo, Passo Fundo RS, Brazil.
[2]Instituto de Neurologia e Neurocirurgia, Passo Fundo RS, Brazil.

Correspondence: Carlos Eduardo Marisco Ceretta; Email: caduceretta@hotmail.com.

Case presentation: Patient A.A.A., 48 years old, male, with no comorbidities and continuous medication, was seen in the emergency department of a tertiary hospital in Rio Grande do Sul (RS) with edema and erythema of the eyelids in the right eye, associated with orbital discomfort, visual clouding and diplopia for 2 days. On physical examination, the right eye showed significant periorbital edema, chemosis, proptosis, and signs of paralysis of multiple cranial nerves on the right (III, IV, and VI). To better elucidate the diagnosis, an angiotomography of the skull and cervical vessels was requested, which showed signs of enlargement of the superior ophthalmic vein, a protruding cavernous sinus with asymmetrical enhancement, suggesting a carotid-cavernous fistula (CCF). The neurosurgical team was asked to evaluate the patient, and cranial arteriography was performed, which confirmed the suspicion of an arteriovenous fistula with multiple afferents, mainly the right external carotid artery draining into the cavernous sinus. Venous embolization was performed, with complete exclusion of the fistula involving the ascending pharyngeal artery and the right inferior petrosal sinus. The patient was discharged from the hospital 2 days after the procedure, with complete improvement of the ocular deficit and signs of cranial nerve palsy. **Discussion:** Carotid-cavernous fistulas are a rare condition involving an abnormal connection between the carotid artery or its branches and the cavernous sinus. CCFs have an incidence of 0.16 to 0.29 per 100,000 adults per year,

mostly due to the rupture of intracavernous aneurysms. The main symptoms are ocular, usually pain, conjunctival injection, and proptosis, with a risk of vision loss. Suspicious clinical findings can be investigated by angiotomography or cranial angiography, which have a sensitivity of 87% and 90% for diagnosing CCF, respectively. The standard approach is endovascular embolization, both arterial and venous. After effective intervention with complete fistula closure, ocular symptoms tend to disappear. **Final comments:** This case clearly illustrates the clinical presentation, diagnostic investigation, and successful therapeutic management of CCFs. It can lead to significant ophthalmic symptoms as a rare and serious condition. Imaging tests play a fundamental role in diagnosis. Venous embolization is a safe and effective therapeutic option, leading to a rapid patient recovery with complete improvement of symptoms.

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Carpal tunnel syndrome with Riche-Cannieu anastomosis: an electrophysiological-ultrasound correlation report

Antonio Edvan Camelo Filho[1], Pedro Lucas Grangeiro Sá Barreto Lima[1], Francisco Luciano Honório Barreto Cavalcante[1], Pedro Vitor Ferreira Rodrigues[1], Oliver Reiks Miyajima[2], Paulo Ribeiro Nóbrega[1], Pedro Braga-Neto[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Antonio Edvan Camelo Filho; Email: edvan.camelo@gmail.com.

Case presentation: A 53-year-old woman who complained of numbness and tingling in both hands was referred for evaluation of possible carpal tunnel syndrome (CTS). There was no atrophy in the thenar muscles, and motor function was preserved during all hand movements. All nerve conduction studies except for median nerve data were bilaterally normal in amplitude, latency, and conduction velocity. Median sensory nerve amplitude potentials (SNAPs) were obtained bilaterally with reduced velocity and normal amplitude values. Right median compound motor amplitude potential (CMAP) had a normal distal latency and markedly reduced amplitude (2mV) without motor block along elbow and axilla stimulations. Left motor median nerve studies were normal. Needle examination was normal, including bilateral abdutor pollicis brevis (APB) muscles, proximal median nerve, and C8T1 territories. These findings supported the diagnosis of a severe right CTS, yet the lack of thenar atrophy and a normal APB myography alerted us to the possibility of an anomalous innervation. We performed muscle and median nerve ultrasounds that confirmed CTS in both hands with a thickened median nerve (R 11 mm2 /L 10 mm2) without abnormalities in the nerve trajectory on both arms. APB muscle echogenicity (Heckmatt 1) and thickness were preserved. The final diagnosis of this patient was the coexistence of a mild bilateral CTS with a Riche-Cannieu anastomosis (RCA) in the right hand. **Discussion:** The Riche-Cannieu anastomosis involves variations in the innervation of intrinsic hand muscles, in which thenar muscles normally supplied by the median nerve are innervated by the ulnar nerve. When complete, this condition is sometimes called the "all ulnar hand". Misinterpretation of RCA may cause several diagnostic problems; for example, in ulnar nerve injury, the presence of signs of denervation in the median-innervated muscles would lead one to suspect the possibility of a C8-Tl or brachial plexus injury. Also, in CTS cases, mild sensory abnormalities may be misclassified as severe CTS and, therefore, indicate surgery. **Final comments:** Electrodiagnosis of peripheral nerves presents challenges due to anatomical anomalies like RCA. This case shows the importance of recognizing anomalous innervations such as the RCA, particularly when interpreting low-amplitude motor responses in the median nerve. Furthermore, integrating neuromuscular ultrasound into clinical practice can significantly enhance diagnostic capabilities. This imaging modality provides real-time visualization of nerves and muscles, offering additional insights into anatomical variations and pathological conditions that may not be fully captured by traditional electrodiagnostic tests alone. By incorporating the awareness of rare anatomical variations like RCA and the use of neuromuscular ultrasound, clinical neurophysiologists can improve accuracy in diagnosis, optimize treatment planning, and ultimately enhance patient care outcomes.

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Case of Wernicke's encefalopathy following hyperemesis gravidarum

Pedro Sanches Corrêa[1], Rodrigo da Franca Santos[1], Ilana Cwajgenberg[1], Crissia Pitanga Malta[1], Maria Eduarda Morgado da Silva[1], Sofia Mermelstein[1], Felipe da Rocha Schmidt[1], Diogo Matheus Terrana de Carvalho[1]

[1]Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.

Correspondence: Crissia Pitanga Malta; Email: crissiapitanga@hotmail.com.

Case presentation: 27 year old female, G4P4A0, diagnosed pregnancy at 31 weeks, due to nausea and intense vomiting, associated with weight loss of 20kg between February and March 2024. In April, fetal death was verified and the patient was submitted to a cesarean section. After 3 days, the patient developed progressive ataxic gait and mental confusion and is readmitted in the fourth day of symptoms at the maternity hospital, where Ampicillin and Aciclovir were administered for 14 days, without clinical improvement. She was transferred to a reference service of Neurology on the 24th day after the cesarean section. At admission, the patient presented with inattention, ataxia with dysmetria, dysdiadochokinesia, intention tremor, saccadic eye pursuit, hypotonia of arms and legs, absent patellar tendon reflex, clonus of the lower limbs, spastic dysarthria and Pseudobulbar affect. Due to the clinical history of hyperemesis gravidarum, Wernicke's Encefalopathy was suspected. Brain MRI demonstrated areas of hyperintensity in T2 and FLAIR with restriction to water diffusion diffusely in the cerebellar cortex with reduced sulcus in the cerebellar folia associated with cytotoxic edema, as well as discrete hyperintensity in FLAIR with restriction to water diffusion in the mammillary bodies and hypothalamus. CSF exam had normal opening pressure, glucose of 59mg/dl, proteins of 31 mg/dl and 15 leukocytes/mm3 (100% lymphocytes). High dose thiamine was initiated orally and intravenously, with progressive improvement of the symptoms. After 1 month, a new brain image showed an exuberant cerebellar atrophy. **Discussion:** Wernicke's encephalopathy is an acute neuropsychiatric condition secondary to thiamine deficiency. Common clinical manifestations include nystagmus or other ocular motility disorders, ataxia and altered mental status. Risk factors include mal-nutrition and excessive vomiting, as in hyperemesis gravidarum. When not adequately treated, it may develop to Korsakoff Syndrome, in which confabulations and amnesia are frequent. Diagnosis may be established by dosing serum thiamine levels or erythrocyte transketolase activity, although these tests have low specificity and are not widely available. Hence, clinical and radiological diagnosis are accepted. Usually, CSF examination is normal and brain MRI usually presents with hyperintensities in T2 in the medial thalamus, mammillary bodies, periaqueductal gray matter and cerebellum. **Final comments:** This case report exemplifies a patient with a probable diagnosis of Wernicke's Encefalopathy following hyperemesis gravidarum, which is very underdiagnosed due to a wide range of possible symptoms and because few patients present with the classic triad. This condition has a high morbidity and mortality rate, and may be easily treated if correctly identified, therefore, must be included as differential diagnosis when risk factors are present.

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Case report - ELOVL4: cerebellar ataxia with axonal sensory-motor polyneuropathy, epilepsy, psychosis and cognitive decline

Kaliny Oliveira Peixoto[1], João Paulo Barile[1], Renan Brandão Rambaldi Cavalheiro[1], Patrícia Marques Mendes[1], Igor Braga Farias[1], Paulo de Lima Serrano[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Kaliny Oliveira Peixoto; Email: kalinyop355@gmail.com.

Case presentation: 52-year-old woman, with adequate neuropsychomotor development up to 3 years old, when she presented the first episode of dysperceptive focal epileptic seizure. Since childhood, she had an intellectual deficit. She had seizures with moderate frequency, becoming rarer after the 30 years old, when she began experiencing visual and auditory hallucinations, heteroaggressiveness and persecutory delirium, associated with cognitive decline. After 2 years, she developed an imbalance, with frequent falls, inability to remain in orthostasis with her eyes closed and impaired fine motor coordination. Two years ago, she began to experience dysphagia for solids and liquids and one year ago, she became wheelchair-bound. She currently has chronic constipation and osteoporosis, with previous ankle fractures. She uses medications to control symptoms. Denies consanguinity. On examination, she presents bradypsychism, dysarthria, slowing of saccades, nystagmus in the horizontal conjugate gaze; appendicular hypotonia; areflexia in lower limbs, hyporeflexia in upper limbs; apalesthesia in the lower limbs, tactile and painful hypoesthesia in the lower limbs; abasia/astasia; cerebellar tests with mild dysmetria in the upper limbs and significant dysmetria in the lower limbs; pyramidal tract signs absent. She underwent electroneuromyography, which showed marked axonal sensory-motor polyneuropathy in the lower limbs (predominance of the sensory component) and absence of the cutaneous-sympathetic reflex in the upper and lower limbs (involvement of fine autonomic fibers). Brain resonance showed bilateral hemispheric cerebellar atrophy, without olivary or pontine atrophy. Deltoid muscle biopsy showed subsarcolemmal mitochondrial proliferation. The genetic panel revealed the presence of the variant c.428T> C (p.Val143Ala) in simple heterozygosity in the ELOVL4 gene. **Discussion:** This gene is involved in the biosynthesis of very long chain fatty acids. Autosomal dominant variants cause spinocerebellar ataxia type 34, classically associated with erythrokeratodermia. Most patients present with axial and appendicular ataxia, dysarthria, and nystagmus. Other manifestations include pyramidal tract signs, autonomic disorders, retinitis pigmentosa, and cognitive impairment. Brain resonance usually reveals cerebellar atrophy and may present Hot Cross Bun Sign, typically associated with Type C Multiple System Atrophy. In our patient's case, we can highlight the absence of erythrokeratodermia, significant neuropsychiatric involvement (psychosis and cognitive decline) and epilepsy as a significant neurological manifestation during evolution. **Final comments:** This report therefore contributes to expanding the spectrum of clinical presentations associated with ELOVL4, whose diagnosis must be considered independently of dermatological manifestations and in the differential approach of cognitive and epileptic syndromes.

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Case report of an autosomal dominant limb-girdle muscular dystrophy due to a heterozygous variant in the CAPN3 gene

Rafael Chaves Claudino de Queiroga[1], Renan Brandão Rambaldi Cavalheiro[2], Bruna Verri de Martino[1], Thomás Hagemann Monteiro[1], Diego Belandrino Swerts[1], Camila Lopes Figueiredo[1], Letícia Machado Dumont[1], Cícero Rodrigues Veloso[1], Ana Luisa Vilela Sá Fortes[1], Melissa Nunes Polaro[1]

[1]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Renan Brandão Rambaldi Cavalheiro; Email: renan.rambaldi@icloud.com.

Case presentation: A 25-year-old man from São Paulo (SP), with no known medical ou neurological familiar history, born to non-consanguineous parents, presented with a 7-year history (onset at age 18) of symmetric proximal lower limb weakness, with progressive worsening. Approximately 2 years later, he noticed weakness in his upper limbs and worsening weakness in his lower limbs, making it impossible for him to run. He started training at a gym, experiencing significant difficulty with upper and lower body resistance exercises and squats. A non-medical professional at the gym advised him to use testosterone, which he found to have some effect on muscle hypertrophy. Physical examination revealed only muscle weakness, as follows: in the upper limbs, grade 4- for shoulder abduction and elbow flexion and grade 5 for wrist extension; and in the lower limbs, grade 3 for hip flexion and knee extension and grade 4 for dorsiflexion of the feet. Laboratory tests (all in UI/L) revealed: creatine phosphokinase 4,213, aldolase 51.4, alanine aminotransferase 198, aspartate aminotransferase 107, and lactate dehydrogenase 474. Electroneuromyography with sensory and motor conduction studies showed no abnormalities; needle examination showed low-amplitude potentials and a sparse recruitment pattern, indicating a myopathic process. MRI of the thighs showed high signal in the posterior compartments on the T1-weighted sequence, indicating fat substitution, with no abnormalities on the T2 and STIR sequences. Finally, a muscle dystrophy panel revealed a heterozygous variant c.2167A> G in the CAPN3 (calpain 3) gene. Based on the clinical and molecular context, a diagnosis of calpain-associated limb-girdle muscular dystrophy was made. **Discussion:** Limb-girdle muscular dystrophies (LGMD) are a heterogeneous group of genetic disorders characterized by proximal muscle weakness. Dozens of subtypes have been described associated with specific gene loci, with dominant forms being less common than recessive forms, accounting for less than 10% of cases. Calpainopathies, also known as autosomal recessive LGMD type 1 (LGMDR1), are considered the most common autosomal recessive LGMD and are characterized by weakness of the proximal muscles of the shoulder and pelvic girdles, with great clinical heterogeneity. Most pathogenic variants in CAPN3 cause loss of function, resulting in recessive disease. In recent years, this exclusive recessive inheritance has been reconsidered with the publication of independent families with an autosomal dominant inheritance pattern. The patient in this study, with no family history of neurological disorders, has a heterozygous variant in the CAPN3 gene, favoring the possibility of inheritance from an unaffected heterozygous relative, resulting in autosomal dominant LGMD. **Final comments:** This case describes a patient with calpain deficiency-related muscular dystrophy with a heterozygous variant. It expands the variability of known genetic presentations.

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Case report of anti-nmda receptor encephalitis in a 51-year-old female: an uncommon presentation

Luciane Ramos Perez Silveira[1], Paula de Oliveira Pinto[2], Samuel Brighenti Bergamaschi[3], Daiane Paola Perin[4], Patrícia dos Santos[5], Ricardo Werner Rocha[3]

[1]Universidade Federal de Pelotas, Pelotas RS, Brazil.
[2]Universidade Federal da Fronteira Sul, Passo Fundo RS, Brazil.
[3]Universidade Federal de Santa Catarina, Florianópolis SC, Brazil.
[4]Universidade do Vale do Itajaí, Itajaí SC, Brazil.
[5]Universidade de Caxias do Sul, Caxias do Sul, Brazil.

Correspondence: Luciane Ramos Perez Silveira; Email: lucianeperez@yahoo.com.br.

Anti-NMDA receptor encephalitis is an autoimmune form of encephalitis often misdiagnosed as viral encephalitis due to varied initial symptoms.. Diagnosis involves EEG, MRI, and lumbar puncture for cerebrospinal fluid analysis. Symptoms include personality changes, autonomic dysfunction, and neurologic deterioration, especially in young females. Treatment includes immunosuppression. We report the case of a51-year-old female patient with a history of bariatric surgery and depression, who was admitted to the emergency room due to acute confusion and seizures. Physical and neurological examination did not reveal any abnormalities. Laboratory tests showed no significant abnormalities. A lumbar puncture that showed WBC of 35, RBC of 20, glucose of 106, and protein of 103; CSF gram was negative. The CSF Herpes virus type 1 PCR was positive, confirming the diagnosis of herpes encephalitis. Brain MRI indicated extensive signal changes affecting cortical and subcortical regions of the mesial temporal lobes, hippocampi, right thalamus, insular regions, anterobasal aspects of the frontal lobes, suggesting viral encephalitis. The ECG was normal. The patient was treated with acyclovir for 21 days. On the 10th day of treatment, she experienced worsening symptoms of headache and fever. A repeat CT scan showed a large intraparenchymal hematoma. Despite this, the patient showed improvement over time, with resolution of seizures. She was discharged on day 41 with a prescription for levetiracetam 1000mg daily. Sixty days after the initial symptoms, the patient returned to the hospital with fever, insomnia, hallucinations, echolalia. Treatment with olanzapine and diazepam was initiated, and the antiepileptic medication was changed to valproic acid. The lumbar puncture revealed WBC of 15, glucose of 56, and protein of 178. A subsequent Brain MRI demonstrated anterior lesions with improved edema and diffusion restriction areas. The patient continued to develop episodes of acute agitation and persistent insomnia. Further investigations, including a transvaginal ultrasound, confirmed the presence of an ovarian teratoma. Finally, CSF results showed testing for anti NMDA receptor antibodies were positive. The patient benefited from intravenous methylprednisolone therapy followed by a cyclophosphamide 1G continued for 9 months. The patient's clinical condition gradually improved. Despite a slow but favorable clinical course with corticosteroid and cyclophosphamide therapy alongside tumor removal, the patient continued to experience some functional deficits at a 10-month follow-up, including episodes of disorganized speech. However, improvements were noted in insomnia and visual hallucinations. **Final comments:** Anti-NMDAR encephalitis typically occurs in young patients with no history of acute psychiatric symptoms. The possibility of this pathology should be considered before diagnosing a patient with a psychiatric illness.

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Case report of Creutzfeldt-Jakob disease in a tertiary center: clinical manifestations and findings on complementary exams

Gabriela Rossini Dantas[1], Caroline de Barros Viegas[1], Dominick E. Monteiro Silva[1], Eduardo Abrão Spinola Rezk[1], Hélio Aquaroni Farao Gomes[1], Itamar Meireles Andrade Santos[1], José Eduardo Trentim Longo[1], João Márcio B. de Oliveira[1], Luciano Ribeiro da Silva[1], Pedro Machry Pozzobon[1]

[1]Universidade Estadual Paulista, São Paulo SP, Brazil

Correspondence: Gabriela Rossini Dantas; Email: rossinidantasg@gmail.com.

Case presentation: Report of 3 cases of probable Creutzfeldt-Jakob disease (CJD). A 69-year-old man, hypertensive and diabetic, started a progressive decline in executive, attentional, and memory functions a month ago, along with inverted sleep-wake cycle, apraxic gait, myoclonus and recently, akinetic mutism. Investigations revealed: Brain MRI with cortical hyperintensity in the left temporal lobe and gyral enhancement in the left parietal and frontal cortex on FLAIR; EEG with moderate baseline activity disturbance and generalized periodic activity (GPD); Cerebrospinal fluid (CSF) showing mild hyperproteinorrachia and presence of 14-3-3 protein. A 59-year-old man, hypertensive and dyslipidemic, developed progressive amnestic, dysexecutive, and aphasic symptoms over 2 months, later accompanied by gait ataxia, abnormal limb movements during sleep, myoclonus and physical examination revealed global hyperreflexia, frontal release signs, and upper limb dysmetria. Brain MRI: cortical enhancement in the left hemisphere and left basal nuclei with associated diffusion restriction; EEG: GPD with sharp wave complexes below 2.5 Hz; CSF: hyperproteinorrachia and presence of 14-3-3 protein. A healthy 59-year-old man presented with progressive spatial disorientation, executive dysfunction, gait ataxia, reduced verbal fluency, and behavioral changes over 2 months, progressing to complete dependence in activities of daily living. Physical examination revealed myoclonus and signs of pyramidal release (bilateral hyperreflexia and Babinski sign). Brain MRI: diffusion restriction in bilateral cortical and basal nuclei; EEG: rhythmic GPD at 1 to 1.5 waves per second. The patient died 3 months after symptom onset. **Discussion:** Sporadic CJD is considered the most common prion disease in humans. With long incubation periods and a rapid clinical course, definitive diagnosis is based on spongiform changes and abnormal prion protein accumulation in brain biopsy. Probable CJD is defined by neuropsychiatric disorder with a positive real-time quaking-induced conversion (RT-QuIC) test; or progressive dementia with two or more clinical features (myoclonus; visual or cerebellar disturbances; pyramidal or extrapyramidal dysfunction; akinetic mutism) and one or more supportive findings on tests (EEG with periodic sharp wave complexes; positive 14-3-3 in CSF with clinical duration less than two years; MRI with hyperintensity in caudate nucleus or putamen and/or in at least two cortical regions on DWI or FLAIR). **Final comments:** Although rare, sporadic CJD should be considered in the differential diagnoses of rapidly progressive dementia when typical clinical, electrophysiological, and neuroimaging features are present. The presence of 14-3-3 protein in CSF is not mandatory for diagnosis. The occurrence of three cases within a relatively short period underscores the importance of early clinical suspicion and detailed investigation for appropriate management of these patients.

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Case report of familial hemophagocytic lymphohistiocytosis in a 5-year-old child: diagnostic and therapeutic challenges in central nervous system involvement

Tomás Tejerina Sellos Costa Bahia de Almeida[1], Guilherme Marques Nogueira[1], Julia Viana Gil de Castro[1], Filipe de Deus Tozani[1], Maria Eduarda Guimarães Porto[1], Weslley Nunes Teles[1], Ana Beatriz Calmon Nogueira da Gama Pereira[1], Ivana Rocha Raslan[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.

Correspondence: Tomás Tejerina Sellos Costa Bahia de Almeida; Email: tejerinatomas@gmail.com.

Case presentation: It is a previously healthy 5-year-old male who was admitted with acute symptoms of imbalance, vomiting, nausea, and headache, that worsened over time. The family history was negative for neurological diseases, except for a brother with an undiagnosed intellectual disability. Physical exam revealed cerebellar ataxia signs, global hyperreflexia with bilateral Babinski and Hoffman signs, and bilateral horizontal nystagmus in all gazes. Brain MRI showed multiple cerebral areas with hyperintense signal on T2 and FLAIR, some with isointense to slightly hypointense signal on T1, with mild contrast enhancement, suggesting with a demyelinating lesion. The patient received pulsotherapy with methylprednisolone with clinical and radiological imaging. Upon tapering the corticosteroid, the patient experienced significant clinical deterioration and was readmitted. After an extensive laboratory investigation, genome test confirmed a PRF1 gene mutation (Het. c.1A> Gp.? and Het.c.529C> Tp.(Arg177Cys) pathogenic variants) associated with Familial Hemophagocytic Lymphohistiocytosis Type 2, with CNS involvement only. The patient underwent chemotherapy with hydrocortisone and intrathecal methotrexate, followed by a bone marrow transplant. The patient has been clinically stable for 2 years, with no signs of disease reactivation. **Discussion:** Hemophagocytic Lymphohistiocytosis (HLH) results from a cytokine storm triggered by various stimuli, causing dysfunction of Natural Killer cells and CD8+ T cells. These cells become overly activated, producing excessive INF-γ and other inflammatory cytokines. HLH can be primary (genetic) or secondary (linked to cancers and infections). Its hallmark is histiocyte build-up in tissues with poor cytotoxic function. Key symptoms include fever, hepatosplenomegaly, and pancytopenia. Neurological symptoms, in 30-73% of patients, include seizures, focal deficits, ataxia, meningismus, and altered consciousness. Diagnosis of HLH involves assessment of clinical presentation, performing tests such as measuring CSF neopterin level, urine B2 microglobulin, liver enzymes, LDH and CPK levels. Nowadays, genetic analysis is an important tool to establish the specific diagnosis. Treatment involves corticosteroids, immunomodulators, intrathecal methotrexate for progressive CNS disease, and hematopoietic stem cell transplantation (HSCT). Despite treatment advances, 25-50% of patients need additional therapies to achieve remission. **Final comments:** Although rare, HLH has high morbidity and mortality, often causing severe neurological damage and death if not promptly diagnosed and treated. Highlighting such cases ensures that physicians consider HLH prior to extensive neurological system involvement, leading to timely intervention and significantly improved patient outcomes.

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Case report of hematomyelia due to cervical intramedullary cavernous hemangioma after plastic surgery

Rafael Chaves Claudino de Queiroga[1], Renan Brandão Rambaldi Cavalheiro[2], Camila Lopes Figueiredo[1], Cícero Rodrigues Veloso[1], Filipe Brito de Ferraz[1], Letícia Machado Dumont[1], Maria Clara Motta Barbosa Valente[1], Francisco Tomaz Meneses de Oliveira[1]

[1]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Camila Lopes Figueiredo; Email: camila04_lopes@hotmail.com.

Case presentation: A 23-year-old woman with a previous medical history of severe scoliosis and bilateral congenital clubfoot developed headache and right hemiparesis shortly after waking up from anesthesia for a reduction mammoplasty. She was discharged from the hospital where the surgery was performed and sought emergency department on the next day. On neurological examination, she had severe scoliosis, Horner's syndrome on the right, ipsilateral hemiparesis, disproportionate, incomplete, predominantly distal and in the upper limb (hand flexion grade 1), with Babinski's sign present, as well as thermal hypoesthesia in the left half of the body. The possibility of spinal cord syndrome was raised, and an magnetic resonance imaging (MRI) of the cervical and thoracic spine was performed, which revealed an extensive cervical intramedullary lesion, located centrally and paramedially to the right, characterized by a heterogeneous signal with hypointensity on T1, hyperintensity on T2/STIR, measuring 2.3 x 0.9 x 0.8 cm, with faint gadolinium impregnation at levels C6 and C7, without restriction to the free diffusion of water molecules in the diffusion sequence images. There were foci of hyperintensity on T1 and hypointensity on T2/STIR, suggestive of hematoma content. Spinal cord edema was associated, characterized by hyperintensity on T2/STIR, extending from C1 to T1. During hospitalization, coagulopathies and systemic neoplasms were ruled out. The patient was referred for outpatient follow-up to undergo rehabilitation (mainly motor physiotherapy) and to investigate family involvement. **Discussion:** Intramedullary cavernous hemangiomas (or cavernomas) are rare vascular malformations, especially when compared to cerebral cavernomas. They can present with progressive and slow neurological decline or with acute impairment that occurs due to a complication of these malformations, which is intraspinal bleeding or hematomyelia. Differential diagnoses include spinal ependymomas, astrocytomas, and spinal arteriovenous malformations. Surgical treatment may be indicated, especially in cases of repetitive hemorrhages to prevent progressive neurological deterioration. The patient in this case presented with a complication of cavernomas in the postoperative period of plastic surgery (reduction mammoplasty) that was performed with general and epidural anesthesia. She presented with lateral spinal cord syndrome, including ipsilateral Horner's syndrome to the lesion. No reports like this were found in the literature. The patient followed up on an outpatient basis and is undergoing rehabilitation, especially with neurofunctional physiotherapy. **Final comments:** Intramedullary cavernomas are rare vascular malformations that can manifest with acute neurological impairment in case of complication (intraspinal hemorrhage). The case in question presents an interesting report of hematomyelia with lateral spinal cord syndrome after plastic surgery.

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Case report of late diagnosis of subependymal heterotopia as a cause of refractory epilepsy in a male with normal neuropsychomotor development

Rafael Chaves Claudino de Queiroga[1], Thomás Hagemann Monteiro[1], Maria Clara Motta Barbosa Valente[1], Letícia Machado Dumont[1], Filipe Brito de Ferraz[1], Cícero Rodrigues Veloso[1], Camila Lopes Figueiredo[1], Vanessa Cristina Colares Lessa[1], Fábio Barreto Cabral[1], Cristine Mella Cukiert[1]

[1]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Cristine Mella Cukiert; Email: cmcukiert@uol.com.br.

Case presentation: A 27-year-old man, with no other medical history besides seizures started at 9 years old. His seizures were characterized as impaired awareness with behavior arrest that sometimes evolved to tonic posture of the right upper limb associated with orofacial automatisms. Neuropsychomotor development (NPMD) and neurological physical exam were unremarkable since that time. He began a follow-up with a neurologist with no etiological cause identified at the time and progressed to refractory epilepsy by 16 years old. At 23 years, as seizures remained uncontrolled, he was referred to a specialized center in the treatment of drug resistant epilepsy for investigation. His electroencephalogram (EEG) showed left temporal posterior sharp waves, sometimes with diffusion to nearby and contralateral areas. A brain magnetic resonance imaging (MRI) revealed nodular subependymal heterotopia (SEH) in the lateral ventricles and nodular inter hypothalamic adhesion. Nowadays, he uses 3 anti-seizure medications (carbamazepine 800 mg/d, clobazam 10 mg/d and levetiracetam 500 mg/d) with a better control of his epileptic events. **Discussion:** SEH is a cortical malformation that can be a cause of epilepsy. Females with SEH typically present with focal seizures starting at the second decade of life, with normal NPMD and neurological examination. Males, on the other hand, can have either X-linked or autosomal inheritance. X-linked SEH, caused by mutations in FLN1 gene on chromosome Xq28, is often associated with visceral and central nervous system anomalies, associated with abnormal NPMD. Autosomal SEH has a similar clinical course to symptomatic females. Diagnosis is made by neuroimaging, with MRI being the most sensitive exam. Treatment is focused on the type of seizure and surgery may be indicated in some cases. Our patient developed seizures at the end of the first decade of life, with progressive refractoriness and delay to referral to a treatment specialized center in drug resistant epilepsy. Only around the age of 25, after the MRI and subsequent diagnosis, he achieved good control with three anti-seizure drugs. **Final comments:** SEH is an established cause of epilepsy, however, it is not always diagnosed at the onset of seizures due to access difficulties or limited resources, demonstrating the importance of assessment in a specialized center. Additionally, it is more common in women and, in men, it can lead to impaired NPMD. This case highlights the importance of considering SEH in the differential diagnosis of epilepsy, particularly in males with normal NPMD and refractory epilepsy.

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Case report of neuro-behçet and intervention with infliximab

Giovanni Caniçali Simões[1], Aldren Thomazini Falçoni Júnior[1], Sephora Sabrina Candido de Almeida[1], Ingrid Lorena da Silva Gomes[1], Guilherme Lago Miranda[1], Tainá Fátima Ramos Gonçalves[1], Henrique de Moraes Bernal[1], Pedro Almeida Martins Pontes[1], Herval Ribeiro Soares Neto[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Giovanni Caniçali Simões; Email: gcanicali@gmail.com.

Case presentation: A 44-year-old woman presented with severe headache, nausea, and photophobia of acute onset, treated with simple analgesics, and discharged with total symptom relief. In three days, she experienced recurrence of symptoms, now including vomiting, asthenia, fatigue, and somnolence, difficulty walking, imbalance requiring a wheelchair, diplopia, and facial numbness. Neurological examination revealed exaggerated deep tendon reflexes in the right upper limb, sensory ataxia with stomping gait. Active search showed erythema nodosum on the lower limbs and arthralgia in the wrists and hips and oral ulcers. MRI and angio-MRI showed hyperintensity in T2 and FLAIR at the pontomesencephalic junction with slight expansion and gadolinium enhancement, without vascular abnormalities. CSF analysis revealed 16 leukocytes (42% lymphocytes, 45% neutrophils, 12% monocytes), with normal protein, glucose and lactate levels. Given the likely inflammatory etiology of the rhombencephalitis, the patient received a 5-day course of 1 gram/day methylprednisolone, resulting in significant clinical improvement. The associated findings of oral ulcers, erythema nodosum, and

joint involvement suggested Behçet's disease, supported by positive HLA-B51/52. For maintenance therapy, infliximab was chosen. The patient has remained stable and symptom-free since. **Discussion:** Behçet's disease (BD) is a rare chronic multisystemic inflammatory condition characterized by recurrent oral and genital ulcers and uveitis, with a relapsing-remitting course. Neurological involvement in BD, known as Neuro-Behçet's disease (NBD), typically occurs years after the onset of systemic symptoms, although it is one of the more severe complications. NBD presents in two distinct forms: parenchymal (80%) affecting the brainstem, brain, spinal cord, and optic nerve, and non-parenchymal (20%), primarily cerebral venous thrombosis. HLA-B51 is a marker for NBD development. Monoclonal antibodies, especially anti-TNF, are recommended for severe or refractory disease. Infliximab has proven most effective for NBD, with substantial literature support. Poor prognostic factors include parenchymal CNS involvement, lesion extent, inflammatory CSF changes, relapses, and early disease progression. **Final comments:** This case call attention to the importance of early diagnosis and appropriate management of BD with neurological involvement. The high diagnostic suspicion of NBD in the presence of rhombencephalitis, along with an active anamnesis focusing on systemic manifestations, was essential for diagnostic confirmation. The therapeutic success, maintaining symptom remission, was guaranteed with infliximab, given the minimal risk of recurrence in a patient with severe manifestation and high functional demand.

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Case report: a new phenotype of ataxia with oculomotor apraxia type 4 in a young woman

João Carlos Cervelin[1], Bruno Wendorff Lucci[1], Sarah Prech e Souza[1], Fernanda Coan Antunes[1], Pricila Bernardi[1], André Dias de Oliveira[1], Gisele Espindola[1]

[1]Universidade Federal de Santa Catarina, Hospital Universitário, Florianópolis SC, Brazil.

Correspondence: João Carlos Cervelin; Email: joaocarloscervelin@gmail.com.

Case presentation: A 21-year-old female patient developed appendicular and gait ataxia at the age of 7, progressively experiencing lower limb weakness. She exhibited normal neuropsychomotor development and lacked a family history of neurological diseases. On neurological examination, she presented with cerebellar dysarthria, gait ataxia (requiring support for walking), mild bilateral dysmetria, distal weakness, distal hypoesthesia, absent deep tendon reflexes, and no oculomotor apraxia. Electroneuromyography (ENMG) revealed sensory-motor axonal polyneuropathy, while brain magnetic resonance imaging (MRI) displayed cerebellar and brainstem atrophy. Laboratory findings included hypercholesterolemia. Genetic testing identified 100% homozygosity for the PNKP gene, confirming the diagnosis of Ataxia With Oculomotor Apraxia Type 4 (AOA4). **Discussion:** Ataxia with oculomotor apraxia encompasses a heterogeneous group of autosomal recessive cerebellar ataxias characterized by a spectrum of clinical manifestations, including cerebellar ataxia, sensorimotor axonal neuropathy, oculomotor apraxia, and extrapyramidal features. AOA4 is a relatively rare subtype, first described in a Portuguese population in 2015. The pathogenesis of AOA4 involves mutations in the PNKP gene, resulting in defective DNA repair mechanisms, particularly in response to oxidative stress. AOA4 typically presents in childhood or adolescence with progressive cerebellar dysfunction, including dysarthria, gait ataxia, and limb incoordination. Our patient's clinical course aligns with these typical features, as she developed appendicular and gait ataxia at a young age, followed by progressive lower limb weakness. The absence of oculomotor apraxia in our patient is noteworthy, as it is a cardinal feature of AOA but may not manifest in all cases. Neuroimaging studies, such as brain MRI, play a crucial role in the diagnosis of AOA4, often revealing cerebellar and brainstem atrophy, as observed in our patient. Additionally, electrophysiological studies, including ENMG, aid in confirming the diagnosis by demonstrating sensorimotor axonal polyneuropathy, as seen in our patient's. Cognitive impairment is a common feature of AOA, affecting a significant proportion of patients with AOA4. However, our patient demonstrated preserved cognitive function, highlighting the variable expressivity of this condition. The absence of cognitive impairment in our case may suggest potential genotype-phenotype correlations or modifier effects influencing the disease presentation. **Final comments:** We report a case of AOA4 in a young woman characterized by cerebellar ataxia, sensorimotor polyneuropathy, and absence of oculomotor apraxia and cognitive impairment. Our findings underscore the clinical heterogeneity and variable expressivity of AOA4, emphasizing the importance of comprehensive phenotypic characterization and genotype-phenotype correlations in understanding the disease pathogenesis.

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Case report: atypical Guillain-Barré syndrome associated with dengue infection

Rafael Bragança Rodrigues Matias[1], Eduardo Mesquita[1], Ana Luisa Vieira de Faria Donadon[1], Amanda Miranda de Mendonça[1], Alex Araujo Fernandes[1], Matheus Luís Braga Munareto[1], Alex Machado Baeta[1], Hennan Salzedas Teixeira[1], Karlla Danielle Ferreira Lima[1]

[1]Beneficência Portuguesa de São Paulo, São Paulo SP, Brazil.

Correspondence: Rafael Bragança Rodrigues Matias; Email: rafaelwaa@gmail.com.

Case presentation: A 20-year-old patient with a history of controlled migraines presented to the emergency room with exacerbation of migraine with photophobia, phonophobia, frontal pain, and associated nausea and vomiting. Neurological examination revealed partial sixth nerve palsy with medial esophoria of the right eye, horizontal diplopia, and weakness while walking. Laboratory tests showed dissociated cerebrospinal fluid (CSF) protein levels (cellularity 3, protein 74), with other laboratory parameters within normal limits. Brain and orbital magnetic resonance imaging demonstrated bilateral optic perineuritis with enhancement of the seventh and eighth cranial nerves, as well as impregnation of the C1 and C2 nerve roots. Additionally, brachial and sacral plexus imaging revealed mild nerve thickening and enhancement. Based on these findings, we hypothesized a variant of GBS, and initiated intravenous immunoglobulin therapy for 5 days. The patient showed satisfactory clinical improvement, including resolution of ophthalmoparesis and ataxia. **Discussion:** Dengue fever is a prevalent mosquito-borne disease affecting millions of people worldwide. While the classic symptoms include high fever, joint pain, and rash, there is growing evidence of neurological involvement in dengue cases. The various neurological manifestations reported include encephalitis, myelitis, Guillain-Barré syndrome (GBS), and myositis. **Final comments:** This case report draws attention to an unusual presentation of a little-recognized neurological syndrome in a context of a dengue epidemic. The syndrome, often overlooked by non-specialists, warrants consideration when encountering patients with neurological symptoms during such outbreaks.

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Case report: atypical meningoencephalitis due to cysticercosis

Lorraine Alves de Souza[1], Kesllin Mariane Gomes Valadão[1], Omar El Sayed[1], Rafael Hofmann[1], Eduardo Augusto Borba[1], Jivago Szpoganicz Sabatini[1], Crislaine Freitas Petla Silva[1], Ana Júlia Kounaris Fuziki[1], Gabriela Alves Jupen[1], Mariana Fonseca[1]

[1]Universidade Estadual de Ponta Grossa, Ponta Grossa PR, Brazil.

Correspondence: Lorraine Alves de Souza; Email: lolorrainealves@gmail.com.

Case presentation: Patient E. A. C., female, 23 years old, with headache and vomiting for 4 months; dysarthria, loss of strength and reduced level of consciousness (CNR), 1 week ago. On admission, she presented bilateral papilledema, paresis of the lateral and medial rectus muscles of the left eye and Glasgow 11. Magnetic resonance imaging (MRI) was performed with intracranial hypertension (ICH), meningoencephalitis, diffuse leptomeningeal enhancement, especially in the occipital lobes and cerebellar hemispheres, and optic neuritis on the left. Cerebrospinal fluid (CSF) with opening pressure: >20 mmHg, leukocytes: 20/mm3, mononuclear: 78%, proteins: 75 mg/dL, glucose: <20 mg/dL, corroborating the hypothesis of ICH with meningoencephalitis, acyclovir was started. When investigating the etiology of meningoencephalitis, the following were requested with negative results serologies for tuberculosis, Epstein-Barr, herpes simplex, parvovirus B19, HIV and hepatitis; PCR for herpes in CSF and autoantibodies for vasculitis. Due to the refractoriness of ICH, the hypothesis of meningoencephalitis due to cysticercosis was raised, and empirical treatment was initiated with dexamethasone and albendazole and anti-cysticerci antibodies in CSF and serum were requested, with a positive result. Despite the treatment being implemented, the patient died. **Discussion:** Neurocysticercosis can manifest in different ways - hypertensive: resulting from obstruction of ventricular drainage; epileptic: convulsive manifestations; meningitic: outbreaks of meningitis. CSF obstruction may progress to ICH. This form is associated with an inflammatory reaction, with arachnoiditis, which can manifest as aseptic meningitis. In this case, the patient presented a compatible clinical picture and MRI with ICH and inflammation of the meninges. Cysticercosis in the optic nerve may present as atypical optic neuritis, as evidenced on MRI in this case. CT and MRI are the gold standard in diagnosis, enabling visualization of the parasite and the reactional process of the lesion, however, on CT, the cysticercus is isodense to the CSF and those in the basal subarachnoid cistern are not detected by these techniques, emphasizing the need for research of IgG anti-cysticerci antibodies in CSF and serum. In this case, although no suggestive findings were found on CT or MRI, the diagnosis of meningoencephalitis due to cysticercosis was concluded, with positive anticysticercal antibodies. Even after treatment with anthelmintics and corticosteroids, the patient died, demonstrating the severity of this form of presentation. **Final comments:** In patients with meningoencephalitis of unknown etiology, it is essential to search for parasitic causes. The diagnosis of cysticercosis was challenging in this case, as imaging tests did not make it possible to identify the parasite. It was necessary to test for anti-cysticercus antibodies, which is an important tool to be included in the list of investigation tests.

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Case report: blue eyes as an initial indicator in the investigation of waardenburg syndrome in a family

Vanessa Gil Humberto dos Santos[1], Amanda Melo Leite Leão[1], Jorge Luiz Alves[1], Poliana Marcela Silva de Farias[1], Lais Felix Teixeira da Cunha[1], Willian de Souza Santos[1], Lais Borneo Moreira[1]

[1]Unigranrio, Duque de Caxias RJ, Brazil.

Correspondence: Vanessa Gil Humberto dos Santos; Email: vanessaterapia@hotmail.com.

Case presentation: In a family from Duque de Caxias, RJ, consisting of nine siblings, five are carriers of Waardenburg syndrome (WS), a genetic disorder characterized by pigmentation anomalies and sensorineural hearing loss. One of the siblings has blue eyes and white hair since birth but does not have deafness. His 29-year-old son was born deaf, with blue eyes and white hair streaks. Two other siblings were born with partial hearing loss, blue eyes, white hair streaks, mental retardation, and clinodactyly, although the latter is minimally noticeable. The two sisters have profound bilateral deafness, blue eyes, and white hair streaks. The father of the family was identified as a carrier of the syndrome. **Discussion:** WS is an autosomal dominant genetic disorder caused by mutations in genes affecting the migration and development of neural crest cells, such as PAX3, MITF, and SOX10. The phenotypic variability within the same family, as observed in this case, can be attributed to the variable expressivity and incomplete penetrance of the genetic mutation. The main diagnostic criteria includes pigmentation anomalies, such as blue eyes or iris heterochromia, and white hair. Hearing loss is a significant, ranging from profound to partial deafness. Other manifestations can include mental retardation and congenital malformations. The blue eye phenotype is particularly notable and common among WS carriers due to a deficiency in melanin production in the irises, resulting from genetic mutations associated with the syndrome. To diagnose of WS, leads clinicians to rely on distinctive clinical features. Audiological exams are essential to determine the degree of hearing loss. Imaging studies, such as computed tomography or magnetic resonance imaging, can be performed to evaluate and confirm malformations. Genetic testing is used to identify specific mutations in WS-related genes, helping to confirm the diagnosis and provide appropriate genetic counseling. There is no significant prevalence of WS between males and females. However, the expressivity of symptoms can vary considerably, even among individuals of the same sex within the same family, as observed in this case. **Final comments:** This case highlights the phenotypic variability of WS within a family with multiple carriers. Identifying WS carriers and understanding the clinical manifestations are crucial for appropriate management and genetic counseling of affected families. Early detection and continuous monitoring can improve the quality of life of the affected individuals, allowing the proper audiological interventions and educational support. This case report contributes to the understanding of the diverse clinical presentations of WS and emphasizes the importance of multidisciplinary follow-up involving geneticists, otolaryngologists, and hearing rehabilitation specialists.

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Case report: cerebellar ataxia of adult onset associated with coenzyme Q10 deficiency

Maria Fernanda Hamamoto Leati[1], Antônio Avelino Mendes Filho[1], Camila Rodrigues Nepomuceno[1], Felipe Cenicio Juliatti[1], João Roberto Ribeiro Pimenta[1], Nayara de Paula Guerreiro[1], Atie Calado Ribeiro[1], Cristiane de Araújo Martins Moreno[1], Julian Letícia de Freitas[1], Maria Sheila Guimarães Rocha[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Maria Fernanda Hamamoto Leati; Email: mfleati@gmail.com.

Case presentation: A 52-year-old female patient from Ceará, Brazil, was referred to the General Neurology outpatient clinic for investigation of motor incoordination. She reported the onset of symptoms around age 30, affecting both her lower and upper limbs insidiously. The condition progressively worsened, leading to impaired gait (feeling drunk), frequent stumbling and falls over the years, and difficulty grasping and frequently dropping objects. She denied other symptoms. Family history revealed consanguinity (parents were cousins) and an older sister with similar lifelong symptoms but without a diagnosis. Neurological examination showed square-wave jerks, vertical gaze paresis, broad-based gait, and altered cerebellar tests (dysmetria and dysdiadochokinesia). Brain MRI revealed diffuse cerebellar atrophy without other abnormalities. Laboratory tests were unremarkable. Genetic testing for Friedreich's ataxia was negative. Whole exome sequencing identified a pathogenic variant c.589-3C> G in the COQ8A gene consistent with coenzyme Q10 (COQ10) deficiency. **Discussion:** COQ10 plays a crucial role in the mitochondrial electron transport chain, and diverse enzymes catalyze the formation of coenzyme Q10 and regulate other biosynthetic proteins. Mutations in the genes (COQ1, COQ2, COQ3, COQ4, COQ5, COQ6, COQ7, COQ8A, COQ8B, COQ9) that code for these enzymes lead to coenzyme Q10 deficiency. The main phenotypes related to coenzyme Q10 deficiency include encephalomyopathy, isolated myopathy, cerebellar ataxia, severe infantile multisystemic form, and steroid-resistant nephrotic syndrome. Clinical manifestations vary, but cerebellar ataxia is a hallmark observed in all patients, and pathogenic variants in the COQ8A gene are the most common genetic cause of COQ10 deficiency-related cerebellar ataxia. Although classically described with onset in childhood, there are cases in the literature with late onset. Epilepsy, cognitive impairment, and movement disorders may accompany the condition. Ocular motor abnormalities such as gaze palsy, ophthalmoplegia, or oculomotor apraxia can also occur. MRI findings typically show cerebellar atrophy, sometimes with diffuse cerebral atrophy in severe cases. **Final comments:** This case is noteworthy not only because the patient presented with adult-onset cerebellar ataxia but also due to the rare finding of a splice proximal mutation in COQ10, a variant scarcely reported in the literature. Moreover, the clinical follow-ups that exceeded ten years showed no disease progression during this period.

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Case report: chronic optic disc edema, unilateral, secondary to vitreopapillary traction syndrome

Theodora Cristina Cruz de Assis[1], Amanda Jardim Diniz da Gama[1], Raphael Cândido Sepúlveda Coutinho[1], Frederico Castelo Moura[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Theodora Cristina Cruz de Assis; Email: t223442@dac.unicamp.br.

Case presentation: Male patient, 57 years old, referred to the Clinical Hospital of the State University of Campinas (HC-Universidade Estadual de Campinas) due to a complaint of amaurosis fugax in the right eye (OD) that lasted fifteen minutes in 2022. He reports that visual acuity (VA) in OD did not return to what it was after this event and that visual blurring got worse during headache episodes. In May 2023, optic disc edema and peridiscal hemorrhage were observed in the OD in an external service. Denied other complaints or previous comorbidities. In evaluation at Universidade Estadual de Campinas in April 2024, he presented corrected visual acuity (VA) of 20/20 in both eyes (OU). Fundoscopy exam revealed optic disc stained in OD with physiological excavation, edema in the nasal region of the optic disc with presence of a superior shunt. Fundoscopy exam without alterations in the left eye (OS). Performed visual field exam (Humphrey campimetry) on 24-2 protocol within normal limits in OD with slight enlargement of the blind spot and exam in normal conditions in OS with nonspecific findings related to learning effect in visual field testing. Optical coherence tomography (OCT) was performed in OD, which showed nasal sectorial optic disc edema, vitreopapillary traction (VPT) in the nasal region of the optic disc, bulging of the peridiscal Bruch's membrane, and peripapillary nerve fiber layer (RNFL) and ganglion cell layer within normal limits. Cranial and orbital magnetic resonance imaging (MRI) within normal limits. Considering clinical and multimodal analysis, the main diagnostic hypothesis was VPT syndrome in OD. **Discussion:** The VPT syndrome is an uncommon condition characterized by increased traction in adhesion points at the vitreopapillary interface, which can be secondary to incomplete posterior vitreous detachment (PVD) in vitreoretinal adhesion, subsequent to previous detachment of the perifoveal (stage 1) and perimacular (stage 2) regions. The pathophysiology of VPT is related to contraction forces associated with liquefaction of the vitreous body with aging, being a case more prevalent among adults aged 50 or older. The strong adherence of the posterior vitreous to the optic nerve head and to peripapillary membranes can primarily lead to hemorrhages, thinning and stretching of ganglion cell axons, decline of visual function and papillary edema formation. In this clinical case, the presence of nasal sectorial edema in the OD and VPT, evidenced in the OCT, in the nasal region of the optic disc confirmed diagnosis of subclinical unilateral optic disc edema in the OD. One of the differential diagnosis was intracranial hypertension due to headache and optic disc edema, although the unilateral edema, which does not represent a common pattern. **Final comments:** The VPT syndrome should be considered in the diagnostic investigation of patients with chronic optic disc edema.

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Case report: clinical and diagnostic challenges in patients with mcardle disease.

Tiago Camilo Eisemberg de Alvarenga[1], Izac Lage Oliveira[1], Sergio Luiz Evangelista Santos[1], Larissa Oliveira Silva Leite Ferreira[1], Rodrigo Alves da Costa[2], Matheus Alvarenga Santos[2], Esthéfane Gonçalves Costa Fernandes[2], Thamires Miguel Soares[2], Ingridi Aparecida Nogueira de Moura[2], Larissa Regina Souza Santos[2]

[1]Hospital Nossa Senhora das Dores, Itabira MG, Brazil.
[2]Centro Universitário Funcesi, Itabira MG, Brazil.

Correspondence: Izac Lage Oliveira; Email: izacoliveira93@hotmail.com.

Case presentation: A 59-year-old female patient sought neurological care with chronic CPK elevation, with values ranging between 200 and 700, with moments of significant elevation associated with metabolic and infectious changes. The patient reported that she had already been evaluated by professionals from different specialties without diagnostic definition (medical clinic, cardiology, endocrinology, rheumatology, neurology and orthopedics). Patient with no known comorbidities, did not use any medication or dietary supplement, her parents are cousins. No neurological changes or deficits were observed, muscle strength and reflexes were preserved. She reported weakness and fatigue since childhood, with difficulty in practicing physical activities. A relevant situation observed during the anamnesis is that she quickly became tired during physical exercise, however, after a brief period of rest, she was able to complete more vigorous activities, compatible with the second wind phenomenon, leading to suspicion due to research into glycogenoses. The genetic study showed variation in the PYGM gene (T> TA) in homozygosity (2 copies). After completing the diagnosis, the patient was advised on non-pharmacological measures, showing good responses. No drug treatment was introduced, genetic guidance was carried out to research the disease in the children. **Discussion:** Myophosphorylase deficiency, historically known as McArdle disease (GDS V), is the most common glycogen storage disease affecting the muscle. In myophosphorylase deficiency, glycogen is not properly broken down in muscle cells, interfering with their function. Myophosphorylase deficiency usually presents in adolescence or early adulthood with exercise intolerance, fatigue, myalgia, cramps, myoglobinuria, poor endurance, muscle swelling, and fixed weakness. However, symptoms

are often seen in childhood and may be misdiagnosed for decades. The disease is suspected on the basis of the history of exercise-induced symptoms. Genetic testing is the most efficient and least invasive means of confirming the diagnosis in patients with clinical findings consistent with the diagnosis. There was no benefit with: D-ribose, glucagon, verapamil, vitamin B6, branched chain amino acids, dantrolene sodium, and high-dose creatine. Minimal subjective benefit was found with low dose creatine and ramipril only for patients with a polymorphism known as the D/D angiotensin converting enzyme phenotype. A carbohydrate-rich diet resulted in better exercise performance compared with a protein-rich diet. Two studies of oral sucrose given at different times and in different amounts before exercise showed an improvement in exercise performance. **Final comments:** Diagnosis of McArdle disease is frequently delayed by many years following the first presentation of symptoms to a health professional. Some simple changes in lifestyle and diet can improve the quality of life of these patients.

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Case report: congenital paramyotonia, a mutation in the SCN4A gene

Ellen Kátia dos Santos Pessoa[1], Hellen Júlia Fernandes Alves[1], Nykassia Rodrigues Coelho[1], Dhulyān Severo Soares[1], Ana Beatriz Vieira Pachêco[1], Thiago Parreira Guzzo[2]

[1]Uninassau, Vilhena RO, Brazil.
[2] Academia Brasileira de Neurologia, São Paulo SP, Brazil.

Correspondence: Ana Beatriz Vieira Pachêco; Email: beatrizpachecoana@gmail.com.

Case presentation: Male patient, 56 years old, started with sustained, non-painful muscle tension in childhood that intensified in winter, associated with loss of limb coordination. At the age of 11 he lost coordination of his lower limbs with limited movement for a period of five months, returning without any intervention. Myotonic crises predominated mainly in the limbs, face and neck. At the age of 54, the weakness in the limbs intensified and after performing the electroneuromyography, acute axonal neuropathy was revealed, starting human immunoglobulin and later mestinon 60mg, but without positive effects. With the persistence of symptoms, a molecular study of the CLCN1 and SCN4A genes was requested, identifying alterations c.4343G> A (p.Arg1448His) in heterozygosity in exon 24 of the SCN4A gene, corresponding to congenital paramyotonia and hypokalemic periodic paralysis, both characterized by a pattern of autosomal dominant inheritance. The patient currently uses phenytoin 100 mg and acetazolamide 100 mg, but still suffers from painful myotonic crises daily. One of his children presents similar symptoms, however, the condition is still being investigated. **Discussion:** Paramyotonia congenita is an autosomal dominant neuromuscular disease, with mutation of the SCN4A gene. The first clinical manifestations begin in childhood, with myotonic crises, induced by physical exertion and exposure to low temperatures. During myotonic crises, the patient has greater difficulty promoting muscle relaxation. There is no cure for PMC, however with appropriate management of diet, lifestyle and medication, patients can maintain a normal life. **Final comments:** The main clinical manifestations of congenital paramyotonia include myotonic crises triggered by cold or repetitive activities, causing muscle weakness and stiffness, which are the main complaints of patients. In principle, other causes can be considered, as paramyotonia congenita is much rarer than myotonia congenita or myotonic dystrophy, highlighting the importance of carrying out a molecular study of the SCN4A gene to conclude the diagnosis. However, more studies on paramyotonia congenita are still needed, especially in relation to the diagnosis early and treatment, in order to improve the patient's quality of life, who, even when using medication, still presents symptoms as in the aforementioned case.

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Case report: cortical blindness in a case of contrast-induced encephalopathy

Fabrício Weinheimer Lippert[1], Khiany Mathias[1], Richard Simon Machado[2], Camila Pezente Dal Toe[1], Ana Paula Benedet de Carvalho[1], David dos Santos[1], Natalli Studnicka[1], Tatiana Pizzolotto Bruch[1], Fabricia Cardoso Petronilho[1], Luiz Pedro Willimann Rogério[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.

Correspondence: Fabrício Weinheimer Lippert; Email: fabriciowlippert@gmail.com.

Case presentation: M.G.C.F, a 57-year-old Caucasian female, was diagnosed with a cerebral aneurysm at the the right posterior inferior cerebellar artery origin during investigation for headaches. The patient had a death family history due to aneurysm rupture. She was admitted for elective embolization. The procedure was performed under general anesthesia via right femoral artery without complications. A 6 French guiding catheter was positioned in the left vertebral artery, and a flow-diverting stent was deployed covering the cerebellar artery origin. Using a microcatheter jailing technique inside the aneurysm, coils were deployed to guide flow. A total of 150 ml of contrast was used during the procedure. Control arteriography and cone-beam CT at the end of the procedure showed no complications. Upon awakening in the intensive care unit, the patient reported bilateral amaurosis. Skull arterial angiography was normal. Subsequent cranial MRI ruled out ischemia, leading to a diagnosis of contrast-induced encephalopathy (CIE). Intravenous corticosteroid therapy was initiated. The patient experienced complete visual deficit improvement over 72 hours. **Discussion:** CIE is an acute neurological disorder, typically reversible, attributed to intra-arterial administration of iodinated contrast medium, occurring in < 1% of procedures. The prognosis is excellent with unique supportive care. However, CIE can potentially cause permanent neurological dysfunction or death, such as persistent visual field defects, retrograde amnesia, and fatal cerebral edema. Under normal circumstances, iodinated contrasts do not cross the blood-brain barrier (BBB). However, they can disrupt the BBB, facilitating their own entry into the brain and causing neurotoxicity. Cortical blindness is characterized by complete loss of visual perception, optokinetic nystagmus with preserved ocular motility, normal pupillary responses, and normal fundoscopic examination. CB is the most common symptom of CIE, likely due to the relatively higher permeability of the BBB in the occipital cortex. Diagnosis of contrast-induced encephalopathy is made by demonstrating typical CT findings such as abnormal cortical enhancement with contrast and cerebral edema, or not, in a symptomatic patient after exclusion of thromboembolic and hemorrhagic complications. Neurological recovery typically occurs within an average of 2.5 days. Treatment of CIE is supportive, usually involving intravenous hydration, systemic corticosteroids, and osmotic diuretics. CIE can recur upon re-exposure to contrast although hydration and low volume of contrast can help in prevention. The risk/benefit ratio of re-exposure to iodinated contrast should be carefully evaluated. **Final comments:** Cortical blindness due to contrast-induced encephalopathy is a rare and potentially serious complication of contrast use, although this case had a favorable outcome.

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Case report: Creutzfeld-Jacob disease in young patient

Erick Dupont[1], Leonardo de Andrade da Cruz[1], Marcele Schettini de Almeida[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Leonardo de Andrade da Cruz; Email: leoandcruz@gmail.com.

Case presentation: A 54-year-old male patient, with no relevant medical history, presents with repeated episodes of behavior arrest, depressive mood, and insomnia. Two months later, with recent memory changes and gait instability, he was hospitalized for investigation. He presented complex visual hallucinations in the hospital and was then diagnosed with Acute Stress Reaction and taken home with Aripiprazole. The condition continued to worsen, especially with severe psychomotor agitation, leading to hospitalization three months after the onset of symptoms. The initial suspicion was autoimmune encephalitis, leading to CSF collection, without relevant changes. Skull Magnetic Resonance Imaging (MRI) demonstrated restriction of the diffusion of water molecules in the thalamus (Hockey stick sign) and cortical regions, especially on the left, a pattern highly suggestive of prion disease. Electroencephalogram (EEG) showed a pattern of short periodic waves, at which time the patient also had myoclonus, especially in the right hemisphere. Patient was discharged with a diagnosis of probable Creutzfeldt-Jacob disease - CJD (sporadic form), using Levetiracetam to control myoclonus, already with severe motor and cognitive impairment. A few weeks after discharge, testing for the 14-3-3 protein and RT-QuiC in the CSF came back positive, further corroborating the diagnosis given. **Discussion:** Prion diseases, such as CJD, when compared to others, are infrequent pathologies in everyday neurology. The disease affects 1 in every 1 million, with a ratio of 1 man to 1 woman. In its pathophysiology, the production of PrPSc (abnormal scrapie prion protein) occurs, which causes cellular changes with consequent neurodegeneration. The most common form is sporadic, which represents 80 to 85% of cases, without identifiable triggers, with the onset age between 55 and 65 years. The genetic form, for 10 to 15% of cases. Symptoms are non-specific, such as: headache, vertigo, fatigue and sleep disturbances. After a period, there is worsening of confusion, disorientation, in addition to symptoms of cooperation. The diagnosis is based on electroencephalographic changes (periodic complexes and triphasic waves), markers in the cerebrospinal fluid, 14-3-3 proteins and RT-QuiC, in addition to changes in imaging tests. Currently, without specific treatment, we prioritize support. The case portrays a patient with behavioral changes who underwent psychiatric and autoimmune diagnoses before prion disease was suspected. Although the definitive diagnosis occurs after a post-mortem biopsy, the sum of the elements presented (history, physical examination, MRI, EEG, and liquid analysis) allows a greater degree of security for these progressive and incurable diseases. **Final comments:** Due to its extremely guarded prognosis, its recognition, especially in cases of not so typical presentation, is essential to offer the best management for the patient and family.

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Case report: diarrhea induced by subthalamic nucleus deep brain stimulation

Glauco Kody Nagata[1], Letícia Bassani Devens[1], Marine Warmling[1], Sheila Trentin[1], Bruno Samuel Fraiman de Oliviera[1], Yuri Ferreira Felloni Borges[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Glauco Kody Nagata; Email: glauconagata@gmail.com.

Case presentation: A 62-year-old right-handed woman presented with predominant left-sided rigid-akinetic Parkinson's disease (PD) at the age of 55. Throughout disease progression, she developed medical refractory

motor fluctuations and dyskinesias and gastrointestinal intolerance to higher doses of Levodopa. Her current medication schedule at the age of 60 was Levodopa 600 mg/day (divided in 6 intakes), Amantadine 400 mg/day; Pramipexol ER 1,5 mg/day and Entacapone 200 mg TID. Therefore, she underwent bilateral Subthalamic Nucleus Deep Brain Stimulation (STN-DBS). During the following 14 months post-operatively, her medication was reduced, as Entacapone was discontinued, Amantadine was reduced to 100 mg/day, and her stimulation settings was L STN1 C+ 1- /5.0 mA/50 mcs/60 Hz; L STN2 C+ 8-/1.5 mA/60 mcs/60 Hz; R STN1 C+ 1-/5.3mA/50 mcs/60 Hz; R STN2 2.0 mA/60 mcs/60 Hz with good outcome. She developed motor fluctuations and bilateral tremor, so bilateral dorsal stimulation amplitude was increased to 1.7 mA on the L STN2 and 2.3 mA on the R STN2 with good symptom control. Although, 4 days after the adjustment, she presented profuse recurrent diarrhea and vomit, requiring three emergency department visits within one month that was persistent despite Ondansetrone 4 mg TID and Loperamide 2 mg 2 tablets a day. She underwent gastrointestinal laboratory tests, abdominal Computed Tomography, endoscopy and colonoscopy that were unremarkable. One month later, after direct contact with her attending neurologist, she was advised to revert the stimulator to the previous program. Two days after this adjustment, the patient reported a complete resolution of gastrointestinal symptoms. **Discussion:** In this reported case, a patient without prior complaints of constipation developed diarrhea following STN-DBS dorsal stimulation. Interestingly, this patient also experienced gastrointestinal side effects with Levodopa doses higher than 600 mg/day. Non-motor symptoms are common and play a significant role in the well-being of PD patients. Abnormal gastrointestinal motility is commonly presented in PD even before the motor features and deteriorates with disease progression. Its physiopathology is likely related to extranigral degeneration due to alpha-synuclein deposition, mainly in the peripheral autonomic nervous system. The effects of STN-DBS on gastric motility are not well understood. Stimulation may improve gastrointestinal functioning by activating fibers that project from or to the hypothalamus. Connections between the hypothalamus and the dorsal motor nucleus of the vagus nerve can strengthen this hypothesis. Red nucleus stimulation can also be involved in autonomic side effects, such as nausea, excessive sweating, and dizziness. **Final comments:** To our knowledge, there is no other case of stimulation-induced diarrhea reported. Further studies with larger cohorts are needed to better understand this mechanism.

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Case report: Foster-Kennedy syndrome associated with optic nerve sheath meningioma

Gilmar Silveira da Silva[1], Thaise Fellini Dal Moro[1], Luiza Toscan[2], Matheus Felipe Kuhn Urnau[2], Alex Roman[1]

[1]Universidade Federal da Fronteira Sul, Hospital São Vicente de Paulo, Passo Fundo RS, Brazil. [2]Universidade de Passo Fundo, Passo Fundo RS, Brazil

Correspondence: Gilmar Silveira da Silva; Email: silveirasilva.gilmar@gmail.com.

Case presentation: A 17-year-old previously healthy patient sought care due to loss of visual acuity on the right side for months. He denied headache or eye pain. Physical examination revealed amaurosis and mydriasis on the right, without direct photoreaction, and only consensual. Direct and indirect fundoscopy with pharmacological pupillary dilation was performed, which showed atrophy of the optic nerve on the right and papilledema on the left. Regarding the complementary tests requested to elucidate the case, the evoked potential showed a reduction in nerve conduction velocity in the visual pathway bilaterally, with accentuation on the right side. MRI of the skull and orbit showed an expansive tumor lesion in the paramedian sellar tubercle on the right extra-axial side, with dimensions of 10×7mm, with compression of the chiasm and extension to the pre-chiasmatic, intracanalicular, and intraorbital right optic nerve up to the middle third, suggesting an extensive meningioma of the optic nerve sheath. Laboratory test results and liquor analyses were unremarkable. After diagnosis, the patient underwent a microsurgical approach to resect the cranio-orbital tumor on the right side, associated with decompression of the optic nerve. The patient's complaints slowly improved over the days following the surgical procedure. **Discussion:** Foster-Kennedy syndrome is fundamentally characterized by atrophy of the optic nerve unilaterally, ipsilateral to an expansile mass, associated with contralateral papilledema due to increased intracranial pressure. It is estimated that the syndrome occurs in 1-2.5% of cases of intracranial tumors, with risk factors potentially associated with the condition: hereditary, female hormones, and previous radiation exposure. Among the tumors, those most associated with the condition are meningiomas, mainly in the subfrontal regions, olfactory groove, and sphenoid wing. Initial evaluation of these patients is usually performed using fundoscopy. Computed tomography and/or magnetic resonance imaging can be performed, the latter being responsible for images that allow a better assessment of the condition. Treatment varies depending on the case and can be carried out through surgical intervention, chemotherapy, radiotherapy, and/or drug therapy, most often using glucocorticoids to reduce peritumoral edema and intracranial pressure. **Final comments:** The case described demonstrates the importance of thorough evaluation of patients with visual changes, such as amaurosis, through fundoscopy, in addition to neuroimaging if a central etiology is suspected. Due to the lack of information about Foster-Kennedy syndrome and its potential complications, it is essential to conduct studies and disseminate greater knowledge on the subject to better define the management of the condition.

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Case report: inflammation related to cerebral amyloid angiopathy

Caroline Dresch Sabadin[1], Emilyane Batalha[1], Luana Lindner Cruz[1], Andressa Luiza Cintra Barbosa[1], Matheus Ramos Pinto[1], Lázaro de Lima[1], Paulo Victor Muller Francescon[1], Paulo Eduardo Mestrinelli Carrilho[1]

[1]Universidade Estadual do Oeste do Paraná, Cascavel PR, Brazil.

Correspondence: Caroline Dresch Sabadin; Email: caroldsabadin@gmail.com.

Case presentation: A 73-year-old female patient was referred to the neurology outpatient clinic of a tertiary hospital due to a headache, emesis, mental confusion and memory loss that began 1 month ago. At that time, the patient developed decreased verbal and motor responses, psychomotor agitation, and agnosia. A cranial computed tomography (CT) scan showed areas of diffuse vasogenic edema. The patient was treated with Phenytoin, Haloperidol, and Risperidone and admitted to the Intensive Care Unit (ICU), with dexamethasone added to the therapeutic regimen. The patient showed clinical improvement, but continued to experience anterograde memory loss. Outpatient follow-up revealed overall improvement, though episodes of mental confusion and memory loss persisted. Angiography revealed a mass effect with multiple foci of sparse subcortical microhemorrhages bilaterally, suggesting inflammatory amyloid angiopathy as one of the main hypotheses. Testing for anti-β-amyloid antibody was not performed due to service unavailability. During outpatient follow-up, ivermectin was initiated along with a plan for pulse therapy with methylprednisolone, followed by an oral taper. Currently, the patient is undergoing the tapering phase without complaints of mental confusion or memory loss. **Discussion:** Cerebral amyloid angiopathy associated with inflammation (CAA-ri) is a rare encephalopathy related to increased cerebrospinal fluid concentrations of autoantibodies directed against the β-amyloid protein deposited in the layers of cortical and leptomeningeal vessels. It primarily affects patients aged 60 to 80 years and is characterized by acute neurological symptoms ranging from cognitive disturbances and headaches to progressive cognitive decline, seizures, altered mental status, and focal neurological deficits. CT findings are characterized by unifocal cortical and subcortical hypodensity. On magnetic resonance imaging (MRI), white matter hyperintensities are observed, indicating vasogenic edema, along with cerebral microhemorrhages. Definitive diagnosis of CAA-ri requires histopathological confirmation. However, recent clinical-radiological criteria and tests for detecting anti-β-amyloid autoantibodies in the cerebrospinal fluid have reduced the need for invasive brain biopsies. Treatment consists of pulse therapy, with or without immunosuppressants. **Final comments:** The atypical case results in a diagnosis of CAA-ri, with initial symptoms of headache, vomiting, mental confusion, and recent memory loss. The final diagnosis was possible only through the clinical-imaging combination. Pulse therapy with methylprednisolone was effective in resolving symptoms. Discussing this case is relevant to broaden the understanding of CAA-ri in similar cases. Demonstrating the clinical-radiological criteria and laboratory testing avoids ineffective investigations, allowing effective treatment. Further research is needed to elucidate the underlying mechanisms of CAA-ri.

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Case report: multidisciplinary management and challenges in treating HTLV-1 in a pediatric patient

Laura Lorenzo Serra[1], Ana Clara de Lemos Guimarães[1], Joanna Sousa da Fonseca Santana[1], Amanda Hellen Sousa Leal[1], Daniel Abreu Santos[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Ana Clara de Lemos Guimarães; Email: anaguimaraes21.1@bahiana.edu.br.

Case presentation: An 11-year-old girl started outpatient treatment for HTLV-1 in 2023. Symptoms began at age 6 with walking difficulties, though developmental milestones were met. Her mother, diagnosed with HTLV-1, died in 2022, while her father has negative serology. She has five siblings, three positive for HTLV-1, one with virus-associated myelopathy. Diagnosed in 2020, she presented with severe urinary issues, including urgency, incontinence, and nocturia, and was diagnosed with neurogenic bladder related to HTLV-1, often linked to tropical spastic paraparesis (HAM/TSP). Psychological challenges included bullying due to her condition. Neurologically, she showed moderate spasticity and scissor gait. Treatment started with oxybutynin for incontinence, pregabalin for pain, and amitriptyline with prednisone for mood and motor symptoms. She experienced partial symptom improvement but continued to suffer frequent urinary infections, managed by infectious disease specialists. Psychological support improved her emotional state and school relationships. By September 2023, there was notable improvement in her depression and bladder function, though she maintained muscle atrophy and spasticity. Medications included amitriptyline 25 mg, prednisone, and oxybutynin 5 mg. A CSF examination for a definitive HAM/TSP diagnosis had not been performed. **Discussion:** Diagnosed at age 11, this case from Bahia – where HTLV-1 is highly prevalent, particularly in Salvador – illustrates the complex nature of this infection. The high transmissibility and severe impact highlighted by her family history underscore the urgency

for effective public health strategies and multidisciplinary approaches to improve life quality and control the spread. **Final comments:** This case emphasizes the importance of a personalized, continuous multidisciplinary approach involving neurology, psychology, infectious diseases, and dermatology to manage HTLV-1 challenges and enhance patient quality of life.

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Case report: multifocal acquired demyelinating sensory and motor neuropathy diagnosis after 15 years of evolution

Leonardo de Andrade da Cruz[1], Erick Dupont[1], Sônia Maria César de Azevedo[1], Mateus Paquesse Pelegrino[1], Davi Vargas Freitas Teixeira[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Leonardo de Andrade da Cruz; Email: leoandcruz@gmail.com.

Case presentation: A 71-year-old female patient with a previous diagnosis of Multifocal Motor Neuropathy (MMN) 15 years ago sought care for progressive dysphagia and dyspnea 2 months ago. Upon admission, in addition to the symptoms reported, she presented a reduction in strength exclusively in her hands, grade III, which was accompanied by significant atrophy and regional hyporeflexia, without other changes on the neurological examination. The investigation revealed paralysis of the right vocal fold and, in speech therapy evaluation, severe dysphagia, thus proving the involvement of bulbar nerves in the condition. Magnetic resonance imaging (MRI) of skulls, cervical spine and neck without clinically relevant changes, as well as liquor, with parameters within normal limits. The electroneuromyography (ENMG) performed at the service showed that, in addition to the motor impairment with conduction block in the upper limbs (UL), already documented in previous exams, there was now involvement of sensory fibers, with a demyelinating, asymmetric pattern and associated axonal degeneration. With these new discoveries, the patient was diagnosed with Multifocal Acquired Sensory and Motor Demyelinating Neuropathy (MADSAM), being discharged with a proposal to undergo Rituximab, but no longer responded to Human Immunoglobulin (IgH). **Discussion:** Even with technological advances in modern medicine, diagnosing peripheral neuropathies is still a challenge. Patient monitoring is crucial in these cases, as it allows for the correct diagnosis and therapy for the disease. The case exemplifies a patient who presented pure motor impairment for a long period, respecting a pattern compatible with MMN. After 15 years, she presented with asymmetric sensory and bulbar involvement, which fit into a new diagnosis of a rare variant of Chronic Inflammatory Demyelinating Polyneuropathy: MADSAM. The condition is characterized by being an acquired chronic demyelinating disease, with symptoms that appear as asymmetric weakness, mainly in the upper limbs, associated with sensory deficits caused by a demyelinating autoimmune response. It is considered a rare disease and is confused with other diagnoses, such as Carpal Tunnel Syndrome and Guyon Syndrome. Unlike Carpal Tunnel Syndrome, in electrophysiological examinations, MADSAM presents conduction block, in addition to asymmetric demyelination of multiple nerves. Furthermore, the 2 syndromes can be differentiated using ultrasound. Early treatment is recommended, as the disease responds well to intravenous human immunoglobulin and corticosteroid therapy. Based on this fact, we noticed that the patient was diagnosed at an advanced stage of the disease and had fewer responses to the therapies used. **Final comments:** The diagnosis of this condition remains a challenge, as it is a rare disease in clinical practice and it is necessary to know its characteristics, as early treatment presents a better prognosis.

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Case report: optic neuritis after dengue virus infection

Tiago Camilo Eisemberg de Alvarenga[1], Izac Lage Oliveira[2], Sergio Luiz Evangelista Santos[3], Larissa Oliveira Silva Leite Ferreira[3], Trícia Carolen de Almeida[2], Skárllatty Bretas Nunes[2], Amanda de Oliveira Tito Viana[3], Clara Dinalli Ornelas Iglesias[3], Ingrid Rodrigues Martins Silva[3], Lorena Rodrigues Dias Martins Silva[3]

[1]Hospital Nossa Senhora das Dores, Itabira MG, Brazil.
[2]Centro Universitário Funcesi, Itabira MG, Brazil.
[3]Hospital Margarida, João Monlevade MG, Brazil.

Correspondence: Tiago Camilo Eisemberg de Alvarenga; Email: tiagocea@hotmail.com.

Case presentation: Female patient, 53 years old, reports headache, prostration, inappetence and nausea lasting 5 days, being investigated and diagnosed with dengue (positive NS1 test). Self-limited condition with spontaneous resolution. However, she states that after 4 days of resolution of the initial condition, she began to notice pain in her eyes associated with a progressive worsening of visual acuity over a few days. Ophthalmological evaluation identified bilateral optic disc edema, with visual acuity restricted to visible figures. She underwent magnetic resonance imaging of the orbits which confirmed the clinical suspicion of bilateral optic neuritis, opting for pulse therapy with Methylprednisolone for 5 days. The patient showed improvement in eye pain after 5 sessions with methylprednisolone, followed by progressive improvement in visual acuity over the following days. MRI of orbits 02/24/24 showed hypersignal on STIR associated with possible gadolinium impregnation in the intra-orbital segments of both optic nerves. After 60 days of the condition, the patient reports that he continues to have daily difficulties related to low visual acuity, without other associated neurological deficits. **Discussion:** Dengue viruses have spread rapidly within countries and across regions in the past few decades, resulting in an increased frequency of epidemics and severe dengue disease, of multiple dengue virus serotypes. Dengue is caused by one of four single-stranded, positive-sense RNA viruses of the genus flavivirus. Infectious virus and the virus-encoded NS1 are present in blood during the acute phase, and high-level early viremia and NS1 antigenemia have been associated with more severe clinical presentations. Dengue has a wide spectrum of neurological complications such as encephalitis, myositis, myelitis, Guillain-Barré syndrome and mononeuropathies.The main ocular changes reported in the literature dengue-related hemorrhages include retinal and macular hemorrhages, peridiscal hemorrhage, Roth spots, diffuse retinal edema, cells in the vitreous, blurring of the margin of the optic disc, serous retinal detachment, choroidal effusion and non-specific maculopathy. Optic neuritis is an inflammatory infectious or demyelinating process of the optic nerve that may manifest as retrobulbar neuritis, papillitis, or neuroretinitis.The prognosis is generally good and resolving spontaneously, although some patients may report relative central scotoma that may persist for months. The use of steroids to treat visual changes is controversial. **Final comments:** The severity of ocular involvement varies greatly according to data observed in the literature. The use of corticosteroid therapy, although controversial, should be considered in severe cases, as well as complementary therapy with plasmapheresis. The knowledge of the various neurological complications helps in looking for the warning signs and early diagnosis for better outcomes.

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Case report: Parkinsonism-hyperthermia syndrome

Alberto Pereira Firmino Filho[1], Maria Weryca de Souza Belo Silva[1], Rodrigo Alencar e Silva[1], Clecio de Oliveira Godeiro Junior[1], Gabriel Soares Dutra de Souza[1], Felipe Toscano Lins de Menezes[1], Mario Teruo Yanagiura[1], Francisco Gil Sarmento Neto[1], Nathália Lindsay Pereira de Queiroz[1], Fátima de Menezes Dantas[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Alberto Pereira Firmino Filho; Email: firminofilho.ap@outlook.com.

Case presentation: A 62 year-old female with Parkinson's disease underwent Deep Brain Stimulation (DBS) placement in October 2023. The device was turned on after six days from surgery with worsening of dyskinesias leading to withdrawal of antiparkinsonian medications. The patient develops behavioral changes and agitation. There is a new DBS adjustment on the 18th post-operative day (POD) followed by the reintroduction of levodopa in low doses. Fluctuations in the level of consciousness persisted, with the appearance of drowsiness. From the 23rd POD onwards she started tachycardia, hypertensive peaks and hallucinations. Investigation for infectious was negative. The patient develops refractory hyperthermia (41 °C), worsening tremors, reduced level of consciousness, increased creatinine and CK levels (maximum 7,000U/L), and thrombocytopenia (54,000 U/mm^3). The hypothesis of Parkinsonism-hyperthermia syndrome complicated with renal failure due to rhabdomyolysis was raised, medications with dopaminergic blockade were suspended, antiparkinsonian medications were escalated, benzodiazepine was reintroduced, dantrolene was started with good control of tremors and correction of hyperthermia. There was no improvement in the level of consciousness and emergency orotracheal intubation was performed and transfer to the ICU. Over the course of days, the patient was able to walk, eat orally and communicate verbally without cognitive complaints. She was discharged with the DBS system on associated with pharmacological therapy. The patient had been using quetiapine regularly since 2019 due to a psychotic episode. **Discussion:** Parkinsonism-hyperthermia syndrome is a rare and potentially fatal complication that occurs when antiparkinsonian drugs are abruptly discontinued, and may also occur when the DBS system functions suddenly stops working. Other risk factors include dehydration, infections and use of neuroleptics. Symptoms are similar to neuroleptic malignant syndrome, with dysautonomia, hyperthermia, rigidity or tremors, and rhabdomyolysis classically within 7 days of changing the dopaminergic drug. Complications such as sepsis, respiratory and renal failure and seizures have also been described. Thrombocytopenia is rarely described in these cases and can be explained by the modulatory role played by dopamine in the platelet response. It is crucial to return to doses of dopaminergic drugs prior to the onset of the syndrome and reactivate the DBS system. The use of bromocriptine and/or amantadine during acute management, as well as the use of dantrolene as an adjuvant to control tremor and hyperthermia, are valid therapeutic options. **Final comments:** The acute worsening of parkinsonian symptoms associated with signs of dysautonomia should raise the suspicion of parkinsonism-hyperthermia syndrome, which may eventually lead to thrombocytopenia. Treatment is based on intensive clinical support, return of medications to usual doses and correction of risk factors.

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Case report: paroxysmal sympathetic hyperactivity syndrome in a patient with fat embolism

Lia Maki Hatisuka Imai[1], Adria Rodrigues da Silva[1], Bruna Beppler[1], Caroline Zapelini[1], Maria Caroline Martins de Araujo[1], Luis Sidonio Teixeira da Silva[1], Lucio Baena de Melo[1], Gabriel Rodrigues Espelho Rossi[1], Juliana Passos de Almeida[1], Luca La Valle Pedrao[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Lia Maki Hatisuka Imai; Email: hatisuka.imai@gmail.com.

Case presentation: A 21-year-old male, with no prior medical history, was admitted to a tertiary hospital after a motorcycle accident, presenting with closed fractures of the right lower limb (tibia, fibula, and femur). There were no signs of cranial trauma, and the initial neurological examination was normal. On the first day of admission, skeletal traction was performed. After two days, the patient suddenly became confused and agitated. Vital signs revealed tachycardia and desaturation, necessitating invasive ventilation on the third day of hospitalization. A chest and cranial CT angiography diagnosed pulmonary thromboembolism and raised the suspicion of pulmonary fat embolism; the cranial examination showed no significant findings. During the ICU stay, the patient developed episodes of hypertension, tachycardia, tachypnea, fever, sweating, and dystonic extension posturing. Initially, these episodes were misinterpreted as seizures, but an electroencephalogram ruled out epileptiform activity during the dystonic posturing. The patient remained febrile since ICU admission despite negative cultures. Cerebrospinal fluid analysis excluded central nervous system infections. Due to the recurrence of these episodes, cerebral fat embolism was suspected. Partial improvement was noted with the administration of morphine, gabapentin and baclofen. **Discussion:** Fat embolism is a rare but potentially life-threatening complication commonly associated with long bone fractures. The classic clinical triad of fat embolism includes respiratory distress, neurological impairment, and petechial rash; however, this patient did not present all these symptoms. Instead, he exhibited signs consistent with Paroxysmal Sympathetic Hyperactivity (PSH), characterized by episodes of hypertension, tachycardia, tachypnea, fever, sweating, and dystonic posturing. PSH can arise secondary to neurological injuries and is often misdiagnosed due to its similarity to seizure activity. The management of PSH in this case involved the use of morphine, gabapentin and baclofen, which partially controlled the symptoms. MRI was instrumental in confirming the diagnosis of cerebral fat embolism, showing multiple small hypointense foci in T2 gradient echo in the supratentorial white matter, predominantly in the subcortical region and corpus callosum (characteristic of fat embolism). **Final comments:** This case underscores the necessity of considering fat embolism and PSH in patients with multiple long bone fractures and acute neurological symptoms. Cerebral fat embolism can present atypically, making differential diagnosis critical for proper management. The use of advanced imaging techniques and a comprehensive therapeutic approach were vital in managing this patient. Effective monitoring and management of complications like PSH are essential for improving patient outcomes and quality of life.

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Case report: pericallosal artery stroke with multiple executive dysfunctions

Robercon Aves do Carmo[1], André Teixeira de Souza e Castro[1], Fernanda Sousa Lima[1], Larissa Lucas Schloicka[1], Rodrigo Torres Sena Pita[1], Carolina Flores Welker[1], André Gustavo Fonseca Ferreira[1], Wagner Afonso Teixeira[1]

[1]Hospital de Base do Distrito Federal, Brasília DF, Brazil.

Correspondence: Robercon Aves do Carmo; Email: robercon025@gmail.com.

Case presentation: A 47-year-old female patient, previously hypertensive, with chronic dialytic kidney disease caused by crescentic glomerulonephritis, atrial fibrillation, and rheumatic mitral valve disease. Admitted in December 2023 with severe expressive aphasia, central pattern facial paralysis on the right, and complete and proportionate right hemiparesis; due to these findings and being within the therapeutic window with unaltered tomography - aspects 10; she underwent chemical thrombolysis with alteplase. Subsequently evolved with difficult-to-control hypertension, remaining for three days on intravenous nitroprusside. A follow-up examination revealed hyperdense image formation compatible with hemorrhagic transformation (PH1-ECASS3) in the left anterior cingulate gyrus. Since admission, the patient had presented with emotional blunting, laconism, however, after 4 days of evolution, the patient began to show logorrhea, disinhibition, with recurrent mood variations, often incongruent with her current state (pseudobulbar affect), also reporting significant difficulty in recalling recent memories and performing calculations. Evaluated during hospitalization, she showed alterations in cognitive assessments (mini-mental state examination - MMSE) in recall memory, working memory, calculation, and attention. After controlling the hypertensive condition, defining the etiology of the stroke (cardioembolic), and initiating appropriate treatment, the patient continued in the neurology outpatient clinic. **Discussion:** Stroke is becoming increasingly important in public health, ranking among the top 10 most prevalent diseases in Brazil. It is important to have broad knowledge about the mechanisms involved in the pathophysiology of the disease and the wide range of symptoms associated with it. Studies show a significant impact on the lives of affected patients, both in motor and cognitive deficits, highly related to functional disability after the disease. **Final comments:** In this presentation, we describe a patient with a pericallosal artery stroke, showing multiple executive dysfunctions after the ischemic insult.

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Case report: progressive encephalopathy associated with BSCL2 gene mutation

Caroline Dresch Sabadin[1], Juliana Dourado Grzesiuk[2], Marta Regina Clivati[3], Matheus Pereira Guarini[1], Amanda Gonçalves Tochettto[1], Camila Fonseca Balcewicz[1], Estela Soares Carniel[1], Gabriel Santos de Almeida[1], Paulo Eduardo Mestrinelli Carrilho[1]

[1]Universidade Estadual do Oeste do Paraná, Cascavel PR, Brazil.
[2]FAG.
[3]Centro Clivati de Neurologia e Desenvolvimento.

Correspondence: Caroline Dresch Sabadin; Email: caroldsabadin@gmail.com.

Case presentation: A 24-year-old female was referred to the neurology clinic at the University Hospital in 2018. She exhibited delayed and regressive neuropsychomotor development – she lost the ability to walk by the age of six and also had an expressive regression on the ability to talk and communicate. At physical examination, she presented with spastic upper and lower limbs with decreased mobility and muscle strength. MRI showed signal changes and signs of volume loss in the cerebral white matter in the fronto-parieto-occipital region and ill-defined signal changes in the deep white matter of the cerebral hemispheres, particularly evident in the periatrial regions and adjacent to the posterior thirds of the lateral ventricles bodies. It also showed global signs of supratentorial and infratentorial brain volume reduction, bilateral and symmetrical, which is early for her age. Fundoscopy revealed slightly pale optic discs in both eyes with good symmetry and sharp borders, and biomicroscopy examination was normal, ruling out late-onset Tay-Sachs disease. Whole exome sequencing detected a heterozygous pathogenic mutation on gene STAC3, which is associated with congenital myopathy 13 when in homozygosity or compound heterozygosity, and also a variant of uncertain significance in the BSCL2 gene which may explain the clinical features of autosomal dominant Silver spastic paraplegia syndrome. The patient remains under outpatient follow-up. **Discussion:** Mutations in the BSCL2 gene, which encodes the seipin protein, can cause various neurological conditions, being 2 recessive: Encephalopathy progressive, with or without lipodystrophy, and Lipodystrophy type 2, and 2 dominant: Neuronopathy, distal hereditary motor, autosomal dominant 13, and Spastic paraplegia type 17 (SPG17), also called Silver spastic paraplegia syndrome. Despite the overlapping phenotypes of the dominant syndromes, Silver's disease causes more spasticity. Seipin is essential for lipid homeostasis; when mutated, it causes endoplasmic reticulum stress, leading to neurodegeneration. In the Silver syndrome, the p.N88S substitution results in cortical white matter anomalies, delayed and regressive neuropsychomotor development, spasticity, and visual and cognitive changes. The condition often manifests in childhood and adolescence with progressive weakness and stiffness of the lower limbs. Management includes physiotherapy and antispastic medications. **Final comments:** The uncertain variant in BSCL2 gene may be related to the spastic tetraparesis presented by the patient, since missense mutations in BSCL2 are associated with autosomal dominant motor neuron diseases and Silver syndrome, causing generalized congenital lipodystrophy, locomotor abnormalities and progressive encephalopathy. Children with Silver syndrome are at increased risk of developing motor, cognitive, speech delays, and learning disabilities. More studies are needed to provide a more accurate prognosis.

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Case report: progressive supranuclear palsy like of vascular etiology

Erick Dupont[1], Leonardo de Andrade da Cruz[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Leonardo de Andrade da Cruz; Email: leoandcruz@gmail.com.

Case presentation: A 71-year-old male patient was hospitalized in 2019 due to a sudden neurological deficit, right hemiparesis, dysarthria, and aphasia. MRI angiography demonstrated intracranial arteries with irregular parietal arteries, mainly in the distal bed. Subsequently, on angiography, these findings were corroborated, leading to a diagnosis of vasculitis involving the central nervous system (CNS). Due to the absence of involvement of other somatic sites, the diagnosis was primary CNS vasculitis. He was discharged with immunosuppressive therapy and returned again in November 2021 with a new deficit after attempting to wean off corticosteroid therapy. On examination, in addition to the sequelae of the previous event, he presented facial hypomimia, rigidity and bradykinesia in the upper limbs, worse on the left, exalted facial axial reflexes and restriction of vertical gaze upwards. New MRI showed several foci of ischemia throughout the brain parenchyma, highlighting an area with restricted diffusion of water molecules in the left cerebellar hemisphere. Furthermore, there was significant midbrain atrophy with numerous lesions with hypersignal on T2/FLAIR, secondary to old ischemic events. In this way, it was concluded that his vasculitis, through ischemia, promoted the intense volumetric reduction of the midbrain, thus demonstrating the clinical syndrome seen in patients with Progressive Supranuclear Palsy (PSP). **Discussion:** As part of tauopathies, PSP is a degenerative clinical syndrome. In these cases, the patient, in addition to the symptoms of a parkinsonian syndrome, also presents restriction of vertical upward gaze and, on imaging tests, intense volumetric reduction of the midbrain, forming the "hummingbird" sign. The case above portrays a patient who presented symptoms and images compatible with the diagnosis of PSP. However, due to his underlying pathology, it was possible to establish that his mesencephalic

atrophy was attributable to an ischemic etiology, and not degenerative, as seen in the classic picture. It is known that PSP of vascular etiology is rare and generally characterized by multiple areas of ischemia in different regions such as the brain stem, basal ganglia, thalamus, frontal lobe, and cerebellum, without presenting anomalous protein deposits, as in tauopathies, with involvement of the cortical-striatum-pallid-nigral-thalamo-cortical pathway, responsible for Parkinsonism. These patients have a high cardiovascular risk and multiple ischemic lesions on imaging tests and do not respond well to high doses of Levodopa. **Final comments:** Even in the face of an apparently classic picture, precise diagnoses cannot be given before the correct investigation of atypical parkinsonian syndromes. The case described, in which a vascular syndrome (vasculitis) mimics a degenerative syndrome (PSP), exemplifies

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Case report: reversible cerebral vasoconstriction syndrome in a young patient using thermogenic supplements and hormonal therapy for high-performance exercise

Thamira Espindola[1], Gabriel Martins Rodrigues[2], Pietra Molin Lorenzzoni[1], Lisandra Farias[1]

[1]Universidade do Sul de Santa Catarina, Florianópolis SC, Brazil.
[2]Hospital Governador Celso Ramos, Florianópolis SC, Brazil.

Correspondence: Thamira Espindola; Email: thamiraespindola@gmail.com.

Case presentation: A 28-year-old male was admitted to a tertiary hospital with an intense, persistent headache lasting over 72 hours. The patient reported a history of pressure-like headaches in the biparietal region, associated with photophobia, triggered by physical exertion and sexual activity. He was using Deposteron, Stanozolol, and Oxandrolone, with recent increases in dosages. The patient had a prior diagnosis of secondary syphilis and had completed a three-dose course of benzathine penicillin 30 days, with a treponemal test titer of 1:64. Cranial MRI demonstrated cerebral volumetric reduction and compensatory dilation of the sulci and cisterns, hyperintense areas on FLAIR and T2 sequences in the bilateral supratentorial white matter, suggesting possible microangiopathy, gliosis, or demyelination. There was irregular contour and slight caliber reduction in the left A1 segment and proximal portions of the right and left M1 segments. Cerebrospinal fluid (CSF) analysis was clear and colorless, with 1 leukocyte/mm^3, 2 red blood cells/mm^3, serum glucose 59 mg/dL, protein 64 mg/dL, and a non-reactive VDRL. Given the headache pattern, the patient was admitted for investigation of central nervous system vasculitis (primary or secondary), with the main differential diagnosis being reversible cerebral vasoconstriction syndrome (RCVS). The patient was treatment with crystalline penicillin G and was initiated for 14 days for possible meningovascular syphilis. Nimodipine 60 mg every 4 hours was also administered during hospitalization. A follow-up CT scan showed no stenosis, occlusions, aneurysms, or vascular malformations. The patient was referred for outpatient follow-up with instructions to discontinue the use of thermogenic supplements and hormones and to monitor for new headache episodes. **Discussion:** RCVS is a clinical syndrome characterized by severe, sudden-onset headaches, often described as thunderclap headaches. The pathophysiology suggests a transient disturbance in the regulation of cerebral arterial tone, often linked to the use of vasoactive substances such as alpha-adrenergic agonists. Diagnosis is based on clinical symptoms, neurological examinations, and cerebral imaging studies, which are used to assess cerebral blood flow and detect arterial vasoconstriction. Cerebral angiography is considered the gold standard for diagnosing RCVS, typically showing uniform narrowing of cerebral arteries. Treatment involves blood pressure control and symptom relief. Most patients recover completely from RCVS. **Final comments:** This case presented a differential diagnosis of RCVS and secondary syphilis, with the possibility that the syndromes might overlap. In this case the patient has no evidence of neurosyphilis in the CSF and has no complications in the control CT in conclusion the diagnosis is most likely a case of RCVS.

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Case report: rhombencephalitis by varicella-zoster virus (VZV) in an elderly and immunosuppressed patient

Patrícia Fagundes[1], Samia Talise El Horr de Moraes[1], Letícia Dall'agnol[1], Gustavo Figueiredo da Silva[1], Alana Bacelar Limeira Sales[1], Matheus Gomes Ferreira[1], Helio Afonso Ghizoni Teive[1], Luis Alfredo Olbertz[1], Lucas Wagner Gortz[1], Maurício de Carvalho[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Alana Bacelar Limeira Sales; Email: alanabls@hotmail.com.

Case presentation: A 68-year-old woman with a history of chronic lymphocytic leukemia, currently undergoing chemotherapy with chlorambucil, diabetes mellitus type 2, hypertension, glaucoma, and recent use of corticosteroids, was admitted for investigation of abdominal pain. Due to a characteristic rash in the dermatome of the right hypochondrium (T7 level), a diagnosis of Herpes Zoster was made. On the fifth day of hospitalization, the first day of acyclovir, she developed a moderate dysarthria, resting and action tremor of moderate amplitude and frequency, negative myoclonus and cerebelar ataxia of the upper limbs. After a syndromic and topographic diagnosis of rhomboencephalitis, further investigation was carried out. Brain MRI showed signs suggestive of microangiopathy, with no other significant changes. EEG showed signs of diffuse cerebral dysfunction; irregular bursts of delta pattern were recorded, as well as a single focal epileptiform activity over the temporal region of the left cerebral hemisphere. Initial CSF analysis showed hypoglycorrhachia and viral PCR with detectable Varicella Zoster Virus (VZV). Other analyses showed no abnormalities. Follow-up CSF on the ninth day of symptoms suggested lymphocytic meningitis: proteins 45 mg/dl, leukocytes 77/mm^3 (90% lymphocytes), glucose 42 mg/dl, and lactic acid: 2.1 mmol/L. Based on these findings, the diagnosis of VZV rhomboencephalitis was confirmed, and IV acyclovir treatment was maintained for 21 days. During hospitalization, the patient showed complete neurological improvement. At the outpatient follow-up after six months, she still had neuropathic pain in the affected dermatome. **Discussion:** Rhomboencephalitis is a clinical syndrome of inflammation of the brainstem and cerebellum, with a mortality rate between 4.3 to 12% depending on the case series and etiology. It can be secondary to infections, autoimmune diseases, and paraneoplastic syndromes, with VZV being a rare cause. Central nervous system infections by VZV have an estimated incidence of 1.02 cases per 100,000 inhabitants, with rhomboencephalitis being an even rarer complication. VZV is neurotropic, with a worse prognosis in immunocompromised patients, due to its ability to reactivate at multiple levels of the dorsal root ganglia and subsequent dissemination. In the above case, we identified several risk factors, such as immunosuppression due to chemotherapy, neoplasia, and corticosteroid use, as well as immunosenescence (40% of VZV reactivation cases occur over 60 years of age). **Final comments:** Rhomboencephalitis secondary to VZV infection is a rare, severe neurological condition with variable semiological presentation. Therefore, in view of the aging population and the development of immunosuppressive therapies, we present this case to highlight the importance of recognizing this condition.

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Case report: sensory neuronopathy secondary to paraneoplastic syndrome

Ana Luisa Rufino de Sousa[1], Amanda Soares Pimenta[1], Daniela Aparecida Lima Viana[1], Elisangela Ferraz Pazinato[1], Giovani Zago Borges[1], João Henrique da Matta Clementino[1], Alex Eduardo da Silva[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Ana Luisa Rufino de Sousa; Email: analuisardesousa@gmail.com.

Case presentation: Female patient, 49 years old, admitted to the emergency room with a history of weakness in lower limbs, associated with subacute onset global paresthesia. History of alcoholism, smoking, and breast cancer treatment. Neurological examination revealed anisocoric pupils (5mm on the right and 2mm on the left) and photoreactive, globally reduced muscle tone and trophism. Mild tetraparesis, tetraparesthesia with reduced tactile and painful sensitivity in upper limbs, lower limbs, and trunk. Hypopallesthesia and reduced osteotendinous reflexes in all segments. Evident sensory ataxia, with significant gait alteration and positive Romberg's sign. Under the syndromic hypothesis of sensory neuropathy (SN), complementary exams showed: normocytic and normochromic anemia, vitamin B12 deficiency, positive tumor marker dosage (CA 125, CA 19-9, and CEA), negative serologies for HIV, HTLV, toxoplasmosis, cryptococcosis. Cerebrospinal fluid biochemical analysis with 2 cells, 174 proteins, glycorrhachia of 55 mg/dL. Imaging exams included cranial and total abdominal computed tomography scans (CT) with no significant findings, chest CT showing marked irregular pleural thickening at the left lung apex associated with para-aortic lymphadenopathy. Electroneuromyography showing sensory neuronopathy. Thoracic magnetic resonance imaging provided clearer definition of the expansive lesion near the subpleural area of the left lung. The patient subsequently underwent pleural biopsy, which confirmed oat cell carcinoma. She was treated with methylprednisolone 1000 mg for 3 days, without satisfactory response, along with nutritional support and correction of vitamin B12 deficiency. **Discussion:** There are various manifestations of neurological syndromes in paraneoplasias. In patients with SN, 80% of cases originate from oat cell lung carcinoma. The immune-mediated etiopathogenesis can be explained by the presence of anti-neuronal nuclear antibody (ANNA-1) or anti-Hu in most cases, acting on intracellular tumor and neuronal antigens. Immunosuppressive treatment is indicated, but the response is not significant. Early diagnosis and treatment of neoplastic disease may stabilize or even regress the neurological condition. **Final comments:** Face of the syndromic picture of SN, it is necessary to establish the relationship with paraneoplastic syndromes and immediately start screening for the main associated primary neoplasms. Multidisciplinary follow-up and planning of symptomatic treatment of patients from a neurological perspective is important. However, diagnosing and initiating oncological treatment is the main modifying factor in the course of the disease for the patient.

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Case report: Takayasu arteritis leading to stroke

Julia Ágata Cardoso Barbosa[1], Samuel Natal Rodrigues[2], Drielle Nogueira Sales[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Julia Ágata Cardoso Barbosa; Email: Juliaa.cardoso12@gmail.com.

Case presentation: A 38-year-old female initially presented with left ankle edema on February 2024 and was evaluated by an orthopedic surgeon, finding no radiological abnormalities. On 03/30/2024, she returned to the hospital with left-sided hemiparesis, nausea, emesis, and facial asymmetry starting 13 hours prior. A cranial CT and CTA on 03/31 showed hypoperfusion in the right posterior cerebral artery territory, occlusion of the right common carotid artery, and critical stenosis of the left common carotid artery. Admitted to the ICU with over 12 hours since symptom onset, she presented left hemiparesis (grade 2 in the left lower limb, grade 1 in the left upper limb), left facial paresis, and mild dysarthria, NIHSS score of 7 indicating moderate neurological deficit. The arteriography revealed brachiocephalic trunk occlusion 2 cm from its aortic origin, severe left common carotid artery reduction, 50% left vertebral artery stenosis, and absent upper third basilar trunk and left subclavian artery visualization. A subsequent brain MRI confirmed right pons ischemia. Positive serum VDRL indicated neurosyphilis investigation. Physical examination revealed upper limb blood pressure discrepancies, stronger left radial, inguinal, and popliteal pulses, with systolic murmurs in the aortic and left subclavian arteries. Symptoms suggested Takayasu arteritis. Primary diagnosis: acute ischemic stroke in the right pons. Secondary diagnoses: critical left common carotid artery stenosis, right common carotid artery occlusion, neurosyphilis, suspected Takayasu arteritis confirmed with 12 points on ACR criteria. Treatment initiated with aspirin 300 mg + clopidogrel 300 mg, followed by aspirin 100 mg and clopidogrel 75 mg daily, corticosteroid therapy for vasculitis control, and penicillin for neurosyphilis. Intensive ICU care monitored ischemic progression, aimed to prevent new occlusions and ischemic events. **Discussion:** Takayasu arteritis, a granulomatous vasculitis affecting aortic and major branches, commonly affecting young women causing vessel stenosis, occlusion, or dilation with varying symptoms, including cerebral ischemia. Clinical and angiographic signs in this case included carotid and vertebral artery stenosis, brachiocephalic trunk occlusion, and absent vertebral and basilar trunk visualization, indicating severe vascular involvement. Neurosyphilis, though not primary, required concurrent treatment due to positive VDRL. **Final comments:** Early diagnosis and multidisciplinary management of Takayasu arteritis are crucial, highlighting improved patient outcomes. Differential diagnoses consideration and coexisting condition management, such as neurosyphilis, are essential for comprehensive care, emphasizing timely identification and treatment to enhance patient prognosis and quality of life.

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Catatonia as first presentation of anti-NMDA receptor encephalitis: a case report

Túlio Marlus Castro Lucena[1], Assis Porfirio Furtado Nogueira[1], Vitor Gustavo Leão Souto[2], Samyra Melo Vital[3], Patrícia Pereira Nunes Ribeiro[1], Maria Júlia Tabosa de Carvalho[2], Geovane Brito Affonso[2], Érika Santos Machado[1], Darlan de Araujo Benevides[1], João Vitor Nunes Sobreira Cruz[1]

[1]Hospital Geral do Estado, Maceió AL, Brazil.
[2]Hospital Universitário Professor Alberto Antunes, Maceió AL, Brazil.
[3]Hospital Metropolitano de Alagoas, Maceió AL, Brazil.

Correspondence: Túlio Marlus Castro Lucena; Email: tuliomarlus.neuro@gmail.com.

Case presentation: A 22-year-old man presenting with behavioral changes, characterized by apathy and catatonia, for approximately one month, was admitted to the Psychiatric Hospital. He developed seizures and fever and was referred for neurological evaluation. Cranial MRI revealed mild diffuse and symmetrical atrophy with slight gliosis of the bilateral hippocampal formations. A lumbar puncture was performed, and cerebrospinal fluid (CSF) analysis showed high CSF protein levels and normal cell count. The patient developed refractory seizures, progressing to status epilepticus, requiring intensive care unit admission and orotracheal intubation. Due to suspiction of immune-mediated encephalitis, plasmapheresis was performed with only partial response, followed by methylprednisolone pulse therapy, which yielded better results. After clinical stabilization, further investigation with CSF collection for immune-mediated encephalitis panel was conducted, which was positive for anti-NMDA receptor encephalitis. The patient was discharged on immunosuppressive therapy with cyclophosphamide and antiepileptic drugs and is currently under outpatient follow-up. **Discussion:** Anti-NMDA receptor encephalitis is a disorder affecting mainly young adults and children, with a higher prevalence in females, particularly in young adults. Symptoms include seizures, decreased consciousness, language disturbances, sleep disorders, and prominent psychiatric manifestations such as disorganized thinking, psychosis, hallucinations, and behavioral changes. Diagnosis is made through CSF analysis and confirmed by detecting IgG antibodies against the GluN1 (also known as NR1) subunit of the NMDA receptor. There is a demonstrated association with herpes simplex virus (HSV) infections and ovarian teratoma. Treatment involves immunomodulatory therapy with methylprednisolone pulse therapy combined with intravenous immunoglobulin or plasmapheresis, and tumor resection when appropriate. Additionally, some patients may benefit from maintenance immunomodulatory therapy to prevent recurrence. **Final comments:** Anti-NMDA receptor encephalitis should always be considered in cases of psychiatric alterations, as seen in this patient. The attending physician should seek signs and symptoms supporting or excluding this diagnosis and perform appropriate investigations. Early therapy improves patient response and outcomes.

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Catatonia due to herpetic encephalitis: a case report

Bruno Nunes Ferraz de Abreu[1], Flávia de Paiva Santos Rolim[1], Kevellyn Cruz Aguilera[1], Paula Camila Alves de Assis Pereira Matos[1], Amanda Vale Catunda[1], Dina Andressa Martins Monteiro[1], Elisa Gabriela Barros Cunha[1], Glauber de Menezes Ferreira[1], Fernanda Martins Maia Carvalho[1]

[1]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Kevellyn Cruz Aguilera; Email: kevellynaguilera@hotmail.com.

Case presentation: We report the case of a previously healthy 46-year-old woman who was admitted to the emergency department with a 24-hour history of new-onset seizures accompanied by fever and headache. Initial neurological assessment revealed mutism, marked waxy flexibility, catalepsy, and stupor, meeting the DSM-V diagnostic criteria for catatonia. She also exhibited nuchal rigidity and mild confusion, with no other neurological findings. A computed tomography scan was normal, and a lumbar puncture revealed an opening pressure of 20 cm H2O. CSF analysis showed 300 cells/μL and elevated protein levels (52.8 mg/dL). Empirical antibiotic treatment was initiated. A few hours after admission, she developed focal seizures with facial automatisms and status epilepticus. The patient was intubated and later transferred to the ICU. Following the initial analysis, a CSF PCR panel was performed, revealing a Herpes simplex type 1 virus infection. Based on this result, acyclovir was continued and the other antibiotics were discontinued. Brain MRI revealed bilateral cortical and subcortical FLAIR hyperintensities in the temporal and frontal lobes. She was treated with intravenous acyclovir for 14 days, showing marked improvement. The patient was discharged home after 40 days due to multiple respiratory infections associated with mechanical ventilation. She had no further seizure episodes after discharge and no motor disabilities. **Discussion:** Catatonia is a complex and not fully understood clinical syndrome marked by speech, behavior and consciousness alterations. Our patient had hallmarks for the diagnosis, waxy flexibility, mutism, stupor and catalepsy. This disorder presents in many psychiatric and clinical disorders. The neurologic disorder most associated with catatonia is autoimmune encephalitis, followed by metabolic disorders and drug abuse. Herpes encephalitis, whatsoever, unusually presents with catatonia, just 4 cases found in literature, with good response to intravenous antivirals. **Final comments:** This report adds to infectious encephalitis to the differential diagnosis when facing a new onset catatonia in the emergency department.

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Cathepsin A-related arteriopathy with strokes and leukoencephalopathy (CARASAL): a systematic review

Francisco Luciano Honório Barreto Cavalcante[1], Leonardo José Rodrigues de Araújo Melo[1], Pedro Vitor Ferreira Rodrigues[1], Lucas Soares Radtke[1], Emanuel de Assis Bertulino Martins Gomes[1], Cássia Caroline Aguiar da Ponte[1], Letícia Gadelha Caldas[1], Pedro Lucas Grangeiro de Sá Barreto Lima[1], Anderson Rodrigues Brandão de Paiva[2], Paulo Ribeiro Nóbrega[3]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Mendelics Análise Genômica, São Paulo SP, Brazil.
[3]Universidade Federal do Ceará, Departamento de Medicina Clínica, Fortaleza CE, Brazil.

Correspondence: Francisco Luciano Honório Barreto Cavalcante; Email: francski2304@gmail.com.

Background: Cathepsin A-related arteriopathy with strokes and leukoencephalopathy (CARASAL) is a rare monogenic disease associated with cerebral small vessel disease (cSVD). As a recently described disease, overall knowledge on CARASAL is still lacking in certain areas. Therefore, it is important to assess this disease's characteristics, such as prevalence, pathophysiology, signs and symptoms. **Objective:** This review aims to summarize the current knowledge on CARASAL, describing its genetic, pathophysiological, epidemiological and clinical aspects. **Methods:** We conducted searches in Pubmed and Medline databases with the terms "CARASAL" and "Cathepsin A-related arteriopathy with strokes and leukoencephalopathy". These searches yielded 17 articles. An 18th article was added due to its relevance, as it contained the possible first descriptions of CARASAL. After screening, 6 articles were included and analyzed. **Results:** CARASAL is a cause of stroke with leukoencephalopathy presenting between the third and fifth decades of life, with only 21 cases described in literature. Most patients were European, hailing from the Netherlands, Britain and France. One patient was Chinese, and another was Brazilian. CARASAL is an autosomal dominant disease associated with a c.973C>T; p.Arg325Cys variant in CTSA, located on chromosome 20q13.12. This gene encodes Cathepsin A (CathA), a protective carboxypeptidase that degrades Endothelin-1 (ET-1), which, in turn, is associated with vasoconstriction and inhibition of the maturation of oligodendrocytes. Astrocytes of CARASAL patients were found to be immunoreactive to CathA and ET-1. Their role in CARASAL's pathogenesis is unclear. Clinically, CARASAL patients present with many neurological findings, such as cognitive decline, migraine, stroke and movement disorders. Brainstem symptoms, such as hearing abnormalities, vertigo, dysphagia and facial pain, were also observed. Non-neurological findings were described, such as resistant hypertension, muscle cramps and sicca syndrome. Neuroimaging usually shows T2 hyperintensities in thalami, basal ganglia, brainstem, subcortical and deep white matter. Such lesions tend to be more widespread in older patients. Infarcts and

microbleeds are present, albeit mild in comparison to the leukoencephalopathy. There's no disease-modifying treatment currently available. Prophylaxis for stroke has been used in some patients. **Conclusion:** CARASAL is a rare cause of cSVD and leukoencephalopathy with very few cases reported in the literature. The actual prevalence of this disease is yet to be estimated. Despite its rarity, CARASAL should be considered as a differential diagnosis in middle-aged patients with stroke, otologic and brainstem signs or cognitive decline, in association with typical imaging findings. Further studies are needed to comprehend the disease's pathophysiology, evaluate prognosis and enable potential treatment strategies.

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Cauda equina syndrome as an initial manifestation of Burkitt leukemia

Carolina Oldoni[1], Matheus Piccoli Machado Schweitzer Klauberg[1], Cesar Augusto Weschenfelder[2], Lorenzo Ricardo de Medeiros Duso[2], Alessandra Aparecida Paz[1], Artur Francisco Schumacher Schuh[1], Pablo Brea Winckler[1]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Carolina Oldoni; Email: carololdoni15@gmail.com.

Case presentation: A 70-year-old male patient with psoriasis under treatment with secukinumab was admitted due to progressive worsening of paresis, which started in the distal left lower limb 1.5 years earlier and slowly progressed to paresis in the right lower limb accompanied by lower back pain, hypoesthesia in the lower limbs (LL) and genital region, constipation, and erectile dysfunction. Weight loss of 10 kg in 2 months following dietary changes was reported. Neurological examination revealed asymmetrical paraparesis, worse distally and bilaterally, hypoesthesia in the L5 and S1 dermatomes, and saddle hypoesthesia. The diagnosis of cauda equina syndrome (CES) was made. Extensive etiological investigation was performed, with initial normal cerebrospinal fluid (CSF) findings. Infections, neoplasms, paraneoplastic, structural causes, rheumatologic diseases, and sarcoidosis were ruled out. There was mild anemia (Hb 11.6), normal total leukocytes but lymphopenia, and an ESR of 80. Hematimetric changes were attributed to 7 monthly doses of SC methotrexate received in another clinic. MRI showed diffuse thickening of the cauda equina roots with focal contrast enhancement. Electroneuromyography was compatible with CES. A second CSF 9 days after the first showed 15 leukocytes (90% lymphocytes), with normal glucose and protein. An immune-mediated CES was considered. The patient was treated with methylprednisolone 1g for 5 days, with slight improvement. Nine months after, with no significant improve, prednisone 60 mg/day was started, which was used for 3 months until rehospitalization due to fever, hypoxemia, worsening of previous neurological deficits, anemia (Hb 9), thrombocytopenia (39,000), acute kidney injury, hyperferritinemia, and hypertriglyceridemia. Bone marrow biopsy diagnosed hemophagocytic lymphohistiocytosis (HLH) secondary to Burkitt leukemia (BL). Etoposide and dexamethasone (HLH-94 protocol) were initiated. After transient clinical improvement, the patient developed septic shock and died. **Discussion:** There are scarce case reports of BL presenting as a cause of CES, with CES occurring after the diagnosis of the neoplasm in those cases. In this case, the diagnosis of the hematological disease occurred afterward. It is inferred that the BL initially presented with CES, but the disease was possibly masked by chronic corticosteroid use. BL is an aggressive form of B-cell lymphoma, accounting for 1-2% of non-Hodgkin lymphoma cases. HLH is also rare, characterized by hyperstimulation of the immune system, leading to inflammation, increased cytokines, and multiple organ failure. HLH usually occurs in association with malignancy (especially lymphoma), immunodeficiency, or autoimmune diseases. **Final comments:** This case aims to report the presentation of Burkitt leukemia with cauda equina syndrome in a patient outside the epidemiological profile for the hematological diseases presented.

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CEBALID syndrome: a rare cause of genetic-based intellectual disability

João Vítor Moreira Nogueira[1], Oliver Reiks Miyajima[2], Alessandra Marangoni Fante[1], Bruna Barbosa Nobre[1], Enzo Lima Alcântara Parente[1], Daniela Vieira Cavalcante[1], Marcelo de Almeida Pinheiro[1]

[1]Unichristus, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: João Vítor Moreira Nogueira; Email: jvmnogueira10@gmail.com.

Case presentation: A 7-year-old male patient, with young, non-consanguineous parents and no family history of intellectual disability or other relevant neurological condition, had a follow-up with a pediatric neurologist due to a learning disorder. The physical neurological examination revealed apparent intellectual disability, significant impairment of expressive language, in addition to microcephaly, flat feet and craniofacial dysmorphisms such as macrotia, micrognathia, retrognathia, oblique palpebral fissure, thick eyebrows, large and pointed nose, high-arched palate and thin upper lip. The investigation identified mild bilateral sensorineural hearing loss, marked hyperopia and moderate intellectual disability with Intelligence Quotient (IQ) of 49. Additional investigation with karyotype, CGH-Array, electroencephalogram and magnetic resonance imaging of the brain showed no alterations. The whole exome sequencing identified a heterozygous mutation in the MN1 gene (c.1347_1348dup;p.[Tyr450Leufs*35]) compatible with CEBALID Syndrome with OMIM entry nº 618774, which does not present structural brain involvement in this case. **Discussion:** The MN1 gene is named as such as it was first identified as an oncogene in meningioma and may be associated with the development of this type of tumor. Another correlation of the pathogenic variants of this gene is with CEBALID syndrome, present in this case, with complex developmental disorder characterized by global developmental delay and variable impairment of intellectual development, craniofacial and structural brain abnormalities. The case presented is typical of this rare syndrome, as the patient presents craniofacial dysmorphisms, sensorineural hearing loss, language impairment and moderate intellectual disability, without brain abnormalities. Although the patient presents no family history indicative of an autosomal dominantly inherited disorder and no consanguinity among parents, whole exome sequencing encountered pathogenic MN1 mutation associated with CEBALID, indicating a de novo mutation. **Final comments:** CEBALID syndrome, related to the pathogenic mutation of the MN1 gene, is a rare autosomal dominant genetic condition, with few cases described in literature, presenting with intellectual disability associated with minor craniofacial dysmorphisms and significant impairment of expressive language. CEBALID syndrome, although rare, should be considered as a possible differential diagnosis in patients fitting in description, even without the presence of parental consanguinity or family history.

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Cenobamate for refractory epilepsy in a Brazilian private practice setting: a preliminary report

Felipe Couto Amendola[1], Rhuan Pimentel Nogueira[1], Matheus Parra Barcelos de Mendonça[1], Bruno Lucca Bonucci Alves Costa[1], Luiz Henrique Martins Castro[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Felipe Couto Amendola; Email: felipe.couto@fm.usp.br.

Background: Refractory epilepsy - a failure to achieve seizure freedom after two adequate trials of appropriately chosen, used and tolerated anti-seizure medication (ASM) schedules (ILAE) - affects up to one-third of epilepsy cases. In 2019, a novel drug, cenobamate, was FDA-approved for the treatment of focal seizures. Different studies have shown promising results of the drug in difficult-to-treat epilepsy. Barbieri et al. (2023) report a seizure freedom rate of 11-28% in treatment-resistant cases, notably higher than those of other ASMs. Cenobamate is currently available in Brazil only through importation, at an approximate R$1,100-R$2,200 monthly cost, depending on dose. **Objective:** To evaluate the impact of add-on cenobamate use in refractory epilepsy cases. **Methods:** This retrospective chart review study included consecutive cases of patients receiving cenobamate for refractory epilepsy in a private clinic setting of São Paulo. We excluded patients without follow-up appointments after cenobamate initiation. Primary outcome was the change in mean monthly seizure frequency after cenobamate introduction. Secondary outcomes were new reported side effects, total daily defined dose (DDD), seizure freedom rate and treatment cost. **Results:** We evaluated 9 patients (5 women), of whom 5 had lesional focal epilepsy, 3 non-lesional focal epilepsy and 1 generalized epilepsy. Mean age was 35,6 (28-48) years. Average cenobamate use duration was 15.4 (2.6-27.3) months. Cenobamate dose was 100mg in 1 patient, 200mg in 4, 300mg in 2, and 400mg in 2. Mean monthly seizure frequency was 5.3 before cenobamate use. After cenobamate introduction, seizure frequency dropped to 0.9 (p=0.004). Five (55.5%) patients achieved seizure freedom. Reported adverse symptoms after cenobamate introduction and change in other ASM doses were dizziness, somnolence, diplopia, ataxia, sedation and memory loss. Before cenobamate use, DDD was 5.5 and mean total treatment cost was R$2,560 (3 patients were undergoing vagus nerve stimulation, not included in the cost). After cenobamate introduction, mean DDD was 3.7 (p=0.018), and mean treatment cost, not including cenobamate cost, was R$1,238 (p=0.007) (only 1 patient was still under VNS). Tapering off of other ASM is still ongoing. **Conclusion:** Cenobamate is already being used in Brazilian private practice settings. As indicated in previous studies, our findings corroborate its impressive effectiveness in refractory epilepsy cases, reducing seizure frequency, total DDD, and, possibly, treatment cost. Cenobamate is a promising option in the current landscape of refractory epilepsy treatment. To our knowledge, this is the first study of cenobamate use in Brazil.

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Central foot drop: a clinical case of a patient with SPG 83

Diógenes Emanuel Dantas da Silva[1], Helmut Kennedy Azevedo do Patrocínio[1], Lucca Ferdinando Queiroz Fernandes[1], Maria Eduarda Medeiros Martins[1], Matheus Araújo de Medeiros[1], Artur Rebouças de Souza[1], Agábio Diógenes Pessoa Neto[1], Matheus Augusto Araújo Castro[1], Mário Emílio Teixeira Dourado Junior[1], Iury Hélder Santos Dantas[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Diógenes Emanuel Dantas da Silva; Email: diogenes.emanuel28@outlook.com.

Case presentation: Female patient, 17 years old, with a history of consanguinity. In 2022, she began to stumble when walking, which intensified in February 2023, when the patient let her shoes fall off her feet, requiring her to walk barefoot and with difficulty, dragging her leg. She also has cramps, paresthesias and progressive fatigue in her legs when walking. No alterations in the upper limbs. In childhood, she had difficulty jumping, as well as learning and reading difficulties. On physical examination, she had a difficult tandem gait, right-sided sputum and calf hypotrophy, increased patellar and achilles reflexes, bilateral Babinski and exhaustible clonus, cavus feet and preserved tone without spasticity. Laboratory tests, neuroimaging and electroneuromyography were unchanged. Exome sequencing found a probably pathogenic variant of the HPDL gene in homozygosity: NM_032756.4:c.553G> T:p.(Gly185Cys), associated with spastic paraplegia type 83. Treatment with baclofen and rehabilitation with motor physiotherapy was started, with symptom relief. **Discussion:** Hereditary Spastic Paraplegia (HSP) is a condition characterized by progressive degeneration of the corticospinal tract, and consanguinity is a risk factor for autosomal recessive forms. The genetic classification is based on the related gene and the associated inheritance pattern. It is numbered according to the chronological order of discovery as SPG1, SPG2 and so on. It shows genetic and phenotypic diversity, with HSP type 83 HPDL as the associated gene, on chromosome 1p34.1. Under normal conditions it encodes a 371 amino acid protein of unknown function, found in mitochondria, and highly expressed in the central and peripheral nervous system. Its phenotypic spectrum can range from neurological non-formation in the fetus to infantile onset spasticity. Its predominant phenotype is pure, with lower limb involvement, muscle weakness, hypotrophy, spasticity and gait alterations. Thus, foot drop is not one of its typical manifestations, as it is commonly related to lower motor neuron syndrome. The diagnosis is made by genetic analysis, when pathogenic variants in homozygosity or compound heterozygosity are found in the HPDL gene, after excluding differential diagnoses by neuroimaging, cerebrospinal fluid analysis, serologies and electroneuromyography. Current treatment is symptomatic. Future treatment depends on a better understanding of the function of the HPDL gene, with genetic counseling being part of the management. **Final comments:** Clinical presentations of pyramidal release, even with the phenotype that may resemble a superimposed lower motor neuron syndrome, should consider SPG in their differential diagnoses. This is important in cases with a family history of consanguinity, such as ours, in which the possibility of a genetic etiology was considered above all because of this characteristic.

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Central pontine myelinolysis not related to rapid sodium correction: a case report

Luiza de Aguiar Lima[1], Catarina Sodré de Castro Prado[1], Izabela Ferreira Pacheco de Oliveira[1], Ana Luisa Utrine Pimentel[1], Johnatan Felipe Ferreira da Conceição[1], Rafael Correa[2], Carolina Bittar Braune[1]

[1]Universidade Federal Fluminense, Hospital Universitário Antônio Pedro, Departamento de Neurologia - EBSERH, Niterói RJ, Brazil. [2]Universidade Federal Fluminense, Hospital Universitário Antônio Pedro, Departamento de Radiologia - EBSERH, Niterói RJ, Brazil.

Correspondence: Luiza de Aguiar Lima; Email: luizadealima@gmail.com.

Case presentation: A 40-year-old male was admitted to the hospital due to a sudden onset binocular visual blurring that had occurred a few days earlier. Furthermore, 3 months ago he began generalized weakness, fatigue, daily fever, anemia, thrombocytopenia and 25kg weight loss. After hospital admission, he evolved with weakness in the lower limbs and left upper limb, resulting in difficulty walking and gripping objects, along with persistent visual blurring and fluctuating consciousness levels. No prior similar medical history was reported, but heavy alcohol and tobacco use were noted. Neurological examination revealed grade 4 paresis in the left upper limb and in lower limbs, symmetric tactile hypoesthesia in legs, bilateral dysmetria in finger-nose and heel-knee tests, worse on the left side, and fluctuating consciousness. The hematology team established the diagnosis of lymphoma based on systemic findings, splenomegaly and bone marrow aspirate. Cerebrospinal fluid analysis showed pleocytosis with predominance of mononuclear cells (83%), mild hyperproteinorrhachia and a positive bacterial culture for Brevebacterium spp., that was regarded as probable contamination. Brain magnetic resonance imaging (MRI) showed a hypointense area on T1 and hyperintense on FLAIR in the central region of the pons, with hyperintensity in DWI sequence, but without restricted diffusion. Considering the clinical picture, brain MRI and CSF without findings suggestive of neuroinfection, the diagnosis of pontine myelinolysis of probable multifactorial etiology was established, including malnutrition, lymphoma and alcoholism. Chemotherapy for B-cell lymphoma was started, and the patient partially improved, being discharged with mild paresis and dysmetria, but clear resolution of level of consciousness. **Discussion:** Central pontine myelinolysis manifests with varied symptoms, mainly consciousness disturbances, cranial nerve dysfunction, and motor deficits. Although classically related to rapid sodium correction, which was not present in our case report, alcoholism, malnutrition, lymphoma and hypoalbuminemia may predispose to the disease. The main differential diagnoses are rhombencephalitis and central nervous system lymphoma, both extremely serious conditions with high mortality rates if not recognized and treated early. Therefore, additional investigation with MRI and cerebrospinal fluid is essential in the workup of these patients, as is follow-up. **Final comments:** This case highlights the importance of considering diverse etiologies beyond rapid sodium correction in pontine lesions. Multidisciplinary management is crucial for improved patient outcomes, despite limited treatment options.

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Central retinal artery occlusion in a young woman associated with isolated ophthalmic artery arteritis

Gustavo Freitas Alves de Arruda[1], João Paulo Borges de Melo[1], Mayko Nascimento Merscher[1], Raíssa Habka Cariello[1], Matheus Spilari[1], Camila Caroso Lobo[1], Danilo dos Santos Silva[1], Wagner Mauad Avelar[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Gustavo Freitas Alves de Arruda; Email: garruda@unicamp.br.

Case presentation: A 43-year-old woman with a history of vitiligo presented with episodes of amaurosis fugax in her right eye over the past 20 days. Initially, it had spontaneous resolution within seconds. However, she developed painless persistent visual loss two days before hospital admission. Neurological examination showed reduced right eye visual acuity, limited to hand movements, and ipsilateral nasal hemianopia. Fundoscopy revealed optic disc pallor and edema with a cherry-red spot at the macula, suggesting a diagnosis of central retinal artery occlusion (CRAO). Fluorescein angiography evidenced hyperfluorescence of the optic disc and three additional points nasal to the fovea without areas of capillary dropout or signs of retinal vasculitis. Computed tomography angiography of the head and neck showed no abnormalities. Extensive laboratory workup, with antinuclear antibodies, rheumatoid factor, infectious serologies, and inflammatory markers, returned negative results. However, tests indicated primary hypothyroidism with elevated thyroid peroxidase antibodies. Magnetic resonance angiography revealed mural contrast enhancement of the right ophthalmic artery on post-contrast T1-weighted images, leading to the diagnosis of focal arteritis. She was treated with oral prednisone 60 mg/day. At 1-month follow-up, there was a slight improvement in visual acuity, with finger counting up to 1 meter. **Discussion:** CRAO is defined by a partial or complete interruption of blood flow through the central retinal artery, leading to retinal ischemia with potential cell death within a few hours. There is a paucity of epidemiological data on the disease in Brazil, but it has an estimated occurrence of 1 case for every 100,000 people in the US and it's related to an increased risk of new strokes, with an incidence of 4-15% by 1-year follow-up. Its etiology is usually related to carotid atherosclerosis or cardioembolic disease, though it may also result from systemic vasculitis, hypercoagulable states, or infectious diseases. This case highlights a rare occurrence of CRAO due to isolated arteritis of the ophthalmic artery, without any evidence of carotid or cardiac emboli nor systemic or retinal vasculitis. The ophthalmic artery supplies both the central retinal and ciliary arteries that nourish the choroid and the posterior portion of the optic nerve. This explains why arteritis in such territory can lead to severe visual impairment. Cases of CRAO associated with isolated ophthalmic artery arteritis are exceedingly rare; the authors found only one similar case in the literature. **Final comments:** In this unique case, CRAO was attributed to isolated ophthalmic artery arteritis, independent of typical etiologies such as embolism or systemic vasculitis. The patient's partial visual improvement following oral prednisone may suggest that this intervention could perhaps offer a better chance for more significant visual recovery in similar cases with earlier diagnosis.

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Cerebellar ataxia as a presentation of neuro-Behçet: case report

Joyce Tolentino Philippi[1], Paulo Eduardo Mestrinelli Carrilho[1], Luisa Bianca Reis Parode[1], Márcio Augusto Nogueira[1], Ana Paula Adame[1], Gabriela Zimmermann[1], Alvaro Joel Santacruz López[1]

[1]Universidade Estadual do Oeste do Paraná, Hospital Universitário do Oeste do Paraná, Cascavel PR, Brazil.

Correspondence: Joyce Tolentino Philippi; Email: joycephilippi@gmail.com.

Case presentation: A 36-year-old female, who recently received treatment for ocular toxoplasmosis due to visual blurring, sought medical attention due to a condition of ataxia, dysarthria, and global paresthesia. Magnetic resonance imaging identified small cystic images in the brainstem and middle cerebellar peduncles (probable dilatation of perivascular spaces) associated with an enhancement focus, diffuse ischemic areas, as well as cerebellar volume loss. The patient was diagnosed with late-onset cerebellar ataxia and while being followed up by a neurologist for two years until she was referred to a rheumatologist. During the initial consultation with the rheumatologist, in addition to ataxia, recurrent oral and genital ulcers that started two years ago were identified, along with erythematous macules on the upper limbs, erythema nodosum, folliculitis, and pustules on the lower limbs. Furthermore, a new ophthalmological evaluation revealed active posterior uveitis. The combination of signs and symptoms, along with the absence of markers suggestive of other rheumatological diseases, led to the diagnosis of neuro-Behçet. The patient was initially treated with azathioprine 150mg/day as per follow-up, leading to a significant improvement in ulcers and other cutaneous lesions. Ten months after the start of treatment, there was a recurrence of posterior uveitis, necessitating a switch to adalimumab, one 40mg vial every two weeks, which resulted in permanent remission of cutaneous-mucosal lesions and visual symptoms. The patient maintained

ataxia and dysarthria, without worsening after the introduction of therapy. **Discussion:** Behçet's syndrome (BS) is a rare systemic vasculitis, with a prevalence of 0.003% in the Brazilian population, classically characterized by a clinical triad that includes recurrent aphthae, genital ulcers, and posterior uveitis. Approximately 10% of patients develop neurological involvement, named as neuro-Behçet's syndrome (NBS), a condition associated with worse prognosis and high morbidity and mortality. Parenchymal-NBS is seen in up to 75-80% and usually presents with an acute-subacute brainstem syndrome and/or a deep hemispheric involvement. The major symptoms and signs of p-NBS include headache, dysarthria, ataxia, and hemiparesis. And generally clear completely within weeks. However, a third of patients evolve with progressive course characterized by dementia, ataxia and dysarthria. **Final comments:** As a multisystemic condition, BS should be considered an important differential diagnosis in patients with neurological conditions, especially with cerebellar symptoms and brainstem involvement. The timely and appropriate diagnosis and treatment are essential to prevent possible irreversible sequelae and promote a better quality of life.

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Cerebellar stroke secondary to bow Hunter's syndrome: a case report

Pedro Cougo Samueli[1], Tauana Caroline Marconato[2], Izadora Celant Miranda da Silva[1], Elora Sampaio Lourenço[1], Ana Carolina de Lima[1], Samara Abdo El Hakim Kadri[1], Emanuel Cassou dos Santos[1], Matheus Kahakura Franco Pedro[1], Caio César Diniz Disserol[1,3,4]

[1]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[2]Universidade Estadual do Centro Oeste,Guarapuava PR, Brazil.
[3]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[4]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Pedro Cougo Samueli; Email: drpedrocougosamueli@gmail.com.

Case presentation: A 46-year-old male with no prior health issues, sought medical attention due to a sudden onset of vertigo, nausea and vomiting, lasting 6 hours. On admission he was asymptomatic and neurological examination was normal. A brain MRI scan revealed an acute stroke in the left cerebellar hemisphere and vermis, and a CT angiography showed an osteophyte at C6 causing stenosis in the V2 segment of the ipsilateral vertebral artery. Initial treatment included a loading dose of dual antiplatelet therapy. Subsequently, a digital subtraction angiography was performed, showing a significant left V2 caliber reduction at C6 level severely exacerbated by left lateral head rotation, confirming an osteophyte-induced stenosis. After stabilization and further evaluation by the neurosurgical team, who opted for a conservative approach, the patient was discharged for outpatient follow-up. **Discussion:** Bow Hunter's syndrome (BHS), also known as rotational vertebral artery occlusion syndrome, is a rare diagnosis with an undetermined true incidence. It represents an uncommon cause of vertebrobasilar insufficiency resulting from dynamic and reversible mechanical compression of the vertebral artery during cervical rotation or extension. Compression occurs due to the presence of abnormal bony structures such as osteophytes, disc herniations, tumors, trauma, or iatrogenic factors. It commonly involves compression of the dominant artery when the contralateral blood supply is limited. Ischemic injury results from blood flow stasis caused by mechanical tension or complete lumen obstruction, leading to endothelial damage and platelet aggregation, culminating in thrombus formation. This can result in transient ischemic attacks and strokes in the posterior circulation. Symptoms include vertigo, nystagmus, syncope, visual disturbances, and infarction in the brainstem and cerebellum. Symptom improvement in the neutral position is considered a pathognomonic sign of BHS. The primary diagnostic methods include CT, MRI and angiography. Currently, dynamic angiography is the diagnostic test of choice, with sensitivity and specificity close to 100%. Treatment with decompression of the affected artery appears to be an option with a low recurrence and complication rate. Other options include arthrodesis or conservative treatment combined with antiplatelet therapy, the latter showing a lower resolution rate. **Final comments:** Although BHS represents an exceptional diagnosis, this syndrome should be considered as a differential in ischemic strokes of the posterior circulation. Emphasis is placed on the importance of thorough investigation to elucidate the etiological factors of mechanical compression

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Cerebral amyloid angiopathy-related inflammation (CAARI): a case report and review

Mateus Coltro Dornelles[1], Jamileh Ferreira Chamma[1], Mariana de Oliveira Trintinalha[1], Liamara Petroli[1], Gabriella Maria Martins Favero[1], Letícia Graniska Salanti[1], Anelise Daiane Carpiné[1]

[1]Pontifícia Universidade Católica do Paraná, Hospital Universitário Cajuru, Curitiba PR, Brazil.

Correspondence: Mateus Coltro Dornelles; Email: mateuscdornelles@gmail.com.

Case presentation: A 56-year-old male, with a history of high blood pressure and dyslipidemia, on losartan and ciprofibrate, was referred to our service after a seizure episode associated with suspected intracerebral hemorrhage (ICH). On admission, he presented with lacunar amnesia. During the investigation, cranial magnetic resonance imaging (MRI) showed numerous spots with low signal on SWI, suggesting microhemorrhage in the periphery of the brain parenchyma, associated with vasogenic edema, mainly in parieto-occipital regions, a pattern suggestive of inflammatory cerebral amyloid angiopathy (CAARI), along with mild proteinorrachia (57.3mg/dL). Pulse therapy was administered with Methylprednisolone 1g/day for 5 days, and, at discharge, he was asymptomatic, being prescribed Phenytoin 300 mg/ day for prevention of further episodes. **Discussion:** Cerebral amyloid angiopathy (CAA) is characterized by the deposition of β-amyloid (Aβ) protein in the leptomeningeal and cortical blood vessels of small and medium caliber. Epidemiological studies on the disease are scarce and estimate a prevalence of 0.13/ 100,000 inhabitants. CAA can be divided into hereditary and sporadic, mainly affecting the elderly (average age of 67 years at diagnosis), with its main clinical manifestations being acute symptomatic lobar intracerebral hemorrhage, cognitive decline, transient focal neurological episodes, or behavioral changes caused by inflammation related to CAA (CAARI). CAARI is characterized by acute or subacute onset of cognitive decline or behavioral changes, and its pathophysiology is explained by three theories: coexistence of vascular Aβ and vascular inflammation, inflammation promoting Aβ accumulation in the vascular wall, and the possibility of Aβ deposition triggering the inflammatory response. Corovic A. et al, 2018, demonstrated the occurrence of proteinorrachia in over 80% of patients, with pleocytosis in 44%. To avoid the need for biopsy, diagnostic criteria for probable CAARI were created: acute or subacute onset of symptoms; age greater than or equal to 40 years; presence of at least one of the clinical features: headache, altered consciousness, behavioral changes, or focal neurological signs and seizures; MRI with irregular or confluent lesions weighted on T2/ FLAIR, presence of previous CAA characteristics (multiple cortical/ subcortical hemorrhages or microhemorrhages and/ or lobar intracerebral hemorrhage), and absence of other causes. Our patient met the proposed criteria and the response to pulse therapy reinforces the diagnostic hypothesis. **Final comments:** We report a case of a patient of atypical age with a clinical and imaging profile highly suggestive of CAARI. The importance of discussions on the subject is highlighted given the importance of early diagnosis for the implementation of therapy, which can modify the course of the disease.

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Cerebral compliance patterns during normal sleep.

Karla Carlos[1], Lívia Jane Galvão Farias[1], Douglas Sousa Soares[1], Ana Flavia Machado Pitz[1], Letícia Lambert[1], Lucas Martins Teixeira[1], Gilmar Fernandes do Prado[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Karla Carlos; Email: ka.carlos1@hotmail.com.

Background: The morphology of the intracranial pressure pulse consists of three waves: P1, corresponding to arterial pulsation; P2, corresponding to cerebral compliance; and P3, corresponding to aortic valve closure. Under normal conditions, P1> P2> P3. Reversal of the P2/P1 ratio represents impaired cerebral compliance (CC) and, indirectly, increased intracranial pressure (ICP). **Objective:** To evaluate cerebral compliance during normal sleep. **Methods:** Cerebral compliance analysis was performed using the Brain4care device placed on patients along with type 1 polysomnography. We evaluated CC during the all-night sleep period and analyzed it according to NREM and REM sleep stages. **Results:** We analyzed CC from 3 participants without sleep disorders. The mean age was 33 years (±3 years). Two participants were female. All participants denied comorbidities and medication use. The mean CC recording time was 363 minutes (minimum = 333; maximum = 385), totalizing 1,088 minutes analyzed. Participant 1. With 370 minutes of CC recording, it exhibited 22.5% REM sleep, 77.5% NREM sleep, and SE of 92%. The mean P2/P1 ratio during sleep was 1.28; 1.29 during REM sleep and 1.30 during NREM sleep. Participant 2. With 385 minutes of CC recording, it demonstrated 14.4% REM sleep, 85.6% NREM sleep, and SE of 84%. The mean P2/P1 ratio during sleep was 0.88; 0.84 during REM sleep and 0.87 during NREM sleep. Participant 3. With 333 minutes of CC recording, showed 28.8% REM sleep; 71.2% NREM sleep, and SE of 87%. The mean P2/P1 ratio during sleep was 0.83; 0.77 during REM sleep and 0.91 during NREM sleep. Only one participant showed P2/P1 ratio values above 1.0 during sleep, suggesting reduced cerebral compliance. We are just starting to understand this new technology, and this sort of result can be somehow explainable by variables that we do not are aware of. After conducting studies comparing invasive and non-invasive methods of ICP assessment, it was possible to establish values to stratify the risk of intracranial hypertension: green zone (P2/P1 0.65 to 1.00); yellow zone (P2/P1 1.00 to 1.20); red zone (P2/P1 >1.21). The higher the ratio, the greater the cerebral compliance disturbance. One of the components influencing ICP is cerebral blood flow, which varies according to the brain's metabolic demand, correlates with the dominant frequency of the electroencephalogram, and is higher during wakefulness. During NREM sleep, blood flow decreases by approximately 10 to 20%. This flow increases by 20 to 35% in REM sleep, compared to wakefulness. The main mechanism justifying this variation is the synergy between cortical activity and perfusion, which represents cerebral metabolism, which is decreased or increased during NREM and REM sleep, respectively. Thus, in normal sleep, an increase in compliance is expected during REM sleep and a reduction during NREM sleep. **Conclusion:** Normal sleep maintains cerebral compliance within the bounds of normality.

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Cerebral metastasis of lung cancer mimicking multiple sclerosis lesions.

Gabriela Lam Brawerman[1], Angelo Pontes de Freitas Campos[1], Denison Pedrosa[1], Luís Filipe Godoy[1], Gustavo Bruniera[1], Rodrigo B. Thomaz[1], Roberta Castro Linhares[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Gabriela Lam Brawerman; Email: brawermangabriela@gmail.com.

Case presentation: A 43-year-old female presented with symptoms of vertical diplopia and paresis in her left lower limb over the past two weeks. She reported no history of tobacco or alcohol use, and her past medical history was significant only for maculopapular lesions on the trunk, arms, and back, which resolved without intervention three weeks before the onset of neurological issues. Neurological examination revealed that she was alert and oriented with fluent language. Her visual acuity and fundi were normal in both eyes. A positive Bielschowsky sign was found on her left side. Based on the Medical Research Council grading, the patient had preserved power in all muscle groups, present deep tendon reflexes in the lower limbs, a positive Hoffman sign in the left upper limb, and bilateral flexor plantar responses. All sensation modalities were normal in all four limbs. Coordination was intact, with no gait disturbance. Meningismus was absent. A contrast-enhanced brain MRI showed a total of 16 lesions distributed in cortical and juxtacortical regions of both hemispheres, the insula, cerebellum, and mesencephalon on T2-FLAIR weighted images, with no oedema observed. Three of these lesions showed a central vein sign. A paramagnetic rim sign was also found. CSF analysis, oligoclonal bands, and kappa light chains were all unremarkable. An orbital MRI showed thickening in the left medial rectus muscle, an atypical finding for MS. A PET scan was performed for neoplastic screening, revealing an abnormal lung nodule and mediastinal lymph nodes. A biopsy confirmed the diagnosis of primary lung adenocarcinoma with ALK fusion. **Discussion:** The McDonald criteria were first introduced for MS diagnosis in 2001, and the latest revision made in 2017 emphasized the risk of misdiagnosis in clinical practice. Although patients could fulfill the criteria of dissemination in time and dissemination in space supported by MRI and oligoclonal bands, it is important to highlight that the MS diagnosis is still clinical, and potential differential diagnoses must be ruled out. Some data indicate that 30% of cases referred for MS Centers are at some point diagnosed with other diseases, and that over 50% of them carried the incorrect diagnosis for at least 3 years. Many different conditions can be confused with MS, such as migraines, cerebral small vessel disease, and neuromyelitis optica spectrum disorder, inflammatory, infectious, and metabolic disorders. Less frequently, breast cancer, colorectal cancer, and lung cancer are also considered in this differential diagnosis. **Final comments:** Although there has been an increase in the sensitivity of the McDonald MS diagnostic criteria over the past years, MS presents with clinical heterogeneity and a lengthy differential diagnosis. An MS misdiagnosis can be associated with serious risks and potential morbidity, and all patients presenting with lesions characteristic of MS must be considered for other potential diagnoses.

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Cerebral perivascular space abnormalities after COVID19

Clarissa Lin Yasuda[1], Benjamin Sinclair[2], Lucas S. Silva[1], John-Paul Nicolo[2], Brunno de Campos[1], Thais Maria Santos Bezerra[1], Terence O'Brien[2], Patrick Kwan[2], Meng Law[2], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Monash University,Melborne, Australia.

Correspondence: Clarissa Lin Yasuda; Email: cyasuda2807@gmail.com.

Background: Although different studies showed structural brain changes after COVID-19, the neurobiological mechanisms are unknown. Perivascular spaces (PVS) form a central part of the proposed glymphatic system, a cerebral analogue to the lymphatic system, responsible for waste clearance in the brain. PVS can be observed on clinical MRI as CSF-isointense vessel-like structures and have been reported to both enlarge and shrink in neurological conditions. We hypothesised patients with LONG-COVID would present with larger volumes of CSF, presenting a potential mechanistic pathway for the neurological and cognitive deficits observed in COVID and long COVID patients. **Objective:** To investigate PVS changes after COVID-19. **Methods:** 392 participants with previous mild COVID-19 and 212 healthy controls with no self-reported symptoms of SARS-COV-2 were scanned on a 3T (patients after 160 days of infection. Perivascular spaces were automatically segmented on the T1w image using an in-house trained nnUNet (a deep learning algorithm). The MNI 152 subject average 1mm T1 template was registered to each patients' T1w image using ANTS symmetric diffeomorphic transformation. The white matter(WM) regions of interest (based on the Harvard Oxford cortical atlases) were used to mask the PVS segmentation masks, and the volume of the WM for each participant calculated. Distinct PVS clusters were counted using a cluster-detection algorithm. To account for total WM volume, counts were divided by the WM total volume to give number of enlarged PVS per mm3 of WM (WM PVS number density: WMPVSd), which was used as the outcome variable. Groups were assessed for differences in outcome variables using a Mann-Whitney U-test. The effect of symptomatic SARS-COV-2 infection on each outcome variable was assessed using a general linear model with PVS as the dependent variable, COVID status as the independent variable of interest, and age and sex as covariates. **Results:** The WMPVSd were 1.61 (1.51-1.71) x 10-4 mm-3 and 1.78 (1.70-1.86) x 10-4 mm-3 in the control and covid group respectively. The COVID group had a significantly higher WMPVSd than the control group in a two-group comparison (U=35846, p=0.005). The general linear model indicated that WMPVSd was not associated with disease group =-8.85e-07 (5.78e-06), t=-0.153, p=0.878, but age was strongly associated with WMPVSd =3.555e-06 (2.166e-07), t=16.408, p<2e-16. A post hoc Mann-Whitney U test of the ages between the control and covid group indicated a significant between group difference (p<0.001). **Conclusion:** Although our data show a significant increase in WM PVS density in COVID patients compared to controls, this association was not significant after regressing out age and sex. These suggest that although the hypothesis of glymphatic involvement in COVID19 is reasonable, PVS as measured on MRI is not a good biomarker in COVID19, and that it is crucial to account for potential confounds such as age when drawing conclusions from PVS .

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Cerebral schistosomiasis due to Schistosoma mansoni infection: a rare case presentation in Brazil

Camila Lopes Figueiredo[1], Carolina Rodrigues Dal Bo[2], Maria Clara Motta Barbosa Valente[1], Leticia Machado Dumont[1], Rafael Chaves Claudino de Queiroga[1], Cicero Rodrigues Veloso[1], Filipe Brito Ferraz da Silveira[1], Antonio José da Rocha[1], Augusto Cesar Penalva de Oliveira[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Camila Lopes Figueiredo; Email: camila04_lopes@hotmail.com.

Case presentation: A 16-year-old female patient presented with a 2-week history of right hemicranial headache, nausea, vomiting, and visual field defect. She had no history of fever, chills, or neck stiffness. She was from the interior of Pernambuco and reported a history of swimming in lakes inhabited by freshwater snails. On examination, the patient had a left homonymous hemianopsia. Cranial computed tomography (CT) revealed a right occipital hypodense lesion with contrast enhancement. Brain magnetic resonance imaging (MRI) revealed cortico-subcortical signal alterations in the right cuneus and precuneus, characterized by intense linear and micronodular enhancement, indicative of the typical pattern observed in cerebral neuroschistosomiasis cases. Serological test for Schistosomiasis mansoni was positive. The patient was treated with praziquantel and corticosteroids. She evolved with complete improvement of symptoms after treatment. **Discussion:** Neuroschistosomiasis is a rare and potentially fatal complication of an infection by larval schistosomes. The primary species responsible for the disease are Schistosoma haematobium, Schistosoma mansoni, and Schistosoma japonicum. In Brazil, Schistosoma mansoni is the prevalent species of parasite responsible for schistosomiasis. It occurs due to the migration of Schistosoma eggs into the central nervous system, where they can lodge and cause granulomatous inflammation. S. mansoni typically affects the spinal cord, while Schistosoma japonicum mainly causes encephalic disease. The clinical presentation can vary widely but typically includes headache, focal neurological deficits, and seizures. Paucisymptomatic cerebral schistosomiasis is more common in patients with chronic S. mansoni infection, as indicated by autopsy studies, and can also manifest as tumor-like lesions. **Final comments:** The case underscores the importance of considering cerebral schistosomiasis in patients from endemic regions presenting with neurological symptoms. Early treatment can reduce the likelihood of disease advancement, thus enhancing overall patient outcomes.

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Cerebral superficial siderosis: challenges in diagnosis

Guilherme Araujo Natal Francisco[1], Nathalia Novaretti[1]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Guilherme Araujo Natal Francisco; Email: guilhermearaujonf@hotmail.com.

Case presentation: 80 years old woman with history of progressive weakness in lower limbs for 10 years, associated with paresthesia and fecal incontinence. Investigation with extensive thoracic spinal cord injury which was treated empirically as neurotuberculosis (CSF: BP=40cmH2O| 2 leukocytes| 56.3 protein| 60 glucose| 113 chloride| VDRL negative| Cultures all negative| CIE fungi negative; syphilis and HIV serology=negative; PPD=25; chest x-ray=calcium nodule in the upper third of the left lung, with a residual appearance, measuring 4mm). The patient remained stable after treatment, with ongoing control of the thoracic spine and improvement in urinary incontinence after implantation of a sacral neuromodulator. 2 years ago, patient sought care again due to worsening gait associated with cognitive impairment and ringing in the ears. On examination: MMSE=27/30, bilateral hearing loss, decreased strength on the left inferior member with hyperreflexia and Babinski's sign, bilateral hypopalesthesia (D>E), gait ataxia and positive Romberg's sign. Screening tests were unremarkable. MRI of thoracic spine (appendix 1): late post-treatment control of myelitis and arachnoiditis, with stable changes in relation to the examination on 08/13/2021. Brain MRI (appendix

2): changes suggestive of mild microangiopathy, characterized by foci of signal changes in the cerebral white matter; findings compatible with superficial siderosis, to a greater extent in the posterior fossa and characterized by superficial areas of marked SWAN hyposignal; cerebellar volumetric reduction. Arterial MRI angiography: small saccular addition image in the communicating segment of the left internal carotid artery suspicious for a small aneurysm; superficial siderosis predominating in the posterior fossa. Spinal and cervical angiography showed no changes. CSF: PA=6.5cmH2O| 5 cells | 8400 red blood cells | 72.9 proteins | 84 glucose | 131 chlorides | 2.9 lactic acid | negative cultures| negative direct mycological examination | negative VDRL). **Discussion:** Superficial siderosis is a rare neurologic disease with prevalence of 0.21% in those aged 50 to 69 and 1.43% in those older than 69, with males affected more often than females It is defined as hemosiderin deposition due to chronic leakage of small amounts of blood into the cerebrospinal fluid. The disease constitutes linear hemosiderin deposits in the leptomeninges and the superficial layers of the cerebrum and the spinal cord. The clinical manifestation of SS includes of progressive sensorineural hearing loss, cerebellar ataxia, and myelopathy. MRI is an important means of diagnosis. **Final comments:** It's a case of infratentorial cerebral superficial siderosis in an elderly woman secondary to a cerebrospinal fluid leak probably secondary to extensive spinal cord injury treated as neurotuberculosis. The importance of the case is to discuss the difficulty of investigation and treatment possibilities in these cases.

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Cerebral tuberculoma as a differential diagnosis for an extra axial lesion in an immunocompetent patient: a case report

Lucca Ferdinando Queiroz Fernandes[1], Matheus Araújo de Medeiros[1], Lúcio Fábio de Assis Arruda Filho[1], Helmut Kennedy Azevedo Do Patrocínio[1], Amanda Lima Sampaio[1], Artur Rebouças de Souza[1], Cláudio de Souza Fernandes Júnior[1], Renner Cassio Nunes de Lucena[1], Hugo Leandro Andrade Paiva[1], Iury Hélder Santos Dantas[1]

[1]Universidade Federal do Rio Grande do Norte, Caicó RN, Brazil.

Correspondence: Lucca Ferdinando Queiroz Fernandes; Email: queirozlucca@gmail.com.

Case presentation: A 26-year-old immunocompetent woman presented with a generalized tonic-clonic seizure. Brain magnetic resonance imaging (MRI) revealed an extra-axial lesion in the right temporal lobe with heterogeneous contrast enhancement, adjacent dural thickening and vasogenic edema. Analysis of the cerebrospinal fluid (CSF) revealed no significant findings. Excisional biopsy of the lesion for pathological analysis identified complete tuberculoid granulomas with central caseous necrosis, suggestive of cerebral tuberculoma. Based on these clinical, laboratory and imaging findings, a diagnosis of cerebral tuberculoma was made. The patient underwent the standard treatment regimen for tuberculosis (RHZE) for one year, with clinical improvement. **Discussion:** Although tuberculosis of the central nervous system is a rare manifestation, especially in immunocompetent patients, it should be considered in the differential diagnosis of expansive intracranial lesions. MRI is a crucial tool in detecting lesions with ring enhancement and perilesional edema, which can easily be mistaken for brain tumors. The diagnosis of cerebral tuberculoma was corroborated by the presence of caseous granulomas in the biopsy. Early treatment with the standard anti-tuberculosis regimen is essential for a good prognosis. **Final comments:** Mycobacterium tuberculosis infection in the central nervous system, manifested as tuberculoma, is rare but significant, especially in areas with a high prevalence of tuberculosis. Diagnosis can be challenging due to the similarity of clinical and radiological signs with other conditions, such as brain neoplasms. The integration of clinical, radiological and anatomopathological data is vital for the identification and effective management of this condition.

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Cerebral vascular accident: epidemiology of hospital admissions for ischemic and hemorrhagic stroke

Rynele Almeida de Fonseca[1], Deborah Araujo Silva[1], Annayza de Oliveira Bessa[1], Larissa Bianca Borsone[1], Amanda Oliva Spaziani[1], João Carlos Bizinotto Leal de Lima[1], Luis Carlos Spaziani[2], Gustavo Henrique da Silva[3], Vanessa Maria Gonçalves de Souza[1], Francieli Gomes Lima[4]

[1]Universidade Brasil, Fernandopolis SP, Brazil.
[2]Fundação Educacional de Fernandopolis, Fernandopolis SP, Brazil.
[3]UPA Santa Fé do Sul, Santa Fé do Sul SP, Brazil.
[4]AME Fernandopolis, Fernandopolis SP, Brazil.

Correspondence: Rynele Almeida de Fonseca; Email: rynelef_@hotmail.com.

Background: Cerebral Vascular Accident (CVA), commonly known as stroke, is a cerebrovascular disease caused by a sudden focal neurological deficit due to the acute absence of blood supply to the brain. This can be due to an occlusion caused by a thrombus, fatty plaques, or reduced cerebral perfusion from a ruptured vessel. It is one of the leading causes of death and disability worldwide, significantly affecting the quality of life of survivors. Stroke has a high mortality rate among cardiovascular diseases. It can be classified as ischemic or hemorrhagic, with the latter occurring when a vessel ruptures, causing blood to leak into the brain. **Objective:** To evaluate the profile of hospital admissions for stroke in Brazil between 2019 and 2023. **Methods:** Data were collected from the Hospital Information System of SUS (SIH/SUS) of the Ministry of Health, publicly available on Tabnet/DATASUS, from January 15 to May 15, 2024. This retrospective, longitudinal study utilized a quantitative, descriptive design. Data were grouped by Brazilian macroregions, type of care, sex, age group, and race/ethnicity. Statistical inference was performed using BioEstat 5.3 software, employing the paired T-test for two related samples to compare groups, with data presented through descriptive statistics, measures of central tendency, and dispersion. **Results:** Between 2019 and 2023, there were 859,733 hospital admissions for stroke in Brazil. Of these, 52.54% (n=451,666) of admissions were predominantly among males (p<0.0001) and 42.10% (n=361,920) among individuals of mixed race (p=0.0564). The majority of cases occurred in individuals aged 70-79 years, accounting for 26.37% (n=226,672; p<0.0001), with the highest concentration in the Southeast region (n = 363,567), representing 42.29% (p=0.0007) of the total, with an average annual cost of R$ 161,411,755.57 for care. Deaths were most frequent in the Southeast, with 42.77% (n=55,473) of fatalities (p=0.0008) and an average mortality rate of 28.17 (p=0.0008). The number of hospitalizations peaked in 2023, comprising 22.79% (n=195,953) of total admissions, compared to 17.88% (n=153,714) in 2020. Most admissions were classified as urgent, accounting for 96.48% (n=829,465; p<0.0001) of cases. **Conclusion:** Stroke admissions are concentrated in the Southeast region of Brazil, with higher proportions of deaths and healthcare costs during the study period. The predominant profile of cases involved males aged 70-79 years of mixed race, with urgent care being the most common type of admission.

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Cerebral vasculitis and pachymeningitis as the first manifestation of rheumatoid arthritis

Gabriel Etienne Brito de Salles[1], Giuliana Gonçalves Moreira Possidente[1], Marcos Martins da Silva[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Gabriel Etienne Brito de Salles; Email: gabrieletienne2009@hotmail.com.

Case presentation: A 60-year-old woman presented with recurrent rotatory vertigo, recent short-term memory loss, severe ataxia, fecal and urinary incontinence, and mild decline in sensorium starting 2 years prior to admission. One year before, she experienced an acute episode of binocular diplopia, left-sided eyelid ptosis and superior rectus muscle paralysis, followed by spontaneous improvement, and another episode of right hemiparesis. She denied previous joint pain or swelling. Neurological examination revealed ataxic gait, torsional nystagmus on left gaze, right hemiparesis, dysarthria, bilateral hyperreflexia with clonus, and a Mini-Mental State Examination score of 28. Laboratory tests were negative for antinuclear antibodies, ANCA, anti-dsDNA, anticardiolipin antibodies, anti-Ro, anti-La, anti-HIV, syphilis, IgG4, anti-HCV, and HBsAg. However, there were slightly elevated levels of anti-CCP (7.70) and rheumatoid factor IgM (5.9, with normal <5), and an elevated erythrocyte sedimentation rate of 46 mm/hour. Cerebrospinal Fluid Analysis revealed elevated protein levels, lymphocytic pleocytosis, and cerebral MRI indicated pachymeningeal and leptomeningeal enhancement, diffusion restriction in the left corona radiata, and lacunar ischemic infarcts in splenium of the corpus callosum, corona radiata, basal ganglia, thalamus and pons bilaterally. Angio MRI and Black Blood MRI showed no vascular abnormalities. **Discussion:** Rheumatoid arthritis (RA) can manifest in the central nervous system as compressive myelopathy from atlantoaxial dislocation, subdural hematoma, hyperviscosity syndrome, rheumatoid meningeal nodules or choroid plexus infiltration, pachymeningitis, leptomeningitis, and rarely cerebral vasculitis. Neurological signs of pachymeningitis and cerebral vasculitis can include focal deficits, epilepsy, cranial nerve involvement, headaches and altered mental status. CSF analysis in meningeal involvement may reveal mild lymphocytic pleocytosis and elevated protein, along with characteristic MRI findings of leptomeningeal or pachymeningeal enhancement, among 28 reviewed case reports, five described patients initially presenting with pachymeningitis symptoms as the first clinical manifestation of the autoimmune disease. Cerebral vasculitis typically affects those with long-standing, active, and erosive disease, MRI findings often include multiple bilateral ischemic lesions affecting cortical and subcortical regions and Anti-CCP, rarely studied in published case reports, tends to be positive, even at low titers. This case underscores the diagnostic challenge posed by these two rare neurological complications of RA. **Final comments:** Diagnosing cerebral vasculitis or pachymeningitis due to RA is challenging due to their rarity and limited literature. This case highlights their potential as initial presentations of RA, emphasizing the importance of early recognition and appropriate management.

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Cerebral venous thrombosis as a cause of subarachnoid hemorrhage and thunderclap headache: a very uncommon presentation

Amauri Pereira da Silva Filho[1], Danielle Cortellazzi Colonna Romano[2], Gabriela Yumi Nakamura[2], Melissa Fernandes Vilela de Freitas[2], Sabrina Borges Batista[2], Taianara Sampaio Reis[3], Natália Rebeca Karpejany[3]

[1]Hospital de Emergência e Trauma Dom Luiz Gonzaga Fernandes, Campina Grande PB, Brazil.
[2]Universidade Nove de Julho, São Paulo SP, Brazil.
[3]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Amauri Pereira da Silva Filho; Email: amauri.filho@yahoo.com.br.

Case presentation: Patient, female, 34-year-old arrived at the ER with a sudden and severe headache, described as the worst pain of her life, with visual blurring, nausea and vomiting, mental confusion, and disorientation. With fair general condition, with normal respiratory and cardiovascular functions, blood pressure: 150/80, no pathological history, and had been regularly taking oral anticoagulants (OAC) for the past 9 years. She had a Glasgow Coma Scale score of 14, isocoric pupils, photoreactive, without appendicular deficits, slight convergent strabismus, with right VI nerve palsy, and neck stiffness +2/+5. A syndromic diagnosis of headache was made, with subarachnoid hemorrhage (SAH) as the main hypothesis. A non-contrast CT scan showed a spontaneous SAH classified as Fisher 2. Angiography revealed no arterial vascular lesions and/or AVM. Venous study showed a significant filling defect in the superior sagittal sinus at the level of the confluence of sinuses. The diagnosis of cerebral venous thrombosis (CVT) was concluded in a patient initially presenting with SAH. **Discussion:** CVT is a neurovascular disease caused by a thrombotic occlusion of either a dural sinus or cerebral vein, and results in a variety of neurological symptoms. CVT can manifest as a single symptom, most common being headache, or it can present as a syndrome. The non-specific clinical picture makes diagnosing CVT difficult. Although the mortality rate of CVT has been significantly reduced by improvements in treatment and diagnostic techniques. The priority of treatment in the acute phase is to stabilize the patient's condition and to prevent or reverse cerebral herniation, surgical removal of the haemorrhagic infarct, or decompressive hemicraniectomy. Another option is anticoagulation with heparin. However, there is a risk to become a SAH. It is a neurologic emergency due to bleeding into the subarachnoid space and is a severe subtype of hemorrhagic stroke associated with a high risk of mortality (can reach 50%) and disability. The clinical presentation is most often in the form of headache, classically defined as maximal at onset and worst of life. It is uncommon for patients to have thunderclap headache (TCH), known as an excruciating headache that reaches maximal intensity within a minute. The principal etiology is SAH. Thus, patients with TCH must be evaluated urgently to identify the underlying cause and initiate prompt therapy. **Final comments:** CVT associated with SAH and ICH is an atypical pathology that requires early diagnosis and treatment. To reduce mortality, anticoagulation, patient stabilization, and multidisciplinary management alleviate symptoms and improve the severe prognosis. It is crucial to raise awareness among healthcare professionals for early diagnosis, and there is a need for more effective new therapeutic strategies in cases of thunderclap headache.

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Cerebral venous thrombosis associated with the use of testosterone implant: a case report

Caroline Zapelini[1], Lia Maki Hatisuka Imai[1], Bruna Beppler[1], Adria Rodrigues da Silva[1], Luis Sidonio Teixeira da Silva[1], Lucio Baena de Melo[1], Gabriel Rodrigues Espelho Rossi[1], Maria Caroline Martins de Araujo[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Caroline Zapelini; Email: zapelinicarol@gmail.com.

Case presentation: A 39-year-old female patient presented to a tertiary hospital due to a generalized tonic-clonic seizure lasting seconds, without sphincter release, and post-ictal drowsiness. The seizure was preceded by holocranial headache. The patient was previously healthy and denied any previous episodes. She reported starting testosterone implant therapy a week ago to enhance sports performance. She denied smoking. Neurological examination revealed no abnormalities. Laboratory tests at admission showed no alterations, and serologies were non-reactive. A CT brain revealed thrombosis of the superior sagittal sinus and bilateral fronto-parietal vein thrombosis, confirmed by a cranial CT angiography. Anticoagulation therapy was initiated, along with investigation for thrombophilias. Testing for factor V Leiden, prothrombin gene mutation, protein S and C, antithrombin, lupus anticoagulant, anti-beta2-glycoprotein, anticardiolipin, and homocysteine were negative. However, the testosterone level was 175 - reference value: 56. **Discussion:** Cerebral venous thrombosis (CVT) represents a diagnostic challenge, given its variability in clinical manifestations and low incidence among stroke cases. It occurs more often in women of reproductive age. The most commonly affected venous sinuses are the superior sagittal sinus and the transverse sinus, with the involvement of multiple veins being common. The etiology is related to hypercoagulability and there are various predisposing factors such as pregnancy, oral contraceptive use, postpartum period, thrombophilias, neoplasms, and autoimmune diseases. Additionally, the use of testosterone for aesthetic purposes, in high doses, is associated with an increased risk of CVT, especially in the early months of use, as seen in the reported case. This leads to an increase in red blood cell production, resulting in blood viscosity and hypercoagulability. Due to the vast etiology, other causes should be investigated, such as thrombophilias, which increase the risk of recurrence. In the reported case, a thorough investigation was conducted, ruling out other etiologies. Treatment should involve anticoagulation, with the duration varying between 3 months and one year depending on the transient or chronic nature of the risk factors. **Final comments:** The report presents a case of cerebral venous thrombosis associated with the use of testosterone. In light of this case, it is important to highlight the risks of testosterone supplementation in unindicated contexts and the role of health education in guiding the population.

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Cerebral venous thrombosis due to ChadOx1-s vaccine against COVID-19: pitfalls of VITT syndrome and response to factor-X inhibitors

Alexandre Kreling Medeiros[1], Marciéli Gerhardt[1], Fernanda Schuh Martins[1], Gabriela Heiden Telo[1], João Eduardo Tonini Bastianello[1], William Alves Martins[1]

[1]Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Marciéli Gerhardt; Email: marcieligerhardt@yahoo.com.br.

Case presentation: We report a previously healthy 24-year-old female who developed severe headache 5 days after vaccination with ChadOx1 nCOV-19. She had no viral symptoms and her PCR for COVID-19 was negative. At this time, she had severe thrombocytopenia (26.000/mm^3) and high levels of D-dimer. Initial CT and MRI of the head showed no sinus thrombosis. Nevertheless, her headache continued to progress, accompanied now by persistent vomiting. Two days later, she had two generalized tonic-clonic seizures and was transferred to our institution. At admission, she was dysnomic but readily responsive and alert, maintaining headache and recurrent vomiting. Head-CT revealed extensive bleeding in the left posterior temporal lobe. Additional exams showed high levels of D-dimer (30.871) and low fibrinogen levels. The diagnosis of Vaccine-Induced Immune Thrombotic Thrombocytopenia (VITT) was immediately considered. A CT venography scan confirmed extensive venous thrombosis involving the left sigmoid and transverse sinuses extending to the left internal jugular vein. Screening for other thrombophilias turned out negative. After considering the risk of worsening the hemorrhagic infarct with anticoagulation with heparinoids, apixaban 5mg BID was introduced. Treatment with IVIG (0.4g/kg/d for 5 days) was immediately started alongside with prednisone (1mg/kg/d). Nevertheless, on the second day of ICU her consciousness deteriorated, and an urgent CT scan showed an increase in intraparenchymal cerebral hemorrhage with global edema. Mannitol was then associated, with the other therapies being maintained, including anticoagulation. She had a neurological improvement in the ensuing days, being discharged from the ICU 5 days after. Follow-up MRI venography showed complete recanalization of the affected cerebral venous sinuses. **Discussion:** COVID-19 vaccine complications are known for rare cases of thrombocytopenia and thrombosis, mostly in cerebral venous sinus or splanchnic veins. This syndrome is called VITT and encompasses thrombocytopenia, high levels of D-dimer, low fibrinogen titles, and high level of antibodies to PF4–polyanion complexes. It has shown remarkable resemblance with heparin-induced thrombocytopenia, including presence of PF4-antibodies in almost all reported cases. Early and aggressive treatment decision in terms of anticoagulation for treating this condition is critical and deals with an expected increased mortality associated with enlargement of superimposed hemorrhage and ischemic injury. Although up-front treatment remains anticoagulation and IVIG, the effective onset time to treat with IVIG and other alternatives such as plasma exchange, which is suggested for refractory VITT, is still unclear. **Final comments:** Our case report shows that maintaining anticoagulation combined with IVIG therapy despite intraparenchymal hematoma expansion may lead to a favorable neurological outcome in VITT syndrome.

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Cerebral venous thrombosis following lumbar puncture: case report and considerations

Adriana Alves Rapp[1], Julia Cordeiro Braga[1], Renan Carvalho Castello Branco[1], Brenda Francisca Cardoso Brito[1], Victor Xavier da Silva[1], Camila Maria Barreto Ribeiro[1], Paulo Ricardo Sampaio de Melo[1], Filipe Nolasco de Souza e Silva[1], Lucas Vergne Cheade Lins[1], Marcel Leal Ribeiro[1]

[1]Hospital Santa Izabel, Salvador BA, Brazil.

Correspondence: Adriana Alves Rapp; Email: adrianarapp15.2@bahiana.edu.br.

Case presentation: A 34-year-old healthy female underwent spinal anesthesia for myoma embolization. Postoperatively, she developed a subacute, moderate headache that worsened when upright. Initially diagnosed with post-puncture headache, she received conservative treatment with partial relief. After two weeks, she returned to the emergency department due to worsening headache (rated 10/10) and vomiting, again worsened by standing. Hospitalized for a blood patch procedure, she experienced mild improvement. However, the headache persisted, now without a postural component. A neurologist consulted, and a brain computed tomography angiography (CTA) revealed extensive cerebral venous thrombosis (CVT) in the superior sagittal sinus. She returned to the emergency department for further investigation and management. She was not taking oral contraceptives and had no family history of thrombotic events, thrombophilias, or cancer. On admission, she was alert, oriented, and had no neurological deficits. Parenteral anticoagulation with enoxaparin was administered for five days, followed by dabigatran. She remained stable and was discharged with good pain control. Tomography and MRI revealed no ischemic areas or intracranial bleeding. **Discussion:** Post-puncture headache affects about 11% of patients after lumbar puncture (LP). While usually benign, cases have shown potential progression to iatrogenic intracranial hypotension with CVT, accounting for up to 2% of CVT cases. This association aligns with the Monroe-Kellie doctrine, where a reduction in cerebrospinal fluid volume leads to compensatory dilation and distortion of venous sinuses due to brain traction, increasing venous

stasis and predisposing to thrombus formation. A transcranial Doppler study showed a 50% reduction in superior sagittal sinus flow velocity post-LP, supporting this theory. Some cases suggest additional risk factors for CVT, questioning the isolated causal role of LP. Literature reviews indicate most individuals present with typical post-puncture headache symptoms, then experience a change in headache pattern, as observed in this patient. **Final comments:** Evidence is insufficient to support LP as an isolated CVT risk factor. However, the growing number of case reports and biological plausibility suggest heightened suspicion for CVT in patients with headache and alarm signs post-LP, particularly with known thrombotic risk factors. Conversely, it is necessary to actively investigate potential pathologies that predispose to CVT in those without risk factors.

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Cerebral venous thrombosis secondary to protein s deficiency: a case report

Bibiana Mayer[1], Chadi Emil Adamo[2]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Chadi Emil Adamo; Email: neurochadi@outlook.com.

L.A, male, 24 years old, anabolic steroid user, presented with a sudden episode of generalized motor dysperceptive crisis one year ago. He underwent a cranial MRI, which showed an extra-axial area of hyperintensity on T2 and hypointensity on T1 in the vicinity of the posterior and bilateral insula, more evident on the left than on the right, with some foci of diffusion restriction. A few days after the episode, hemorrhagic transformation was observed on the left, and he underwent drainage of the extra-axial hematoma. Several months after the episode, a venous phase arteriography was requested, which revealed thrombosis of perisylvian cortical veins bilaterally with partial recanalization. Subsequently, the patient presented with epileptic seizures nosologically categorized as non-dysperceptive and non-motor, focal, with pars opercularis syndrome, characterized by brief motor aphasias lasting seconds. The electroencephalogram indicated an epileptogenic focus with downward reversal in the T5 region. The patient also phonemic paraphasia errors, with increased effort for proper articulation. The following tests were performed: Laboratory tests: Hemoglobin 16.06 | Platelets 199,400 | Beta 2 glycoprotein 1 IgG 1.4 IgM 1.1 | Complement C3 177/ C4 29.0 | Homocysteine 8.91 | Anticardiolipin IgG 2.3 and IgM 9.4 | Lupus Anticoagulant 1.19 and 1.31 | Prothrombin Gene Mutation G2A21A negative | Functional Protein S 36% | Protein C Resistance negative | Factor V Leiden Mutation negative | Coagulation time for extrinsic, intrinsic, and common pathways within normal range. The protein S tests were repeated, as well as the quantitative value, and protein S deficiency was documented. Therefore, apixaban 5mg every 12 hours was chosen, and phenytoin was replaced with lamotrigine 300 mg/day due to memory complaints. The patient showed a satisfactory response. In this context, a diagnosis of cortical venous thrombosis with hemorrhagic complication was admitted. As a sequela, the patient developed language errors and epilepsy secondary to structural alteration. The most probable cause in this context is protein S deficiency. Additionally, the patient has a family history of thrombosis in his mother at the age of 20 and a sister with a previous miscarriage. Cerebral venous thrombosis accounts for 0.5% to 3% of all strokes. The most vulnerable populations include young individuals, women of reproductive age, and patients with a prothrombotic state. Predisposing factors for cerebral venous thrombosis (CVT) include oral contraception and hormonal therapies which can increase CVT risk nearly 8-fold, with obesity possibly adding synergistic effects. Other well-established risk factors are acquired thrombophilias (like antiphospholipid antibody syndrome), malignancies (particularly myeloproliferative disorders), autoimmune diseases and genetic thrombophilias (such as protein C and protein S deficiency, factor V Leiden, and prothrombin G20210A polymorphism).

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Cerebrospinal fluid AD biomarkers in patients with suspected normal pressure hydrocephalus

Vinícius Ribeiro Jeunon[1], Isabela Resende Silva[1], Vinícius Tinarelli Silva[1], Stephanny Pinto de Oliveira Lima[1], Jessica Abdo Gonçalves Tosatti[1], Jessica Diniz Pereira[1], Karina Braga Gomes Borges[1], Paulo Caramelli[1], Leonardo Cruz de Souza[1], Elisa de Paula França Resende[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Vinícius Ribeiro Jeunon; Email: vjeunon@hotmail.com.

Background: Alzheimer's disease (AD) and idiopathic Normal-Pressure Hydrocephalus (iNPH) are conditions that affect the elderly. AD and iNPH share several clinical features, which makes the differential diagnosis difficult. Additionally, the co-occurrence of neurodegenerative disease is controversial and may compromise the effectiveness of the ventriculoperitoneal shunt response. The detection of AD biomarkers in the cerebrospinal fluid (CSF) may be a possible resource to help distinguish these two conditions. **Objective:** To investigate and describe the profile of CSF AD biomarkers in patients with iNPH in a tertiary neurological service. **Methods:** We included eleven patients from an outpatient neurological service between 2022 and 2024. The inclusion criteria were the presence of at least one clinical manifestation of iNPH and the analysis of AD biomarkers in CSF. The biomarkers assessed were β-amyloid 1-42 (Aβ(1-42)), β-amyloid 1-40 (Aβ(1-40)), phosphorylated tau (p-tau), and total tau. We used the following cut-off values 1,100 pg/mL for β-amyloid 1-42 (Aβ(1-42)), >24 pg/ml for phosphorylated tau (p-tau), and >266 pg/ml for total tau to access the ATN status. We performed a descriptive statistical analysis to describe the main characteristics. The values of age and CSF biomarkers will be presented as mean ± standard deviation. **Results:** The group consisted of 63.6% men, with a mean age of 75.01±6.6 years, and a mean educational level of 11.6±6.0 years. The average time between the symptoms onset and the clinical visit was 5.1±4.6 years. The most frequent symptom was cognitive impairment (100%), followed by urinary incontinence (70.0%), and gait apraxia (63.6%). Only 54.5% of patients manifested all three symptoms simultaneously. Regarding AD biomarkers, the mean level of Aβ(1-42) was 609.2±292.3pg/mL (A-), of p-Tau was 62.6±45.3pg/mL (T+), of total tau was 348.9±45.3pg/mL (N+), and of Aβ(1-40) was 1861.39±3233.0pg/mL. Only 9.1% of patients were Aβ(1-42) positive, whereas 75% were t-tau positive, 77.8% were p-tau positive, and 100% were Aβ(1-42)/Aβ(1-40) positive. **Conclusion:** Patients with suspected iNPH that presents to a tertiary neurological unity have cognitive impairment as the most common symptom, with the classical triad in half of cases. The AD CSF biomarkers profile was overall not consistent with AD (A+) suggesting that the patients had non-AD neuropathological changes (SNAP) and maybe a good chance of responding to ventricular shunt.

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Cerebrospinal fluid HIV viral escape

Bruna Andrade de Oliveira[1], Brenno Tavares de Vasconcelos Brandão[1], Filipe Britto de Matos[2], Giuliana Gonçalves Moreira Possidente[2], Marco Antonio Lima[2,3]

[1]Universidade Federal do Rio de Janeiro, Instituto de Neurologia Deolindo Couto,Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Fundação Oswaldo Cruz, Rio de Janeiro RJ, Brazil.

Correspondence: Bruna Andrade de Oliveira; Email: brunaado97@gmail.com.

Case presentation: A 53-year-old woman, diagnosed with HIV infection two years ago and pulmonary tuberculosis at the same time and treated properly presented to the infection diseases clinic with memory difficulties and slowness in thinking, that had begun around one year and a half ago and remained despite the antiretroviral treatment for approximately a year. Her last blood HIV load was 30 copies/mL and T cell CD4+ count of 166 cells/mm^3. A brain MRI two months ago showed extensive areas of high signal on T2 and FLAIR diffusely distributed throughout the white matter, notably in the semiovale centrum, corona radiata and non-specific periventricular regions. She was admitted for investigation one month later, when she underwent a new MRI and lumbar puncture. Neurological examination showed cognitive dysfunction without focal deficits, but neuropsychological assessment was not available. Her MRI showed an increase in the areas of high signal on T2 and FLAIR, hippocampi with preserved signal and slightly reduced dimensions (MTA 2), and prominent cortical grooves, fissures and brain cisterns. The cerebrospinal fluid (CSF) results were: glucose 56 mg/dL, protein 50,4 mg/dL, cell count 12 white cells/mm^3 (all mononuclear), VDRL negative, CRAG negative and GeneXpert test for tuberculosis negative. The viral load in CSF was 8.960 copies/mL. The neurological findings associated with the neuroimaging pattern and the CSF HIV load exceeding the plasma HIV load were indicative of CSF viral escape leading to HIV encephalopathy. The antiretroviral drugs were changed to optimize treatment. **Discussion:** Encephalopathy is a common complication of HIV infection and should be suspected in the presence of cognitive dysfunction associated with brain atrophy and bilateral diffuse white matter abnormalities on brain MRI. The treatment consists in prompt administration of antiretroviral therapy in patients who have not yet started or were abandoning treatment or changing the antiretroviral regimen in those who do not have adequate control of the disease. An important issue is the best course of action in patients with cognitive complaints despite low or undetectable plasma viral load in good use of adequate antiretroviral treatment. Patients living with HIV presenting with new onset neurological symptoms and low or undetectable viral load in plasma benefit from the analysis of viral load in cerebrospinal fluid to assess CSF escape. CSF escape is a rare but important phenomenon because the antiretroviral treatment can be optimized based upon drug susceptibility and penetration in the central nervous system, leading to better outcomes. **Final comments:** When HIV encephalopathy is suspected in patients with a low or undetectable plasma viral load, it is imperative to assess the CSF viral load to search for CSF viral escape.

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Cerebrospinal fluid hypotension presenting atypically and with different types of headache: a case report

Apolônio Peixoto de Queiroz[1], Alex Tiburtino Meira[1], Rafael de Souza Andrade[1], Gilmar Leite Pessoa Filho[1], Ana Luísa Castelo Branco Gomes[1], Silvia Helena de Sousa Silva[2], Gabrielly Lopes Rodrigues[2], Yan Carlos de Sousa Diniz[2], Maira Monteiro Amorim[2], Joan Ravy Sousa Lacerda[2]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[2]Centro Universitário de Patos, Patos PB, Brazil.

Correspondence: Apolônio Peixoto de Queiroz; Email: apolopxq@gmail.com.

Case presentation: Female patient, 68 years old, hypertensive and without other comorbidities, presented clinical signs of cognitive impairment and headache. The clinical history began with forgetfulness and inattention in the last year. This was followed, months later, by orthostatic thunderclap headache and then mild motor incoordination with gait imbalance. In the last three months the headache pattern has changed, becoming mild, restricted to the occipital region, unrelated to position, described as a pressure-like pain, relieved by simple analgesics and worsening with the Valsalva maneuver. She was seen in an outpatient clinic, where anxiety, mild dysarthria, dysexecution on the clock test and dysmetria in the left leg were noted on neurological examination. Neuraxial magnetic resonance imaging (MRI) revealed typical signs of hypotension in the cerebrospinal fluid (CSF). Caffeine 300mg/day was started, resulting in small improvement. Cisternal scintigraphy did not locate any fistula. Empirical blood patching was performed. After the procedure and in outpatient follow-up appointments, she reported no headaches, with improvement in cognitive symptoms, according to family members. Post-procedure neurological examination showed improvement in gait, appendicular ataxia and cognitive tests. **Discussion:** Hypotension is a disorder of intracranial pressure, often caused by CSF leakage due to a fistula, which may be traumatic, spontaneous, or iatrogenic. The typical clinical presentation is an orthostatic headache. This patient initially presented cognitive symptoms, with a predominance of executive dysfunction, only later rising a typical headache but which changed its pattern, going from initially orthostatic and thunderclap to tension-type headache, unrelated to orthostasis. She denied any mild head trauma. Magnetic resonance imaging (MRI) of the neuraxial and cisternal scintigraphy were performed, which demonstrated classic signs of CSF hypotension: caudal descent of the cerebellar tonsils beyond the margins of the foramen magnum and extra-axial lamellar subdural collections in the convexities of the cerebral hemispheres, but not the possible leak site. After a joint clinical decision between neurology and neurosurgery teams, she underwent empirical blood patching with a clear improvement in all symptoms. **Final comments:** This case emphasizes the presence of different types of headache as a symptom of CSF hypotension, reinforcing the possibility of atypical clinical presentations even in the face of typical imaging findings. Furthermore, the patient's positive evolution highlights the importance of a rapid diagnosis, a multidisciplinary approach and adequate management in cases of CSF hypotension, ensuring a significant improvement in the quality of life of affected patients.

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Cerebrotendinous Xanthomatosis: electrophysiological and ultrasonographic characterization in a Brazilian study

Antonio Edvan Camelo Filho[1], Pedro Lucas Grangeiro Sá Barreto Lima[1], Paulo Ribeiro Nóbrega[1], Pedro Braga-Neto[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Antonio Edvan Camelo Filho; Email: edvan.camelo@gmail.com.

Background: Cerebrotendinous Xanthomatosis (CTX) is a rare autosomal recessive lipid-storage disorder related to bile acid biosynthesis pathways with pathogenic variants in CYP27A1. It leads to the increased formation and storage of abnormal lipid content, mainly cholestanol, in several tissues, especially tendons, eye lenses, and peripheral and central nervous systems. CTX has been recently recognized as a considerably underdiagnosed disease, a fact that might be related to a highly heterogeneous clinical presentation with a wide range of symptoms, severity, and age of onset. Peripheral neuropathy is a frequently reported and disabling feature of CTX. Assessing the peripheral nervous system plays a crucial role in the diagnosis and the ongoing monitoring of CTX. Peripheral nerve ultrasound is a valuable tool for diagnosing and monitoring hereditary neuropathies, but little is known regarding CTX. Recently, nerve enlargement was shown in one case report and one case series of 4 patients in CTX. **Objective:** To describe the patterns and extent of peripheral nerve involvement in Brazilian patients with CTX using nerve ultrasound, EMG, clinical manifestations, and laboratory biomarkers. **Methods:** Included in this study were 5 genetically diagnosed patients. The participants have been examined and received electrophysiological work-up, including NCS and nerve ultrasound, accordingly to a published protocol with a high-resolution probe (15 MHz linear probe, GE Logiq). Nerve ultrasound covered peripheral nerves on the right body site (median, ulnar, tibial, fibular, and sural nerve, as well as the C5 and C6 nerve roots and supraclavicular plexus), assessing the cross-sectional area (CSA) of peripheral nerves. **Results:** Of the 5 patients evaluated, the average age was 31.2 years, with an average diagnostic delay of 21.2 years, and a male-to-female ratio of 3:2. Four out of 5 (80%) of the patients presented with xanthomas. All patients had either sensory or motor complaints in the lower limbs, but only 2 of 5 (40%) had a polyneuropathy on EMG. All polyneuropathies were demyelinating (100%). All patients, including those with normal electroneuromyography, showed thickening of at least 1 nerve evaluated, with the supraclavicular plexus and median nerve at the elbow being the most accurate. **Conclusion:** CTX is an important condition to recognize and should be included in the differential diagnosis of any neuropathy of unknown etiology, even in patients without xanthomas. It is a treatable condition with chenodeoxycholic acid (CDCA), which inhibits cholestanol accumulation by reducing bile acid production. Our findings show the presence of proximal nerve enlargement in patients of CTX even before changes in EMG. Nerve ultrasound emerges as a promising biomarker for early disease progression detection and monitoring and establishing standardized endpoints for future clinical trials in CTX and other hereditary neuropathies.

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Cerebrovascular diseases by place o death: investigating associated factors and trends in an ecological time series study

Victor Hugo Palhares Flávio-Reis[1], Yago Marcos Pessoa-Gonçalves[1], Chamberttan Souza Desidério[1], Marlos Aureliano Dias-Sousa[2], Carlo José Freire Oliveira[1]

[1]Universidade Federal do Triângulo Mineiro, Instituto de Ciências Biológicas e Naturais, Laboratório de Imunologia e Bioinformática, Uberaba MG, Brazil. [2]Universidade Federal do Triângulo Mineiro, Hospital de Clínicas, Departamento de Neurologia, Uberaba MG, Brazil.

Correspondence: Yago Marcos Pessoa-Gonçalves; Email: yagompg98@gmail.com.

Background: Cerebrovascular diseases (CD) have been described as one of the leading causes of mortality today. An important factor associated with this spectrum of diseases is the location where the patient dies. This outcome is underexplored in the literature and deeply connected to several determinants such as the response time of emergency medical services, sociodemographic factors that influence the pursuit and access to emergency medical care, and disparities in transportation availability between urban and rural areas in accessing healthcare services, among others. **Objective:** To analyze deaths by CD based on the location of patient death and examine the factors and trends associated with this issue. **Methods:** This is an ecological time-series study. Data on the place of death were obtained from the National Mortality Information System, with sociodemographic variables analyzed for the period from 1996 to 2022. Data for determining the number of deaths per 100.000 inhabitants were obtained from the Brazilian Institute of Geography and Statistics. Subsequently, the data were tabulated using Microsoft Excel version 2309 from 2019, and statistical analyses were conducted using GraphPad Prism version 8.0.0 and Joinpoint Trend Analysis Software version 5.2.0. **Results:** A total of 2,565,168 deaths from CD were recorded in Brazil between 1996 and 2022. The majority (44.2%) were caused by stroke. 80.9% took place in hospitals and other health establishments, 16.9% at home and public places represented 0.45%. The analysis showed a statistically significant decreasing trend in the number of deaths in hospitals and other health establishments over the years (Tau = -0.276, p=0.04536). The federative unit with the highest number of hospital deaths per 100,000 inhabitants was Rio de Janeiro, with 57 deaths per 100.000 inhabitants. The state with the highest number of home deaths was Piauí, with 23 deaths per 100.000 inhabitants. Individuals aged 80 years or older presented the highest mortality rates in hospitals (787/100.000 inhabitants), households (32/100.000 inhabitants), and in public roads (2/100.000 inhabitants). Individuals with 9 to 11 years of education represented 0.3% of deaths, while individuals with no education represented 20% of deaths. Finally, regarding race, the Black population showed the highest mortality rate in hospitals (42/100.000 inhabitants), while Asians presented the highest mortality rate at home (8/100.000 inhabitants). **Conclusion:** It is concluded that the majority of deaths occur in hospitals, but a considerable number of deaths occur in other locations, such as at home. Black men aged 80 years or older, with low educational attainment, and residents of states in the southeastern region of Brazil accounted for the majority of deaths due to stroke in hospitals. Therefore, further studies need to be conducted to better elucidate the relationships between deaths and the place of death of patients.

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Cervical and dorsal pain are more prevalent in individuals with headache: preliminary data from a Brazilian cohort

Fabíola Dach[1], Heloisa Betiol[1], Viviane Cunha Cardoso[1], Marco Antônio Barbieri[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Fabíola Dach; Email: fabioladach@usp.br.

Background: Studies show the existence of comorbidity between headaches and other painful conditions. Patients with headaches are twice as likely to report musculoskeletal pain than those without headaches. Another study showed that 83% of headache patients have neck pain, with high frequency and moderate intensity. The factors involved in the comorbidity between painful conditions are obesity, physical inactivity, anxiety, depression, stress, occupational factors, and genetic factors. **Objective:** To verify the prevalence of pain in another topographies in individuals with headache, whether there is an association between pain in other topographies and the presence/frequency (episodic/chronic) of headache, and to check if there is correlation between the frequencies of pain in another topographies and headache. **Methods:** An analytical and descriptive study from a cohort of live births (1978/79) in Ribeirao Preto/SP. The variables of interest were: presence/frequency of headache in the last three months. Pain in another topography and the place of greatest pain considering the last three months. The regions were separated: cervical, dorsal, lumbar, upper limbs, lower limbs, abdomen, thorax and pelvis. **Results:** 1775 individuals attended this interviewed (average age:38.13±0.579, 52.3% female). Sixty three percent reported headache in the last 3 months (16.3% chronic headaches). Among patients with headaches, 51% had pain in another topography. Among patients with headache and pain in another topography, pain in the cervical

region was observed in 12.1%, dorsal region in 8.7%, lumbar region in 37.4%, upper limbs in 8.7%, lower limbs in 24.6%, abdomen in 3.2%, chest in 0.9% and pelvis in 4.4%. There was an association between headache and pain in another topography [X2(1)=21.743, p=0.001], with the cervical (82.9% versus 17.1%) and dorsal (79% versus 21%) regions being more prevalent among individuals with headache [X2(7)=17.778, p=0.013]. There was an association between pain in another topography and chronic headache [X2(1)=5.079, p=0.024]. There was no correlation between the frequencies of headache and pain in another topography (rs=0.165, p<0.001). **Conclusion:** The prevalence of pain in another topography was 51% among individuals with headaches. An association was observed between pain in another topography with headaches and chronic headaches. Pain in the cervical and dorsal regions was significantly more prevalent in individuals with headaches. There was no correlation between the frequencies of pain in another topography and headache.

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Cervical compressive myelopathy due to ossification of the posterior longitudinal ligament: a case report

Rayanne Pereira Mendes[1], Saulo Barros Teixeira[1], Marco Túlio Sette dos Santos[1], Atilio Gomes Romani[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Rayanne Pereira Mendes; Email: rayannepmendes@gmail.com.

Case presentation: A 63-year-old female patient presented with a six-month history of progressive motor weakness and sensory alteration in all four limbs, worse on the left side. She was admitted to the hospital following a fall from standing height, which worsened her pre-existing symptoms. On admission, she was alert and oriented in time and space, with mildly asymmetric spastic tetraparesis: motor strength was grade 3 in the left upper limb, grade 2 in the left lower limb, and grade 4 in the right side, with no other neurological abnormalities. Cervical spine MRI showed extensive calcification of the posterior longitudinal ligament, causing cervical canal stenosis from C2 to C6, extending over 5.5 cm, most notably at C4-C5 with over 50% stenosis, and associated with a signal alteration in the spinal cord at C4-C5. The diagnosis was Cervical Compressive Myelopathy due to Ossification of the Posterior Longitudinal Ligament. The patient underwent C3-C4-C5-C6 laminoplasty with immediate postoperative improvement in motor potentials and long-term segmental strength improvement. **Discussion:** Ossification of the posterior longitudinal ligament occurs as heterotopic bone forms in the posterior longitudinal ligament, resulting in neural compression, myelopathy, and radiculopathy, most commonly in the cervical spine. It is more frequent in East Asian populations, with prevalence rates of 1.9% to 4.3% reported in Japan, and is rare in other populations, with prevalence ranging from 0.1% to 1.7%. Patients with neurological symptoms may require surgical decompression. Cervical laminoplasty and laminectomy are two surgical approaches used to treat spinal cord compression often caused by spinal stenosis, herniated discs, or other degenerative pathologies. Cervical laminoplasty involves reconstructing the lamina, a bony part of the spine, to widen the spinal canal and relieve pressure on the spinal cord and nerves, preserving vertebral structure and stability, being less invasive, and potentially offering faster recovery and lower long-term deformity risk. It is more suitable for patients needing multisegmental decompression. Laminectomy may be preferred when complete removal of the lamina is necessary to relieve symptoms. **Final comments:** In cases of progressive spastic tetraparesis, cervical spine MRI is crucial for diagnosis and treatment planning, with Ossification of the Posterior Longitudinal Ligament being a condition to consider, especially in extensive longitudinal lesions. Proper surgical technique can provide a good prognosis.

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Cervical dural arteriovenous fistula presenting as acute extensive longitudinal transverse myelopathy: a case report

Camila Lorenzini Tessaro[1], Catarina Dantas Corrêa[1], Gustavo Figueiredo da Silva[1], Henrique Lacerda Lage Lopes de Oliveira[1], Maria Cláudia Hahn Ferrucio[1], Luana Antunes Maranha Gatto[1], Paulo José Lorenzoni[1], Caio César Diniz Disserol[1], Marcos Christiano Lange[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Catarina Dantas Corrêa; Email: catarinadcorrea@gmail.com.

Case presentation: A 20-years-old female with no known comorbidities presented with ascending paresthesias in the lower limbs associated with progressive gait impairment, evolving into wheelchair-bound over 45 days. She was admitted to another hospital and discharged without specific investigation or treatments, with the hypothesis of Guillain-Barré syndrome, because she partially improved and regained the ability to walk with support. After five days the patient experienced acute tetraplegia, T2 sensory level, dysphonia, dysphagia, and urinary incontinence. She was admitted at our hospital for investigation. Due to bulbar symptoms and signs of respiratory failure, the patient was intubated and transferred to the Intensive Care Unit. Brain CT revealed hypoattenuation of the cervical spinal cord, and cranial and cervical CT angiography revealed sigmoid sinus thrombosis associated with venous engorgement in the posterior fossa and around the cervical spinal cord. Spinal cord MRI showed extensive longitudinal transverse myelopathy from the medulla to T1 level, and prominent flow voids along the cervical and thoracic spinal cord segments. Serum and CSF analysis were normal. Anti-aquaporin 4 antibodies were negative. Digital angiography diagnosed and treated a dural arteriovenous fistula supplied by the middle meningeal, occipital, and pharyngeal arteries, draining into the sigmoid sinus. The fistula was successfully embolized without complications. The patient remains under intensive care with partial improvement. **Discussion:** Spinal dural arteriovenous fistulas (SDAVFs) are the most common type of vascular malformation affecting the spinal cord. They usually occur spontaneously but can be secondary to cerebral venous thrombosis. Due to venous congestion and edema a progressive myelopathy may occur, which can mimic polyneuropathy/radiculopathy due to the ascending motor and sensory symptoms. This case is peculiar because acute presentations are quite rare and because the most common location for SDAVFs is the thoracic spine, while cervical locations are extremely rare. Commentary The authors report a rare case of extensive longitudinal myelopathy secondary to a dural arteriovenous fistula at the bulbocervical junction, treated by endovascular embolization.

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Challenges and decisions at the start of Pompe disease treatment: case report

Camila Irini Oliveira Perivolaris[1], Melissa Pereira de Oliveira[1], Maria Izabel Alves Rodrigues[1], Fernanda Broch[1], Giovana Ribeiro[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Camila Irini Oliveira Perivolaris; Email: camilairini@gmail.com.

Case presentation: Child, 11 years old, male, born in Machado - MG, in routine examinations with a pediatrician in 2022, an increase in transaminases was observed. During follow-up, laboratory changes persisted. Associate had elevated creatine phosphokinase and decreased alpha-glucosidase dosage. Genetic testing was requested, which revealed a heterozygous pathogenic variant in the GAA gene. Young man, 15 years old, male, born in Jundiaí- SP, was undergoing regular routine monitoring when he observed increased creatine phosphokinase in three consecutive doses, associated with an increase in alanine aminotransferase, aspartate aminotransferase and lactate dehydrogenase. Asymptomatic patient. A broad panel of neuromuscular diseases was carried out with the following result: two heterozygous variants in the GAA gene. **Discussion:** Pompe disease, a metabolic myopathy, is caused by mutations in the gene that codes for acid alpha-glucosidase (GAA), the enzyme that breaks down glycogen. There are two types of involvement - based on the onset of symptoms and the presence or absence of cardiomyopathy. If the disease occurs early in life, the disease manifests itself with more severe symptoms, known as classic infantile-onset Pompe disease. It is characterized by a clinical condition that begins before 12 months of life, with hypertrophic cardiomyopathy, respiratory distress, muscle weakness and left ventricular outflow obstruction. Another group of patients may have late-onset Pompe disease, which manifests itself after 12 months of life, and may have cardiac involvement, with milder consequences. The progression of symptoms is generally slower, with a poor outcome. For the treatment of the disease, enzyme replacement therapy is available - human recombinant alpha-glucosidase acid. Indicated in the first months of life for cases with early onset and in late cases but with symptoms present. **Final comments:** Currently, there are promising drug therapies capable of preventing the progression of the neurodegenerative condition when used in the early stages of the condition. It is important to highlight that treatment must be individualized as the severity of the disease varies. The progressive nature of the disease that affects the cardiac and skeletal muscles, resulting in death from respiratory insult, must be considered. It is known that starting early treatment in symptomatic patients helps preserve motor function and slow the progression of the disease. In asymptomatic carriers, the indication of drug treatment should be considered. There is still a lack of data to prove better long-term results compared to starting treatment after symptoms appear. Factors such as age, patient wishes and family must be taken into consideration. An obvious harm with early diagnosis is the psychological consequence. Knowing about the disease years before it appears can be devastating. The beginning of psychological therapy should be considered in these patients.

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Challenges in diagnosing of inclusion body myositis (IBM) in Brazil: revisiting the new diagnostic criteria

Carolina Braga Moura[1], Nicole Mancen Freire Rosa[1], Gabrielle Silva Sales[1], Izabela Ferreira Pacheco de Oliveira[1], Izabela Jardim Rodrigues Pitta[2], Caroline Bittar-Braune[2]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.
[2]Universidade Federal Fluminense, Hospital Universitário Antônio Pedro - EBSERH, Departamento de Neurologia, Niterói RJ, Brazil.

Correspondence: Gabrielle Silva Sales; Email: gabrielle.sales1@unemat.br.

Case presentation: A 53-year-old female patient presented with a six-years history of progressive dysphagia, including an episode of bronchoaspiration pneumonia requiring intensive care unit admission. Slowly after the onset and progression of dysphagia, she noted an evolution of muscle weakness, with cervical involvement, distal involvement of the upper limbs and facial paresis. She was unaware of her family history because she had been adopted. Neurological examination showed grade

4 strength for cervical flexion and shoulder abduction, asymmetric grade 4 (worse on the right) for flexion of only the index fingers and hip flexion (also worse on the right); grade 1 deep tendon reflexes, with abolished achilles; paresis of orbicularis oculi and mouth, characterizing facial diparesis. Additional tests showed elevated creatine phosphokinase, myoglobin and lactate dehydrogenase. Electroneuromyography showed fibrillations and positive sharp waves in all muscles evaluated, an increased number of myopathic potentials and the presence of some repetitive complex discharges. Previous esophagography revealed digitiform narrowing in the upper third of the esophagus secondary to hypertrophy of the cricopharyngeal muscle. Considering the hypothesis of atypical presentation of inclusion body myositis, a left medial thigh muscle biopsy was requested and revealed slight endomysial fibrosis in sections stained with Gomori's trichrome, presence of degenerative fibers with influx of macrophages, fibers undergoing regeneration and endomysial lymphocyte influx, all findings compatible with the diagnosis of an inflammatory myopathy, often described in polymyositis and inclusion body myositis. **Discussion:** Inclusion body myositis (IBM) is the most frequent myopathy affecting adults older than 45 years, being more common in men. The typical clinical presentation is asymmetric and slowly progressive muscle weakness, characteristically affecting knee extensors and deep finger flexors. However, atypical forms with varying patterns of muscle weakness are recognized, including axial weakness, foot drop, isolated dysphagia, facial diplegia and proximal limb weakness. The patient presented in our case was a woman and exhibited an atypical pattern of muscle weakness. In 2024 the diagnostic criteria for IBM were revised, in order to make the diagnosis simpler and inclusive. According to the new criteria, and considering our patient, the diagnosis of IBM would be confirmed if, in addition to the atypical presentation and the mandatory biopsy finding, there were additional supporting criteria (positive anti-cN1a antibody, typical imaging pattern), both of which are limited resources in our setting. **Final comments:** IBM should always be considered as an etiology in adult-onset myopathies. Despite the premise of simplification and inclusiveness of the new criteria, there are still difficulties in confirming the diagnosis of atypical forms in our country.

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Challenges in differential diagnosis of demyelinating diseases: a clinical and laboratory review and comparison of four cases

Victor Hugo de Souza Silva Gomes[1], Helen Luiza Silvestrini[2], Leticia Lage Silva[2], Albérico Rocha Lima Neto[1], Beatriz Fulador Maciel[1], Rafaella Resplande Xavier[1], Heloise Helena Siqueira Borges[1], Rhaymysom Jasmy Gomes Abreu[1]

[1]Hospital Geral de Cuiabá, Cuiabá MT, Brazil. [2]Universidade de Cuiabá, Cuiabá MT, Brazil.

Correspondence: Helen Luiza Silvestrini; Email: helensilvest@gmail.com.

Background: Demyelinating diseases are characterized by inflammation and immune-mediated attacks on components of the central nervous system (CNS), with an autoimmune, progressive, and chronic nature. The main representatives are multiple sclerosis (MS) and neuromyelitis optica spectrum disorder (NMOSD). MS is characterized by chronic demyelination with loss of oligodendrocytes and axonal damage, leading to sensory-motor deficits with variable recovery during relapse and remission phases. In contrast, NMOSD is based on the presence of AQP4-IgG autoantibodies targeting astrocytes, manifesting with severe acute episodes of optic neuritis and longitudinally extensive transverse myelitis (LETM), as well as area postrema syndrome. Due to the broad spectrum of symptoms that can mimic other conditions, differential diagnoses such as Susac Syndrome and CLIPPERS must be considered. Additionally, there is an association with systemic autoimmune comorbidities such as Systemic Lupus Erythematosus (SLE) and Sjogren's syndrome (SjS). **Objective:** To analyze the profile of patients who manifested symptoms suggestive of demyelinating diseases like MS or NMOSD and were later diagnosed with other conditions that have a similar course. **Methods:** This is an observational and descriptive study of 4 patients followed in the Neuroimmunology Outpatient Clinic. Data were obtained through medical record review and literature review, information collection, and delivery of the Informed Consent Form (ICF). **Results:** All patients were female, with an average age of 26 years, and presented clinical and neuroimaging findings compatible with the diagnostic hypothesis (DH) of CNS demyelinating diseases. All underwent pulse therapy with variable responses. Case 1 presented LETM; however, after positive ANA 1:640, Anti-SSb, and Anti-SSa antibodies, a diagnosis of Sjogren's was established. Case 2 began with headaches, hemiparesis, and decreased auditory and visual acuity, with MRI indicating callosal and pericallosal lesions, compatible with Susac Syndrome. Case 3 began with headaches, vertigo, diplopia, strabismus, and MRI showing right nucleocapsular, midbrain, pons, and cerebellum alterations, with complete remission after corticosteroids, suggestive of CLIPPERS. Case 4, already diagnosed with SLE, presented with right optic neuritis and LETM. After pulse therapy, significant recovery was noted, with AQP4-IgG negative, suggestive of neurological involvement by SLE. **Conclusion:** It is possible to verify the importance of anamnesis, neurological examination, and complementary tests, given the difficulty in establishing a diagnosis when dealing with autoimmune diseases. The difficulty patients face in accessing the required tests, which must be regular and may present false-negative results, is also highlighted. Despite the challenges, proper management of these conditions directly influences the quality of life and correct long-term treatment direction.

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Challenges in managing acute hepatic porphyrias with comorbid conditions: a case series

Daniel Delgado Seneor[1], Paulo de Lima Serrano[1], João Paulo Barile[1], Kaliny Oliveira Peixoto[1], Patrícia Marques Mendes[1], Renan Brandão Rambaldi Cavalheiro[1], Igor Braga Farias[1], Wladimir Bocca V. R. Pinto[1], Acary Souza Bulle Oliveira[1], Paulo Victor Sgobbi de Souza[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Daniel Delgado Seneor; Email: danielseneor30@gmail.com.

Acute hepatic porphyrias (AHP) are rare metabolic disorders characterized by deficiencies in enzymes of the heme biosynthesis pathway, leading to the accumulation of toxic precursors. In spite of showing some chronic manifestations, these disorders typically present with acute neuropathic symptoms, exacerbated by triggers such as emotional stress, fasting, hormonal fluctuations, recreational drugs and unsafe medications. **Case presentation:** Patient 1: A 65-year-old female with hereditary coproporphyria, diagnosed at age 34. She has a history of chronic neuropathic pain and multiple comorbidities, including Tropical Spastic Paraparesis (HTLV-I) an nephrolithiasis. The patient uses tizanidine (safety profile not yet classified) and nitrofurantoin (porphyrinogenic), which may trigger porphyria attacks. Patient 2: A 58-year-old male with variegate porphyria, diagnosed at age 30. He has a history of Charcot-Marie-Tooth disease type 2P (CMT2P - LRSAM1), chronic alcohol abuse and other comorbidities. He experienced an acute porphyria exacerbation triggered by alcohol. Patient 3: A 55-year-old female with variegate porphyria, diagnosed at age 33, who is also HIV-positive. She uses lamivudine, zidovudine (both probably not porphyrinogenic, but may trigger attacks due to inappetence and nausea), and atazanavir (probably porphyrinogenic) as antiretroviral therapy. **Discussion:** Each patient's case illustrates the complexity of managing patients with AHP and superimposed conditions. Patient 1's case highlights the need to consider safety profile when choosing antimicrobial and antispastic medications. Patient 2's case shows that alcohol is an important trigger for porphyria attacks and that a concurrent unrelated cause of neuropathy may interfere with diagnosis and management of AHP. Patient 3's case demonstrates the complexities of managing such condition in HIV-positive patients, since antiretroviral medications must be chosen cautiously. Managing patients with acute hepatic porphyrias is particularly intricate when they have addiction disorders or concurrent conditions requiring pharmacological treatments, as these can exacerbate the disease. Additionally, in patients with comorbidities that cause peripheral nerve damage, distinguishing whether worsening neuropathy stems from porphyria or the coexisting condition poses a significant challenge, complicating their care. **Final comments:** This case series illustrates the multifaceted challenges presented by patients with AHP, including addiction disorders, superimposed diseases, and the use of potentially unsafe medications. Individualized patient management and thoughtful medication selection are of utmost importance for minimizing the risk of attacks, while maintaining an acceptable treatment efficacy. Continued education and awareness can improve the quality of care for these complex patients.

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Challenges in the immunocompromised patient: a case of ureaplasma meningoencephalitis

Jackeline Neves Pereira[1], Clara Moreira Gonzaga[1], Luccas Almeida de Andrade[1], Guilherme Antonio Silva Ribeiro[1], Raphael Gusmao Barreto[1], João Vitor Gerdulli Tamanini[1], Gabriel Augusto Wandekoken[1], Fernanda Ferreira de Abreu[1], José Luiz Pedroso[1], Fabiano Moulin de Moraes[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Jackeline Neves Pereira; Email: jac.nevesp@gmail.com.

Case presentation: A 61-year-old male has been regularly treating HIV since 2009, with an undetectable viral load since 2010, and a T CD4+ cell count of 267/mL. He presented an acute headache, vertigo, diplopia, and ataxia, reaching the Hospital two days after the onset of the symptoms, admitted with a blood pressure of 200x120mmHg. A brain computerized tomography (CT) scan revealed a hemorrhagic stroke in the right cerebellar hemisphere. On the following day, due to obstructive hydrocephalus, brain surgery was performed for blood drainage and external ventricular shunt. The patient had a poor recovery, needed invasive ventilatory support, had fever, and a new CT revealed an ischemic area in the left occipital lobe. Cerebrospinal fluid (CSF) had a high Neutrophils and protein count, and low glucose; cultures, GRAM staining, and molecular tuberculosis testing were all negative. A brain magnetic resonance (MR) demonstrated several ischemic lesions, with true diffusion restriction, particularly along vascular border zones. The patient was empirically treated with Vancomycin, Meropenem, and later Polimixin B, without improvement. Metagenomic analysis of the CSF was positive for Ureaplasma urealyticum (UU), confirmed through Polymerase Chain Reaction (PCR). Before the final results, the patient had worsened systemic and neurological parameters and was declared dead. **Discussion:** UU is a bacteria part of the genito-urinary tract microbiota, more commonly associated with newborn central nervous system infections, with scarce literature in adults. In this case report, the patient first

had a hemorrhagic stroke, with subsequent ischemic lesions. It was extremely challenging to reason whether the hemorrhage was the first manifestation of an infectious vasculitis or a separated event from an infectious vasculitis secondary to a surgical wound complication. The patient did not respond to empirical antibiotic therapy, and the pathogen was only identified through molecular testing. All the other infectious tests, including CSF, blood, and cerebellar tissue culture, were negative. Meningitis due to UU was reported in a patient with adult-onset immune deficiency known as Good's syndrome, the diagnosis was made by Next-Generation Sequencing. In another case, a brain abscess related to UU mastoiditis in a hypogammaglobulinemia patient following Rituximab use was also reported, routine culture was negative, and the diagnosis was through 16S rRNA gene PCR. The same method was used to diagnose the first reported case of UU meningitis in an adult, a complication after a kidney transplant, published in 2008. **Final comments:** All taken together, Ureaplasma urealyticum seems to be of particular importance as a cause of encephalomeningitis in immunocompromised adults who do not respond to empirical antibiotics. It is noticeable that molecular methods, which are not ubiquitously available, play a central role in the correct diagnosis, meaning that this condition might be more frequent.

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Challenging visual acuity measurement in a child with MOGAD syndrome: a case report

Jonas Leite de Souza Filho[1], Jonatas Paulino da Cunha Monteiro Ribeiro[1], Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Luma Rodrigues da Silva[1], José Henrique de Melo Feitosa[1], Pedro Henrique Ximenes Ramalho Barros[1], Sabrina Ruthiele Santos de Carvalho[1], Tiago José Do Nascimento Teixeira Leite[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Jonas Leite de Souza Filho; Email: jonaslf25@gmail.com.

Case presentation: Female, 5 years old, admitted due to frequent falls associated with visual impairment and trouble identifying colors. The patient complained of worsening her visual acuity and bilateral blurriness. This symptom begun 14 months ago. Previous comorbidities and continuous medication use were denied. After an ophthalmological evaluation, bilateral vision loss was diagnosed. On physical examination, she presented isochoric and photoreactive pupils. Because the patient did not cooperate in measuring visual acuity, a subjective assessment was conducted. She was tasked with identifying toys, games, pictures on a monitor, and colors. The patient struggled to identify the items, suggesting a reduction in peripheral vision bilaterally. Muscle strength was preserved (grade V), without pathological plantar reflex. No alterations in balance, sensitivity, and coordination were diagnosed. Brain MRI disclosed gadolinium enhancement, without expansive characteristics, affecting the intracanalicular segment of the optic nerves and the intraorbital segment of the right optic nerve, suggesting bilateral optic neuritis (ON). Retinography showed bilateral papilledema. Thus, hospitalization was recommended to perform pulse therapy with Methylprednisolone (20 mg/kg/day for 5 days). Since then, she has been scheduled for Albendazole and pulse therapy, and tests were requested for anti-MOG (positive) and anti-aquaporin IV (non-reactive). After 10 days of hospital treatment, the patients visual acuity was reassessed, showing an improvement in her condition, except for color identification ability. She is followed up by neurology and undergoing rehabilitation for better color visualization. She is on Azathioprine and Prednisolone, as well as Paracetamol for intense headaches. She had unremarkable signs for autoimmune diseases. **Discussion:** We presented a case of a child with MOGAD, a recently recognized inflammatory demyelinating disorder of the central nervous system. The most common findings of MOGAD attacks are severe impairment on visual acuity with a good recovery after an early management. In this case, the patient presented with isolated bilateral ON, with almost complete improvement of visual impairments, despite challenging measurement of visual acuity. We used alternative visual skills to measure the visual acuity in a child with MOGAD optic neuritis. **Final comments:** The diagnosis of MOGAD is necessary for appropriate management to be established. However, the difficulty of clinically assessing visual acuity can hinder this early diagnosis.

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Changing the natural course of hereditary transthyretin amyloidosis: the importance of early diagnosis

Ligia Rocha Andrade[1], Ana Siquara[2], Clarissa Neves Spitz[1,2], Eduardo Davidovich[1], Marcelo Imbroinise Bittencourt[1], Salim Lázaro Balassiano[1], Larissa Bittencourt de Carvalho[1,2], Robson Teixeira Vital[1], Rafael Oliveira Vidon[1], Marcia Rodrigues Jardim[1,2]

[1]Universidade do Estado do Rio de Janeiro, Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.
[2]Fundação Oswaldo Cruz, Rio de Janeiro RJ, Brazil.

Correspondence: Ligia Rocha Andrade; Email: ligiaandradee@gmail.com.

Background: Hereditary transthyretin amyloidosis (ATTRv) has a heterogeneous clinical presentation, and the symptoms are mainly neuropathic, including autonomic, sensory, and motor impairment. In addition, it may be associated with gastrointestinal, cardiac, renal, and ocular involvement. Diagnosis in the early stages is essential to allow appropriate treatment and to prevent or slow the progression of the disease. **Objective:** Early detection of the disease in asymptomatic ATTRv carriers. **Methods:** A cross-sectional study of asymptomatic individuals with ATTRv was conducted at the neuromuscular diseases outpatient clinic of Hospital Pedro Ernesto in Rio de Janeiro from April 2019 to November 2023. **Results:** 4 out of 16 individuals (25%) were diagnosed with the disease through salivary gland biopsy, after presenting alterations in at least two complementary tests related to the evaluation of small-fiber neuropathy, even if those individuals remained completely asymptomatic. **Conclusion:** Small-fiber neuropathy was the earliest sign of disease onset, occurring in all four individuals diagnosed and the tests that proved to be more sensitive were sympathetic skin response (SSR) and quantative sensory testing (QST)

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Charcot-Marie-Tooth disease in a patient with long-chain 3-hydroxyacyl-coa dehydrogenase (LCHAD) deficiency

Maria Rodrigues Lima de Souza Pessoa[1], Ariele Amaral Alves[1], Marilaine Menezes de Matos Ferreira[1], Marcela Camara Machado Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Maria Rodrigues Lima de Souza Pessoa; Email: mariapessoa20.1@bahiana.edu.br.

Case presentation: A 9-year-old male patient, with brown skin color, natural childbirth at term without further pregnancy information, started follow-up at the center on June 8, 2022, with a history of motor delay. This was recognized due to a lack of gait, by his parents at 12 months old. The parents were unable to recognise other motor milestones. He developed distal atrophy of the upper and lower limbs, muscle weakness, retraction of the fingers and toes, and bilateral cavus feets, as well as eyelid ptosis, which worsened in multiple episodes of rhabdomyolysis secondary to infections, requiring hospitalization. He had previously been observed at another center where a clinical diagnosis of Charcot-Marie-Tooth disease was made at 22 months of age. His father was observed to have cavus foot and calf atrophy, as well as electroneuromyography with evidence of axial sensory-motor polyneuropathy. Physical examination revealed distal atrophy and retraction of the calcaneus and toes, scoliosis, level 4 limbs strength, global level 1 osteotendinous reflexes, no gait but able to stand. Sensitivity was preserved. At that time, he was using 5 milligrams of baclofen twice a day and was not followed by a multidisciplinary team. The main suspect at the time was long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency and a genetic panel was conducted, confirming the diagnosis. He was followed at the center and began nutritional support and physical therapy. The neurological status remained unchanged, with no ophthalmic or cardiac changes to date, but with new episodes of rhabdomyolysis associated with the occurrence of infections. **Discussion:** Given the clinical presentation, the progressive, muscle weakness with atrophy and limbs dysmorphism associated with a compatible family history stands out. These factors favor the diagnosis of Charcot-Marie-Tooth disease, considering the clinical features and the frequency of the hereditary nature of the neuropathy. In view of rhabdomyolysis secondary to infection and associated muscular neuropathy, especially in the lower limbs, there is support for a LCHAD case, considering the onset in early childhood. **Final comments:** The patient presents two rare diseases in which the overlapping of the pathologies may have significantly worsened the clinical manifestations, especially those motor in nature.

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CHD8 and autism spectrum disorder: genomic advances and clinical implications for neurological practice

Tereza Raquel Xavier Viana[1], José Eduardo Martinelli[1]

[1]Faculdade de Medicina de Jundiaí, Jundiaí SP, Brazil.

Correspondence: Tereza Raquel Xavier Viana; Email: espacotr@hotmail.com.

Background: Autism Spectrum Disorder (ASD) is a complex neurodevelopmental condition characterized by deficits in social communication and repetitive behaviors. Recent advances in genomics have revealed the CHD8 gene as an important factor in the etiology of ASD, with loss-of-function mutations often associated with variable clinical manifestations of the condition. Understanding the specific role of CHD8 in ASD is crucial to developing more accurate diagnostic strategies and targeted therapies. **Objective:** To analyze recent studies on the CHD8 gene and its implications in ASD in order to provide a comprehensive overview of the latest genomic findings and discuss their clinical implications in neurological practice. **Methods:** This is an exploratory and descriptive systematic review conducted according to the PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) and PICO (Population, Intervention, Comparison, Outcome) guidelines. Searches were carried out in the PubMed, SciELO, Cochrane Library and BVS databases, using the descriptors: "CHD8" AND "Genomics" AND "Autism Spectrum Disorder". We included studies published in the last 5 years (2019 - 2024), in English and Portuguese, which investigated the associations between the CHD8 gene and ASD, exploring genomic advances and their clinical implications in neurological practice. Experimental

studies, controlled clinical trials and systematic reviews were considered for inclusion in the analysis. **Results:** During the initial search, 26 articles were identified. After reviewing the titles, abstracts and conclusions, 21 were excluded because they did not meet the inclusion criteria. Five studies were selected for full reading and inclusion in this research. These studies offer significant advances in the understanding of CHD8 in ASD, highlighting its robust association with the pathogenesis of this condition and its influence on specific clinical phenotypes. The research reviewed reveals that mutations in CHD8 are frequently found in individuals with ASD, contributing significantly to the phenotypic variability observed in the condition. In addition, they elucidate the molecular mechanisms by which CHD8 affects synaptic function, gene expression and chromatin compaction, providing important insights for molecular diagnosis and the potential development of personalized gene-based therapies. **Conclusion:** This study underlines the importance of the CHD8 gene in the etiology of ASD and highlights recent genomic advances that are shaping the understanding of the condition. Integrating these findings into neurological practice may offer new opportunities for early diagnosis, more effective clinical management and the development of targeted therapeutic interventions. Future studies should continue to explore the functional and clinical implications of CHD8, aiming to improve outcomes for individuals affected by ASD related to mutations in this specific gene.

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Chediak-Higashi syndrome presenting as pure juvenile Parkinsonism

Matheus Gomes Ferreira[1], Renata Cristine Alves[1], Luis Eduardo Borges de Macedo Zubko[1], Christelle Thesson[2], Thomas Courtin[2], Alexis Brice[2], Francisco Manoel Branco Germiniani[1], Helio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[2]Sorbonne Université, Paris Brain Institute, Institut du Cerveau et de la Moelle épinière, Paris, France.

Correspondence: Renata Cristine Alves; Email: renata_cristinealves@hotmail.com.

Case presentation: Here we present a Brazilian patient with juvenile parkinsonism as the sole manifestation of a new LYST mutation. His parents were consanguineous and he had a sister with clinical diagnosis of Chediak-Higashi syndrome based on bleeding diathesis, immunodeficiency and inclusion bodies in peripheral blood leukocytes. He began with asymmetrical parkinsonism when he was 15 years old characterized by bradykinesia, rigidity and resting tremor. By that time, he began treatment with levodopa with a good clinical response. During follow-up the patient developed motor fluctuations, an important impulse control disorder and cognitive decline. His current clinical evaluation had an UPDRS-III score of 64 and a 11/30 on the MoCA test. He was submitted to whole-exome sequencing, with the finding of a homozygous missense mutation p.R3107H in the LYST gene. **Discussion:** Chédiak-Higashi syndrome is a lysosomal storage disorder caused by mutations in the LYST gene with an autosomal recessive inheritance. It is characterized by the disruption of transport and fusion of lysosomes, leading to a myriad of clinical signs and symptoms. Clinically patients manifest variable degrees of partial oculocutaneous albinism, bleeding diathesis and immunodeficiency. Depending on the age of symptoms onset, cases manifesting during childhood have mainly hematological symptoms, while the ones who present in adolescence/early-adulthood usually display neurological symptoms. The latter may include parkinsonism, cerebellar ataxia, dystonia, dementia, neuropathy and epilepsy, with a prolonged clinical course and slow deterioration. **Final comments:** As far as we know, this is the first reported case of homozygous missense mutation p.R3107H in the LYST gene, with an atypical presentation characterized by early-onset parkinsonism without associated hematological disease.

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Chorioretinitis, bilateral optic neuritis and pachymeningitis as manifestation paraneoplastic probable of esophageal squamous cell carcinoma

Renan Macionil Cotrim[1], Itamar Meireles Andrade Santos[1], João Márcio Borgue de Oliveira[1], Bianca Latance da Cruz[1], Pedro Enrico Cyprestes Sant Anna[1], Gabriela Rossini Dantas[1], Eduardo Abrão Spinola Rezk[1], Helio Aquaroni Farao Gomes[1], Ana Beatriz Marangoni Baston[1], Laura Cardia Gomes Lopes[1]

[1]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Renan Macionil Cotrim; Email: renan.cotrim@unesp.br.

Case presentation: Man, 61 years old, smoker, started in February 2024 progressive visual impairment of the left eye and holocranial headache in "pressure". Evolutionarily, he presented a left clavicle fracture. On physical examination, the patient didn't have neurological deficits, except for the left eye with visual acuity 0.3 and fundoscopy with optical disc edema. Bone scintigraphy detected diffuse radiopharmaceutical uptake, including the skullcap, left clavicle, and costal arches, and the neuroimaging study showed hypersignal on T2/FLAIR sequences, and contrast uptake in the retina, optic nerve on the left, and meningeal enhancement at the base of the brainstem, and anterior cranial fossa. The visual evoked potential test demonstrated bilateral optic neuropathy, worse on the left, and rachicentesis, pleocytosis with lymphomonocytic predominance and hyperproteinorrachia, latex for bacterial or fungal agents, and culture negative. Neoplastic cells on cytopathological examination absent, serology for HTLV, HIV, CMV, HBV , HCV and syphilis negative. Pulse therapy was performed with methylprednisolone 1 g/day for 5 days, with improvement in the condition. Esophagogastroduodenoscopy with histopathology positive for esophagogastric transitional squamous cell carcinoma, palliative chemotherapy with carboplatin and paclitaxel is initiated. Later, the worsening conditions, associated with aggressiveness and convulsive crises of focal onset, the new cytopathological examination of cerebrospinal fluid detects atypical cells, suggesting meningeal carcinomatosis. Admitted to a palliative care ward, he developed a headache that was refractory to optimized analgesia and worsened his behavioral condition, resulting in death. **Discussion:** Paraneoplastic neurological syndromes represent 1 in every 300 cancer patients, and result from the production of antibodies with target organ dysfunction. The diagnosis is based on the understanding of clinical, oncological, and immunological heterogeneity, with the need to exclude other causes, such as meningeal carcinomatosis or brain metastasis, but its treatment is still uncertain. The diagnostic criteria are supported by a panel called PNS-Care that includes high-risk phenotypes, such as limbic encephalitis, and intermediate-risk phenotypes, when the onset is rapidly progressive, with inflammatory findings on cerebrospinal fluid or neuroimaging. Although uncommon in tumors of the gastrointestinal tract, a literature review carried out between 1970 and 2021 about esophageal cancer described phenotypes for neurological paraneoplastic syndromes. **Final comments:** Paraneoplastic neurological manifestations are heterogeneous, we report a case of choreoretinitis, bilateral optic neuritis and pachymeningitis as the initial manifestation of esophageal squamous cell carcinoma, signaling it for further studies, and alerting clinical reasoning in the face of similar conditions that occur in others centers.

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Chronic Chagas heart disease presented as comma-shaped stroke

Ana Carolina Leite Ribeiro[1], Brenno Tavares de Vasconcelos Brandão[1], Bárbara Pina Aiello[1], Giuliana Gonçalves Moreira Possidente[1], Marcos Martins da Silva[1], Carolina Rouanet de Albuquerque[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Ana Carolina Leite Ribeiro; Email: anacarolinaneuro@gmail.com.

A previously healthy 52-year-old woman from Minas Gerais was admitted with subacute left-sided motor and sensory hemiparesis and dysarthria. A head computed tomography scan revealed a large infarct in the right lenticulostriate artery territory. Intracranial and cervical vessel studies showed occlusion of the left MCA segment M1 with preserved cortical vascularization. Brain magnetic resonance imaging revealed similar findings. An electrocardiogram identified atrial fibrillation and right bundle branch block. A 24-hour Holter monitor detected nonsustained ventricular tachycardia. Transthoracic echocardiogram revealed dilated cardiomyopathy with diffuse hypokinesis and an ejection fraction of 36%. The thrombophilia screen was negative. Given the dilated cardiomyopathy in the absence of cardiovascular risk factors or a history of alcoholism, Chagas disease was suspected and confirmed by two serological tests. Large deep striatocapsular infarctions account for 1-6% of ischemic strokes. The basal ganglia and parts of the caudate nucleus, internal capsule, putamen, and globus pallidus are supplied by the lenticulostriate arteries, the artery of Heubner, and the anterior choroidal artery. Large striatocapsular infarctions, previously termed giant or super lacunae, occur due to MCA trunk occlusion affecting all lenticulostriate arteries, typically caused by cardioembolism or atheroembolism. The cortex is often spared due to collateral artery formation, resulting in comma-shaped infarctions. Clinical manifestations include motor and/or sensory hemiparesis and may include dystonia, involuntary movements, aphasia, and hemineglect. Bladin and Verkovic described two mechanisms for preserving cortical perfusion: thrombotic occlusion at the carotid T-junction with cortical blood supply via anastomoses, and embolism at the carotid T-junction obstructing lateral striate arteries while allowing some cortical perfusion. The patient presented with this unusual form of stroke, likely due to chronic Chagas cardiomyopathy, previously asymptomatic. Full anticoagulation and heart rate control were initiated as secondary prophylaxis. Electrical cardioversion was considered after resolving contraindications. The possibility of etiological treatment for Chagas disease was discussed, which is reserved for the chronic indeterminate form in adults under 50 years old. Identifying stroke etiology and understanding the involved pathophysiology, especially in rare presentations, is crucial for establishing secondary prophylaxis and treating the underlying disease. Advanced vessel imaging studies were particularly useful for understanding the pathophysiological and anatomical mechanisms.

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Chronic dengue virus encephalitis diagnosed by metagenomic analysis: a case study

Gabriela Abrahão Allioni[1], Gabriel Samir Martins de Souza[1], Julia Haddad Labello[1], Gabriel Saboia de Araújo Torres[1], Yuri Reis Casal[1], Amaro Nunes Duarte Neto[1], Luiz Henrique Martins Castro[1], Gustavo Bruniera Peres Fernandes[2], João Renato Rebello Pinho[2], Bruno Fukelmann Guedes[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Gabriela Abrahão Allioni; Email: gabriela.allioni@hc.fm.usp.br.

Background: The dengue virus (denV) can lead to acute encephalitis during the initial infection. Chronic encephalitis with progressive dementia is rarely documented. **Methods:** We present a novel case of chronic encephalitis causing rapidly progressive dementia, in which a probable denV brain infection was identified through metagenomic next-generation sequencing of brain tissue. Brain pathology and molecular diagnosis are detailed. **Results:** A 20-year-old male from São Paulo began experiencing symptoms of rapidly progressive dementia, including speech apraxia and apathy, starting in June 2022. Initial cognitive testing in January 2023 revealed severe impairment across multiple domains, with a score of 18 points on the Mini-Mental State Examination. A brain MRI performed six months after symptom onset showed white matter abnormalities and brain atrophy. Cerebrospinal fluid (CSF) analysis indicated elevated white blood cells, increased proteins, and the presence of oligoclonal bands. An extensive diagnostic work-up ruled out common infectious and autoimmune causes of encephalitis. Over a six-month period, the patient's condition continued to deteriorate, resulting in progressive impairment of executive functions and language, accompanied by the onset of tremors and myoclonus. In August 2023, the patient was readmitted for further diagnostic investigation, revealing reactive denV IgG and non-reactive IgM in an arboviruses immunology panel, prompting additional investigation. Due to the lack of an etiological diagnosis and the severity of the case, a brain biopsy was performed. Histopathological examination of the brain biopsy suggested a chronic infectious etiology unrelated to bacteria, supported by the frequent presence of CD8+ T lymphocytes, plasma cells, and glioneuronal alterations. Next-generation sequencing and phylogenetic analysis of brain tissue identified DENV-1 genotype V, with the recovered sequence clustering within a clade of Brazilian sequences from 2000 to 2022. # **Conclusion:** This case highlights the importance of unbiased techniques like metagenomics in diagnosing encephalitis of unknown etiology, providing a potential etiological explanation for the patient's chronic encephalitis. While dengue virus is occasionally described as a cause of acute encephalitis/encephalopathy, this case broadens the clinical spectrum of the infection, demonstrating that it may also cause chronic encephalitis.

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Chronic inglammatory demyelinating polyneuropathy (CIDP): a case series from a tertiary center

Mariane Panka[1], Paulo José Lorenzoni[1], Renata Dalprá Ducci[1], Otto Hernandes Fustes[1], Paula Raquel Rodrigues[1], Rosana Herminia Scola[1], Cláudia Suemi Kamoi Kay[1]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Mariane Panka; Email: mari_panka@hotmail.com.

Background: Chronic inflammatory demyelinating polyneuropathy (CIDP) represents a spectrum of treatable chronic immune-mediated disorders affecting peripheral nerves and nerve roots. CIDP typically presents with symmetric motor deficits, including proximal and distal weakness and areflexia. Sensory symptoms such as paresthesia and hypoesthesia are often more pronounced than pain and temperature alterations. The disease course can be progressive or marked by remitting-relapsing patterns. Treatment generally involves immunosuppressants or immunomodulators such as glucocorticoids, intravenous immunoglobulin, or plasma exchange. Diagnosis relies on electrodiagnostic evidence of peripheral demyelination, supplemented by laboratory tests, cerebrospinal fluid analysis, neuroimaging, and occasionally nerve biopsy. Distinguishing CIDP from other chronic demyelinating neuropathies is critical due to differing treatment approaches that significantly impact patient outcomes. **Objective:** To assess clinical features, electroneuromyographic findings, cerebrospinal fluid analysis, and imaging results in patients undergoing follow-up at our institution. **Methods:** A retrospective review of medical records, electroneuromyography, laboratory tests, and imaging studies was conducted. **Results:** Seventeen patients meeting CIDP diagnostic criteria were identified from our neuromuscular diseases out patient clinic. Coexisting conditions included well-controlled diabetes mellitus in two patients, Waldenströmma croglobulinemia in one, neurofibromatosis in another, and exacerbation post-influenza vaccination in one case. Cerebrospinal fluid analysis indicated elevated protein levels in all but one patient, averaging 326.37mg/dL (median: 345mg/dL, range: 36 – 964.3mg/dL). Electroneuromyography demonstrated demyelinating and axonal polyneuropathy in 11 patients, with demyelinating patterns in five and axonal patterns in one. Magnetic resonance imaging revealed root thickening in three out of tem patients tested. Genetic testing for hereditary neuropathies or the TTR gene panel was negative in all seven patients screened. Three patients showed poor response to treatment, including one with CIDP associated with Waldenström macroglobulinemia. **Conclusion:** This case series confirms the presence of CIDP in all patients based on diagnostic criteria and consistent electroneuromyographic findings. Most patients exhibited elevated cerebrospinal fluid protein levels, and imaging studies were crucial in diagnosing and managing uncertain cases. Treatment out comes were generally favorable, underscoring the efficacy of current therapeutic modalities.

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Chronic inflammatory demyelinating polyradiculoneuropathy related to neurofascin-155 IgG4 with response to rituximab: case report

Luma Elem Soares Batista[1], Patrícia Pereira Nunes[1], Gabriela de Castro Cavalcante Mendonça[1], Amanda de Araújo Cravo[1], Carla Fernanda Veiga Pimentel[1], Marianna Kelly de Araújo Souza Barbosa[1], Gabriel Barroso Cunha[2], Mikaella de Lima Pereira Moraes[1], Paula Raphaella da Silva Melo[1]

[1]Centro Universitário de Maceió, Maceió AL, Brazil.
[2]Universidade Estadual de Ciências da Saúde de Alagoas, Maceió AL, Brazil.

Correspondence: Gabriel Barroso Cunha; Email: gabrielbarrosocunha@gmail.com.

Case presentation: Male, 35 years old, with paresthesia in the hands and feet associated with difficulty walking and progressive neuropathic pain in the lower limbs, for approximately 6 months. He underwent intravenous immunoglobulin (IgEv), showing partial improvement in complaints. Physical examination revealed facial diparesis, vibratory hypoesthesia in the malleoli (left better than right). Grade 4 strength in finger flexion and thumb opposition, in addition to plantar flexion and dorsiflexion (4+ in L and 4 in R). Positive Romberg, heel gait with wide base. Electroneuromyography indicating chronic, slightly asymmetric acquired demyelinating peripheral sensory-motor polyradiculoneuropathy, magnetic resonance imaging of the lumbar spine with contrast compatible with inflammatory radiculitis, cerebrospinal fluid with normal cellularity and hyperproteinorrhaquia with a positive Anti-NF-155 test (1/8100 - predominant IgG4 subclass). He underwent two more cycles of IgEv with improvement in pain and sensitivity (improving MIE hypoesthesia and facial weakness), but maintaining ataxia. We chose to undergo three cycles of intravenous corticosteroid therapy and oral immunosuppressant (azathioprine), with partial improvement of all symptoms, with the exception of ataxia. One month later, the patient returned, maintaining previous changes and then underwent 8 plasmapheresis sessions, also with a slight improvement in symptoms. Rituximab was started, on a schedule of 4 weekly infusions of 500 mg, progressing with significant improvement in neurological examination, as well as in functionality. Patient continues to be monitored for discussion of medication dose progression. **Discussion:** Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an immune-mediated disease that affects roots and peripheral nerves, with a decrease or absence in motor and sensory amplitudes. Around 10% of patients have autoantibodies (mainly IgG4) against nodal and paranodal proteins (NF-155). Because this group of patients with specific antibodies share clinical characteristics and response to treatment different from those of typical CIDP, this disease is included in the recent diagnostic category of autoimmune nodopathy, therefore being a distinct classification from CIDP. In these cases, there is greater distal than proximal muscle weakness, positive sensory symptoms and gait ataxia. Compared to other node-paranodopathies, they have higher recurrence, morbidity and younger age of onset. It is refractory to IgEv, with a favorable response to second-line (Rituximab) and third-line immunosuppressants, and a prolonged time until clinical improvement, up to two years. **Final comments:** Autoimmune node diseases have a broad clinical spectrum, which makes establishing an effective treatment challenging. Thus, serology for specific autoantibodies makes it possible to use treatment based on the most recent evidence, providing an individualized and effective therapeutic plan.

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Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS): case presentation

Ana Luísa Castelo Branco Gomes[1], Davi Telécio Firmino[2], Gilmar Leite Pessoa Filho[1], Luciano Gonçalves Do Nascimento Júnior[1], Felipe Nunes de Araújo[1], Rafael de Souza Andrade[1], Luíza Alves Monteiro Torreão Villarim[1], Bianca Etelvina Santos de Oliveira[3], Juliana Magalhães Leite[1], Alex Tiburtino Meira[1]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[2]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[3]Conselho Regional de Medicina da Paraíba, João Pessoa PB, Brazil.

Correspondence: Ana Luísa Castelo Branco Gomes; Email: analuisa_gomes@hotmail.com.

Case presentation: 52-year-old female patient, previously healthy, presented to the ataxia outpatient clinic with a history of progressive imbalance for 8 years. She developed paresthesia in her extremities in the past 4 months, and dysphagia for liquids and solids in the last 2 months. The physical examination revealed dysarthria, Parinaud's syndrome, Collier's sign, global cerebellar ataxia, hyperreflexia in the lower limbs and bradykinesia at alternating movements. Magnetic Resonance Imaging (MRI) revealed a punctate and curvilinear T2/FLAIR hyperintensity on the central portion of the pons and extending to the midbrain with gadolinium enhancement. Cerebrospinal fluid analysis, electroneuromyography and electroencephalography showed no abnormalities. Due to the clinical and laboratory findings, the diagnosis of Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS) was suggested. The patient was treated with methylprednisolone 1g/day for 5 days, progressing with moderate improvement of

the imbalance, dysarthria and dysphagia. **Discussion:** CLIPPERS is a rare chronic inflammatory disorder of the central nervous system (CNS). The disease can occur in all age groups, and both men and women can be affected. Its etiology and pathogenesis remain unknown and most patients have no specific triggering factors. Diagnosis is based on clinical, radiological and pathological criteria. Clinical findings include subacute pontocerebellar dysfunction such as ataxia, diplopia and facial paresthesia, as well as clinical responsiveness to corticosteroid therapy. MRI of the pons shows punctate images with a "salt and pepper" appearance with contrast enhancement. Biopsy, when performed, shows infiltration of the perivascular spaces by T cells. The cornerstone of the treatment is a corticosteroids pulsotherapy, which typically leads to significant improvement in symptoms. No study has shown that CLIPPERS can be cured, so, long-term steroid maintenance therapy is necessary to prevent recurrence of the disease. **Final comments:** Clinical, radiological and pathological findings of CLIPPERS are non-specific and occur in several corticosteroid-sensitive neurological diseases and there is still a lack of specific biomarkers for its diagnosis. Therefore, diagnosis of CLIPPERS is challenging and is a disease yet to be explored due to its rarity. This diagnosis should be kept in mid in patients with subacute or chronic history of cerebellar ataxia.

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Chronic meningitis neurobrucellosis: case report

Ahmad Kassem El Zein[1], Lucas Henrique de Moura Rogerio Garcia[1], Matheus Rodrigues Nogueira[1], Joana Laura Aihara Barbosa[1], Josenice Gomes Trindade Seidel[1], Julia Martins Brunelli[1], Gabriela Almeida Pimentel[1], Thiago Gonçalves Guimarães[1], Lucas D'Andrea Pereira Sousa[1], Bruno Fukelmann Guedes[1]

[1]Hospital Sírio-Libanês, São Paulo SP, Brazil.

Correspondence: Ahmad Kassem El Zein; Email: ahd-95@hotmail.com.

Case presentation: A 46-year-old man presented with a 4-month history of headache and fatigue, a 10-kilogram weight loss over the previous 3 months, low-grade fever, night sweats, and blurred vision. He had recently traveled to Lebanon, where he consumed unpasteurized dairy and raw meat. Upon examination, the patient was alert but confused, exhibiting bilateral papilledema without other neurological deficits. Brain magnetic resonance imaging revealed gadolinium enhancement in the leptomeningeal compartment, particularly pronounced in the basal cisterns, accompanied by hydrocephalus and signs of CSF transudation. Cerebrospinal fluid analysis showed an opening pressure of 31 cm H2O, 149 white cells/mm3 (70% lymphocytes), low glucose (24mg/dl), high lactate (30mg/dl), and elevated total protein (160mg/dl). A comprehensive diagnostic workup, including metagenomic next-generation sequencing of CSF, was negative for syphilis, sarcoidosis, tuberculosis and other infectious and autoimmune etiologies. The patient was started on empirical anti-tuberculosis treatment, prednisone, and acetazolamide. Brucella agglutination tests were positivein the CSF (titer, 1:20) and serum (1:40). Tuberculostatics were discontinued, and replaced with an empirical anti-brucella treatment with ceftriaxone, doxycycline, and rifampicin. The patient's symptoms improved steadily over the 3-week course of antibiotics, which paralleled improvement of CSF parameters. The patient was discharged with a good clinical recovery **Discussion:** Brucellosis is a bacterial infection caused by Brucella spp, the neurological presentation is varied and may include meningitis, encephalitis, cranial neuropathies, radiculitis, and peripheral neuropathy. The clinical presentation can be very similar to tuberculous meningitis. The main route of transmission is through the consumption of unpasteurized or contaminated dairy products. The diagnosis of neurobrucellosis is based on suggestive symptoms, positive cultures of cerebrospinal fluid for Brucella spp., or positive IgG agglutination titers for Brucella in the blood or CSF, presence of lymphocytic pleocytosis and increased protein levels in cerebrospinal fluid, and positive response to specific antibiotics. Radiological aspects and cerebrospinal characteristics can aid in diagnosis, but should be differentiated from other conditions causing similar symptoms, presentations of neurobrucellosis can also be diverse, with 45% of cases showing meningeal inflammation with post-contrast enhancement. The treatment of neurobrucellosis typically involves prolonged antibiotic therapy with Ceftriaxone, rifampicin and doxycycline. The duration of therapy may be extended to 4 to 6 months, and other regimens are available. **Final comments:** Given the atypical clinical presentation of neurobrucellosis, diagnosis can be challenging, and clinical suspicion is essential in patients presenting with chronic meningitis and a history of exposure in endemic regions.

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Chronic pelvic inflammatory context leading to migraine exacerbation: a case report

Jovana Gobbi Marchesi Ciriaco[1], Lucas Grobério Moulim de Moraes[1], Laissa Fiorotti Albertino[1], Bianca Garcia Sardi[1], Brenda Comper[1], Breno Sousa Paiva[1], Beatriz Gomes da Fonseca[1], Carlos Eduardo de Nadai[1], Amanda Reis Flores[1], Ana Luíza dos Santos Machado Costa[1]

[1]Universidade Federal do Espírito Santo, Hospital Universitário Cassiano Antonio Moraes, Vitória ES, Brazil.

Correspondence: Lucas Grobério Moulim de Moraes; Email: lucasgromoulimdm@hotmail.com.

Case presentation: Woman, 48 years old, infertile, with dysmenorrhea, episodic headache since the age of 35 with weekly attacks and worsening related to the menstrual cycle. In 2020, she developed a sudden onset of dyspnea and chest pain, during menstruation which progressed to pneumothorax. A biopsy of the right upper lung lobe identified subpleural emphysema which, associated with the clinical picture of intense dysmenorrhea and magnetic resonance imaging of the pelvis, was presumptively diagnosed as a pulmonary focus of endometriosis. The patient has been menopausal since 2023 with worsening of migraine symptoms. In gynecological investigation, signs of hypoestrogenism were noted, vulvar atrophy associated with fissures in the vestibule after physical examination, cervix posterior and lateralized to the left, hypotrophic vaginal walls, shortening of the left parametrium painful upon mobilization, thickening of the endopelvic fascia and intense dysmenorrhea. After the pneumothorax, the dysmenorrhea maintained the same pattern with the headache becoming daily, located in the right temporo-occipital region, intense and pulsatile, radiating to the right hemiface with intra and extracranial allodynia, followed by progressive course, accompanied by aura with nausea, blurred vision, scotomas, photophobia and phonophobia. In the post-crisis period, there is a change in mood and hyporexia. **Discussion:** Endometriosis is a chronic gynecological condition characterized by the presence of endometrial tissue outside the uterus resulting in chronic pelvic pain and infertility. Endometriosis affects 60% of women and adolescents with pelvic pain and up to 50% of women with infertility. Migraine is a common neurological disorder characterized by recurrent attacks of intense headache often accompanied by nausea, vomiting and photophobia. The highest incidence occurs among women of reproductive age. A population-based study indicated that migraines are 1.7 times more common in women with endometriosis than in those without the disease, in addition to demonstrating that endometriosis is an independent predictor of migraines. One of the factors described in the literature on the pathophysiological pathways of endometriosis that may explain its apparent relationship with migraine is the activation of sensory fibers in ectopic endometrial tissue that can lead to neuronal hyperactivity throughout the central nervous system (CNS). Pulmonary manifestation demonstrates a possible exacerbation of neuronal activity in the CNS causing worsening of migraine. The scientific literature lacks prevalence rates of pulmonary endometriosis and migraines. **Final comments:** The role of female sex hormones in the pathogenesis of migraine is well established. The prevalence of migraine in patients with endometriosis is high and healthcare professionals need to think about both diagnostic hypotheses when caring for a patient with dysmenorrhea.

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Clinical acute manifestations in the multiple sclerosis onset associated with worse functional outcomes

Felipe Asaf Borges Millard[1], João Pedro Gonçalves[1], Gabriel Nunes Medrado[1], Marivaldo Yslan Silveira Leal[1], Luísa Dias Moreno Aravena[1], Gabriel Souza Suzart[1], Pedro Antonio Pereira de Jesus[1,2], Bruno Bacellar Pedreira[2], Thiago Gonçalves Fukuda[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil. [2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazilo.

Correspondence: Felipe Asaf Borges Millard; Email: felipemillard@ufba.br.

Background: Multiple sclerosis (MS) is a chronic autoimmune disease with a wide variability of neurological manifestations in the first acute episode. Some symptoms may be associated with long term neurological dysfunction and functional dependence in the patient's daily activities. The Expanded Disability Status Scale (EDSS) and the Multiple Sclerosis Functional Composite (MSFC) are complementary and aim to quantify the degree of disability of patients with MS. In this sense, data regarding the incidence and liability implications of the first acute manifestation are still lacking. This knowledge may help re-evaluate therapeutic protocols in MS. **Objective:** To identify the clinical syndromes in MS onset that are associated with worse functional outcomes. **Methods:** This study is a cross-sectional design with patients diagnosed with MS, according to the McDonald 2017 criteria, who were followed up at a neuroimmunology reference center in Bahia, from January 2019 to May 2024. This study is part of an ongoing cohort study that evaluates the clinical features of patients followed in this reference center. The clinical manifestations of the first acute episode were collected through a clinical interview and compiled into seven major syndromes: visual, motor, sensory, ataxic, postrema area, cranial nerves and autonomic. Functional outcomes were assessed using the EDSS (assessed by neurologists and/or neurology residents) and the MSFC (assessed by trained researchers).Patients with MS and a relapse in the last three months were excluded. The Mann-Whitney test was used to assess the association of the first clinical syndrome and worse functional outcomes. Linear regression was performed to address the effect of each clinical syndrome in both functional scales, considering only syndromes that were associated in univariate analysis with worse functional outcome. **Results:** A total of 274 patients were included in the study, of whom the majority were female (71.5%), non-white (71.9%) and complete secondary education (79.6%). Our sample's mean age was 42 (SD±13) with a median length of disease of 8 years (4-13), a median EDSS of 2 (0-4.5), a global MSFC of -0,6 (-1.6 to 0.0), and relapsing-remitting clinical form as the most common (87.6%). Worse EDSS and global MSFC scores were identified in those patients who presented motor ($p< 0.01$), ataxic ($p<0.01$), and autonomic syndrome ($p<0.01$) in the first acute episode.In multivariate analysis considering EDSS as the dependent variable, motor and autonomic syndrome remained associated with worse functional outcome ($p<0.001$, B=0.373;

p0,05, B=0.163, respectively). Regarding MSFC, we found that motor, ataxic, and autonomic syndromes remained associated (p<0.05, B=-0.206; p<0.05, B=-0.188; p<0.05, B=-0.156). **Conclusion:** Among the possible symptoms in the first acute episodes of patients with Multiple Sclerosis, motor, ataxia and dysautonomic symptoms may be associated with worse functional status.

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Clinical and electroneuromyographic assessment in lepromatous leprosy: the confirmation of an asymmetrical and non-length-dependent pattern.

Lucas Kenzo Ozera[1], Caio Luiz Nazar Cunha[1], Ana Clara Nogueira Cezar[1], Ariane Aparecida Correa de Miranda[1], Laura Caroline Felipe de Freitas[1], Eduardo William Farinha Carvalho[1], Isabella Sabião Borges[1], Douglas Eulálio Antunes[1], Isabela Maria Bernardes Goulart[1], Diogo Fernandes dos Santos[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Lucas Kenzo Ozera; Email: lucaskenzoozera@gmail.com.

Background: Electroneuromyography (ENMG) is the gold standard test for evaluating neural impairment in leprosy. However, as it is a spectral disease and especially in advanced cases, the neurophysiological pattern is variable. It can be confused with numerous etiologies presenting a symmetrical polyneuropathy pattern. **Objective:** To describe electroneuromyographic findings in patients with lepromatous leprosy (LL). **Methods:** The study is a retrospective observational analysis of leprosy patients undergoing ENMG at a National Reference Center in Brazil. **Results:** 57 patients with LL underwent ENMG at diagnosis. On physical examination, all patients presented an asymmetrical distribution pattern of sensory symptoms, compatible with truncal and intradermal involvement in all cases. 89.5% had neural thickening, and 64.9% had motor symptoms. Concerning the ENMG findings, 417 altered nerves were identified, with an average of 7.9 (±4.2). 100% showed abnormalities in at least one sensory nerve, with an average of 5.5 (±2.3), and 89.0% in at least one motor nerve, with an average of 2.4 (±1.3). 3.6% had mononeuropathy, and 96.4% had multiple mononeuropathy, with asymmetry present in all cases. 100% presented sensory axonal impairment and 40.0% motor axonal impairment. 70.9% showed signs of demyelinating. The most commonly observed electroneuromyographic pattern was an axonal sensory-motor neuropathy, predominantly sensory, with focal myelin impairment in 61.9%. No patient presented a symmetrical neurophysiological pattern compatible with the diagnosis of polyneuropathy. **Conclusion:** The term polyneuropathy defines a diffuse, length-dependent, symmetrical involvement with distal predominance. Therefore, although confluent, the term polyneuropathy should not be used in this disease that presents an essentially asymmetrical involvement, even in LL.

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Clinical and epidemiological aspects of patients attended at a reference center for movement disorders

Caio Fontes d Nascimento[1], Maria Eduarda Messias Vassoler[1], Gabriel Souza Medrado Nunes[1], Ayla Morais Vitório Guerra[1], Fernanda Garcia Silva Pereira Nascimento[1], Gabriel Teles de Oliveira Piñeiro[1], Roberta Borges Gomes Kauark[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Caio Fontes do Nascimento; Email: caiofontesdn@hotmail.com.

Background: Movement disorders are neurological conditions characterized by involuntary movements that significantly impact patients' quality of life. These disorders are commonly divided into hyperkinetic and hypokinetic categories. Hyperkinetic disorders include a variety of uncontrollable movements such as dystonias, tremors, myoclonus, and athetosis. Hypokinetic disorders are primarily characterized by parkinsonian syndromes, with Parkinson's disease (PD) being the most prominent and the second most common neurodegenerative disease. The prevalence of movement disorders in the general population is low and specific data on the Brazilian population are scarce, so studies exploring the characteristics of affected populations are crucial for developing better therapeutic strategies. **Objective:** To describe the frequencies of the different diagnoses among patients at the movement disorders clinic and their clinical manifestations, as well as their epidemiological data. **Methods:** This is a retrospective descriptive epidemiological study that analyzed medical records of patients seen at a referral movement disorders outpatient clinic from January 2019 to May 2024. **Results:** Among the 279 patients followed during the period, 57.6% were male, with a mean age of 65.1 years (±14.4). The most frequent diagnosis was PD (48.7%), followed by other forms of parkinsonism (11%), essential tremor (3.6%), Huntington's disease (2.2%), cervical dystonia (1.8%), Segawa's disease (1.1%), Wilson's disease (1.1%), and other diagnoses (9.3%). Additionally, 59 patients (21.2%) remained under investigation. Of the PD patients (136), 94.1% had bradykinesia, 91.1% were rigid, 73.5% had rest tremor and 29.4% had gait block. Regarding non-motor symptoms: 35.2% of patients had REM sleep behavior disorder, 28.6% had insomnia, 33% had constipation and 16% had orthostatic hypotension. Tremor was reported as the first symptom by 53 of these patients (38.9%). The mean Levodopa Equivalent Daily Dose (LEDD) was 638 mg (±677), with 64.3% of patients using levodopa-benserazide. **Conclusion:** The gender distribution and average age of the sample are in line with known epidemiological trends for PD, which suggests an accurate representation of the patient population. The predominance of PD among the diagnoses reflects its high prevalence. The high proportion of undiagnosed cases indicates the challenges of diagnosing movement disorders and scarce resources. Tremor was the most frequently reported initial symptom, which highlights its importance in early diagnosis and patient management. The frequencies of rigidity, bradykinesia, postural instability, insomnia, and REM sleep behavior disorder indicate a relevant prevalence of conditions that can potentially affect patients' quality of life. The average LEDD reflects typical therapeutic practices, and its variability suggests different stages of the disease, necessitating personalized therapeutic approaches.

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Clinical and epidemiological aspects of patients with myasthenia gravis in a tertiary center

Beatriz Carneiro Gondim Silva[1], Eduardo de Paula Estephan[1], Ana Flávia Pincerno Pouza[1], André Macedo Serafim da Silva[1], Antonio Alberto Zambom[1], Edmar Zanoteli[1], Sara Caixeta de Souza[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Beatriz Carneiro Gondim Silva; Email: beatrizcgondim@gmail.com.

Background: Myasthenia gravis (MG) is a rare autoimmune disease with variable incidence depending on the geography and over the years. In the literature, there are few studies describing the incidence of MG in centers specializing in neuromuscular diseases and neuromuscular junction disorders, as well as the epidemiological and clinical profile of the population evaluated for this condition. **Objective:** To describe clinical and epidemiological aspects of patients diagnosed with MG at the neuromuscular diseases in a large tertiary service. **Methods:** A retrospective analysis was carried out of medical records of patients admitted and diagnosed with MG between the period 2020 and 2024. Data were collected regarding age, sex, ethnicity, onset of symptoms, severity, presence of antibodies, thymectomy, time since thymectomy and response afterwards, number of myasthenic crises, need to use intravenous immunoglobulin and/or plasmapheresis, in addition to associated comorbidities. **Results:** 217 patients were studied, of whom 152 were females (70%) and 65 were male (30%), with an average age of 51.4 years, ranging from 11 to 90 years. Age of onset of the first symptoms varied from 3 - 69 years (average of 31.7). Generalized myasthenia gravis was the most common clinical presentation, with MGFA IIA, IIIA and IIIB being the most common in the early pre-treatment phase. From those tested for anti-acetylcholine receptor (AChR) and anti-Musk antibodies 88% were AChR positive and almost 6% anti-MuSK. Thymectomy was performed in 107 patients (49%), with an average time to perform thymectomy of 4.3 years and pathology demonstrating thymoma in 16%, with the type B2 being the most common. After thymectomy, 23 patients presented myasthenic crisis requiring immunoglobulin and/or plasmapheresis. Concomitant comorbidities evaluated, most patients had high blood pressure and diabetes. In addition, 4 patients were found with associated rheumatoid arthritis, 1 with multiple sclerosis and 1 with Parkinson's disease. **Conclusion:** In this single center study, the data analyzed are similar with other studies in the literature, but with a higher percentage of thymectomized patients. However, comparing to another Brazilian cohort reported, from a different region of the country, the antibodies profile is quite different, and main reflect regional influences (genetic and/or environmental) on immunopathogenesis of MG.

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Clinical and epidemiological characteristics of patients with Parkinson's disease being investigated for obstructive sleep apnoea: an observational analysis in the Brazilian Amazon

Marcello Facundo do Valle Filho[1], Diego Monteiro de Carvalho[1], Carlos Maurício Oliveira de Almeida[1], Marcus Vinicius Della Coletta[1], Alvaro Siqueira da Silva[1], Raíssa Costa Said[1], Carlota Assayag Mendes[2], Yenly Gonzalez Perez[1], Luma Santos Arruda[2], Ana Carolina Freire Nogueira[2]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Faculdade Metropolitana, Manaus AM, Brazil.

Correspondence: Marcello Facundo do Valle Filho; Email: marcello_valle@outlook.com.

Background: Parkinson's disease (PD) is the second most common neurodegenerative disease in the elderly, representing about 1% of the population over 60 years old. Literature data demonstrate that in PD, sleep disorders are observed in up to 90% of patients. Obstructive sleep apnoea (OSA) is diagnosed in more than 50% of these patients, and its presence in PD can impact the quality of life by increasing daytime sleepiness, thereby negatively influencing both motor and non-motor symptoms of the disease. Understanding the clinical and epidemiological characteristics of PD patients being investigated for OSA is essential to improve the diagnosis and management of these concomitant conditions. **Objective:** To analyse the clinical and epidemiological characteristics of

PD patients being investigated for OSA. **Methods:** This is a cross-sectional observational study, with a sample composed of PD patients from the neurology clinic at the Adriano Jorge Hospital Foundation (FHAJ), Manaus-AM. The study was approved by the local Ethics Committee under Certificate of Presentation of Ethical Consideration No. 58684622.2.0000.0007. Patients underwent otorhinolaryngological and neurological evaluations, had their physical data and vital signs collected, and their clinical and epidemiological data were gathered after signing the Informed Consent Form. Patients were also subjected to the Mini-Mental State Examination (MMSE) for cognitive screening. **Results:** Of the 33 patients, the minimum and maximum ages were 31 and 88 years, respectively, with a minimum disease duration of 1 year and a maximum of 33 years, with an average disease duration of 8.1 years, a coefficient of variation of 79.8%, and a 95% confidence interval (CI) of 5.7%-10.3%. Regarding neck circumference, the average was 34.5cm, with a minimum of 20cm and a maximum of 44cm, a coefficient of variation of 5.2%, and a 95% CI of 5.7-10.3%. The majority were male (57.6%-19 patients) and 42.5% (14 patients) were female. Most individuals were married (66.7%), 21.2% were single, 9.1% divorced, and 3% widowed. Considering the body mass index, 48.5% of the patients were overweight and 12.1% had grade 1 obesity, while the remaining 39.4% were eutrophic. Only 3% of the patients had postgraduate education, 24.2% had completed higher education, 57.6% had completed secondary education, and 15.2% had only primary education. **Conclusion:** The majority of PD patients in this study are male, with an average age of 62 years, married, overweight, and with an average disease duration of 8 years. There was no association with low education levels. This information is crucial for a better understanding of the characteristics of PD, thereby contributing to the identification of risk factors, improving diagnosis, and developing more effective management strategies for this population.

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Clinical and epidemiological characterization of myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) in a tertiary center in Amazonas

Andreia Lira de Oliveira[1], Nise Alessandra de Carvalho Sousa[1], João Victor de Chagas Rossin[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Andreia Lira de Oliveira; Email: andreialira.med@gmail.com.

Background: Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) is the most recently characterized inflammatory disorder of the central nervous system (CNS). It is a demyelinating disease in which an inflammatory response targets this glycoprotein, which is uniquely present on the surface of CNS myelin sheaths. Clinical presentation is diverse and includes optic neuritis, myelitis, brainstem syndrome, acute disseminated encephalomyelitis (ADEM), and FLAIR-hyperintense lesions in anti-mog associated encephalitis with seizures (FLAMES). Diagnosis is made based on compatible clinical and radiological findings, along with the presence of anti-MOG IgG in serum or cerebrospinal fluid. **Objective:** To characterize cases diagnosed with MOGAD managed at the Getúlio Vargas University Hospital of the Federal University of Amazonas (HUGV/Universidade Federal do Amazonas). **Methods:** A retrospective study was conducted by reviewing the medical records of patients diagnosed with MOGAD between 2019 and 2024. **Results:** 6 cases of MOGAD managed by the service were identified during the study period, with 4 cases presenting initially after the age of 20 years. One case occurred in a 7-year-old child. There was no gender difference in the distribution of cases. The most common presentations were optic neuritis, myelitis, and optic neuritis associated with myelitis, with only 1 case of ADEM. Only 1 patient had a preceding infection before symptom onset, and none had been vaccinated in the days preceding the onset. None of the patients had associated autoimmune diseases. All patients showed hyperintense lesions on T2-weighted MRI, and all received acute phase treatment, albeit delayed, as well as ongoing chronic immunosuppression. Five cases experienced symptom recurrence, with myelitis being the most common recurrent clinical presentation. **Conclusion:** Given the recent recognition of MOGAD as a distinct clinical entity, it is crucial to characterize identified cases in different populations. In our study population, a notable finding was that recurrence predominantly manifested as myelitis, contrasting with current literature that mentions optic neuritis as the primary recurrent presentation. Further studies are needed to confirm this characteristic.

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Clinical and epidemiological characterization of patients with Guillain-Barré syndrome (gbs) attended at a university hospital, 2014-2018

Lilian Yatiyo Nakagawa Dittmar[1], Isabela de Carvalho Florencio[1], Thiago Dias Fernandes[1], Rodrigo Ibañez Tiago[2], Dalton Santos Pinheiro[1], Rodolfo Antonio Corona[1]

[1]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.
[2]Universidade Federal de Mato Grosso do Sul, Faculdade de Medicina, Campo Grande MS, Brazil.

Correspondence: Isabela de Carvalho Florencio; Email: isabelacflorencio@hotmail.com.

Background: Guillain-Barré syndrome (GBS) is an inflammatory polyradiculopathy characterized by acute or subacute symmetrical weakness, areflexia, with mild sensitive symptoms and autonomic manifestations. The diagnosis is essentially clinical, although further examinations are useful for differential diagnosis. Early diagnosis and treatment can modify the disease's natural progression. **Objective:** Tto describe clinical and epidemiological aspects of GBS in a university public hospital of Mato Grosso do Sul state, Brazil. **Methods:** Cross-sectional descriptive study of GBS patients admitted to a university hospital, in Campo Grande/MS from January 2014 to December 2018. **Results:** Between January 2014 and December 2018, 29 patients were admitted with GBS. The incidence was 0.2/100,000 inhabitants per year. The men/women ratio was 1.6 with no statistical significance (p=0.1). Eighteen patients were admitted in the summer-spring period and 11 in the winter-autumn (p=0.1). Twenty-nine patients (93.1%) presented the classic subtype. Sixteen patients presented facial involvement (58.6%), and of these, six (37.5%) needed orotracheal intubation (p=0.1). Autonomic manifestations such as heart rate and blood pressure changes, intestinal constipation, and urinary retention were present in 62.1%. Albumin-cytological dissociation of cerebrospinal fluid (CSF) was observed in eighteen patients with a mean protein value of 103.4 mg/dL. The average duration of hospitalization was 19,7 days and 34.5% needed intensive care. Full recovery occurred in 52.4% of patients, evaluated 60 days after discharge. **Conclusion:** The data obtained are in accordance with the literature. This is the first study for understanding the profile of GBS patients assisted in Mato Grosso do Sul state of Brazil.

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Clinical and epidemiological evaluation of sleep disorders in chronic kidney patients at a dialysis center in Amazonas, Brazil

Ana Carolina Souza Galvão[1], Eduarda Kei-lan Tavares Pierre[1], Mariane Marques Queiroz[1], Lucas de Souza Ribeiro[1], Gabrielle Logatto Ignacio de Souza[1], Virgínia Souza Galvão[1], Gabriel Vinicius Silva de Carvalho[1], Ana Matilde Schramm[1], Carlos Maurício Oliveira de Almeida[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Eduarda Kei-lan Tavares Pierre; Email: ektp.med20@uea.edu.br.

Background: Sleep disorders range from simple insomnia to more severe conditions, affecting overall bodily function. In Brazil, about 20 million people report some form of sleep disorders. Studies show a strong relationship between sleeping shorter or longer than normal with the prevalence and severity of chronic diseases such as hypertension, diabetes and cardiovascular diseases. Among patients with chronic kidney disease (CKD), up to 80% prevalence of sleep disorders can occur, which occurs both in patients in the early stages and in patients on maintenance dialysis. Therefore, it becomes relevant to gather clinical and epidemiological data from patients with CKD undergoing dialysis and correlate them with sleep disorders to assess whether there is evidence of a worse prognosis in these patients. **Objective:** Investigate the main sleep disorders in chronic kidney patients at a reference center for kidney diseases in Amazonas. **Methods:** The proposed study is a cross-sectional observational type and will be conducted at the Amazonas Kidney Disease Center, in Manaus, from December 2023 to March 2024. The sample will include CKD patients from the clinic, selected after approval by the Ethics Committee and signing of the Informed Consent Form. Patients underwent extensive data collection, including medical information, completion of sleep questionnaires such as Mini Sleep Questionnaire, Epworth Scale, Stop bang questionnaire and the diagnosis of Restless Legs Syndrome (RLS) that followed clinical criteria. **Results:** 100 CKD patients were evaluated, 63 (63.0%) of whom were male, 51 (51%) aged between 50-69 years, and 43 (43%) in the overweight range. Considering the MSQ, 60 (60%) have highly altered sleep; 43 (43%) have abnormal daytime sleepiness according to the Epworth scale; and, according to the Stop-Bang questionnaire, 41 (41%) are at high risk of developing sleep apnea. According to a previous assessment, of the 100 patients, seven (7%) were diagnosed with RLS. Of the seven patients, one (14.3%) had a mild degree, one (14.3%) had a moderate degree, four (57.1%) had a severe degree, and one (14.3%) had a very severe degree of RLS. There was no statistically significant evidence that sleep quality is related to the presence of RLS in chronic kidney disease patients (p> 0.05), considering the 5% level of significance. **Conclusion:** In this study, 60% of CKD patients through MSQ had alterations in sleep patterns, suggesting complexity in sleep conditions, with a variety of challenges ranging from daytime sleepiness to potential risks of sleep apnea. These findings underscore the importance of a holistic approach in managing the sleep of these patients, necessitating further studies for deeper exploration of this topic.

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Clinical and epidemiological profile of hospital admissions due to cerebral venous thrombosis in a reference hospital

Joanna Sousa da Fonseca Santana[1], Amanda Hellen Sousa Leal[1], Ana Clara de Lemos Guimarães[1], Catarina Cavalcanti da Silva[1], Katarina Brito Gama[1], Brena Lima Cunha[1], Lucas Paulo Andrade Borges[1], Maria Clara Bastos de Sá[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Joanna Sousa da Fonseca Santana; Email: joannasantana20.2@bahiana.edu.br.

Background: Cerebral venous thrombosis (CVT) is a condition where a clot causes occlusion of the cerebral sinus or vein, affecting cerebral blood drainage. About 15% of cases result in death or permanent disability, with the most common occlusion sites being the transverse sinus, superior sagittal sinus, and sigmoid sinus. Given its prevalence and impact on healthcare, understanding the sociodemographic profile of CVT patients is crucial in brazilian reality. **Objective:** To describe the clinical and epidemiological profile of hospital admissions due to cerebral venous thrombosis. **Methods:** This descriptive observational study uses preliminary data from a sample of 75 patients and was conducted based on medical records from a reference hospital in Bahia from January 2015 to August 2023. The sample is a non-probabilistic convenience sample. Inclusion criteria: all patients diagnosed with CVT in a reference hospital in Bahia between January 2015 and December 2023 were included. Patients without demographic, clinical, and pathological data available for the study were excluded. Variables of interest were analyzed using SPSS (Statistical Package for the Social Sciences). The study was approved by the Research Ethics Committee. **Results:** Data from 39 admissions were analyzed, with 3 (7.6%) deaths. The mean age was 35 years, ranging from 20 to 42 years. Of the patients, 30 (76.9%) were female. Imaging reports from 38 patients revealed that the most affected vessels were the transverse sinus (32), sigmoid sinus (29), and superior sagittal sinus (28). The number of affected vessels per patient varied: 4 vessels (12 cases, 30.7%), 3 vessels (11), 2 vessels (8), 1 vessel (5), 5 vessels (3), and 6 vessels (1). Among the 30 women, 17 (56.6%) used oral contraceptives. Additionally, 14 (35.9%) had pre-existing risk conditions: 5 with thrombophilias, 4 with previous CVT, 2 with recent surgeries, 1 during pregnancy/puerperium, 1 with hyperhomocysteinemia, and 1 with a family history of CVT. The most common symptom was headache (85%, 33 cases), followed by visual changes (38%, 15), motor symptoms (28%, 11), sensory symptoms (21%, 8), and cognitive symptoms (13%, 5). Focal deficits were present in 31% of cases. No patient was admitted during a seizure, but 6 (15.4%) had seizures during hospitalization. Laboratory data showed an average MAP of 98.6 mmHg, with maximum value of 133.3 mmHg. Regarding interventions, 33 (84.6%) patients required anticoagulation during hospitalization. **Conclusion:** The study's findings align with existing literature regarding the patient profile and emphasize the significant impact on neurological care, including the risks of intracranial hypertension and seizure development. Effective preventative strategies and early interventions targeting high-risk groups are essential to improve outcomes for CVT patients. By focusing on these areas, healthcare providers can better manage and reduce the incidence and impact of CVT.

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Clinical and functional profile of patients with ischemic stroke secondary to arterial dissections

Lealdo Rodrigues de Andrade Filho[1], Yasmin Gabrielle Pereira Pacheco Cavalcanti de Albuquerque[2], Rayane Barbosa Do Amaral[2], Arthur Cavalcanti Pereira da Silva[2], Elizabeth Costa Ferreira de Sousa[2], Ana Beatriz Leite Amando[2], Sara Raquel Ataíde de Oliveira[2], Nicole Maria dos Santos Silva[2], Marcelo Ataíde de Lima[1]

[1]Hospital da Restauração, Recife PE, Brazil.
[2]Centro Universitário Maurício de Nassau, Recife PE, Brazil.

Correspondence: Lealdo Rodrigues de Andrade Filho; Email: leroanfilho@gmail.com.

Background: Cervicocephalic arterial dissection is the loss of integrity of the arterial wall. It is the main cause of stroke in young people identified in industrialized countries, being responsible for approximately 2% of all strokes and 20% in the age group under 45 years, being an important condition to incapacity for adjusted life years. **Objective:** To describe clinical-epidemiological characteristics, in addition to exploring the functional outcome after 90 days of patients diagnosed with cervicocephalic arterial dissection. **Methods:** Observational, descriptive study, based on the review of patient records from the reference tertiary hospital in the State of Pernambuco and referenced health regions, Brazil, between January 2013 and January 2023. **Results:** In this retrospective collection of 10 years in reference hospital, the total number of patients was 73. The average age is 47.6±14.1 years. The absence of comorbidities was present in 40% of patients. There was greater trauma exposure in younger people when compared to patients without associated trauma (40.3±15.4 vs 49.9±11.9 years, p=0.035). There was a significant predominance of patients with a favorable functional outcome after 90 days with posterior circulation involvement compared to an unfavorable outcome (91% vs 42.9%, p=0.003). It was also observed that patients with unfavorable scores were admitted with lower Glasgow Coma Scale scores, more bleeding, as well as longer hospital stays, but all variables were not statistically significant. **Conclusion:** Younger patients were associated with a precipitating factor when compared to older patients, being a critical condition for greater loss of years of life due to disability. Posterior circulation strokes have an apparent better outcome compared to anterior circulation, questioning the choice of tool to assess functionality.

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Clinical and genetic characterization of limb-girdle muscular dystrophy 2G/R7 in a large cohort of Brazilian patients

Jonas Alex Morales Saute[1,2], Eduardo B. U. Cavalcanti[3], Paulo Lorenzoni[4], Paulo V. S. de Souza[5], Marcondes Cavalcante França Jr[6], Cláudia F. R. Sobreira[7], Elmano H. T. Carvalho[8], Alzira A. S. Carvalho[9], Tobias Gaviraghi[12]

[1]Hospital de Clínicas de Porto Alegre, Serviço de Genética Médica, Porto Alegre RS, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Serviço de Neurologia, Porto Alegre RS, Brazil.
[3]Rede SARAH de Hospitais de Reabilitação, Brasília DF, Brazil.
[4]Universidade Federal do Paraná, Hospital de Clínicas, Departamento de Medicina Interna, Divisão de Neurologia, Serviço de Doenças Neuromusculares, Curitiba PR, Brazil.
[5]Universidade Federal de São Paulo, Departamento de Neurologia e Neurocirurgia, Divisão de Doenças Neuromusculares, São Paulo SP, Brazil.
[6]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Departamento de Neurologia, Campinas SP, Brazil.
[7]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Departamento de Neurociências, Ribeirão Preto SP, Brazil.
[8]Rede SARAH de Hospitais de Reabilitação, Belo Horizonte MG, Brazil.
[9]Centro Universitário Saúde ABC, Santo André SP, Brazil.
[10]Lgmd Brazilian Consortia Additional Authors.
[11]Universidade Federal do Paraná, Hospital de Clínicas, Departamento de Medicina Interna, Curitiba PR, Brazil.
[12]Universidade Federal do Rio Grande do Sul, Pós-Graduação em Medicina: Ciências Médicas, Porto Alegre RS, Brazil.

Correspondence: Jonas Alex Morales Saute; Email: jsaute@hcpa.edu.br.

Background: Limb-girdle muscular dystrophy type 2G/R7 (LGMD2G/R7) is an ultra-rare condition caused by bi-allelic pathogenic variants in the TCAP gene that was initially identified within the Brazilian population. **Objective:** We aimed to expand clinical and genetic information about this disease, including its worldwide distribution. **Methods:** A multicenter historical cohort study was performed at 13 centers in Brazil from Middle west, South and Southeast in which data from index cases and their affected relatives from consecutive families with LGMD2G/R7 were reviewed from July 2017 to August 2023. Additionally, a systematic literature review was conducted to identify case reports and series of the disease worldwide. **Results:** Forty-one LGMD2G/R7 cases were described in the Brazilian cohort, being all subjects homozygous for the c.157C> T/(p.Gln53*) variant in TCAP. Survival curves showed that the median disease duration before individuals required walking aids was 21 years. Notably, women exhibited a slower disease progression, requiring walking aids 13 years later than men. LGMD2G/R7 was frequently reported not only in Brazil but also in China and Bulgaria, with 119 cases identified globally, with possible founder effects in the Brazilian, Eastern European, and Asian populations. **Conclusion:** These findings are pivotal in raising awareness of LGMD2G/R7, understanding its progression, and identifying potential modifiers. This can significantly contribute to the development of future natural history studies and clinical trials for this disease.

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Clinical and genetic profile of patients with chanalopathies in a reference center for neuromuscular diseases: retrospective observational study

Izadora Vieira Bolzani Lopes Lima[1], Felipe Franco da Graça[1], Anamarli Nucci[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Izadora Vieira Bolzani Lopes Lima; Email: izadoralima7@gmail.com.

Background: Channelopathies are rare muscle disorders caused by mutations in genes that encode ion channels. The most common phenotypes are congenital myotonia, paramyotonia and periodic dyskalemic paralysis. The definitive diagnosis is based on genetic tests and patients can take years to have a defined diagnosis, often undergoing unnecessary and high-cost interventions. The relative low availability of clinical studies on this topic in Brazil brings the need for research in order to establish standards and make possible future efforts to find pharmacological treatments that help in symptomatic control. **Objective:** To describe clinical, genetic and electrophysiological characteristics in patients with muscle channelopathies diagnosed by next sequencing generation tests in a specialized outpatient clinic of a high-complexity hospital based on an observational, retrospective study and to describe the diagnostic journey of these patients. The study was approved by the local Ethics Committee. **Methods:** Data were collected from the medical records of 29 patients who presented with signs and symptoms of channelopathy. We excluded 5 patients with genetic mutation in the RYR1 gene without compatible clinical findings and also 9 patients who had a suggestive phenotype without genetic proof. We compared clinical features between patients according to genetic variations using non-parametric tests, given the sample size. The Mann-Whitney test for continuous variables and Fisher's exact test for categorical ones. **Results:** Fifteen patients met inclusion criteria and had diagnostic confirmation through an NGS panel for pathogenic variants related to channelopathies. We identified 22 variants in 3 different genes: SCN4A (6 cases, 40%), CLCN1 (8 cases, 53.3%) and KCNJ2 (1 case, 6.66%). Of these patients, ten (66.6%) presented an AD inheritance pattern. Missense mutations in all positive panels accounted for 72.7% of mutations. It was noted greater difficulty in establishing a diagnosis in sodium channelopathies, with the highest percentage of misdiagnosis in these cases (83.3% versus 37.5%). We

also observed the long journey until diagnostic definition (median 34.5 years). The presence of warm up phenomenon was present in all patients with chloride CLCN1 and paramyotonia was present in most patients with SCN4A, indicating the role of the active search for these phenomena in the differentiation between conditions (p=0.015). Weakness on physical examination, hypertrophy, presence of cramps, periodic paralysis, seizures, increased CK and diet have not shown statistical significance between the groups. We have found a new variant for the CLCN1 mutation (c.2284+1G> A) and for the KCNJ2 (c.788t> G - p.Leu263Arg) not previously described in literature. **Conclusion:** NGS panel has proven to be a relevant diagnostic tool in the assessment of channelopathies and the knowledge of clinical variables linked to these diagnoses enables better clinical practice.

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Clinical and genetic spectrum of the ZFYVE26 gene: an analysis of 7 patients

Renan Brandão Rambaldi Cavalheiro[1], Kaliny Oliveira Peixoto[1], João Paulo Barile[1], Patrícia Marques Mendes[1], Igor Braga Farias[1], Paulo de Lima Serrano[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Renan Brandão Rambaldi Cavalheiro; Email: renan.rambaldi@icloud.com.

Background: The hereditary spastic paraplegias (HSPs) are a heterogeneous group of monogenic diseases characterized by length-dependent corticospinal tract degeneration that manifests as bilateral lower limb spasticity, hyperreflexia and extensor plantar responses. They can be further divided into pure and complex, based on additional clinical features, such as epilepsy or cognitive decline. Their genetic classification uses the designation spastic paraplegia gene (SPG). SPG15/Kjellin syndrome, caused by biallelic pathogenic variants in the ZFYVE26 gene, is the second most common autosomal recessive HSP. While a complex HSP phenotype is described, the clinical spectrum of the ZFYVE26 has expanded over the years, with movement disorders and motor neuron disease being reported as possible clinical features. **Objective:** Describe the clinical characteristics of 7 patients with genetically proven SPG15/jALS. **Results:** The average age at onset of symptoms was 16.4 years (range: 9-24 years) and the average age at diagnosis was 27.6 years (range: 14-45 years). Consanguinity was observed on 4 patiens. Two main phenotypes were observed: 3 patients had a clinical presentation compatible with juvenile amyotrophic lateral sclerosis (jALS), while the other 4 patients were diagnosed with a HSP phenotype. Distinguishing features between the two groups were the presence of cerebellar ataxia (75% in the HSP group, none in the jALS group), muscle fasciculations (66% in the jALS group, absent in the HSP group) magnetic resonance imaging (corpus callosum thinning was observed in all patients; the classic "ears of the lynx" sign present in 100% of the HSP patients, more diffuse cortical atrophy and white matter changes in the jALS group). Retinal "yellowish flecks" were observed on two patients (one on each group). Cognitive decline was observed on five patients, ranging from moderate dysexecutive changes on 3 patients in the HSP to dementia resembling frontotemporal lobar degeneration (1 patient in each group). Dystonia was observed as the sole movement disorder in two patients, while a combination of dystonia and parkinsonism was observed in one patient. **Conclusion:** The ZFYVE26 spectrum ranges from a complex HSP to a disorder resembling FTD-ALS; additional clinical features include dystonia, parkinsonism and cerebellar ataxia. MRI remains an useful diagnostic tool to distinguish between ZFYVE26 HSP/ ALS from other genetic o acquired disorders.

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Clinical and genotypic characteristics of patients with Nonaka distal myopathy at a specialized clinic in Salvador, BA

Ananda Vieira de Lima Almeida[1], Rafael Amaral de Matos Albuquerque[1], Maria Carolina Freitas Ferreira[1], Laís Brandão Mota[1], Guiniver Couto Almeida[1], Marcela Câmara Machado Costa[1], Marilaine Matos de Menezes Ferreira[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Rafael Amaral de Matos Albuquerque; Email: rafaelalbqq@outlook.com.

Case presentation: Distal myopathy of the Nonaka type is a rare genetic disorder, with an estimated prevalence of 1 in 1,000,000. It is progressive and inherited autosomally recessively, caused by mutations in the GNE gene. This gene encodes the enzyme UDP-N-acetylglucosamine 2-epimerase/N-acetylmannosamine kinase, vital for sialic acid biosynthesis. The disease typically manifests in the third decade of life with predominant distal muscle weakness. We describe five patients from four distinct families diagnosed with GNE myopathy at a specialized clinic in Salvador, BA. The group comprises three women and two men, averaging 38.6 years old. All had distal lower limb muscle weakness starting at around 25 years old, yet the average time to genetic diagnosis was 9.6 years. All five patients showed foot drop as a primary clinical feature from anterior leg compartment weakness. Two patients had consanguineous parents. One initially presented with dropped head syndrome, an atypical late manifestation, currently using a cervical collar. This patient is the only one in this series with respiratory involvement, experiencing respiratory effort and dyspnea. Genetic analysis revealed the following GNE mutations: c.829C> T (p.Arg277Trp) in homozygosity, c.766G> A (p.Asp256Asn) in heterozygosity, and G> A in homozygosity. The c.829C> T variant is present in three patients and is one of the most common pathogenic variants associated with this disease. The c.766G> A (p.Asp256Asn) variant, present in one patient, is documented in the literature and considered probably pathogenic. Muscle biopsies from four patients showed autophagic vacuoles and cytoplasmic inclusions consistent with the disease. Three patients use wheelchairs, with a mean onset of 5.6 years after molecular diagnosis. All patients are currently receiving multidisciplinary outpatient care following their confirmed diagnosis. **Discussion:** This case series is crucial for advancing scientific understanding of the genotype-phenotype correlation of this disease. One case presents atypically with the onset of dropped head syndrome, a manifestation typically seen only in advanced stages due to the progressive distal-to-proximal muscle involvement affecting the neck flexors. The usual clinical presentation is distal leg weakness, evident as foot drop, followed by slow progression to upper musculature. Respiratory involvement, rare in literature, occurs in advanced stages. All identified mutations have been previously documented. Most mutations are seen across multiple families or within a single family with several cases, impacting symptom onset, presentation, severity, and disease progression. **Final comments:** Understanding clinical-genotypic aspects of Nonaka distal myopathy is crucial for comprehending disease behavior. As a rare condition with limited global research, this series can enhance knowledge through investigating genotypic and phenotypic characteristics.

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Clinical and imaging findings in language-predominant dementia syndromes: a case series

Marcela Maria Cabral Ribeiro[1], Pedro Jatobá Arteiro[2], Alonso Arlen Rodrigues Pimentel de Almeida[2], Maria Regina Vendas Carneiro Leão[2], Victoria Hellen da Silva Barbosa[2], Gabriel Felipe Almeida de Barros[2], Raphaelly Ribeiro Campos[2], Tatiana Caldas Neves da Silva[2], Rodrigo Cavalcanti Machado da Silva[2], Breno José Alencar Pires Barbosa[2]

[1]Universidade de Pernambuco, Recife PE, Brazil. [2]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Marcela Maria Cabral Ribeiro; Email: mmcabralribeiro@gmail.com.

Background: Primary progressive aphasia (PPA) is a rare neurodegenerative disorder, with estimated prevalence of 3-4 per 100.000 population, characterized by predominant language impairment, alongside prejudice to daily activities and without equivalent disturbance on behavior and episodic memory during symptoms emerge. While PPA is the prototype of a language-predominant dementia syndrome, aphasia could be the presenting symptom of other conditions such as Corticobasal Syndrome, defined as a combination of cortical and subcortical signs usually affecting the parietal lobes asymmetrically. **Objective:** Describe five PPA cases of patients followed at a cognitive neurology reference center in Recife. **Methods:** Case series with thorough revision of health-records, imaging and laboratory tests of all patients classified with language-predominant dementia syndromes between 2019-2023 in a tertiary center. **Results:** Five female patients were described, with ages from 56 to 65 years (average 60.5 years). Language impairment was the first isolated manifestation observed in 80% of cases. Associated symptoms were difficulty in lexical access during speech (n=3), paraphasia (n=3), echolalia (n=2), writing deficits (n=2), apraxia of speech (n=1), dysexecutive (n=1) and stuttering (n=1). Two patients (40%) referred preference for low nutritional foods and one patient (20%) evolved with mutism. Atypical parkinsonism is present in 40% of the cases, raising the possibility of PPA secondary to corticobasal syndrome. Magnetic resonance imaging (MRI) revealed a more pronounced volumetric reduction in the parietal (60% of cases) and frontotemporal (40% of cases) lobes, with a predominance on the left. Metabolic positron-emission tomography (FDG-PET) showed glycolytic hypometabolism compatible with metabolic pattern of frontotemporal dementia (n=3), corticobasal degeneration (n=1), and Alzheimer's disease (n=1). The hypothesized variants were semantic (n=3), nonfluent (n=1), and logopenic (n=1). Multidisciplinary intervention with speech therapy had great adherence in 80% of cases, and selective serotonin reuptake inhibitors (SSRIs) were prescribed for symptoms in 80% of cases. **Conclusion:** PPA is a diagnosis to be considered in middle adulthood and elderly patients with language impairment. The differential diagnosis among other cognitive disorders is subtle and may request imaging. Owing to PPA's insidious nature, the outcome is progressive cognitive decline with language predominance in all cases, associated with movement disorders in 40% and with mutual predominance of behavior disorders in another 40% of cases. These patients' identification allows the early institution of a multidisciplinary therapeutic plan, which aims to retard the progression of dementia and to improve patients' and caregivers' quality of life.

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Clinical and laboratory profile of thrombotic microangiopathy following gene therapy for 5q-spinal muscular atrophy in Brazil

Clara Gontijo Camelo[1], Rodrigo Holanda Mendonça[1], Cristiane de Araújo Martins Moreno[1], Adriana Banzzatto Ortega[2], Vanessa van der Linden[3], Rejane de Souza Macedo Campos[4], Helio van der Linden[5], Natalia Spinola Costa da Cunha[6], Janaina Monteiro Chaves[7], Edmar Zanoteli[8]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

[2]Hospital Pequeno Principe, Curitiba PR, Brazil.
[3]Raros, Recife PE, Brazil.
[4]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.
[5]Instituto de Neurologia de Goiânia, Goiânio GO, Brazil.
[6]Hospital da Criança de Brasília, Brasília DF, Brazil.
[7]Hospital Base do Distrito Federal, Brasília DF, Brazil.
[8]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Edmar Zanoteli; Email: edmar.zanoteli@usp.br.

Background: Spinal muscular atrophy linked to chromosome 5 (SMA-5q) is a progressive genetic disease caused by deletions or disease-causing variants in the survival motor neuron 1 (SMN1) gene. Onasemnogene abeparvovec is the first gene replacement therapy (GT) approved to treat SMA-5q. SMN1 gene transfer therapy through AAV9 aims to restore normal levels of functional SMN protein. More recently, new safety concerns have arisen, such as hepatotoxicity, thrombotic microangiopathy (TMA), and prolonged use of corticosteroids, underscoring the importance of continuous post-marketing drug safety monitoring. **Objective:** This study aimed to show the frequency of TMA in patients with SMA-5q treated with onasemnogene abeparvovec in a large multicenter Brazilian cohort and to describe the clinical and laboratory profile. **Methods:** This retrospective multicenter observational study evaluated all patients who developed TMA following treatment with onasemnogene abeparvovec for SMA-5q across Brazil from October 2020 to January 2024. Data were collected from medical records and from results presented at conferences (OPHELIA study). **Results:** Among 150 patients with SMA-5q treated in Brazil from October 2020 to January 2024, seven developed TMA. Six patients were type 1 SMA-5q, and one type 2 SMA-5q. The average age at the time of gene therapy administration was 20.8 months (range: 12 - 37 months), with an average weight of 11.4kg (range: 8.0 - 16.7kg). Notably, only one patient had a documented infection before gene therapy. No patient had received a vaccine within one month before treatment. All patients had antibody titers ≤1:50 before infusion. Patients exhibited the first symptoms of TMA between the first and second week post-infusion (mean 7.8 - range: 6-10 days). Thrombocytopenia in all patients was severe, with values below 50,000 (mean 17,000 - range: 5,000 to 31,000). Six patients exhibited both proteinuria and hypertension, and three experienced oliguria. Additionally, six patients showed increased creatinine levels, indicating signs of acute kidney injury. Five patients required treatment for TMA; two underwent plasmapheresis, and three patients had an increase in the corticosteroid dose to 2mg/kg/day. One patient treated with plasmapheresis developed hematuria, kidney and liver failure, and died. All other patients recovered after TMA. The occurrence of TMA was associated with prolonged corticosteroid use (mean 194 days - range: 70-390). No patient received Eculizumab or any other immunomodulator. **Conclusion:** TMA is a severe life-threatening side effect. Early identification and appropriate treatment are essential for a positive outcome.

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Clinical and molecular profile of Duchenne muscular dystrophy in a single center in Brazil

Karlla Danielle Ferreira Lima[1], Marco Antônio Veloso Albuquerque[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Karlla Danielle Ferreira Lima; Email: karlla.dfl@gmail.com.

Background: Duchenne muscular dystrophy (DMD), a disease with X-linked recessive inheritance pattern, is one of the most severe forms of inherited muscular dystrophies, caused by mutation in DMD gene and leading to progressive muscle fiber degeneration and weakness. Progressive weakness results in loss of ambulation early, and death from respiratory impairment or cardiac dysfunction. **Objective:** To evaluate the clinical profile of patients with DMD, followed from 2000 to 2024. **Methods:** This is a cross-sectional and retrospective study carried out at the Muscle Section of a reference center in Brazil. **Results:** The medical records of 143 patients were reviewed. Patients had a mean age of 11 years old (2-23 years old). The average age at onset of signs/symptoms was 3 years of age (ranging from 6 months to 9 years). Patients had their diagnosis confirmed at an average of 6 years of age (ranging from 6-16 years). The mean first baseline serum CPK was 12,936. The main signs/symptoms of initial manifestation were: falls (40%), motor developmental delays (15%) and change in gait (10%). 3% had language developmental delay as first symptoms and 5% had serum enzymes elevation as first manifestation even without symptoms. Regarding the molecular profile, 62% had deletion of 1 or more exons, 14% duplication of 1 or more exons, 10% nonsense variant, 10% frameshift and 4% at the splice site. Regarding corticosteroids, the average starting age was 6.9 years old; 70% were using deflazacort while 25% were using prednisolone/prednisone, and 5% were not using it or did not have data in the medical record. 33% of patients had already lost the ability to walk, with an average age of loss of ambulation of 9.6 years old. 39% of patients had neurodevelopmental disorder: 49 (32%) patients had some degree of intellectual disability, 21 (13%) had Attention Deficit Hyperactivity Disorder (ADHD), 10 (6%) autism spectrum disorder (ASD), 4 (2%) oppositional defiant disorder (ODD). 13% had more than one neurodevelopment disorder. Only two patients had epilepsy, one of which was associated with another genetic variant. **Conclusion:** The study demonstrated a profile of young patients, with 76% of patients presenting deletion or duplication of one or more exons, a molecular profile similar to that described in the literature. The average age of diagnosis was late compared to other studies, and the period between the onset of symptoms and molecular diagnosis was relatively long, reflecting the difficulty for physicians in recognizing the condition in its earliest stages. Finally, there was a higher prevalence of cognitive/neurodevelopmental disorders in this cohort.

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Clinical and molecular spectrum of TK2-deficiency: a large Brazilian cohort

Cristiane Araujo Martins Moreno[1], Mariana Cunha Artilheiro[1], André Macedo Serafim da Silva[1], Alulin Tácio Quadros Santos Monteiro Fonseca[1], Michelle Abdo Paiva[1], Clara Gontijo Camelo[1], Andre Luiz Santos Pessoa[2], Eduardo de Paula Estephan[1], Maria da Penha Ananias Morita[3], Anna Paula Paranhos Miranda Covaleski[4]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[2]Hospital Infantil Albert Sabin, Fortaleza CE, Brazil.
[3]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[4]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Cristiane Araujo Martins Moreno; Email: cristianeammoreno@gmail.com.

Background: Biallelic pathogenic variants at TK2 lead to a severe and progressive myopathy (TK2d). For a scenario with unspecific clinical findings, added to the possibility of supplementation therapy that changes the natural history of the disease, highlighting clinical features that increase suspicion and accelerate diagnosis is essential. **Methods:** Clinical (N=32) and genetic findings (N=36) of patients with TK2d from several centers across Brazil were identified and presented in this work. Genotype-phenotype correlation was performed for recurrent and novel variants. Motor and respiratory assessments were systematically performed in 10 patients. **Results:** Nine patients with the infantile form, 19 with childhood form and 5 with late-onset form were described. Extramuscular features were present in 30% of the cohort. Neuropathy and encephalopathy were clinically predominant for some patients. Four variants were recurrent, and one was novel, allowing genotype-phenotype correlations. T108M patients presented a milder presentation and slower progression when compared to the H121N group. R183T might be associated with peripheral nerve involvement and c.536_538+8del with encephalomyopathic involvement. A two-year follow-up of three patients showed decreased motor, bulbar, and respiratory function, compared to a dramatic improvement in treated patients. **Conclusion:** TK2d is a very debilitating disease. Early diagnosis is essential because a potential treatment can change the natural history of the disease. High diagnostic suspicion is imperative, and extramuscular involvement might play an important role for diagnostic strategies, starting with the inclusion of the TK2d in the differential diagnosis for neuropathies and epilepsy

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Clinical and motor functional evaluation of patients with bi-allelic disease-causing variants in SMN1 gene and two copies of SMN2 treated in the pre-symptomatic stage

Raquel Diógenes Alencar Sindeaux[1], Rodrigo de Holanda Mendonça[1], Joemir Jabson da Conceição Brito[1], José Pedro Baima[1], Graziela Jorge Polido[1], Eduardo Vital de Carvalho[1], Leandro Augusto de Almeida[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.

Correspondence: Eduardo Vital de Carvalho; Email: duvital1983@gmail.com.

Background: Spinal Muscular Atrophy linked to chromosome 5 (SMA-5q) is a rare genetic disorder that leads to progressive muscle weakness and atrophy. Patients with SMA-5q can be diagnosed in a pre-symptomatic phase through newborn screening or when there is a sibling with SMA-5q. Early diagnosis enables prompt intervention and improves patient outcomes. **Objective:** To describe the experience of a specialized neuromuscular center in the treatment of pre-symptomatic SMA-5 patients and to analyze the clinical aspects during a follow-up period up to 57 months (range 1-57; average 16.3 months). **Methods:** Six SMA-5q patients started treatment in a pre-symptomatic phase and have been monitored in a neuromuscular center specialized in motor neuron diseases, from August 2019 to May 2024. Neurological examination, motor scale and respiratory assessments have been performed by a multidisciplinary team at the diagnosis, every 3 months during the first year of follow-up and every 6 months thereafter. Information on adverse effects have also been collected. **Results:** All six pre-symptomatic patients had homozygous deletion in SMN1 gene and 2 copies of SMN2. Four patients were diagnosed by newborn screening and have been followed from 1 to 10 months old (mean 5.75 months). They started risdiplam between 25 and 29 days of age (mean 26.75 days). One of them started risdiplam recently and remains asymptomatic at 2 months old, but still does not have a comparative motor scale. The other three risdiplam-treated patients achieved maximum score on Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) at 4 months old. All risdiplam-treated patients have adequate development for their age, but two of them switched treatment to zolgensma at 6 and 9 months old because it is a single dose therapy. Two patients were younger siblings of SMA-5q patients and had an intrauterine diagnosis. They started nusinersen between 2 and 7 days old (mean

4.5 days) and have been followed from 18 to 57 months (mean 37.5 months). They both changed treatment to zolgensma because they were experiencing weakness in their limbs. One of them received zolgensma at 5 months old, reached maximum CHOP INTEND score at 6 months old and started walking at 1 year and 4 months old. The other patient received zolgensma at 1 year and 6 months old, reached maximum CHOP INTEND score at 1 year old and started walking at 1 year and 10 months old. None of the patients presented dysphagia or need for ventilatory support. Patients who received zolgensma reported these adverse effects: one had fever and vomiting, one thrombocytopenia and two elevated hepatic transaminases. One patient who received risdiplam had anemia. **Conclusion:** The positive outcomes observed in our cohort underscore the importance of early diagnosis and intervention. By demonstrating consistency with clinical trials, our findings further support the efficacy of current treatment approaches for SMA-5q.

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Clinical and neuroimaging features of corticobasal degeneration

Carolina Rocha da Silveira Brügger Cardoso[1], Bruna Andrade de Oliveira[1], Natália da Costa Chiote Pinheiro[1], Luiz Felipe Vasconcellos[1]

[1]Universidade Federal do Rio de Janeiro, Instituto de Neurologia Deolindo Couto, Rio de Janeiro RJ, Brazil.

Correspondence: Carolina Rocha da Silveira Brügger Cardoso; Email: carolinarbrugger@gmail.com.

Case presentation: Case 1: 81-year-old man started 3 years ago with rigidity in right lower limb leading to progressive impairment of walking with falls within 6 months and wheelchair bound in 1 year. Later he also presented rigidity of right upper limb. Concomitantly, he presented cognitive impairment and bradyphrenia. Neurological exam: right side bradykinesia, global rigidity, slow velocity of vertical saccades and involuntary elevation of the right arm (alien limb phenomenon). MRI: bilateral hippocampal atrophy. Case 2: 59-year-old woman started at the age of 54 memory impairment, executive dysfunction and motor apraxia. Physical exam: gait apraxia, global rigidity, hyperreflexia with Babinski and Hoffman signs in the right side, resting tremor in the right upper limb, polyminimyoclonus, severely cognitive impairment with global aphasia, slow velocity of vertical saccades and right alien limb phenomenon. MRI: asymmetric frontoparietal atrophy, well-marked on the left. Case 3: 58-year-old woman started with dyscalculia at 54 years, evolving with progressive aphasia, cognitive impairment and gait apraxia. Neurological exam: gait apraxia, global rigidity, myoclonus, dystonic posture of the trunk and upper limbs and severe cognitive impairment. MRI: asymmetric frontoparietal and hippocampal atrophy. Case 4: 68-year-old woman presented with gait impairment and postural instability. The symptoms started at the age of 65 with dystonic movement of the right hand evolving to ipsilateral tremor and motor apraxia. Physical exam: parkinsonian gait, myoclonus, motor apraxia, global aphasia, dysphagia, right alien limb phenomenon. MRI: global parenchymal atrophy, worse on left frontoparietal cortex. **Discussion:** Corticobasal degeneration (DCB) is a rare cause of atypical parkinsonism (1). The terms "corticobasal syndrome" (CBS) and "corticobasal degeneration" (CBD) represent distinct conditions. The former denotes the clinical phenotype, whereas CBD can only be confirmed by postmortem anatomopathological analysis (2). Classically, motor symptoms occur asymmetrically and include akinetic-rigid parkinsonism, dystonia, myoclonus and the alien limb phenomenon. Cognitive symptoms include apraxia, aphasia, cortical sensory deficits (2). In neuroimaging, magnetic resonance imaging typically reveals asymmetric cortical atrophy, predominating on the posterior frontal and parietal cortex (3). However, this pattern is observed around 50% of cases, depending mainly on disease stage. **Final comments:** In this report, we present four cases of CBS/CBD, through its clinical evolution with motor and cognitive findings, describing aspects of neuroimaging. The definition of CBD is only possible by pathological analysis, but neuroimaging could be a useful tool for differential diagnosis.

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Clinical and neurophysiological characterization of patients with ganglionopathies: case series

Andreia Lira de Oliveira[1], Pablo Vinícius Silveira Feitoza[1], Luiziane Maria da Silva Alves[1], Tiago Breno de Farias Brito[1], João Victor de Chagas Rossin[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Andreia Lira de Oliveira; Email: andreialira.med@gmail.com.

Background: Neuronopathies or ganglionopathies constitute a group of relatively rare sensory neuropathies, resulting from involvement of the dorsal root ganglion. This condition was first described in 1948 and has a distinct clinical presentation from other sensory neuropathies, mostly affecting deep sensitivity with a non-length-dependent pattern. It typically manifests with sensory ataxia and areflexia. **Objective:** To characterize clinically, epidemiologically and neurophisiologically the cases of ganglionopathies observed in our service. **Methods:** A retrospective study was conducted by reviewing the medical records of ganglionopathy cases treated between 2018 and 2024. **Results:** 10 diagnosed cases were identified with ganglionopathy during the study period. The average age of the patients was 59.8 years, with a predominance among women (60%) and symptom onset within 5 years in 70%. Disease presentation was chronic in 90% of cases, initially affecting lower limbs in 70% of cases, although with disease progression, 90% of patients eventually presented involvement of both upper and lower limbs. Half of the patients reported paresthesia as the initial symptom, while others reported ataxia (30%) and pain (20%). Only 30% of patients had pure deep sensitivity impairment, even though all of them predominantly exhibited this type of sensory deficit. Two patients presented with associated cerebellar involvement, 90% of cases presented with hyporeflexia or areflexia, mainly in the lower limbs. Etiologically, half of the cases remained idiopathic, while 2 cases were associated with Sjogren's syndrome, 2 cases with Paraneoplastic syndrome and 1 case with vitamin B12 deficiency. The majority of cases (90%) were treated with immunosuppressive therapy or vitamin b12 supplementation, but only 5 patients reported symptom improvement after treatment, mostly ataxia improvement. Among the 5 cases that showed symptom improvement, 4 patients (80%) received pulse therapy with methylprednisolone. Regarding electroneuromyography, there was a predominance of asymmetric and severe axonal impairment of nerves, with 60% of cases characterized as sensory axonal neuropathy and 20% as sensory-motor axonal neuropathy. **Conclusion:** Our aim is to contribute to the characterization of this relatively rare and understudied condition, encouraging further research in this area, especially its etiology with the identification of autoantibodies potentially associated with this condition, as well as uncertainties regarding appropriate therapeutic choices.

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Clinical and neurophysiological epidemiology of brain death

Tarcísio Nunes Alvarenga[1], José Eduardo Trentim Longo[1], Júlio César Moreira[1], Marina Carvalho Acosta Cleto[1], Thalita Luiza Madoglio[1], Laércio Martins de Stefano[1], Lilian Audi Goulart[1], Rebeca Aranha Barbosa Sousa[1], Yasmim Nadime José Frigo[1], Luiz Eduardo Gomes Garcia Betting[1]

[1]Universidade Estadual Paulista, São Paulo SP, Brazil

Correspondence: Tarcisio Nunes Alvarenga; Email: tarcisionalvarenga@gmail.com.

Background: Brain death (BD) is the complete and irreversible loss of brain functions. EEG is a diagnostic test that is low-cost and relatively accessible. In Brazil, the estimated rate of BD is 90-100 cases per million people per year. **Objective:** This study is an epidemiological survey and neurophysiological results of individuals with suspected BD in the region of the Regional Health Department (DRS-VI). **Methods:** Retrospective study reviewing EEG reports of all BD protocols opened in DRS-VI over a 1-year period. This region comprises 68 municipalities with approximately 1.8 million people. All tests are performed by the OPO team at the Hospital das Clínicas, Faculty of Medicine of Botucatu - UNESP. After a compatible clinical analysis, the complementary test of choice is the EEG. This test is performed according to SBNC recommendations derived from ACNS. Basically, tests are performed with video-EEG recordings, electrode placement according to the international 10-20 system, and lasting 30 minutes including 2 painful stimuli. **Results:** 107 BD protocols were opened. The estimated incidence was 57 cases per million people. The individuals had a mean age of 48±18 years (min=10 – max=86, 46 women). Regarding etiology: 61% had stroke (n=8 ischemic stroke, n=48 hemorrhagic stroke, and n=9 unspecified), 22% traumatic brain injury, 7% ischemic encephalopathies, 5% neoplasms, 4% neuroinfections, and 2% others. The EEG was compatible with the diagnosis in 90% of cases. In 11 individuals, the test needed to be repeated. In these cases, it was confirmatory in 8 cases, remained inconclusive in 2, and in only 1 patient presented cerebral electrical activity. **Conclusion:** The incidence of BD in the region is lower than the estimated national rate. EEG helped to rule out one case in 107 and was inconclusive in 2. These findings confirm its importance in the protocol currently used in the country.

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Clinical and polysomnographic characteristics associated to excessive daytime sleepiness in obstructive sleep apnea

Eder Leandro da Silva Dantas[1], Eduardo Cordeiro Vitor Martins[1], Fernando Gustavo Stelzer[1], Alan Luiz Eckeli[1]

[1]Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.

Correspondence: Eder Leandro da Silva Dantas; Email: ederleandro.sdantas@gmail.com.

Background: Obstructive sleep apnea (OSA) is characterized by recurrent episodes of reduction or cessation of upper airway airflow during sleep. One of the most frequent symptoms related to OSA is excessive daytime sleepiness (EDS), with prevalence ranging from 35 to 87%. EDS has elevated morbidity, being related, for example, to increased risk of motor vehicle and work-related accidents and impaired cognitive and work performance. However, there are few studies evaluating epidemiology and polysomnographic characteristics of EDS among individuals with OSA. **Objective:** To evaluate prevalence of EDS and to assess the association of clinical and polysomnographic characteristics of EDS among adult patients with OSA. **Methods:** This was a cross-sectional study, including adult individuals referred for diagnostic polysomnography (PSG) between March, 2014 and

March, 2015. The main inclusion criteria were confirmed OSA diagnosis and age ≥18 years old. Exclusion criteria were participation refusal, age <18 years old, moderate to severe depression symptoms, chronic use of sedative-hypnotic drugs, and cognitive impairment. All participants provided written informed consent forms. EDS was assessed by Epworth Sleepiness Scale (ESS) and defined as a score >10. Depressive and anxious symptoms were evaluated through Beck Depression Inventory and Beck Anxiety Inventory, respectively. All participants were submitted to full-night polysomnography. **Results:** Data from 392 participants was analyzed. EDS prevalence was 45.7%, being more prevalent in men and in the economically active group. There was a positive correlation between body mass index and ESS score and there was a negative correlation between ESS score and age. Prevalence of depressive and anxious symptoms was higher in the EDS-group. EDS was more prevalent in severe OSA. Regarding polysomnographic findings, EDS was associated with shorter sleep duration and REM sleep latency and greater apnea-hypopnea index (AHI), sleep efficiency, sleep fragmentation, and total sleep time in comparison to non-EDS ones. EDS was not associated to oxygen saturation parameters. **Conclusion:** Prevalence of EDS in OSA patients was within mid-range from previous studies. There is a higher prevalence of EDS between economically active young men. Besides, our findings suggest that sleep fragmentation due to respiratory events in OSA might play a role in EDS pathophysiology.

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Clinical and radiological improvement of oligodendrocyte myelin syndrome with combined treatment of rituximab and plasmapheresis

Maria Luiza Levindo Coelho Martinis[1], Bárbara Luisa Coraspe Gonçalves[1], Luiz Guilherme Gerhardt Fernandes[1], Marina Brasileiro Cesar Leitão[1], Bárbara França Amorim[1], Osvaldo Pantoja de Oliveira[1], Luiz Fernando Guimarães[1], Ana Carolina Andorinho de Freitas Ferreira[1], Cristiane Borges Patroclo[1], Marcelo Cagy[1]

[1]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.

Correspondence: Maria Luiza Levindo Coelho Martinis; Email: mlevindocoelho@gmail.com.

Case presentation: Female patient, 22 years old, started in 2015 with paresthesia in the left lower limb. At time she didn't have her symptoms evaluated by a neurologist. Her symptoms were considered to be of psychogenic origin, and she was treated with an anxiolytic drug. In 2019, the patient developed bilateral optic neuritis (ON) and was referred to a neurology service. She underwent magnetic resonance imaging (MRI) which showed several demyelinating lesions, as well as a positive oligoclonal band. Her optical coherence tomography showed bilateral changes. She underwent intravenous methylprednisolone 1000mg per day for five days, with slight clinical improvement, repeating the treatment after a month. The patient was initially diagnosed with multiple sclerosis (MS). Interferon was started and, despite this, progressed to recurrent optic neuritis outbreaks and sensory and motor symptoms. The new MRI shows disseminated demyelinating lesions in the spinal cord. Antibody - aquaporin-4 (anti-aqp4) and myelin oligodendrocyte glycoprotein andibody (anti-MOG) research was performed (cell base assay method) and showed a negative anti-aqp4 antibody and positive anti-MOG. The patient, while using interferon, developed tetraparesis, spasticity in the lower limbs, worsening gait, requiring a wheelchair. After confirming the positivity of the anti-MOG, the patient underwent plasmapheresis (twice a year) and the use of rituximab, showing significant improvement in his gait (walking more than 100 m with a walker) and reducing spasticity after one year of treatment. **Discussion:** Myelin oligodendrocyte glycoprotein (MOG) is present on the surface of oligodendrocytes and the myelin sheath of the nervous system and the MOG antibody-associated disease (MOGAD) is the most recently studied inflammatory demyelinating disease of the central nervous system. The presence of MOG antibodies was found in a small portion of a group of patients with multiple sclerosis, with a more severe disease course, with higher rates of recurrence and no therapeutic response to disease-modifying therapies. Typical symptoms of MOGAD include ON and myelitis, which overlap with MS and neuromyelitis optica (NMO). In profile of the patient, MOGAD has less sex or age predilection than MS or NMO. The above patient presented reduced clinical and radiological activity with concomitant treatment with rituximab and plasmapheresis **Final comments:** The case presented in this article shows a female patient that had the MS diagnosis but the latest investigation showed anti-mog positive. Thus, the anti-MOG research shows it importance in cases of MS with non responsive treatment. In Brazil we have limited access to this type of exam, making diagnostic confirmation difficult. There is no specific medication to treat this disease. Treatment with rituximab and plasmapheresis for the patient resulted in clinical and radiological improvement.

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Clinical and ultrasonographic evaluation of dysphagia in patients with LAMA2-CMD

Clara Gontijo Camelo[1], Pedro Henrique Marte de Arruda Sampaio[1], Ana Lucila Moreira[1], Cristiane de Araujo Martins Moreno[1], Mariana Cunha Artilheiro[1], André Macedo Serafim da Silva[1], Umbertina Conti Reed[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Clara Gontijo Camelo; Email: claragc@gmail.com.

Background: LAMA2-CMD is a rare autosomal recessive disease characterized by hypotonia, muscle weakness, and the inability to achieve independent gait. It imposes a considerable disease burden, including difficulty in weight gain and dysphagia. **Objective:** However, this disease burden has not been thoroughly assessed in the medical literature, and this study aims to address this gap. **Methods:** Patients with genetically confirmed LAMA2-CMD were cross-sectionally evaluated. Clinical assessments included analysis of dysphagia, NdSss scale, weight measurement, and the necessity of gastrostomy. Patients received ultrasound evaluations of tongue muscles, and the muscle abnormalities were classified using the modified Heckmatt scale. The ultrasound M mode assessed the hyoid elevation movement during swallowing. Qualitative analysis of ultrasound findings and correlations with clinical presentations were conducted. **Results:** 20 patients with LAMA2-CMD were evaluated, ranging from 3-27 years old (mean 7.7). 8/20 patients (40%) weighted below the 3rd percentile. All patients experienced some degree of dysphagia, with a maximum score of 7 on the NdSss scale. 17/20 patients (85%) reported taking over 30 minutes to finish a meal. 4/20 patients (20%) had gastrostomy tubes. The geniohyoid muscle was the most affected among the three tongue muscles evaluated. 14/ 20 patients had markedly hyperechoic muscle. The genioglossus muscle was the second most affected, with 9/20 patients having markedly hyperechoic muscle. None of the patients had normal geniohyoid or genioglossus muscles. Lastly, the least affected muscle was the digastric, with 7/20 patients having markedly hyperechoic muscle. 16/20 patients required an increased number of hyoid elevation movements and had low amplitude movements during swallowing. A correlation was observed between the patient's age and the severity of ultrasound abnormalities. The genioglossus and geniohyoid muscles appeared more preserved in patients under 5 years old than older patients (p=0.005 and p=0.017, respectively). Similarly, there was a correlation between extremely low weight (p<3) and markedly hyperechoic genioglossus muscle (p=0.0176). **Conclusion:** Patients with LAMA2-CMD experience greater difficulty in weight gain and exhibit some degree of dysphagia. Assessing tongue muscles via ultrasound is a valuable tool for comprehending functional abnormalities in these patients, aiding in clinical management, and potentially serving as a marker for disease progression

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Clinical challenges and multidisciplinary management in Takayasu's arteritis: a case report

Rhanice Soares Huguenin Câmara[1], Bruno Castelo Branco Rodrigues[1], Iuri Sena Melo[1], Júlio César Iaia[1], Tatiane Thaís Santos Silva[1], Sophia Keiko Inuzuka[1], Maria Fernanda de Abreu Rodrigues[1], Roberto Pereira Santos[1], Ana Lucia Zuma de Rosso[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Rhanice Soares Huguenin Câmara; Email: rhaniceshcamara@gmail.com.

Case presentation: A 43-year-old female patient presented with an ischemic stroke (CVA) in September 2020, resulting in right hemiplegia and aphasia. CT angiography revealed severe bilateral carotid lesions, unusual in extent given hypertension as the only comorbidity. Suspecting Takayasu's arteritis, we requested ESR, which was elevated, confirming the diagnosis. After starting corticosteroid treatment, ESR levels reduced to 6 mm and are now at 3 mm. The patient is on continuous treatment with prednisone (5mg/day), azathioprine (100mg/day), aspirin (100mg/day), and phenytoin (100mg every 8hours). She reports anxiety and anhedonia. Recent lab results show platelets at 199,000, hemoglobin at 14.9g/dL, LDL at 95mg/dL, and ESR at 3mm. Clinically, she presents with hypertension (BP 137/105mmHg), tachycardia (HR 112bpm), hemiplegic gait, grade 4 paresis, mild spasticity in the right upper limb, hyperreflexia, right-sided hypoesthesia, positive Hoffman sign on the right, and central paralysis of the right seventh cranial nerve. **Discussion:** This clinical case highlights the complexity of managing a patient with Takayasu's arteritis, a rare disease affecting large arteries and potentially leading to severe complications like ischemic stroke. The reduction in ESR levels after starting corticosteroids indicates inflammatory control. However, significant stroke sequelae persist, including hemiplegia, aphasia, and neurological signs (hyperreflexia, hypoesthesia, and Hoffman sign), while hypertension and tachycardia require careful management to prevent further complications. Treatment includes prednisone and azathioprine as immunosuppressants, aspirin for thromboembolic event prevention, and phenytoin for seizure prevention. Anxiety and anhedonia underscore the importance of psychological support, making a multidisciplinary approach essential. **Final comments:** This case underscores the complexity of Takayasu's arteritis. The multidisciplinary therapeutic approach has proven effective in reducing inflammatory markers and stabilizing the patient clinically. However, neurological sequelae persist, impacting the patient's quality of life and functional capacity, highlighting the importance of early diagnosis. Thus, it is crucial to identify the disease before complications arise and to conduct longitudinal studies on the progression of Takayasu's arteritis in patients treated with different therapeutic regimens to determine long-term efficacy and side effects. Sharing this clinical experience aims to enhance the understanding of Takayasu's arteritis and serve as a tool for students and healthcare professionals.

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Clinical characteristics of patients with positive genetic testing for peripheral neuropathies at a tertiary Brazilian neuromuscular outpatient clinic in an university hospital

Carolina Braga Moura[1], Ana Luiza Morgado Costa[1], Gabriel Thomaz Candido[1], Camila Castelo Branco Pupe[1], Osvaldo Jose Moreira Do Nascimento[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Ana Luiza Morgado Costa; Email: anamorgado@id.uff.br.

Objective: To determine prevalent clinical and epidemiological characteristics among patients with positive genetic testing for peripheral neuropathies. Background: Diagnosing peripheral neuropathies frequently necessitates genetic testing. Yet, there is no established protocol to guide neurologists in discerning the appropriate circumstances for requesting genetic testing in a brazilian population. **Methods:** Seventy-one patients were tested at an outpatient clinic for neuromuscular diseases in Brazil. Only patients with positive genetic tests were included. The prevalence of mutations was registered. Clinical characteristics were retrieved from electronic records (i.e., symptoms, physical exam, and nerve conduction study). Descriptive analyses were conducted. **Results:** Thirty patients had a positive genetic test. All patients had a Charcot-Marie-Tooth (CMT) disease phenotype. The mean age of diagnosis was 46 years old. Most patients had a mutation in the PMP22 (9/30, 30%), ALMS1 (4/30, 13%), and SH3TC2 (3/30, 10%) genes. Of the nine patients with PMP22 mutation, six (66%) presented numbness, six (66%) had imbalance, and five (55%) presented weakness. Pes cavus and distal atrophy were each reported by two patients (2/9, 22%). **Conclusion:** Our study highlights key clinical characteristics among patients with positive genetic testing for hereditary neuropathies. We found CMT phenotype was the most prevalent among study participants, with numbness, imbalance, and weakness being recurrent clinical manifestations. Notably, most patients only disclosed diagnosis later in adulthood. This result emphasizes the need to rapidly identify prevalent mutations, symptoms, and associated conditions to order genetic testing for hereditary neuropathies.

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Clinical evaluation of dysphagia as a prognosis factor in stroke patients: preliminary results

Victor Vale da Silva[1], Joanna Sousa da Fonseca Santana[1] , Amanda Hellen Sousa Leal[1], Ana Clara de Lemos Guimarães[1], Carla Loureiro Alvarez[1], Catarina Cavalcanti da Silva[1], Brena Lima Cunha[1], Felipe Oliveira Costa, Júlia Ribeiro Oliveira Campos[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Afya Faculdade de Ciências Médicas de Itabuna, Itabuna BA, Brazil.

Correspondence: Victor Vale da Silva; Email: vitor.futebolclube@hotmail.com.

Background: Dysphagia is a complication of stroke that frequently affects patients, with diagnosis occurring in 40-70% of cases within the first 3 days. This condition can prolong hospitalization, increase the risk of mortality and increase healthcare costs. Furthermore, some patients will experience prolonged dysphagia, which increases risks such as pneumonia, dehydration and reduces quality of life. Therefore, it is essential to clinically evaluate dysphagia as a prognostic factor in stroke patients, considering the severity of dysphagia, in order to improve medical care for these patients. **Objective:** To analyze dysphagia as a prognostic factor in patients who have suffered a stroke. **Methods:** This work is an observational and retrospective cohort study, descriptive and was based on the partial collection of secondary data from electronic medical records of patients previously admitted to a reference hospital in the city of Salvador, Bahia. The study population consists of patients diagnosed with stroke, aged 18 years or older, who were admitted to the hospital between January 2021 and December 2022. Variables of interest include: sex, age, date of admission, infection, length of hospital stay, complications and mortality. Excel and SPSS software were used for data analysis. The study was approved by the Research Ethics Committee. **Results:** To date, the study sample size is 178 patients and, among them, 86 (48.3%) patients presented dysphagia. Of the total number of patients, 128 were aged >60 years (median 75 years), with 68 (53.1%) having dysphagia (43 women [63.2%] and 25 men [36.8%]). Regarding the number of patients who cont racted infection during hospitalization: 7 (25%) of patients with severe dysphagia developed infection, 5 (18.5%) of patients with moderate dysphagia developed infection and 7 (22.6%) of patients with mild dysphagia developed infection. And in relation to length of stay, patients with dysphagia had an average of 20.4 days of hospitalization (standard deviation 21.4) and a maximum length of stay of 150 days. Patients without dysphagia had an average of 11.5 days (standard deviation of 11.9) and a maximum hospitalization of 66 days. **Conclusion:** It is clear that dysphagia is a prevalent and significant condition in patients who have suffered a stroke, considerably impacting the prognosis of these individuals. The majority of stroke patients evaluated had some degree of dysphagia, significantly increasing hospitalization time, with more severe cases being associated with a higher incidence of infections during hospitalization. Therefore, early identification and appropriate treatment of dysphagia are essential to optimize healthcare and clinical outcomes for these patients.

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Clinical factors associated with constipation in patients with Parkinson's disease: a Brazilian cross-sectional study in Bahia

Gabriel Teles de Oliveira Piñeiro[1], Maria Eduarda Messias Vassoler[1], Ayla Morais Vitório Guerra[1], Gabriel Souza Medrado Nunes[1], Fernanda Garcia Silva Pereira Nascimento[1], Caio Fontes do Nascimento[1], Roberta Borges Gomes Kauark[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Ayla Morais Vitório Guerra; Email: aylaguerra1@hotmail.com.

Background: Constipation is a well-known non-motor symptom that can occur during Parkinson's disease (PD). However, there is still a lack of studies exploring the associations between constipation and other commonly observed characteristics of PD patients. **Objective:** Investigate clinical characteristics of Parkinson's disease patients and their association with constipation at a reference center for movement disorders. **Methods:** Cross-sectional study carried out between 2023-2024, patients who reported defecating less than three times per week were classified as constipated. Clinical, social, and epidemiological data were collected, in addition to the application of the following scales: Unified Parkinsons Disease Rating Scale-Part III (UPDRS-III) for severity of motor symptoms, Beck Depression Inventory (BDI) for depression assessment, and the Hoehn and Yahr scale (HY) for functional disability. PD subtypes used were Tremor Dominant (TD), Akinetic-Rigid (AR), and Mixed, calculated using the UPDRS. Kolmogorov-Smirnov was performed to assess the normality of the data, Chi-square, Mann-Whitney, and T-test for correlation. **Results:** The study included 70 PD patients, of whom 67.1% were men, with a mean age of 65.45 (±9.28) years. Regarding clinical status, the mean disease duration was 9.60 (±5.16) years, and the severity of motor symptoms, as assessed by UPDRS III, had a mean score of 43.46 (±16.12). The median BDI score and Levodopa Equivalent Daily Dose (LEDD) were 13 (IQR: 4-20.75) and 1000 (IQR: 600-1460), respectively. Among all patients, 69 (98.6%) were using Levodopa and only 4 (5.7%) were using anticholinergics. The most frequent subtype was AR (45.7%), followed by TD (18.6%) and Mixed (8.6%). Constipation was identified in 19 (27.1%) patients, with a mean age of 66.9 (±8.5) years, and its presence was significantly associated with higher UPDRS III (p=0.026). Furthermore, the Mixed subtype was more associated with constipation than AR, while a negative association between TD and constipation was observed (p=0.001). There were no associations between constipation and disease duration LEDD, HY, or BDI (p>0.05). **Conclusion:** Constipation was found to be associated with more severe motor symptoms and Parkinson's subtypes, particularly with the Mixed form, but no correlations were observed with other clinical factors. However, further studies are necessary to better understand this relationship.

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Clinical features and quality of life of patients with amyotrophic lateral sclerosis at a specialized care service in Salvador-BA

Marie Lidio dos Santos Galvão Ribeiro[1], Maria Rodrigues Lima de Souza Pessoa[1], Maria Luiza Pereira Falconery[1], Stefania Zingone Andrade Carvalho[1], Marilaine Matos de Menezes Ferreira[1], Marcela Câmara Machado-Costa[1], Têmis Maria Félix[2], Lorena Alves dos Santos Pereira[1], Luana Souza Vasconcelos[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Marie Lidio dos Santos Galvão Ribeiro; Email: marielidiogr@gmail.com.

Background: Amyotrophic lateral sclerosis (ALS) is a rare and progressive neurodegenerative disease that affects motor neurons in the spinal cord, brainstem, and motor cortex. Often misdiagnosed or undiagnosed until its later stages, there are only palliative treatments, leading to a fatal prognosis. Consequently, it is a condition that burdens healthcare systems worldwide, causing loss of productivity and quality of life for patients and their caregivers. In Brazil, there is a lack of research on its clinical profile and epidemiology, which makes it challenging to analyze the disease in the country. **Objective:** To describe the clinical features and quality of life of ALS patients treated at a specialized care service in Salvador, Bahia. **Methods:** This was a cross-sectional observational study involving primary data collection. Patients with a diagnosis of definite ALS (by El Escorial criteria) followed up at the service from February 1, 2022, to July 31, 2023, were included. The following data was collected: gender, age, race, city of residence, disease stage (by Kings College staging), clinical phenotype, onset symptom location, main clinical signs, and complications. Furthermore, a series of questionnaires were applied, including the ALSFRS-R, EQ-5D, and WPAI-GH. The variables were summarized and subjected to descriptive statistical analysis. **Results:** A total of 28 patients were included in the study, with a mean age (SD) of 57 years (12). Most patients were male (64.29%) and brown (64.29%). Salvador represents the primary city of residence for 46.43% of patients. Spinal ALS was present in 78.57% of patients, with initial symptoms occurring more frequently in the upper limbs (60.71%). The most frequently reported symptoms were pain (64.29%), fasciculations (57.14%), and dysphagia (57.14%). Although 57.14% of the population was at stage 4, 75% of the patients

did not use any feeding device, and only 35.71% used non-invasive ventilation. Moreover, only 25% of patients underwent gastrostomy, while 3.57% underwent tracheostomy. The functionality score (ALSFR-R), utility value (EQ-5D), and productivity score (WPAI-GH) had a mean value (SD) of 20.39 (9.85), 0.178 (0.211) and 93.21 (14.92), respectively. **Conclusion:** The ALS profile observed in this study is consistent with that reported in the scientific literature: it shows a male predominance, with greater involvement from the 5th decade of life onwards and an urban setting. Spinal ALS is the main clinical phenotype, and the ethnic profile differed from that presented in the literature due to miscegenation. The population exhibits advanced ALS progression, with significant functional impairment, a wide range of associated symptoms, and a greater dependence on caregivers and family members. Quality of life declines as the disease progresses, but anxiety and depression are the least reported symptoms, suggesting effective psychological coping mechanisms to manage the diseases progression.

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Clinical features on Friedreich's ataxia patients in São Paulo, Brazil

Daiana Suelen Machado[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Daiana Suelen Machado; Email: daianamachado@gmail.com.

Background: There are few reports characterizing Friedreich´s Ataxia (FRDA) phenotype beyond Europe and USA. **Objective:** Herein, we describe the clinical profile of a representative FRDA cohort from São Paulo, Brazil. **Methods:** Patients were invited to answer an online survey comprising 24 to 32 questions, which was sent by physicians and the Brazilian Association of Acquired and Hereditary Ataxias via email and releases. We collected data on sex, current age, age at diagnosis, affected relatives, parental consanguinity, ability to walk, the need of ambulatory assistive device or wheelchair and presence of diabetes, cardiomyopathy, and scoliosis. Descriptive statistics was used to report the results - means, proportions, and Standard Deviations (SD). Age at loss of ambulation (LOA) was assessed both for classical and late-onset (LOFA) groups using Kaplan-Meier curves. **Results:** We gathered 104 answers from FRDA patients or legal guardians, being 79 (75.9%) patients with classical FRDA and 25 (24%) with late-onset (LOFA) . There were 64 (61.5%) women and 40 (38.5%) men. Mean age and age at diagnosis were 37.3±13.8 and 24.1±11.5 years, respectively. Parental consanguinity and familial disease recurrence were seen in 24 (23%) and 50 (48.7%) of the patients, respectively. Fifty five out of 104 patients lost ambulation at the mean age of 29.4±11.5 years. Those who are still ambulatory but need assistive devices, first required them at a mean age of 27.1±13.1 years. As expected, patients with classical FRDA had LOA earlier than LOFA (25.5±7.7 vs 47.8±10.2 years). The frequency of diabetes, cardiomyopathy, hearing impairment and scoliosis in this cohort was 36±10.9, 23.6±14.2, 32.5±8.8 and 18.5±9.1, respectively. **Conclusion:** The overall phenotype of FRDA in Brazil is similar to previous European and US reports, but age at LOA was slightly higher. Further studies are needed to better understand such difference.

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Clinical outcomes and safety of repetitive transcranial magnetic stimulation in the treatment of Alzheimer's disease: a scoping review

Jéssica Yumi Lago Matsuda[1], Liria Matsuzawa Figueiredo[1], Thelma Yuri Lago Matsuda[1]

[1]Faculdades Pequeno Príncipe, Curitiba PR, Brazil.

Correspondence: Jéssica Yumi Lago Matsuda; Email: jlagomatsuda@gmail.com.

Background: Alzheimer's disease (AD) is a chronic, progressive neurodegenerative disorder and accounts for half to 75% of all dementia cases. Considering that current treatments using pharmacotherapies, such as acetylcholinesterase inhibitors, still present limitations in improving cognitive performance, the investigation of alternate therapeutic methods is needed. In this context, repetitive Transcranial Magnetic Stimulation (rTMS) is emerging as a non-invasive promising therapy for AD. It uses electromagnetic induction to generate a pulsed magnetic field, which can stimulate the brain and modulate cortical excitability and neural activity. **Objective:** To identify in the literature about the clinical outcomes and safety of repetitive transcranial magnetic stimulation in the treatment of patients with AD. **Methods:** A scoping review was conducted based on the Preferred Reporting Items for Systematic reviews and Meta-Analyses extension for Scoping Reviews (PRISMA-ScR) statement and was duly registered on the Open Science Framework, via the link: https://osf.io/8y6r3/. Searches were selected from PubMed, ScienceDirect and Biblioteca Virtual em Saúde (BVS) databases and the descriptors "Alzheimer's disease" and "repetitive transcranial magnetic stimulation" were used. Inclusion criteria involved: studies published in the last 5 years, studies involving humans and written in english. **Results:** In total, 406 studies were identified, of which 17 were included in the review. 14 studies were randomized controlled trials involving a total of 550 patients who were randomly divided into two groups to either receive real rTMS of the dorsolateral prefrontal cortex (DLPFC) or sham stimulation. The protocols varied in the duration of treatment, from 2 to 24 weeks and the frequency of stimulation, from 10 Hz to 20 Hz. The outcomes were evaluated by score changes in the Alzheimer's Disease Assessment Scale (ADAS-cog) and Mini-Mental State Examination (MMSE). The results indicated that, compared to the sham group, the patients who received rTMS scored significantly higher on the MMSE and showed notably better performances on the ADAS-cog measures. Moreover, a recent meta-analysis analyzed the clinical advantages and safety of rTMS treatment over current pharmacotherapy. 15, 548 individuals with AD in 57 randomized clinical trials were included in the study. Compared with multiple pharmacological interventions, rTMS was more effective in improving global cognition, had the highest likelihood to improve MMSE score and showed notably fewer adverse effects. **Conclusion:** Studies demonstrated that rTMS can improve global cognitive function and may be safely used in patients with Alzheimer's disease. Furthermore, fewer adverse events were observed when compared with the current clinical drug therapy, showing broad prospects in treating AD. However, the long-term effectiveness of rTMS in AD is still little known and should be explored in future studies.

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Clinical spectrum of the PLA2G6 gene

Renan Brandão Rambaldi Cavalheiro[1], Kaliny Oliveira Peixoto[1], João Paulo Barile[1], Patrícia Marques Mendes[1], Igor Braga Farias[1], Paulo de Lima Serrano[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Renan Brandão Rambaldi Cavalheiro; Email: renan.rambaldi@icloud.com.

Background: The PLA2G6 gene encodes the enzyme A2-Phospholipase, responsible for phospholipid metabolism. This process is vital for the maintenance of the cell membrane. Disruption of this process leads to accumulation of cell products and development of spheroid bodies in the axons. This, in turn, leads to axonal loss and neuronal death. Pathogenic variants in the PLA2G6 can produce at least 3 distinct clinical entities: Infantile neuroaxonal dystrophy (NAD), atypical neuroaxonal dystrophy (aNAD), and the adult-onset PLA2G6-related dystonia-parkinsonism. These conditions are grouped under the term PLA2G6 associated neurodegeneration (PLAN), one type of NBIA (neurodegeneration with brain iron accumulation) syndromes. The NBIAs are a group of ultra-rare (less than 1 per million people) disease; their recognition is utmost importance in order to provide patients and caregivers adequate support and treatment. **Objective:** To describe the clinical spectrum of 4 cases PLA2G6-associated neurodegeneration in a neuromuscular center results of the 4 patients included in this study, 2 patients had a phenotype of PLA2G6- dystonia-parkinsonism. In addition to dystonia and parkinsonism, these patients presented with negative myoclonus and facial-faucial- finger myoclonus. The other 2 patients presented with a motor-neuron presentation. All patients had myoclonus and 3 of 4 patients had generalized epilepsy. Characteristic brain magnetic resonance imaging findings of low signal on T2/Flair weighted-imaging was observed in only one patient (the other 3 patients had cortical atrophy as the only imaging abnormality). None of the patients had attention deifficts or a dysexecutive syndrome. Genetically, all patients were compound heterozygous for the condition. **Conclusion:** Pathogenic variants in the PLA2G6 can cause a devastating disease with varied clinical presentations and an unmet need of disease-modifying treatments. The present work aims to alert clinicians to these clinical features in order to provide prompt diagnosis and support to the patients and their caregivers.

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Clinical study of patients with familial amyloidotic polyneuropathy at Pedro Ernesto University Hospital

Marcia Maria Jardim Rodrigues[1,2], Ligia Andrade[1], Larissa Bittencourt de Carvalho[1], Julia Chaves[1], Camila Tales[1], Felipe Cerqueira Matheus[1], Clarissa Spitz[1], Marcelo Imbroinise Bittencourt[1]

[1]Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.
[2]Fundação Oswaldo Cruz, Rio de Janeiro RJ, Brazil.

Correspondence: Marcia Maria Jardim Rodrigues; Email: jardim.mm@gmail.com.

Background: Hereditary transthyretin amyloidosis (ATTRv) is a rare autosomal dominant disease associated with mutations in the transthyretin gene. Patients present with various symptoms related to sensory, motor and autonomic neuropathy, as well as gastrointestinal, ocular, cardiac, renal and orthopedic symptoms, resulting from the deposition of transthyretin amyloid fibrils in multiple organs. The progressive nature of ATTRv amyloidosis requires pre- and post-onset monitoring of the disease. It is a rare genetic disease considered endemic in some countries. There is phenotypic variability, making it a rare and very heterogeneous disease. **Method:** This is a retrospective study, through the analysis of the database of the Peripheral Neuropathy outpatient clinic of the Pedro Ernesto University Hospital that has ATTR gene mutation. Data from symptomatic and asymptomatic patients followed at the service who underwent genetic testing for the ATTR gene mutation between June 2012 and June 2024 were included. All patients underwent clinical evaluation, including neurological examination, disability score (Polyneuropathy disability score, PND), clinical cardiological Evolution, and neurophysiological and imaging studies. **Results:** A total of 60 individuals, over 18 years of age, with the ATTR gene mutation were

included. Forty individuals (66.6%) were female, mean age 49 years (18-77 years). Forty-four (73.3%) individuals were symptomatic, of which 4 (6.5%) had already died at the time of analysis. The most frequent mutation was val30met (38.3%) followed by val122ile (26.6%), ile 127val (13.3%), Phe84leu (8.3%). Eight individuals (13.3%) had a history of consanguinity. Patients registered at the outpatient clinic. Twenty-three patients (52.2%) had a mixed phenotype. PND 0 36.9%, PND I 32.6%, PND II 7.4% and PND III 19.5%. The first symptoms were neurological (76.9%), with 20.5% presenting paresthesia in the upper limbs, 55.8% paresthesia in the lower limbs, neuropathic pain in 26.4%, cardiac pain occurred in 13% of cases, dysautonomia in 6.5% and weight loss in 5.8%. ATTRv has a heterogeneous clinical presentation, varying between individuals and geographic locations. Brazil is a country with a large number of ATTRv cases. Still, there are no epidemiological studies that could help in public policies to improve early diagnosis and indicate high-cost treatments. Currently, we can conduct research through genetic studies more easily. We have demonstrated the presence of mutations that seem to be more frequent than previously determined. **Conclusion:** The description of epidemiological data from centers monitoring patients with ATTRv is extremely important to understand the nature of this rare disease in Brazil.

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Clinical variant of Nasu-Hakola disease associated with frontotemporal dementia and absence of osteodysplasia: case report

Isabelly de Arruda Cardoso Slavec[1], Thais Laura Almada da Fonseca[1], Yasmin Coelho Patrial[1], Aline Mizuta Kozoroski Kanashiro[1], Elder Yanaze Oda[1], Eña Rezende Bispo do Nascimento[1]

[1]Hospital Regional de Mato Grosso do Sul, Campo Grande MS, Brazil.

Correspondence: Isabelly de Arruda Cardoso Slavec; Email: isa_mhta@hotmail.com.

Case presentation: Patient L.S., female, 46 years old, healthy until the age of 43, housewife. In 2020, a slow and gradual progression of cognitive decline began, characterized by memory loss, impairment of executive function, spatial disorientation, as well as inability to judge, disinhibition, inattention and mood swings. It evolved into behavioral changes with infantilization, psychosis, and episodes of tonic-clonic epileptic seizures began. The patient started to neglect her own hygiene, besides loss of autonomy for functional activities and sphincter continence. No reports of movement disorders. Positive family history, with 5 brothers with a similar condition, absence of family consanguinity, and genetic investigation for Huntington's disease was negative. Exome examination revealed the presence of a homozygous variant in the TREM2 gene, an autosomal recessive inheritance condition. In the last outpatient consultation, the patient presented with significant cognitive worsening, with temporo-spatial disorientation, severe speech impairment, with anarthria, dysphagia and need for a gastrostomy for feeding. In addition, she had an apraxic gait, inexhaustible glabellar reflex, palmar grip and graping were present. On magnetic resonance imaging of the skull, significant global volumetric reduction, thinning of the corpus callosum, periventricular, semi-oval and cerebral hemispheres hypersignal on T2/FLAIR, preserved CSF flow. **Discussion:** This condition is rarely reported in the medical literature, with only 8 case reports found in Pubmed, it occurs due to the presence of a pathogenic variant in the TREM2 gene, associated with Nasu-Hakola disease or type 2 sclerosing leukoencephalopathy, which primarily causes changes and bone deformities, with late neurological manifestation, in the case described this last change was of greater severity. **Final comments:** According to the scarce literature on the subject and the age at which symptoms began, it is of great relevance to consider this pathology as a differential diagnosis of early-onset dementia.

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Clinical, epidemiological, and genetic profile of late onset Pompe disease in a tertiary hospital in the Northeast of Brazil

Caio Viana Botelho[1], Guilherme Mota Passos[1], Matheus Rolim Mendes de Alencar[1], Natália Merten Athayde[1], Antônio Edvan Camelo Filho[1], Hermany Capistrano Freitas[1], Lara Albuquerque de Brito[1], Keyla Rejane Frutuoso de Morais[1], Eduardo Braga de Oliveira[1], Cleonisio Leite Rodrigues[1]

[1]Hospital Geral de Fortaleza, Serviço de Neuromuscular, Fortaleza CE, Brazil.

Correspondence: Caio Viana Botelho; Email: caiobotelhoneurologia@gmail.com.

Background: Late-onset Pompe disease (LOPD) is a genetically inherited autosomal recessive metabolic myopathy associated with a deficiency of the enzyme acid alpha-glucosidase. It presents with symptoms of proximal appendicular, axial, and respiratory muscle weakness. Although rare, it is a treatable condition, where the early introduction of enzyme replacement therapy (ERT) in symptomatic individuals is crucial to reduce morbidity and mortality. **Objective:** To describe the clinical, epidemiological, and laboratory profile of patients diagnosed with LOPD and medically monitored at a reference center in the state of Ceará, Brazil. **Methods:** A descriptive cross-sectional analysis was performed on 11 patients with LOPD at the neuromuscular diseases outpatient clinic of Hospital Geral de Fortaleza-CE. Data recorded included sex, origin, age of symptom onset, genotype, phenotype, CPK levels, acid alpha-glucosidase enzyme activity, neurophysiological pattern in electromyography, and therapy employed. **Results:** Between 2016 and 2024, 11 patients (from 4 families) were identified with a clinical and molecular diagnosis of late-onset Pompe disease, corresponding to about 31.4% of the patients monitored for genetically determined myopathy in the outpatient clinic (excluding patients with Steinert myotonic dystrophy). There was an equivalent prevalence regarding sex (about 1:1). The average age of symptom onset was around 28 years. The most frequent variant was c.-32-13T>G, present in all patients, and often associated with other pathogenic mutations (compound heterozygosity). The most common phenotype was proximal weakness, occurring in 9 patients. CPK levels were altered in 10 patients (90.9%), with an average diagnostic value of 612 U/L and a maximum of 1457 U/L. Neurophysiological studies showed findings consistent with chronic myopathy in all patients, with the presence of myotonic discharges in 8 of them (72%). Acid alpha-glucosidase enzyme activity was measured in 9 patients and was considered low in all, with an average value of 0.45µmol/L/h (reference value: above 2.1µmol/L/h). Almost all of them (9 out of 11) were on ERT with alglucosidase alfa. For clinical follow-up evaluation, strength grading scales (MRC), grip strength measurement with a dynamometer, 10-meter walk test, and inspiratory and expiratory maximum pressure measurement with a manovacuometer were used. **Conclusion:** To our knowledge, this is the largest series of LOPD cases described from a single neuromuscular disease reference center in our country. The most frequent pathogenic variant was the intronic c.-32-13T> G, along with the phenotypic pattern of proximal weakness and EMG showing proximal myopathy with electrical myotonia. The vast majority of patients are receiving ERT and are being monitored with functional motor exams, respiratory parameters, and 10-meter walk tests.

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Clinical, epidemiological, serological, and neurophysiological data of patients with myasthenia gravis from a reference center in Northeast of Brazil

Natália Merten Athayde[1], Matheus Rolim Mendes de Alencar[1], Caio Viana Botelho[1], Hermany Capistrano Freitas[1], Lara Albuquerque de Brito[1], Eduardo Braga de Oliveira[1], Cleonisio Leite Rodrigues[1]

[1]Hospital Geral de Fortaleza, Serviço de Neuromuscular, Fortaleza CE, Brazil.

Correspondence: Caio Viana Botelho; Email: caiobotelhoneurologia@gmail.com.

Background: Myasthenia gravis (MG) is the most common autoimmune disease of the neuromuscular junction. Its hallmark is the fluctuating muscle weakness affecting ocular, bulbar, facial, limb, and/or respiratory muscles. Clinical manifestations of MG can vary from mild, focal weakness in some patients to severe tetraparesis with respiratory failure. It is a treatable condition in most cases but can result in significant morbidity and even mortality. **Objective:** To describe the main epidemiological, clinical, and neurophysiological characteristics of MG patients and their serological correlation in a reference service for neuromuscular diseases in the Northeast. **Methods:** A cross-sectional and retrospective study was conducted by reviewing the medical records of MG patients followed at the Neuromuscular Diseases Outpatient Clinic of Hospital Geral de Fortaleza (HGF). The diagnosis of MG was based on clinical characteristics, positive serology, neurophysiological findings (repetitive nerve stimulation test - RNS, and jitter study), and treatment response. **Results:** A total of 154 patients were included, 93 (60.6%) female and 61 (39.3%) male, aged 17 to 84 years. The vast majority of patients had the generalized form (90.2%), while the pure ocular form was the minority (9.74%). Of the total analyzed patients, 18.18% experienced myasthenic crisis, with the great majority occurring in the first 18 months of disease evolution. All of them were treated with plasmaferesis or IV immunoglobulin. Regarding antibody levels, patients were categorized as positive for anti-acetylcholine receptor antibodies (anti-AChR) (63.63%), positive for anti-muscle-specific tyrosine kinase antibodies (anti-MuSK) (5.19%), double seronegative (21.42%), and with pending results (9.74%). The double seronegative patients had typical clinical presentation, abnormal RNS or jitter analysis and good treatment response. Some of them had negative genetic molecular screening. It was observed that the majority of patients who experienced myasthenic crisis were anti-ACh positive (71.4%). Of the 154 patients analyzed, 151 underwent RNS testing, of which 84.7% had abnormal results. Among patients with normal RNS who underwent jitter analysis (N=17), 100% had abnormalities in this method. Regarding drug treatment, approximately 80.5% of patients showed clinical improvement with first-line treatment, but with high scores on quality of life questionnaires though. **Conclusion:** This study provides a comprehensive overview of the clinical, epidemiological, serological, and neurophysiological findings of 154 patients with MG. The results are generally consistent with those described in the literature, highlighting the percentage of patients who experienced a myasthenic crisis, which, compared to some references, was higher than expected. This finding may be attributed to possible delays in diagnosis and difficulties in accessing healthcare services during the diagnostic phase of the disease.

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Clinical-epidemiological aspects of patients with myasthenia gravis in an outpatient of the public health system in northeast of brazil

Marivaldo Yslan Silveira Leal[1], Luisa Dias Moreno Aravena[1], Antonio Lázaro Santos Soares Júnior[1], Marcel Leal Ribeiro[2], Bruno Bacellar Pedreira[2], Alice Silva Oliveira[2], Jamary Oliveira Filho[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil. [2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Luísa Dias Moreno Aravena; Email: luisaaravena@ufba.br.

Background: Myasthenia Gravis (MG) is an autoimmune disease that affects the neuromuscular junction, resulting in muscle weakness with fatigability and fluctuating pattern. The study of clinical and epidemiological aspects in MG is essential to enhance therapy, especially in the context of the public healthy system, in which the majority of users have low income and education, in addition to less accessibility to comprehensive medical care. **Objective:** To describe the clinical and epidemiological factors in patients with MG treated at a reference outpatient clinic of the public health system in Salvador (Bahia). **Methods:** Descriptive and analytical, single-center study, with patients over 18 years diagnosed with MG. Collection was conducted using oral questionnaires and consultation of medical records. **Results:** 60 patients participated in the study. The majority of whom were female (73.3%), more specifically black women (61.7%). The average age of symptom onset was 28.5 (SD±11.98) years. 46.7% of patients presented with the generalized form, 35% with the ocular form and 18.3% with the bulbar form. 7 out of 10 did not have access to a neurologist in the first month. For diagnostic tests, 92.9% of patients underwent the anti-rACh antibody test, which was reactive in 66.7%, and 22.4% the anti-MUSK test, which was reactive in 30% of cases. 96.4% were able to undergo at least one electroneuromyography. The average number of crisis after the initial manifestation was 2.42 (SD±1.60) and half of the participants had a myasthenic crisis. On admission, the generalized form was the most prevalent (68.3%), followed by ocular (20%) and bulbar (11.7%). The median MGADL was 3 (IQ 1 - 6.75) and the median MGQoL was 9 (IQ 4.25 - 15). Anticholinesterase is the medication most used by 96.7%, while 46.7% corticosteroids and 50% azathioprine. 48.3% underwent thymectomy. 58.3% patients have some associated comorbidity, with 30% of these having high blood pressure, 21.7% diabetes mellitus and 20% dyslipidemia. 70% are overweight or obese and only 26.7% practice physical activity. 1 in 5 patients has another autoimmune disease, with Graves' disease (8.3%) and Hashimoto's thyroiditis (6.7%) being the main ones. 86.4% were studying or working before the onset of symptoms and only 36.7% continued to work or study after the diagnosis of MG. 6 out of 10 participants did not complete high school. 61.7% live on a family income of less than 2 minimum wages and 52.2% receive some form of social assistance. 62.7% of public health system users reported difficulty in accessing the public network, especially when it came to making appointments and taking tests. **Conclusion:** The analysis of clinical and epidemiological characteristics is necessary to conduct diagnostic and therapeutic approaches in patients with Myasthenia Gravis in the context of the health system. Thus, it may help guiding the applicability of public health policies in order to minimize treatment delay.

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Clinical-epidemiological profile and correlations with arboviruses of inpatients with Guillain-Barré syndrome at a hospital in the Northeast of Brazil

Matheus Rolim Mendes de Alencar[1], Fernanda Martins Maia Carvalho[1], Hermany Capistrano Freitas[1], Eduardo Braga de Oliveira[1], Lara Albuquerque de Brito[1], André Borges Ferreira Gomes[1], Paula Camila Alves[1], Gláuber de Menezes Ferreira[1], Déborath Lúcia de Oliveira Diniz[1], Cleonísio Leite Rodrigues[1]

[1]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Matheus Rolim Mendes de Alencar; Email: matheus_alencar_1@hotmail.com.

Background: Guillain-Barré syndrome (GBS) is an acute polyradiculoneuropathy characterized by weakness, sensory deficits and sometimes cranial neuropathy, autonomic dysfunction and respiratory insufficiency. Diagnosis is usually based on clinical features, electrodiagnostic studies and laboratory findings. Timely diagnosis is crucial to stablish the correct treatment and try to avoid disability. **Objective:** To describe the clinical-epidemiological profile of patients with GBS and their correlation with arboviruses infections treated at a public tertiary hospital in the last 3 years. **Methods:** This is a retrospective cohort study of patients with acute polyradiculoneuropathy, who were evaluated for GBS at the time of hospitalization in a reference hospital in Northeast Brazil, between January 2022 and May 2024. Data such as age, gender, functional status, and other clinical and epidemiological information were analyzed from medical records. The test used for arbovirus evaluation was PCR from serum and cerebrospinal fluid, which was not widely available for all patients. **Results:** A total of 64 suspected patients were evaluated. Of these, 57 inpatients were diagnosed with GBS, while 7 had others diagnoses. At the time of hospitalization, patients ranged in age from 18 to 82 years (mean of 46.7 years). Most patients were male (68.4%), and the mean time from symptom onset to hospital admission was 8.22 days (SD 9.11 days). A history of infectious diseases or vaccinations was present in 70% of patients. Arbovirus assays were available for 37 patients (65%), of which 16% tested positive for arboviruses, notably dengue and chikungunya virus; no tests for Zika virus were positive. The main clinical pattern of GBS was classic sensorimotor (79%), but other variants such as Miller-Fisher Syndrome (7%), Bickerstaff brainstem encephalitis (3.5%), paraparetic (3.50%), and bilateral facial palsy with paraesthesias (1.75%) were also present. Albumino-cytological dissociation was present in 74% of patients, and the main electrodiagnostic patterns observed were AIDP (54%) and AMSAN (30%), with sural sparing in 37% and H reflex abnormalities in 98% of the sample. Regarding severity and functional status, 37% of inpatients required assisted ventilation in an intensive care unit. The mean Hughes scale score at hospital admission was 3.61 (SD 0.87), and the Polyneuropathy Disability Scale at hospital discharge was IV (28%) and IIIa (19%). Intravenous immunoglobulin (33%) and plasma exchange (56%) were the chosen treatments; other patients may not have needed treatment either due to loss of the therapeutic window or good functional status. **Conclusion:** The classic AIDP pattern of GBS was the most common in our sample. Arbovirus infections continue to play a significant role as triggers for GBS in our region. Preventive measures to control these infections are crucial, as they could reduce the incidence of future cases of GBS.

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Clinical-radiological mismatch in patient with moyamoya syndrome and seronegative rheumatoid arthritis

Vitor Gustavo Leão Souto[1], Bruna Acioly Leão[2], Nayra Roberta Sales Salvador[2], Juliana Oliveira de Almeida[2], Patrícia Pereira Nunes Ribeiro[2], Túlio Marlus Castro Lucena[2], Assis Porfirio Furtado Nogueira[2], Maria Júlia Tabosa de Carvalho Galvão[1], Geovane Brito Affonso[1], Letícia Januzi de Almeida Rocha[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil. [2]Hospital Geral do Estado, Maceió AL, Brazil.

Correspondence: Vitor Gustavo Leão Souto; Email: gustavosoutoo11@gmail.com.

Case presentation: A 70 year old female with seronegative rheumatoid arthritis (RA), hypertensive, dyslipidemic and coronary heart disease presented in 2016 with a sudden onset of weakness and headache. In the investigation there was clinical and radiological inconsistency due to complete obstruction of the internal carotid artery in non-symptomatic territory. Between 2016 and 2024, it evolved with improved functionality and motor deficits, but there was progression of intracranial stenosis and the emergence of the moyamoya pattern. There are complete occlusion of the ACID, ACMD M1, ACME M1, ACAD, stenosis less than 50% in several vascular territories and small cerebral aneurysms, with no previous or current history of rupture. Despite the extensive vasculopathy, the skull resonance only showed an ischemic gap in the left internal capsule. **Discussion:** Moyamoya disease is a primary vasculopathy of the brain that causes chronic and progressive steno-occlusion of the internal carotid arteries, forming an abnormal collateral vascular network at the base of the brain known as "moyamoya vessels". When secondary, it is called Moyamoya syndrome (MMS). SMM is associated with atherosclerosis, brain tumor, meningitis, sickle cell disease, thyrotoxicosis and autoimmune diseases. The clinical manifestations of MMS may be related to recurrent stroke, of the ischemic or hemorrhagic type, occurring in up to 25% of patients. Female sex is an independent factor for disease progression, and inflammatory diseases such as RA can contribute to the progression of atherosclerosis. Compared to other autoimmune conditions such as Lupus and Sjögren's syndrome, the association between MMS and RA is less well described. The literature highlights progression to cognitive deficits, visual changes, intracranial hemorrhage, generalized seizures and other neurological manifestations, but in this case only headache and minimal motor deficit without functional impairment have been found so far. **Final comments:** Given the significant risk of morbidity, this case demonstrates the importance of managing comorbid autoimmune conditions that may predispose to the development of MMS. The pathophysiology of this association still remains uncertain. It is believed that the immune system is responsible for amplifying the inflammatory cascade, triggering chain vessel stenosis, but more studies are needed so that secondary prophylaxis for these patients is truly effective.

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Clinicogenetic characterization and response to disease-modifying therapies in spinal muscular atrophy: real-world experience from a reference center in Southern Brazil

Ana Letícia Amorim de Albuquerque[1], Júlia Kersting Chadanowicz[1], Isabela Possebon Bevilacqua[1], Ana Lucia Portella Staub[2], Pablo Brea Winckler[2], Thayne Woycinck Kowalski[2], Karina Carvalho Donis[2], Michele Michelin Becker[2], Jonas Alex Morales Saute[2]

[1]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Ana Letícia Amorim de Albuquerque; Email: analeticia.neurologia@gmail.com.

Background: Spinal muscular atrophy (SMA) is an autosomal recessive neurodegenerative disease characterized by progressive proximal muscle atrophy and weakness. **Objective:** This study addresses the scarcity of research on novel disease-modifying therapies for SMA in Latin America by reporting a real-world experience in Southern Brazil. **Methods:** This is a single-center historical cohort that included all patients diagnosed with spinal muscular atrophy at a Regional Reference Service for rare diseases. **Results:** Eighty-one patients were included, of whom 7 died during follow-up. Of the remaining 74 patients, 5.4% were identified as pre-symptomatic, 24.3% with SMA type 1, 28.4% with type 2, 36.5% with type 3, and 5.4% with type 4. Approximately 42% of these patients received specific disease-modifying therapy, of these, 96.8% received Nusinersen, with 19.35% transitioning to gene therapy using Onasemnogene Abeparvovec, and 6.4% starting Risdiplam. Most patients with SMA type

1 were on disease-modifying treatment, whereas only slightly over a third of patients with type 2 and about 10% of those with type 3 were receiving such treatments. Among treated patients, 80% demonstrated improvement in motor performance during the follow-up, with a lesser therapeutic response being associated with late initiation of treatment and low motor function scores at baseline. **Conclusion:** This real-world study reinforces the effectiveness of disease-modifying therapies for SMA in Brazil within the context of low- and middle-income countries, which is greater the earlier and the better the patient's functional status.

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CLIPPERS syndrome: case report

José Víctor Filho de Oliveira[1], Stephania Neves Scapim[1], Iasmim Portela Maifrede[1], Leopoldo Antonio Pires[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Juiz de Fora MG, Brazil.

Correspondence: José Víctor Filho de Oliveira; Email: victorr_oliveira@hotmail.com.

Case presentation: A 74-year-old man, with a benign prostatic hyperplasia, presents with significant unintentional weight loss combined with progressive tetraparesis over a one-year period, leading to confinement to a wheelchair. The physical examination revealed pseudobulbar affect, mild dysarthria, tetraparesis with distal predominance in the upper limbs and proximal predominance in the lower limbs with signs of upper motor neuron involvement. In addition, there was significant bilateral appendicular and truncal ataxia, intentional tremor and hyperreflexia of axial facial reflexes. A brain magnetic resonance imaging (MRI) revealed multiple areas of heterogeneous enhancement with predominantly perivenular pattern in the white matter of the centrum semiovale, periventricular regions, nucleocapsular areas, brainstem, with predominance in the lower part of the pons and medulla, and midbrain cerebral peduncles. There was also enhancement along the ependymal surface of the lateral ventricles, especially near the atrium, posterior and temporal horns. High signal areas on T2 and FLAIR sequences were associated with no apparent diffusion restriction in the centrum semiovale and confluent periventricular white matter of both cerebral hemispheres, mainly adjacent to the occipital horns of the lateral ventricles. Cervical and thoracic MRI demonstrated signs of cervical myelitis, with scattered areas of irregular enhancement on contrast imaging. Initial hypotheses were autoimmune disorders , lymphoproliferative disease, and chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids (CLIPPERS). Cerebrospinal fluid (CSF) analysis revealed elevated protein levels and mild lymphocytosis without other positive infectious markers, and oligoclonal bands of pattern 3. Tests for autoimmune, inflammatory, and systemic granulomatous diseases were negative. Pulse therapy with methylprednisolone was prescribed, with improvement of symptoms. **Discussion:** CLIPPERS is a rare central nervous system condition centered in the pons. Its pathophysiology involves T cells and shows a good response to glucocorticoids. Patients typically present with subacute symptoms such as gait ataxia, diplopia, and dysarthria. MRI reveals a distinctive pattern of punctate and curvilinear perivascular gadolinium enhancement in the pons and adjacent regions. Pathogenesis remains unknown but an immune-mediated process against a perivascular antigen is a possible explanation. Diagnosis is based on clinical findings, radiological features, and response to GCS. The diagnosis is by exclusion. MRI findings show a characteristic pattern of lesions, which include the possibility of extending into the cervicothoracal spinal cord. **Final comments:** Neurologists should recognize the typical conditions and presentations, as prompt and appropriate treatment with GCS shows significant improvement. Maintenance with immunosuppressive agents is often necessary to prevent relapses.

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Cluster analysis reveals diversity among the neuropsychological phenotypes of SARS-Cov-2 infected subjects

Ítalo Karmann Aventurato[1], Mateus Henrique Nogueira[1], José Flávio Becchelli[1], Marjorie Cristina Rocha da Silva[1], Ana Paula Beppler Lazaro Lino[1], Leila Camila Santos Silva[1], Lucas Scardua Silva[1], Marina Koutsodontis Machado Alvim[1], Fernando Cendes[1], Clarissa Lin Yasuda[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Ítalo Karmann Aventurato; Email: italo.kar.av@gmail.com.

Background: while cognitive dysfunction in post-COVID-19 patients has been documented, a comprehensive exploration of neuropsychological subgroups is yet to be undertaken. **Objective:** This study, however, takes a novel approach, aiming to delineate the neuropsychological profile of various subgroups using a data-centric hierarchical clustering method. **Methods:** we analyzed the data from the NeuroCovid cohort, comprising different neuropsychological tests (covering multiple cognitive domains) and neuropsychiatric scales. Subjects were recruited through online forms and on-campus ads. After outlier removal, a factor analysis was performed to derive the latent cognitive factors in the data. Factor-transformed data was renormalized according to the expected population mean and standard deviation and used in hierarchical analysis with Ward's method. The number of clusters was chosen by Monte-Carlo simulations using a multivariate Gaussian distribution with the same covariance as the sample. Subjects were compared between clusters regarding demographics, illness severity, depression and anxiety scores. **Results:** among more than 5000 subjects screened, 569 were evaluated in person, and complete neuropsychological data was obtained from 265. Sixteen subjects were identified as outliers using the Isolation Forest algorithm. We applied factor analysis to neuropsychological data from the remaining 249 subjects. Data was deemed proper for factor analysis (Kayser-Meyerz-Olkin statistic 0.63), and the number of factors was determined to be six by parallel analysis. Factors regarding memory, executive functions (EF), automatic processes (AP), motor, visuospatial construction (VSP) and visuospatial construction speed were identified and showed congruence with current cognitive theory. One of the six clusters identified was excluded due to insufficient subjects (n=2). The remaining clusters were (A) unimpaired (n=86), (B) visuospatial impaired (n=18), (C) memory impaired (n=83), (D) AP impairment (n=24), and (E) EF impairment (n=36). Cluster C comprised older subjects, whereas subjects were younger in Cluster E. Depression scores (Beck's Depression Inventory-II) were increased in clusters C and E. Remaining demographic and neuropsychiatric scales did not show statistically significant differences. **Conclusion:** our data shows that the individuals with persistent symptoms after mild COVID present a heterogeneous neuropsychological profile with subgroups showing impairments in different cognitive domains. The findings align with recent publications showing that post-COVID subjects in different age ranges and disease severity presented distinct cognitive profiles. An increased score in depressive symptoms is also consistent with previous reports of neuropsychiatric illness in those previously infected by SARS-Cov-2.

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Cluster phenotyping and genomic correlation of motor neuronopathies

Rodrigo Siqueira Soares Frezatti[1], Pedro José Tomaselli[1], Gustavo Maximiano Alves[1,2], Jasper Morrow[3], Mike G. Hanna[3], Mary M. Reilly[3], Cláudia Ferreira da Rosa Sobreira[1], Antônio Carlos dos Santos[1], Wilson Marques Júnior[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Departamento de Neurologia, Ribeirão Preto SP, Brazil.
[2]Icgnmd Consortium.
[3]University College London, Queen Square Institute of Neurology, Department of Neuromuscular Diseases, London, UK.

Correspondence: Rodrigo Siqueira Soares Frezatti; Email: frezattirodrigo@hotmail.com.

Background: Spinal muscular atrophy (SMA) is mainly linked to biallelic loss-of-function variants in the SMN1 gene located on 5q13. In recent years, with the advance in next-generation sequencing (NGS), several genes outside of chromosome 5 have been recognized. This subgroup, known as non-5q SMA, in linked to a wide phenotypic variability and, albeit a growing list of potentially causative genes, there is a low diagnostic yield amongst them. There is an unmet need to better characterize and molecularly approach these patients. **Objective:** We sought to characterize a Brazilian cohort of non 5q SMA patients through clinical and paraclinical analysis and evaluate NGS performance in two main groups: pure and complex. **Methods:** Cases with non-length dependent neurogenic weakness were included. Patients were categorized regarding a pure lower motor neuron involvement or within a complex phenotype. Molecular analysis was conducted considering initial assessment and included single gene test (ATXN2, VWA1, AR, VAPB), WES or WGS in whom initial analysis was negative. **Results:** The cohort was predominantly composed by sporadic cases (54%). Mean age of onset was 15 years old. So far, sixty-seven probands were tested, most of them with WES (58/67) and in some cases with bespoken single gene test (6/67). Muscle MRI was performed upon structured approach for cases with confirmed molecular diagnosis. Overall, thirty-two patients (49%) had an initial positive result and nineteen different genes have been identified: AR (n=1), AARS1 (n=1), COL6A2 (n=1), COL6A3 (n=1), TK2 (n=2), SIGMAR1 (n=1), VRK1 (n=5), BICD2 (n=2), DYNC1H1 (n=1), FBX038 (n=1), HEXA (n=2), VAPB (n=5), TBCK (n=1), FUS (n=2), SLC52A (n=1), MME (n=1), PRPS1 (n=3), COQ7 (n=2), MYH7 (1).Complex phenotype patients achieved a higher diagnostic yield (60% x 22%) and dysautonomia is, so far, the most predictive feature to positive diagnosis as it was present in 80% of our VAPB cases and not in any other patient. **Conclusion:** This is an ongoing work with still incomplete molecular assessment and muscle MRI analysis. Nevertheless, so far, we highlight the highest diagnostic yield in complex phenotype, the importance of muscle MRI in the deep phenotyping effort, the diversity of encountered genes and the unexpected frequent relation with "myopathic" genes: MYH7,COL6A3, COL6A2, SGCA and TK2. Overall, a comprehensive and clustered phenotyping, using muscle MRI muscle fatty infiltration pattern identification seems to positively influence diagnostic outcome.

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Cluster tic syndrome: the duel of two excruciating headaches

Sofia Mônaco Gama[1], Luccas Almeida Andrade[1], Gabriel Pinheiro Martins de Almeida e Souza[1], João Vitor Gerdulli Tamanini[1], Guilherme Antonio Silva Ribeiro[1], Flávio Moura de Rezende Filho[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Sofia Mônaco Gama; Email: smonacogama@gmail.com.

A 50-years-old woman presented with a 3 year-history of severe facial pain that was characterized by paroxysmal episodes of shock-like pain in the right maxillary and mandibular regions, lasting seconds and triggered by a

breeze or light touch in the face. Further investigation with brain magnetic resonance imaging (MRI) demonstrated neurovascular conflict on the right side of the trigeminal nerve. The diagnosis of classical trigeminal neuralgia was made, and treatment with carbamazepine was initiated, resulting in a good response. However, the patient developed a cutaneous allergic reaction and the treatment was replaced with gabapentinoids. Two years ago, the patient began experiencing a new pattern of severe unilateral headache in the right retro-orbital and hemicranial regions, lasting 10 to 40 minutes with a significant sense of restlessness. Concurrently, there was a worsening of the previous facial pain, frequently occurring before this new headache. Additional investigations were unremarkable and optimizing gabapentinoid treatment did not result in improvement. One year ago, both types of pain worsened. Therefore, neurovascular decompression surgery was performed, resulting in improvement of the shock-like pain in the subsequent weeks. Nevertheless, the patient developed a new exacerbation of the right headache, occurring up to 8 times per day. The diagnosis of cluster tic syndrome was made. Treatment with verapamil was initiated, as well as oxygen during crises, resulting in significant improvement of the symptoms. Cluster tic syndrome is a rare neurological condition that combines features of both cluster headaches and trigeminal neuralgia. The first description was in 1978 and since then only approximately 40 patients have been reported. The headache initially presents as trigeminal neuralgia or as cluster headache separately or as a combination of both. The pain is unilateral and severe. In cluster headache episodes, the pain is localized in the periorbital or temporal region, associated with autonomic symptoms. In trigeminal neuralgia, the pain is localized in territories innervated by the trigeminal nerve and characterized by paroxysm sharp, shock-like pain triggered by facial or intraoral stimuli. Treatment of the acute phase is with oxygen therapy, triptans and prednisone in cluster headache attacks. In exacerbations of trigeminal neuralgia, management is with triptans, intravenous phenytoin, lidocaine or local procedures. Maintenance treatment is based on oral medications used for both types of headache, such as verapamil, lithium, carbamazepine, lamotrigine and gabapentin. Surgical decompression of trigeminal nerve is indicated for refractory cases.

- Cluster tic syndrome is characterized by trigeminal neuralgia and cluster headache simultaneously.
- This condition is important to be recognized because both must be treated for the patient to become headache free.

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c-NORSE score identifies patients with cryptogenic NORSE in a Brazilian cohort

João Henrique Fregadolli Ferreira[1], Anna Maria Gomes[1], Ana Carolina Zetehaku[1], Taíssa Ferrari Marinho[1], Fabio Fieni Toso[1], Caio César Diniz Disserol[1], Luís Otavio Caboclo[1], Livia Almeida Dutra[1]

[1]Hospital Israelita Albert Einstein, Instituto do Cérebro, São Paulo SP, Brazil.

Correspondence: João Henrique Fregadolli Ferreira; Email: jhfregadolli@gmail.com.

Background: New-Onset Refractory Status Epilepticus (NORSE) is a severe condition requiring extensive investigation and long hospital stays. Most cases remain without a cause, termed cryptogenic (c-NORSE). Data with inconsistent antineuronal antibody testing showed that autoimmune encephalitis (AE) is the most common identifiable cause of NORSE in adults. Second-line treatment for AE and c-NORSE differs, with drugs targeting innate immune pathways, such as anti-IL1 and anti-IL6, recommended for c-NORSE. Recently, a score based on clinical findings and complementary investigation helped identify c-NORSE at early stages. **Objective:** To describe a cohort of Brazilian NORSE patients and evaluate the applicability of c-NORSE score. **Methods:** We retrospectively evaluated patients with Status Epilepticus (SE) or RSE included in the Brazilian Autoimmune Encephalitis Network from January 2017 to November 2023. Patients were investigated with brain MRI, CSF analysis and herpes virus PCR, followed by testing for antineuronal antibodies using TBAs, CBAs, and immunoblots in serum and CSF. Patients were allocated into two groups according to C-NORSE score: high score (≥5) or low score (<5). Groups were compared using Mann-Whitney, Fisher's exact test or chi-square test. p-value<0.05 was considered significant. Sensitivity and specificity were calculated using contingency table analysis. **Results:** Of 81 SE or RSE patients, 40 met NORSE criteria and were selected. Eighteen (40%) were children ≤13y and 52% were female. Based on c-NORSE score, 11/40 (27%) patients were in the high score (HS) group and 29/40 (73%) were in the low score group (LS). HS group had more bilateral and symmetric MRI findings (HS 100% x LS 35%, p=0.003), and fewer memory or behavioral symptoms (HS 27% x LS 83%, p=0.002). No difference was observed in gender, prodromic fever, or dyskinesia. LS group had alternative diagnosis: seropositive AE (n=7), MOGAD (n=2),c-NORSE (n=8), HSV encephalitis (n=4), seronegative AE (n=1), encephalomyelitis (n=1), and other causes (n=5). All patients from the HS group remained as c-NORSE. The sensitivity of the c-NORSE score was 58%, and the specificity was 100%. About 22% NORSE patients had AE, and 47% were cryptogenic. c-NORSE score had lower sensitivity than previously reported, and high specificity. Thus, at bed side, patients with ≥5 points probably present c-NORSE, and treatment decision may favor IL-6 or 1L-1 blocking drugs. Our findings differ from prior reports mostly because (1) we did not consider patients with Graus criteria for probable AE as a final diagnostic category, (2) samples were tested with two different techniques (TBAs and CBAs), and only antibodies with well stablished clinical pathogenicity were considered. Our data confirms that AE can manifest as NORSE, but other findings suggestive of AE other than seizures must be actively searched. **Conclusion:** c-NORSE score is a valuable tool in identifying cryptogenic cases in our population.

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Coexisting anti-NMDAR and MOG antibody-associated encephalitis: case report

Santhiago Calvelo Graça[1], Maria Clara Motta Barbosa Valente[2], Cícero Rodrigues Veloso[2], Rachel Leirner Argelazi[1], Mauro Gualano da Costa Abreu[1], Mariana Akemi Takegawa Miyahara[1], Rafael Chaves Claudino de Queiroga[2], Annelise Akemi Higa Lee[2], Keila Narimatsu[2], Francisco Tomaz Meneses de Oliveira[2]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Santhiago Calvelo Graça; Email: santhiagocalvelo@hotmail.com.

Case presentation: A 29-year-old patient presented to the emergency with a history of epileptic seizure associated with headache, paresthesia and paralysis of the right upper limb. In the previous week, the patient had experienced their first seizure, characterized by sudden appearance of scotomas in the central field of vision of the right eye, associated with abduction of the right eyeball, disperception and progressing to a tonic-clonic seizure. On the days following the initial event, the patient reported episodes of headache after transient episodes of paresis and paresthesia of the right arm and the trunk, alongside motor aphasia. These episodes persisting despite the use of 50 mg/day of lacosamide. Neurological examination revealed slurred speech and atypical gait, without other abnormalities. The patient had previous results from magnetic resonance imaging (MRI) performed on another service, that showed focal lesions suggestive of central nervous system infection and involvement of the leptomeninges. An electroencephalogram revealed right centroparietal epileptic paroxysms. The initial treatment included: acyclovir, dexamethasone, and maintenance of lacosamide. During investigation, a molecular panel for meningitis/encephalitis (FilmArray) was negative for various agents, including varicella-zoster and other herpes viruses. The cerebrospinal fluid was clear with lymphocytic pleocytosis (56 cells with 96% lymphocytes), negative for bacteria and fungi and the autoantibodies test indicated positive anti-NMDA. Given the neuroimaging findings and clinical presentation, the possibility of an overlap with MOGAD was considered. Therefore, anti-MOG antibodies were requested, and the result was positive, confirming the clinical-radiological hypothesis and aiding in the management and delineation of the prognosis. During follow-up, the patient showed improvement in seizures. A new MRI showed improvement of the initial lesions. In this context, the patient was diagnosed with coexisting anti-NMDAR and MOG Antibody-Associated Encephalitis and immunotherapy was introduced. **Discussion:** The presence of antibodies against both NMDA receptor and MOG represents a recently described entity: overlapping autoimmune encephalitis (OAE). This condition can manifest with a distinct clinical phenotype compared to single-antibody encephalitis. In our case, following the positive anti-NMDAR antibody test, the possibility of co-existing anti-MOG antibodies arose based on neuroimaging findings. OAE should be suspected in atypical cases differing from classic anti-NMDAR or MOGAD presentations. Fortunately, after the treatment the patient has maintained a stable clinical course with no relapse. **Final comments:** This case highlights the importance of investigating autoantibodies in patients with encephalitis of probable autoimmune etiology, especially when the clinical is atypical. Early diagnosis and initiation of appropriate immunotherapy are crucial for improving prognosis.

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Cognition and endothelial function in hypertensive individuals with and without a history of COVID-19: a cross-sectional study

Pedro Igor Lustosa Roriz[1], Ádrya Aryelle Ferreira[1], Anderson Bispo Coelho[1], Ellen Cristinni Maciel Canuto[1], Antônio Marconi Leandro da Silva[1], Gerlene Grudka Lira[1], Matheus Sobral Silveira[1], Anna Cristina da Silva Santos[1], Armele de Fatima Dornelas de Andrade[2], Victor Ribeiro Neves[1]

[1]Universidade de Pernambuco, Petrolina PE, Brazil.
[2]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Pedro Igor Lustosa Roriz; Email: pedroilroriz@gmail.com.

Background: Patients with systemic arterial hypertension (SAH) are at higher risk of developing a more severe course of SARS-CoV-2 viral infection, in addition to potentially developing lasting cognitive sequelae. Current literature provides evidence that the mechanism underlying the worse outcome of COVID-19 and the emergence of cognitive impairments in hypertensive individuals is endothelial dysfunction. Flow-mediated dilation (FMD) is a technique that has been used for several decades to assess endothelial function. **Objective:** To evaluate the effect of exposure to mild forms of COVID-19 on the cognitive status and endothelial function of patients with SAH. **Methods:** A cross-sectional study compared cognitive status, assessed through neuropsychological tests, and endothelial function, assessed through FMD, in 52 patients diagnosed with SAH who were previously exposed or not to mild forms of COVID-19. Patients were allocated into two groups: SAH with COVID-19 exposure (SAHEXP_COVID) and SAH without COVID-19 (SAH). Assessment was documented between six months and eighteen months post-infection. **Results:** No difference in FMD% was observed between the studied groups (p=0.69): for the SAH group, FMD% was 6.7 (IQR 6.1) and

for the SAHEXP_COVID group, FMD% was 5.4 (IQR 7.6). No differences in cognitive performance were observed when comparing the groups, nor was there a relationship between cognition and endothelial function. **Conclusion:** Although the study did not show differences between the groups, this finding may provide evidence that many lesions caused by the SARS-CoV-2 virus, in cognitive and endothelial impairment, may be self-limited in nature, especially in cases of mild infection.

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Cognitive and neuropsychiatric analysis of patients with myotonic dystrophy type I

Daniel Delgado Seneor[1], Vitor Dias Gomes Barrios Marin[1], Carlos Alberto Castro Teixeira Júnior[1], Fernanda Lustosa Cabral Gomez[1], Lucas Toshio Uenishi[1], Lara Santos Machado[1], Igor Braga Farias[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Daniel Delgado Seneor; Email: danielseneor30@gmail.com.

Background: Myotonic dystrophy type 1 (DM1) is a genetic disorder that affects multiple systems, including the central nervous system. While motor impairments in DM1 are well-documented, the neuropsychiatric profile of these patients, particularly in the Latin American context, is still being delineated. **Objective:** This study aims to evaluate the cognitive and neuropsychiatric profile adult DM1 patients and correlate clinical data with neuropsychiatric test outcomes. **Methods:** Fifty adult patients diagnosed with DM1 were recruited from the Myopathy Unit, Division of Neuromuscular Diseases at Universidade Federal de São Paulo. They were submitted to a comprehensive neuropsychological battery comprising 17 tests that assess emotional status and multiple cognitive domains, including attention, language, executive function, and memory. The patients' performance was systematically compared with that reported in previously conducted population studies documented in the medical literature. Furthermore, correlative statistical analyses were performed to determine whether there is association between performance in each test and the degree of motor impairment in the same individual (quantified by the Medical Research Council Scale for Muscle Strength). **Results:** Cognitive assessments revealed impairments across various domains, notably in the clock-drawing test and the cube drawing test (three-dimensional visuoconstructive praxis). Interestingly, patients did not show difficulties in copying two-dimensional images such as a rhombus, circle, and Rey's semi-complex figure. Despite such alterations in visuoconstructive praxis, executive functions remained unaffected. However, abstract reasoning exhibited apparent difficulty. Additionally, domains such as attention, working memory, and episodic memory appeared largely preserved. Correlation analyses did not reveal a clear association between cognitive impairments and motor function. **Conclusion:** This study highlights the intricate neuropsychiatric manifestations observed in adult DM1 patients, emphasizing visuoconstructive praxis as the most affected cognitive domain. Importantly, the study findings show that cognitive impairment in DM1 does not correlate with motor impairment. These findings reinforce the concept of DM1 as a systemic disease with complex central nervous system manifestations, which seem to be independent of the motor aspect of the disease.

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Cognitive and theory of mind impairment in sporadic and familial amyotrophic lateral sclerosis patients

Danilo Jorge da Silva[1], Sophia Calabria da Silveira[1], Fernanda Franklin Pedroso Moreira[1], Nadia Shigaeff[1], Marcelo Maroco Cruzeiro[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Danilo Jorge da Silva; Email: danilomachadosilva@gmail.com.

Background: Amyotrophic lateral sclerosis (ALS) is a rare neurological disease that leads to impairment of both lower and upper motor neurons, resulting in progressive weakness, muscle atrophy and respiratory and swallowing dysfunction. Despite well-documented motor symptoms, non-motor manifestations remain underexplored. **Objective:** To investigate the cognitive function in individuals with ALS in both sporadic and familial forms and its relationship with disease progression. **Methods:** Observational, cross-sectional, controlled study based on primary data of ALS patients over 18 years old, without a prior diagnosis of dementia. Functionality and neuropsychological assessment battery were measured by the following instruments: ALS Functional Rating Scale Revised (ALS-FRS-R), ALS Cognitive Behavioral Screen (ALSCBS), semantic verbal fluency test, phonemic verbal fluency test, digit span subtest from the Wais III scale, Hopkins Verbal Learning Test-Revised (HVLT-R), five-point test, and Mini-Mental State Examination (MMSE). We also applied the social cognition/theory of mind tests of Faux Pas and Theory of Mind – 15 (ToM-15). **Results:** Twenty-two ALS patients and 24 healthy controls were recruited. Among the patients, nine (40.9%) had the sporadic form and 13 (59.1%) had the familial form. Significant differences were observed in MMSE and ALSCBS scores between the ALS and control groups, with worse scores associated with the ALS group, and these differences were more evident among those with the familial form when compared to controls. Digit span, HVLT-R (total recall), five-point test, and ALSCBS were inversely related to ALS-FRS-R scores suggesting worse cognitive impairment in individuals with greater functional impairment related to ALS. Significant differences were also observed between the ALS and control groups in the performance of social cognition tasks measured by the Faux-Pas intention, comprehension, false beliefs, empathy, and total score. The ALS group (more evidently the familial ALS patients) had worse performance in social cognition tasks. This same pattern was also observed in the ability to detect false beliefs using the ToM-15 test with no evidence of significant relationship between ALS-FRS-R and social cognition impairment, suggesting a possible early compromise of theory of mind abilities. **Conclusion:** Individuals with ALS tend to show alterations in multiple cognitive domains, not all of them directly related to functional decline. Cognitive impairment was more pronounced in individuals with the familial form compared to the sporadic form, and there is evidence that social cognition impairment may occur early in the disease in this population.

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Cognitive assessment in patients with spinocerebellar ataxia type 6

Fernanda Mary Machado[1], José Luiz Pedroso[1], Orlando Graziani Povoas Barsottini[1], Marcondes Cavalcante Franca Júnior[2]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Fernanda Mary Machado; Email: marypsico.nanda@gmail.com.

Cerebellar cognitive-affective syndrome proposes that patients with cerebellar lesions present not only ataxic symptoms, but also important deficits in cognition, affect, and behavior. Cognitive, behavioral, and affective deficits, found in cerebellar cognitive-affective syndrome, may occur due to a process of diaschesis. Spinocerebellar ataxia type 6 (SCA6) is described as the prototype of a pure cerebellar ataxia. The present study aims to provide data on the contribution of corticocerebellar functional connections to clinical manifestations in a population of subjects with SCA6. It is a qualitative-quantitative, observational and cross-sectional research. 20 subjects (10 with ataxia and 10 controls) will be evaluated. All of those evaluated had an IQ greater than 70. Subjects will be matched by age group and education. This work is ongoing, therefore, 6 individuals from the control group and 6 individuals with a confirmed diagnosis were selected. In terms of intelligence quotient, 5 of the 6 patients evaluated with a diagnosis of type 6 ataxia obtained results below those of the control group. On average, there was a difference of 18.8 points. Regarding selective attention, worse results were obtained in the group of diagnosed, the differences ranged from 1 second to 7.63 seconds, with a mean value of 3.38 seconds. The evaluation of inhibitory control indicated a longer response time in the group of diagnosed, with a mean difference of 105.48 seconds versus 45.69 seconds, respectively. On average, patients diagnosed with ataxia had a difference of 8.16 raw points versus 12.33 in verbal long-term memory. When obtaining the average result, in relation to logical reasoning ability, there were higher results among the control group, in which they obtained an average of 13.3 points versus a result of 11.3 compared to the group of diagnosed patients. Regarding perception, the control group also obtained results higher than those obtained by the diagnosed group, which were 18.83 points versus 14.83 points, respectively. In relation to visuoconstructive ability, there were higher scores in the control group, in which they obtained an average of 45 raw points versus 30.6. In terms of cognitive flexibility, there was a score of 6.22 mean points versus 4 points between the diagnosed group and the control group. Regarding image naming, the control group obtained 56 points and the ataxia group 52.66 points. There was a difference in relation to the pertinent result to the working memory ability of the visual sphere. The control group achieved an average score of 8.66, compared to an average score of 7.33 points in relation to the diagnosed group. The presented data suggest cognitive affective syndrome of the cerebellum in type 6 ataxia. Which leads to the question of whether type 6 ataxia is in fact entirely pure, since it underscores a variety of cognitive changes in different areas.

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Cognitive changes in patients with concentric Baló sclerosis: a systematic review

Regina Maria Papais Alvarenga[1], Diana Góes de Souza[1]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Diana Góes de Souza; Email: dianagoes.cortex@gmail.com.

Background: Baló's concentric sclerosis (BCS) is a rare subtype of idiopathic inflammatory demyelinating disease of the central nervous system, characterized by a pattern of alternating and concentric layers of white matter preservation and demyelination, shaped like "onion rings". The disease affects young individuals, preferably women and has radiological and clinical presentations close to those of multiple sclerosis (MS). Since the emergence of magnetic resonance imaging (MRI), an increasing number of BCS cases have been diagnosed at an early stage and it has become possible to quickly identify the disease, with effective treatments using corticosteroids, which prolonged the survival of patients with BCS. **Objective:** To integrate the findings of a systematic literature review and describe clinical characteristics of BCS cases with

cognitive impairments. **Methods:** An electronic search was carried out in the PubMed database with the terms "Balo Concentric Sclerosis or concentric sclerosis or Balo's disease and Magnetic Resonance" between the years 1989 and 2024, written in English, French, Italian and Spanish, in article format original or case study, with humans. 217 articles were initially identified in the search and 34 were sellected. **Results:** What is generally described in case studies with BCS patients are the clinical symptoms, the age at onset of the disease, the monophasic or recurrent course, drug treatments performed and the MRI findings. Among the different symptomatological presentations of the disease is cognitive impairment. 19 patients were identified with cognitive decline among 65 BCS case reports reviewed (29%), which mainly presented language impairments, memory deficits and attentional changes. An SPSS analysis indicated that man predominated (n=11; 58%) and the mean age of onset was 34.47 years, ranging from 10 to 54 years. The recurrent clinical course was identified in 5 cases (26.3%) and 14 were monophasic (73.7%). Clinical treatment with corticosteroids was administered in 15 cases (79%) and 4 patients did not take any medicine (21%). Regarding the outcome, 13 cases improved (68.4%), 5 had sequels after the events (26.3%) and 1 had no information about the evolution of the symptoms. Among the cases reviewed with cognitive decline, language impairments occurred in 13 cases (68.4%), memory deficits in 4 (21%) and attentional changes in 2 (10%). **Conclusion:** This research achieved the main objective since all 19 cases reviewed that presented with cognitive declines could be identified per type (language, memory and attention), which reinforces the importance of future studies investigating phenotypes of cognitive changes in patients with BCS. It was observed that, differing from socio demographic research with BCS patients, in this systematic review male cases predominated and it could be investigated if cognitive impairment occurs more in man.

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Cognitive decline in patients with Chagas and non-Chagas etiology heart failure: a prospective cohort study

Israela Souza Brito Santos[1], Lucy Rodrigues Ribeiro[1], Cárita Victória Carvalho de Santana[1], Bruna Helena Ribeiro Araújo dos Santos[1], Janaina Brito de Pinho[1], Júlia Barreto de Farias[1], Bruna Guilherme Martendal[1], Beatriz Malaquias Neves[1], Graziele Carolina Araújo Soares[1], Jamary Oliveira Filho[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Israela Souza Brito Santos; Email: israela.souza@ufba.br.

Background: Cognitive impairment in Chagas disease (CD) remains understudied. Patients with chronic CD may exhibit varying levels of cognitive dysfunction; however, the longitudinal evolution of this dysfunction over time is not well understood. **Objective:** To investigate the association between CD and cognitive dysfunction in patients with heart failure (HF), and to assess cognitive decline over time. **Methods:** Prospective cohort study involving adult individuals with heart failure from 2021 to 2024. Exclusion criteria included history of cancer (except skin), stroke, end-stage renal or hepatic disease, and other neurodegenerative conditions. Cognitive assessment tools included: Mini-Mental State Examination, Brief Cognitive Screening Battery, Luria's test of motor sequence, Rey Complex Figure Test, and Digit Sequencing Task, grouped into four cognitive domains (Z-scores): memory, visuospatial, executive, and global. A repeat cognitive assessment identical to the first was conducted at least 2 years later. Z-scores ≤1.5 indicated cognitive dysfunction in each domain. Logistic regression adjusted for age, sex, and education level compared the proportion of cognitive dysfunction between individuals with and without CD. **Results:** A total of 86 participants were included, with 54 having Chagas etiology HF and 32 having other etiologies. Compared to participants without CD, those with CD were predominantly older (53.2±12.7 without CD vs. 57.1±7.6 with CD), female (15 [48%] without CD vs. 39 [84.7%] with CD), and had incomplete primary education (14 [45%] without CD vs. 31 [67.3%] with CD). Cognitive dysfunction was predominantly observed in Chagas patients (vs. non-Chagas) in the executive function domain, present in 37.0% vs. 15.6%, respectively, p=0.034. After a mean follow-up of 6.4 (±3.1) years, executive function domain dysfunction persisted in 29.6% vs. 12.5% of these patients, respectively. **Conclusion:** Patients with Chagas etiology HF exhibit greater cognitive deficits, predominantly in the executive function domain, compared to patients with other cardiomyopathies. However, the degree of cognitive dysfunction appears to remain stable over time.

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Cognitive impairment following leptospirosis encephalitis: case report and literature review

Nathan Augusto Müller[1], Lucas Ziegler Dalenogare[1], Nathália Oberto Câmara[1], Kathy Aleixo Marcolin[1], Mauricio Scopel Hoffmann[2], Daniel Pascotto de Oliveira[2], Kelly de Oliveira Harada[1], Juliana Oliveira Freitas Silveira[3], George Vasconcelos Calheiros de Oliveira Costa[3], Alba Letícia Peixoto Medeiros[4]

[1]Universidade Franciscana, Santa Maria RS, Brazil.
[2]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.
[3]Hospital Universitário de Santa Maria, Santa Maria RS, Brazil.
[4]Centro Universitário Cesmac, Maceió AL, Brazil.

Correspondence: Juliana Oliveira Freitas Silveira; Email: julianaofs@yahoo.com.br.

Case presentation: A 67-year-old woman was admitted to the hospital after episodes of epileptic seizures and mental confusion. She reported anxiety, disorientation, and headaches for the past 15 days. On initial examination, she was confused, drowsy, with signs of meningeal irritation. The cranial nerves were preserved. No alteration in strength or reflexes were found. Glasgow score was 13. A cranial MRI revealed extensive areas of T2/FLAIR hypersignal, severe microangiopathy, and pial enhancement by contrast in the right temporo-parieto-occipital region, in addition to positive Ig M serology for leptospirosis. She was treated with ampicillin and sulbactam. On the 3rd day of admission, there was a worsening of consciousness level, recurrent epileptic seizures, hyponatremia, and worsening renal function, leading to orotracheal intubation and mechanical ventilation for status epilepticus treatment. After 30 days hospitalized, she showed clinical and neurological improvement and was discharged with phenytoin and levetiracetam medication. Prior to hospitalization, she used lithium and sertraline to treat her bipolar affective disorder (BAD) diagnosed in her youth. After discharge, she began to presente cognitive complaints. During the neurologist consultation, the Montreal Cognitive Assessment (MoCA) test was 18 out of 30 points. The diagnostic hypothesis was multifactorial cognitive impairment secondary to deterioration of BAD associated with sequelae of leptospirosis encephalitis and vascular lesions. **Discussion:** Encephalitis is an acute inflammation of the central nervous system (CNS), specifically of the brain parenchyma, often caused by viruses such as influenza, herpes simplex, arboviruses, and in endemic areas like Brazil, dengue, Zika and Chikungunya viruses can also be pathogenic agents. Less commonly, bacterial pathogens such as Leptospira species can lead to this clinical manifestation. Leptospirosis is a zoonosis of significant global concern, prevalent in tropical climates and associated with lower economic levels. The disease is transmitted through contact of an open wound with the urine of the transmitting animal and can manifest in various clinical forms, although neurological presentation is uncommon (about 10-15% of Leptospirosis cases). The CNS can be affected by this zoonosis either through direct infection of the brain parenchyma or secondarily through hepatorenal involvement. This CNS involvement may manifest as meningitis or encephalitis. **Final comments:** Therefore, due to encephalitis being a medical emergency and this manifestation of Leptospirosis being considered rare, many physicians may not consider this possibility and may mismanage the patient or even miss the appropriate time to start treatment to prevent more severe sequelae. It is essential that, especially in the most prevalent areas of this significant zoonosis, this clinical manifestation must be adequately investigated and treated promptly.

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Cognitive impairment: clinical presentation of an immunocompetent patient with neurosyphilis

Paulo Henrique Martinelli Oliveira[1], Amanda Michelucci dos Santos[1], Larissa Miyashiro[1], Raderi Luiz Cardoso Do Santos[1], Lucas Silva Dias[1], Laiz Helena de Pádua[1], Gustavo Carvalho Costa[1], Rebeca Aranha Barbosa Sousa[1]

[1]Hospital Municipal Dr. José de Carvalho Florence, São José dos Campos SP, Brazil.

Correspondence: Paulo Henrique Martinelli Oliveira; Email: paulohmoliveira96@gmail.com.

Case presentation: Patient R.P.S, male, 47 years old, previously diagnosed with depression. About two years ago, sought care with a headache, temporal and spatial disorientation, and cognitive impairment, evolving with inability to accomplish activities of daily living without assistance. The patient was diagnosed with syphilis (serologic VDRL 1/64) and received treatment for late latent stage, with benzathine penicillin G intramuscularly in 3 weekly doses. After treatment, the patient persisted with cognitive impairment and infatilized behavior. On May, 2024, was brought to the emergency care unit after first epileptic seizure with secondary generalization. There was no recurrence of seizure since admission. Montreal Cognitive Assessment (MoCA) was performed, with score 15/30. The initial brain computerized tomography was normal. The laboratory revealed syphilis rapid diagnostic test positive, serologic VDRL 1/32, anti-HIV negative, and reactive VDRL in cerebrospinal fluid. The patient underwent treatment with intravenous crystalline penicillin for 21 days. After therapy, his MOCA score was 19/30. He was discharged and referred to the Cognitive Neurology clinic. **Discussion:** Neurosyphilis can occur at any stage of syphilis. Clinical manifestation range from asymptomatic, meningeal, meningovascular, to tabes dorsalis and general syphilitic paresis. Recognizing the neurological manifestations is important, as the treatment differs from that of syphilis without neurological involvement. Failure to diagnose and inadequately treat neurosyphilis can cause severe and permanent damage to the patient. Without proper treatment, the infection can progress, resulting in severe neurological symptoms such as headaches, confusion, loss of vision, hearing, and motor control. The patient may develop dementia, paralysis, personality changes, and psychiatric problems. These complications can lead to a significant reduction in quality of life, inability to work, and increased dependence on medical care and social support. In extreme cases, untreated neurosyphilis can be fatal. **Final comments:** Neurosyphilis can be a diagnostic challenge due to its variety in clinical presentation, and it should be considered as a differential diagnosis in patients with cognitive impairment.

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Cognitive multisensory rehabilitation (cmr perfetti): improving motor function in chronic stroke patients

Rafael Correa Fantato[1], Julian Pablo Gorosito[1], Mauro Cracchiolo[1], Mara Cristina Santos Melo[1], Fernanda Devecchi Prado[1], Patricia Simone Lopes de Souza[1], Ivana Brito[2]

[1]Instituto Avencer
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Rafael Correa Fantato; Email: tnc@institutoavencer.org.

Motor recovery following a stroke is a multifactorial and dynamic process influenced by various factors including age, ethnicity, socioeconomic factors, genetic traits, initial injury severity, and time since the stroke occurred. Neurorehabilitation strategies play a pivotal role in facilitating recovery but must account for the considerable variability among patients, making a standardized intervention protocol challenging to develop. Therefore, it is essential that rehabilitative reasoning prevails to guide individualized approaches to rehabilitation. Advancements in our understanding of neural plasticity have transformed the prognosis for post-stroke patients, emphasizing continuous improvement. Cognitive multisensory rehabilitation (CMR), also known as the Perfetti method, is a motor learning model that emphasizes high-level cognitive function and occurs through the integration of perception-cognition-emotional processes, such as motor regulation. Activation of the brain's cognitive process, which represents perception, memory, attention, language, and decision-making, is the core of the training. **Case presentation:** Case 1: A 37-year-old woman, previously healthy, developed spastic hemiplegia on the right following an ischemic stroke caused by dissection of the left internal carotid artery in June 2015. She received conventional physiotherapy treatments until February 2018, after which she underwent CMR Perfetti treatment until July 2023. Case 2: Man, 51 years old, with congenital visual impairment, presents with ataxic tetraparesis with brachial and left predominance, due to hemorrhagic cerebellar stroke in January 2015. He was followed up with conventional physiotherapy treatments until November 2020, when he began treatment with CMR Perfetti. For Case 1: There was a notable 22.2% improvement in MESUPES-hand and a 5% improvement in MESUPES-arm scores, indicating enhanced quality of movement in the paretic upper limb. Tardieu scale showed increased range of motion was observed in shoulder flexors, extensors, adductors, and medial rotators, as well as elbow flexors and extensors. Thumb opponency improved by 2cm. For Case 2: There was a significant 22.5% improvement in MESUPES-arm score, reflecting enhanced quality of movement in the affected arm. Range of motion gains exceeded 50% for shoulder adductors, wrist extensors, and index finger flexors (Tardieu). Physical domain of quality of life (WHOQOL) improved by 10%. **Discussion:** These improvements highlight the effectiveness of CMR Perfetti treatment in enhancing motor function, range of motion and quality of life in both cases, despite their distinct initial conditions and challenges. **Final comments:** Advancements in our understanding of neural plasticity, along with the recognition of the importance of consciously constructing the necessary information for action, have transformed the prognosis for post-stroke patients, emphasizing continuous improvement.

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Cognitive symptoms in long COVID patients: clinical and epidemiological analysis

Emanuel de Assis Bertulino Martins Gomes[1], Antônio Alves Sobreira Neto[2], Chiara Gübel Portugal[2], Lorenzo Marinho Morais[2], Samuel Cavalcante Marinho[2], Álissa Elen Formiga Moura[1], Thaís de Maria Frota Vasconcelos[1], Marcello Holanda de Andrade[1], José Wagner Leonel Tavares Júnior[1], Pedro Braga Neto[1]

[1]Universidade Federal do Ceará, Complexo Hospitalar, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Emanuel de Assis Bertulino Martins Gomes; Email: emanueldeassis@alu.ufc.br.

Background: During the COVID-19 pandemic, some patients presented neurological symptoms in the acute phase of the disease, such as dizziness and headaches. Subsequently, many patients reported long-term cognitive effects of SARS-CoV-2, a phase known as "Long Covid", with mild to severe effects on quality of life. **Objective:** To describe and compare clinical and epidemiological data from patients who developed long-term cognitive complaints after COVID-19. **Methods:** This is a prospective, cross-sectional study of patients previously diagnosed with COVID-19. We used data from the project "Rede Nordeste de Neurocovid Longo", a cohort investigating neurological alterations in "Long Covid". All patients signed an informed consent form to take part in this research. The participants were evaluated at a university hospital complex in Fortaleza city by a neurologist, using scales for cognitive and functional assessment, such as the Mini-mental state examination and the Pfeffer scale. In addition, blood samples were taken to analyze APOE gene and neurodegeneration markers. Data analysis was conducted using Jamovi software. Mann-Whitney U and Kruskal-Wallis tests were used for comparative analysis. **Results:** From March 2023 to April 2024, 136 patients were assessed, with a mean age of 42.79 years (±13.21) and a predominance of women (77.2%). Cognitive complaints were reported by 128 (94.11%) participants, who presented an average age of 42.93 years (±13.26) and 78.12% female patients. In the cognitive assessment, 5.88% of the patients showed no alterations, 86.76% had subjective cognitive decline (SCD), 5.14% had mild cognitive impairment (MCI) and 2.20% had dementia. Regarding related symptoms, cognitive complaints were mainly associated with sleep disorders (76.56%), anxiety (61.71%) and headaches (37.50%). In relation to total patients, 40 (29.41%) had more than one SARS-CoV-2 infection and 22 (16.17%) were hospitalized for COVID-19, of which 16 (11.76%) were admitted to the Intensive Care Unit (ICU). A comparative analysis was carried out between the information described and the clinical cognitive assessment, in which patients were described ordinally as "Unaltered", "SCD", "MCI" or "Dementia". A significant association was seen in comparison with the variables Hospitalization (p=0.022), ICU admission (p=0.007) and Anxiety (p=0.023). **Conclusion:** The epidemiological profile of patients with "Long Covid and cognitive complaints consisted mainly of women and presented a mean" age of 42.93 years. In this group, the majority of participants had Subjective Cognitive Decline according to the clinical assessment. In addition, cognitive symptoms were mainly accompanied by sleep disorders and anxiety. Finally, comparative analyses showed an association between cognitive decline and hospitalization, ICU admission and anxiety.

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Cognitive training associated with transcranial direct current stimulation in the treatment of age-related mild cognitive impairment: a systematic review of randomized clinical trials

Felipe Miranda Camboim[1], Daniel Macedo de Oliveira[1], Flávia Blank[1], João Guilherme Gomes Pedrosa[1], João Vitor Andrade Fernandes[1], Rebeca Oliveira da Silva[1], Isabella Araujo Mota Fernandes[2]

[1]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[2]Hospital Universitário Lauro Wanderley, João Pessoa PB, Brazil.

Correspondence: Felipe Miranda Camboim; Email: felipemirandacamboim@gmail.com.

Background: Cognitive training (CT) is an established therapy for cognitive decline in ,ild cognitive impairment (MCI), however, there are other non-pharmacological alternatives, such as transcranial direct current stimulation (tDCS). A novel approach suggests combining both CT and tDCS, in an attempt to augment its effects on patient cognitive performance. **Objective:** To synthesize evidence on the efficacy of combined cognitive training and transcranial direct current stimulation treatment of age-related mild cognitive impairment. **Methods:** A systematic review was conducted using the following databases: Medline, Embase, Cochrane, Web of Science, and BVS. The search terms utilized included "Transcranial Direct Current Stimulation," "Mild Cognitive Impairment," and "Adult," as well as their entry terms, combined with Boolean operators OR and AND. Only randomized controlled trials (RCTs) were included in the review. No language or date filters were applied during the search. The studies were independently analyzed by two reviewers according to eligibility criteria. The articles were screened using a computerized review software and the data obtained was organized in an online spreadsheet. The inclusion criteria were randomized controlled trials that compared the use of tDCS combined with CT versus sham-tDCS in adult patients with age-related MCI. The intervention group in the selected studies involved the use of tDCS with electrodes positioned in the left dorsolateral prefrontal cortex with a direct current of 2mA, on average, combined with computerized cognitive training. **Results:** A total of 381 articles were found, of which 12 studies were included, according to eligibility criteria, for analysis, totaling 440 patients. None of the studies included were conducted in South America. Overall, the tDCS + CT group obtained a greater improvement in the parameters analyzed (everyday memory, executive functions, working memory) compared to the sham tDCS group. However, most of the studies did not find statistically significant differences between the groups analyzed. **Conclusion:** Although tDCS combined with cognitive training was able to improve patient's overall cognitive performance, these results were not statistically significant in comparison to controls. However, study methods diverged substantially, especially regarding the protocol for application of active and sham tDCS.

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Coinfection of neurotuberculosis and neurocryptococcosis in an immunocompetent patient: a case report

Lucca Ferdinando Queiroz Fernandes[1], Maria Eduarda Medeiros Martins[1], Diógenes Emanuel Dantas da Silva[1], Matheus Araújo de Medeiros[1], Artur Rebouças de Souza[1], Tiago Lameque de Sousa e Silva[1], Helmut Kennedy Azevedo Do Patrocínio[1], Lucas Medeiros Dunga[1], Iury Hélder Santos Dantas[1], Fernanda Gurgel de Oliveira[1]

[1]Universidade Federal do Rio Grande do Norte, Caicó RN, Brazil.

Correspondence: Lucca Ferdinando Queiroz Fernandes; Email: queirozlucca@gmail.com.

Case presentation: A 52-year-old woman with no comorbidities, presented with asthenia and weight loss, accompanied by paresis in the right lower limb, initially diagnosed as depressive disorder. After 3 months, developed tetraparesis, gait ataxia, dysarthria, fever and psychomotor slowing. Nuclear magnetic resonance (NMR) with contrast showed leptomeningeal enhancement in the basal cisterns, Sylvius and interhemispheric fissure, brainstem and cerebellar sulci, with extension to the spinal cord and some roots of the cauda equina. Diffusion restriction in T2/FLAIR was observed in the pontine region on the left and the nucleocapsular region on the right. Findings suggestive of neurotuberculosis with ischemic events due to associated vasculitis. The cerebrospinal fluid (CSF) showed mild pleocytosis with a lymphomononuclear

predominance, hyperproteinorrachia and hypoglycorrachia. Fungoscopy, bacilloscopy, TRM-TB in the CSF were negative. A chest CT revealed a calcified granuloma in the middle lobe, with no signs of active pulmonary tuberculosis (TB). Treatment for TB with RHZE and corticotherapy was started, with clinical improvement and discharge after 2 months. One week after discharge, she was readmitted with worsening motor skills, dysarthria, fever and headache. A new MRI showed increased leptomeningeal enhancement. Serial lumbar punctures revealed increased opening pressure, mild lymphomononuclear pleocytosis, hyperproteinorrachia, hypoglycorrachia, negative bacilloscopy and TRM-TB, but positive fungoscopy. A Lateral Flow Assay for cryptococcal antigen (LF-CrAg) was carried out and the serum and CSF were reactive. Liposomal amphotericin B and flucytosine were started, as well as daily CSF punctures for relief, with a good response. Control MRI showed a slight reduction in leptomeningeal enhancement and no new ischemic events. Serologies for HIV and HTLV during hospitalization were non-reactive, with an undetectable HIV viral load. She continues to be followed up as an outpatient, clinically stable, under treatment with RH, fluconazole and weaning off corticosteroids. **Discussion:** Although both neurotuberculosis and neurocryptococcosis are diseases with a high incidence in HIV-AIDS patients, coinfection is rare. Even rarer in immunocompetent patients. This case reports the coinfection of these two infections in a patient with no known immunosuppression. The clinical-laboratory presentation and imaging findings were initially suggestive of TB, a diagnosis reinforced by the initial response to treatment with RHZE and corticotherapy. The subsequent clinical worsening, however, raised the suspicion of a second infection, confirmed by positive fungoscopy and a reactive LF-CrAg test. **Final comments:** Coinfection of neurotuberculosis and neurocryptococcosis is rare, especially in immunocompetent patients. Both infections can share similar epidemiological and clinical characteristics, requiring high clinical suspicion for the diagnosis of coinfection.

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Collet-sicard syndrome: a clue to occult malignancy in an elderly female - a case presentation

Guilherme Lago Miranda[1], Erick Dupont[1], Ingrid Lorena da Silva Gomes[1], Henrique de Moraes Bernal[1], Giovanni Caniçali Simões[1], Pedro Almeida Martins Pontes[1], Tainá Fátima Ramos Gonçalves[1], Matheus Reis Rosa[1], Marcele Schettini de Almeida[1], Sonia Maria Cesar de Azevedo Silva Moura Magalhães Gomes[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Guilherme Lago Miranda; Email: lagomiranda.g@gmail.com.

Case presentation: A 69-year-old female patient with no previous relevant diseases was admitted to the emergency room of a tertiary hospital complaining of twinging neck pain that had been ongoing for 2 days. She also mentioned dysphagia, and noticed slurred speech and hoarseness. On neurological examination, she had dysphonia and dysarthria that were worse for palatal sounds; the palate was asymmetrical, with a dropped velum on the right and reduced gag reflex on the same side (cranial nerves - CN IX and X); the sternocleidomastoid and trapezius on the right had IV+ strength grade, and contralateral strength was preserved (XI CN); at rest, her tongue was deviated to the left, with deviation to the right on protrusion (XII CN). The rest of the exam was unremarkable. A brain MRI revealed a lesion involving the bones of the right skull base (occipital and small adjacent inferior temporal area), with an erosive nodular image with heterogeneous enhancement by the contrast that measured 2.5 x 2.0 cm. Her head CT revealed a 28 mm paramediastinal nodule in the upper lobe of the right lung associated with a subpleural 14 mm nodule. Hilar and mediastinal lymph node enlargements and osteolytic lesions in bilateral costal arches were also seen, as well as right hemilaryngeal paralysis. **Discussion:** With the data above, the involvement of CN IX, X, XI and XII on the right side was established, which is consistent with the diagnosis of Collet-Sicard syndrome. It is a rare neurological condition resulting from skull base lesions that affect the posterior lateral condylar space, injuring the bulbar cranial nerves. It can be linked to neoplastic, infections, trauma and vascular causes. In this case, the lung nodulation was biopsied and found to be a carcinoma with neuroendocrine differentiation, and the occipital lesion was considered a probable site of metastasis. The patient had no known history of oncological diseases, and the syndrome described above was the manifestation that led her to seek the health service, allowing the diagnosis of the underlying disease and the beginning of treatment. **Final coments:** This case presentation demonstrates the value of considering Collet-Sicard syndrome in patients presenting with the characteristic combination of cranial nerve palsies. Even more importantly, the syndrome's role in unveiling an occult malignancy is further emphasized.

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Communication problems are not associated with cognitive decline in people with Parkinson's disease

Camila Pimentel Piemonte[1], Katia Cirilo Costa Nobrega[1], Isaíra Almeida P. S. Nascimento[1], Natalia Cardoso Penha[1], Clara Greif Cerveira de Paula[1], Maria Elisa Pimentel Piemonte[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Camila Pimentel Piemonte; Email: camila.piemonte@usp.br.

Background: Parkinson's disease (PD) is the second neurodegenerative disorder more prevalent globally, typically presenting around age 60. The progressive neurodegeneration process in basal ganglia leads to motor and non-motor symptoms responsible for decreased quality of life. Due to the degenerative process, people with PD present progressive communication difficulty associated with motor symptoms, such as hypokinetic dysarthria and voice variation. Besides the voice dysfunction, cognitive alterations and depression have also been associated with communication impairments. Aim: The present study aims to investigate the association between communication function and motor symptoms severity and mental alterations in people with PD. **Methods:** A cross-sectional study with a convenience sample of 423 (201 women) people with idiopathic PD, mean age 57.98±10.9, in early to advanced disease stages (I-IV stages according to H&Y rating scale) and 4.21±0.96 years of schooling. All participants were evaluated in individual sections by trained researchers. The self-reported communication problems were assessed by the Communication Domain of the Parkinson's Disease Questionnaire (PDQ-39); the global cognitive capacity by Montreal Cognitive Assessment (MoCA), depression by Geriatric Depression Scale (GDS), and motor symptoms severity by Unified Parkinson's Disease Rating Scale (UPDRS III). **Results:** There was a significant difference in self-reported communication problems between people with PD in advanced (H&Y IV) and moderate and early disease stages (H&Y I-III). The self-reported communication problems were correlated with UPDRS III but not age, gender, MoCA, and GDS. **Conclusion:** The communication impairments increase in advanced stages of PDD and are associated with the severity of motor symptoms but not with cognitive decline and depression. Interventions to improve communication skills in people with PD should focus on motor alterations associated with the phonatory process.

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Companion's knowledge on stroke and symptoms-to-door time in reference hospital

Bárbara Nascimento Moutinho[1], Mário Luciano de Mélo Silva Júnior[1], João Pedro Wanderley Soares[1], Mariana Leandro Silva[2], Karyne de Souza Nobre[3], Renata Maria de Melo Moraes[3], Teresa Virgínia Macedo de Aquino[3], Yasmin Gabrielle Pereira Pacheco Cavalcanti de Albuquerque[3], Ilo de Vasconcelos Ximenes Júnior[1], Bárbara Francisca Do Nascimento[3]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil.
[2]Faculdade de Medicina de Olinda, Olinda PE, Brazil.
[3]Centro Universitário Maurício de Nassau, Recife PE, Brazil.

Correspondence: Bárbara Nascimento Moutinho; Email: barbara.moutinho@ufpe.br.

Background: Cerebrovascular diseases were responsible for over 107,000 deaths in Brazil in 2022, according to DATASUS data. Rapid recognition of a stroke can increase the likelihood of reaching medical services within a narrow therapeutic window for effective treatment. Previous studies indicate that patients' prior knowledge of stroke was not a predictor of faster arrival at the emergency department, however, studies examining the relationship between companions' prior knowledge of stroke and the time taken for patients to reach reference services after stroke symptoms are scarce. **Objective:** To evaluate previous knowledge and clinical-epidemiological characteristics of companions of patients with ischemic or hemorrhagic stroke and its relation with the symptoms-to-door time. **Methods:** Cross-sectional study involving companions of patients diagnosed with ischemic or hemorrhagic stroke, admitted in a tertiary care center. Data collection was performed using an electronic form developed by our team and applied to the patients' companions, with objective questions about sociodemographic data, personal history, knowledge of warning signs, risk factors for stroke, Mobile Emergency Service (SAMU) telephone number, and we also collected the symptom-to-door time. We considered an optimal symptom-to-door time when patients arrived in up to 270 minutes. **Results:** The sample consisted of 129 companions of patients with acute stroke [98 women, 76%; median age = 45 (IQR=32.5-54) years, with 12 (IQR=7-12) years of formal education, and 24 (18.6%) had health insurance]. About 2/3 (88/129, 68.2%) were the patients' children or partners. Regarding knowledge of stroke, 59 (45.7%) knew the organ involved in a stroke; 74 (57.4%) knew the SAMU hotline number; and 69 (53.3%) knew at least one risk factor for stroke. Thirty-nine (30.2%) patients/companions arrived in optimal time. We did not find any difference between arriving in or out the optimal time regarding sex, having health insurance, education level, having previously cared for someone with a stroke, knowing the organ damaged in a stroke, the SAMU number, or warning signs for stroke; there was also no difference in the NIHSS or Glasgow scores at admission, nor in the perception of disease severity. Knowing at least one risk factor was more frequent in the optimal time group of companions (71.8% vs 45.6%, p=0.007), and this group was older (median 48 vs 42 years, p=0.025). **Conclusion:** Knowledge of risk factors and older age of the companions are associated with a shorter arrival time of patients with acute stroke. Other knowledge about stroke (damaged organ, SAMU number, and warning signs) was not related to arrival time.

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Comparative analysis of emergency assistance for stroke in the interior of Pernambuco to national guidelines

Maria Laura Lima da Silva[1], Maria Vitória Do Nascimento Silva[1], Jéssica dos Santos Coelho[1], Joan Lins Serafim[1], Thiago Bezerra dos Anjos[1], Ana Luiza Sampaio Lopes[1], Américo Danúzio Pereira de Oliveira[1], Rita Di Cassia Oliveira Angelo[2]

[1]Universidade de Pernambuco, Serra Talhada PE, Brazil.
[2]Universidade de Pernambuco, Garanhuns PE, Brazil.

Correspondence: Maria Vitória do Nascimento Silva; Email: vitoria.nsilva@upe.br.

Background: Stroke is a medical emergency associated with significant morbidity and mortality, considered the second leading cause of death and disability worldwide. Given the urgency of stroke care, several protocols have been established. In Brazil, the Ministry of Health recommends the use of guidelines from the Brazilian Society of Cerebrovascular Diseases (SBDCV) for emergency stroke care. **Objective:** To describe the emergency care provided to stroke cases at an emergency service in Serra Talhada, PE, and to conduct a comparative analysis with SBDCV guidelines. **Methods:** Cross-sectional descriptive study, including stroke cases admitted at a state public health hospital, a reference in Serra Talhada, PE. Data collection utilized individual registration forms and self-reported health condition questionnaires from the Unified Health System (SUS), as well as an evaluation form developed according to the acute stroke emergency protocol recommended by SBDCV. **Results:** The sample of 194 patients, predominantly male (n=106; 54.6%), Black race/ethnicity (n=140; 72.2%), aged between 61-80 years (n=99; 51%), farmers (n=91; 46.9%), and retirees/pensioners (n=138; 71.1%). Regarding comorbidities and lifestyle habits, the majority were hypertensive (n=158; 81.4%), diabetic (n=97; 50.0%), sedentary (n=170; 87.0%), and did not follow a balanced diet (n=153; 78.9%). Approximately 36% (n=70) reported a previous stroke. The clinical parameters assessed during initial care, the majority had their blood pressure measured (n=180; 92.8%); however, capillary blood glucose (n=72; 37.1%), oxygen saturation (n=70; 36.1%), and temperature (n=103; 53.1%) were not consistently evaluated. Among laboratory tests, complete blood count was requested for 88.6% (n=168), but creatinine (n=51; 26.3%), electrolytes (n=54; 27.8%), fasting blood glucose (n=131; 67.5%), and coagulation studies (n=173; 89.2%) were often omitted. Regarding etiological investigation, electrocardiograms were requested in 68.6% (n=133) of cases. However, trans-thoracic echocardiograms were not assessed in 83.0% (n=88) and carotid Doppler ultrasound in 74.7% (n=84). In terms of treatment, the majority used statins (n=136; 70.6%) and antihypertensive drugs (n=130; 67.0%). Antiplatelet therapy was administered using acetylsalicylic acid (n=131; 68.0%) and clopidogrel (n=16; 8.2%). A minority required anticoagulation; when administered, warfarin was used in only 1 case (0.5%) and new oral anticoagulants in 6 (3.1%). No patients underwent reperfusion therapy as the service is not equipped for it. **Conclusion:** Emergency stroke treatment has advanced significantly, but unevenly across Brazilian territory. In the interior of Pernambuco, the analyzed healthcare service facility deviates from the majority of SBDCV guidelines recommendations and require adjustment.

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Comparative analysis of prognosis between patients with wake-up stroke and patients with acute ischemic stroke with defined onset time

Gracchо Cardoso da Silva Neto[1], Joanna Sousa da Fonseca Santana[1], Ana Clara de Lemos Guimarães[1], Amanda Hellen Sousa Leal[1], Maria Clara Bastos de Sá[1], João Lucas Lima Diogo[1], Victor Vale da Silva[1], Brena Lima Cunha[1], Gabriela Morais de Medeiros Dias Melo[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Gracchо Cardoso da Silva Neto; Email: gracchoneto@icloud.com.

Background: The wake-up stroke is characterized by being a stroke that occurs without a precise onset time, as symptoms manifest upon awakening, representing 25% of ischemic stroke cases. Based on this, it's necessary to analyze the prognosis of these patients and compare it with patients with ischemic stroke with a defined ICTUS time, in order to reduce hospitalization costs with more appropriate treatments, to develop better diagnostic plans and reintegration into society. **Objective:** To analyze the differences in the prognosis of patients with wake-up stroke and patients with ischemic stroke with defined ictus times beyond the thrombolysis window. **Methods:** It's a retrospective and descriptive cohort study with 6,993 patients. The study was conducted based on secondary data extraction from electronic medical records of patients previously admitted to a public state hospital with focus on emergency services in Salvador, BA. The study population comprises patients diagnosed with stroke who were admitted between January 2022 and December 2022. The included variables were: age, sex, length of hospital stay, NIHSS at admission and discharge. We received approval from the Ethics Committee. The analysis was performed using SPSS and Excel, in a descriptive manner. This is a partial result of the STRIVE (Stroke Treatment and Recovery Investigation with Vascular Evaluation) research. **Results:** A total of 72 patients were analyzed, with 62 (86.1%) diagnosed with ischemic stroke beyond the window (iSBW) and 10 (13.9%) with ischemic stroke with unknown onset (Wake-Up). Regarding the primary outcome analysis, 9 patients in the iSBW (14.5%) and 2 in the wake-up group (20%) died. Concerning the length of hospital stay, patients in the iSBW group had a median stay of 9 days (minimum of 1 day and maximum of 150 days), while patients in the wake-up group had a median stay of 12.5 days (minimum of 3 days and maximum of 51 days). Regarding the NIHSS, patients were stratified as follows: minor stroke (MI; ≤4); moderate stroke (MO; >4 - ≤15); moderate severe stroke (MOS; ≥16 - <21); and severe stroke (S; >21). In the iSBW, there were 24 MI patients (38.71%), 11 MO (17.74%), 4 MOS (6.45%), and 4 S (6.45%). In the wake-up stroke group, there were 3 MI patients (30%), 4 MO (40%), 1 MOS (10%), and no MS patients. **Conclusion:** In our limited sample, patients with wake-up stroke had a higher mortality rate and a longer median hospital stay compared to patients in the iSBW group. Therefore, it is important to recognize the specific characteristics of wake-up stroke and develop tailored protocols to improve clinical outcomes and management of these patients. These findings are consistent with those observed in other studies, which also highlight the unique challenges and poorer prognoses associated with wake-up stroke. Further research is needed to better stratify these two groups of patients.

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Comparative analysis of the clinical and epidemiological profile of patients with ischemic stroke according to age in a referral hospital in Bahia

Amanda Hellen Sousa Leal[1], Joanna Sousa da Fonseca Santana[1], Ana Clara de Lemos Guimarães[1], Katarina Brito Gama[1], Gabriela Morais de Medeiros Dias Melo[1], Carla Loureiro Alvarez[1], Paula Beatriz Azevedo Marques[1], Bruno de Brito Guimarães[1], Larissa Nunes Almeida dos Santos[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Joanna Sousa da Fonseca Santana; Email: joannasantana20.2@bahiana.edu.br.

Background: Stroke is a leading cause of disability globally and the second leading cause of death in people over 60. Although it predominantly affects the elderly, the incidence among younger adults is increasing. Approximately 15% of strokes occur in adults under 45 years, termed early-onset stroke. This study aims to compare the clinical profiles of young and elderly stroke patients in Brazil to improve understanding and management of this condition. **Objective:** To compare the clinical profiles of stroke patients younger than 45 years with those aged 45 and older. **Methods:** This is a retrospective cohort. Data were collected from electronic medical records of patients previously admitted to a reference hospital in Salvador, Bahia, with a diagnosis of stroke between April 2015 and August 2023. The study included all patients diagnosed with ischemic stroke by a neurologist with available data. The variables of interest collected and analyzed using the software SPSS (Statistical Package for the Social Science). The study was approved by the Research Ethics Committee. **Results:** Among the patients aged 45 and older, there were 646 individuals, of which 47% were male and 52% female, with a median age of 69 years and a maximum age of 106 years. The average hospital stay for this group was 11.9 days. The most commonly affected arteries were the middle cerebral artery (19%), anterior cerebral artery (16%), and posterior cerebral artery (5%). Thrombolysis was performed in 9% of cases. The most common etiologies were undetermined causes (61%), followed by cardioembolic (18%), small artery occlusion (9%), large artery atherosclerosis (8%), and other etiologies (2.7%). Hypertension was prevalent in 83% of patients, diabetes mellitus in 45%, heart disease in 15.6%, and 19% had a history of previous stroke. For group under 45 years, there were 130 individuals with ages ranging from 14 to 45 years, with a median age of 40 years. This group consisted of 56.2% males and 43.8% females. The middle cerebral artery was most commonly affected (35%), followed by the anterior cerebral artery (3%) and posterior cerebral artery (4%). In terms of comorbidities, 33.1% of young patients had none, 50% had hypertension, 12% had diabetes mellitus, 3.1% had heart disease, and 6.9% had a history of previous stroke. **Conclusion:** The study reveals significant differences and similarities between young and elderly stroke patients. In young patients, stroke onset occurs between 14 and 45 years, with a median age of 40, and the middle cerebral artery being the most commonly affected. The prevalence of hypertension was high in both groups, while diabetes mellitus was more common among older patients, and the absent of comorbities more comun among the younger group. These findings underscore the necessity of tailored stroke management strategies that consider age-specific clinical profiles and risk factors to improve patient outcomes.

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Comparative analysis of the management and prognosis of ischemic cerebral stroke patients according to age in a referral hospital in Bahia

Joanna Sousa da Fonseca Santana[1], Amanda Hellen Sousa Leal[1], Ana Clara de Lemos Guimarães[1], Katarina Brito Gama[1], Gabriela Morais de Medeiros Dias Melo[1], Carla Loureiro Alvarez[1], Paula Beatriz Azevedo Marques[1], Larissa Nunes Almeida dos Santos[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Joanna Sousa da Fonseca Santana; Email: joannasantana20.2@bahiana.edu.br.

Background: Stroke remains a major health concern globally, especially affecting the elderly, yet the incidence in younger populations is rising. Early-onset stroke, defined as occurring in individuals under 45 years, presents unique challenges in terms of management and prognosis. However, the literature still lacks of enough

information about this condition. **Objective:** To compare the prognostic factors and management outcomes of stroke patients younger than 45 years with those aged 45 and older. **Methods:** This is a retrospective cohort study, derived from a larger stroke study. Data were collected from electronic medical records of patients previously admitted to a reference hospital in Salvador, Bahia, with a diagnosis of stroke between April 2015 and August 2023. The variables of interest collected were: sex, age, mortality, NIHSS score at admission, hospitalization time, stroke etiology and dysphagia. Data were analyzed using the software SPSS. The study was approved by the Research Ethics Committee. **Results:** In the elderly group (45 years and older), the sample included 646 patients with 47% males and 52% females. The median age was 69 years, with a maximum age of 106. Mortality was reported at 16%. Dysphagia data were incomplete for 20% of patients; 21% had mild, 17% moderate, and 13% severe dysphagia. The average hospital stay was 11.9 days. Stroke etiology was undetermined in 61% of patients, cardioembolic in 18%, small artery occlusion in 9%, large artery atherosclerosis in 8%, and other causes in 2.7%. Stroke severity was classified as minor in 18.8%, moderate in 38.5%, moderate to severe in 4.7%, and severe in 8.2%. Thrombolysis was performed 9% of cases. Among the under 45 years group, there were 130 individuals, predominantly male (56.2%), with ages ranging from 14 to 45 years and a median age of 40. The mortality rate was 10.8%. Hospital stays averaged 10.8 days. Dysphagia assessment was missing in 13.8%, absent in 52%, mild in 17.7%, moderate in 10.8%, and severe in 5.4%. Thrombolysis was performed in 13% of cases, and craniectomy in 8%. The primary stroke etiologies were undetermined (60.8%), cardioembolic (17%), large artery atherosclerosis (7%), and small artery occlusion and other causes (each 6.9%). Stroke severity was classified as minor in 23.1%, moderate in 42.3%, moderate to severe in 4.6%, and severe in 7.7%. Thrombolysis was performed 13% of cases. **Conclusion:** Younger patients exhibited a lower mortality rate and shorter hospital stays. Dysphagia was less prevalent in the younger group, with a significant number showing no signs of it. The severity of strokes was predominantly moderate in both age groups. Thrombolysis were more possible among the younger group. These findings highlight the importance of age-specific management strategies and the need for further research to better understand the prognostic factors affecting different age groups, ultimately improving stroke care and patient outcomes.

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Comparative analysis of the spatio-temporal distribution of acute ischemic stroke treatment with thrombolytic therapy in Brazil from 2019 to 2023

Paola Carolina Santos Foinkinos[1], Karolliny Amador Kzam , Matheus Prado dos Santos[1], Bruno Kauê Rodrigues Vilhena[1], Vinícius Adriano Coelho Amaral[1], Luan Nascimento Pereira de Amorim[1], Fernando Gabriel Rodrigues Baia[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.
[2]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.

Correspondence: Paola Carolina Santos Foinkinos; Email: paola.csfoinkinos@aluno.uepa.br.

Background: The treatment of acute ischemic stroke with the use of thrombolytic therapy is a therapeutic approach that aims to dissolve the blood clot responsible for the obstruction of cerebral vessels, using recombinant tissue plasminogen activator (rt-PA). It is crucial that the stroke is diagnosed quickly to confirm that it is an ischemic stroke and not hemorrhagic, as thrombolytic therapy is contraindicated in cases of cerebral hemorrhage. By restoring blood flow, thrombolytics can reduce the extent of damage to brain tissue, improving the chances of functional recovery. Patients treated with thrombolytics within the appropriate time window have better results in terms of neurological and functional recovery. The main risk associated with the use of thrombolytics is bleeding, including intracerebral hemorrhage and, although rare, allergic reaction to the drug. **Objective:** To comparatively analyze the spatial distribution of thrombolytic treatment for ischemic stroke in Brazil between 2019 and 2023. **Methods:** This is a descriptive, cross-sectional and quantitative study in which secondary data was collected from the Department of Informatics of the Unified Health System (DATASUS) from 2019 to 2023. The following variables were analyzed: deaths per procedure, approvals per hospital admission and total amount spent. **Results:** From 2019 to 2023, with regard to the variable deaths per procedure, there were a total of 2,320 deaths in the treatment of acute ischemic stroke with the use of thrombolytic therapy, with an increasing number of deaths. In terms of the country's regions, the North had the fewest deaths, with only 2, 1 in 2020 and 1 in 2023. Regarding the region with the highest number of deaths, the Southeast region stands out, with 800 cases, with the state of São Paulo having the highest number, 485 cases. As for the variable approvals for hospital admissions, there were a total of 22,056 admissions, with the Northeast region standing out with 7,872 admissions. In addition, in terms of the total amount spent on the procedure, there was a total of R$67,024,999.05, with the region that spent the most being the Southeast with R$25,198,904.56. **Conclusion:** It can therefore be seen that there is a considerable discrepancy in the distribution of stroke treatment with thrombolytics in Brazil, with the Southeast receiving the largest share of cases, while the less developed regions (North) still need effective investment in this therapy to avoid stroke complications. The same can be seen with regard to the costs of this therapeutic practice by region of the country, with the Southeast concentrating almost 40% of the total expenditure during the study period. As it represents a good prognosis in the treatment of strokes, the use of thrombolytics should be disseminated so that it can be effectively applied nationwide.

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Comparative efficacy of different pharmacotherapies for muscular dystrophy: a systematic review and network meta-analysis

Artur Menegaz de Almeida[1], Maria Eduarda Cavalcanti[2], Francisco Cezar Aquino Moraes[3], Fernando Luiz Westphal Filho[4], Paulo Roberto Moss Lopes[4], Fernanda Moraes Tamashiro[5], Celso de Almeida Souza Miranda[6], Jorge Henrique Cavalcanti Orestes Cardoso[2], Francinny Alves Kelly[7]

[1]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[2]Universidade de Pernambuco, Recife PE, Brazil.
[3]Universidade Federal do Pará, Belém PA, Brazil.
[4]Universidade Federal do Amazonas, Manaus AM, Brazil.
[5]Universidad de Buenos Aires, Buenos Aires, Argentina.
[6]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[7]DPIC, São Paulo SP, Brazil.

Correspondence: Artur Menegaz de Almeida; Email: arturomenegaz@gmail.com.

Background: Muscular dystrophies (MDs) are rare neuromuscular disorders that lead to progressive muscle degeneration and early death. MDs like Duchenne have a poor prognosis associated with an unclear preferred treatment. Despite several new agents being assessed in clinical trials, no network meta-analysis (NMA) has been conducted to date evaluating different treatment options for MDs. **Methods:** MEDLINE, EMBASE, and Cochrane databases were searched for randomized and non-randomized clinical trials testing different agents in patients with any type of MD. The NMA was performed under a Frequentist framework using RStudio software version 4.3.2 and extension packages "netmeta" and "dmetar" for all statistical analysis. Our primary outcome was the improvement in 6-minute walk distance (6MWD) and the change in time to run/walk 10 m (TTRW). The NMA pooled all the direct and indirect evidence and simultaneously compared several treatments using the network relationship through a model for continuous outcomes. **Results:** 17 studies and 1,905 patients were included, of whom 1,802 (94.59%) were diagnosed with Duchenne muscular dystrophy. Among the included studies, 11 different drugs were evaluated for their efficacy in improving disease progression. Ataluren and drisapersen significantly increased the 6-minute walk distance in comparison with placebo: (SMD -21.08; 95% CI -23.08 to -19.08) and (SMD -12.91; 95% CI -15.32 to -10.51), respectively. Additionally, no benefit was shown with the administration of tamoxifen or recombinant human insulin-like growth (rhIGF), while the placebo group was favored in comparison with tadalafil treatment. Concerning the change in TTRW, ataluren (SMD 1.07; 95% CI 0.91 to 1.22) and drisapersen (SMD 1.73; 95% CI 1.54 to 1.92) also showed a statistically significant benefit, in addition to viltolarsen (SMD 0.97; 95% CI 0.32 to 1.62) and vamorolone (SMD 0.08; 95% CI 0.03 to 0.13), which showed a slight improvement in the score. **Conclusion:** Our findings suggest that ataluren and drisapersen are both associated with a significant improvement in disease progression, possibly reducing muscle deterioration as seen in the 6MWD and TTRW. Additionally, viltolarsen and vamorolone were also associated with benefits in patients with MDs.

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Comparative spatial analysis of leprosy cases in the North and Southeast regions

Fernando Gabriel dos Santos Santiago[1], Rafael Hendrick Freitas de Paula[1], Douglas Ferreira Pereira[1], Savio Roberto Silva Costa[1], Danusa Neves Somensi[1], Mariangela Moreno Domingues[1], Emanuel de Jesus Soares de Souza[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Fernando Gabriel dos Santos Santiago; Email: fernando_gsantiago@outlook.com.

Background: Leprosy represents a significant obstacle for Brazil, which is the second country in the number of leprosy cases in the world. In the meantime, the northern and southeastern regions of the country show significant differences in socioeconomic indices and access to health services, which are determining factors for the disparity in the spread of infection in these regions. Therefore, understanding the epidemiological differences between the regions, aiming to identify distribution patterns, is essential for the development of prevention and control strategies. **Objective:** To compare the epidemiological profile and clinical variables of leprosy between the North and Southeast regions of Brazil. **Methods:** This is an epidemiological and cross-sectional study, with data collected from the Notifiable Diseases Information System, referring to notifications of leprosy cases in the North and Southeast regions of Brazil, from 2014 to 2023. The variables analyzed included: sex, race, education, age, operational class, clinical form, and reactional episode. **Results:** The data analysis revealed a higher predominance of cases in the North region with 181,542 and 118,636 cases in the Southeast. The average incidence rate per 100,000 inhabitants was 100.64 in the North region and 13.58 in the Southeast.The prevalence of males was higher in both regions. The brown race was predominant in the North, corresponding to 72.3. However, in the Southeast region, there was a slight predominance of the white race over the brown, with 40.8% and 40.1% respectively. Leprosy was more prevalent in patients with elementary education, portraying 48.3%-52.11% of cases. The prevalent age group was above 15 years in both regions, corresponding

to about 90.3%. The most found operational class was multibacillary in both regions, being 76,5-77.2%. The most present clinical form was dimorphic, with 53.4% of cases in the North, followed by virchowiana with 16%. In the Southeast, the percentages were 41.8% and 27.8% respectively. In both cases the main reactional form was type 1, corresponding to 10,7-11.1% of cases. **Conclusion:** The significant regional disparities in terms of incidence rate, which may be related to different socioeconomic levels among the regions. The epidemiological profile showed similar in the studied regions, being male, elementary education with age over 15 years, except for the greater predominance of brown color in the northern region, which is related to the larger population quantity of brown race in this region compared to the Southeast. There were no significant differences between the reactional forms among the regions. The greater predominance of the multibacillary operational classification indicates delay in disease identification, so it is necessary to emphasize the importance of public policies aimed at early identification, especially in the North region due to higher incidence of cases.

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Comparison between conventional physiotherapy rehabilitation and virtual reality in Parkinson's disease: a systematic review

Maria Eduarda Scheffer Valverde[1], Tacio Rafael Santos Batista[1], Thais Oliveira Bellavinha Martins[1], Virgílio Andrade dos Santos Maciel[1], Paula de Sousa Silva[1], Rafael Henrique Neves Gomes[2]

[1]Faculdade de Ciências Médicas e da Saúde de Juiz de Fora, Juiz de Fora MG, Brazil.
[2]Universidade Federal de Juiz de Fora, Hospital Universitário, Juiz de Fora MG, Brazil.

Correspondence: Maria Eduarda Scheffer Valverde; Email: mariascheffer1418@gmail.com.

Background: Virtual reality (VR) has become a useful tool in physiotherapeutic rehabilitation for patients with Parkinson's disease (PD), particularly in alleviating balance deficits and gait impairments. **Objective:** To compare conventional physiotherapy with VR-based rehabilitation in PD through a systematic review. **Methods:** A search was conducted on PubMed in May 2024 using the keywords "Virtual reality" and "Parkinson Disease," along with their MeSH variations, and filters including "Humans," "Randomized Controlled Trial", "Clinical Trial", "Multicenter Study" and "5 years." Among the 20 studies found, the three most relevant were selected for this review. The PRISMA scale was used to improve reporting quality. **Results:** A total of 111 patients were analyzed across the studies. Inclusion criteria encompassed scales assessing PD progression (UPDRS), motor dysfunctions (POMA, DASH, TUGT), balance impairments (BSS, FGA, DGI), dementia (CDR, MCS) and psychosocial impacts (PIADS). Treatment durations varied among studies, ranging from 5 weeks (Maranesi et al.) to 12 weeks (Feng et al.). Various VR programs were utilized, such as the Tymo system (Maranesi et al.), featuring exergames and motion analysis equipment, and the NIRVANA system (Pazzaglia et al.), employing markerless infrared optoelectronic devices for immersive sensory and audiovisual exercises. All studies demonstrated superior improvements in balance among patients undergoing VR rehabilitation compared to control groups ($p<0.05$). Pazzaglia et al. and Maranesi et al. also noted enhancements in mental health status, while Feng et al. highlighted improvements over conventional methods in UPDRS scores, which assess tremor, rigidity, speech, posture, and gait, indicating broader benefits of VR rehabilitation beyond motor dysfunctions. **Conclusion:** VR proves effective in enhancing functional parameters for PD patients and could serve as a valuable alternative in physiotherapeutic rehabilitation protocols. However, further studies are needed to optimize treatment duration and session frequency for optimal outcomes.

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Comparison of disability level between early and late onset Parkinson's disease using WHODAS-2

Isaíra Almeida Pereira da Silva Nascimento[1], Kátia Cirilo Costa Nóbrega[1], Maria Elisa Pimentel Piemonte[1], Isabela Carneiro Barone[1], Giovanna Checchio[1], Vitória Pereira Ponciano[1], Clara Greif Cerveira de Paula[1], Arieni Nunes Possani[1], Natália Cardoso Penha[1], Bruno Rafael Antunes de Souza[2]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Kátia Cirilo Costa Nóbrega; Email: katiacirilo@usp.com.

Background: Parkinson's disease (PD) is a degenerative neurological disorder that usually affects people over the age of 60. However, 10% – 20 % of patients have an early onset of PD (EOPD). **Objective:** To compare disability levels according to the World Health Organization Disability Assessment Schedule 2.0 (WHODAS-2) between people with EOPD and those with late-onset PD (LOPD). **Methods:** We conducted a cross-sectional study with 95 EOPD patients (mean-age 44.51±4.63, H&Y 1.93±0.93) and 255 LOPD patients (mean-age 63.01±7.99, H&Y 2.02±0.95). Demographic information, clinical characteristics, cognitive evaluation by Telephone-Montreal-Cognitive-Assessment (T-MoCA), functionality self-evaluation by WHODAS-2 and the Unified-Parkinson's-Disease-Rating-Scale (MDS-UPDRS), parts I and II, were documented for each patient by an individual remote interview. **Results:** Analysis showed a statistically significant difference between EOPD and LOPD in two domains of WHODAS-2 only: cognition (Z-adjusted = 2.60; p-value adjusted <0.009) and activities of daily living related to work/school (Z-adjusted = 2.34; p-value adjusted <0.01). T-MoCA scores confirmed more impaired cognition capacity in LOPD (Z-adjusted = 2.42; p-value adjusted <0.01). The two groups had no significant differences in levodopa daily dosage, Hoehn and Yahr (H&Y) stages, disease time duration, and MDS-UPDRS I and II scores. **Conclusion:** People living with EOPD face similar disability levels as those with LOPD, except for cognition, where LOPD patients exhibited higher levels of disability than EOPD and for work activities where the EOPD exhibited higher levels of disability than LODP. These results highlight the challenges faced by people with EOPD in interacting with society and living with the disease for a longer time. The WHODAS-2 can be a useful tool to assess disability and tailor interventions for people with PD of different age groups.

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Comparison of electrochemical skin conductance and skin biopsies tests for detection of neuropathy in patients with hereditary transthyretin amyloidosis-polyneuropathy (ATTRv-PN)

Mateus Gustavo Favaro[1], Carolina Lavigne Moreira[1], Marcus Vinicius Simoes[1], Pedro Manoel Marques Garibaldi[1], Pedro Jose Tomaselli[1], Gustavo Maximiano Alves[1], Wilson Marques Junior[1], Manoella Guerra de Albuquerque Bueno[1], Matheus Compart Hemerly[1]

[1]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Mateus Gustavo Favaro; Email: mateusgustavofavaro@gmail.com.

Background: The diagnosis of neuropathy, especially small fiber type, has been a challenge in ATTRv-PN. Many methods were described, but the gold standard remains the skin biopsy. However, they are invasive and scarcely available in poor countries. The evaluation of the sudomotor function, measured by the electrochemical skin conductance in the palms of the hands and soles of the feet, is a non-invasive method aimed at identifying peripheral nerve involvement. However, Its use in ATTRv-PN has not been properly investigated. In this project we compared the efficacy of the electrochemical skin conductance test against the results of the intraepidermal nerve fiber density (IENFD). **Objective:** To compare the efficacy of the electrochemical skin conductance on the feet of patients with ATTRv-PN with the IENFD of the distal leg. **Methods:** We studied 28 patients ATTRv-PN patients with positive genetic test.. Skin biopsies were performed on the lower limb, and the intraepidermal nerve fiber density (IENFD) was calculated based on normal values published bty the European Federation of Neurology (2010). Electrochemical skin conductance was evakluated with SDOSCAN, a commercial machine. Results of the conductance test were divided into normal or abnormal (categorical variables). Analysis were carried with the Concordance test (Kappa test) and the association test (Chi-squared). Also, the absolute numbers (quantitative variable) from the conductance study were compared using the Wilcoxon/Mann-Whitney non-parametric test. Later, the ROC curve was calculated. SPSS 22.0 was used for statistical tests, and REDCap for data storage. **Results:** We studied 28 patients There is a weak concordance (Kappa=0.25; I.C. 95 [0.0 - 0.56], p=0.13) and no association (Chi-squared p=0.22; Mann-Whitney p=0.42) between the electrochemical skin conductance of the lower limb with IEFND in patients with ATTRv-PN in our center. Also, the skin conductance test has low accuracy 59.1% I.C.95% (37.5%-80.7). **Conclusion:** In this study we did not find a good correlation between nerve fiber density and the electrochemical skin conductance. A possible explanation is that skin biopsy does not distinguish any specific type while the electrochemical skin conductance just measure the pos-synaptic sympathetic fibers. In addition, nerve biopsy identify the fibers, but not their their functional integrity. This result indicates that the diagnosis of small fiber neuropathy must be sought through varied approaches, until the identification of a gold standard method.

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Comparison of Kappa index and oligoclonal bands detection positivity in cerebrospinal fluid samples of patients with suspected multiple sclerosis (MS)

Renan Domingues[1], Irineu Massaia[1], Myrna Monteiro[1], Marcio Vega[1], Camila Spinelli[1], Lais Guerra[1], Daiane Salomão[1], Carlos Senne[1]

[1]Senne Liquor Diagnóstico, São Paulo SP, Brazil.

Correspondence: Renan Barros Domingues; Email: contato@renandomingues.med.br.

Background: The evaluation of the intrathecal production of IgG is useful in the evaluation of MS suspected cases. The qualitative detection of oligoclonal bands (OCBs) in cerebrospinal fluid (CSF) and serum and the Kappa Index are currently the best methods to detect intrathecal production of IgG. Besides their diagnostic value it is possible that OCBs and Kappa Index have also prognostic value. **Objective:** To evaluate the results of the Kappa Index and the oligoclonal bands (BOCs) in samples from patients with suspected multiple sclerosis (MS). We also evaluated if patients with 8 or more OCBs have higher Kappa index values. **Methods:** Patients with suspected MS underwent cerebrospinal fluid (CSF) analysis. CSF samples were submitted to the detection of BOCs and kappa index. These comparisons were carried out with Chi Square test. The number of OCBs was classified as 2-7 and ≥8. The comparison of the median Kappa Index between these two groups were carried out with Mann-Whitney test. **Results:** Two eighty-five serum and CSF

samples were included, 115 of them had positive CSF BOCs and 138 were positive with Kappa index. Overall, 148 patients had positive results with at least one of the methods and Kappa index was more sensitive than BOC (93.2% x 77.7%, P<0.0001). 37 patients had 2-7 OCBs and 72 had ≥8 OCBs. The median Kappa index values were: 2-7 OCBs group = 18.23 (range 3.02 to 457.9) and ≥8 OCBs group = 92.49 (range 12.1 to 1117) (P<0.0001). **Conclusion:** The Kappa index showed to increase the sensitivity for detecting intrathecal IgG detection. However, there were cases in which BOC was the only positive method for the detection of intrathecal IgG production. Thus, it remains important to perform both methods to increase the CSF diagnostic performance in MS suspected cases. Patients more OCBs had higher Kappa index values, suggesting that Kappa Index may be a prognostic MS marker. Future studies including clinical data should address this question.

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Comparison of mortality rate and length of stay between intracranial aneurysm treatment by endovascular surgery vs. microsurgery in Northeastern Brazil over the past 5 years

Rosa Carolina Santos de Oliveira[1], Felipe de Jesus Gois[2], Christian Wallace Santos Meneses[2], Clayton de Jesus Barbosa[1], Eduarda Tavares da Silva[2], André Luiz Borges Silva[2], Gabriel Valentim dos Santos Menezes Siqueira[2], Helton Benevides Santana de Oliveira[2]

[1]Universidade Tiradentes, Aracaju SE, Brazil.
[2]Universidade Federal de Sergipe, Aracaju SE, Brazil.

Correspondence: Rosa Carolina Santos de Oliveira; Email: rosa.oliveira@souunit.com.br.

Background: This study analyzes the mortality rate and average hospital stay of patients with intracranial aneurysms treated by endovascular surgery versus microsurgery under Brazil's public health system (SUS) in Northeastern Brazil over the last five years. Comparing these approaches is crucial for understanding their specific efficacies, providing relevant data for healthcare optimization and aiding decision-making. **Objective:** To compare the mortality rate and average hospital stay of intracranial aneurysm treatments performed in Northeastern Brazil over the last 5 years by endovascular surgery versus microsurgery. **Methods:** The study utilized DATASUS data to analyze mortality rate and hospital stay averages between endovascular and microsurgery treatments for intracranial aneurysms. Analysis covered the general population and segmented results by Brazilian regions from 2019 to 2023. Data was analyzed using Excel, presenting values in both absolute and percentage terms. **Results:** Temporal analysis of DATASUS data from 2019 to 2023 revealed the following findings: The average hospital stay for microsurgery was 16.2 days in 2019, 14.7 days in 2020, 15.6 days in 2021, 15.4 days in 2022, and 12.8 days in 2023, resulting in an overall average of 15.1 days. The mortality rate associated with microsurgery per 100,000 inhabitants was 12 in 2019, 13.64 in 2020, 9.02 in 2021, 14.18 in 2022, and 9.2 in 2023, with a total average of 11.76. For embolization procedures, the average hospital stay was 10.2 days in 2019, 9.3 days in 2020, 9.7 days in 2021, 10.4 days in 2022, and 10.1 days in 2023, totaling an average of 10 days. The mortality rate per 100,000 inhabitants associated with embolization was 5.6 in 2019, 6.7 in 2020, 8.31 in 2021, 5.75 in 2022, and 5.02 in 2023, resulting in an average of 6.28. Comparing data from 2019 to 2023, a 21% reduction in average hospital stay for microsurgery and a 1% reduction for embolization were observed. Regarding mortality rate, there was a 23.33% decrease in overall mortality for microsurgery and a 10.3% decrease in mortality associated with embolization. **Conclusion:** Both mortality rate and hospital stay duration are lower in patients undergoing intracranial aneurysm treatment via endovascular surgery compared to conventional microsurgery. However, both techniques remain necessary and are not mutually exclusive, requiring individual assessment based on patient and aneurysm characteristics when choosing the method. Furthermore, this study underscores the need for long-term comparative studies, cohorts, and randomized clinical trials to establish the real benefits and indications of each procedure.

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Comparison of open-loop and closed-loop transcutaneous auricular vagus nerve stimulation on autonomic control in healthy individuals: new directions for epilepsy treatment

Kevin Gustavo dos Santos Silva[1], Gabriella Maria de Faria[2], Viviane Fajardo Filgueiras[2], Henrique Amorim[2], Lívia Bruni de Souza[2], Fabiana Gomes Ferreira[2], Nádia Bertoncello[2], Karina Rabello Casali[2], Tatiana Sousa Cunha[2]

[1]Universidade Anhembi Morumbi, São José dos Campos SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Kevin Gustavo dos Santos Silva; Email: kevingustavo30@gmail.com.

Background: Non-invasive transcutaneous auricular vagus nerve stimulation (taVNS) can activate autonomic control and elicit therapeutic effects on various pathological conditions, especially epilepsy. The significant clinical interest in using such devices has opened the way for new variations seeking to optimize the effects evoked on vagal modulation and expand possibilities in the treatment of patients with epilepsy unresponsive to conventional therapies. In this sense, studies have shown that stimulation applied in synchronization with respiration can enhance the neuromodulation effect of taVNS. Our research group has developed a closed-loop taVNS prototype that aligns stimulation parameters with respiratory rhythms. **Objective:** This study aims to compare the effects of vagus stimulation on the autonomic control of healthy individuals using a commercial open-loop taVNS device (Vitos) and our closed-loop taVNS prototype. **Methods:** A total of 6 healthy volunteers of both sexes (22-48 years old) were included in this blinded controlled study and randomized into two groups: open-loop device (OL; n=4) and closed-loop prototype (CL; n=2), both at 25Hz. Cardiac data was collected at three points: at rest, during intervention (20 minutes of use of OL or CL taVNS), and after intervention, using a continuous and non-invasive heart rate monitor (Polar V800 heart rate monitor). The autonomic nervous system was assessed by quantifying sympathetic (SM) and vagal (VM) modulations, estimated through spectral analysis applied to tachograms. The study was approved by the Research Ethics Committee. **Results:** The results revealed that taVNS, in both protocols, caused a significant increase in VM compared to baseline, in normalized units (nu) (OL_basal = 13.1±5.9; OL_during = 24.9±7.9; OL_post = 25.9±12.8; CL_basal = 13.6±0.8; CL_during = 46.7±3.5; CL_post = 25.1±2.9, p=0.001). CL taVNS produced a substantially greater increase in vagal activity during the maneuver compared to the OL device (p=0.006). In conclusion, both vagus nerve stimulation through OL and CL taVNS resulted in a significant increase in vagal activity during the session, demonstrating the effectiveness of the developed prototype. Additionally, CL had a more pronounced effect on immediate autonomic nervous system activation, reinforcing existing literature that suggests optimization and enhancement of vagal modulation effects. **Conclusion:** The prototype showed a significant effect on vagal modulation, indicating potential for clinical validation and market application. A randomized clinical trial will be conducted for proper certifications involving healthy subjects and patients with epilepsy.

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Comparison of quality indicators in a stroke unit during and after the COVID-19 pandemic

Laiza Milena de Araújo[1], Nicole Rampani Franzoni[1], Luana Miyahira Makita[1], Gustavo de Oliveira Morais[1], Ana Carolina Pelek[1], Amanda Paixão Peron[1], Paola Cristine Ramos Santos[1], Luiz Felipe Fontana Triches[1], Renata Dal-Prá Ducci[1], Marcos C. Lange[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Luiz Felipe Fontana Triches; Email: luizfelipefontri@gmail.com.

Background: Health quality indicators are essential for monitoring the quality of care provided by healthcare services. In this regard, there are numerous indicators available to measure the quality of care in stroke management. However, during the COVID-19 pandemic, healthcare services faced significant challenges, further highlighting the importance of these indicators. **Objective:** To assess, through quality indicators, whether there have been changes in the care provided to stroke patients during and after the COVID-19 pandemic. **Methods:** This is a cross-sectional, retrospective observational study using data analysis from electronic medical records. The study sample comprised patients admitted to the Stroke Unit of the Clinical Hospital of the Federal University of Paraná for ischemic stroke between November 2020 and May 2024, divided into groups during and after the COVID-19 pandemic. The criterion for dividing the periods was the date of the end of the global emergency proposed by the WHO, on 05/05/2023. A total of 554 patients were studied according to variables such as sex, age, door-to-needle time, imaging exams performed, treatment, evaluation by the multidisciplinary team (physiotherapy, speech therapy, and occupational therapy), and length of hospital stay. **Results:** Overall, 554 individuals diagnosed with ischemic stroke were admitted to the Stroke Unit of the Clinical Hospital during (n=408) and after (n=146) the pandemic. The mean age, percentage of men, and therapeutic strategy were similar (p>0.05) at both time points. The mean door-to-needle time was 43.7±40.8 and 47.4±39.9 minutes in patients admitted intra and post-Covid-19, respectively. Although the data suggest an increase in the number of patients with door-to-image time <30 minutes after the pandemic, a large percentage of patients have unknown data (47.9%). Hence, making concrete statements on the matter becomes unfeasible. Regarding prescription for secondary prevention, there was an increase in the prescription of anticoagulants (25% vs 32.2%, p<0.001) and antiplatelet agents (55.1% vs 57.5%, p<0.001) following the pandemic. Although, there are also many patients with unknown data in this category. Multidisciplinary care, including physiotherapy (84.1% vs 74%, p<0.05) and occupational therapy (82.6% vs 53.4%, p<0.05) were more frequent during the pandemic. Mean hospital stays were similar between intra and post-pandemic individuals (11.8±4.4 days vs 11.1±4.7 days, p>0.05). **Conclusion:** Therefore, although COVID-19 impacted healthcare services, there is no evidence to suggest a worsening of health indicators. Additionally, there is a reduction in the availability of occupational therapy and physiotherapy services. This is due to the reallocation of professionals and an increasing demand for services at the hospital. Indicators are valuable tools for advocating improvements in care. Thus, it is important to accurately record information to reflect reality reliably.

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Comparison of stroke awareness between Aparecida (SP) and São Paulo (SP): a cross-sectional study from 2023 and 2024

Beatriz Barbosa Nishigima[1], Mariana Rachas Reis[1], Anna Luísa Mennitti[1], Fernanda Yumi Abe[1], Rubens José Gagliardi[1]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.

Correspondence: Beatriz Barbosa Nishigima; Email: beatriz.nishigima@gmail.com.

Background: Stroke poses substantial health risks globally, emphasizing the critical importance of early symptom recognition for improving outcomes. Disparities in stroke awareness between metropolitan and smaller urban areas warrant investigation. This study compares stroke knowledge between Aparecida/SP and São Paulo/SP, highlighting regional differences and underscoring the need for target educational efforts. **Objective:** To assess and compare stroke awareness between the population of Aparecida/SP, located in a semi-urban setting, and São Paulo/SP, situated in a metropolitan area. **Methods:** This is a descriptive and comparative cross-sectional study utilizing data from a structured questionnaire available on the REDCap platform. It was administered to participants in two distinct settings: firstly, to random passers-by at subway stations in the city of São Paulo in October 2023; and secondly, to patients of a community outreach program in Aparecida/SP, a semi-urban city of São Paulo, in January 2024 . The study sample included individuals aged 18 years or older who signed the term of consent. Participants in the Aparecida/SP group were also required to be registered for outpatient appointments. The questionnaire collected demographic data and included inquiries about stroke, such as its definition, symptoms, prevention methods, appropriate actions upon suspicion and acquaintance with someone who had experienced a stroke. **Results:** The sample evaluated in Aparecida/SP consisted of 276 individuals, while in São Paulo/SP it comprised 370, totaling 646 answers. Among the interviewees in Aparecida, 62.7% were women, and the most prevalent age group ranged from 60 to 69 years (28.6%), with 31.8% of them having incomplete primary education. In the São Paulo sample, 57.3% were women, the most prevalent age group was also 60 to 69 years (26%) and 28.6% had completed secondary education. In the sem-urban sample, 69.9%% knew someone who had a stroke and 59.8%% couldn't define the disease. Comparatively, in the capital, these numbers were 77.3% and 60%. Regarding symptoms, the most mentioned one in both Aparecida and São Paulo samples was paresthesia in the face, arm or leg (54.6% and 49.2% respectively), while the least cited one was vision impairment (14.3% and 11% respectively). The most frequently cited preventive measure in the small town was physical activity (69.2%), whereas in the big city it was healthy eating (64.3%). In terms of response to witnessing a stroke, in Aparecida, 62.7% would call an emergency service and 0.72% would check the time, and in São Paulo these percentages were 95.7%% and 10.5%. **Conclusion:** This study underscores significant disparities in stroke awareness between Aparecida/SP and São Paulo/SP. Despite varying levels of awareness, both regions exhibit gaps in knowledge about the subject. Targeted educational efforts are essential to enhance public awareness and mitigate the impact of stroke across diverse urban settings.

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Comparison of the efficacy of monotherapy with bevacizumab versus combined therapies or monotherapies with other drug classes in the treatment of recurrent glioblastoma

Isabela Lara Marcondes[1], Evelyn de Castilho Brandt[1], Stefany Vitória Schneider[1], Laura Cristina Lazzari[1], Kennyel Andre Veloso[1], Paola Patrícia Dias de Farias[1], Gabriela Tartarotti Godoy[1], Gustavo Colombo Dal-Pont[1]

[1]Universidade Alto Vale do Rio do Peixe, Caçador SC, Brazil.

Correspondence: Isabela Lara Marcondes; Email: belinhamarc@hotmail.com.

Background: Glioblastoma (GBM) is an aggressive type of primary brain tumor that has a mean overall survival (OS) of less than 2 years. GMB causes mutations in Vascular Endothelial Growth Factor (VEGF), leading to exacerbated angiogenesis and tumor malignancy. The standard treatment for newly diagnosed GMB is resection of the tumor, followed by radiotherapy and chemotherapy with temozolomide. However, the tumor almost always reappears after an average of 8 months. Bevacizumab (BEV) is a VEGF antagonist Monoclonal Antibody (MaB) and was approved in 2009 for use in recurrent glioblastoma (rGBM) by the Food and Drug Administration, after studies demonstrated that its use improves progression-free survival (PFS) within 6 months. **Objective:** To compare the use of BEV as monotherapy versus combination therapies or monotherapy with other drug classes for the treatment of rGBM. **Methods:** This is a systematic review of the literature, carried out using the PRISMA method, in the PubMed database in search of clinical trials, controlled and randomized tests from the last 10 years (2014-2024), in the English language. The works included were articles that exclusively analyzed rGBM and the pharmacological treatment with Mabs in combination or as monotherapy. The articles were analyzed blindly by the authors, using Rayyan® software. **Results:** With the search strategy, 117 studies were found, of which 9 met the inclusion criteria. The association of BEV with vorinostat, lomustine, RO5323441, VB-111 or other MaBs, such as pembrolizumab and dasatinib, did not demonstrate benefits in PFS compared to the drug alone in patients with rGMB. On the other hand, new radiotherapy sessions, associated with the use of BEV, proved to be interesting in a randomized phase II study, further increasing PFS. In comparative analyzes of the use of different drugs alone, there is no statistical relevance in the mortality rate with the use of nivolumab versus BEV. In contrast, a phase II study comparing BEV with fotemustine demonstrated the superiority of BEV in objective response rate, showing it to be an effective option for reducing tumor size. However, fotemustine showed a trend towards greater OS, increasing life expectancy by approximately 6 months after the start of treatment, in 62% of patients treated with BEV and 73% of those treated with fotemustine. **Conclusion:** Limiting factors such as death or progression of GMB in patients are a challenge in the search for effective therapies for the treatment of rGMB. Currently, there are studies that show an improvement in PFS in the combined use of BEV with re-irradiation. Although, the use of BEV alone remains the main therapy used. However, fotemustine presents itself as a promising drug, increasing OS compared to BEV. Nevertheless, more research is needed to better understand these mechanisms.

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Comparison of the incidence and mortality of intracranial trauma in children under 1 year old in the Southeast region of Brazil during 2020 and 2021

Aline Marques Barreto Scalioni[1], Thiago Luvizon Mollo[2], Karen Paes Mello[3], Polyany Batista de Moura Lemos[4], Guilherme de Andrade Ruela[5]

[1]Universidade Nove de Julho, Guarulhos SP, Brazil.
[2]Universidade Nove de Julho, Bauru SP, Brazil.
[3]Universidade Brasil, Fernandópolis SP, Brazil.
[4]Faculdade CET, Teresina PI, Brazil.
[5]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Aline Marques Barreto Scalioni; Email: alinemscalioni@gmail.com.

Background: Traumatic brain injury (TBI) is one of the main causes of pediatric emergencies, responsible for a high incidence of hospital admissions with alarming mortality and morbidity rates. This type of trauma is characterized by any traumatic injury that results in anatomical damage, such as cranial fractures or scalp injuries, or functional impairment of the meninges, brain, or its vessels. Pediatric TBI is considered a public health problem. **Objective:** To analyze and compare the incidence and deaths resulting from Traumatic Brain Injury in patients under 1 year old in the southeastern region of Brazil in 2020 and 2021. **Methods:** This was an ecological, retrospective, descriptive, and quantitative study using data from the Department of Informatics of the Unified Health System (DATASUS). The study analyzed the number of hospitalizations and deaths, as well as the mortality rate, due to intracranial trauma in 2020 and 2021 in the southeastern region of Brazil, including the age group "under 1 year". Any other categories not specified were excluded. **Results:** According to the data collected, there were 2,387 hospitalizations for pediatric TBI in the age group of 0-1 year in the southeastern region of Brazil between 2020 and 2022. There were 30 deaths, representing a mortality rate of approximately 2.51%. The year with the highest incidence of TBI among children under 1 year old was 2021, with 1,173 hospitalizations (and 17 deaths). The state with the highest rate was São Paulo, with 1,467 hospitalizations and 19 deaths. In contrast, Espírito Santo had the highest mortality rate among the states in the region in both years (2.70% and 3.23% in 2020 and 2021, respectively). Despite the increase in hospital admissions, there was no substantial increase in deaths during the analyzed years, keeping the mortality rate low. It is worth noting that there are indications of underreporting and incorrect filling out of Hospitalization Authorizations (AIH) and Death Certificates (DO), which may result in the creation of false epidemiological profiles due to deaths from undefined causes, making it difficult to develop a strategic plan to prevent these deaths. **Conclusion:** The data collected demonstrate the importance of preventive measures and awareness regarding the severity of TBI and its prognostics, due to its potential to cause severe consequences and permanent side effects. Findings from the present study showed high numbers of hospitalizations in this vulnerable age group, which can result in severe outcomes if the patient is not managed correctly by a multidisciplinary team. This highlights the importance of legal guardians taking measures to protect children and ensure their safety at various levels.

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Comparison of the prevalence of hospitalizations due to Alzheimer's disease in the federal regions of Brazil and the mortality rate

Guilherme Galardino Santos[1], Bernardo Caetano Novaes[1], Samuel José Feres Ozava[1], Myllene Rodrigues Silva[1], Victor Bruno dos Santos Teixeira[1], Davi Melo de Hollanda Cordeiro[1]

[1]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.

Correspondence: Guilherme Galardino Santos; Email: guilhermegalardino@gmail.com.

Background: Alzheimer's disease (AD) is a neurodegenerative disease that primarily affects older adults. It is considered the most common cause of dementia and one of the leading sources of dependency, disability, and morbidity and mortality in the elderly population. Estimates suggest that currently, 44 million people worldwide live with this dementia, and projections for the year 2050 indicate that these numbers will triple. The main neuropathological features of AD are the accumulation of diffuse and neuritic plaques, marked by the extracellular deposition of beta-amyloid, which induces the spread of neurofibrillary tangles of hyperphosphorylated tau protein in the brain tissue. Although its pathogenesis remains uncertain, the most essential and frequently encountered clinical manifestation of AD is the selective impairment of memory and cognition. While there are treatments that can improve some symptoms of the disease, there is no cure, and the disease progresses

inevitably in all patients. **Objective:** To determine the frequency of hospitalizations due to AD in the different federal regions of Brazil and compare this frequency with the mortality rates in the same regions over the past five years. **Methods:** In this study, the Hospital Information System (HIS) was used to collect data from January 2019 to December 2023, while population data from the 2022 IBGE census were used to calculate prevalence. Mortality rates were extracted from the HIS for the period from January 2019 to December 2023. **Results:** During the study period, there were 7866 hospitalizations due to AD in Brazil. The federal region with the highest prevalence (per 100,000 inhabitants) was the South (6.72), followed by the Southeast (4.33), Central-West (3.17), Northeast (2.56), and North (1.57). Despite the higher prevalence of cases in the South, the annual total processing mortality rate was highest in the Southeast (30.07), followed by the Central-West (27.96), South (19.36), Northeast (16.24), and North (15.07). **Conclusion:** Based on the data, the South region (6.72) recorded the highest number of hospitalizations for AD, followed by the Southeast (4.33), Central-West (3.17), Northeast (2.56), and North (1.57). However, the annual total processing mortality rate was highest in the Southeast (30.07). These regional discrepancies suggest the need for a more in-depth analysis of the factors contributing to the prevalence and outcomes of AD in different areas of the country. Furthermore, the importance of early diagnosis and management strategies for AD across the national territory is highlighted. Equally crucial is ensuring equitable access to quality healthcare in all regions of the federation.

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Comparison of the prevalence of hospitalizations due to Parkinson's disease in the federal regions of Brazil and the mortality rate

Samuel José Feres Ozava[1], Guilherme Galardino Santos[1], Bernardo Caetano Novaes[1], Victor Bruno dos Santos Teixeira[1], Davi Melo de Hollanda Cordeiro[1], Myllene Rodrigues Silva[1]

[1]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.

Correspondence: Guilherme Galardino Santos; Email: guilhermegalardino@gmail.com.

Background: Parkinson's disease (PD) is the most common progressive neurodegenerative movement disorder, characterized by numerous motor, non-motor, and neuropsychiatric symptoms that can impact an individual's life to varying degrees. It is caused by the progressive degeneration of dopamine-producing neurons in the substantia nigra of the midbrain. PD is considered a syndrome that manifests with resting tremor, bradykinesia, rigidity, and loss of postural reflexes, which are regarded as the cardinal signs of the disease, thereby impairing the patient's function and quality of life. **Objective:** To determine the frequency of hospitalizations due to PD in the different federal regions of Brazil and compare this frequency with the mortality rates in the same regions over the past five years. **Methods:** In this study, the Hospital Information System (HIS) was used to collect data from January 2019 to December 2023, while population data from the 2022 IBGE census were used to calculate prevalence. Mortality rates were extracted from the HIS for the period from January 2019 to December 2023. **Results:** During the study period, there were 7866 hospitalizations due to PD in Brazil. The federal region with the highest prevalence (per 100,000 inhabitants) was the South (4.32), followed by the Southeast (2.37), Central-West (1.58), Northeast (1.21), and North (1.09). Despite the higher prevalence of cases in the South, the annual total processing mortality rate was highest in the North (9.52), followed by the Northeast (9.60), Central-West (9.34), Southeast (7.58) and South (6.81). **Conclusion:** Based on the data, the South region (4.32) recorded the highest number of hospitalizations for PD, followed by the Southeast (2.37), Central-West (3.17), Northeast (2.56), and North (1.57). However, the annual total processing mortality rate was highest in the North (9.52). These regional discrepancies suggest the need for a more in-depth analysis of the factors contributing to the prevalence and outcomes of PD in different areas of the country. Furthermore, the importance of early diagnosis and management strategies for PD across the national territory is highlighted. Equally crucial is ensuring equitable access to quality healthcare in all regions of the federation.

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Comparison of years of life lost due to ischemic stroke between two countries with a public health system: 5-year analysis

Luan Victor Silva Ribeiro[1], Hálisson Ferreira Freitas Filho[1], Izabella Carolline Menescal Ferreira[1], Larissa Suzan Basilio e Silva[1], Felipe José Silva e Silva[1], João Victor Souza de Barros[1], Igor Moreira Miguez Godoy[3], Kaliana Kennedy Oliveira Calixto[1], Antônio Marcos da Silva Henriques[1], Nathália Coimbra Coêlho[1]

[1]Afya Faculdade de Ciências Médicas, Abaetetuba PA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.
[3]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.

Correspondence: Luan Victor Silva Ribeiro; Email: luanvictorsilvaribeiro21@gmail.com.

Background: Ischemic stroke occurs when there is arterial obstruction, causing paralysis of brain areas, which prevents the passage of oxygen due to the lack of blood circulation. It is one of the main causes of death worldwide, increasing early mortality in several countries with different realities, such as Brazil and Canada. **Objective:** To compare the rate of years of life lost (YLLs) due to ischemic stroke between two countries with public health systems over five years, identifying differences and trends in early mortality, providing data to guide public health policies and prevention strategies. **Methods:** This is an observational, descriptive epidemiological study in which data on years of life lost (YLLs) due to stroke from 2017 to 2021 were obtained from the Institute for Health Metrics and Evaluation (IHME) platform, in the Global Burden of Disease (GBD) section. Information on cerebrovascular disease was selected and data on annual ischemic strokes was filtered out. The YLLs were calculated by multiplying the difference between the country's life expectancy and the age at death by the number of people who died from the disease in question. In order to make the appropriate correlation with the years of the fourth decade of life and calculate the rate (per 100,000 inhabitants), the age groups of both sexes, 30 to 34 years and 35 to 39 years, classified as age group 1 and age group 2, respectively, were selected. **Results:** In both countries, age group 2 had the highest rates in all the years analyzed. In the first year, 2017, men in Canada had a rate of 6.86 and women 7.6. In Brazil, in the same year, the male rate was 37.09 and the female rate was 31.34. In 2018, the second year of analysis, the male rate in Canada was 11.23 and the female rate was 8.77. In Brazil, the rate for men was 40.63 and for women 35.69. In 2019 in Canada, the male rate was 12.02 and the female rate was 9.70. In Brazil, the male rate was 40.99, and the female rate was 38.19. In the penultimate year, 2020, in Canada, the male rate was 12.38, and the female rate was 9.56. This year in Brazil, the male rate remained at 40.99, and the female rate increased slightly to 38.42. Last year, for 2021, in Canada, the male rate was 11.96, and the female rate was 9.10. In Brazil, the male rate was 42.98 and the female rate was 40.16. **Conclusion:** It was concluded that age group 2 had the highest rates in all the years analyzed in both Canada and Brazil. There was a general increase in rates in both countries over the period analyzed, with an increase of 74.3% in Canadian men, 19.7% in Canadian women, 15.8% in Brazilian men and 28.1% in Brazilian women in 2021 compared to 2017, with Brazil showing significantly higher rates than Canada.

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Compensated idiopathic intracranial hypertension due to traumatic cerebrospinal fluid fistula: a case report

Apolônio Peixoto de Queiroz[1], Breno Bezerra Arruda Câmara[1], Yan Carlos de Sousa Diniz[2], Ana Clara Melo de Medeiros[2], Joan Ravy Sousa Lacerda[2], Maira Monteiro Amorim[2], Ana Karoline Alves de Brito Marques[2], Rhawana Lorrana Oton Guimarães[2], Maria de Fátima Trigueiro Silva[2], Regina Dayana de Morais Santos[2]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[2]Centro Universitário de Patos, Patos PB, Brazil.

Correspondence: Apolônio Peixoto de Queiroz; Email: apolopxq@gmail.com.

Case presentation: A 33-year-old obese female was followed in an outpatient headache clinic. In 2018, she presented with progressive holocranial headache, worsening with dorsal decubitus, and transient visual darkening. She was diagnosed with idiopathic intracranial hypertension (IIH) after cerebrospinal fluid (CSF) collection and exclusion of other causes. She was treated with topiramate and made lifestyle changes, including regular physical activity, dietary adjustments, and weight loss, leading to significant symptom improvement for several years. In January 2023, she suffered a mild traumatic brain injury (TBI) after hitting her head on a cabinet. Days later, she experienced intense, almost daily headaches different from her previous ones. The new headaches worsened or appeared when upright and were accompanied by clear, colorless, odorless, constant rhinorrhea in the right nostril, ear fullness, nausea, and occasional vomiting. She stopped her medication, resulting in partial symptom improvement. During a follow-up appointment, she reported these new symptoms. An MRI with cerebrospinal fluid flow study revealed a frontoethmoidal meningoencephalocele through a bone defect in the cribriform plate. After consulting with the Neurosurgery team, the decision was made to proceed with a ventriculoperitoneal shunt instead of correcting the fistula. Post-procedure, she showed significant improvement and was discharged without symptoms. **Discussion:** Disorders of intracranial pressure, such as intracranial hypertension (IH) and cerebrospinal fluid hypotension (CSFH), cause headaches and can significantly impair quality of life. These conditions often present with transient visual loss, tinnitus, and gait and balance disturbances, with positional headache being the main symptom. This patient was initially diagnosed with IIH, which was well-managed with medical treatment and lifestyle changes for several years. However, the mild TBI in 2023 caused a notable change in her headache pattern and additional symptoms, including rhinorrhea and orthostatic headache, indicative of a cerebrospinal fluid leak. MRI findings confirmed a frontoethmoidal meningoencephalocele, a rare complication post-head trauma leading to cerebrospinal fluid fistula. **Final comments:** This case demonstrates a secondary headache due to IIH that was well-controlled for years but exacerbated by a mild TBI, leading to a new headache pattern and additional symptoms. The diagnosis of cerebrospinal fluid fistula and meningoencephalocele was confirmed through imaging studies. Opting for a ventriculoperitoneal shunt instead of surgical fistula correction was effective, providing significant symptom relief. In conclusion, this case highlights the importance of thorough evaluation and appropriate management in patients with changes in headache patterns following head trauma.

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Complex case of trismus, ischemic stroke and neuromuscular disorder: a diagnostic challenge

Larissa da Cruz Cardoso[1], Isadora Ribeiro Doria[1], Beatriz Palladino Ferreira[1], Augusto Costa Ataides[1], Sandy Karoline Carnaúba dos Santos[1], Matheus Henrique Fonseca Melo[2], Agnaldo Queiroz Plaster[3], Maria Luísa Ribeiro Lima[4], Evellyn Fatima Rasia[1], Mireli Martins Montijo[1]

[1]Universidade do Estado de Mato Grosso, Cáceres MT, Brazil.
[2]Faculdade de Ciências Médicas, Itabuna BA, Brazil.
[3]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.
[4]Centro Universitário Governador Ozanam Coelho, Ubá MG, Brazil.

Correspondence: Larissa da Cruz Cardoso; Email: lari_osodrac@hotmail.com.

Case presentation: The patient, MGSB, is a 58-year-old female with no known allergies to medications. She denies smoking or drinking alcohol. She sought hospital care after experiencing multiple ischemic strokes, trismus for 3 days, and tongue protrusion in a lateral frontal bite. Upon physical examination, she was in good general condition, alert, and hydrated. The neurological examination revealed a Glasgow Coma Scale score of 15, with reduced strength in the right upper limb, changes in motor coordination, aphasia, and photoreactive pupils. The diagnosis evolved with a new episode of subacute ischemic stroke, bronchoaspiration, and severe oropharyngeal dysphagia. A cerebrospinal fluid examination was performed, which was clear and colorless, with no significant changes. A head CT scan showed gliosis in the left temporoparietooccipital region and the right precentral gyrus, as well as intracranial atheromatosis. MRI revealed areas of encephalomalacia bordered by gliosis in the left fronto-parietal transition and the left occipital lobe. To investigate the hypotheses of trismus and other neuromuscular changes, laboratory tests were conducted to probe metabolic, infectious, and structural etiologies. However, the test results did not provide reasonable evidence, making the case an important scientific concern. The tests obtained were: non-reactive ANA, negative Anticardiolipin IgM and IgG, and non-reactive Factor V Leiden. The only notable change observed was an elevated Antithrombin III level at 132%. **Discussion:** The clinical picture presents a heterogeneous neurological syndrome, making the case difficult to classify. The result of the lupus anticoagulant was weakly positive and will need to be repeated after six months, indicating Systemic Lupus Erythematosus as a diagnostic possibility. Clostridium research was also conducted to search for botulism, along with diagnostic tests for myasthenia gravis and cephalic tetanus, all of which were negative. The current diagnostic hypothesis centers around autoimmune encephalitis. To justify this diagnosis, analysis of CSF and anti-GAD serum was requested. This research is based on the patient's clinical condition, which includes signs of cerebellar ataxia, epilepsy, and the neuromuscular disorder trismus. As a therapeutic test, immunoglobulin will be used to observe the clinical condition and to determine if there is an association with autoimmune encephalitis. **Final comments:** The patient's clinical condition points to a variety of possibilities that are not supported by the test results obtained. The difficulty in establishing a reliable diagnosis prevents therapeutic decisions from being made, resulting in longer hospital stays and clinical worsening of the patient. This challenge hinders early identification and appropriate management, which are essential to reduce the morbidity and mortality associated with the clinical picture.

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Complex dementia case: overlapping symptoms of vascular, Alzheimer's, and frontotemporal dementia with adult leukoencephalopathy

Lucas de Andrade Saraiva[1], Paulo Caramelli[1], Elisa de Paula França Resende[1], Leonardo Cruz de Souza[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Lucas de Andrade Saraiva; Email: lucasdeandradesaraiva@gmail.com.

Case presentation: Mr JAS, 73, married, from Betim, 4 years of schooling, a baggage handler, admitted to the neurology clinic on 09/23. Since the pandemic, the patient has been experiencing severe irritability, which escalated to self-harm by hitting himself in the head. His behaviour deteriorated further, displaying uninhibited behaviour such as disclosing sensitive information about a past relationship with a prostitute to his family, heightened verbal aggression, and paranoid jealousy toward his wife. The patient's wife reported that he had a history of learning difficulties and displayed long-term uninhibited behaviour, including verbal aggression, racism, inappropriate jokes; and delusional jealousy towards his wife. Despite these behaviours, he was able to work as a baggage handler at the bus station and use public transport independently. The patient reported obsessive thoughts about buying a coffin, as well as exhibiting compulsive behaviour related to ordering. The wife also mentioned discreet social apathy and mild dysarthria in the last 2 months. Comorbidities: history of cardiac arrhythmia. Family history: His mother had epilepsy, Alzheimer's disease (AD) at age 75, and multiple strokes. His father had AD at age 70. Physical examination findings: reduced visual fields in both eyes, cogwheel joint stiffness, and mild heightened reflexes in the lower limbs. Head CT: showed diffuse atrophy, mild bilateral frontal and temporal predominance and white matter lesions (Fazekas 3). Brain MRI: showed extensive multifocal lesions, with hypersignal on T2/FLAIR and hyposignal on T1. Hippocampi showed moderate atrophy (MTA 3). Brief Cognitive Screening Battery (BCSB): poor performance in naming, learning memory, and delayed memory. **Discussion:** The patient exhibits characteristics indicative of behavioral variant frontotemporal dementia (bvFTD), including behavioral disinhibition, compulsive behavior, and apathy, supported by CT scan findings of consistent atrophy patterns. While bvFTD is likely, AD cannot be ruled out due to episodic memory impairment and moderated hippocampal atrophy (MTA 3). Recent onset of dysarthria, coupled with cardiac arrhythmia and white matter lesions (Fazekas 3), suggest a mixed dementia presentation with bvFTD/AD and cerebrovascular disease, warranting consideration. The latest MRI findings indicate atrophy patterns consistent with adult Leukoencephalopathy, a condition marked by movement disturbances, vision impairments, balance issues, memory loss, behavioural alterations, and attention deficits, some of which the patient displays. In certain instances, this condition may advance to dementia. **Final comments:** In summary, Mr. LFC's case presents a diagnostic challenge due to symptoms suggestive of multiple dementia etiologies. Further assessments are needed to accurately identify the underlying pathology and guide management strategies for his cognitive decline.

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Complex neurological manifestations in a case study of narcolepsy and autosomal dominant ataxia

Jonathas Taumaturgo Soares[1], Danilo Lemes Reis[1], Diogo Rabelo de Lima[1], Victor Hugo Soares dos Santos[1], Carlos Maurício Oliveira de Almeida[1], Marcus Vinicius Della Colletta[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Jonathas Taumaturgo Soares; Email: soaresjonathas2004@gmail.com.

Case presentation: A 54-year-old female patient of Italian descent was referred for neurological evaluation due to tremors. At the age of 40, she began experiencing tremors in her left upper limb, which progressed to her right upper limb within a month, accompanied by difficulty in writing. At 44, she noticed progressive hearing loss, and by 46, she had difficulty walking. She also reported episodes of inappropriate daytime naps since the age of 25, agitated sleep with nocturnal vocalizations, and excessive movements during sleep. She described episodes of falls initiated by trunk and lower limb weakness without loss of consciousness and episodic in nature. Upon physical examination, mild bradykinesia, bilateral resting and postural tremor, ataxic gait, and slight dysmetria were noted. Epworth Sleepiness Scale: 13/24. RBDSQ-Br Questionnaire: 5/13. Pittsburgh Sleep Quality Index (PSQI): 8. Audiometry: Bilateral hearing loss. Brain MRI: Hypo signal in T2 in bilateral globus pallidus and midbrain. PANK2 negative. Polysomnography with Multiple Sleep Latency Test (MSLT): AHI 14/hour, PLMI 52/hour; Sleep efficiency 96%, presence of REM without atonia, average latency of 4.64 minutes, and presence of 3 SOREMPs. EEG: Normal; CSF: Normal; Molecular Test: p570 Ala-Val mutation in Exon 21 of the DNMT1 gene. **Discussion:** Narcolepsy is an autoimmune and genetic disorder and is the most common cause of central hypersomnia (ICSD-3). Narcolepsy is caused by the selective loss of orexinergic/hypocretinergic neurons in the lateral hypothalamus, which promote wakefulness. There are two described forms: Narcolepsy type I (with cataplexy) and type II (without cataplexy), and its diagnosis requires PSG with MSLT demonstrating a mean sleep latency of less than 8 minutes and 2 or more SOREMPs, or reduced hypocretin levels in CSF for Narcolepsy type I. The DNMT-1 gene is crucial for DNA methylation and is located on chromosome 19. Three phenotypes have been described for DNMT-1 mutation, all with an autosomal dominant pattern: Hereditary Sensory Neuropathy with Dementia and Hearing Loss (Exon 20), Ataxia, Deafness, and Narcolepsy (Exon 21), and the third with overlapping manifestations. The initially described cases were observed in Italian immigrants, like our patient. The described case represents one of these phenotypes. **Final comments:** Narcolepsy can be one of the manifestations of the DNMT-1 gene mutation. In this case, the presence of deafness and ataxia, in addition to narcolepsy, represents one of the recently described phenotypes.

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Complication of herpetic encephalitis: Klüver-Bucy syndrome: clinical case

Daniele da Costa da Silva[1], Carolina Bonifácio Gomes[1], Ana Carolina Grande[1], Ivo Marcos Darella Lorenzin Fernandes Neto[2], Eduarda Basso Badalotti[2], Angelo Marcelo Wosniacki Filho[2], Gabriel Angelo Garute Zenatti[2], Renato Endler Iachinski[2], João Luis Corso Bandeira[2], Antonio Carlos de Andrade Soares[2]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Fundação Hospitalar São Lucas, Cascavel PR, Brazil.

Correspondence: Daniele da Costa da Silva; Email: dani.costadasilva09@gmail.com.

Case presentation: A 51-year-old female patient was admitted with progressive confusional status that started one day ago. On arrival, the patient presented on postictal status after two convulsive episodes and no other neurological abnormalities on clinical examination. On further investigation, a CSF and a magnetic resonance with spectroscopy were performed showing an area of signal alteration affecting part of the left cerebral hemisphere, with restriction to the diffusion and with slight leptomeningeal enhancement in the temporal pole. During the exam, a lowering of the level of consciousness was observed and the patient was transferred to the ICU after intubation. Suspected of herpetic encephalitis and started IV Acyclovir. During hospitalization, she developed visual agnosia, placidity, apathy to external stimuli, and

amnesia, leading to a diagnosis of partial Kluver-Bucy syndrome. Discharged after twenty-eight days, presenting with expressive and receptive aphasia and showing partial improvement of mental confusion. **Discussion:** Nervous system infections' clinical manifestations vary according to the pathogen involved, the severity, and the location of the lesion. Initially, fever, changes in the level of consciousness, and convulsions can be acute or subacute. Diagnosis is carried out through lumbar puncture. Imaging exams and electroencephalogram can be performed. Herpes simplex encephalitis generally affects the temporal lobes and adjacent limbic systems, being a common change to meningeal congestion. Behavioral syndromes, such as Klüver-Bucy syndrome, amnesia, and hypomania, may be associated. Treatment is carried out with acyclovir, which is the only medication recommended for this viral condition. Furthermore, KBS is associated with temporal lobe lesions and is characterized by hyperorality, hypersexuality, eating disorders, placidity, visual agnosia, and amnesia. The diagnosis is clinical, and the progression of the syndrome varies in each patient, with treatment focusing on symptom management. **Final comments:** Herpes simplex encephalitis is considered a neurological emergency that must be investigated and treated due to its high morbidity and mortality rate. Lumbar puncture and initiation of treatment with intravenous acyclovir must occur early to ensure a better prognosis. Additionally, continuous monitoring of patients and their families is essential to assist in the rehabilitation phase after treatment.

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Comprehensive analysis of the clinical and genetic features of RYR1-related myopathies

Alulin Tácio Quadros Santos Monteiro Fonseca[1], Clara Gontijo Camelo[1], André Macedo Serafim da Silva[1], Cristiane de Araújo Martins Moreno[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Alulin Tácio Quadros Santos Monteiro Fonseca; Email: alulin.fonseca@gmail.com.

Background: The RYR1 gene encodes the skeletal muscle ryanodine receptor, an ion channel responsible for regulating the release of calcium stored in the sarcoplasmic reticulum. Variants in the RYR1 gene are responsible for a wide variety of phenotypes in myopathies, with autosomal dominant (AD) and autosomal recessive (AR) inheritance patterns. **Objective:** We present the clinical and genetic data of 24 patients (from 23 families) with RYR1-related myopathies with either AD or AR inheritance patterns, aged between 1 and 61 years (mean age 16.3 years). **Methods:** Clinical, histological, and molecular data were collected. Clinical information from patients was gathered cross-sectionally between November 2022 and June 2024. Muscle biopsy data were evaluated retrospectively. The RYR1 gene was analyzed using next-generation sequencing. **Results:** An AR inheritance pattern was observed in 13 patients (54%). All variants with AD inheritance patterns were missense. In the AD group, two variants (p.Arg4861His and p.Ile4898Thr) were observed in two patients each. Among the 12 AR families, 50% had truncating variants (splice site or frameshift) associated with a missense variant. Except for one patient who began experiencing weakness in adulthood, all others had a congenital myopathy diagnosis. Eight patients (33%) had reduced intrauterine movement. One patient had bilateral congenital hip dislocation. The mean age at gait acquisition was 1 year and 11 months, with three patients aged between 6 and 19 years never acquiring gait. The oldest age for gait acquisition was 5 years. Eyelid ptosis was observed in 54% of patients. Ophthalmoparesis was seen in 46% of AR cases and in none of the AD cases. Facial weakness was present in 45% of cases, and 70% had a high-arched palate. Trophic changes of the tongue were observed in 69% of patients. Scapular winging was present in 6% of cases. Muscle weakness of the cervical flexors and rectus abdominis was noted in most cases. Joint contractures were more prominent in AR patients, particularly affecting the Achilles tendon. Spinal deformities and rib cage deformities were seen in 45% and 58% of patients, respectively. Two patients required a g-tube, and three were on ventilatory support. Prominent dynamic symptoms (fatigue, myalgia, and cramps) were reported in 37.5% of patients. No patient had a history of rhabdomyolysis or malignant hyperthermia. CPK levels ranged from 32 to 2396 IU/L. Muscle biopsy was performed in 16 patients (66%). A central core pattern was observed in the AD group. In AR patients, the most prominent histological findings were nuclear centralization, mild dystrophic changes, and uneven oxidative staining. **Conclusion:** RYR1-related myopathies are a heterogeneous group of diseases, mostly presenting congenitally. AR forms exhibit more prominent ophthalmoparesis, trophic tongue changes, and joint contractures. Histological changes are more diverse in the AR group.

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Comprehensive step-by-step clinical assessment for managing action-induced focal dystonia after bilateral gpi deep brain stimulation for Parkinson's disease

Flávia de Paiva Santos Rolim[1], Verônica Tavares Aragão[2], Joanisson Rubens Gomes Diniz[2], Ana Silvia Sobreira Lima Verde[2], André Borges Ferreira Gomes[2], Fernanda Martins Maia Carvalho[1]

[1]Universidade de Fortaleza, Fortaleza CE, Brazil.
[2]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Flávia de Paiva Santos Rolim; Email: flaviarolim@unifor.br.

Case presentation: We report a case of a PD male patient diagnosed at 32 years-old, who underwent bilateral Globus Pallidus internus (GPi) DBS at 43 years-old due to severe motor complications. Despite excellent response to levodopa, he experienced incapacitating ON dyskinesias. Post-DBS, he achieved complete remission of dyskinesias. However, levodopa dosage optimization was necessary to manage wearing off phenomena, despite DBS adjustments. Few weeks after a regular programming session, he reported unpredictable involuntary muscle contractions during right biceps activation, which impaired functionality but disappeared at rest. Movement phenomenology resembled focal dystonia, triggered solely by flexion and extension of the elbow during tasks such as brushing teeth, eating, writing, or other activities requiring biceps activation. This initially led us to hypothesize that the symptom might be related to stimulation, so we opted to progressively reduce stimulation in the left GPi. However, this resulted in worsening of parkinsonism. We also attempted to change the stimulated contacts based on previous monopolar review. A trial of amantadine was started in an attempt to cover a possible peak dyskinesia, but with no improvement after reaching a dose of 300mg. Despite different approaches and adjustments, the involuntary movement persisted. A structured troubleshooting process was initiated to differentiate between potential causes: 1) OFF Medication, ON Stimulation: Baseline assessment conducted 12 hours without dopaminergic medication. 2) OFF Medication, OFF Stimulation: Assessment without pharmacological or DBS influence. 3) ON Medication, OFF Stimulation: Evaluating the isolated effect of levodopa. 4) ON Medication, ON Stimulation: full therapeutic state assessment. During the third phase (ON Medication, OFF Stimulation), the previously unobserved action-induced dystonic movement emerged. This indicated that the dystonic movement was likely related to medication (levodopa-induced). We then decided to reduce the total daily levodopa dose, with partial clinical improvement, in parallel with image reconstruction and neuroimaging-based DBS programming. **Discussion:** This case highlights the need for meticulous clinical evaluations for PD patients with DBS. Understanding medication and stimulation interactions is decisive for managing complex motor symptoms. GPi's somatotopic organization places the face and arm posteriorly and ventrally, and the leg more centrally and dorsally. Deviating from the targeted stimulation area could led to adverse effects, such as the focal dystonia seen in our patient. **Final comments:** This case underscores the intricate balance required in managing advanced PD with DBS. Step-by-step evaluation and neuroimaging-based troubleshooting process provides a comprehensive framework for distinguishing between medication-induced and stimulation-induced movement disorders, ensuring precise and effective therapeutic strategies.

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Concomitant elevation of pro-inflammatory cytokines IL-6, IL-10 and neurodegeneration biomarkers neurofilament light chain (NFL) as potential biomarkers for severe COVID-19 outcomes

Helena França Alcaraz Ferreira[1,2], Andreza Salvio Lemos[1], Renan Amphilophio Fernandes[1], Isabela Stornelli Teixeira[1], Carlos Henrique Ferreira Ramos[2], Carlos Henrique Ferreira Ramos Filho[3], Wagner Luís da Costa Nunes Pimentel Coelho[4], Luciane Almeida Amado Leon[4], Soniza Vieira Alves-Leon[1,2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[4]Fundação Oswaldo Cruz, Rio de Janeiro RJ, Brazil.

Correspondence: Helena França Alcaraz Ferreira; Email: helena.faf33@gmail.com.

Background: The COVID-19 pandemics has left some consequences in the way we understand host-antigen interactions, despite the great advances during the period, there is still much to be understood about the extension of this disease's impacts on the human body. The cytokine storm resulting from the host's exacerbated response to the virus, as well as its innate ability to infect tissues beyond its point of entry, gives the disease inflammatory and systemic properties. **Objective:** To investigate the concomitant association between the interleukins 6,10 (IL-6, IL-10) and the neurofilament light chain (NfL) level with the death outcome. **Methods:** Peripheral blood had been collected in order to investigate the relationship between the protein biomarker of neurological damage and the anti-inflammatory cytokines. Statistical analysis using the Kruskal Wallis and Mann-Whitney U test with a cohort of 32 critical COVID-19 patients were made. The court was divided between the outcome Deceased and Survivor, and Presence (PNS) or Absence (ANS) of Neurological Symptoms. Each of these groups was compared with its homologous biomarker from a healthy control group. The data evaluation was performed using the SIMOA platform for the NfL, and LUMINEX 200 system for the cytokines analysis. **Results:** The results showed there was a significant difference between the deceased and control patients for both IL-6 and IL-10 expression (p=0.01022) when comparing deceased to the survivor group (p=0.02036). Of the cohort of 32 patients, 9 were classified as PNS, of which 6 were also part of the Decease group with high NfL rates compared to Survivor and control groups (NfL Decease = 805.6±2,696pg/mL; NfL Survivor = 109.9±192.3pg/mL; NfL Heath control = 12.49±5,210pg/mL). **Conclusion:** It is already established from previous studies that an exacerbated immune response may be related to the worsening of the condition, as well as the increase in NfL which has been associated with the death outcome. These variations, combined with the clinical features of the patients, may be an indication of the relevance of these biomarkers for the monitoring of COVID-19 evolution, as well as the important role of inflammatory response in COVID-19 severity.

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Congenital insensitivity to pain due to SCN9A gene mutation: a case report

Josiane de Souza[1], Daniel Almeida do Valle[1], Jaqueline Souza Chaves Taniguchi Leite[2], João Pedro Tejerina Bader Maluf[2], Julianna Rodrigues Beltrão[2], Lucas Schiavon[2], Maria Valentina Denardini[2], Marina Faria Negrão[2]

[1]Faculdade Pequeno Príncipe, Curitiba PR, Brazil.
[2]Pontifícia Universidade Católica do Paraná, Curitiba PR, Brazil.

Correspondence: Marina Faria Negrão; Email: mfnegrao@outlook.com.

Case presentation: The patient is a male, the only child of a non-consanguineous couple, with no significant changes related to gestation and appropriate neuropsychomotor development throughout childhood. The investigation of the patient's clinical condition began at 7 months of age and continued for the following 6 years after multiple hospitalizations due to episodes of self-aggressiveness, mutilation, fractures, and recurrent osteomyelitis. The guardians reported episodes of biting and abrasions on multiple parts of the body, resulting in deep wounds with heavy bleeding, as well as occurrences such as direct contact of the skin with fire or motorcycle exhaust. In none of the described events did the patient show signs of pain, crying, reflex, or discomfort. During the years of the patient's follow-up, possible diagnoses such as storage disease, rheumatic diseases, and immunodeficiencies were investigated, without success in defining the etiology. The diagnosis of congenital insensitivity to pain was made at the age of 7, after performing a whole genome sequencing test revealing the pathogenic variant c.2723G> A, p.(Trp908Ter) in exon 16 of the SCN9A gene in homozygosity. This alteration is responsible for generating a premature stop codon and consequently a truncated protein. In the medical record documented at the age of 7 years and 3 months, the patient had good school performance with preserved intellect, which is consistent with the phenotype of the SCN9A variant presented by the patient. **Discussion:** Congenital insensitivity to pain is a rare autosomal recessive neuropathy characterized by damage to small-caliber sensory C fibers and/or A-delta fibers of the peripheral nerves. This alteration results in impaired perception of nociceptive, mechanical, and thermal stimuli. Although pain insensitivity is a universal symptom, each pathogenic variant presents specific peculiarities related to the affected gene, resulting in multiple phenotypes of the disease. Consequently, children with this condition lose an essential defense mechanism, potentially leading to self-mutilation habits and recurrent injuries. Furthermore, this condition results in significant psychological challenges, such as the need for continuous monitoring of the patient, affecting the child's self-esteem and social interactions. Lastly, it is important to highlight the relevance of genetic counseling in this case, which aims to clarify the risks of recurrence of the mutation, enabling the family to make informed decisions regarding future pregnancies. **Final comments:** The reported case involves a patient with a rare mutation in the SCN9A gene, suspected and identified through episodes of self-mutilation and hospitalizations for recurrent fractures. Understanding the molecular, clinical, and genetic characteristics not only provides a comprehensive view of the disease but also contributes to new diagnostic and therapeutic strategies.

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Congenital limb-girdle muscular dystrophy: case report with molecular diagnosis

Ana Beatriz Landim Ribeiro[1], Giovana Rodrigues França[1], Luciana Amorim de Matos[1], Carlos Alexandre Gomes Farias[1], Paulo Victor Zattar Ribeiro[2], Lisandra Mesquita Batista[2], Lucca Biolcati Palavani[3]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Hospital das Clínicas, Ribeirão Preto SP, Brazil.
[3]Max Planck University Center, Berlim, Germany.

Correspondence: Ana Beatriz Landim Ribeiro; Email: ablr2016@icloud.com.

Case presentation: Male patient, currently 3 years old, with a history of fetal exposure to alcohol and cocaine, without complications during delivery. At two months of age, he presented with generalized hypotonia and dysphagia. Developmentally, delays in neuro psychomotor milestones were noted, particularly delayed acquisition of walking. Upon physical examination, a prominent forehead, apparent brachycephaly, anteverted nares, bilateral knee flexion contractures, and elbow region contractures were observed, with more prominent appendicular hypotonia in the upper limbs. In 2020, a brain MRI revealed diffuse T2 hyperintensity in white matter, confluent, affecting fibers, pronounced T1 hypointensity, and spectroscopy indicating reduced neuronal viability, findings suggestive of metachromatic leukodystrophy at the time. Additionally, he has a normal ECG and an absence of pathogenic variants in the spinal muscular atrophy (SMA) gene. Therefore, complete exome sequencing was performed, revealing compound heterozygous variants in the intronic region following exon 36 of the LAMA2 gene. The patient continues to be followed up, with appropriate screenings up to date. **Discussion:** Congenital muscular dystrophy type 1A (MDC1A) is an autosomal recessive inherited myopathy, typically characterized by peripheral hypotonia, respiratory insufficiency, loss of muscle mass, and spontaneous movement, all symptoms observed in the case. It may also be associated with gastroesophageal reflux and recurrent thoracic infections. MDC1A manifests from birth or early childhood, with an estimated prevalence of 1 in 30,000 individuals. The pathophysiology of the disease results from pathogenic variants in the LAMA2 gene, which encodes the alpha-2 chain of laminin. Diagnosis can be made through a muscle biopsy, where a deficiency of merosin in muscle and skin can be detected. In this case, the patient exhibits feeding difficulties with neuro psychomotor causes. This can be explained as the disease progresses, weakness of facial muscles, temporomandibular joint contractures, and macroglossia can impair feeding and speech. **Final comments:** Mutations in the LAMA2 gene affect the production of the α2 subunit of laminin-211 (= merosin) and result in either partial or complete laminin-211 deficiency. Complete merosin deficiency, as seen in our patient, is typically associated with a more severe congenital muscular dystrophy (CMD), clinically manifested by hypotonia and weakness at birth, the development of contractures of large joints, and progressive respiratory involvement. So, the diagnosis of this condition should be remembered in cases like ours, primarily when severe muscle weakness, hypotonia, and white matter abnormalities in brain imaging are presented.

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Congenital miopathy presenting as mitocondrial miopathy: the relevance of genetic testing and semiological findings in chronic progressive external ophtalmoplegia

Yasmin Ramalho Mandarino[1], Luana Cristina Stefanes[1], Pedro Vitor Fortunato Silveira[1], Hannah Bang[2], Natália Cardoso Darabas[2], Pietra Cani Linzmeier[2], Thábata Emanuelle Martins Nunes[2], Maria Eduarda Angelo de Mendonça Fileti[2], Carla Heloísa Cabral Moro[2], Alexandre Luiz Longo[2]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Yasmin Ramalho Mandarino; Email: yasminramalhoo@gmail.com.

Case presentation: A 56-year-old female presented with progressive weakness in her lower and upper limbs since childhood, with description of neonatal hypotonia. She also reported dysphagia and dyspnea on exertion. Her medical history included hemochromatosis, type 2 diabetes, fibromyalgia, and a history of lumbar fracture. Neurological examination revealed grade IV muscle strength in the proximal lower limbs, limited bilateral eye abduction, and impaired upward gaze. Deep tendon reflexes were hypoactive. Electromyography and nerve conduction studies showed borderline myopathic findings. MRI indicated severe liposubstitution of the gluteus maximus and tensor fasciae latae muscles bilaterally. A genetic panel revealed a variant of uncertain significance in the ANO5 gene and a pseudodeficiency allele in the GAA gene. Genetic testing confirmed proximal myopathy and ophthalmoplegia associated with the MYH2 gene. Initially, mitochondrial myopathy was suspected due to the presence of external ophthalmoplegia, however, genetic test results were negative for this condition. **Discussion:** Congenital myopathies constitute a heterogeneous group of diseases in terms of clinical manifestations and genetic mechanisms, characteristic findings can be observed in muscle biopsy. The prevalence of congenital myopathy in the general population is 1.50 per 100,000. Hypotonia and weakness are common clinical findings in congenital myopathy typically observed in the first year of life, accompanied by hip dysplasia, hypomimia, and motor development delays. The presence of specific histopathological changes in muscle biopsy distinguishes these conditions from other neuromuscular disorders. The pathophysiology is complex, but the genes commonly involved affect calcium ion homeostasis, muscle contraction mechanisms, and thin and thick filament structures. More than 30 genes have been associated with congenital myopathies. The diagnosis is made through muscle biopsy, muscle MR and genetic tests. The disease is incurable, and treatment primarily focuses on providing supportive care. A possible differential diagnosis in this case would be progressive external ophthalmoplegia. However, unlike the patient described above, in this condition, patients always have ptosis. In cases of congenital myopathy with ophthalmoplegia, patients do not have associated ptosis. **Final comments:** Congenital myopathies constitute a heterogeneous group of diseases in terms of clinical manifestations and genetic mechanisms. Prognosis and severity vary according to the type of mutation involved, resulting in either a stable or progressive disease course. The absence of specific clinical symptoms and genetic diversity can make molecular diagnosis so complex that even specialist physicians often face difficulties in reaching an accurate diagnosis. The use of new molecular genetic sequencing techniques is essential for the development of future therapies.

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Congenital myasthenia gravis: a case report from the perspective of neurology

Camila Yukari Yokota Barbosa[1], Maria Eduarda Barbosa Stecca[1], Maria Luiza Azambuja Locatelli[1], Luiz Felipe Azambuja Locatelli[1]

[1]Centro Universitário Mauricio de Nassau, Cacoal RO, Brazil.

Correspondence: Camila Yukari Yokota Barbosa; Email: camilayukari2005@gmail.com.

Case presentation: A 3-year-old male patient, went to the doctor's office on 04/12/2023, with his grandmother, who reported eyelid ptosis and lower limb asthenia. After the consult, an anti-acetylcholine receptor antibody and a genetic test were solicited, due to suspicion of myasthenic gravis syndrome (MGS). The presence of anti-acetylcholine receptor antibodies was confirmed, reinforcing the diagnostic hypothesis of congenital myasthenia gravis. New muscle exams and genetic testing were required. fluoxetine hydrochloride was initiated. 15 days post-treatment, the exams did not suggest muscle

disease, maintaining fluoxetine hydrochloride for two months and then starting salbutamol sulfate (2.5 ml, 3x/day). On the last consult, genetic tests were negative, and a chest tomography showed no signs of thymoma. The patient was referred to a geneticist, but the consult did not occur due to geographic and socioeconomic issues. This report was approved by the CEP, opinion 6.902.166. **Discussion:** The patient presents typical signs and symptoms of congenital myasthenic syndrome (CMS), such as muscle weakness, eyelid ptosis that worsens throughout the day, and early age onset. As it is an uncommon disorder, diagnosis is usually late, leading to a worsening of clinical aspects. This case is considered rare because the diagnosis was established in early childhood. CMS is a rare disease involving the neuromuscular junction, characterized by Ach receptor defects, which can occur due to autosomal dominant, recessive, or sporadic mutations. For the diagnosis, it is necessary to assess the age at which symptoms appear, delay in psychomotor development, weakening muscle, paralysis or asthenia in eye muscles, skeletal deformities, inefficiency of immunosuppressive therapy, fatigable weakness, and ptosis. According to Abicht (2003), the most common characteristic of CMS is fluctuating muscle weakness, which worsens by use and improves at rest, as seen in this case. Hence, for an accurate diagnosis, it is crucial to rely on physical examination, laboratory findings, repetitive pharmacological stimulation tests, tests that demonstrate the presence of antibodies, response to anticholinesterase therapy, family history, and genetic testing. **Final comments:** In this report, the exams revealed anti-acetylcholine receptor antibodies, indicating MGS. Although the genetic consult had not occurred, clinical symptoms and laboratory findings strongly suggested the condition. The diagnostic complexity and the need for multifaceted therapeutic approaches highlight the uniqueness of this case and the importance of careful and personalized management for patients with CMS in pediatrics. Thus, the exclusion of MGS was necessary based on negative results of genetic and general exams, requiring better follow-up with a geneticist. Identification of genetic defects that lead to CMS is crucial for adequate treatment as they can occur in different locations of the neuromuscular junction.

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Congenital myopathy due to a mutation in the ACTA1 gene: case report

Jéssica Cristina Silveira Damasceno[1], Laura Altomare Fonseca Campos[1], Bruna Queiróz Vieira[2], Marcelo Sobrinho Mendonça[1], Thiago Cardoso Vale[1]

[1]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.
[2]Fundação Hospital Estadual do Acre, Rio Branco AC, Brazil.

Correspondence: Jéssica Cristina Silveira Damasceno; Email: drajessicadamasceno@gmail.com.

Case presentation: A 22-year-old man, previously asthmatic, with a history of delayed motor development, having only started walking at the age of 2, noticed proximal weakness in all 4 limbs at the age of 7. At 17 years old, he began experiencing episodes of short-term blurred vision and diplopia that resolved spontaneously. At 20 years old, he developed frequent episodes of liquid dysphagia and dyspnea on medium exertion, when he sought medical assistance due to the greater functional impairment. His family history was that his mother and sister have undiagnosed proximal weakness in the lower limbs. Laboratory investigation, including creatine phosphokinase, was normal. Electroneuromyography demonstrated signs of global myopathy with proximal predominance with (brief) myotonic bursts. Due to the suspicion of congenital myopathy, a broad DNA panel for neuromuscular diseases was carried out in July 2022 with the presence of heterozygosity for the actin alpha 1 (ACTA 1) gene, suggesting a diagnostic hypothesis of nemaline myopathy (NM). The patient was last seen at the end of 2022, when he remained with a grade 4 strength in the limbs without facial weakness and did not require ventilatory assistance. Until that time, no muscle biopsy had been performed. **Discussion:** Mutations in the ACTA1 gene most frequently cause NM, but they can cause other forms of congenital myopathy less frequently. Among these, the main differential diagnosis from NM is myopathy due to disproportion of muscle fibers. Myopathy due to disproportion of muscle fibers occurs due to a selective atrophy of type 1 fibers, which are up to 80% smaller than type 2 fibers, presenting a clinical picture of significant proximal, ocular, facial and cervical weakness that begins at birth. NM, on the other hand, has great clinical heterogeneity, ranging from conditions with the onset of severe hypotonia at birth and even forms with a later onset and mild muscle weakness. However, in most cases of NM, forms with moderate and mild symptoms predominate. In NM biopsy, there are nemaline bodies or rods in the sarcoplasm of the muscle fibers, although fiber disproportion may also occur. Moreover, if the fibers are very small, it may be difficult to detect nemaline bodies. Given that the diagnosis of forms of congenital myopathy requires a muscle biopsy, in the present case it was not possible to determine the patient's histopathological diagnosis. **Final comments:** In short, the patient presents mild proximal appendicular weakness, absence of ophthalmoparesis, facial and bulbar weakness detected on neurological examination, despite reports of diplopia and dysphagia. All of these elements make the diagnosis of fiber disproportion myopathy less likely than the diagnosis of nemaline myopathy. However, due to the lack of histopathological confirmation, the patient remains classified as having congenital myopathy due to a mutation in the ACTA1 gene.

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Contrast-induced encephalopathy: a mimicker in the ICU

Gabriel Pinheiro Martins de Almeida e Souza[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Lara Santos Machado[1], Daniel Delgado Seneor[1], Marcio Pinheiro Lima[1], João Victor Cabral Correia Ferrer[1], Victor Cardoso de Faria[1], Ruan Gambardella Rosalina de Azevedo[1], Andre Felipe Candêas Amorim[1], João Brainer Clares de Andrade[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Gabriel Pinheiro Martins de Almeida e Souza; Email: gabrielpinheiromas@gmail.com.

Case presentation: A 57-year-old woman presented with speech disturbance and right-sided weakness during an elective arteriography for a cerebral aneurysm located at the M1-M2 bifurcation of the right middle cerebral artery. She did not report experiencing headaches, nausea, or vomiting. The patient presented lethargy, mild confusion, motor aphasia, gaze deviation to the left, and right hemiparesis. Computed tomography revealed spontaneous hyperdensity in the subarachnoid space of the left hemisphere and mild cerebral edema. She received intravenous hydration and was admitted to the intensive care unit for monitoring. After several hours, she experienced a seizure and was treated with intravenous phenytoin. The patient fully recovered after 48 hours, with resolution of symptoms and imaging findings. **Discussion:** Contrast-induced encephalopathy (CIE) is a rare neurological complication associated with the administration of contrast agents. The incidence of CIE ranges between 0.3% and 2%, depending on the patient characteristics (older age, hypertension diabetes mellitus, impaired renal function, cardiovascular disease) and type of contrast agent (larger volume, hyperosmolarity, higher concentration, and ionic contrast agents). The pathophysiological mechanism is not fully understood but it is thought to involve a direct neurotoxic effect of the contrast agent, leaked in the brain parenchyma and subarachnoid space due to blood-brain barrier disruption and endothelial dysfunction. Clinical manifestations usually develop shortly after administration of the contrast agent and are typically transient. Symptoms include altered mental status, agitation, seizures, headache, visual disturbances, nausea and vomiting. The diagnosis is suspected in cases of clinical deterioration during or shortly after procedures using contrast agents that cannot be explained by the original ischemic area, reperfusion injury, recurrent stroke, or hemorrhagic transformation. Neuroimaging findings can support the diagnosis, such as cerebral edema beyond the infarct core and contrast staining in the brain parenchyma or subarachnoid space. The treatment of this condition is uncertain due to the small number of cases described. Management is symptomatic and individualized. Although there is no consensus, the systematic review by Mariajoseph et al. (2024) showed that intravenous hydration and glucocorticoids were the most used therapies in practice. The clinical course of CIE is usually favorable with full recovery in most patients within 48-72 hour. **Final comments:** Post-contrast encephalopathy is a differential diagnosis to be considered in cases of clinical worsening during or immediately after procedures that used contrast. Recognition of this condition avoids unnecessary additional contrast studies such as CT angiography or perfusion and inadequate treatment.

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Contribution of nerve biopsy in leprosy and other neuropathies: 26 years of experience at a reference center

Ana Caroline Siquara de Sousa[1], Mariana Hacker[2], Robson Teixeira Vital[3], Izabela Jardim Rodrigues Pitta[4], Clarissa Spitz[3], Leila Maria Cardão Chimelli[5], Sergio Luiz Gomes Antunes[2], Euzenir Nunes Sarno[2], Roberta Olmo[2], Marcia Rodrigues Jardim[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Fundação Oswaldo Cruz, Rio de Janeiro RJ, Brazil.
[3]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[4]Universidade Federal Fluminense, Niterói RJ, Brazil.
[5]Instituto Estadual do Cérebro, Rio de Janeiro RJ, Brazil.

Correspondence: Ana Caroline Siquara de Sousa; Email: anasiquara@gmail.com.

Background: Brazil has the second-highest number of leprosy cases in the world, with around 25,000 cases reported in 2022 alone. There are specialized centers in the country for the diagnosis, treatment, and monitoring of leprosy. **Objective:** To describe the experience of nerve biopsies at an outpatient clinic specializing in leprosy. **Methods:** We performed a retrospective analysis of the nerve biopsies database spanning 26 years, from 1997 to 2023. Each tissue sample was preserved in glutaraldehyde, 10% formalin, and frozen in liquid nitrogen. For those embedded in paraffin, the sections were stained with hematoxylin and eosin, Wade, and Gomori's trichrome. Semi-thin sections were stained with Toluidine Blue. A total of 31 histopathological parameters were studied. **Results:** The ratio of male to female biopsies was 1.7 to 1. The patients' ages ranged from 11 to 89 years, with an equal mean and median age of 51 years. The most commonly biopsied age group was 50-59 years. Out of 850 nerve biopsy records, 31 biopsies did not contain peripheral nerve material (3.7%). Of the remaining 819 cases, 332 (40.5%) were diagnosed as leprosy. For more than 50 patients with a suspected pure neural form of leprosy (without dermatological lesions), the disease was confirmed. Other diagnoses included vasculitis (5.4%), diabetic neuropathy (3.7%), compression neuropathy (2.4%), amyloidosis (2%), CIDP (0.7%), alcoholic neuropathy (0.4%), MADSAM (0.2%), among others. About 277 cases (33.8%) had no definitive diagnosis, only histopathological description. The diagnosis rate using nerve

biopsy, other complementary tests, and clinical sessions was 66.2%. The most biopsied nerves were the cutaneous branch of the ulnar nerve, the sural nerve, and the superficial peroneal nerve. At the initial evaluation, 68.4% of patients were already symptomatic, and predominant symptoms included sensitivity disturbance (85.7%), paresthesia (70.3%), and decreased muscle strength (59.4%). Pain was reported by 30.5% of the patients. The most frequent findings in ENMG were multiple mononeuropathy (in 84,2% of sensory nerves and 42,7% of motor nerves); motor nerves with mononeuropathy (28,1%); and normal exam (22% of motor nerves and 6,6% of sensitive nerves). **Conclusion:** Nerve biopsy was found to be useful for detecting leprosy, especially in the pure neural form, and it remains a valuable tool for reaching a diagnosis, ruling out differential diagnoses, and guiding treatment in uncertain cases. The clinical-pathological correlation was essential for the final diagnosis of the patients. To optimize its utility, we recommend considering the exam in cases with extensive clinical investigation, asymmetric clinical presentation, and progressive deterioration. Our main indication for performing a nerve biopsy was in a patient with multiple mononeuropathy, as the clinical condition was not sufficient to make the diagnosis of pure neural leprosy.

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Co-occurrence of multiple sclerosis and autism spectrum disorder: a case series of 9 patients

Eduardo Souza Cardoso[1,2,3], Jéssica de Andrade Ribeiro Lima[2], Giovanna Santana Silva Borges[2], Pedro Lucas Bomfim Figueirêdo Farias[2], Maiêva Pereira Ribeiro[2], Gabriel Eduardo Pereira Cardoso[4], Grace Patricia Santana Pereira Cardoso[5]

[1]Universidade Estadual de Feira de Santana, Feira de Santana BA, Brazil.
[2]Universidade do Estado da Bahia, Salvador BA, Brazil.
[3]Clinica IBIS, Salvador BA, Brazil.
[4]Faculdade Zarns, Salvador BA, Brazil.
[5]Hospital Portugues, Salvador BA, Brazil.

Correspondence: Gabriel Eduardo Pereira Cardoso; Email: gabrielwer204@gmail.com.

Background: Multiple sclerosis (MS) and autism spectrum disorder (ASD) are distinct neurological conditions that rarely coexist. This study presents a series of 9 patients initially diagnosed with MS, in whom level 1 ASD was observed during follow-up. **Objective:** To document the co-occurrence of MS and level 1 ASD in a series of cases, describe the clinical features observed in these patients, and discuss the diagnostic challenges and management strategies for this combination of conditions. **Methods:** A retrospective review was conducted on the medical records of 9 patients diagnosed with clinically defined MS. During neurological follow-up, characteristics of level 1 ASD were identified in these patients. The diagnosis of ASD was based on detailed clinical assessments using DSM-5 criteria and standardized screening tools. **Results:** The study included 9 patients, consisting of 7 females and 2 males, with a mean age of 34 years. All patients initially presented with typical MS symptoms, including demyelinating lesions visible on MRI, recurrent neurological relapses, and various neurological deficits. During follow-up, signs suggestive of ASD, such as difficulties in social communication, restrictive and repetitive behaviors, and specific interests, were observed. Subsequent evaluations confirmed the diagnosis of level 1 ASD in each case. The patients received multidisciplinary management, which included neurological treatment for MS and specialized support for ASD. The co-occurrence of MS and ASD presents significant diagnostic challenges as symptoms can overlap and affect clinical presentation. Early detection of ASD in patients with MS is crucial for implementing appropriate and personalized interventions. This study highlights the need for a collaborative approach between neurologists and ASD specialists to effectively manage these conditions. The identification of ASD in patients with MS requires vigilance and a comprehensive assessment strategy to ensure both conditions are adequately addressed. **Conclusion:** The co-occurrence of MS and level 1 ASD, although rare, requires special attention to ensure integrated and optimized management. Early identification and combined treatment can significantly improve the quality of life for these patients. The findings of this case series underline the importance of awareness and interdisciplinary collaboration in managing patients with complex neurological conditions.

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Co-occurrence of nmo spectrum disorder (nmosd) anti- AQP-4positive and a history of low-grade glioma in childhood: a case report

Letícia da Silva Alves[1], Arthur Costa Nascimento[1], Rogério de Rizo Morales[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Letícia da Silva Alves; Email: draleticiasilvaalves@gmail.com.

We present the case of a 17-year-old patient, male, who had a history of low-grade glioma in childhood at the medullary level of C3-T1, which was partially treated with surgery in 2012, with the interruption of adjuvant therapy at the family's option in February 2013. The patient developed paresis of the lower limbs and plegia of the upper limbs when, in 2016, he was hospitalized with pneumonia, experienced cardiorespiratory arrest after hypoxemic acute respiratory failure, and was discharged from the hospital with a tracheostomy and invasive mechanical ventilation. In August 2022, he had the first episode of bilateral optic neuritis with a good response to pulse therapy with corticosteroids. An etiological investigation then confirmed the diagnostic criteria of NMOSD with the presence of anti-aquaporin-4 (AQP-4) antibodies, and treatment with azathioprine was initiated. In October 2024, he presented with further worsening of visual acuity and underwent new pulse therapy with methylprednisolone in December 2022, but experienced a headache followed by a seizure during administration on the fourth day of metilprednisolone. The patient received high-dose corticosteroids to manage acute NMOSD exacerbations and plasmapheresis (PLEX) was scheduled, but suspended after 1 session due to clinical complications and patient choice. Treatment was escalated to biannual rituximab, with good tolerance and no new evidence of disease activity. The patient did not show significant improvement in visual function, but did not experience recurrence of disease attacks. During imaging monitoring, there were no signs of tumor recurrence. Given the concern for immunosuppression, close monitoring of the neoplasm recurrence was maintained. In a literature review, there are few case reports involving NMOSD with the presence of anti-aquaporin-4 (AQP-4) antibodies associated with neoplasms. The coexistence of NMOSD and a history of low-grade glioma in a pediatric patient is rare, and the exact relationship between these two conditions remains unclear; potential mechanisms may involve shared immune dysregulation pathways. Further research is warranted to explore the potential interplay between autoimmune disorders and prior malignancies.

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Correlation between Guillain-Barré syndrome and COVID-19 infection or vaccination in patients undergoing therapeutic plasmapheresis

Eduarda Leitzke[1], Maria Eduarda Broco[1], Ana Luiza Fontana[1], Manuela Spagnol[1], Mariana Gatti Altafini[1], Jaqueline Solda Palaoro[2], Bruna Accorsi Machado[2], Aline Ferreira de Miranda[2], Gisele de Oliveira Doring[2], Cristiane da Silva Rodrigues de Araujo[1]

[1]Universidade de Passo Fundo, Passo Fundo RS, Brazil.
[2]Hospital São Vicente de Paulo, Passo Fundo RS, Brazil.

Correspondence: Eduarda Leitzke; Email: leitzkeeduarda@gmail.com.

Background: Guillain-Barré syndrome (GBS) is a clinical condition characterized by immune system disruption and subsequent dysregulation. It can be triggered by antigen exposure through infections or even vaccinations. **Objective:** To analyze the possible correlation between the development of GBS and exposure to the COVID-19 virus through infection or vaccination in patients undergoing therapeutic plasmapheresis at the hemotherapy service of a tertiary hospital. **Methods:** Cross-sectional study with a retrospective approach of patients reported with GBS from March 2020 to March 2022, who underwent the therapeutic plasmapheresis procedure. **Results:** A total of eleven patients were diagnosed with GBS. The average age was 49 years, with six males and five females. Among these, only one patient developed COVID-19 infection, while ten patients did not contract the infection. Additionally, out of the eleven patients, only eight had been vaccinated against COVID-19. **Conclusion:** The relationship between the development of GBS and infection or vaccine against COVID-19 is rare. However, an association is possible, as contact with the virus can trigger an exaggerated and unregulated immune response, contributing to the development of GBS.

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Correlation between median motor latency and complementary sensory tests on the 1st and 4th fingers for early diagnosis of carpal tunnel syndrome

Juliana Calvet Kallenbach Aurencao[1], Mariana Quintanilha Deslandes[2], Claudia Cristina Ferreira Vasconcelos[3]

[1]Hospital Federal dos Servidores do Estado Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Neurologia e Neurofisiologia do Rio de Janeiro.
[3]Universidade Federal do Estado do Rio de Janeiro, Hospital Gaffree e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: Juliana Calvet Kallenbach Aurencao; Email: julianacka@hotmail.com.

Background: Carpal tunnel syndrome (CTS) was first described in 1854 and is characterized by pain, paresthesia and tingling in the hands, and in advanced cases, weakness with atrophy of the thenar region, being a frequent cause of disability and absence from work activities. Electroneuromyography is an accurate tool for diagnosis, making it possible to identify cases in the early stages. **Objective:** To detect patients in the early stages of the disease, through the correlation between the distal motor latency of the median nerve and complementary sensory tests of latency difference of the median nerve in the 1st and 4th fingers. **Methods:** Data from 159 exams were collected, with different indications. Motor neuroconduction of the median and ulnar nerves, and sensory neuroconduction of median, radial and ulnar nerves were performed. Exams with diagnosis of polyneuropathy were excluded. The SPSS program was used for statistical analysis. ROC curves and accuracy tests for diagnostic were performed. **Results:** The shortest motor latency of the median nerve found, with an accuracy of 81%, in the analysis of both the 4th and 1st fingers, was 3.3ms. On the 2nd finger, with an accuracy of 74%, it was

3.5ms. There is a large number of diseases that can simulate the symptomps of CTS, and several other illnesses can be associated with this disease. The electroneuromyography examination routinely begins with the motor neuroconduction of the median nerve, which is then, the first parameter of the examination to be analyzed. Then, followed by examination of the median sensory nerve in the 2nd finger. Complementary sensory tests are usually performed only in patients with high suspicion or indication for the diagnosis of CTS. The results above demonstrate that, when performing stimulation of the median motor nerve, latencies of 3.3ms demonstrated high specificity and sensitivity for detecting positivity in the latency difference tests between median and radial and ulnar nerves in the 1st and 4th fingers respectively. In the 2nd finger, the shortest motor latency for diagnosis was 3.5ms, being then affected later than in the other two tests above. **Conclusion:** When identifying a motor latency in the Median nerve of 3.3ms, complementary comparative tests must be performed, with the aim of identify cases of mild CTS, thus enabling early diagnosis and, consequently, early intervention.

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Correlation between meningitis vaccination and mortality rates in Brazil: a retrospective analysis

Antonia Cardoso Silva[1], Júlio César Soares Barros[1], Larissa da Silva Almeida[1], Guilherme Carvalho de Souza[1], Raissa Nogueira de Souza[2], Mariana Reis Prado[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.
[2]Centro Universitário UniFTC, Salvador BA, Brazil.

Correspondence: Antonia Cardoso Silva; Email: antonia.silva@famed.ufal.br.

Background: Meningitis is an inflammation of the meninges, the membranes that surround the brain and spinal cord. The disease can be caused by viruses, bacteria, fungi, or parasites, posing a significant public health challenge due to high mortality rates and neurological complications. Vaccination, available in both public and private healthcare systems, is the most efficient way to prevent meningitis. However, the incidence of meningitis cases remains high. **Objective:** To evaluate the correlation of meningitis mortality with vaccination by region against serotypes A, C, W, Y, and 135 available in the public health system in Brazil from 2020 to 2022. **Methods:** The analysis was conducted through a cross-sectional, retrospective, and descriptive epidemiological study using data from the Mortality Information System (SIM/DATASUS) from 2020 to 2022. The assessment of meningitis mortality was performed by analyzing the variables "Region," "Outcome," "Immunobiologicals," and "Serogroup." **Results:** Between 2007 and 2009, the years prior to the implementation of meningitis vaccines in the vaccination schedule of the Unified Health System (SUS), there were 7,541 meningitis-related deaths in the country. In contrast, from 2020 to 2022, the same period after the implementation of the Meningococcal Conjugate - C and Meningococcal ACWY135 vaccines, the number of deaths was 2,861, a 62.06% overall decrease: 50% in the North, 64.97% in the Northeast, 63.94% in the Southeast, 55.26% in the South, and 66.60% in the Center-West, demonstrating the effectiveness of vaccination. The Center-West region had the highest proportional reduction, followed by the Northeast and Southeast, with variations possibly influenced by factors such as vaccination coverage, access to healthcare services, and socioeconomic conditions. Despite the undeniable effectiveness of vaccination, a notable finding was the linear increase in the number of deaths from 2020 to 2022, with 723 deaths in 2020, 785 in 2021, and 1,308 in 2022. This trend was associated with significant declines in vaccination rates (9,748,789 in 2020, 8,124,523 in 2021, and 8,681,157 in 2022), highlighting the need to strengthen vaccination campaigns. Additionally, there remains a considerable number of deaths associated with serogroups for which no vaccination is available in SUS, particularly serogroup B, responsible for 35 deaths from 2020 to 2022. Although these numbers are substantially lower compared to those covered by vaccination, they still represent an area of concern. **Conclusion:** This study evaluates the effectiveness of meningitis vaccination campaigns in reducing mortality, highlighting the significant reduction in deaths. It reinforces the importance of maintaining and expanding vaccination programs to cover additional serogroups in the vaccination schedule and strengthening efforts in surveillance, education and public health infrastructure in the fight against meningitis.

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Correlation between microvascular changes in the retina and choroid with disease duration, severity, and cognitive deficits in migraine with and without aura

Luiz Felipe Mota Freitas[1], Leonardo Provetti Cunha[2], Mauro Eduardo Jurno[3], Márcio Alves da Cruz Júnior[1], Thiago Cardoso Vale[1], Lígia Barros de Oliveira[3]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Juiz de Fora MG, Brazil.
[2]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[3]Fundação Hospitalar do Estado de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Luiz Felipe Mota Freitas; Email: luizfelipemotafreitas@gmail.com.

Background: The thickness of the retina and choroid, as measured by optical coherence tomography (OCT), may reflect neurovascular changes in migraine patients. **Objective:** To evaluate the relationships between the changes identified by OCT and optical coherence tomography angiography (OCTA) in patients with migraine with aura (MA) or migraine without aura (MWA) and the duration and severity of the disease using the Headache Impact Test-6 (HIT-6) questionnaire and cognitive assessment through the Montreal Cognitive Assessment (MoCA). **Methods:** In this cross-sectional study, 16 patients with MA and 15 patients with MWA were compared with 16 healthy age-matched controls. **Results:** Foveal choroidal thickness was lower in individuals with migraine than in individuals in the control group ($p<0.05$). Statistically significant negative correlations were found between disease duration and total macular thickness ($p=0.037$; compared with the average thickness of the MWA group), ganglion cell complex (GCC) thickness ($p=0.017$; compared with the average thickness of the MWA group) and choroidal thickness ($p=0.039$; compared with the average thickness of the MWA group), as well as the HIT-6 score and the peripapillary retinal nerve fiber layer (pRNFL) thickness ($p=0.027$; compared with the mean thickness of the MWA group). **Conclusion:** Compared with controls, individuals with migraine showed a significant reduction in choroidal thickness. Our results showed that the longer the disease duration was, the thinner the total macula, GCC and choroidal thickness were. Additionally, the thickness of the pRNFL layer showed an inverse correlation with the disability caused by migraine.

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Correlation between motor development and cognitive development in children with cerebral palsy

Môyra Aloia Romero[1], Jordan Paes de Almeida[1], Julia Mirella de Souza Monteiro Silva[1], Nicole Fernanda Bernardino[1]

[1]Centro Universitário UniMetrocamp Wyden, Campinas SP, Brazil.

Correspondence: Jordan Paes de Almeida; Email: almeidapjordan@gmail.com.

Cognitive capacity is closely linked to learning and adaptability in humans, serving as a fundamental component for the survival and evolution of the species. It provides full access to mental functions, which, in turn, enable adaptation to the environment through the monitoring and shaping of behaviors, conduct, and interactions. According to Vygotsky, cognitive development is directly related to social interactions and mental functions. Therefore, an individual does not socialize due to their cognitive development; rather, cognitive development occurs through socialization. Consequently, it is impossible to separate cognitive development from social interactions. Considering that motor development influences cognitive development through curiosity, feedback, reactions, and interactions with the environment, many authors advocate the correlation between the ability to learn and the ability to perform. Children with cerebral palsy (CP), also known as non-progressive chronic encephalopathy of childhood, are inserted in this context. Children with CP who have severe motor impairments (levels III to V on the GMFCS) are often pre-judged as intellectually disabled due to their motor disability. The lack of cognitive assessment methods for children with severe motor disabilities, especially those with significant language deficits, can lead to misinterpretations of their true cognitive abilities. This raises the question of what the actual cognitive development of children with CP might be. Knowing that learning is influenced by a stimulating environment, we must consider the cognitive stimuli present in their surroundings. Given this scenario, this research aims to propose the creation of two instruments to measure the intellectual capacity of children with CP: 1. An environmental analysis tool for school-aged children with CP to assess the daily cognitive stimulation affordances provided; 2. Assessment tools for evaluating the cognitive performance of children with CP. Children with CP can have normal or even above-normal intelligence, or they may have intellectual delays, not only due to brain lesions but also primarily because of a lack of experience and interaction with their environment. The delay in psychomotor development among disadvantaged children is largely attributed to growing up in environments lacking facilitative stimuli. In schools, there is often confusion between motor and cognitive deficits. In the inclusion process, educators may attribute learning difficulties to motor challenges as if they also indicate cognitive issues. Therefore, it is crucial to improve access to and understanding of the cognition of this group, promoting adequate interaction with the environment to positively impact the development and learning of these children.

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Correlation between motor, non-motor, and cognitive functions with optical coherence tomography (OCT) in Parkinson's disease

Víctor de Oliveira Costa[1], Daniel de Oliveira Araujo[1], Leonardo Provetti Cunha[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Juiz de Fora MG, Brazil.

Correspondence: Victor de Oliveira Costa; Email: victordeoliveiracosta82@gmail.com.

Background: Patients with Parkinson's disease (PD) exhibit a wide range of visual disturbances throughout the course of the disease. Many of these visual deficits in PD patients occur due to mechanisms involving changes in retinal cells and layers. Optical coherence tomography (OCT) can non-invasively study different retinal layers and their thicknesses, showing a correlation with PD. **Objective:** To assess the total macular thickness and the

inner and outer retinal layers' thickness in patients with PD and to verify the correlation between these parameters with motor symptoms and cognitive function. **Methods:** Forty-six eyes of 23 PD patients and 40 eyes of 20 healthy controls (HC) were included. Cognitive, functional, and non-motor symptoms were assessed on all participants using the Katz Index of Independence and Pfeffer's of Activities for Daily Living (ADLs), Mini-Mental State Exam (MMSE), the Frontal Assessment Battery (FAB), Schwab & England (S&E) staging scales, and the Movement Disorders Society Nonmotor Symptoms Scale (MDS-NMS). Total, inner and outer OCT macular thickness measurements were recorded. We assessed the correlation between OCT parameters and cognitive, functional, and non-motor symptoms. **Results:** The values of the Katz, Pfeffer's ADL, and MMSE tests were significantly lower in PD than in HC. Total macular measurement was thicker in PD patients. The temporal and inferior outer sectors were significantly greater for the GCC thickness in patients. There was a significant correlation between total macular thickness and MDS-UPDRS-III values. Conversely, there was a negative correlation between outer macular thickness parameters and MDS-UPDRS-III values. There was a significant correlation between the total macular thickness and GCC thickness with MMSE, S&E, FAB, Katz's, and Pfeffer's ADL scores. In addition, the S&E scale correlates with outer macular thickness measurements. **Conclusion:** The total and inner macular thickness measurements at temporal and inferior outer sectors were higher in PD patients when compared to the control group. These findings suggest that macular measurements may be thicker in PD, especially when associated with mild motor symptoms. Additionally, we demonstrated a significant correlation between many total, inner, and outer OCT parameters with motor and non-motor symptoms and cognitive function impairment.

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Correlation between postural control and sleep quality in patients with parkinson's disease: a cross-sectional study

Samuel Lidione da Silva[1], Alessandra Cattaneo Estrada Melanda[2], Rogério José de Souza[2], Amanda Lima Nogueira dos Anjos[2], Hayslenne Andressa Gonçalves de Oliveira Araújo[2], Milena Marie Matias Martins[1], Heloísa Freiria Tsukamoto[1], Suhaila Mahmoud Smaili[2]

[1]Centro Universitário Filadélfia, Londrina PR, Brazil. [2]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Samuel Lidione da Silva; Email: lidione67@gmail.com.

Background: Postural instability is an integral part of the characteristic motor tetrad of Parkinson's disease (PD) and significantly compromises the functional capacity, independence, and autonomy in this population. Among the various non-motor symptoms, sleep disturbances are very common, difficult to manage, and can also negatively impact physical, cognitive, and social domains. However, little is known about the relationship between postural control and sleep quality in individuals with PD. **Objective:** To correlate postural control and sleep quality in individuals with PD. **Methods:** This is a cross-sectional study, comprising of 42 individuals of both sexes, with mild to moderate disease staging, as assessed by using the modified Hoehn & Yahr scale. For the assessment of signs and symptoms, the Movement Disorder Society Unified Parkinson's Disease Rating Scale (MDS-UPDRS) was applied, and the cognitive status was assessed using the Mini-Mental State Examination. Postural control assessment was performed using the Mini Balance Evaluations Systems Test (MiniBESTest), and sleep quality as well as daytime sleepiness were evaluated using the following instruments: the Parkinson's Disease Sleep Scale (PDSS-2), the Pittsburgh Sleep Quality Index (PSQI), and the Epworth Sleepiness Scale (ESS). Pearson and Spearman correlation analyses were performed, according to data normality, considering a significance level of 5%. This study was approved by the Research Ethics Committee (CAAE: 45459521.6.1001.5188). **Results:** Participants had a mean age of 64.10±9.75 years, H&Y stage of 2.0 [2.0-2.5], and MDS-UPDRS II and III of 41.81±15.39, indicating mild to moderate severity. The final scores of postural control and sleep quality variables were: MiniBESTest = 22.62±2.47 - indicating good postural control (the maximum score is 28 points), PDSS-2 = 15.5 [7.8-22.5] - indicating good sleep quality (the maximum score is 60, and higher scores indicate poorer sleep quality), PSQI = 7.24±3.74 - indicating poor sleep quality (score >5 points), and ESS = 10.67±4.61, indicating excessive daytime sleepiness (score ≥10 points). Additionally, no statistically significant correlations were found between postural control and sleep quality. **Conclusion:** The study did not find any correlation between postural control and sleep quality. Although the PSQI suggested poor sleep quality, the PDSS-2, designed specifically for assessing sleep in PD individuals, did not indicate significant sleep impairment among the participants in this study. Similarly, no significant impairment in postural control was detected. Thus, an investigation into individuals in the mild to moderate phase of PD did not uncover any links between these outcomes. Further research is warranted in individuals in more advanced PD stages and utilizing objective sleep quality and postural control measures, to deepen our understanding of the relationship between sleep and postural control in individuals with PD.

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Correlation between severity of ischemic stroke and levels of disability and functional dependence: a cohort study

Jéssica dos Santos Coelho[1], Maria Vitória Do Nascimento Silva[1], Maria Laura Lima da Silva[1], Joan Lins Serafim[1], Thiago Bezerra dos Anjos[1], Ana Luiza Sampaio Lopes[1], Américo Danúzio Pereira de Oliveira[1], Rita Di Cassia Oliveira Angelo[2]

[1]Universidade de Pernambuco, Serra Talhada PE, Brazil. [2]Universidade de Pernambuco, Garanhuns PE, Brazil.

Correspondence: Maria Vitória do Nascimento Silva; Email: vitoria.nsilva@upe.br.

Background: Stroke is the second leading cause of death globally and the third leading cause of loss of disability-adjusted life years. In Brazil, it is estimated that more than 2 million people are affected by stroke, of which nearly a quarter will be left with severe disability. The degree of functional disability may be associated with several factors, including the severity of the cerebrovascular event in the first 24 hours post-stroke. Therefore, prompt recognition of the magnitude of neurological damage and rapid therapeutic interventions can help modify the functional course of the disease. **Objective:** To analyze the relationship between the level of severity of ischemic stroke at hospital admission up to 24 hours post-stroke, and the course of functional capacity in three months, in patients at an emergency service in the city of Serra Talhada-PE . **Methods:** Cohort study, carried out between 2021 and 2023, which included patients diagnosed with stroke admitted to a public hospital in Serra Talhada-PE. Assessments took place at three stages: admission, discharge and after three months. The instruments used were the SUS individual registration form and the assessment scales NIHSS, TOAST, Modified Rankin Scale (ERm) and the Modified Barthel Index (IBM). **Results:** The sample consisted of 157 patients, predominantly male (n=89; 56.7%), aged between 61-80 years (n=86; 54.8%) and hypertensive (n=130; 82 .8%). Of these, 10.2% had moderate to severe disability (ERm≥3) and 11% had severe neurological deficit (NIHSS>22) on admission. The comparative analysis of the levels of disability and functional dependence between the moments of discharge and after three months, showed significant maintenance (p<0.001) of the moderate to severe level of disability and the moderate level of functional dependence, with a median IBM equal to 28 [10-45] in both evaluations. There were strong and significant correlations (p<0.001) between the level of stroke severity at admission and the degrees of functional dependence at discharge (p=0.77) and after three months of follow-up (p=0.61). It should be noted that none of the patients received reperfusion therapy or thrombectomy. **Conclusion:** There is a bidirectional and proportional relationship between the level of stroke severity at admission and the degree of functional dependence and post-stroke disability at three months of follow-up. Patients undergoing reperfusion therapy with thrombolytics or mechanical thrombectomy have a greater chance of a favorable outcome, both in the NIHSS at discharge and in the Rankin scale score after three months. The establishment of a specialized stroke unit can be a useful tool in the effective and efficient in-hospital treatment of stroke, reducing the morbidity and mortality of this event. Such results demonstrate the importance of implementing strategies to reduce the impact of this event on the lives of affected patients.

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Correlation between sleep and kinematic gait parameters in people with parkinson's disease

Nathalia de Oliveira Franco[1], Alessandra Cattaneo Estrada Melanda[1], Andressa Leticia Miri[1], Amanda Lima Nogueira dos Anjos[1], Amanda dos Santos Siqueira[1], Denise Bolonhezi Ribeiro[1], Suhaila Mahmoud Smaili[1], Rogério José de Souza[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Nathalia De Oliveira Franco; Email: nnatfrancos@outlook.com.

Background: Sleep disorders are highly prevalent in individuals who have Parkinson's disease and negatively impact their quality of life in addition to various other symptoms such as gait freezing, cognitive disorders, mood disorders, and fatigue. However, little is known about the relationship between sleep quality and objective gait parameters. **Objective:** To correlate sleep disorders with the gait of individuals with Parkinson's disease. **Methods:** This is a cross-sectional study that included 24 individuals with a confirmed diagnosis of idiopathic Parkinson's disease according to the London Brain Bank criteria, in mild to moderate staging, and without cognitive deficits. The kinematic gait evaluation was conducted using the Optitrack system under simple gait and cognitive dual-task conditions. The gait parameters analyzed were step length, step width, cadence, maximum ankle angle, vertical center of mass displacement, and gait speed. Sleep quality was assessed using the Parkinson 's Disease Sleep Scale-2 (PDSS-2), Pittsburgh Sleep Quality Index (PSQI), and the Epworth Sleepiness Scale (ESE). The study was approved by the ethics and research committee (CAAE: 72474923.3.0000.5231). The significance level adopted was 5%. **Results:** Regarding simple gait, moderate correlations were found between the following variables: step width with the disturbed sleep domain of the PDSS-2 (r=-0.42; p=0.04); vertical center of mass displacement with the total PDSS-2 score (r=-0.51; p=0.01); and gait speed with the sleep duration domain of the PSQI (r=-0.57; p<0.01). For dual-task gait, moderate correlations were observed between the following variables: maximum ankle angle and the total PDSS-2 score (r=-0.45; p=0.02); vertical center of mass displacement with the total PDSS-2 score (r=-0.46; p=0.02); step width with the daytime dysfunction domain of the PSQI (r=-0.43; p=0.03); gait speed and the sleep duration domain of the PSQI (r=-0.55; p<0.01); and gait speed and the total PSQI score (r=-0.42; p=0.03). No correlation was found between daytime sleepiness, as

assessed by the ESE, and gait parameters. **Conclusion:** Individuals with Parkinson's disease who have poorer sleep quality exhibited worse parameters in both simple and dual-task gait, such as reduced step width, speed, and vertical center of mass displacement. Thus, further studies are needed to investigate the relationship between gait and sleep, especially longitudinal cohort studies. The importance of adequate investigation of sleep disorders in this population for effective treatment is emphasized.

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Correlation between vitamin d deficiency and post-stroke depression: an updated meta-analysis

Pietra Amorim Cerquinho Oliveira[1], Gustavo Carreira Xavier[2], Lucas Drummond Savassi Stehling[2], Luisa Rezende Batista[2], Artur Menegaz de Almeida[3], Ítalo Barros Andrade[4], Michele Kreuz[5]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal de Viçosa, Viçosa MG, Brazil.
[3]Universidade Federal do Mato Grosso, Cuiabá MT, Brazil.
[4]Faculdade Santo Agostinho de Vitória da Conquista, Vitória da Conquista BA, Brazil.
[5]Universidade Luterana do Brasil.

Correspondence: Lucas Drummond Savassi Stehling; Email: lucasstehling0541@gmail.com.

Background: Depression is the most common psychiatric repercussion in patients after a stroke. **Objective:** To analyze the relationship between serum vitamin D levels and post-stroke depression (PSD), which is not yet fully understood. **Methods:** MEDLINE, Scopus and Web of Science databases were systematically searched until august 02, 2023. Data were examined using the Mantel-Haenszel method and 95% CIs. Heterogeneity was assessed using I2 statistics. Meta regression analysis was conducted to evaluate a possible link between vitamin D deficiency and post-stroke depression. R, version 4.3.2, was used for the statistical analysis. **Results:** Nine studies and 637 patients were included in this meta-analysis. In the Hamilton Rating Scale for Depression (HAMD-17) vitamin D deficiency was statistically correlated with post-stroke depression (MD = -7.62 [-13.02; -2.23]; p<0.01; I2 = 83%). Conversely, in the general analysis with Beck's Depression Inventory (BDI-II), Montgomery Asberg Depression Rating Scale (MADRS), Patient Health Questionnaire (PHQ-9) and HAMD-17 there was no significant correlation between vitamin D deficiency and post-stroke depression (MD = -3.12 [-6.74; 0.49]; p=0.090421; I2 = 89%). **Conclusion:** In this updated meta-analysis, no significant correlation between vitamin D deficiency and post-stroke depression was found. Nevertheless, the sub-group HAMD-17 analysis showed a correlation between these two groups. Consequently, further studies are necessary to solve this controversy regarding different depression scales in order to identify the real role played by vitamin D in post-stroke depression.

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Cortical correlations of CSF neurogranin, VILIP-1, alpha-synuclein, and NFL in SCD and MCI according to amyloid status

Emanuel Rodrigues Guimarães[1], Brunno Machado de Campos[1], Isadora Cristina Ribeiro[1], Ítalo Karmann Aventurato[1], Marjorie Cristina Rocha da Silva[1], Liara Rizzi[2], Marcio Luiz Figueredo Balthazar[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]UCFS University of California, San Francisco, United States of America.

Correspondence: Emanuel Rodrigues Guimarães; Email: emanuelrg.inst@gmail.com.

Background: Alzheimer's disease (AD) pathophysiology is complex and not completely known. Emerging new biomarkers that evaluate synaptic function (VILIP-1, neurogranin), co-pathology (alpha-synuclein), and neurodegeneration (NFL) are potential candidates to be incorporated into the early AD diagnosis. **Objective:** To better understand the relevance of these biomarkers, we evaluated the correlations between their CSF concentrations with whole-brain grey matter volumes in SCD and MCI, according to their amyloid status (A- or A+). **Methods:** 75 participants diagnosed with SCD or MCI were included, 30 A- and 45 A+. They all underwent comprehensive neuropsychological assessment, CSF analyses (Roche Elecsys), and volumetric 3T MRI (Philips Achieva). Voxel-based morphometry was used to quantify brain volumes through CAT12 running on MATLAB 2019b. Partial correlation analysis between CSF biomarkers and cortical volumes was performed with SPSS 22, adjusting for age and sex. **Results:** Significant correlations were found in the A- but not in the A+ group. NFL and hippocampus (r=0.40, p=0.028); VILIP-1 and superior temporal gyrus (r=-0.38, p=0.035), and superior frontal gyrus (r=-0.40, p=0.025); alpha-synuclein and hippocampus (r=-0.39, p=0.036), middle frontal gyrus (r=-0.375, p=0.049), and superior temporal gyrus (r=-0.478, p=0.01). **Conclusion:** We found different patterns of correlations between brain anatomy and emerging biomarkers, according to amyloid status, in the very early stage of the neurodegenerative process. Interestingly, these correlations were found only in the amyloid-negative group, which might suggest that different pathological processes involving synaptic function, neurodegeneration, and co-pathology occur in suspected non-Alzheimer pathophysiology cases.

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Cortical laminar necrosis and its characteristics in a patient with systemic hypoperfusion: a case report

Diogo Rabelo de Lima[1], Agatha Lopes Azedo[1], Giovanna Bruna Real Antonio[1], Eduarda Kei-lan Tavares Pierre[1], Fernando Luiz Westphal Filho[2], Matheus Cardeal Clos[1], Victor Hugo Soares dos Santos[1], Caio Felipe Tourinho de Souza Barbosa Obando[1], Tales de Oliveira Junior[1], Vanise Campos Gomes Amaral[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Diogo Rabelo de Lima; Email: diogo.rabelodelima@gmail.com.

Case presentation: E.S.M., a 66-year-old male with a history of diabetes, hypertension, and chronic kidney disease, was admitted on May 21, 2023, presenting with advanced peripheral arterial disease and sepsis due to secondary bacterial infection in the left foot. He developed acute kidney injury and hypotension, requiring high doses of noradrenaline due to inadequate blood pressure improvement. On June 10, 2023, he was transferred to the ICU following a seizure episode. A cranial CT scan revealed bilateral cortical lesions with gyral distribution in the frontoparietal region, characteristic of cortical laminar necrosis (CLN). **Discussion:** Cortical laminar necrosis involves neuronal destruction due to decreased oxygen and glucose supply to the nervous tissue, typically occurring in conditions such as cardiac arrest, generalized hypoxia, hypoglycemia, hematologic disorders, and status epilepticus. Cortical neurons are particularly vulnerable due to their high metabolic activity compared to glial cells and adjacent white matter, with the third layer of the cortex composed of pyramidal cells being especially susceptible to metabolic stress. It is important to differentiate CLN from other conditions such as cortical hyperintensities in T1-weighted MRI or cortical calcifications following ischemic stroke, as CLN exclusively affects the cortex without involvement of subcortical white matter. On CT imaging, CLN appears days after the event as a hyperdensity following the gyri of the cerebral cortex in a linear pattern, devoid of hemorrhage or calcification. In MRI, T1 hyperintensity is characteristic due to denatured proteins and lipid-laden macrophages, distinguishing it from other pathologies. T2/FLAIR sequences typically show isointensity to hyperintensity. **Final comments:** The cerebral cortex integrates sensory inputs and initiates voluntary motor responses, underscoring its pivotal and intricate role. Therefore, understanding the pathophysiology and neuroimaging characteristics of cortical laminar necrosis is crucial. This knowledge facilitates accurate differential diagnosis in cases with similar clinical and radiological features, particularly in emergency and intensive care settings where cerebral energy depletion is common. Identification of CLN serves as a prognostic indicator for neurological outcomes, highlighting its significant implications in patient management.

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Cost analysis of amyotrophic lateral sclerosis according to clinical protocols and therapeutic guidelines compared to real-world clinical practice: application of time-driven activity-based costing methodology in a specialized service

Laís Brandão Mota[1], Marie Lidio dos Santos Galvão Ribeiro[1], Maria Rodrigues Lima de Souza Pessoa[1], Stefania Zingone Andrade Carvalho[1], Temis Maria Felix[2], Marcela Câmara Machado Costa[1], Marilaine de Matos Menezes Ferreira[1], Camila Azevedo[3], Marcelo Nita[3], Myrianne Gilsar[3]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[3]MAPES Solutions, São Paulo SP, Brazil.

Correspondence: Laís Brandão Mota; Email: bmota.lais@gmail.com.

Background: The Clinical Protocol and Therapeutic Guidelines (PCDT) is a technical guide that establishes criteria based on scientific evidence for the diagnosis, treatment, and monitoring of specific diseases and health conditions. This work was part of the Journey of Value for Patients with Rare Diseases (JAV-RARAS) project. The JAV-RARAS study presents an innovative approach to understanding the real costs of rare disease care in Brazil. **Objective:** To map the care journeys of patients with Amyotrophic Lateral Sclerosis (ALS) in a specialized healthcare facility and compare them with the PCDT recommendations established by the Ministry of Health. For this, the Time-Driven Activity-Based Costing (TDABC) methodology was used. **Methods:** JAV-RARAS employs the TDABC methodology to assess the costs associated with the management of rare diseases. Through interviews and process analyses, the actual annual costs of managing patients with ALS in the specialized service were identified. The costs defined by the PCDT were compared to the current clinical practice observed in the service. This made it possible to evaluate the adherence of real costs to the standards established in the PCDT and identify opportunities to optimize ALS management. **Results:** Preliminary results showed that the actual annual cost at the institution was $7,506.20 per patient per year, while the estimated cost according to the PCDT was $8,054.67. Furthermore, the estimated cost of treatment by the PCDT ($7,262.04) is also close to the actual cost charged by the institution ($7,238.15). Regarding the source of

the resources, it was found that 98.3% were funded by the Unified Health System (SUS), 0.80% by the patient, and 0.88% by the institution itself. These results indicate an alignment between the costs estimated in the PCDT and those observed in clinical practice. **Conclusion:** The average annual cost per ALS patient in the specialized service was close to the value reported in the study, but with differences in the composition of these costs. This is mainly due to variations in the use of drug therapies and the need to refer certain surgical procedures to other centers, which can generate additional costs not accounted for locally. The TDABC methodology demonstrates its potential to support the review of policies and protocols, aiming in the efficient allocation of resources in the care of rare diseases such as ALS.

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Costs of hospitalizations for the treatment of acute ischemic and hemorrhagic stroke in the state of São Paulo between 2019 and 2023

Ana Clara Lacerda Freitas[1], Aline Teixeira[1], Laysa de Souza Maia[2], Andressa Gonçalves Vicente[3], Ana Alice Lemos Lima[4], Gabriela Hernandez Dumani[5], Ana Cristina Carvalho e Silva[3], Estefany Moreno de Souza[6], Geovana Franklim Gomes e Silva[7], Guilherme de Andrade Ruela[4]

[1]Centro Universitário Funorte, Montes Claros MG, Brazil.
[2]Centro Universitário Aparício Carvalho, Jaru RO, Brazil.
[3]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[5]Universidade Franciscana, Santa Maria RS, Brazil.
[6]Universidad de Aquino Bolivia, Santa Cruz de La Sierra, Bolívia.
[7]Centro Universitário de Patos de Minas, Patos de Minas MG; Brazil.

Correspondence: Ana Clara Lacerda Freitas; Email: anaclf15@yahoo.com.br.

Background: Stroke, whether ischemic or hemorrhagic, is one of the leading causes of morbidity and mortality worldwide. Despite the identification of modifiable risk factors such as hypertension, smoking, and obesity, stroke continues to pose a significant challenge for healthcare systems, especially in terms of resource management and economic impact. In the state of São Paulo, the most populous region in Brazil, the high incidence of stroke necessitates a thorough understanding and analysis of hospitalizations and associated treatment costs, highlighting the importance of an organized and efficient care system for successful treatment outcomes. **Objective:** To analyze healthcare costs related to stroke in the state of São Paulo between 2019 and 2023. **Methods:** This ecological study utilized data obtained from the Hospital Information System of the Brazilian Unified Health System (SIH/SUS) through the Department of Informatics of the Brazilian Unified Health System (DATASUS). Data acquisition, organization, and tabulation were performed using Google Sheets, encompassing the number of cases, duration of hospitalizations due to stroke in São Paulo, average cost per hospitalization, total cost, and mortality rate from 2019 to 2023. **Results:** The analysis included a total of 208,190 hospitalizations, with an average hospital stay of 8 days, irrespective of ward or ICU admission, resulting in an average cost of R$ 1,769.63 per hospitalization. Consequently, direct costs alone amounted to R$ 368,419,830.80 for the state. Additionally, 35,162 patients died, with a mortality rate of 16.89%. There was a 3.2% reduction in the number of cases in 2020 (38,616 cases) compared to 2019 (39,907 cases). However, costs increased from R$ 58,015,826 in 2019 to R$ 60,886,962.95 in 2020, representing a 4.95% increase. This cost increase continued despite the minimal increase in the number of cases, reaching R$ 94,721,716.99 in 2023, with 45,651 cases, the year with the highest number of hospitalizations. **Conclusion:** The study revealed a discrepancy between the number of stroke cases and the associated treatment costs in the state of São Paulo between 2019 and 2023. Despite the reduction in the number of cases in 2020 compared to 2019, there was a consistent increase in direct costs, possibly due to the increased complexity of cases, inflation in medical supply prices, and changes in hospital management practices in response to the COVID-19 pandemic. The high mortality rate underscores the severity of stroke, emphasizing the importance of effective public policies for prevention, focusing on risk factors to reduce hospitalizations. Moreover, it is imperative that health policies be adjusted to optimize the cost-effectiveness of stroke treatment, ensuring the sustainability of the healthcare system and improving patients' quality of life.

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Could coinfection accelerate htlv-1-associated myelopathy/tropical spastic paraparesis (HAM/TSP) onset?

Ana Flávia Novaes[1], Davi Tanajura Costa[1], Fernanda Khouri Barreto[1], Qesya Rodrigues Ferreira[1], Jéssica Oliveira de Souza Nascimento[1], Carolina Souza Santana[1], Ariana Leal Borges da Cruz[1], Rebeca Leão Amorim[1], Alice Palmeira Nascimento Cerqueira[1], Felipe de Oliveira Andrade[1]

[1]Universidade Federal da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Ana Flávia Novaes; Email: anaflavia635@gmail.com.

The human T-cell lymphotropic virus type 1 (HTLV-1) is a retrovirus that infects human cells and causes severe diseases. It is estimated that more than 5 to 10 million people worldwide are infected with this virus. This viral infection can affect the central nervous system, leading to HTLV-1-associated myelopathy/tropical spastic paraparesis (HAM/TSP). Additionally, as a sexually transmitted infection, there is a prevalence of coinfections with other viruses. However, the relationship between coinfections and neurological clinical outcomes has not yet been well studied. This quantitative cross-sectional study was carried out with people living with HTLV-1 and with another infection (HIV, HBV, HCV, or syphilis), monitored in a Sexually Transmitted Infections Reference Center in Bahia, Brazil, from March 2023 to May 2024. Data were obtained by analyzing medical records and during neurological examination. The statistical significance analyses were performed using Fisher's exact test with SPSS software v. 27. A total of 110 patients were evaluated, with 82 (74.5%) classified as non-HAM/TSP and 28 (25.4%) as HAM/TSP. The mean age of the non-HAM/TSP patients was 48.66±16.78 years, with 65 (80%) female. The mean age of the HAM/TSP patients was 56.46±19.75 years, with 21 (75%) being female. From this sample, 32 (29%) patients were coinfected. Half of the HAM/TSP patients (14, or 50%) exhibited coinfection, with a statistically significant difference between groups ($p<0.0001$). Syphilis was the most prevalent co-infection in the HAM/TSP group (25%), followed by HBV (18%), tuberculosis (14%), HIV (10%). HCV and HTLV-2 (3%). with a statistically significant difference noted between groups ($p<0.01$) in those co-infected with syphilis, HBV, and tuberculosis. The presence of diabetes was observed in 1 (3%), hypertension in 7 (25%), memory impairment in 6 (21%). and muscular pain in 7 (25%), all of which were proportionally higher in the myelopathy group, with statistical significance noted in the pain category ($p<0.01$). The role of each co-infection in HAM/TSP outcome is not yet completely clarified. Diseases caused by different pathogens, including HTLV-1, may involve an exacerbated immune response. Studies have shown that syphilis is an independent risk factor for HTLV-1 infection in males and females. In a Brazilian study, tuberculosis was associated with an increased risk of HAM/TSP development, maybe because of the change in immunological response. Some studies suggested that patients with HTLV-1 or HTLV-2 and HIV coinfection have worse clinical outcomes compared to other coinfections, in addition to having a higher mortality rate, also increasing the likelihood of developing neurological manifestations such as HAM/TSP. This study sheds light on the potential role of coinfections in amplifying the risk of HAM/TSP development. Data regarding co-infections' impact on HTLV-1 outcome is extremely limited and more studies are needed.

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COVID-19 and vascular complications: case analysis of ischemic stroke in the artery of percheron

Melissa Caroline Godoi Prestes[1], Luan Yudi Prando Kasuga[1], Bruna Beppler[1], Ádria Rodriges da Silva[1], Lia Maki Hatisuka Imai[1], Caroline Zapelini[1], Gabriel Rodrigues Espelho Rossi[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Melissa Caroline Godoi Prestes; Email: melissa.caroline@outlook.com.

Case presentation: An 83-year-old woman with a history of hypertension presented with mild diarrhea, rhinorrhea, cough, asthenia, and decreased functionality for three days. She was brought to a tertiary care center by emergency medical services due to a decreased level of consciousness. Family members reported that the patient fell asleep in the afternoon and did not wake up. On physical examination, she scored 3 on the Glasgow Coma Scale, exhibited global hyporeflexia, and had bilateral lung crackles. She was diagnosed with Covid-19 following a positive rapid antigen test. Chest computed tomography (CT) revealed bilateral consolidative foci. Cranial CT indicated a bilateral thalamic ischemic stroke, with well-defined hypodense areas in the medial thalami and the rostral midbrain, suggestive of recent ischemic lesions. The patient developed bilateral convergent strabismus, right hemiparesis, fluctuating consciousness levels, episodes of anisocoria and static gaze, dysphagia, productive cough, and progressively worsening lung crackles. She underwent a prolonged hospital stay, requiring intensive care, and experienced significant clinical deterioration, leading to a decision to limit support, resulting in death after approximately three months of hospitalization. **Discussion:** The artery of Percheron is a rare anatomical variation where a single artery supplies the paramedian thalamus and the majority of the rostral midbrain bilaterally. Occlusion of this artery results in bilateral thalamic ischemic stroke, characterized by severe symptoms. In Covid-19 patients, acute cerebrovascular diseases are relatively common, especially in individuals with pre-existing vascular risk factors, such as hypertension. Ischemic strokes are most frequently associated with Covid-19 due to pathophysiological mechanisms that include hypercoagulable states, vasculitis, and cardiomyopathy. **Final comments:** Ischemic stroke of the artery of Percheron is rare in the general population and poorly documented in Covid-19 patients. Considering the influences of Covid-19 infection, it is a significant risk factor for thrombotic events. Therefore, this case underscores the importance of recognizing extrapulmonary symptoms in patients with severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2).

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COVID-19 pandemic and sleep quality in medical students at university of Ribeirão Preto

Lara Helena Graña Uzun[1], Luisa Hasimyan Ferreira[1], Nathália Novaretti[1]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Lara Helena Graña Uzun; Email: anauzun@gmail.com.

Background: Among young adults, medical students have a special place in the populations with the highest risk factors for worse mental health, sleep deprivation, and sleep disorders. In this context, the impact of social isolation and the magnitude of the COVID-19 pandemic may have deteriorated pre-existing mental disorders and the physical health of these young people. However, there are still few Brazilian studies about the impact of the pandemic on this population. **Objective:** Evaluate the impacts of the pandemic on sleep quality and its related factors among medical students. **Methods:** Analytical cross-sectional study, in a single center. Information was collected from a questionnaire created using Google Forms and applied to medical students at the University of Ribeirão Preto between June and August 2021. **Results:** We collected 212 responses. The vast majority of students reported a drop in academic performance (76.4%) and mental health (82.5%), while 71.7% correlated this last topic to a drop in quality of life and/or academic performance. It is identified that the decrease in academic performance was mainly due to difficulty concentrating in front of computer screens (51.8%), lack of motivation (36.9%), external interference during classes (10.1%) and others (1.2%). 9.4% of the students also brought as challenges the difficulty in not picking up the cell phone during classes, workloads at night, lack of perspective in the world, fear and insecurity and a demotivating environment. Among the future doctors, only 39.2% continued to practice physical activities during the pandemic. Among those interviewed, 65.5% reported worsening of previous sleep disorders, diagnosed or not, with insomnia being the most prevalent. Regarding sleep hygiene, only 41.5% didn't know the method, while 59.1% performed it when they were unable to sleep. 58.9% confirmed the use of stimulants to stay awake. There was also evidence of a worsening in mental health (82.5%) during the period of social isolation and, despite this, 66.1% did not receive psychological counseling. It is also believed that the worsening of mental health is closely linked to the worsening of quality of life and/or academic performance during the isolation period (71.7%). **Conclusion:** The consequences of the pandemic on the quality of sleep and health of medical students are evident, and are bidirectionally associated with the physical and mental health of students. Therefore, not only their academic performance was affected, but the quality of our future professionals is at risk, making this a public health problem. New studies will be needed to assess whether this change was temporary or not, so that support and guidance measures should be applied in medical schools.

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Cranial nerve syndrome in a MOG-AB positive patient

Camille Freitas de Araujo[1], Gabriel Lisbôa Pereira[1], Pedro Ferreira Bastos[1], Cleonice Alves de Melo Bento[2], Priscila Mendonça Sacramento[1], Regina Maria Papais Alvarenga[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Camille Freitas de Araujo; Email: camillefreitasa9@gmail.com.

Case presentation: A 43-year-old white man presented with diplopia in July 2021 while looking in multiple directions. The patient was diagnosed with paralysis of the VI nerve on the right, and the ophthalmologist indicated treatment with orthoptics without resolution of the diplopia. She went to a neurologist and performed cerebrospinal fluid (CSF) analysis and magnetic resonance imaging (MRI). CSF examination on 30/07/2021 detected oligoclonal IgG band with excluded infections. An MRI scan in August 2021 showed a lesion with hypersignal on T2/FLAIR sequences in the integment of the pons in the topography of the nucleus of the VI cranial nerve on the right. As a result, he sought a specialist in demyelinating diseases with the suspicion of Multiple Sclerosis. Neurological examination showed recovery of ocular motility, and a deviation of the labial commissure was observed on the left, resulting in facial nerve involvement. Given the atypical location of the lesion, the biomarkers anti-aquaporin 4 (AQP4-AB) and anti-oligodendrocyte myelin glycoprotein (MOG-AB), positive for MOG-AB, were requested. On 15/01/2022, a new MRI showed an elongated hypersignal plaque with enhancement located in the middle of the cervical cord at the level of C5-C6, extending longitudinally for 8.5mm. In November, she presented a new outbreak with myelitis, with paresthesia and hypoesthesia in the right lower limb ascending to the abdomen. A new MRI was performed, which showed an increase in the cervical plaque and dorsal lesion at the level of D7. The patient underwent corticosteroid therapy with complete remission of spinal cord symptoms. In 2024, he started preventive treatment with Azathioprine 100mg/day. At the last follow-up on this occasion, the neurological examination was normal. **Discussion:** This study aims to report a case of MOG-AB positive disease (MOGAD) with rare brainstem (pons) involvement associated with myelitis in a context of differential diagnosis with multiple sclerosis. MOGAD is a recently described disease in patients with neuromyelitis optica and high-risk syndromes (transverse myelitis and optic neuritis) that are AQP4-AB negative but MOG-positive. It is a rare disease with an estimated annual incidence of 1.6 to 3.4/million people worldwide. The clinical manifestation presented in this case report is a rare presentation of MOGAD that is present in only 8-10% of reported cases. An article published in 2019 presented 3 case reports with cranial nerve syndrome in Mog-IgG positive patients, among them an association of the sixth and seventh cranial nerves with pons involvement in the nucleus region of the respective pairs. **Final comments:** Because it is a rare disease, it is extremely important to recognize atypical symptoms such as cranial nerve involvement associated with unilateral myelitis in cases of MOGAD. In the case reported here, we presented a patient with MOGAD initially characterized by cranial nerve injury with recurrent course and myelitis.

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Cranial nerves palsy due to pneumococcal meningitis: a case report

Yan Victor Araújo Rodrigues[1], Leandro Alves Mendes[1], Juliana Gaigher Gonçalves[1], Eliaquim Ribeiro de Oliveira[1], Beatriz Borba Casella[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Yan Victor Araujo Rodrigues; Email: yanvictorped@gmail.com.

A previously healthy 1 year and 2 months-old male child presents with fever and phlegm cough. He is taken to a pediatric visit on the fourth day of symptoms, when a macrolide and an antipyretic are prescribed, under the hypothesis of bacterial tonsillitis. On the sixth day of illness, he is again evaluated by a pediatrician, due to persistence of fever, and a restriction in the abduction of the left shoulder. Through an ultrasonography, an infectious arthritis was ruled out. The following day, the infant is once again taken to medical evaluation, for repeated vomiting, being once more discharged, this turn with antiemetics. After less than 24 hours, the patient develops fluctuation between drowsiness and irritability, neck stiffness and altered eye movement, which carries out hospitalization due to the hypothesis of acute meningoencephalitis. The initial neurological assessment exhibited wakefulness and behavior oscillations, mydriasis, divergent strabismus and right-sided ptosis. Furthermore, there was an infranuclear facial paralysis on the right side and a left hemiparesis with crural predominance, with an ipsilateral Babinski sign. In addition, behavior arrest seizure types are witnessed, requiring intravenous benzodiazepines Prior to initiating broad coverage for acute infectious meningoencephalitis (ceftriaxone, vancomycin and acyclovir), cerebrospinal fluid was collected, with results compatible with bacterial meningitis: pleocytosis of 2,240 cells/mm^3 (61% being neutrophils), high protein level (184mg/dL) and low glucose count (<5mg/dL). The bacterioscopic examination of the CSF showed gram-positive diplococci and, in spite of negative culture testing, the real-time polymerase chain reaction (RT-PCR) detected Streptococcus pneumoniae. Brain and cervico-thoraco-lumbar magnetic resonance imaging, performed when the patient already was on the twelfth day of antibiotics, showed dilation of the supratentorial ventricular system, with bilateral pyogenic ventriculitis. The electroencephalogram was normal. With regards to previous medical history, the child had 2 doses of 10-valent pneumococcal conjugate vaccine, at 2 and 4 months of age. However, he had not yet received the third (booster) dose scheduled for 12 months-old. During the subsequent days after initiation of antibacterial therapy, the child first improved mydriasis, divergent strabismus and peripheral facial paralysis. Thenceforth, the infant showed resolution of right hemiparesis. Upon being discharged from the hospital, after completing 14 days of ceftriaxone and vancomycin, however, a discreet eyelid ptosis on the right side remained.

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Creation of a university EEG study group to collaborate with medical education in electroencephalography with the democratic Republic of Congo, Africa

Maria Emilia Cosenza Andraus[1,2], Prince Kazadi[3], Augusto Cesar Marques Santiago[2], Ange-quentin Bedel Togbe[2], Candy Mbuyamba[2], Möise Lubendo[3], Daniel Tshiyuk[3], Nazaire Kimponto[3], Cesar Fantezia Andraus[1,2], Arjune Sen[4,5]

[1]Universidade Federal do Rio de Janeiro, Instituto de Neurologia Deolindo Couto, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Rio de Janeiro, Centro de Ciências da Saúde, Faculdade de Medicina, Rio de Janeiro RJ, Brazil.
[3]Epilepsy Awareness Foundation, Democratic Republic of Congo.
[4]University of Oxford, Oxford Epilepsy Research Group, Nuffield Department of Clinical Neurosciences, Oxford, United Kingdom.
[5]University of Oxford, Wolfson College, The Centre for Global Epilepsy, Oxford, United Kingdom.

Correspondence: Maria Emilia Cosenza Andraus; Email: cosenzaandraus@yahoo.com.

Background: The Democratic Republic of Congo (DRC), a large country located in Central Africa, has a population of around 105 million people. It is the 14th most populous country in the world and it is estimated that more than a million Congolese people have epilepsy. According to data from the Epilepsy Awareness Foundation for the DRC (ASLEK), the country has only ten neurologists, who concentrate their work mainly in large cities. There is a marked lack of equipment, such as electroencephalograms (EEGs), to aid the diagnosis of epilepsy. Previously, there were only five EEG machines to serve the entire population. In addition, there was a lack of local training and continuing education related to performing and interpreting EEG, making it necessary for clinicians to seek this from outside the country. **Objective:** To collaborate with continuing Medical Education in EEG with the DRC through the creation of a University Study Group. **Methods:** From May to July 2023, after contact from a group of doctors from the DRC requesting collaboration in training on performing and interpreting EEG, a University Extension Action was carried out under the title "Extension Course on Electroencephalography

and Video-Electroencephalographic Correlation of the Main Epileptic Syndromes in Adults and Children". The Action, registered with the Pro-Rectory of Extension of the Federal University of Rio de Janeiro (PR-5/UFRJ), Brazil, was delivered remotely to overcome barriers relating to travel and other costs. Training was directed to this group of doctors, who serve people with epilepsy in rural and poor setting areas and who already had basic working knowledge relating to EEG. **Results:** A Study Group was created, enlisting three undergraduate medical students already enrolled at our Faculty of Medicine in Brazil - one from the DRC, one from Benin (both French-speaking African countries) and one from Brazil. The final group was coordinated by a professor of clinical neurophysiology (MA) and included three professors (from the areas of neurology, clinical neurophysiology and neurosurgery), three undergraduate medical students and four doctors from the DRC (neurology and general medicine). The course was taught in English and the medical students helped in facilitating communication between MA and the French-speaking participants when this was necessary. Two-hour remote theoretical classes covered topics from the neurophysiological bases of EEG to its applications and clinical interpretation. The total time spent delivering the course was 24 hours. Also, through this Action, a Brazilian EEG company donated an EEG equipment to the DRC and a virtual workshop to use this equipment was held. **Conclusion:** The Extension Action has allowed the creation of a University Study Group that has been collaborating effectively with doctors in the DRC to deliver sustainable Medical Education in EEG.

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Creutzfeldt-Jakob disease (CJD): two-cases report

Elisangela Ferraz Pazinato[1], Alex Eduardo da Silva[1], Ana Luisa Rufino de Sousa[1], Giovani Zago Borges[1], Luiza Barbosa de Oliveira[1], Amanda Soares Pimenta[1], Daniela Aparecida Lima Viana[1], João Henrique da Matta Clementino[1]

[1]Universidade Federal do Triângulo Mineiro, Hospital de Clínicas, Uberaba MG, Brazil.

Correspondence: Elisangela Ferraz Pazinato; Email: elisangelapazinato@gmail.com.

Case presentation: Case 1: Female, 57 years old, 3 months before inpatient admission, presented progressive drowsiness, mental confusion, gait ataxia, myoclonus, tremors of the extremities and dysarthria. Magnetic resonance imaging (MRI) of the skull showed areas with restricted diffusion in the thalamus, putamen, caudate and globus pallidus bilaterally, as well as in the frontal neocortex. The electroencephalogram (EEG) showed a rapid temporal rhythm on the right hemisphere, sometimes bilateral, and diffuse disorganization of the tracing. Cerebrospinal fluid (CSF) analysis was positive for search of 14-3-3 protein. Case 2: Male, 77 years old, 18 days before admission, began with blurred vision associated with memory changes followed by slowed speech, difficulty walking and involuntary movements of the right upper limb with progression to the left upper limb. Skull MRI performed at an external service was reported without changes. The 14-3-3 protein was positive in CSF analysis. **Discussion:** Creutzfeldt-Jakob disease (CJD) is a rare, incurable neurodegenerative pathology with a poor prognosis and one of the causes of rapidly progressive dementia. In addition to cognitive impairment, patients with CJD may present with cerebellar, pyramidal and extrapyramidal motor, visual, and psychiatric symptoms. The sporadic form is the most common. Complementary exams such as cranial MRI, EEG and CSF analysis for the 14-3-3 protein are useful in investigating possible and probable CJD, but the definitive diagnosis is obtained from histopathological analysis of the central nervous system. **Final comments:** Although CJD is rare, it should always be considered as a differential diagnosis when suspected of rapidly progressive dementia.

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Creutzfeldt-Jakob disease Heidenhain variant: case report with progressive cognitive decline recorded by videos

Rafael Santalena Brambilla[1], Rosana Carandina-Maffeis[2], Hilton Mariano da Silva Júnior[1], Caroline Moraes Tapajós Bolzani[2], Pedro Neves Fortunato[2]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[2]Hospital Municipal Dr. Mário Gatti, Campinas SP, Brazil.

Correspondence: Rafael Santalena Brambilla; Email: rafael.bramba@gmail.com.

Case presentation: A 67 year old woman was admitted to the emergency room with a report of "seizures and dementia for 1 month". The condition started, in August 2020, with a decrease in visual acuity. Her son recorded the evolution of the disease in videos on his mobile phone. The patient got worse with inability to reach and to identify objects, optical apraxia, psychomotor slowing, abnormal repetition of acts and impairment of judgment. The first nuclear magnetic resonance (MRI) of the brain was normal and the electroencephalogram (EEG) was performed, with diffuse slowing of the baseline activity. She progressed with intermittent myoclonus and abulia. Extensive investigation for encephalitis was normal. Treatment with acyclovir and pulsetherapy with methylprednisolone were performed. A new brain nuclear magnetic resonance showed restricted area in the diffusion and T2 / FLAIR hypersignal exclusively cortical involving both hemispheres. Also new electroencephalogram showed generalized periodic discharges. The 14-3-3 protein was detected in her cerebrospinal fluid (CSF). The patient died 4 months after the onset of the condition due to infectious complications. **Discussion:** Creutzfeldt-Jakob disease (CJD) is a rapidly progressive neurodegenerative disorder. Heidenhain's variant has isolated visual symptoms that persist even without any cognitive decline for a few weeks. The video recording of the patient allows a detailed analysis of the clinical evolution and becomes an important complementary diagnostic tool. **Final comments:** The Heidenhain variant of CJD should be considered a differential diagnosis in all patients who have isolated visual complaints, especially if associated with cognitive complaints.

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Creutzfeldt-Jakob disease in Brazil: a review of its profile and notification mechanisms

Vinícius Bacelar Ferreira[1], Érica Giovanna de Souza Biváqua[1], Felipe Alves de Paiva[1], Pietra Amorim Cerquinho Oliveira[1], Riesly de Oliveira Vasconcelos[1], Ronilson Ferreira Freitas[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Vinícius Bacelar Ferreira; Email: viniciusbacelarferreira@gmail.com.

Creutzfeldt-Jakob disease (CJD) is a rare and fatal neurodegenerative condition, characterized by a rapidly progressive dementia. CJD is on compulsory notification list of diseases by the Health Ministry in Brazil and, even though it appears as a potential health risk, it lacks specific data about the disease in Brazil. This project analyzed the CJD profile in Brazil, with the objective to broaden the knowledge of its epidemiological pattern in the country and its distinct characteristics, utilizing as search engine PubMED e SciELO to search for case reports in the country, while the incidence of the disease in Brazil was analyzed based in data accessible through the "Sistema de Informação de Mortalidade" from DATASUS, using the mortality rates of CID A81 that englobes Creutzfeldt-Jakob Disease and other 4 types of neurological diseases, meaning that those data are just an approximation. In the research: initially 35 articles were selected to analyze, from which only 15 filled out the criteria established with 18 cases total. The DATASUS results revealed that CJD affects predominantly white females with greater incidence in the age range of 60 to 69 years, while in global reports the while males are more affected. The southeast region of Brazil is the most affected, principally the states of São Paulo, Rio de Janeiro e Minas Gerais. Meanwhile the case reports in Brazil reveal a younger profile with most case is white males, showing a divergence between what can be found in the databases and what it's been reported by physicians. Furthermore, it was possible to established that feature the disease as compulsory notification has been beneficial to raise notification, as has been show by the raise in numbers after 2018, when it was considered compulsory notification, even though there still are some divergences in the national protocol, if compared with the international standard, as consequence from Brazil's lack of specialized centers and professionals in the area, those divergence may impact and harden this disease's diagnosis. With all the data gathered, it is possible to affirm that Brazil may have a different epidemiological profile that the international one, also the protocols and data access related to CJD are still not optimized, in DATASUS it is impossible to access it specifically, meanwhile the Brazilian notification protocol are unspecific and not well defined, with a lack of specialized centers and personal, the diagnosis in Brazil are even harder than already is. Ultimately, a better notification and data access mechanism must be developed, as more specialized centers are needed.

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Creutzfeldt-Jakob disease, a series of cases in a public hospital in Paraíba, Brazil

Gilmar Leite Pessoa Filho[1], Ana Luisa Castelo Branco Gomes[1], Juliana Magalhães Leite[1], Tiago César Costa da Silva[2], Rafael de Souza Andrade[1], Matheus Gurgel Saraiva[1], Luciano Gonçalves do Nascimento Júnior[1], Felipe Nunes de Araújo[1], Rafael Gonçalves Duarte Cunha[1], Alex Tiburtino Meira[1]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[2]Universidade Federal da Paraíba, João Pessoa PB, Brazil.

Correspondence: Gilmar Leite Pessoa Filho; Email: gilmarpessoa@gmail.com.

Background: Creutzfeldt-Jakob disease (CJD) is a rare, transmissible and fatal neurodegenerative disease, caused by abnormally folded prion protein accumulation. Typically, patients present as a rapidly progressive dementia, associated to visual symptoms, movement disorders, cerebellar and pyramidal signs. The global incidence of CJD is unclear, although literature reports to be around 1-2 cases per million per year. Between 2005 and 2021, 10 suspected cases of CJD were reported in Paraíba, Brazil, four of them were confirmed. **Objective:** Herein we report a high incidence of CJD cases in a short period of time from a neurology quaternary hospital in Paraíba, Brazil. **Methods:** We report a series of 5 cases of patients with rapidly progressive dementia syndrome associated with other clinical and imaging signs that meet criteria for probable CJD, between 2022 and 2023 in a public hospital in Paraíba, Brazil. **Results:** Five cases of a rapidly progressive dementia suspected to be CJD related were referred to our service from April 2022 to September 2023. The average age was 63 years (ranging from 46 years old to 75 years old). Two (40%) of them were male. Four (80%) of these patients lived in the metropolitan area of João Pessoa, and only one lived in the rural area. All the cases were sporadic. All of them exhibited

dystonia, myoclonus, appendicular ataxia, rigidity, and global hyperreflexia. Seizures and akinetic mutism were observed in 4 (80%) of them. Magnetic Resonance Images findings were typical in all cases: bilateral and symmetrical hyperintensity in the basal ganglia and diffuse and asymmetrical hyperintensity in cortex, on DWI and T2/FLAIR, with no Gadolinium enhancement. Three patients (60%) showed the hockey stick sign. Three patients undergone Electroencephalogram, all of which showed triphasic waves. Four cases were confirmed by cerebrospinal fluid (CSF) real-time quaking-induced conversion (RT-QUIC), but CSF RT-QUIC of one of the cases was unavailable. Four individuals progressed to death, within less than 4 months of onset and only 1 for 10 months. **Conclusion:** Between April 2022 and September 2023, 5 suspected cases were recorded in just one neurology service, four of which were confirmed by RT-QUIC. Interestingly, 80% of these new patients resided in the same metropolitan region, although no association was found among them. The observed incidence appears to be higher than that described in the global literature, becoming a serious public health problem.

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Creutzfeldt-Jakob disease: a case report

Marina de Assis Brasil Mendes Monteiro[1], Carollyna Scherer Brum[1], Gabriel Bess Geier[1], Kelly de Oliveira Harada[1], Juliana Oliveira Freitas Silveira[2], Fernando Von Bock Bolli[3]

[1]Universidade Franciscana, Santa Maria RS, Brazil.
[2]Hospital Universitário de Santa Maria, Santa Maria RS, Brazil.
[3]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.

Correspondence: Fernando Von Bock Bolli; Email: fernandobolli@hotmail.com.

Case presentation: A 71-year-old man presents with a 6-month history of progressive cognitive impairment, disrupting his activities of daily living. He has been under treatment for heart disease and depression. Medications include desvenlafaxine, metoprolol and rivaroxaban. He denies any neurological disorder in his family. A Mini-Mental State Examination (MMSE) was performed during this evaluation, scoring 10 out of 30. Blood tests for measurement of lipid panel, vitamin B12, HbA1c, glucose, blood count and VDRL were normal, apart from TSH levels sligthly increased (6.22mUI/L). The MRI scan showed restriction to diffusion on frontal, temporal, parietal and occipital córtex. The patient was told about his guarded prognosis and lack of a specific therapy for his condition. **Discussion:** Creutzfeldt-Jakob disease (CJD) is a fatal spongiform encephalopathy caused by the transmission of prions, which are misfolded proteinaceous infectious particles. The concentration of prions in CJD is elevated in the retina, optic nerve and central nervous system, resulting in visual symptoms, cerebellar ataxia, extrapyramidal signs, cognitive impairment and myoclonus. CJD accounts for 60% of the main causes of rapidly progressive dementia and there are three major forms that human prion diseases can be manifested: sporadic (accounting for 85% to 90% of CJD cases), genetic (caused by mutations in the prion protein gene PRNP, with a prevalence of 10-15%) or acquired. Pathologically, CJD is characterized by spongiform degeneration of subcortical and cortical gray matter structures, nerve cell loss and gliosis. Definitive diagnosis requires the presence of scrapie associated fibrils in histopathological analysis by brain biopsy ou identification of the protease-resistant PrPSc with immunohistochemistry or Western blot methods. Clinical diagnostic criteria are based on evidence of rapidly progressive dementia with at least two typical features (myoclonus, visual symptoms, or cerebellar and extrapyramidal signs), compatible findings on MRI (diffusion restriction on basal ganglia, or at least two of temporal, parietal, and occipital cortical regions), sharp-wave complexes on EEG or dosage of 14-3-3 protein in the liquor. **Final comments:** The presented case reveals a challenging scenario of progressive cognitive decline in an elderly patient, whose symptoms lead to suspicion of CJD, a rare and fatal neurodegenerative condition. The diagnosis was based on the rapid progression of cognitive complaints associated, especially, with characteristic changes on MRI. Early recognition of these cases is crucial for guiding appropriate clinical management, despite the lack of specific therapies available, allowing for the implementation of supportive measures and palliative care aimed at improving the patient's quality of life and providing support to family members facing a reserved prognosis.

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Creutzfeldt-Jakob disease: a case report

Tairine Luiza Esteves da Costa Silva[1], Gabriela Flor Nimer[1], Caroline Bittar Moreira da Silva Amaral[1], Rafael Correia[1]

[1]Universidade Federal Fluminense , Niterói RJ, Brazil.

Correspondence: Gabriela Flor Nimer; Email: gfnmed1@gmail.com.

A 61-year-old male patient began experiencing difficulty naming words and forgetting passwords. Simultaneously, family members reported a slight alteration in his gait, with frequent stumbles. About 2 months later, he started experiencing spatial disorientation and emotional lability, becoming aggressive towards family members. He began experiencing visual and auditory hallucinations and also developed gait difficulty and falls. Approximately 6 months after the onset of symptoms, falls became frequent, and he developed dysphagia for solids and myoclonus. Due to injuries from a fall, the patient was hospitalized, and an investigation was initiated. Cranial MRI revealed hypersignal in T2 and FLAIR in the caudate nucleus, lentiform nucleus, thalamus, as well as involvement of the cerebral cortex including the cingulate gyrus bilaterally; also in the brainstem, insula, and higher portion of the superior frontal gyri, and left temporo-parietal region. Cerebrospinal fluid: positive for 14-3-3 protein. After discharge, he began outpatient follow-up at a tertiary hospital. On examination, he presented with myoclonus, mutism, and lack of cooperation with the examiner. He had spasticity and hyperreflexia, worse in the lower limbs. Levetiracetam was initiated during the consultation, and the indication for gastrostomy was discussed. Two months later, there was partial improvement in myoclonus, and the family accepted the placement of a gastrostomy tube. Creutzfeldt-Jakob disease is very rare, approximately one to two new cases occur per 1,000,000 in worldwide distribution. Clinical findings can be heterogeneous, but must be considered in case of rapidly progressive dementia. For the investigation, complementary exams are available that allow differentiation from other dementias, autoimmune or iatrogenic discordes. The patient presents clinical, radiological, and laboratory evidence confirming the diagnosis of Creutzfeldt-Jakob disease. This case illustrates the diagnostic approach and follow-up of a rare pathology and in addition, it presents high mortality. After diagnosis, it is important supportive and symptomatic treatment and a discussion about palliative care.

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Creutzfeldt-Jakob disease: a Heidenhain variant case report

Mayara Zavatini Cansini[1], Guilherme Menezes Mescolotte[2], Elaine Fernanda Dornelas de Souza[2], Pedro Henrique Queiroz de Oliveira[2], Maria Teresa Fernandes Castilho Garcia[2]

[1]Universidade do Oeste Paulista, Presidente Prudente SP, Brazil.
[2]Faculdade de Medicina de Presidente Prudente, Presidente Prudente SP, Brazil.

Correspondence: Mayara Zavatini Cansini; Email: mayazavatini@gmail.com.

Case presentation: We present a 64-year-old man, referred from a private clinic with a history of ataxia, diplopia, right upper eyelid ptosis, and sporadic episodes of mental confusion. Miller-Fisher syndrome, demyelinating syndrome, and encephalitis were considered initial diagnostic hypotheses. After further examination, he started with ocular apraxia, with moments when he reported not being able to open his eyes, and needed to use his hands to open it. He was diagnosed with prion disease with positive 14-3-3 protein in the cerebrospinal fluid (CSF) and radiological findings such as high signal intensities in the cerebral cortex and basal ganglia.From the onset of symptoms until diagnosis, there was a progressive worsening of the clinical picture leading him to be admitted to the ICU where he died due to complications of a pulmonary thromboembolism after 2 months of hospitalization. **Discussion:** Progressive neurological signs are common in SCJD, but ocular motor apraxia is not frequently reported and it is named Heidenhain variant Creutzfeldt-Jakob.A definite diagnosis requires the identification of the cardinal neuropathological features of sCJD (spongiform change, neuronal loss, and astrocytic gliosis) at postmortem or rarely brain biopsy. RT-QuIC and 14-3-3 protein in CSF can help in diagnosis. The most common findings of sCDJ on neurological examination include rapidly progressive dementia, visual disturbances and cerebellar ataxia, which are non-specific symptoms and can be present in a variety of neurological diseases.However, in the Heidenhain variant, the classic manifestation is cortical blindness resulting from impairment of the parieto-occipital cortex, and myoclonus and progressive dementia may be present. **Final comments:** Although sCJD is incurable and there is no generally accepted treatment currently available, it is important to make an early and accurate diagnosis because the early notice can help the family and the patient to prepare for the expected course of the condition and consider a better palliative treatment. In this case, visual changes were of utmost importance for clinical diagnosis, since their rarity helps to identify the condition and variant

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Crossed cerebellar diaschisis: a report of presentation with tardive ataxia and diagnosis with PET-CT and tractography

Evelliny Gomes da Silva[1], Eduardo Souza Melo[1], Luziany Carvalho Araújo[1], Arthur Cesário de Holanda[1], Bruno Henrique Carneiro Costa Filho[1], Allefy Beltrão Albano[1], Fabrício Martins Veras[1], Letícia Klabinske Marques Monteiro[1], Marcelo Palmares Oliveira e Silva[1], Maiza de Oliveira Tenório[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.

Correspondence: Evelliny Gomes da Silva; Email: evellinygomes1@hotmail.com.

Case presentation: Female, 76 years old, monitored at the Neurology outpatient clinic of the Hospital das Clínicas of the Federal University of Pernambuco, due to a history of a stroke in 2014, began to develop, in 2021, with global cerebellar ataxia (with an appendicular component more evident at the left), of a progressive nature, characterized by dysmetria in the index-nose and heel-knee maneuvers on the left, slurred speech, nystagmus, limb-trunk dyssynergia, tendon dance and drunken gait, in addition to mild motor sequelae in the right side of a previous stroke. She underwent extensive investigation

for sporadic cerebellar ataxia and received the diagnosis of crossed cerebellar diaschisis after excluding other etiologies of ataxias and undergoing confirmatory positron emission tomography (PET-CT) and magnetic resonance tractography. The patient's PET/CT showed an area of occipital, temporal and parietal hypometabolism on the right, compatible with an area of previous stroke and a cerebellum with hypouptake on the left, compatible with a diagnosis of CCD, and tractography confirmed the absence of the right-left corticopontocerebellar tract. **Discussion:** Crossed cerebellar diaschisis (CCD) is a rare phenomenon, initially described by Constantin von Monakow in 1914, and is characterized by reduced hemispheric cerebellar blood flow and metabolism that can occur contralateral to the supratentorial lesion. It is believed that the mechanism of CCD is due to the interruption of corticopontocerebellar tracts, and may be observed in patients with cerebral infarction, supratentorial tumors, epilepsy or encephalitis. Thus, it is noteworthy that only some patients with CCD may present functional or structural impairment. However, CCD can occur with hemiataxis and it is suggested that it results not only from the interruption of the corticopontocerebellar pathway, but also the dentatorubrothalamic pathway, justifying the patient's clinical description. It has also been shown that functional CCD leads to crossed cerebellar atrophy, that is, structural CCD, over time. This phenomenon was not seen in this case and can be justified by the fact that CCD cerebellar atrophy was more associated with younger patients. The classic diagnosis and finding on PET is, in fact, described as contralateral cerebellar and supratentorial hemispheric hypometabolism. **Final comments:** Indeed, this case illustrates clinical and imaging manifestations of an uncommon but important neurological clinical entity. Therefore, it is important to consider the importance of considering crossed cerebellar diaschisis in the differential diagnosis of ataxias, especially in older patients with previous structural brain damage, especially when secondary to a vascular accident.

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Crucial role of the ApoE4 variant in the pathogenesis of Alzheimer's disease in homozygous patients: a systematic review

Luiz Eduardo Ferreira Mazzanti[1], Nicolly Borges Costa Silva[2], Laís de Souza Rodrigues[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil. [2]Centro Universitário de Valença, Valença RJ, Brazil.

Correspondence: Luiz Eduardo Ferreira Mazzanti; Email: dudumazzanti4@gmail.com.

Alzheimer's disease (AD) is a progressive neurodegenerative condition that mainly affects patients' memory, thinking and behavior. It is one of the most common forms of dementia and accounts for the majority of cases in the elderly, placing a huge burden on the health system and families. In this context, it has been noted that individuals who inherit a copy of the apoE allele, a protein involved in the transportation and metabolism of lipids in the body, have an increased risk of developing Alzheimer's. The aim of this research was to analyze through a systematic review, the relevance of the ApoE4 allele in the development of AD and the consequences that can be caused in carriers of this allele. In this way, the PubMed databases and the regional portal of the virtual health library were taken into account with the descriptors "apoE4", "alzheimer" and "homozygous adults", using the Boolean operator "AND". In this context, the search reported 26 studies. The inclusion criteria were articles available in English and Portuguese, published between 2019 and 2024. The criteria for exclusion were articles that were not pertinent to the subject or duplicated. After using the exclusion and inclusion criteria, a total of 19 articles were selected for this systematic review. All of the articles were unanimous regarding the presence of the ApoE4 allele in homozygous patients with AD, because the presence of this apolipoprotein is associated with a greater accumulation of beta-amyloid protein plaques, which is the first step towards the onset of Alzheimer's. However, some studies have shown that the presence of this allele only has an impact on cognitive performance and does not influence cognitive performance. Nevertheless, some research has shown that the presence of the allele only has an impact on cognitive performance and has no impact on motor performance. It is also important to note that AD is multifactorial and is influenced by a combination of genetic and environmental factors, so not all individuals affected by the pathogenesis who have the ApoE4 allele will develop the disease.

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Cryptococcal meningoencephalitis in an immunocompetent patient: case report

Airton Sena Oliveira Sales[1], Danielli Regina da Silva[1], Laisy Bárbara Souza Silva[1], Larissa Batista Xavier[1], Lorenzo Pereira de Almeida Melo Montes[1], Guilherme Victor Rocha Pereira[1], Gabriel Vinícius de Andrade Rodrigues[1], João Pedro Lopes Geraldi[1], Emerson Oliveira Milhorin[1], Luís Carlos Oliveira Resende[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Airton Sena Oliveira Sales; Email: d202120376@uftm.edu.br.

Case presentation: A 64-year-old immunocompetent male with a medical history of diabetes, hypertension, and an unspecified pulmonary mass presented with neuropsychiatric symptoms for approximately one month. These symptoms included mood changes (irritability and emotional lability), cognitive dysfunction (memory impairment), and behavioral changes. Due to the progressive worsening of these symptoms, he was admitted to the hospital for further investigation. In addition to the presenting symptoms, he reported intermittent positive visual scotomas (red light flashes), bilateral frontotemporal headache refractory to common analgesics, and unquantified weight loss. During the investigation, cranial magnetic resonance imaging (MRI) revealed eight nodular lesions of uncertain etiology with adjacent hemorrhage, located near the cerebellar vermis, left occipital lobe, and right frontotemporal region. Initial differential diagnoses included primary or metastatic neoplasia and neuroinfection, leading to the decision to perform a biopsy of the lesions. The diagnosis of neuroinfection due to Cryptococcus gattii was confirmed, and treatment with Amphotericin B was initiated. The patient was discharged but returned to the emergency department eight weeks later with refractory septic shock and subsequently died. **Discussion:** Cryptococcus gattii meningoencephalitis predominantly affects immunocompetent individuals and can be associated with pulmonary involvement. Inflammatory lesions of the meninges and brain can impair cranial nerves (causing visual changes), cognition, and consciousness levels. Physical examination frequently reveals signs of meningeal irritation, focal neurological deficits, as well as nausea and vomiting.Diagnosis should be based on compatible clinical and epidemiological history supported by complementary exams such as MRI, cerebrospinal fluid (CSF) analysis, fungal cultures, and biopsy of lesions. The recommended treatment consists of intravenous administration of Amphotericin B combined with Fluconazole. **Final comments:** Fungal meningoencephalitis is rare in immunocompetent individuals but can cause severe manifestations. The clinical presentation and MRI findings are nonspecific and can be easily confused with other conditions, such as neoplasms and metastases, especially in the elderly. Therefore, it is important to consider the epidemiology of infections in tropical regions, which can provide valuable clues for accurate and early diagnosis.

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CSF1R-related leukoencephalopathy: case report

Luciane Ramos Perez Silveira[1], Paula de Oliveira Pinto[2] , Pedro Henrique Gargioni de Andrade[3]

[1]Universidade Federal de Pelotas, Pelotas RS, Brazil. [2]Universidade Federal da Fronteira Sul, Passo Fundo RS, Brazil. [3]Universidade Estadual de Maringá, Maringá PR, Brazil.

Correspondence: Luciane Ramos Perez Silveira; Email: lucianeperez@yahoo.com.br.

Hereditary diffuse leukoencephalopathy with spheroids (HDLS) is a rare autosomal dominant disorder characterized by giant neuroaxonal spheroids in the cerebral white matter (WM). Symptoms are variable and may encompass cognitive, psychiatric, and motor dysfunctions. **Case presentation:** Female patient, 28 years old, was admitted to the hospital emergency department with a progressive gait disturbance over the past three months, associated with behavioral changes, movement disorders, and episodes of urinary incontinence. She initially developed avolition and blunted affect at approximately 27 years of age, managed for depression with sertraline by the psychiatric team without improvement. Prior to symptom onset, she lived independently, worked without difficulty, and cared for her child. **Discussion:** There is no reported history of neurodevelopmental abnormalities or delays in the family. Family history includes non-consanguineous parents, a deceased brother at 9 years old from unknown causes, and a father who died at 30 years old in a car accident. On physical examination, she presented with a dysmorphic face, bradykinetic but oriented. Generalized preserved strength, choreoathetoid movements predominantly on the left side, hyperreflexia, and limb rigidity were noted. No skeletal abnormalities were observed. Laboratory tests were unremarkable. Cerebrospinal fluid analysis revealed 10 cells, 60 erythrocytes, glucose 59, protein 29, gram-negative staining. Brain MRI showed extensive signal abnormalities predominantly affecting periventricular white matter in both hemispheres, characterized by T1 hypointensity and T2/FLAIR hyperintensity, with scattered hemorrhagic foci suggesting leukoencephalopathy, vasculitis, inflammatory, or infectious processes. Treatment with methylprednisolone 1g/day was initiated without improvement. Comprehensive investigations including CT angiography of cerebral vessels, chest, abdominal, and pelvic CT scans revealed no significant findings. Further extensive screening for hematologic, infectious, or rheumatologic disorders was negative. Genetic testing for leukodystrophy revealed heterozygosity in the CSF1R (colony stimulating factor 1 receptor) gene. Over an 18-month follow-up period, the patient's condition progressed to cognitive and motor dysfunction, currently requiring a wheelchair, showing disengagement from others, speech and fluency disturbances, spasticity, hyperactive tendon reflexes, pathological reflexes, and recently developing dysphagia. **Final comments:** Accurate disease knowledge is crucial for correct diagnosis. Further studies are needed to elucidate disease classification and potential future treatments.

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Cutaneous anaplastic large cell lymphoma in a multiple sclerosis patient receiving natalizumab

Letícia da Silva Alves[1], Arthur Costa Nascimento[1], Rogério de Rizo Morales[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Letícia da Silva Alves; Email: draleticiasilvaalves@gmail.com.

A 23-year-old patient, who in 2021 presented with bilateral optic neuritis, which improved after pulse therapy with methylprednisolone. He met McDonald's 2017 diagnostic criteria and was diagnosed with relapsing-remitting multiple sclerosis (MS) in April 2021. Glatiramer was initiated until the release of high-potency medication, and he was switched to natalizumab in February 2022. However, due to high titers for the JC virus, therapy was changed to ocrelizumab in July 2023. In 2023, during outpatient follow-ups, lymph node enlargement was noticed in the cervical region, with slow and progressive growth. A biopsy performed in June 2023 revealed cutaneous anaplastic large cell lymphoma, with positive CD30 and negative ALK, and he was subsequently referred for oncological treatment. Ocrelizumab was suspended after the first dose for the treatment of lymphoma. Anaplastic cell lymphoma is a rare subtype of non-Hodgkin lymphoma. In the literature, there is a case report of a patient with multiple sclerosis using fingolimod, with a similar diagnosis. There is also an association of this type of non-Hodgkin's lymphoma with the use of glatiramer (GA). There are no reports in the literature of similar neoplasms in patients using natalizumab. GA has a mechanism of action that is not yet fully understood, but which involves activation of both the innate and adaptive immune systems, upregulation of anti-inflammatory M2 monocytes, Th2 cells and T regulatory cells (Tregs). The shift to Th2 immunity is potentially tumorigenic. Natalizumab acts as a barrier drug preventing the transmigration of lymphocytes by blocking cell adhesion through alpha4beta1 integrin. Several types of neoplasms have been reported with the use of natalizumab, including diffuse large B-cell lymphoma, with a probable mechanism associated with reduced migration of T cells to tumor sites, due to the blockade of alpha4 integrin that interferes with the activation of antigen-specific T cells. The discussion surrounding disease modifying therapies (DMTs) in MS is extensive. There is a consensus that immunosuppressive therapies with some drugs can increase the risk of neoplasia in patients with MS. However, the most commonly used DMTs have data indicating both their efficacy as anti-cancer medications and post-marketing reports of neoplasias, which do not confirm, but raise hypotheses regarding the nature of the association between them. Case reports involving neoplasms and patients undergoing treatment for autoimmune diseases are important in understanding the possible long-term effects of immunomodulatory treatments, as well as the risk-benefit ratio, and to aid in therapeutic decision-making and patient management, such as extra surveillance in relation to neoplasms, especially but not only in those with acquired or non-modifiable risk factors.

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Cyclic stereotypic catatonia revealing anti-nmda encephalitis

Victor Alfonso Garcia Ortiz[1], Manuella Edler Zandona, Lucas Spanemberg, William Alves Martins

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Victor Alfonso Garcia Ortiz; Email: victorgarcia.or@gmail.com.

Case presentation: The case involves a 42-year-old man with a three-month history of psychosis, stereotyped movements, and confusion, complicated by past drug abuse and moderate cranioencephalic trauma. Initially admitted for evaluation of seizures and psychiatric symptoms, he presented with agitation, delusions regarding harm from his girlfriend, and a history of recent cannabis and cocaine use cessation. His symptoms included auditory and visual hallucinations, leading to a bipolar disorder diagnosis despite treatment with multiple antipsychotics. After discharge against medical advice, his behavior deteriorated, exhibiting bizarre actions like rolling around lights and neglecting self-care. Neurological examination revealed frontal dysfunction, supported by MRI findings of encephalomalacia in the left pars opercularis. Treatment with various antiepileptic drugs for suspected seizures and psychosis showed no improvement. Upon admission to Hospital São Lucas, he displayed frontal dysfunction signs, including echolalia and delusions, despite treatment with olanzapine, carbamazepine, clobazam, and valproic acid. A 'washing out' phase of medication exacerbated his agitation and stereotypies, with catatonic episodes responding to lorazepam. Diagnostic tests revealed no new MRI findings but indicated suspicion of autoimmune encephalitis due to atypical progression and CSF abnormalities, including leukocytosis and elevated protein levels. Confirmation came from CSF testing showing anti-NMDA antibodies. Despite initial mental status improvement, he developed dysarthria, left arm dystonia, and parkinsonian features, partially alleviated by levodopa but complicated by bilateral convulsive seizures requiring diazepam. Treatment escalated to rituximab and cyclophosphamide, resulting in gradual recovery. Discharged on carbamazepine and levodopa, he continues rituximab infusions every six months. **Discussion:** Anti-NMDA receptor encephalitis involves autoantibodies targeting NMDA glutamate receptors, causing receptor dysfunction and severe neurological symptoms such as psychosis, hallucinations, seizures, and movement disorders. Frequently misdiagnosed initially as psychiatric illness, particularly in young adults, timely recognition is crucial to prevent severe morbidity and mortality. **Final comments:** This case underscores the challenges and complexities of diagnosing and managing autoimmune encephalitis, emphasizing the importance of comprehensive neurological evaluation and targeted immunotherapy for optimal patient outcomes. Cyclic catatonia may represent one of the many faces of anti-NMDA encephalitis and should prompt neurological assessment.

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Cytotoxic lesions of the corpus callosum (CLOCC): a neurological complication of dengue fever

Amanda Monteiro Viagi[1], Renato Pierre Lima[1], Rodrigo Meirelles Massaud[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Amanda Monteiro Viagi; Email: amandavalec@gmail.com.

Case presentation: A 25-year-old man presented with acute slurred speech and imbalance. He reported flu-like symptoms, including cough and diarrhea, starting six days prior, without fever. Three days before admission, he began azithromycin for respiratory symptoms. His history includes bipolar disorder and dyslexia, with regular use of lithium carbonate and divalproex sodium, without recent dose adjustments or misuse. During the neurological examination, he was awake, oriented, with slurred speech. Muscle strength was preserved, and cranial nerve examination was normal. Sensitivity was intact, but cerebellar testing revealed axial and bilateral appendicular ataxia, worse on the left, with dysmetria and dysdiadochokinesia. The NIH Stroke Scale score was 3, indicating ataxia and dysarthria. Immediate neuroimaging revealed a partially well-circumscribed lesion in the splenium of the corpus callosum on cranial MRI. The lesion appeared hyperintense on T2-weighted images and FLAIR sequences, with patchy DWI lesions, suggestive of cytotoxic lesions of the corpus callosum (CLOCC). The DWI lesion was extensive and larger than the lesion visible on FLAIR and T2-weighted sequences. The lesion did not enhance with gadolinium contrast. The TOF sequence in MRI did not demonstrate occlusion of major arterial trunks. Further investigation, including serum lithium levels, serological analyses, cerebrospinal fluid examination, PET-MRI with FDG imaging, electroencephalogram, chest and abdominal CT scans, and a respiratory panel with PCR for viruses and bacteria, revealed normal findings, except for a positive dengue IgM, with non-reactive NS1 antigen and IgG. During hospitalization, the patient's neurological deficits worsened, including severe dysphagia and dysarthria, tetraparesis and bilateral Babinski sign. Treatment with human immunoglobulin for 5 days followed by pulse therapy with 1000 mg of methylprednisolone for 5 days resulted in partial improvement. Follow-up MRI showed a reduction in lesion size and intensity in the splenium of the corpus callosum, with new multiple small foci of microbleeds, despite a normal platelet count. **Discussion:** We present a case of CLOCC secondary to dengue fever in an oligosymptomatic and afebrile patient at the onset of neurological signs. This condition can result from severe viral or bacterial infections and may also be associated with drug withdrawal, toxins, vascular conditions, metabolic disorders, drug toxicity, and seizures. A notable aspect of this case was the clinical manifestation of appendicular ataxia and pyramidal symptoms, along with dysphagia, dissociated from the cytotoxic lesions observed in the corpus callosum on MRI. PET MRI with FDG revealed normal findings, suggesting no metabolic alterations in other areas. **Final comments:** This case highlights several atypical features of CLOCCS secondary to dengue, including the patient's presentation and radiological progression, which are uncommon in CLOCCS.

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Dabigatran reduced the length of hospital stay for cerebral venous thrombosis

Camila Carneiro[1], Matheus Kahakura Franco Pedro[1], Vanessa Rizelio[1], Caroliny Trevisan Teixeira[1], Zuly Mary Briceno Moreno[1]

[1]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.

Correspondence: Camila Carneiro Ferreira; Email: camilasilcarneiro@gmail.com.

Background: From the current available evidence, it is reasonable to transition to direct oral anticoagulants or warfarin after a period of lead-in parenteral anticoagulation in the treatment of cerebral venous thrombosis (CVT). But whether 5 to 15 days to do the transition to oral anticoagulants is a safer or more effective strategy than shorter periods is not known. **Objective:** In this study we demonstrate the safety and effectiveness of the transition carried out over an average period of 3,6 days. **Methods:** A retrospective, single-center, longitudinal study evaluating data from the electronic medical records of patients of both sexes hospitalized for CVT from 2014 to 2019 (Warfarin group) and from 2019 to 2023 (Dabigatran group). Statistical analyses were carried out using SPSS (Statistical Package for the Social Sciences) software. **Results:** A total of 137 patients were included: 73 in the warfarin group (WG) and 64 in the dabigatran group (DG). In both groups, patients received 3 days of low molecular weight heparin. After this phase, WG received 10 mg of warfarin until they reached an INR of 2 to 3, and the DG started on 150 mg of dabigatran twice a day and could be discharged. In the warfarin group, there were 57 women (78%), the mean age (SD) of the patients was 36.7 (±10.7). 38 (52%) were using some hormonal therapy, and 16 (22%) were found to have thrombophilia. On admission, 7 (9.6%) had already been with venous infarction, and 13 (17.8%) had refractory symptoms of intracranial hypertension. Of the 73 patients, only 58 (79.5%) were followed up for more than 3 months after hospitalization. None of these patients experienced anticoagulant-related bleeding, 5 (8.6%) had a recurrence of CVT (over 5 years of follow-up), and all achieved recanalization during follow-up. In the dabigatran group, there were 41 women (64%). The mean age (SD) of the patients was 38.6 (±11.8). Twenty-five (39%) were using some hormonal therapy, and 11 (17%) were found to have thrombophilia. On admission, 11 (17%) had already been with venous infarction, and 19 (30%)

had refractory symptoms of intracranial hypertension. Of the 64 patients, 42 (65%) were followed up for more than 3 months after hospitalization. None of these patients experienced anticoagulant-related bleeding, 1 (2.3%) had a recurrence of CVT (over 2 years of follow-up), and all achieved recanalization during follow-up. The mean (SD) length of hospital stay in the WG was 5.42 days (±2.2), and in the DG it was 3.69 days (±1.2). The difference between the means was 1.73 days, which was statistically significant (T= 5.76; p< 0.001) using the Student's t-test. **Conclusion:** The transition from heparin to oral anticoagulants within 5 days was safe and effective in this coorte. Dabigatran can reduce the length of hospital stay even further, as it does not require dose adjustments according to the international standard of 2 to 3, which is necessary with warfarin.

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DBS of the Meynert basal nucleus for patients with Lewy body dementia: a meta-analysis of randomized clinical trials

Jonatas Paulino da Cunha Monteiro Ribeiro[1], Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Dário Correia Negreiros[1], Márcio ítalo Santos de Oliveira[1], Társis Vinícius Cronemberger de Carvalho Moura Mendes[1], Guilherme Victor Antas Cronemberger[1], Thaís de Negreiros Neves[1], Luciano Barbosa de Sousa Santos Filho[1], João Luís Reis Freitas[2], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Jonatas Paulino da Cunha Monteiro Ribeiro; Email: jonatas.ribeiro@ufpi.edu.br.

Background: Lewy body dementia (LBD) is a neurodegenerative disease clinically characterized by motor and cognitive symptoms. Its pathophysiology is based on the development of abnormal clusters of defective proteins called alpha-synuclein, which settle in neurons and interneuronal spaces. Currently, LBD has no disease-modifying therapy, with drug treatment offering only moderate improvement in symptoms. Deep brain stimulation (DBS) of the nucleus basalis of Meynert (NBM) is not used therapeutically in these cases, however, considering the impact of this disease and the limitation of pharmacological therapy, it is important to evaluate the effects of procedures such as this in the cognition of patients with LBD. **Objective:** To evaluate the cognitive and neuropsychiatric impact of DBS of the nucleus basalis of Meynert in patients with LBD. **Methods:** Two investigators systematically searched Embase, Cochrane and PubMed for Randomized Controlled Trials (RCTs), with no publication year limit, that compared DBS of the basal nucleus of Meynert with "sham" procedures in patients with LBD. Outcomes of interest included were the change in the Mini-Mental State Examination (MMSE) and Neuropsychiatric inventory (NPI) scales after the intervention. Statistical analysis was performed using the R Studio 4.3.2 software. Heterogeneity was calculated based on the I2 statistic. **Results:** Three RCTs were included in the statistical analyses, it was observed that the NPI and MMSE variables are reported as secondary outcomes. A total of 18 patients, all males aged between 35 and 80 years, 6 patients from each study and 9 for each arm (DBS or sham procedure). For the MMSE measure, there was no statistical significance, with a mean of -0.658 [-3.670, 2.353]. For the NPI measure, there was no statistical significance, with a mean of -6.37 [-16.16, 3.42]. The results are the same for fixed and variable effect methods, and this is due to the fact that there are few studies. Thus, the estimate for tau squared is not reliable. **Conclusion:** Evaluating the cognitive and neuropsychiatric impact of DBS of the basal nucleus of Meynert in patients with LBD, there was no statistical significance for the clinical improvement in the cognitive segment, based on the change in the MMSE and NPI scales.

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Deaths to epilepsy in Brazil: a comparison between sex and age group from 2018 to 2022

Izabelly Constatinov Lemos[1], Gabriel Cordova[1], Emanuel Henrique dos Santos[1], Anne Beatriz Ricardo Amaral[1], Sofia Beatriz Andrade Manfio[1], Kauan Correia Derbli[1], Julia Farto Viana Oliveira[2], Gabriel André Silvério[1]

[1]Centro Universitário Campo Real, Guarapuava PR, Brazil.
[2]Universidade Estadual do Centro Oeste, Guarapuava PR, Brazil.

Correspondence: Izabelly Constantinov Lemos; Email: med-izabellylemos@camporeal.edu.br.

Background: Epileptic seizures are abnormal brain states caused by a period of uncontrolled synchronous excitation. The ion channels of the Central Nervous System, involved in communication through the release of neurotransmitters, appear unbalanced in the pathophysiological process of the disease. Studies mention that epileptic channelopathies are derived from genetic mutations, acting on voltage-sensitive channels such as Na+, K+, Ca2+, and Cl-, as well as the nicotinic acetylcholine receptor and GABAA. The high morbidity and mortality rate of the epileptic state is characterized by recurrent seizures, lasting for at least 5 minutes without return to normal physiological characteristics. Considering all this information, the need for investigative research on the epidemiological characteristics of mortality from epilepsy in Brazil becomes clear, especially the comparison between sex and age group, covering the period from 2018 to 2022. **Objective:** To determine the number of deaths (n) from epilepsy in Brazil, associating and comparing the most affected age group and sex between 2018 and 2022. **Methods:** Observational epidemiological study of a descriptive nature based on data extracted from the Department of Epidemiological Analysis and Surveillance of Non-Communicable Diseases (DAENT) referring to the number of deaths from epilepsy in Brazil in the period from 2018 to 2022. The constants used were: place of occurrence, coverage, deaths by residence, notification of deaths, without restriction of race/color. And as variables: sex and age group (0 to ages above 80 years). The ignored category was included in the total number of deaths, since it has little impact on the study results. The indicator used was G40.9 epilepsy, not specified. Subsequently, an analysis was carried out to identify which age groups and sex are most affected. **Results:** Of the total number of deaths (n=14,379), male patients (n=9,322) had the highest number of deaths from epilepsy between 2018 and 2022, accounting for 64.8% of deaths. In comparison, female patients had a significant number of deaths, totaling 5,054 individuals during this period. In addition, the most affected age group was 50 to 59 years old, with 18% (n=2,592) of the total analyzed. **Conclusion:** The number of deaths increased in the period from 2018 to 2022, mainly among men in the age group of 50 to 59 years old. Consequently, it is noted that more research is essential to understand the development of epilepsy in the affected age group and sex, with the aim of ensuring an accurate epidemiological perception and promoting appropriate multidisciplinary care for the profiles presented.

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Deciphering the TBK1 mutation: a key to unlocking the mysteries of frontotemporal dementia and als in Brazilian families

Rachel Leirner Argelazi[1], Santhiago Calvelo Graça[1], Pedro Vilaça Thomazoni[1], Yngrid Dieguez Ferreira[2], Diogo Haddad Santos[2]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Rachel Leirner Argelazi; Email: racheleirner@gmail.com.

Case presentation: In April 2021, two brothers, JLS1, 43 years, and GLS, 45 years, presented at the neurology outpatient clinic with a significant history of cognitive decline starting in their early twenties. Both brothers exhibited progressive symptoms leading to dependence on daily life activities. JLS1 began to show behavioral changes at 36, leading to job loss and increasing social isolation. His condition worsened over time, with symptoms including refusal of personal care, aggression, tremors, dysarthria, and cognitive decline. By 2018, JLS1 was admitted to Dom Pedro Hospital at São Paulo, where he remained socially isolated, experiencing psychomotor agitation and aggression. MRI scans revealed diffuse brain volume reduction, particularly in the caudate nuclei, suggesting a neurodegenerative disorder such as Huntington's disease. GLS, also from São Paulo, had a similar trajectory. He experienced depressive episodes and behavioral changes that intensified after losing his job in 2014, leading to a suicide attempt. Over the years, his symptoms included psychosis, cognitive decline, hallucinations, and severe motor deficits. Despite testing negative for Huntington's disease, MRI scans showed significant brain atrophy and signs of encephalomalacia/gliosis. The brothers' condition reflected a troubling pattern within their family. The youngest brother, JLS2, at 36, exhibited early symptoms and lived in poor conditions. The family history included similar symptoms in their father, uncle, grandmother, and a cousin, starting around age 20 and leading to severe cognitive and motor deficits by their 40s. In May 2022, genetic testing of GLS identified a variant in the TBK1 gene (c.965A> T), linked to frontotemporal dementia (FTD) and amyotrophic lateral sclerosis (ALS). This suggested a genetic basis for the family's symptoms, though the variant's pathogenicity remains uncertain. Both GLS and JLS1 passed away, while the status of JLS2 and other family members remains unknown. **Discussion:** Mutations in the TBK1 gene have been linked to an increased risk of developing both FTD and ALS. The phenotype is diverse, including behavioral variant FTD, primary progressive aphasia, and pure ALS. Studies have found a higher prevalence of TBK1 mutations in European patients compared to Asian patients. In Brazil, only one case report was published on the gene and clinical manifestation. **Final comments:** Despite its rarity, frontotemporal dementia accompanied by motor deficits is significant for neurologists due to the guarded prognosis and potential familial impact on the patient's descendants. In this study, we aim to write a case report on this rare, hereditary neurodegenerative disease that results in rapid neurodegeneration. Our goal is to contribute to the scientific understanding of frontotemporal dementia.

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Deep learning for neuroimage classification in Alzheimer's disease: barriers to clinical relevance

Caio Hussid de Góes[1], Diogo Haddad Santos[2]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Caio Hussid de Góes; Email: caio.goes@aluno.fcmsantacasasp.edu.br.

Background: As new pharmacological treatment options emerge for Alzheimer's disease (AD), reliable early diagnosis escalate in importance. In the clinical setting, neuroimaging methods in AD face problems related to the diversity of brain degeneration patterns, as well as overlapping of Alzheimer's characteristics with other dementias.

A possible approach is the development of image classification algorithms through Deep Learning models, a subtype of Machine Learning. Thus, study's **Objective:** was to carry out a literature review to clarify the barriers for clinical use of Deep Learning models capable of identifying patients who will undergo conversion from Mild Cognitive Impairment (MCI) to Alzheimer's. **Methods:** The methodology was organized according to the PRISMA guideline of 2020. Article search was conducted in Pubmed, between 2007 and 2023, while the MeSH terms were chosen based on the pertinent topics for the study: clinical presentations, Deep Learning models, and Neuroimages. After the search, articles were sorted for inclusion based on abstract. Then, the full text of the articles was analyzed, followed by data extraction. Articles that only mentioned Deep Learning, articles that did not predict the conversion of MCI into Alzheimer's and that used neuroimaging sample amplification methods were excluded, as were articles not available in English and studies without access to the full text. **Results:** The initial search resulted in N=178 articles. Of these, 128 were excluded, and of the remaining 50, n=34 fit the inclusion criteria. The most frequent model used was a 3D Convolutional Neural Network (17), followed by Deep Belief Network combined with Support Vector Machine (2), and the remaining 15 models being used by a single article. All articles used the Alzheimer's Disease Neuroimaging Initiative dataset for models, with other 4 much smaller dataset used in 4 different articles. A total of 9 different categories of additional input data were used alongside neuroimages across 14 articles, and 6 articles performed model cross validation. Analyzing different Deep Learning architectures proved to be complex. Inconsistent reporting of important steps in the model's construction was found in the pre-processing protocols for input data, the different processes for obtaining model hyperparameters, and a tendency of association of different architectures to obtain robust models, hurting the study's reproducibility and validation. Moreover, the results reveal a lack of diversity in datasets used for model training, limiting the applicability of the models in real world scenarios despite the relatively high performance reported in most models, which is made worse by the lack of model cross validation. **Conclusion:** Ultimately, it can be concluded that the applicability of Deep Learning models for neuroimaging classification is still in its infancy, and while it shows potential, more studies with robust methodology reports are needed for effective real-world usage.

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Deep phenotyping of hereditary amyloidosis I107V in a Brazilian cohort

Jemima Araujo da Silva Batista[1], Wilson Marques Jr[2], Cleonisio Leite Rodrigues[3], Lara Albuquerque de Brito[3], Adilson Junior Pinto Galvão[4], Renata Amaral Andrade de Mendonça[5], Anna Paula Paranhos Miranda Covaleski[6], Marcela Camara Machado Costa[7]

[1]Universidade Federal do Vale do São Francisco, Petrolina PE, Brazil.
[2]Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[3]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.
[4]Hospital Regional Dr. Mário Dourado Sobrinho, Irecê BA, Brazil.
[5]Hospital Pelopidas Silveira, Recife PE, Brazil.
[6]Serviço de Referência em Doenças Raras de Pernambuco, Recife PE, Brazil.
[7]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Jemima Araujo da Silva Batista; Email: jemimaaraujods@gmail.com.

Background: Hereditary amyloidosis is a multisystemic and heterogeneous disease whose clinical presentation and prognosis varying according to the genotype. The I107V is a rare variant that has been more frequently described in Brazil. However, its precise phenotypic characterization is still incipient, which hinders both its early recognition and optimal clinical management. **Objective:** To conduct an in-depth phenotyping of the I107V variant (ATTRI107V). **Methods:** This is an observational, retrospective, multicenter study that included carriers of ATTR I107V treated in four centers in Brazil between 2020 and 2023. Asymptomatic individuals and those with additional causes for neuropathy were excluded. Demographic and clinical data were analyzed. Quantitative data were described as means and standard deviations, while categorical variables were expressed as frequencies. **Results:** A total of 88 individuals with the I107V variant were identified, of whom 51 were excluded due to additional causes of neuropathy or incomplete evaluation. Thirty-seven individuals were included, 59% of whom were female. The average age of symptom onset was 55±12 years (range: 22-70). The most frequent initial presentation was small fiber neuropathy (SFN) (38%), followed by SFN associated with sensorimotor and autonomic neuropathy (24%). Isolated autonomic neuropathy occurred in 5% of cases, and cardiac onset in 3%. During the disease, 76% presented with dysautonomia, with orthostatic hypotension being the most frequent symptom (50%). Muscle weakness occurred in 59% of patients, with an average onset time of 2.43±1.25 years. The interval between symptom onset and bed restriction was 4.7±1.5 years. Cranial nerve involvement was observed in 64% of patients, with the most frequent manifestations being dysgeusia (47%), followed by tongue atrophy (36%) and dysphonia (33%). Carpal tunnel syndrome (STC) was identified in 82% of cases and preceded polyneuropathy by about 3.4±4.5 years (0.5-24). Electroneuromyography demonstrated axonal polyneuropathy in 62%, mixed in 4%, demyelinating in 4%, and was normal in 8%. Tafamidis was the indicated treatment for most individuals (73%). Only 23% exhibited cardiac symptoms; however, 86% had ECG or Holter alterations, and 43% had cardiopathy on echocardiogram. Cataracts were identified in 15% and dry eyes in 13% of cases. No individual presented with renal impairment. The mortality rate was 16%, and the average survival was 9.3±2.3 years (7-13). **Conclusion:** The ATTRI107V presented a mixed phenotype, with a predominantly neurological initial manifestation, late disease onset, and rapid progression. Dysautonomia and STC were frequent and may serve as warning symptoms for early diagnosis. Cranial nerve involvement is prevalent and should be recognized in these individuals. Cardiac involvement is frequent and can be asymptomatic, underscoring the importance of systematic cardiac investigation and multidisciplinary follow-up.

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Delirium phenotype in acute stroke

Gabriel Souza Suzart[1], Luiz Paulo Oliveira Martins[1], João Pedro Fernandes Gonçalves[1], Marivaldo Yslan Silveira Leal[1], Luísa Dias Moreno Aravena[1], Thais Barbosa de Oliveira[1], Vinícius Bessa Mendez[1], Alex de Cerqueira Silveira Figueiredo[1], Gabriel Souza Medrado Nunes[1], Milena Fernandes de Oliveira[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Luiz Paulo Oliveira Martins; Email: luizpom2000@gmail.com.

Background: Delirium is a common complication in acute stroke. The literature is scarce in studies evaluating underlying delirium phenotype in this setting. **Objective:** Determining the prevalence of delirium phenotypes and its association with long term functional outcome. **Methods:** Prospective cohort of stroke patients admitted to a brazilian reference stroke unit. Subjects included were older than 18 years admitted to the stroke unit within 72 hours of symptoms onset. Delirium was assessed during hospitalization by the Confusion Assessment Method for the Intensive Care Unit. Delirium phenotypes were septic, hypoxic, sedative-related, metabolic, multiple causes or undetermined. The main outcome was functional dependence or death in 90 days after stroke, determined by the modified Rankin Scale >2 points. **Results:** A total of 213 patients were included. Our sample's mean age was 66 (±13) years, with a majority of first ever stroke (74.3%) and ischemic subtype (93.9%). Delirium occurred in 47 (22.1%) patients, from which its phenotype was determined in 15 (31.9%) cases. The septic delirium was the most common phenotype (14.9%), followed by sedative-related (6.4%), multiple causes (6.4%), metabolic (2.1%) and hypoxemic (2.1%), being undetermined in the remaining episodes (68.1%). Delirium patients had a higher rate of poor outcome (odds ratio, 2.2 [IC 95%, 1.6-2.9; p<0.001). Worse outcomes were less frequent in sedative-related and undetermined delirium compared to other phenotypes (0% vs 80% vs 100%). **Conclusion:** Delirium is a clinical predictor of poor prognosis in acute stroke settings. Septic, metabolic and hypoxic delirium phenotypes might have worse effect over functional outcome compared to sedative-related and undetermined causes. To our knowledge, this is the first study to explore delirium phenotypes in acute stroke. Higher sample size is needed to confirm our findings.

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Delirium post-stroke: does the cognitive phenotype impact the patient's functional outcome?

Milena Fernandes de Oliveira[1], Nara Samara Silva dos Santos[1], Gabriel Souza Medrado Nunes[1], Thais Barbosa de Oliveira[1], Maria Eduarda Messias Vassoler[1], Luiz Paulo Oliveira Martins[1], Alex de Cerqueira Silveira Figueiredo[1], Antonio Lázaro Santos Soares Junior[1], Marivaldo Yslan Silveira Leal[1], Pedro Antônio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Milena Fernandes de Oliveira; Email: milenafer.oliv@gmail.com.

Background: Delirium, an acute cognitive disorder with attention fluctuations, can lead to worse outcomes in post-stroke patients. As for its phenotypes, it is traditionally classified according to the presented level of agitation-sedation, using the Richmond Agitation Sedation Scale (RASS), however, some patients only show fluctuations of inattention and disorganized thoughts, without alteration in the level of consciousness. Therefore, it is important to investigate whether these presentations have a different impact on the functional outcome. **Objective:** To identify if altered levels of consciousness in delirium lead to worse functional outcomes in post-stroke patients. **Methods:** Prospective cohort study with patients over 17 years old admitted to a semi-intensive care unit, from July/2023 to March/2024, in a reference center in Bahia, Brazil. We collected clinical and demographic data through bedside interviews. The level of altered consciousness was classified according to RASS and the presence of disturbances in the content of consciousness was assessed according to the Confusion Assessment Method for the ICU (CAM-ICU) every 48 hours of hospitalization. As for the functional outcome, patients were classified according to the Modified Rankin Scale (mRS) using a bedside approach or through calls to family members after 30 days of the stroke episode. We considered mRS 0-2 as a favorable outcome and mRS 3-6 as a poor outcome. The chi-square test was used for the statistical analysis. **Results:** A total of 213 patients were admitted, of whom 46 suffered from delirium during their hospitalization. Of these 46, 3 were excluded from the analysis due to mRS missing data. Of the patients included in the study, 51.2% were female, as for ethnicity, 30.2% were white, 67.4% were African descent, and 2.3% were Asian descent. Regarding age, the average was 68.9 (±1.96). As for the delirium phenotype, 29 patients had at least one episode of RASS other than 0 concomitant with attention disturbance; 7 (24%) had

favorable outcomes and 22 (66%) had poor outcomes. Regarding the other 14 individuals who presented delirium with only attention disturbance, 4 (28.5%) had favorable outcomes, and 10 (71.5%) had poor outcomes. In the statistical analysis, there was no difference between the proportions of bad outcomes in patients with RASS other than 0 and those with 0 (p=1.000). **Conclusion:** In the studied sample, altered consciousness in delirium did not significantly impact functional outcomes. These results emphasize the importance of addressing delirium in patients without altered consciousness, highlighting the need for prevention, treatment, and further studies with larger samples to better understand these cases, considering that delirium is often underreported in the ICU, particularly in patients without significant changes in consciousness.

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Dementia in public health in Brazil: epidemiological analysis regarding hospitalizations and mortality between 2012 and 2022

Ana Laura Capel Sanchez[1], Thiago Oscar Goulart[2], Nathália Novaretti[1]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Harvard University, MA, USA.

Correspondence: Ana Laura Capel Sanchez; Email: analauracapelsanchez@gmail.com.

Background: Dementia is characterized as the association of cognitive and functional impairment. Alzheimer's disease (AD) and vascular dementia (VD) are the main etiologies. Their prevalence has increased worldwide due to aging and demographic changes. Thus, analyzing its epidemiology and its socioeconomic impact is of utmost importance. **Objective:** To perform an epidemiological analysis of dementia syndrome and its burden on the Brazilian public health system. **Methods:** We collected data from DATASUS, a public and open database, searching for the ICD-10 related to Dementias (F00 - F03). Then, we did the statistical analysis with Excel and RStudio regarding the number of hospitalizations and deaths due to dementia syndromes. **Results:** Between 2012 and 2022, Vascular Dementia (F01) exhibited fluctuating death tolls, with 252 deaths reported in 2013 and 461 in 2022. Unspecified Dementia (F03) emerged as the leading cause of death, surpassing other categories and accumulating 29,168 deaths over the decade. However, the data, sourced from death certificates that record only the underlying cause, may underestimate the full impact of dementia-related fatalities. The G30 category experienced a surge in deaths from 13,142 in 2012 to 29,429 in 2022, totaling 221,004 deaths. Altogether, there were 286,524 dementia-related deaths from 2012 to 2022. Mortality rates in the F03 category varied regionally, notably showing a consistent rise in the Southeast region, especially in 2021. In contrast, mortality rates in the F01 category fluctuated across regions, with the Southeast exhibiting higher and relatively stable rates, peaking in 2019. Dementia hospitalization trends over the decade reflected distinct patterns among regions, where the Southeast recorded the highest number of hospitalizations (17,889), while the North Region reported the lowest hospital admission count (512), indicating regional disparities in healthcare utilization for dementia cases. **Conclusion:** Overall, the analysis reveals significant variations in dementia-related deaths and hospitalizations over the past decade, highlighting the notable impact of Vascular Dementia (F01) and Unspecified Dementia (F03) in mortality rates. The data underscore the regional disparities in mortality trends, with the Southeast region demonstrating higher rates and consistent increases. These findings emphasize the importance of targeted interventions and regional healthcare strategies to address the varying burden of dementia across different areas.

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Dementia syndrome in multiple sclerosis

Andreia Costa Rabelo[1], Denise Sisterolli Diniz[1], Kamylla Camilo Araujo de Oliveira Diniz[1], Amanda Rabelo Mendonça[2], Leonardo Ferreira Caixeta[1]

[1]Universidade Federal de Goiás, Goiânia GO, Brazil.
[2]Escola Superior de Ciências da Saúde, Brasília DF, Brazil.

Correspondence: Amanda Rabelo Mendonça; Email: amandarm.13@gmail.com.

Background: Dementia syndrome is a clinical condition that results in cognitive decline and impairs patient activities. Cognitive impairment (CI) in Multiple Sclerosis (MS) is a prevalent and disabiliting symptom. Memory, attention and information processing speed are the most common affected cognitive domains, followed by spatial vision, executive functions and verbal fluency. The spectrum of cognitive symptoms in MS may vary in intensity from mild cognitive impairment (MCI) to installed dementia syndrome. **Objective:** To describe finds of dementia syndrome in people with MS (PwMS) in a reference center. **Methods:** The sample included 117 participants with MS randomly selected. All patients were diagnosed and followed up by a neurologist with expertise in MS. Inclusion criteria: patients with MS, <18 years old, vision acuity at least 0.5 in both eyes with or without lens correction, depression only if treated and EDSS <6.5. Exclusion criteria was presence of motor or visual deficit that interferes in tests response. The neuropsychological and behavioral assessment occurred all in a day conducted by a single neuropsychologist using the following instruments in order of application: Brief Repeatable Battery of Neuropsychological Tests (BRB-N), Beck Depression Inventory-2ª edition and Beck Anxiety Inventory. A neuropsychiatrist specialized in cognitive disorder and behavioral changes diagnosed them according to DSM-5 criteria to dementia syndrome. The study was approved by a local Ethics Committee. **Results:** The sample was 80.3% female, 70.1% had ≥10 years of schooling and mean age 42 years old (SD= ±13.3). Regarding clinical profile, 83% presented RRMS, 59.8% was diagnosed >6 years, disease time 7,85 years (SD±5.47) and EDSS <3.0. Were found 20 PwMS that met dementia syndrome's criteria, 7 males and 17 females, 50% with <9 years education, only 25% with EDSS >6, 70% of them 49 year old or younger and 50% presented Bipolar Disorder. The dementia cases divided into fenotypes: 60% like Frontotemporal lobar degeneration (50% apathetic and 50% uninhibited), 20% Alzheimer's disease and 20% none specific. Cognitively, language, semantic memory and executive functions, sensitive to temporal and frontal lobe dysfunctions, was the most impaired performance, following by evocation of visual memory and processing speed. Patients with dementia syndrome's performance was lower and indicate severe clinical deficits: 13 of them had 5 compromised domains and the other 7, 3 or 4 domains. **Conclusion:** MS causes CI in most patients and may increase the possibility of dementia syndrome. Motor severity, age, EDSS and disease time are risk factors for cognitive worsening and for possible dementia syndrome. Emphasis in the find of half the patients diagnosed with dementia were also diagnosed with bipolar disorder.

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Dementia syndrome secondary to postherpetic encephalitis: a sequelae lesion

Geovanna Barros Rocha[1], Fredson Lucas de Almeida Soares[1], Denis Antonio Nascimento da Costa[1], Matheus Prado dos Santos[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Geovanna Barros Rocha; Email: geovannarocha@outlook.com.

Case presentation: M.R.S, female, 65 years old, mixed race, retired teacher, completed higher education, is hypertensive, has type 2 diabetes mellitus, using Metformin 850 mg, Losartan 50 mg, Escitalopram 10 mg. The patient reports that around 1 year ago she began to experience frequent forgetfulness, difficulty remembering recent events and changes in behavior, including episodes of irritability and social disinhibition. These symptoms began approximately 6 months ago, after being diagnosed with herpetic encephalitis treated with Acyclovir. She presents cognitive and behavioral symptoms: difficulty forming new memories and remembering recent past events; Episodes of disorientation in time and space; Difficulty finding words and understanding language; Personality changes with irritability, social disinhibition and inappropriate behavior. On physical examination, she appeared in a regular general condition, conscious, but with episodes of confusion, oriented in person, but disoriented in time and space. Cognitive Tests: Minimental 12/30; Verbal fluency 7; Clock test: correct circle drawing, numbers in incorrect position, does not include the 12 numbers and hands in wrong positions. **Discussion:** This is a unique case of dementia secondary to herpetic encephalitis, given the rarity of herpetic encephalitis and its high mortality (70%). The clinical picture of an adult with this dementia syndrome is quite variable, which includes a combination of neurological, cognitive and behavioral symptoms. Herpetic encephalitis, caused by HSV-1, can result in significant damage to the central nervous system, especially the temporal lobe and hippocampus, areas critical for memory and cognition. As a consequence, the patient usually presents with an acute onset of neurological symptoms followed by a gradual development of cognitive and behavioral impairments. **Final comments:** The patient presented serious behavioral changes, which may be the result of the damage resulting from herpetic infection in the brain. The case represented a diagnostic challenge, due to the absence of changes in imaging studies and due to the rarity and complexity of the syndrome, as well as the lack of support in the literature. The patient remains under outpatient follow-up, with progressive cognitive and behavioral improvement. It is important that more studies are carried out in order to provide greater access to information about this syndrome, as well as elucidate the diagnosis and appropriate management of affected patients.

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Demographic, genetic and clinical profile of patients with hereditary transthyretin amyloidosis from a reference center in Brazil

Caroline Bittar Braune[1], Fábio de Souza[2], Cibele Franz[2], Kelma Macedo Pohlmann Simões[3], Yuri Vianna Fernandes da Silva[2], Ana Clara Farias de Negri[2], Regina Maria Papais Alvarenga[2,4]

[1]Universidade Federal do Estado do Rio de Janeiro, Hospital Gaffrée e Guinle, Departamento de Neurologia, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Faculdade de Medicina, Rio de Janeiro RJ, Brazil.
[3]Universidade Federal do Estado do Rio de Janeiro, Hospital Gaffrée e Guinle, Departamento de Oftalmologia, Rio de Janeiro RJ, Brazil.
[4]Universidade de Vassouras, Vassouras RJ, Brazil.

Correspondence: Caroline Bittar Braune; Email: caroline.bittar@gmail.com.

Background: Hereditary transthyretin amyloidosis (ATTRv) is a rare, progressive, and fatal disease caused by pathogenic variants in the transthyretin (TTR) gene. TTR is a protein mainly produced in the liver that circulates as a stable tetramer involved in the transport of vitamin A and thyroxine. Pathogenic TTR variants dissociate into unstable monomers, that misfold to form amyloid deposits, which ultimately accumulate and damage multiple

tissues, including nerves, heart, gastrointestinal tract, and musculoskeletal structures. ATTRv amyloidosis is thus a multisystem disease with phenotypic heterogeneity depending on factors such as geographic location, underlying mutation, age of onset of symptoms, among others. Until recently, liver transplantation was the only therapeutic option, but in the last decades there has been a notable increase in the range of disease-modifying treatment strategies. **Objective:** Brazil is an endemic country for ATTRv amyloidosis, but data on the disease are limited to a few reference centers. The present study aims to describe the demographic, genetic and clinical characteristics of patients with ATTRv amyloidosis followed at a reference center in the southeastern region. **Methods:** This was a cross-sectional evaluation, with descriptive analysis, including patients with pathogenic variants in the TTR gene. All patients were evaluated by a neurologist and cardiologist. Data regarding demographic aspects, genotype and phenotype were collected from medical records. **Results:** A total of 78 patients were evaluated, 33 (42%) of whom were male, with a median age of 56 years (IQR 45-66). Genotypes Val30Met, Val122Ile and Ala39Asp occurred in 50 (64%), 26 (33%), and 2 (3%), respectively. 54% of patients were symptomatic, with a median age at symptom onset of 50 years (IQR 37-62). 29% had undergone liver transplantation, 20 (48%) had pure neurological phenotype, 1 (2%) had pure cardiac phenotype, and 20 (48%) had a mixed phenotype. Neuropathic impairment score (NIS) was available in 39 patients, with a median score of 34 (IQR 8-63). 90% of patients had dysautonomia, and the genotypes without dysautonomia (n=4) were 3 Val122Ile and 1 Ala39Asp. 78% of symptomatic patients were in Coutinho stage I, and 22% were in stages II (17%) or III (5%). **Conclusion:** In the present study, we present a large cohort of patients with ATTRv amyloidosis. Val30Met was the predominant mutation, consistent with that described in Brazil, and a rare mutation (Ala39Asp) also occurred. Most patients exhibit a mixed phenotype of polyneuropathy and cardiomyopathy, including Val122Ile patients, and NIS demonstrated relevant neuropathy in symptomatic patients. 90% of patients had dysautonomia, a known red flag for this disease, and most were in Coutinho stage I. Despite this, 22% were in stages II and III, indicating that we need to increase awareness about the disease even in our endemic country, since many patients still present in more advanced stages.

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Demyelinating impairment in leprosy and the importance of the differential diagnosis with inflammatory neuropathies

Iago Resende Carvalho[1], Isabella Sabião Borges[1], Pedro Henrique Sirotheau Corrêa Alves[1], Fernanda de Oliveira Cirino[1], Douglas Eulálio Antunes[1], Raquel Campos Pereira[1], Marcus Vinicius Magno Gonçalves[2], Isabela Maria Bernardes Goulart[1], Diogo Fernandes dos Santos[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.
[2]Universidade da Região de Joinville, Joinville SC, Brazil.

Correspondence: Iago Resende Carvalho; Email: iagorcarvalho42@gmail.com.

Background: Leprosy neuropathy (LN) is a chronic condition that starts as an infection of the Schwann cells by the bacilli Mycobacterium leprae, that may evolve into a demyelinating neuropathy that worsens during leprosy reactions (LR), with patterns not being clear, as both demyelination and axonal degeneration are common. Type-1 LR (T1LR) occurs in patients with active cell-mediated immune response against M. leprae and Type-2 LR (T2LR) occurs in multibacillary cases. Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an acquired immune-mediated neuropathy, which can be typical or present as atypical variants, such as multifocal acquired demyelinating sensory and motor neuropathy (MADSAM). In some situations, although the patients fill the neurophysiological and clinical criteria for CIDP, the differential diagnosis of LN should be considered. **Objective:** This study aimed to describe how to interpret the demyelinating impairment in LN and verify if those patients fulfilled the criteria for CIDP. **Methods:** Retrospective observational analysis of patients with leprosy in a National Reference Center in Brazil between 2014-2023. 117 patients underwent neurophysiological examinations that showed a demyelinating pattern defined by prolonged distal motor latency, reduction in motor conduction velocity, and presence of conduction block (CB) and/or temporal dispersion (TD). All cases were analyzed according to the European Academy of Neurology/Peripheral Nerve Society (EAN/PNS) to establish a differential diagnosis of CIDP with neurophysiological and clinical criteria. **Results:** 82.0% had CB and 96.6% TD. 78.6% had both. 46.1% had T1LR and 17.9% T2LR. 81.3% of the patients with LR and 83.3% without LR had CB (p=0.78). 96.0% of LR group and 97.6% in the group without LR had TD (p=0.64). 83.3% of T1LR and 76,2% of T2LR had CB (p=0.47), and 94.4% of T1R and 100.0% of T2LR had TD (p=0.27). 92.3% (108/117) of patients had NCS compatible with CIDP, but none met the criteria for clinical CIDP, as none had proximal muscle weakness of limbs, nor absent or reduced tendon reflexes. On the other hand, 46.3% (50/108) of the patients with compatible NCS met clinical criteria for atypical CIDP, notably MADSAM. **Conclusion:** Leprosy is a spectral disease that may present different patterns of demyelination and axonal damage. The patterns observed in ENMG observed in the LN patients of our study are also observed in other inflammatory neuropathies, especially typical and atypical forms of CIDP. Therefore, an isolated analysis of ENMG does not allow for a correct differential between both conditions. This shows that the diagnosis of LN might be delayed with an incorrect diagnosis of CIDP, especially in developed countries. Hence, a careful clinical evaluation and detailed neurological exam are essential to establish the diagnosis of diseases with similar neurophysiological patterns.

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Dengue encephalitis and the differential diagnosis of bilateral thalamic lesions

João Márcio Borgue de Oliveira[1], Itamar Meireles Andrade Santos[1], Caroline de Barros Viegas[1], Jerônimo Moda[1], Renan Macionil Cotrim[1], Camila Bonfanti Baima[1]

[1]Universidade Estadual Paulista, Faculdade de Medicina de Botucatu, Hospital das Clínicas, Botucatu SP, da Brazil.

Correspondence: João Márcio Borgue de Oliveira; Email: jmarcio.bo.md@gmail.com.

Case presentation: Woman, 20 years old, with attention-deficit/hyperactivity disorder, hospitalized due to encephalopathy, with fever, headache, asthenia, and myalgia that started 3 days ago. She appeared drowsy, disoriented, with reduced verbal fluency, partially understanding commands, with dysarthria, dysphagia, incomplete and disproportionate hemiparesis with brachial predominance on the left, Babinsk sign on the left, bilateral palmomental reflex, absent meningeal signs, and normal fundoscopy. Acyclovir was started due to the possibility of herpetic encephalitis. The serum NS1 antigen test was positive. Computed tomography (CT) of the brain with bilateral hypodensity in the thalamus and posterior arm of the internal capsule on the right. Arterial and venous CT angiography without occlusions, stenoses, or other signs of vasculopathy. Cerebrospinal fluid (CSF) with hyperproteinorrachia (68mg/dL) was maintained during evolution (60mg/dL). Electroencephalogram with baseline activity level disturbance. Magnetic resonance imaging of the brain with hyposignal on T1 and hypersignal on T2/FLAIR in the thalamus, posterior arm of the internal capsule, and clear globe on the right, without restriction to diffusion with heterogeneous gadolinium impregnation and intervening microhemorrhages. Acyclovir was suspended after findings not suggestive of herpetic encephalitis, with a negative viral panel in CSF for HSV-1, HSV-2, EBV, VZV, and CMV and a negative IgM for dengue. Throughout hospitalization, the patient progressed with improvement of encephalopathy, dysarthria, and dysphagia, being discharged after 14 days, maintaining temporal disorientation, and partial improvement in hemiparesis. **Discussion:** Patient with dengue in the acute phase, with criteria for probable encephalitis (change in mental status, fever, new focal sign, and parenchymal changes on imaging examination), with neuroimaging findings suggestive of arbovirus encephalitis, with other causes of viral encephalitis being excluded, and although the absence of IgM for dengue in the liquor has low sensitivity for diagnosis, Given the acute evolution and bilateral thalamic involvement, it is important to consider the following differential diagnoses: Stroke due to occlusion of Percheron's arterie–bilateral paramedian thalamic infarction with or without involvement of the midbrain and anterior thalamus; deep cerebral vein thrombosis (CVT); acute disseminated encephalomyelitis (ADEM) – generally start after remission of the febrile period with predominantly white matter involvement. **Final comments:** The diagnosis of dengue encephalitis is based on criteria for encephalitis, suspicion of dengue, laboratory confirmation of systemic infection, neuroimaging changes with or without CSF changes, and the exclusion of other etiological agents for the encephalitis. Furthermore, the presence of bilateral thalamic lesions and acute evolution makes it necessary to exclude stroke, CVT, and ADEM.

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Dengue encephalitis: a reality in an endemic country

André Luiz Pereira Martins[1], Olga Cassol Silva[1], Joana Laurindo da Silva[1], Mariana Costa Borges[1], Maria Eduarda Andreatta[1], Carolina Aparecida Donofrio Santos Costa[1], Gabriela da Silva Cremonese[1], Cristine Koehler[1], Rafael Marques Mendes[1], Gladys Lentz Martins[1]

[1]Hospital Governador Celso Ramos, Florianópolis SC, Brazil.

Correspondence: André Luiz Pereira Martins; Email: martinsandrelp@gmail.com.

Case presentation: A eighty year-old woman was transferred to the emergency room in a tertiary hospital at Florianópolis (Santa Catarina, Brazil), due to the report that she woke up with tonic bilateral arms flexion and global aphasia. She had no neurological deficits the night before. As comorbidities, only hypertension and diabetes (either well controlled) and no limitations on daily activities (modified Rankin scale of zero). In the last three days, the patient presented general malaise, myalgia, fever and retro-orbital headache. Evaluated by a neurologist in the emergency, presence of mental confusion and aphasia, without focal neurological deficits. Entry laboratory tests showed leukopenia and thrombocytopenia, without other changes. An imaging investigation was performed with tomography and angiotomography of the skull and neck, with no changes. The patient evolved with lowering of the sensorium. CSF examination showed mild proteinorrhaquia, low cellularity, normal glucose; HSV and CMV investigation, bacterioscopy and bacterial culture were negative. MRI of the skull was unremarkable. Because of the dengue endemic, associated with clinic symptoms, leukopenia and thrombocytopenia, NS1 for dengue was requested; and an electroencephalogram (EEG) investigation was carried out due the altered mental status plus neurological symptoms. Laboratory of NS1 was positive; EEG showed basic activity with severe disorganization and periodic lateralized discharges with greater right fronto-temporal projection - these findings are associated with non-specific diffuse encephalopathies and non-convulsive status epilepticus; furthermore, periodic lateralized

discharge may be associated with encephalitis of infectious etiology, such as viral. A loading and maintenance dose of levetiracetam was administered. The patient progresses with gradual improvement and is discharged with her neurological status recovered. **Discussion:** According to data from the Ministry of Health's Arbovirus Panel, Brazil recorded, in the first half of this year, an unprecedented number of more than 5 million cases (confirmed and probable) of dengue. The endemic - and epidemic - situation is constant in our territory. Neurological manifestations are increasingly diagnosed in patients with dengue infection. In an Asian study, neurological complications occurred in 0.5% – 5.4% of confirmed dengue cases; in other studies, up to 20% of encephalitis cases presented in hospitals were diagnosed with dengue fever. **Final comments:** Dengue infection emerges every year in Brazil, as well as mortality and serious illnesses caused by the disease. Therefore, it is necessary to share experiences of the different forms of neurological presentations of dengue infection, given the current lack of defined diagnostic criteria. Consequently, attention will be given to assessing patients in a holistic manner, correlating clinical and epidemiology data, obtaining the best care for neurological patients.

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Dengue fever and neurological complications: a report of acute disseminated encephalomyelitis (ADEM) with good response to plasmapheresis

Bruna Beppler[1], Adria Rodrigues da Silva[1], Caroline Zapelini[1], Lia Maki Hatisuka Imai[1], Luis Sidônio Teixeira da Silva[1], Lúcio Baena de Melo[1], Priscila Audibert Nader[1], Maria Caroline Martins de Araújo[1], Jéssica Camila Fizinus[1], Lívia Souza Primo[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Bruna Beppler; Email: brunabeppler@hotmail.com.

Case presentation: A 19-year-old previously healthy male was admitted to a tertiary hospital due to symptoms that began 10 days prior with fever, myalgia, and loss of appetite. Subsequently, he developed decreased level of consciousness, spastic tetraparesis, paresthesia of the lower limbs, and urinary retention. On neurological examination, the patient was lethargic and minimally responsive, with grade 0 muscle strength in the lower limbs, grade III in the upper limbs, normal global reflexes, and no cranial nerve abnormalities. Dengue IgM serology was positive. Cerebrospinal fluid (CSF) analysis showed 1013 cells (56% lymphocytes), glucose level of 56, and protein level of 223. Oligoclonal bands were absent. Viral panel in CSF was negative. The anti-aquaporin (AQP4) serum test was non-reactive Neuroaxis magnetic resonance imaging revealed bilateral and symmetrical T2 hyperintensity in the corticospinal tracts, splenium of the corpus callosum, brainstem, cerebellar peduncles, and dentate nuclei. The spinal cord was continuously affected from the cervical to T11 levels. Intravenous methylprednisolone 1g/day was initiated on admission and continued for 5 days. Due to lack of clinical response to intravenous methylprednisolone, plasmapheresis was performed. After 5 sessions, the patient was discharged with complete improvement of tetraparesis and consciousness level, although still experiencing urinary retention requiring intermittent catheterization. Follow-up showed no neurological progression or new lesions on neuroimaging. **Discussion:** Dengue fever is the most common arboviral disease affecting humans. Neurological manifestations, although rare, are mostly severe. Acute disseminated encephalomyelitis (ADEM) is one of the described complications, characterized by a rare, monophasic, inflammatory-demyelinating condition affecting the central nervous system. Typically triggered by an infection or prior immunization, clinical features may include fever, headache, altered consciousness, optic neuritis, and focal neurological deficits. Neuroimaging in the acute phase reveals diffuse, extensive lesions primarily involving white matter. Differential diagnosis from Multiple Sclerosis can be challenging initially. Treatment options include high-dose corticosteroids, Human Immunoglobulin, and in certain cases, plasmapheresis. **Final comments:** This case illustrates a severe presentation of ADEM following dengue fever, with favorable outcomes following plasmapheresis after inadequate response to intravenous methylprednisolone. It highlights the importance of aggressive treatment in a potentially disabling and life-threatening condition.

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Dengue meningoencephalitis: a case of neurological manifestations and diagnostic challenges in pediatrics

Sandra Souza Meirelles[1], Luiza de Azevedo Freitas Giles[2], Thalita Freitas Machado[1], Thiely Guedes Giles[1]

[1]Centro Universitário do Espírito Santo, Colatina ES, Brazil.
[2]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Luiza de Azevedo Freitas Giles; Email: luiza.azevedo.freitas@gmail.com.

Case presentation: A 12-year-old female patient presented to the emergency department with a one-day history of fever and myalgia, which had progressed to incoherent speech and disorientation and retrograde amnesia. On physical examination: fair general condition, alternating between lucidity and espatiotemporal disorientation, afebrile, and without meningeal signs. Considering the hypothesis of meningoencephalitis, she was admitted to the pediatric ward and treated with acyclovir and ceftriaxone. Blood tests showed thrombocytopenia and leukopenia, leading to a positive NS1 test for dengue virus. Cranial computed tomography (CT) showed no abnormalities, and cerebrospinal fluid (CSF) was clear, with 2 leukocytes, 40 mg/dL glucose, 48 mg/dL proteins, and a non-reactive VDRL. Blood cultures were negative, and ceftriaxone was discontinued. During the course of hospitalization, the patient exhibited psychotic episodes, febrile responses, cutaneous rash, nausea, and headache. By the 5th day, cranial MRI showed bilateral and symmetrical hypersignal in the anteromedial portions of the thalami. CSF analysis confirmed Dengue via RT-PCR (serotype DEN-1) by the 10th day, with no additional microbial evidence. The clinical symptoms improved slightly by the 5th day, and she was considered fit for discharge on the 10th day, with outpatient follow-up with a pediatric neurologist. Post-discharge, the patient reported a satisfactory condition with no significant impacts, although family members noted headaches and impulsivity as potential sequelae following dengue meningoencephalitis. **Discussion:** Dengue virus, a Flaviviridae family arbovirus, is generally considered non-neurotropic. However, in recent years, the incidence of neurological manifestations in severe forms of the disease has increased, occurring in 0.5 to 21% of cases. In dengue meningoencephalitis CT may reveal hyperdense parenchymal foci and hypodensities in the thalamus and basal ganglia, while MRI is more accurate, showing hypersignal on T2-weighted images. In this case, cranial CT was unremarkable, but MRI showed thalamic hypersignal. There is still no specific treatment for dengue meningoencephalitis, supportive care is standard. Corticosteroids, antibiotics, and antivirals lack proven efficacy but are often used empirically, as in this case. **Final comments:** This case highlights the complexity and severity of dengue's neurological manifestations. A comprehensive diagnostic approach is crucial for early and accurate clinical management. MRI proved superior to CT, emphasizing its role in evaluating suspected neurological complications of dengue virus. The patient's gradual improvement suggests a favorable prognosis, but neurological sequelae indicates the need for long-term follow-up. This case emphasizes the importance of recognizing neurological manifestations of dengue virus, even if rare, and conducting thorough investigations to ensure accurate diagnosis and appropriate treatment.

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Dengue virus-induced encephalitis leading to new-onset refractory status epilepticus

Valmir Vicente Filho[1], Caroline Link[1], Isabelle Pastor Bandeira[1], Camila Emi Fujiwara Murakami[1], Marlon Wycliff Caeira, Helio Afonso Ghizoni Teive[1], Cláudia Suemi Kamoi Kay[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Valmir Vicente Filho; Email: valmirvicentef@gmail.com.

Male, 74 years, with functional status, previous hypertension, hypothyroidism, ex-smoker. The initial symptoms were anorexia, diarrhea, vomiting, headache, and fever. Three days post onset, the patient experienced a decline in consciousness level, right lower limb contraction, and subsequent bilateral tonic-clonic seizures. At the Emergency Care Unit, a positive NS1 serum test confirmed dengue virus infection. The patient was transferred to a reference hospital for advanced management due to the necessity of ventilatory support for airway protection caused by altered consciousness. Serial electroencephalography (EEG) studies in the intensive care unit (ICU) revealed severe diffuse cerebral dysfunction, predominantly in the left temporal region, leading to refractory status epilepticus unresponsive to initial phenytoin infusion. The condition required loading doses of phenobarbital and ketamine for seizure control. Magnetic resonance imaging (MRI) of the brain did not show acute changes, aside from mild diffuse cortical atrophy. Cerebrospinal fluid (CSF) analysis showed normal cellularity but elevated protein levels. Type 3 oligoclonal bands were identified. RT-PCR for dengue virus in the CSF was negative, but dengue IgM antigen was confirmed at serum. The patient received a 5-day course of methylprednisolone and intravenous human immunoglobulin. His level of consciousness improved. During hospitalization, he developed ventilator-associated pneumonia and Clostridioides difficile diarrhea. With motor and speech rehabilitation, the patient was discharged with a patent airway, no stomas, ambulatory without assistance, and without cognitive sequelae. Dengue encephalitis can manifest as altered levels of consciousness or personality, seizures, focal neurological signs, reactive IgM dengue antibody, NS1 antigen, or positive dengue PCR in serum and/or CSF, and the exclusion of other causes of viral encephalitis or encephalopathy. This case exemplifies new-onset refractory status epilepticus (NORSE) associated with dengue encephalitis.

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Description of a cohort of patients with Baló concentric sclerosis from Rio de Janeiro/Brazil

Vanderson Carvalho Neri[1], Marina Papais Alvarenga[1], Marcos Papais Alvarenga[2], Claudia Vasconcelos[3], Diana Goes[3], Regina Papais Alvarenga[3]

[1]Hospital da Lagoa, Neurologia, Rio de Janeiro RJ, Brazil.
[2]Clinica NeuroRio, Rio de Janeiro RJ, Brazil.
[3]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Vanderson Carvalho Neri; Email: vandersoncn@yahoo.com.br.

Background: Baló Concentric Sclerosis(BCS) is a rare demyelinating disease of the central nervous system characterized pathologically by large alternating lamellae of demyelinated and myelinated white matter arranged in a concentric pattern. BCS is considered an unusual variant of multiple sclerosis (MS) by some authors. **Objective:** To describe the clinical and neuroimaging characteristics of patients in the State of Rio de Janeiro/Brazil. Define the pattern of neurological involvement that matches differential diagnoses with other demyelinating diseases, cerebrovascular diseases, and brain tumors. **Methods:** A case series with an anti-mortem diagnosis of BCS performed by magnetic resonance imaging between 1985 and 2023 was selected from the base date of a public hospital (Hospital Federal da Lagoa), a University hospital (Gaffrée-Guinle/Universidade Federal do Estado do Rio de Janeiro), and a private clinic (NeuroRio). **Results:** Twenty cases of BCS were selected from a sample of 1,500 patients diagnosed with the inflammatory demyelinating disease. 14 women and 6 men were evaluated; the Average age: was 32.05. 6 cases (30%) had a monophasic course, 14 (70%) had a recurrent course 5 cases (20%) had manifestations during childhood. Monophasic cases: 100% presented symptoms acutely - symptoms: hemiparesis/hemiparesthesia: 6 (100%), dysarthria: 2 (33.3%), language disorder: 2 (33.3%), hemianopsia: 1 (16.7%); 3 patients (50%) showed complete improvement, 3 (50%) maintained neurological sequelae. Recurrent cases: 13 (92.9%) with acute manifestation, 1 (7.1%) progressive symptoms. Outbreaks: mean 3 (min.2, max. 17); Symptoms: hemiparesis/hemiparesthesia: 7 (50%), dysarthria: 1 (7.1%), language disorder: 1(7.1%), hemianopsia: 1 (7.1%); optic neuritis: 1 (7.1%), seizure: 1 (7.1%), 1 (7.1%) chorea/athetosis. 11 (78.6%) cases showed complete improvement, 2 (14.2%) maintained sequelae, and 1 (7.1%) died. One case of NMO-like with AQP4+ and two pseudotumoral/ADEM-like has been described. Neuroimaging: 20 (100%) had a characteristic lesion of BCS in the periventricular white matter, 5 (25%) had a lesion also outside the brain: 5 cases had a lesion in the brainstem, and one case in spinal cord. **Conclusion:** Characteristic lesions were predominant in the periventricular white matter. Symptoms related to long pathways, ascending and descending, were the predominant symptoms. 70% of cases had a recurrence of symptoms. 78% of recurrent cases and 50% of monophasic cases had complete recovery of symptoms, with no neurological sequelae. Therefore, it can be said that BCS is not a variant of MS.

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Description of prevalence of hospitalizations for brain neoplasms and the mortality among Brazilian federative regions

Guilherme Galardino Santos[1], Bernardo Caetano Novaes[1], Victor Bruno dos Santos Teixeira[1], Samuel José Feres Ozava[1], Heloisa Magda Resende[1]

[1]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.

Correspondence: Guilherme Galardino Santos; Email: guilhermegalardino@gmail.com.

Background: Brain tumors are neoplasms originating from various cells of the central nervous system (CNS) or from metastases of other primary cancers. The global incidence is low, around 3 cases per 100,000 people, with 11,000 new cases annually in Brazil. Although rare, they are highly incapacitating and have high morbidity and mortality, with a 5-year survival rate of 37%. **Objective:** To calculate the prevalence of hospitalizations due to brain neoplasms (primary and metastatic lesions) in the federative regions of Brazil and to compare these prevalences with the mortality rates in these regions over the last 5 years. **Methods:** The methodology of this study involved data collection from the Hospital Information System (SIH) from January 2019 to December 2023. To calculate prevalence, we used population data from the 2022 census by IBGE. Mortality rates were also obtained from the SIH for the same period. All data were based on the place of residence and year of service. **Results:** From 2019 to 2023, there were 75,799 hospitalizations due to brain neoplasms in Brazil. The federative region with the highest hospitalization prevalence (per 100,000 inhabitants) was the South (58.68), followed by the Central-West (40.09), Southeast (37.5), Northeast (29.66), and North (21.22) regions. Although the highest prevalence of cases was concentrated in the South, the highest annual mortality rate was in the North (15.32), followed by the Southeast (14.82), Northeast (12.88), South (12.25), and Central-West (12.11). The average mortality rate for hospitalizations due to brain neoplasms in Brazil was 13.60. **Conclusion:** Brain tumors, while rare, represent a significant public health challenge in Brazil due to their high morbidity and mortality. Between 2019 and 2023, there were 75,799 hospitalizations for brain neoplasms, with the South region having the highest prevalence (58.68 per 100,000 inhabitants) and the North region the lowest (21.22 per 100,000 inhabitants). However, the highest mortality rate was observed in the North (15.32). The national average mortality rate was 13.60, highlighting the need for interventions to improve the diagnosis and treatment of brain tumors, especially in regions with higher mortality, in addition to enhancing healthcare infrastructure.

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Description of the prevalence of epilepsy hospitalizations in the federative regions of Brazil and the mortality rate

Bernardo Caetano Novaes[1], Huyla Pereira de Almeida Lima[1], Lara Valentim Vale[1], Fabiana Aparecida Eller[1], Elder Machado Sarmento[1]

[1]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.

Correspondence: Bernardo Caetano Novaes; Email: bernardo16.novaes@gmail.com

Background: Epilepsy is a neurological disorder characterized by recurrent seizures caused by abnormal neuronal discharges in the brain. The global prevalence is estimated at 621.5 cases per 100,000 inhabitants, including primary and secondary cases. Although treatable, many patients achieve remission. However, epilepsy can have an unfavorable prognosis, leading to death, especially in middle- and low-income countries. **Objective:** To calculate the prevalence of epilepsy hospitalizations in the federative regions of Brazil and to compare the prevalence rates with the mortality rate in these regions over the last 5 years. **Methods:** The methodology employed in this study involved data collection from the Hospital Information System (SIH) from January 2019 to December 2023, and population data from the 2022 Brazilian Institute of Geography and Statistics (IBGE) census were used to calculate the prevalence. Mortality rates were also obtained from the Hospital Information System in the same study period. **Results:** During the studied period, there were 285,241 hospitalizations for epilepsy in Brazil. The federative region with the highest prevalence (per 100,000 inhabitants) was the South (191.1), followed by the Central-West (150.65), Southeast (134.86), Northeast (133.52), and North (92.76). Despite the higher prevalence of cases in the South, the total annual processing mortality rate was highest in the Southeast (3.12), followed by the Northeast (2.88), North (2.35), Central-West (1.96), and South (1.88). **Conclusion:** Based on the data, the South region (191.1) recorded the highest number of epilepsy hospitalizations, followed by the Central-West (150.65), Southeast (134.86), Northeast (133.52), and North (92.76). However, the total annual processing mortality rate was highest in the Southeast (3.12). These regional discrepancies suggest the need for a more in-depth analysis of the factors contributing to the prevalence and outcomes of epilepsy in different areas of the country. Additionally, they highlight the importance of prevention strategies, early diagnosis, and management of epilepsy nationwide. Equally crucial is ensuring equitable access to quality healthcare across all regions.

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Description of the prevalence of migraines and other pain syndromes hospitalizations in the federative regions of brazil and the mortality rate

Bernardo Caetano Novaes[1], Davi Melo de Hollanda Cordeiro[1], Victor Bruno dos Santos Teixeira[1], Guilherme Galardino Santos[1], Samuel José Feres Ozava[1], Myllene Rodrigues Silva[1]

[1]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.

Correspondence: Bernardo Caetano Novaes; Email: bernardo16.novaes@gmail.com.

Background: Migraines and other pain syndromes are neurological disorders characterized by recurrent episodes of intense, debilitating pain. Syndromes such as tension-type headache, cluster headache, and neuropathic pain also contribute to the global burden of chronic pain. Although effective treatments exist, their efficacy varies, and many patients suffer from uncontrolled chronic pain. These syndromes and their socioeconomic implications are acute in low- and middle-income countries, where access to specialized care is limited. Studies using the International Headache Society (IHS) criteria indicate that 15-18% of women and 6% of men are affected by migraines, primarily between ages 25 and 55. Migraine varies in severity and disability and is underdiagnosed worldwide. **Objective:** To investigate the prevalence of hospitalizations for migraine and headache syndromes in different regions of Brazil and to correlate these prevalences with regional mortality rates over the past five years. **Methods:** This study was based on data collected from the Hospital Information System (SIH) between January 2019 and December 2023. Population data from the 2022 IBGE census were used to calculate prevalences. Mortality rates for the same period were obtained from the SIH. **Results:** During the period from January 2019 to December 2023, there were 51,269 hospitalizations for migraine and other headache syndromes in Brazil. The highest prevalence was observed in the South region, with 44.23 cases per 100,000 inhabitants, followed by the Northeast (28.98), North (24.52), Central-West (18.21), and Southeast (17.65). The highest mortality rate was recorded in the North region (2.89), followed by the Northeast (1.61), Southeast (0.4), Central-West and South (0.28). The average mortality rate in Brazil is approximately 1.12. **Conclusion:** Headaches are among the most common medical complaints, significantly impacting people's professional lives. This study investigated the prevalence of hospitalizations for migraine and headache syndromes in Brazil, correlating these with regional mortality rates over the past five years. From 2019 to 2023, there were 51,269 hospitalizations. The South region had the highest prevalence (44.23 cases per 100,000 inhabitants) and the lowest mortality rate (0.28 per 100,000). In contrast, the North and Northeast regions had significantly higher mortality rates (2.89 and 1.61 per 100,000, respectively). Although the overall mortality rate is relatively low, it is crucial to investigate the factors contributing to these regional disparities, both to understand the high mortality in the North and Northeast and to explore why the South region, with the highest prevalence, has the lowest mortality rate.

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Descriptive analysis of cerebrovascular disease patients at a public hospital in São Paulo - Ipiranga Hospital

Vitória Karoline Justino dos Santos[1], Gabriela Simões Noel da Silva[1], Isabella Guimarães Silva Alípio[1], Rafael Alves Banzati Viana[1], Eduardo Lengruber Oliveira[1], Levy Werneck de Almeida Menezes[1], Vitor Henrique Justino dos Santos[2], Beatriz Azevedo Godke Veiga[1]

[1]Hospital Ipiranga, São Paulo SP, Brazil.
[2]Universidade Anhembi Morumbi, São Jose dos Campos SP, Brazil.

Correspondence: Vitória Karoline Justino dos Santos; Email: vitoriakj25@gmail.com.

Background: Cerebrovascular Disease (CVD) patients have a high prevalence of hypertension (70-80%) and diabetes (30%), particularly in ischemic stroke. Mortality is higher in hemorrhagic stroke (40%) due to the severity of cerebral hemorrhages. Infections such as pneumonia occur in about 30% of cases, complicating the clinical outcome and increasing patient mortality and morbidity. **Objective:** This study aims to evaluate CVD patients' epidemiology at Ipiranga Hospital. **Methods:** We selected 290 CVD patients treated at one center from January 1, 2022 to March 1, 2024. Data collected included gender, age, CVD type, comorbidities such as prior stroke, myocardial infarction, hypertension, diabetes, and whether they were alcoholics or smokers. We collected data on mortality and additional diseases during hospitalization. We described continuous variables in medians (±interquartile range) and categorical variables in percentages. We used the Mann-Whitney test for continuous variables and the chi-square test for categorical variables. The p-value considered statistically significant was <0.05. **Results:** The sample consisted of 290 patients, with 50.3% being men. Of these, 12.17% had a hemorrhagic CVD (CVDh), and 87.2% had an ischemic CVD (CVDi). The median (±IQR) age of the study group was 68 years (±17). The comorbidities frequencies were 71.38% systemic arterial hypertension (SAH), 28,61% diabetes mellitus (DM), 20% previous CVD (CVDp), and 7.23% previous myocardial infarction (MIp). The infection and mortality rates were 11.8% and 12.5%, respectively. The average National Institutes of Health Stroke Scale (NIHSS) score at admission was 6; at discharge, it was about 3 (±7 for both). **Conclusion:** This study suggests CVD patients treated at Ipiranga Hospital have a predominance of comorbidities such as hypertension and diabetes, which is consistent with the existing literature. Therefore, it is imperative to focus on stabilizing these primary conditions through primary and secondary prevention measures to reduce the incidence of such events.

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Descriptive analysis of ischemic stroke patients undergoing thrombolysis in a São Paulo metropolitan public hospital

Gabriela Simões Noel da Silva[1], Vitória Karoline Justino dos Santos[1], Isabella Guimarães Silva Alípio[1], Rafael Alves Banzati Viana[1], Eduardo Lengruber Oliveira[1], Levy Werneck de Almeida Menezes[1], Vitor Henrique Justino dos Santos[2], Beatriz Azevedo Godke Veiga[1]

[1]Hospital Ipiranga, São Paulo SP, Brazil.
[2]Universidade Anhembi Morumbi, São José dos Campos SP, Brazil.

Correspondence: Gabriela Simões Noel da Silva; Email: gabisnoel@gmail.com.

Background: Cerebrovascular disease (CVD) is the second leading cause of death in Brazil, with a mortality rate of 51.3%. Currently, in ischemic stroke cases, venous thrombolysis is an effective acute-phase treatment option that reduces mortality and is widely available through the Brazilian public health system. **Objective:** This study evaluates the epidemiological profile of CVD patients who underwent intravenous thrombolysis at a public hospital in São Paulo, and compares the prevalence of this treatment with rates from previous studies. **Methods:** Retrospective analysis of medical records from 267 CVD patients between January 2022 and March 2024. Logistic regression was used to assess factors influencing the likelihood of undergoing intravenous thrombolysis. **Results:** Among 267 ischemic CVD, 30 underwent intravenous thrombolysis (11%), one patient was excluded due to insufficient data in the medical record. Of these, 62.1% were male, with a median age of 64,5 years (±30.5). High blood pressure was present in 50%, and diabetes mellitus in 14.3%. Additionally, 68.9% reported prior exposure to smoking, of which 34.5% were active smokers; 17.2% reported alcoholism. Only one patient (1.8%) with a history of prior stroke underwent thrombolysis. Regarding etiological investigation, the majority of events had an undetermined cause (44.8%), followed by cardioembolism (27.6%), atherosclerosis (24.1%) and lacunar stroke (3.4%). Logistic regression analysis demonstrated that cardioembolic etiology was associated with a 4.3 times higher risk of undergoing thrombolysis compared to atherosclerotic etiology. The anterior circulation was affected in 72% of cases. The mean National Institutes of Health Stroke Scale (NIHSS) on admission was 13.2 (±11) and 6.56 (±9) at discharge. Each point increase in NIHSS on admission correlated with a 22% increase in the likelihood of undergoing thrombolytic therapy. Among the patients who underwent this procedure, 20% died. **Conclusion:** The profile of patients with ischemic stroke who underwent thrombolysis was predominantly composed of men in their seventh decade of life, with multiple comorbidities and a high rate of involvement of the anterior cerebral artery territory. Data analysis indicated a lower occurrence in patients with reduced NIHSS, stroke of atherosclerotic cause, or those with a history of previous stroke. Although the prevalence of thrombolysis was similar to that in some developed countries (11% of patients), such as the United States (8.8%), it is still far from the rates observed in countries like Austria (16.8%) and the Czech Republic (23.8%). Given these data, it is essential to identify potentially reversible factors, such as delays in seeking early medical care and failures in the intra-hospital process, to facilitate access to this therapeutic modality.

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Desmopressin effects in patients with antiplatelet associated intracranial haemorrhage: a systematic review and meta-analysis

João Arthur Cerqueira Taumaturgo[1], Hugo Lorran Souza Costa[2], Natalia Cinopolli Gomas[3], Anna Lydia Machado Silva[3], Artur Menegaz de Almeida[4], Ítalo Barros Andrade[5], Lilianne Rodrigues Fernandes[6], Michele Kreuz[7]

[1]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[2]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[3]Universidade Cidade de São Paulo, São Paulo SP, Brazil.
[4]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[5]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[6]Faculdade de Ciências Médicas, Abaetetuba PA, Brazil.
[7]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: João Arthur Cerqueira Taumaturgo; Email: arthurotaumaturgo@gmail.com.

Background: Desmopressin is an antidiuretic hormone used to treat central diabetes insipidus (CDI), nocturnal enuresis and bleeding disorders, enhancing platelet adherence and aggregation on the vascular endothelium. Patients on antiplatelet therapy are at an increased risk of intracranial haemorrhage. Therefore, desmopressin administration in patients with intracranial haemorrhage secondary to antiplatelet therapy is often considered. **Objective:** We aimed to perform an updated systematic review and meta-analysis assessing desmopressin safety and efficacy in treating intracranial haemorrhage in patients under antiplatelet therapy. **Methods:** MEDLINE, Scopus, and Web of Science databases were systematically searched until 17 May 2024. We considered the risk ratio and odds ratio for dichotomous outcomes and the mean difference for continuous outcomes. Data were examined using the Mantel-Haenszel method and 95% CIs. Heterogeneity was assessed using I^2 statistics. R, version 4.3.2, was used for statistical analysis. **Results:** A total of 10 studies and 7017 patients were included, of whom 882 (12%) received desmopressin and 6135 (88%) received a placebo. Only the length of hospital stay showed significant statistical results comparing both analysed groups (RR 0.56; 95% CI 0.09-1.03; p=0.019534; I^2=6%). No statistically significant difference between groups was seen regarding levels of haematoma expansion 20% (RR 0.96; 95% CI 0.44-2.13; p=0.929678; I^2=77%) and 33% (RR 0.90; 95% CI 0.48-1.71; p=0.756357; I^2=71%) and thrombotic events incidence (RR 0.66; 95% CI 0.33-1.30; p=0.227244; I^2=0%). **Conclusion:** In this updated meta-analysis, our results suggest that desmopressin administration was not associated with more beneficial outcomes in patients with intracranial haemorrhage undergoing anti-platelet treatments, as only one of the seven outcomes analysed presented a statistically significant difference tending for desmopressin administration.

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Desmopressin in hematoma expansion in patients with intracranial hemorrhage and prior use of antiplatelet agents: a systematic review and meta-analysis

Gilberto José de Melo Neto[1], Isabella Melo Soares[1], Isadora Fregonese Antunes Salomão[2], Carolina Moresi Vieira[3], Anna Luíza Soares de Oliveira Rodrigues[4], Isabela Dernandes Scabello[4], Jaqueline Simone de Oliveira Bianchi[5], Arquimedes Cavalcante Cardoso[6], Kelson James Silva de Almeida[6], Leonardo Halley Carvalho Pimentel[6]

[1]Centro Universitário Facid Wyden, Teresina PI, Brazil.
[2]Hospital Regional Unimed Missões, Departamento de Anestesiologia, Santo Ângelo RS, Brazil.
[3]Universidade Federal do Amazonas, Manaus AM, Brazil.
[4]Centro Universitário de João Pessoa, João Pessoa PB, Brazil.
[5]Universidade Municipal de São Caetano do Sul, São Caetano SP, Brazil.
[6]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Gilberto José de Melo Neto; Email: gjdemeloneto@outlook.com.

Background: Intracranial hemorrhage (ICH) is a severe condition that presents high levels of morbidity and mortality in patients who are either traumatized or suffer from poorly controlled hypertension. Its severity is notable, affecting various brain regions and requiring prompt interventions to halt its progression and prevent additional complications. However, the treatment of ICH, especially in patients on antiplatelet therapy, poses significant challenges. In this context, the hypothesis emerged that desmopressin (DDAVP), a synthetic analog of vasopressin, might have some beneficial effect on hematoma expansion (HE) in these patients. **Objective:** In light of this scenario, the present study aimed to qualitatively and quantitatively evaluate, through a systematic review and meta-analysis, the effects of desmopressin on important clinical outcomes such as HE and length of hospital stay, two variables that can significantly affect patient prognosis. **Methods:** This study was a systematic review and

meta-analysis registered via PROSPERO (registration number CRD42024525639) and conducted and presented according to the Preferred Reporting Items for Systematic Review and Meta-analysis (PRISMA) guidelines, which govern all scientific productions of this type. **Results:** Five outcomes were analyzed with the use of desmopressin, of which two showed a slight reduction with desmopressin use (HE and thromboembolic events), and three favored the control group (mortality, unfavorable neurological outcome, and length of hospital stay). However, none of these data reached significance, with p-values >0.05. Hematoma expansion: OR=0.85, CI: 0.51 to 1.42, I^2=69%; Mortality: OR=1.05, CI 0.68 to 1.60; Thromboembolic events: OR = 0.62, CI 0.29 to 1.29; Unfavorable neurological outcome: The combined OR of 1.73 (CI 0.99-3.03) suggests a trend towards increased risk of unfavorable neurological outcomes with desmopressin, but the CI is very close to not including 1, indicating this result is on the borderline of statistical significance. Heterogeneity of I^2=14%, indicating low heterogeneity among the studies. Length of hospital stay: The overall mean difference was 0.49 days in favor of the control (CI [-0.20 to 1.17]; p=0.17). Subgroup analysis among patients with ICH of traumatic or spontaneous etiology showed no difference in HE. Additionally, the epidemiological profile of ICH patients was outlined, with men being the most affected and the mean age among studies ranging from 56.5 to 77 years. **Conclusion:** The present study does not support the standardized use of desmopressin for all patients with intracranial hemorrhage. Its use should be critically evaluated based on the clinical context to determine which individuals may benefit the most from this pharmacological strategy.

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Detailed neurological examination in people living with HTLV-1

Qesya Rodrigues Ferreira[1], Ana Flávia Novaes[1], Ariana Leal Borges da Cruz[1], Carolina Souza Santana[1], Larissa Alves Fernandes[1], Jéssica Oliveira de Souza Nascimento[1], Alice Palmeira Nascimento Cerqueira[1], Luiz Guilherme Alves Pinheiro Faria[1], Fernanda Khouri Barreto[1], Davi Tanajura Costa[1]

[1]Universidade Federal da Bahia, Instituto Multidisciplinar em Saúde, Vitória da Conquista BA, Brazil.

Correspondence: Qesya Rodrigues Ferreira; Email: rodriguesqesya@gmail.com.

Background: The Human T-lymphotropic Virus Type 1 (HTLV-1) was the first retrovirus identified and it is estimated that 5 to 10 million people in the world live with the virus. HTLV-1 is the etiological agent of pathologies including HTLV-1 Associated Myelopathy/ Tropical Spastic Paraparesis (HAM/TSP), characterized as an immune-mediated disease of the nervous system. In HAM/TSP, the hip flexor and adductor muscles are particularly affected by spasticity and paresis, causing difficulties in locomotion and a reduction in quality of life. **Objective:** To characterize the neurological examination in patients living with HTLV-1 in the Brazilian hinterland, based on clinical diagnosis. **Methods:** This is an observational study that evaluated patients treated at an HTLV-1 reference center in Bahia, Brazil, between February 2023 and May 2024. Data were obtained by analyzing medical records and neurological consultations. Analyses of statistical significance were carried out with Fisher's exact test using the SPSS v. 27 program. **Results:** 110 participants were included, 82 classified as non-HAM/TSP and 28 as HAM/TSP. The mean age and standard deviation were 50.1±17.9 years, and 86 (78.2%) of the patients were female. Regarding to the neurological physical examination, 1 (1%) of patients classified as non-HAM/TSP and 11(40%) of those classified as HAM/TSP showed weakness in the upper limbs (p<0.001), while 9 (11%) of non-HAM/TSP patients and 24 (86%) of HAM/TSP patients showed weakness in the lower limbs (p<0.001). Analysis of deep tendon reflexes in the upper limbs showed a higher prevalence of exalted reflexes in the HAM/TSP group in relation to the bicipital and brachioradialis reflexes (p<0.05). Regarding the reflexes seen in the lower limbs, 21 (25.6%) patients classified as non-HAM/TSP and 18 (64.28%) classified as HAM/TSP had an adductor reflex (p=0.007). There was also a higher prevalence of exaggerated patellar and ankle jerk reflexes in the HAM/TSP group (p<0.05). As for pathological reflexes, 6 (7.31%) non-HAM/TSP patients had Babinski's sign and 6 (7.31%) had Hoffman's sign, while in the HAM/TSP group, the prevalence of such reflexes was 16 (58%) and 11 (39%)respectively (p<0.01). As for sensitivity, there was a higher prevalence of decreased vibration and pain sensitivity in the HAM/TSP group (p<0.05). There was no difference in the coordination tests performed by the two groups (p=0.159). **Conclusion:** Given the data observed, we conclude that HTLV-1 is a multisystemic virus with heterogeneous clinical manifestations. Despite the physical examination showing more significant findings in HAM/TSP patients, we observed important neurological changes in patients not classified with the disease, which could be related to an incomplete presentation or other neurological conditions. The presence of the adductor reflex appeared to be a good predictor of HAM/TSP in this study.

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Detection of latent tuberculosis infection in patients at a reference center for demyelinating diseases in Recife-PE

Bruno Paulo Teles Chaves[1], Vanessa Cristina Fragoso Cassiano Alencar[1], Marcilio Oliveira Filho[1], Nathalia Saraiva Ramos e Cruz[1], Mauricéa Novaes Costa Pereira[1], Lidiane Alves de Macedo Souza[1], Maria Iris Moraes Machado[1], Maria Lucia Brito Ferreira[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Bruno Paulo Teles Chaves; Email: brunopaulo.c@gmail.com.

Background: Latent tuberculosis infection (LTBI) is a persistent immune response triggered by Mycobacterium tuberculosis antigens without evidence of clinical manifestations of active tuberculosis. In recent decades, it is estimated that approximately 5-10% of infected patients develop the active form of the disease within the first five years after infection. Patients with multiple sclerosis (MS) and other neuroimmunological diseases who undergo treatments with immunosuppressants can develop active tuberculosis, making it necessary to carry out tests to detect LTBI before starting such treatments. **Objective:** To establish the prevalence of LTBI in a group of patients with multiple sclerosis and other neuroimmunological diseases undergoing immunosuppressive therapy at the reference center for demyelinating diseases at Hospital da Restauração. **Methods:** 139 medical records of patients who underwent a tuberculin skin test (PPD), defined as reactor when ≥5mm and non-reactor <5mm, and IGRA (interferon-gamma release assay), defined as positive or negative, were analyzed. **Results:** Of the 139 patients, 102 were female (73.4%). The average age was 46 years (range 17 and 74 years). One hundred fourteen were RRMS (82%), 17 SPMS (12.2%), 1 PPMS (2.2%), and 5 Neuromyelitis Optica Spectrum Disorder (3.6%). PPD was performed in 132 patients (95%), and 7 underwent IGRA (5%). Of those who underwent PPD, 37 were classified as reactor (26.6%) and 95 non-reactor (68.3%); of those who underwent IGRA, three were positive (2.2%) and four negative (2.9%). One patient had active tuberculosis. The prevalence of LTBI in this sample was 28,8%. **Conclusion:** We consider essential to perform LTBI detection tests in groups of patients at high risk for developing active tuberculosis. It was possible to observe a high incidence of LTBI in the initial sampling of this study, making it imperative to systematically carry out tests to detect LTBI in order to reduce morbidity and mortality in patients undergoing immunosuppressive therapies.

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Development of an instrument for the assessment of compensatory strategies in individuals with stroke

Pedro Henrique Sousa de Andrade[1], Caroline Rodrigues Osawa[1], Luciane Aparecida Pascucci Sande de Souza[1], Gustavo José Luvizutto[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Pedro Henrique Sousa de Andrade; Email: pedrohsandradeft@gmail.com.

Background: Essential motor components are lost due to the impairments caused from stroke, leading individuals to acquire new strategies to produce movement, to compensate the lack of the motor components. Since stroke is considered the main cause of chronic disability in adults, the evaluation of their compensatory strategies, mainly during the performance of daily activities (core tasks), is essential to outline a full assessment and treatment plan after stroke. **Objective:** To develop a new biomechanical analysis tool for individuals with stroke, based on the Glittre-ADL test: the Glittre - Assessment of Biomechanical Strategies (Glittre-ABS), with a scoring system that will provide an index for the evaluation of movement quality for each core task. **Methods:** This is a cross-sectional study and literature review. A literature search was performed in the main databases with terms related to sit to stand, stand to sit, gait, stair climbing, 180º turn, postural balance, adaptive strategies and stroke and their synonyms in addition to a compilation and evaluation of patient videos and expert opinions in the field of stroke rehabilitation, to produce a preliminary version of the instrument with the most common compensatory strategies used after stroke. Content validation was performed by a panel of experts to calculate the content validity coefficient (CVC), which was considered acceptable if its value was ≥0.8. **Results:** Thirty-eight studies were selected through a literature search, videos of 5 stroke patients performing the Glittre-ADL test were analyzed and opinions from 10 physiotherapists with experience in the stroke rehabilitation were collected. One table was assembled with the possible compensatory strategies adopted by stroke individuals gathered from these 3 sources of information. These strategies were sent to a panel of 10 specialists for scoring and a CVC of 0.83 for relevance and 0.75 for clarity was obtained for the sit to stand domain, 0.82 and 0.78 for the gait domain, 0.84 and 0.78 for the step up stairs domain, 0.82 and 0.80 for the step down stairs domain, 0.82 and 0.76 for the dynamic balance while standing domain, 0.81 and 0.75 for the 180º turn domain and 0.85 and 0.79 for the stand to sit domain, respectively. **Conclusion:** This data suggest that the strategies defined to compose the instrument are relevant, but their clarity are still uncertain, except for the step down stairs domain.

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Diabetes insipidus associated with infundibuloneurohypophysitis: case report

Gilmar Silveira da Silva[1], Thaise Fellini Dal Moro[1], Ana Cláudia Martins Ferreira[1], Marlon Costa Ferreira[1], Luiza Toscan[2], Matheus Felipe Kuhn Urnau[2], Wesley Alfredo Gawlinski de Arruda[1], Victor Emanuel Angeliero[1], Jéssica Vieira[1], Jéssica Maldaner Lui[1]

[1]Universidade Federal da Fronteira Sul, Hospital São Vicente de Paulo, Passo Fundo RS, Brazil. [2]Universidade de Passo Fundo, Passo Fundo RS, Brazil.

Correspondence: Gilmar Silveira da Silva; Email: silveirasilva.gilmar@gmail.com.

Case presentation: A 23-year-old patient, 30 days prior, started experiencing polydipsia, drinking approximately 20 L/day of water, in addition to nausea, vomiting, and weight loss of 17 kg. Smoker (3 pack-years) had anxiety disorder and depression as comorbidities. On physical examination, the patient presented with muscle weakness due to hypokalemia. A tomography of the skull was performed, demonstrating calcification located in the fourth ventricle, measuring 0.4 x 0.3 cm, in addition to a rounded supraclinoid lesion. MRI of the skull and sella turcica demonstrated neurohypophysis without hypersignal on T1, suggesting a relationship with diabetes insipidus, thickening of the pituitary stalk (approximately 5 mm), homogeneous contrast enhancement in the pituitary stalk and pituitary gland, suggesting infundibuloneurohypophysitis, and mild hypersignal in the corpus callosum, probably due to cytotoxic edema, suggesting CLOCS (secondary to hydroelectrolyte disorders). Laboratory tests revealed suppressed TSH (<0.1), hypernatremia (160), and hypokalemia (2.0). After correction of the hydroelectrolyte disorder, the patient recovered muscle strength. The patient was discharged from the hospital after the condition stabilized with endocrinological monitoring. **Discussion:** Infundibuloneurohypophysitis is an autoimmune and inflammatory condition caused by lymphocytic infiltration that selectively affects the neurohypophysis and pituitary infundibulum. In most cases, it manifests as osmotic symptoms, such as polyuria and polydipsia, associated with central diabetes insipidus, in the absence of signs of involvement of the anterior pituitary gland. In addition to confirming the presence of hydroelectrolyte disorders and diabetes insipidus, diagnostic investigations must include complementary tests capable of identifying commonly associated causes, such as granulomatous, xanthomatous, systemic diseases related to immunoglobulin G4, and neoplastic and autoimmune diseases. The neuroimaging of choice is magnetic resonance imaging, with typical findings being the loss of the "bright spot" in the posterior pituitary gland, associated with thickening of the infundibulum (>4 mm). The gold standard for diagnosis is transsphenoidal biopsy with an immunohistological study. Most studies suggest that spontaneous remission occurs, and the management of the condition is controversial, with no benefits demonstrated with glucocorticoids. Long-term follow-up is mandatory through serial neuroimaging examinations and hormonal assessments. Even after radiological remission of the disease, most patients remain with central diabetes insipidus, requiring prolonged desmopressin use for its management. **Final comments:** The case description above highlights the importance of additional investigation of diabetes insipidus, as it has numerous possible etiologies, with infundibuloneurohypophysitis being a commonly associated, but possibly underdiagnosed condition.

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Diagnoses of overlapping cases of neuromuscular junction disorders: myasthenia gravis and Lambert Eaton syndrome

Beatriz Carneiro Gondim Silva[1], Filipe Tupinambá Di Pace[1], Nícolas Ruan dos Santos Cavalcante[1], Eduardo de Paula Estephan[1], André Macedo Serafim da Silva[1], Rodrigo de Holanda Mendonça[1], Antonio Alberto Zambon[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Beatriz Carneiro Gondim Silva; Email: beatrizcgondim@gmail.com.

Background: Myasthenia gravis (MG) and Lambert Eaton myasthenic syndrome (LEMS) are autoimmune disorders of the neuromuscular junction. In MG the acetylcholine receptor (AChR) and the postsynaptic membrane are the targets, while in LEMS the epitope is located on the voltage-gated calcium channels, constituting a presynaptic disorder. The coexistence of LEMS-MG seems to be very rare and remains controversial. **Objective:** Aims to describe 4 cases of LEMS-MG overlap focusing on pitfalls in the workup diagnosis. **Results:** Case 1: 47-year-old woman with positive AChR antibody classic MG diagnosed in 2019 evolved with clinical remission after two years of onset. In 2022 she presented with weakness in the lower limbs and areflexia. The neurophysiological study (NPS) showed maintance of pathological decrement (> 10%) in low-frequency repetitive stimulation (already present on earlier study) and an increment greater than 100% after maximum voluntary contraction. The antibody test for voltage-gated calcium channels (anti-VGCC) was positive. She was put on prednisone and pyridostigmine, reaching minimal manifestation status. Case 2: 63-year-old man presented with a history of bulbar and upper limbs weakness, global areflexia with facilitation phenomenon. NPS showed a pathological decrement (> 10%) at low-frequency and a pathological increment (163%) at high-frequency repetitive stimulation. He had positive antibodies to AChR and anti-VGCC, compatible with LEMS-MG overlap. Case 3: 41-year-old woman with no medical history, was admitted with ventilatory insufficiency, ptosis, dysphagia, and global weakness. The investigation demonstrated a pathological increment on NPS only in repetitive stimulation at 50Hz and positive anti-VGCC, the AChR antibody was negative, leading to the diagnosis of LEMS with respiratory crisis, mimicking a myasthenic crisis. Case 4: 90-year-old woman with proximal tetraparesis, worse in the lower limbs, areflexia, hypopalesthesia, and axial ataxia. A first NPS demonstrated an axonal and demyelinating sensorimotor neuropathy and the diagnosis of chronic inflammatory demyelinating polyradiculoneuropathy was made. A second NPS was performed demonstrating low CMAP amplitudes and an increment of more than 400% in high-frequency repetitive stimulation, and a diagnosis of LEMS was subsequently made. Anti-VGCC is not yet defined. She had a good response to intravenous human immunoglobulin and prednisone. **Conclusion:** LEMS-MG association may be an underrecognized condition in clinical practice. It must be suspected not only if signs of both disorders appear simultaneously, but also if they are added to an already established disorder or if the clinical features change.

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Diagnosis and treatment of a case of functional (conversion) disorder: the importance of neurology-psychiatry collaboration

Roberto Pereira Santos[1], Bruno Miceli Gonzalez Nogueira[1], Carmem Zapata[2], Soniza Vieira Alves-leon[1], Nathalia Borges Carmo[1], Leonardo Siqueira de Araújo Pinto[1], Adriana Fiszman[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Roberto Pereira Santos; Email: r.santos.ufrj@gmail.com.

Case presentation: A 60-year-old woman complained of stiffness, pain, and weakness in her lower limbs. She also reported imbalance and difficulty finding words. Neurological examination revealed instability during standing and walking, with the patient falling in a point and support test (incongruence), but not falling with distraction (inconsistency). The shoulder tap test, highly specific for conversive postural instability, was positive (incongruence). Expressive aphasia and phonemic substitutions disappeared when the patient expressed sadness (inconsistency). The patient assumed a posture of thigh extension and painful leg with bilateral plantar flexion (dystonia). A suggestion technique called torpillage was applied: a vibrating tuning fork was placed on the patient's painful points, suggesting that the vibration "penetrated her muscles and led to their relaxation". The painful/dystonic episode was completely reversed by torpillage. **Discussion:** We present a case of functional (conversive) disorder to emphasize the importance of a multidisciplinary approach, particularly involving neurology and psychiatry. The neurologist is pivotal in making the diagnosis, as highlighted by Jon Stone, who underscores the importance of explaining incongruences and inconsistencies leading to the diagnosis. For him, explaining the diagnosis is one of the most critical aspects of treatment. Neurologists should also employ suggestion techniques both to confirm and treat conversive symptoms, particularly in acute cases. Torpillage maneuver is commonly used for these purposes. According to DSM-5 criteria, the patient presented with mixed symptoms of conversion disorder and persistent depressive disorder with obsessive-compulsive symptoms. Pharmacological treatment included paroxetine 80 mg/day alongside psychotherapy based on psychoanalytic principles. **Final comments:** Psychiatric evaluation is crucial to identify diagnoses beyond conversion disorder. It has been demonstrated that for many patients, conversion symptoms are clinical expressions of underlying psychiatric diagnoses. This appeared to be the case for the patient presented here, who received a psychiatric diagnosis of persistent depressive disorder with obsessive-compulsive symptoms. Treatment not only improved this diagnosis but also addressed the conversion symptoms effectively. This case illustrates that an integrated neurological and psychiatric approach is highly effective for diagnosing and treating conversion disorders. Specialized multidisciplinary centers are essential for managing these patients effectively.

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Diagnosis and treatment of anaplastic supratentorial cortical ependymoma in an adult patient: a case report

José Nilo Ribeiro Neto[1], Yuri Victor Barbosa Silva[1], Carolina Diniz Furtado[1], Márcio Luís Soares Ferreira[2], Sanzylly Soares França Ferreira[3], Roseliny de Morais Martins Batista[4], João Guilherme de Brito Rodrigues[1], Lael Mendonça Moraes[1], Pedro Marcelo Azevedo Barroso[1]

[1]Centro Universitário UNDB, São Luís MA, Brazil.
[2]Universidade Federal do Maranhão, São Luís MA, Brazil.
[3]Universidade Estadual do Maranhão, São Luís MA, Brazil.
[4]Universidade Ceuma, São Luís MA, Brazil.

Correspondence: José Nilo Ribeiro Neto; Email: josenilo999@gmail.com.

Case presentation: G.A.J., 54 years old, male, of mixed ethnicity, originally from and residing in the city of São Paulo (Brazil). Chief Complaint: "Fatigue in the left arm and leg" for the past 10 days. History of Present Illness: The patient reported a progressive onset of weakness 10 days prior to admission at the Primary Health Care Unit, accompanied by paresthesias in the left hemibody. This condition progressed to memory loss for recent events, behavioral changes, and mental confusion. Initially, the patient sought care at a Primary Health Care Unit and was subsequently transferred to a public hospital in São Paulo, where a cranial Computed Tomography scan was performed, revealing an intracranial expansile lesion. The patient denies weight loss, changes in bowel habits, seizures, and headaches. Physical Examination: The patient was in good general condition, eupneic, anicteric, acyanotic and normal-colored. Diagnosis: After topographic and microscopic investigation, the patient was diagnosed with anaplastic supratentorial cortical ependymoma. Treatment: As treatment, the patient underwent tumor resection. **Discussion:** Ependymomas are rare central nervous system tumors that originate from ependymal cells lining the ventricles of the brain and the central canal of the spinal cord. These tumors can occur in both children

and adults, being more common in the brain in children and in the spinal cord in adults (Rudà et al, 2022). These tumors arise from ependymal cells that line the ventricular system, the choroid plexus, the central canal of the spinal cord, and the filum terminale. However, ependymomas can also be found along the neural axis, even outside the boundaries of the ventricular system. Such entities are known as supratentorial extraventricular ependymomas, which exhibit a low incidence and an unclear etiopathogenesis (Cuoco et al, 2022). The treatment of anaplastic supratentorial cortical ependymoma involves complete surgical resection of the tumor as the primary approach. This procedure is essential to improve the patient's prognosis and minimize the chance of recurrence, followed by detailed postoperative evaluation to determine the need for additional therapies, such as radiotherapy or chemotherapy, based on the tumor's molecular characteristics and initial response to treatment (Lin et al, 2022). **Final comments:** Given the studies presented, it is evident that anaplastic supratentorial cortical ependymoma in adult patients is extremely rare. The recommended primary surgical approach is complete tumor resection, which is crucial for improving prognosis and reducing the chance of recurrence. Additionally, radiotherapy has proven effective as an adjuvant therapy, while chemotherapy, though less effective in recent diagnoses, still plays a significant role in the substantial clinical improvement of patients under 3 years of age. These combined treatments are fundamental for the safe management and recovery of patients.

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Diagnosis and treatment of myasthenia gravis in a pregnant patient with suspected head and neck tumor: case report

Matheus Jorand Cecatto[1], Roberto Pereira Santos[1], Rita de Cássia Leite Fernandes[1], Liene Duarte Silva[1], Tatiane Thais Santos Silva[1], Marco Antonio Sales Dantas de Lima[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Matheus Jorand Cecatto; Email: matheuscecatto@gmail.com.

Case presentation: A 23-year-old Black female patient presented with a progressive onset of dysphonia, dysphagia, and significant weight loss, leading to suspicion of a head and neck tumor. Further investigation at the time was inconclusive. Two years later, she developed sudden dyspnea. She underwent tracheostomy and was transferred to Hospital Universitário Clementino Fraga Filho for further investigation. Upon examination, the patient exhibited dysphonia, hypomimia, asymmetric bilateral ptosis with a positive ice pack test, Cogan's sign, curtain sign, ocular paresis on supraversion, and proximal weakness of the upper limbs. Electroneuromyography showed a decremental pattern with repetitive nerve stimulation, and Acetylcholine Receptor (AChR) antibodies were detected in the serum, confirming the diagnosis of myasthenia gravis (MG). Treatment with prednisone, pyridostigmine, and intravenous human immunoglobulin was initiated. To identify and stage a possible thymic tumor, a chest and abdomen computed tomography (CT) was requested. The patient denied any possibility of pregnancy, even if unintended. However, an intrauterine gestational sac was identified on the CT scan, and a 6-week pregnancy was confirmed by transvaginal ultrasonography. The patient showed partial improvement of symptoms after 5 days of immunoglobulin. She was discharged with outpatient follow-up by neurology and referred to high-risk prenatal care. **Discussion:** MG is an autoimmune neurological disease that affects the postsynaptic structures of the neuromuscular junction of skeletal muscles, causing weakness and fatigue in ocular, facial, bulbar, axial, and appendicular muscles. The global incidence rate of MG is 5.3 per million person-years. The most common onset age for MG in women is between 20 and 39 years, coinciding with childbearing age. The concomitant occurrence of pregnancy not only modifies the prognostic factors but also the therapeutic options, given the risk profile of adverse effects on the fetus. Up to 41% of pregnant women will experience exacerbation. There is still no consensus regarding indiscriminate pregnancy screening. **Final comments:** This case demonstrates how predominantly bulbar onset MG can mimic other conditions, delaying the diagnosis and timely treatment of the disease. It is noteworthy to consider the possible benefit of laboratory pregnancy screening in hospitalized female patients who are sexually active and of childbearing age.

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Diagnosis journey of Brazilian Friedreich's ataxia patients

Daiana Suelen Machado[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Daiana Suelen Machado; Email: daianamachado@gmail.com.

Background: Friedreich´s Ataxia (FRDA) is the most common recessive ataxia worldwide, but its diagnosis is often delayed for multiple reasons. This impacts the timely introduction of therapeutic interventions and therefore, prognosis. **Objective:** Herein, we sought to characterize the diagnostic journey of FRDA in São Paulo, the most populous state in Brazil. **Methods:** Patients were invited to answer an online survey comprising 24 to 32 questions, which was sent by physicians and the Brazilian Association of Acquired and Hereditary Ataxias via email and releases. We recorded information on age at first symptoms, age at diagnosis, first symptom observed, time since first symptoms to diagnosis, medical specialty first sought, number of physicians visited up to the diagnosis and misdiagnoses. Descriptive statistics was used to report the results - means, proportions, and standard deviations (SD). Comparison between classical (onset <25yo) and late-onset - LOFA (onset >25yo) diagnostic journey was accomplished using parametric tests. **Results:** One-hundred four FRDA patients or legal guardians answered the survey. There were 79 (75.9%) classical and 25 (24%) LOFA FRDA. Mean age at first symptoms was 17.6±9.2 years, whereas mean age at diagnosis was 24.1±11.5 years. Overall, diagnostic delay was around 7.8±6.7 years, for the whole group. There was no difference between classical and LOFA subgroups (7.6±6.8 vs 7.6±6.4 years, p=0.978). Unsteadiness was the most frequent heralding symptom (50%). Neurologists (35.8%) closely followed by orthopedical surgeons (30.8%) were the most frequent medical specialties first sought. On average, 6 different physicians were visited until diagnosis was reached. Misdiagnoses were reported by 38 (46.3%) of patients. **Conclusion:** In Brazil, FRDA is characterized by impressive diagnostic delay and frequent misdiagnoses, with no relevant differences between classical and LOFA patients.

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Diagnosis of Chediak-Higashi syndrome in the neurology ward

Crissia Pitanga Malta[1], Pedro Sanches Corrêa[1], Rodrigo da Franca Santos[1], George da Silva Divério Junior[1], Maria Eduarda Morgado da Silva[1], Diogo Matheus Terrana de Carvalho[1], Sofia Mermelstein[1], Felipe da Rocha Schmidt[1]

[1]Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.

Correspondence: Crissia Pitanga Malta; Email: crissiapitanga@hotmail.com.

Case presentation: This is a 35-year-old man, born in Rio de Janeiro, Brasil. His maternal grandparents were first cousins, and there was no known history of neurological disease in the family. Since childhood, he attracted attention due to his grayish hair color and hypopigmented skin. He also had a learning disability, leaving school in fourth grade. When he was around 20 years old, he started presenting recurrent skin infections, accompanied with progressive paresis, global areflexia, trunk and limb ataxia and visual acuity deficit. He has a 30-year-old sister, with a similar condition of skin hypopigmentation, grayish hair, recurrent bacterial lung infections, cerebellar ataxia, areflexia and amaurosis. On physical examination, what stood out was the presence of paraparesis of the lower limbs, requiring assistance from a wheelchair for mobility, bilateral amaurosis, bilateral internuclear ophthalmoplegia, global ataxia, areflexia and hypopalesthesia also in lower limbs. Magnetic resonance imaging of the skull revealed cortical atrophy, and electroneuromyography showed severe sensorimotor polyneuropathy. Presence of giant cytoplasmic granules in leukocytes was seen in a peripheral blood smear. Evaluation by ophthalmology exhibited a pale optic disc and dermatology observed diffuse hypopigmentation and grayish facial hair and capillaries. Genetic testing was carried out, disclosing a LYST gene mutation. Family screening was carried out and the 30-year-old sister, with amaurosis and ataxia, also had giant cytoplasmic inclusions. **Discussion:** This case report concerns a patient admitted to the Neurology ward of a teaching hospital in Rio de Janeiro, diagnosed with Chediak-Higashi syndrome, a rare genetic entity, with less than 500 cases reported worldwide. It is an autosomal recessive condition, which occurs due to mutations in LYST gene. The presence of giant cytoplasmic granules in leukocytes and platelets in a peripheral smear are considered pathognomonic of the disease. Clinical manifestations can range from oculocutaneous albinism, immunodeficiency with recurrent bacterial infections, coagulation defects and hemophagocytic syndrome to progressive neurological dysfunction. Regarding neurological symptoms, they often present cognitive and degenerative alterations. In childhood, they may present learning difficulties. During late infancy other symptoms appear, such as worsening of cognitive impairment, cerebellar dysfunction, weakness, neuropathy and neuronopathy, areflexia and parkinsonian signs. In imaging studies, neurodegenerative findings stand out. **Final comments:** This is a case of Chediak-Higashi syndrome diagnosed in the neurology ward, due to the patient's severe cognitive and motor impairment. Rare, difficult-to-diagnose genetic condition that presents a wide range of varied and non-specific neurological characteristics.

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Diagnosis of exclusion in progressive idiopathic ganglionopathy: a case report

Lucas Ferreira Alves[1], Ana Carolina Araujo de Oliveira[1], Leonardo Jadyr Silva Rodrigues Alves[2], Pedro Maia Nobre Rocha Saffi[2], Lorena Oliveira Carneiro[2], Ana Beatriz de Matos Berg Abrantes[2], Maria Cecília Mendes Resende[1], Ingrid Faber de Vasconcellos[2], Keila Rejane Ferreira Galvão[2], Luciano Talma Ferreira[2]

[1]Universidade de Brasília, Brasília DF, Brazil. [2]Hospital Universitário de Brasília, Brasília DF, Brazil.

Correspondence: Ana Carolina Araujo de Oliveira; Email: anacarolinaestudos08@gmail.com.

Case presentation: A 54-year-old female presented with hypoesthesia and paresthesia in the lower limbs seven months ago. Three months ago, she experienced

worsening paresthesia and hypoesthesia in the lower limbs, in addition to muscle tension, which altered her gait. She also reported pain in right thigh for the past month that appears late at night, worsens with right lateral decubitus, and subsides spontaneously after a few hours. Physical examination revealed hypoesthesia and loss of vibration sense in the lower limbs, predominantly in the left lower limb, with abolished Achilles reflex and diminished left patellar reflex. She had a high-stepping gait with a positive Romberg sign. Consequently, magnetic resonance imaging (MRI) of the cervical, thoracic, and lumbosacral spine was requested and showed no signs of compressive myelopathy. Peripheral blood tests showed negative inflammatory markers. Electroneuromyography (ENMG) of the upper and lower limbs showed sensory axonal neuropathy, with amplitudes decreased by 80% in the radial nerves, 85% in the superficial fibular nerves, 20% in the median and ulnar nerves, and the sural nerves were non-excitable. A lumbar puncture was performed, with normal cerebrospinal fluid (CSF) results. The patient was diagnosed with progressive idiopathic ganglionopathy. Three sessions of corticosteroid pulse therapy were prescribed due to the symptoms, followed by Duloxetine, Gabapentin and Citoneurin. **Discussion:** Progressive idiopathic ganglionopathy is a rare sensory neuropathy characterized by primary degeneration of the dorsal root ganglia without an identified cause. The incidence of progressive idiopathic ganglionopathies in the general population ranges from 0.2 to 0.9%, increasing to 2% in the elderly. The most common initial clinical manifestations are hypoesthesia, paresthesia, ataxia, and pain. The disease manifests slowly and gradually, which makes early diagnosis crucial for a good prognosis. The diagnosis consists in exclusion, eliminating other possible causes, such as Guillain-Barré syndrome, sarcoidosis, diabetes, or infections, and involves ENMG and quantitative sensory autonomic testing to assess the patient's thermal and electrical sensitivity. The pathophysiology of the disease involves disruption and demyelination of nerve fibers, primarily Aβ and Aδ fibers. Treatment for progressive idiopathic ganglionopathy includes corticosteroids to prevent autoimmune attacks on the myelin sheath, intravenous immunoglobulin injections, and methotrexate. **Final comments:** The prognosis varies depending on the severity of symptoms, the patient's age, and the response to treatment. Thus, the prognosis can be quite positive when symptoms are adequately controlled and remyelination is effective, or it can result in irreparable sensory losses depending on the level of nerve damage involved.

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Diagnostic and treatment pathways for adult patients with brain cancer in bahia: a comparative analysis in 2013 and 2023

Thiago Costa Oliveira[1], Antônio Felipe Rocha Brandão[1], Bruna Rios Beserra[1], Bruno Levy de Souza Amaral Maia[2], Cauã Araujo de Carvalho[2], Clara Carvalho Dantas Muniz[1], Ellen Santos Barreto[2], Fernanda Almeida da Silva de Sá Oliveira[1], Maria Clara Moysés de Siqueira[1], Cleydson Santos[3,4]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Universidade Federal da Bahia, Salvador BA, Brazil.
[3]Hospital Mater Dei Salvador, Salvador BA, Brazil.
[4]Obras Sociais de Irmã Dulce.

Correspondence: Thiago Costa Oliveira; Email: thiagocoliveira23.1@bahiana.edu.br.

Background: Primary malignant brain neoplasms are the 10th most common type of cancer in Brazil. The brain is the most frequent location, accounting for 88% of cases. Despite their low incidence, the mortality rate is high, with 84% of individuals succumbing to the disease, necessitating prompt and efficient care as mandated by the "60-Day Law," which stipulates a maximum of 60 days for treatment initiation following diagnosis. This process often requires patients to travel to cities with High Complexity Oncology Care Units (UNACON) or High Complexity Oncology Care Centers (CACON), which provide the necessary diagnosis and treatment. Bahia has 11 hospitals providing such care, with seven located in Salvador and the others in Feira de Santana, Vitória da Conquista, Ilhéus, and Itabuna, thus facilitating the treatment flow for patients within the state. **Objective:** To analyze and quantitatively compare the diagnosis and treatment flows of adult patients (aged 20 and above) diagnosed with primary malignant brain neoplasms in municipalities of Bahia in the years 2013 and 2023. **Methods:** This retrospective study was conducted using the PAINEL - Oncologia platform available on DATASUS, considering secondary data of patients diagnosed with malignant brain neoplasms residing in the state of Bahia in the years 2013 and 2023. **Results:** In 2013, there were 62 cases of this pathology recorded in 26 municipalities in Bahia, with 35 (56.45%) in Salvador and 25 (40.32%) in other regions. Regarding treatment, most occurred where the disease was confirmed: 36 (58.00%) in Salvador, 11 (17.74%) in Itabuna, 8 (12.9%) in Feira de Santana, 5 (8.00%) in Vitória da Conquista, and 1 (1.61%) each in Aracaju and Petrolina. In 2023, for the same age group, there were 196 cases of primary brain cancer recorded in 102 municipalities, with 132 (67.35%) in Salvador and 64 (32.65%) in other locations. Concerning treatment, of the 125 recorded cases, 65 (52.00%) were treated in Salvador, 25 (20.00%) in Feira de Santana, 10 (8.00%) in Itabuna, 9 (7.20%) in Vitória da Conquista, 4 (3.20%) in Porto Seguro, 4 (3.20%) in Santo Antônio de Jesus, 1 (0.80%) in Teixeira de Freitas, and 5 (4.00%) in other states. **Conclusion:** Given the high diagnostic and therapeutic complexity of primary malignant brain neoplasms, which require neurosurgical, oncological, and radiotherapeutic care, along with hospital support from a skilled multidisciplinary team and intensive care units, many patients still need to travel to reference centers, most of which are in the capital and major cities. The referral and counter-referral process can be slow, contradicting the "60-Day Law" and negatively impacting clinical outcomes, including resectability potential and overall and progression-free survival. Decentralizing and expanding specialized services to the interior could improve access, optimize clinical outcomes, and reduce the economic, physical, and psychological burden of traveling to major centers.

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Diagnostic and treatment pathways for pediatric patients with brain cancer in Bahia: a comparative analysis in 2013 and 2023

Thiago Costa Oliveira[1], Antônio Felipe Rocha Brandão[1], Bruna Rios Beserra[1], Bruno Levy de Souza Amaral Maia[2], Cauã Araujo de Carvalho[2], Clara Carvalho Dantas Muniz[1], Ellen Santos Barreto[2], Fernanda Almeida da Silva de Sá Oliveira[1], Maria Clara Moysés de Siqueira[1], Cleydson Santos[3,4]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Universidade Federal da Bahia, Salvador BA, Brazil.
[3]Hospital Mater Dei Salvador, Salvador BA, Brazil.
[4]Obras Sociais de Irmã Dulce.

Correspondence: Thiago Costa Oliveira; Email: thiagocoliveira23.1@bahiana.edu.br.

Background: Worldwide, primary malignant brain neoplasms are the fourth most common type of cancer and have the highest mortality rate among individuals up to 19 years of age. In Brazil, it is the most common solid tumor in children and adolescents. In this population, therapeutic modalities and their impact on quality of life and long-term consequences are particularly relevant. According to Law 14.238/21, which mandates special attention for children, their transportation to High Complexity Oncology Care Units (CACON) and High Complexity Oncology Care Centers (UNACON) should be optimized. Bahia has 11 referenced hospitals, seven of which are in Salvador, the only municipality with a UNACON that serves pediatric patients. The remaining four units, located in Vitória da Conquista, Itabuna, Ilhéus, and Feira de Santana, are restricted to adult patients. **Objective:** To analyze and quantitatively compare the diagnostic and treatment flows of pediatric patients (0-19 years) diagnosed with brain cancer in the municipalities of Bahia in the years 2013 and 2023. **Methods:** This retrospective study was conducted using the PAINEL - Oncology platform available on DATASUS, considering secondary data of patients diagnosed with malignant brain neoplasms residing in the state of Bahia in the years 2013 and 2023. **Results:** In 2013, 35 cases of primary malignant brain neoplasms were recorded in pediatric patients across 23 municipalities in Bahia: 31 (88.57%) in Salvador, 2 (5.71%) in Feira de Santana, and 2 (5.71%) diagnosed in Recife (PE) and Barretos (SP). Additionally, 31 patients had treatment records, with 29 (93.55%) treated in Salvador and 1 (3.23%) each in Recife and Barretos. By 2023, 31 cases were recorded in 22 municipalities: 19 (61.29%) diagnosed in the capital, 7 (22.58%) in Feira de Santana, 3 (9.68%) in Barretos, and 1 (3.23%) in Recife. Of the 28 patients treated, 20 (71.42%) were treated in Salvador, 6 (21.43%) in Feira de Santana, and 2 (7.14%) in Barretos. **Conclusion:** The data highlights the significant role of the capital city, Salvador, in diagnosing and treating pediatric patients with primary malignant central nervous system neoplasms in the state. Given the overall low incidence, it is expected that patients are directed to specialized high-complexity services with substantial expertise in managing these cases, leading to better clinical outcomes, including overall survival. Optimizing diagnosis and access to excellent services within or outside the state is crucial for achieving better results. Additionally, the decentralization of specialized care services can reduce the physical, psychological, and economic burden of patient and caregiver travel to major centers. This study contributes to a better understanding of the dynamics and challenges faced by this specific group and underscores the need for improved policies and medical training to enable rapid diagnosis and more effective treatment.

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Diagnostic challenge in neuromyelitis optica in the pediatric population: case report

Paulo Henrique Martinelli Oliveira[1], Rebeca Aranha Barbosa Sousa[1], Amanda Michelucci dos Santos[1], Larissa Miyashiro[1], Raderi Luiz Cardoso do Santos[1], Lucas Silva Dias[1], Laiz Helena de Pádua[1], Gustavo Carvalho Costa[1], Alexandre Coelho Marques[1]

[1]Hospital Municipal Dr. José de Carvalho Florence, São José dos Campos SP, Brazil.

Correspondence: Paulo Henrique Martinelli Oliveira; Email: paulohmoliveira96@gmail.com.

Case presentation: Patient F.A.P.R, 12 years old, previously healthy. Approximately 20 days after vaccination for SARS-Cov-2 (Pfizer), she developed flaccid, areflexic and asymmetric crural paraparesis (worse on the left). Over the following week, she developed tetraparesis, dysarthria, dysphagia and bilateral appendicular ataxia (worse on the left). Upon hospital admission, the hypothesis of acute polyradiculoneuritis (Guillain-Barré syndrome? Miller-Fisher syndrome?) was suggested, with intravenous immunoglobulin being administered for 5 consecutive days, without improvement in the condition. During hospitalization, the patient developed signs of pyramidal signs, with central nervous system involvement being suggested. After receiving 5 days of intravenous methylprednisolone, she showed partial improvement in symptoms. CSF assessment without significant changes. Electroneuromyography found signs of polyradiculoneuritis in a hyperacute phase, being

unable to define the classification as myelinic or axonal. Cranioencephalic resonance imaging revealed multiple areas of signal alteration compromising the calloseptal interface on the left side, bilateral thalamus, mesial aspect of the temporal lobes, hippocampi, cerebral peduncles, pons and medulla oblongata (images with the appearance of an inflammatory/demyelinating substrate). Serum evaluation revealed a reactive Anti-Aquaporin 4 IgG antibody. At the time of hospital discharge, the patient maintained tetraparesis with a predominantly spastic component, with signs of pyramidal release, being able to walk without support, in addition to oral food intake. **Discussion:** Neuromyelitis optica spectrum disorder (NMOSD) cases in the pediatric population make up approximately 3-5% of all NMOSD records. The median age range is around 14 years (7-17 years) and there is a significant female predominance (around 80% of cases). In addition to the diagnostic difficulty, excluding differential diagnoses of NMOSD is also arduous. Among the main differences are metabolic etiologies (such as late-onset biotinidase deficiency), as well as inflammatory, post-infectious and systemic nosologies (such as MOG antibody disease, sarcoidosis, flaccid myelitis and systemic lupus erythematosus). **Final comments:** Although it is a rare condition, the number of NMOSD cases in the pediatric age group has increased significantly. This finding is mainly due to the evolution and availability of diagnostic imaging methods, in addition to the possibility of serum assessment of the Anti-Aquaporin 4 antibody. However, due to the wide spectrum of clinical presentations, early diagnosis in the face of an acute evolution continues to be a great challenge.

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Diagnostic challenges in Behçet's disease: a case report with multisystemic manifestations

Vanessa Gil Humberto dos Santos[1], Amanda Melo Leite Leão[1], Laís Borneo Moreira[1], Jorge Luiz Alves[1], Pollyana Marcela Silva de Farias[1], Lais Felix Teixeira da Cunha[1]

[1]Universidade Unigranrio Afya, Duque de Caxias RJ, Brazil.

Correspondence: Vanessa Gil Humberto dos Santos; Email: vanessaterapia@hotmail.com.

Case presentation: A 29-year-old female patient from Duque de Caxias presented with two months of amenorrhea, urinary incontinence and retention, mild redness eye, oral ulcers, episcleritis, and hypotension. She denied dermatological issues, genital ulcers, pathergy, thrombosis, and cirrhosis, though she had arthritis. In 2017, she reported sudden visual loss, dysphagia, difficulty walking, and dysarthria. At the time, the patient was advised to undergo clinical follow-up with neurology. In 2014, she iniciate experiencing bilateral tremors, more prevalent at the right side, exacerbated by fine motor tasks. When asked about past history she previously presented with pleural effusion and four episodes of pneumonia at the age of 3. She was admitted for screening, including serologies and two MRIs. Initially was suspected by Wilson's disease, after that she was diagnosed with Behçet's disease. Upon receiving the diagnosis, she began treatment with three days of pulse therapy evolving with cervical hypotonia and sialorrhea with improvement of symptoms after second day of treatment. She was discharged with MRI showing a 70% reduction in inflammatory activity, for clinical follow-up discharge with prescription of azatioprine. During the pandemic, she stopped treatment for four years, recently returning the follow-up with physiotherapy and adjunctive therapies and medicine. She's presenting significant clinical improvements, such as motor skills, dysphagia, dysarthria, and tremor. **Discussion:** Behçet's disease is clinically diagnosed based on the criteria of the International Study Group for Behçet's Disease (ISGBD), requiring recurrent oral ulcers and at least two of the following: recurrent genital ulcers, ocular lesions (anterior/posterior uveitis, cells in the vitreous humor, retinal vasculitis), skin lesions (erythema nodosum, pseudofolliculitis, acneiform eruptions), and a positive pathergy test (formation of a papule or pustule after a sterile needle prick).Steps for diagnosis include a detailed clinical history investigating symptoms. Behçet's disease is a rare systemic vasculitis that requires a multidisciplinary approach due to its wide range of clinical manifestations. The diagnosis is based on specific clinical criteria and the exclusion of other pathologies. Early and accurate diagnosis is essential for effective management and improving patients' quality of life.The treatment requires a multidisciplinary approach. Pulse therapy reduced acute inflammation, and azathioprine maintained long-term symptom control. **Final comments:** This case illustrates the diagnostic and therapeutic complexity of Behçet's disease, highlighting the need for a multidisciplinary approach and regular follow-up. It contributes to the understanding and management of rare and complex conditions, emphasizing the importance of health policies that ensure continuous treatment and adequate support, promoting better prognosis and quality of life.

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Diagnostic challenges of longitudinally extensive transverse myelitis: unveiling an uncommon culprit

Johnatan Felipe Ferreira da Conceição[1], Olivia Gameiro de Souza[1], Ana Luísa Utrine Pimentel[1], Luiza de Aguiar Lima[1], Caroline Bittar Braune[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Johnatan Felipe Ferreira da Conceição; Email: johnatanfelipe@id.uff.br.

Case presentation: A 43-year-old male was admitted with a history of insidious onset of ascending paresthesia and hypoesthesia, starting in the feet. After 1 month, he noticed urinary and fecal retention, as well as difficulty walking due to weakness, along with worsening of sensory symptoms. An orthopedic evaluation established the diagnosis of a herniated disc and intramuscular corticosteroids were prescribed. He showed a significant worsening of symptoms after the administration of this medication, progressing with escalation of sensory symptoms and tetraparesis with rapid progression. He was admitted to the emergency department of an outside hospital and neuraxis magnetic resonance imaging showed a longitudinally extensive transverse myelitis between C1-C7. Cerebrospinal fluid analysis and laboratory analysis were normal. The diagnosis of probable inflammatory myelitis was established, a 5-day pulse therapy regimen with methylprednisolone was defined, resulting in new neurological deterioration, and then the patient was transferred to our university hospital and evaluated by neurology team. Upon admission, neurologic examination showed grade 3 paresis in the upper limbs and grade 1 in the lower limbs, global hyperreflexia with bilateral Babinski sign and achilles clonus, hypertonia in the lower limbs and tactile, thermal and painful hypoesthesia. Considering the etiology of vascular myelopathy, the investigation was continued with an arteriography, which showed a dural arteriovenous fistula at the bulbar-cervical junction. A successful occlusion procedure was performed on the fistula, but the patient showed little functional gain. **Discussion:** Arteriovenous fistula (AVF) is a vascular malformation characterized by anomalous communications between meningeal arteries and dural or subarachnoid veins. It can occur in intracranial or spinal cord topographies, making it the most common spinal vascular malformation. It usually affects individuals in their fifth and sixth decades of life, with a higher frequency of occurrence in the mid-thoracic and lumbar spinal cord. The pathophysiological process consists of congestion and spinal venous hypertension, leading to edema and progressive ischemia in the affected spinal cord levels. Our case illustrates the diagnostic challenge of the approach to longitudinally extensive myelitis. A diagnostic clue was the worsening with corticosteroid therapy, a classic finding described in AVF. Furthermore, the patient was not in the typical age range, as well as the unusual topography of the fistula, all possible confounding factors for diagnosis which may then have contributed to the unfavorable prognosis. **Final comments:** Our case highlights the importance of clinical reasoning and diagnostic clues in the diagnostic approach of longitudinally extensive myelitis. Although AVF is an infrequent etiology, its early recognition can lead to specific therapeutic interventions, with potential impact on prognosis.

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Diagnostic imaging of spontaneous intracranial hypotension: a case series

Samara Abdo El Hakim Kadri[1], Camila Carneiro Ferreira[1], Mohamad Ali Hussein[1], Pedro André Kowacs[1], Bernardo Correa de Almeida Teixeira[1], João Victor de Oliveira Souza[1], Lucas Bernardi Garcia[1], Eduarda Nakoula[1], Ana Júlia Caetano Ferreira[1], Gabriella Mara Arcie[1]

[1]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.

Correspondence: Samara Abdo El Hakim Kadri; Email: samara.kadri@hotmail.com.

Background: Imaging plays a central role in the initial diagnosis of spontaneous intracranial hypotension (SIH) and in its subsequent investigation and management. **Objective:** In this paper, we describe the main findings of brain MRI scans of patients admitted due to SIH at a neurology reference center. **Methods:** A retrospective, single-center, longitudinal study evaluating data from the electronic medical records of patients of both sexes hospitalized for SIH from 2019 to 2024. Statistical analyses were carried out using SPSS (Statistical Package for the Social Sciences) software. **Results:** A total of 16 patients were included, of whom 7 (44%) were female and 9 (56%) were male. The average age of the patients was 48 years (SD ±17.5). The youngest patient was 17 years old, and the oldest was 79 years old. Among the imaging findings, the most prevalent was pachymeningeal enhancement, and the least prevalent was epidural collection. A total of 12 (75%) patients had pachymeningeal enhancement, 9 (56%) had an engorged pituitary gland, 7 (44%) had engorged dural sinuses, 8 (50%) had a reduced nipple-point angle, 9 (56%) had cerebellar herniation, 7 (44%) had thinning of the ventricles, 6 (37%) had subdural collections, none (0%) had epidural collections, 4 (25%) had enlarged venous plexuses, and 1 (6%) had superficial siderosis. **Conclusion:** In this cohort, as previous literature has already shown, pachymeningeal enhancement was the most common imaging finding among SIH patients, while spinal findings were less prevalent.

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Diagnostic pathway of Noonan-like syndrome with loose anagen hair 1: case report

Matheus Sathler de Souza, Ivana Rocha Raslan[1], Ana Beatriz Calmon Nogueira da Gama Pereira[1], Ynnaê Côrtes da Silva Neri[1], Clarissa Laurindo de Oliveira[1], Raquel Mendes Moreira[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.

Correspondence: Matheus Sathler de Souza; Email: usoaternos@gmail.com.

This report describes a male patient (BSV) aged 8 years, followed since the age of 6 months due to global developmental delay. Parents were not consanguineous and pregnancy and birth were uneventful. Motor development milestones were delayed. Walking was achieved at the age of 3, with nonfunctional speech, cognitive deficits and epilepsy. The disease is typically nonprogressive and nonfluctuating. Dysmorphic features, capillary abnormalities, convergent strabismus, hypotonia and ataxic gait are the major clinical findings. The etiology could not be established despite comprehensive investigation (serum lactate, copper and ceruloplasmin measurement, analysis of biotinidase activity, amino acid chromatography, acylcarnitine profile analysis, chromatographic analysis of glycosaminoglycans and urinary oligosaccharides and hexosaminidase A testing). Cardiovascular and ophthalmological examination, auditory evoked potentials and abdominal ultrasound were unremarkable. Ventricular dilation was detected on brain MRI. G-banded karyotyping failed to detect aneuploidies. Microarray (CGH) analysis revealed a deletion on chromosome 8, p22p21.3, region 18483764_19418072, a variant of uncertain significance. Exome sequencing identified a monoallelic pathogenic variant in the SHOC2 gene NM_007373.5 (SHOC2):c.44A>G;p.(Ser2Gly). Hence, Noonan-like syndrome with loose anagen hair 1 (OMIM: #607721) was diagnosed. Noonan syndrome is a clinically and genetically heterogeneous, autosomal dominant rasopathy characterized by short stature, congenital heart defects and facial dysmorphism. It is caused by mutations in components of the RAS-MAPK signaling pathway and affects 1:1,000 to 1:2,500 live births. Rasopathies are genetic diseases caused by germline mutations in multiple genes of the ERK1/2 pathway. Loose anagen hair (NS/LAH; OMIM 607721) is a variation of the Nooan syndrome, with a single DNA nitrogenous base mutation in the scaffold protein SHOC2. Affected individuals are typically short and have comparatively more severe cognitive deficits, distinct behavior and easily pluckable, sparse, thin and slow-growing anagen hair lacking inner and outer root sheaths. Genetic counseling is advised given the autosomal dominant nature of the disease and probability of transmission. Patients should be monitored on a regular basis due to complications inherent to the syndrome, such as growth hormone deficiency, heart disease (especially pulmonary valve dysplasia and septal defects) and cognitive deficits.

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Diagnostic tools in the diagnosis of pure neural leprosy

Clarissa Neves Spitz[1,2], Izabela Jardim Rodrigues Pitta[1], Larissa Bittencourt de Carvalho[1,2], Ligia Rocha Andrade[1,2], Ana Caroline Siquara de Sousa[1], Mariana de Andrea Vilas-Boas Hacker[1], Anna Maria Sales[1], Roberta Olmo Pinheiro[1], Euzenir Nunes Sarno[1], Marcia Rodrigues Jardim[1,2]

[1]Fundação Oswaldo Cruz, Laboratório de Hanseníase, Rio de Janeiro RJ, Brazil. [2]Universidade Federal do Estado do Rio de Janeiro, Programa de Pós-Graduação em Neurologia, Rio de Janeiro RJ, Brazil.

Correspondence: Marcia Maria Jardim Rodrigues; Email: jardim.mm@gmail.com.

Background: Pure neural leprosy (PNL) is described as a clinical neurological lesion in the absence of cutaneous lesions, with an atypical presentation. Sensory cutaneous impairment, numbness, paresthesia, nerve pain, and nerve thickening are the most common symptoms and signs of PNL. Some authors claim that a definitive diagnosis of PNL can only be achieved by performing a biopsy of the periphery nerve. However, confirmation is still difficult, which encourages the search for the availability of other diagnostic methods. **Objective:** Correlate neurological clinical findings, nerve conduction studies (NCS) and peripheral nerve ultrasound (US) with peripheral nerve biopsy for the diagnosis of the PNL. **Methods:** Twenty-ninepatients were evaluated over a period of 18 months and were selected of possible PNL diagnosis. All of them had already been seen by dermatology and there were no suspicious lesionand negative bacilloscopy. In neurological assessment, patients were asked about the time of onset of symptoms, neural pain and triggering factors, sensory and/or motor symptoms, as well as neurological examination and detecting thickening neural, sensory-motor changes. NCS was performed to topography sensory and/or motor lesios and the type of impairment: axonal or demyelinating. Finally, US to evaluated neural thickening, morphological changes and fasciular echogenicity, flow on Power Doppler or muscular morphostructural changes. Thus, a peripheral nerve biopsy was performed and histopathologically analyzed for the presence of acid-fast bacilli (AFB) as well as the PCR technique. **Results:** Sixty percent of patients were male, median age of 56 years. The most frequently affected nerve was the ulnar and the average time of presentation was 2.5 years. In the clinical evaluation, none of the signs and symptoms were statistically significant. Regarding the NCS, all showed an abnormality in the sensory response and 25 also in the motor; the most common pattern of involvement was mixed (17), followed by axonal (7) and demyelinating (5) - all with conduction block. US: neural thickening in 72% of patients, hypoechogenicity and/or changes in fascicular morphology in 52%. The homogeneous fascicular pattern was the most frequently seen (20 patients). Neither the US nor the NCS method showed statistical significance. Of the 29 cases, 11 had the PNL form confirmed by biopsy, with p<0.05, for AFB and PCR technique. Of the 18 unconfirmed cases, all are still being followed up, with 2 diagnosed with familial amyloid polyneuropathy and 2 with autoimmune neuropathy. **Conclusion:** NLP is still a very common cause of neuropathy in many countries, such as Brazil, but it is still poorly recognized and understood. This work highlights that although we are looking for patterns of disease presentation and characteristics of complementary exams, peripheral nerve biopsy continues to be the exam capable of confirming PNL.

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Diagnostic value of intraepidermal nerve fiber density (IENFD) in lower limb skin biopsies of patients with hereditary transthyretin amyloidosis-polyneuropathy (ATTRv-PN) in a Brazilian centre

Mateus Gustavo Favaro[1], Carolina Lavigne Moreira[1], Gustavo Maximiano Alves[1], Marcus Vinicius Simões[1], Pedro Manoel Marques Garibaldi[1], Pedro José Tomaselli[1], Manoella Guerra de Albuquerque Bueno[1], Matheus Compart Hemerly[1], Wilson Marques Junior[1]

[1]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Mateus Gustavo Favaro; Email: mateusgustavofavaro@gmail.com.

Background: Hereditary transthyretin amyloidosis (ATTRv) is an autosomal dominant genetic disease that can lead to severe polyneuropathy and death. Many types of peripheral nerve involvement were described, including sensory-motor, sensory, motor, upper-limb onset, CIDP-like, radiculopathy, and small-fiber neuropathy. Identification of disease onset in pre-symptomatic carriers is a crucial step in order to identify disease onset and initiate early treatment. However, the best instrument to identify disease onset is still a point of debate. This project aims to determine the diagnostic value of intraepidermal nerve fiber density (IENFD) in lower limb skin biopsies in a Brazilian center. **Objective:** Identify sensitivity and specificity of intraepidermal nerve fiber density (IENFD) in lower limb skin biopsies of patients with Hereditary Transthyretin Amyloidosis-Polyneuropathy (ATTRv-PN) in a Brazilian center. **Methods:** 28 patients with pathogenic TTR variants followed at the the Amyloidosis Centre-HC-FMRP-USP were analyzed. These patients were pre-symptomatic carriers that that presented questionable complakints and/or abnormalities on neurological examination. Skin biopsies were performed on the lower limb, 10 cm above the lateral malleolus according to the European Federation of Neurological Societies/Peripheral Nerve Society Guidelines and established normal values (2010) Statistical analysis was performed at SPSS 22.0, and data was stored on REDCap. **Results:** Among this small Brazilian cohort of ATTRv-PN, the test's sensitivity was 69%, with a 100% specificity, a positive predictive value of 100%, and a negative predictive value of 41%. **Conclusion:** The sensitivity of IENFD for the identification of overt polyneuropathy in a Brazilian ATTRv population was lower than previously reported in American and European populations. These results can be related to the standardized values of IENFD, which could be different between populations around the globe, as a lower than expected sensitivity of this biomarker was also reported in a Chinese population

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Diagnostic value of voxel-based morphometry in MRI negative epilepsies and focal cortical dysplasia: correlation with electrocorticography and surgical results

William Alves Martins[1], Eliseu Paglioli[1], Ricardo Paganin[1], Thomas More Frigeri[1], Rafael Paglioli[1], Ricardo Bernardi Soder[1], Natalie da Silveira Donida[1], Tais Michele Werle[1], Bibiana Liberman Thomé[1], Andre Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: William Alves Martins; Email: walvesm.br@gmail.com.

Background: Epilepsy surgery is still the only hope of achieving seizure freedom for a significant number of patients with epilepsy. However, a visible MRI epileptogenic lesion, the best predictor of surgical success, is not found in a substantial portion of patients. Voxel-based morphometry (VBM) is an emerging method that may help identify and determine the extent of cortical abnormalities unseen in conventional imaging. **Objective:** To evaluate the diagnostic value for seizure control of VBM-based technique (Morphometric analysis program [MAP]) in patients with normal MRI or focal cortical dysplasia (FCD) undergoing epilepsy surgery. **Methods:** In this single center retrospective study, all patients submitted to epilepsy surgery between 2006 and 2020 at the Porto Alegre Epilepsy Surgery Program were screened . We included patients >2 years of age in which MRI was either normal or positive for FCD. Concordance of the extent of VBM-MAP+ lesions with electroclinical hypothesis and electrocorticography (ECoG) was evaluated, and completeness of resection of such lesions was correlated with surgical outcome using Engel classification. **Results:** Of the 141 patients included, 62 had a nonlesional MRI and 79 had a lesional MRI. MAP was positive in 93 (65.9%) patients, 66 lesional and 27 nonlesional. Resection of MAP+ regions was associated with seizure freedom in both in the whole cohort (76.6% x 26.1%; P=0.001) and in nonlesional patients (72.7% x 25%; p=0.02). In addition, resection of ECoG/MAP concordant regions led to a higher rate of seizure-freedom (75.8% x 16.6%, p=0.001). For non-lesional patients, MAP showed a sensitivity of 40% (CI 95% 19-64) and a specificity of 93% (CI 95% 80-98), with accuracy of 76% (CI 95% 63-86) for localizing the epileptogenic zone. **Conclusion:** VBM-MAP provides useful information in planning epilepsy surgery even in the absence of invasive extra-operative investigations. Combining a strong electroclinical hypothesis with ECoG guided resection of MAP+ regions may be a feasible strategy in low resource settings.

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Diagnostic yield of autosomal dominant cerebellar ataxia by whole exome sequencing in an undetermined ataxia cohort

Thiago Yoshinaga Tonholo Silva[1], Orlando Graziani Povoas Barsottini[1], Marcondes Cavalcante França Jr.[2], Flávio Moura Rezende Filho[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.
[2]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Thiago Yoshinaga Tonholo Silva; Email: thiago.yoshinaga@gmail.com.

Objective: To evaluate the diagnostic yield of whole exome sequencing (WES) in identifying autosomal dominant cerebellar ataxias (ADCA) caused by point mutations. Preamble: Currently, there are over 50 different subtypes of ADCA, mainly comprising spinocerebellar ataxias (SCA). The most frequent ADCAs are caused by nucleotide repeat expansions; however, most can be attributed to point mutations. Preliminary studies have demonstrated the utility of WES in the etiological identification of undetermined genetic ataxias. Therefore, WES has potential in the etiological diagnosis of undetermined ADCA after excluding repeat expansions. **Methods:** WES was performed on 130 patients with undetermined ataxia. A subset of the sample, comprising 12 cases, consisted of undetermined SCAs (ataxia with autosomal dominant inheritance and negative tests for the most common repeat expansions: SCA 1, 2, 3, 6, 7, 10, 12, 17, DRPLA). The remaining 118 cases were sporadic adult-onset undetermined ataxia without relevant family history. **Results:** Twenty-four patients were identified with ADCA related to point mutations: 3 patients with SCA28 (AFG3L2), 3 patients with SCA14 (PRKCG), 3 patients with SCA5 (SPTBN2), 1 patient with SCA15 and 2 patients with SCA28 (ITPR1), 2 patients with SCA45 (FAT2), 2 patients with SCA42 (CACNA1G), 2 patients with SCA19 (KCND3), 1 patient with SCA23 (PDYN), 1 patient with SCA26 (EEF2), 1 patient with SCA48 (STUB1), 1 patient with SCA25 (PNPT1), 1 patient with Alexander disease (GFAP), 1 patient with dominant ataxia related to CACNA1A, and 1 patient with SCA21 (TMEM240). **Conclusion:** WES was diagnostic in 18.46% (24 out of 130) of the cases in the Brazilian cohort of undetermined ataxia, identifying ADCA caused by point mutations. The most common point mutation SCAs in the series were SCA5, SCA14, SCA15/29, SCA19, SCA23, SCA28, SCA42, SCA45, and SCA48. These results may have implications for diagnosis, genetic counseling, and potentially treatment.

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Diagnostic yield of distinct next generation sequencing approaches in adult patients with suspected genetic myopathy

Alexandre Motta Mecê[1], Alberto Rolim Muro Martinez[1], Anamarli Nucci[1], Marcondes Cavalcante França Júnior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Alexandre Motta Mece; Email: amottamece@gmail.com.

Background: Myopathies in adults have diverse causes, but a substantial number of cases stem from genetic variations in over 200 identified genes. Whole-exome sequencing (WES), whole-genome sequencing (WGS), and genetic panels (GP) have been utilized as diagnostic tools in this context, showing varying rates of success. Variables such as age, ethnic background, and phenotypic characterization of patients appear to impact the precision in diagnosing myopathies. In this study, we examined the diagnostic yield of WES, WGS, and GP in a heterogeneous group of Brazilian patients with suspected myopathy. **Objective:** To determine the accuracy of WES, WGS and commercial GP in a population of patients from a reference neuromuscular center with suspected genetic myopathy. **Methods:** A total of 161 patients visiting a specialized neuromuscular center were enrolled. Despite physical examinations, muscle biopsies, electrophysiology, and laboratory tests, a definitive diagnosis remained elusive. WES was conducted according to established protocols for 30 patients, while GP was undertaken for 121 patients, and genome sequencing for 10 patients. Variants classified as pathogenic or likely pathogenic were identified following ACMG guidelines. Subsequently, the diagnostic efficacy of WES and GP was assessed across the entire cohort and within subgroups defined by major phenotypic characteristics. **Results:** A definitive genetic diagnosis was established in 12 out of 30 patients in the WES group (40% diagnostic accuracy), 53 out of 121 in the genetic panel group (43%), and 3 out of 10 in the genome sequencing group (30%), resulting in an overall diagnostic yield of 42% in this cohort. Pathogenic variants were identified in genes including SCN4A, ACADS, COL6A2, SGCB, CAPN3, POLG, TWNK, RYR1, TRIM32, TTN, EMD, SLC25A4, TPM3, DOK7, DMD, GAA, SGCB, PYGM, LAMA2, ANO5, CHRNE, GAA, ADSSL1, CLCN1, PABPN1, PYGM, PNPLA, HACD1, NEB, DPAGT1, MYH7, FKTN, and TRAPPC11. **Conclusion:** WES, WGS and GP are valuable diagnostic tools for adult patients presenting with undiagnosed myopathy. In this cohort, the diagnostic yield reached 42%, uncovering variants across multiple genes. Our findings indicate that the diagnostic yield of WGS is not different than that of WES in this population.

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Did Thomas Willis describe narcolepsy?

Fernanda Lustosa Cabral Gomez[1], Karla Carlos[1], Lucas Martins Teixeira[1], Letícia Lambert[1], Fernando Morgadinho Santos Coelho[1], Afonso Carlos Neves[1], Gilmar Fernandes do Prado[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Fernanda Lustosa Cabral Gomez; Email: fernandalustosacg@gmail.com.

Background: Narcolepsy is a disorder characterized by chronic daytime sleepiness, sudden sleep attacks and cataplexy. Westphal is often credited for first describing narcolepsy in 1877 and Gélineau for naming it in 1880. However, descriptions by Thomas Willis in the 17th century suggest that the disorder might have been recognized two centuries before. In this article, we explore the possibility of Willis (1621-1675), known as the founder of clinical neuroscience, having described narcolepsy in his works. **Objective:** Analyze Willis' descriptions and identify any indications that he described narcolepsy. Compare Willis' observations with modern understanding of narcolepsy. **Methods:** A group of sleep medicine specialists performed a review of Willis' publications, especially the English translation of "De Anima Brutorum" by Samuel Pordage. We compared Willi's descriptions with the current definition of narcolepsy, altogether with a review of interpretations of Willis' work by later scholars, such as Lennox. **Results:** In one of his most influential works, "De Anima Brutorum," Willis describes a condition he named "continual somnolence" where patients presented with uncontrollable sleep episodes during activities such as talking, walking or eating. Willis described these individuals maintained normal functions between these episodes, aligning with narcolepsy's intermittent sleep attacks . However, Willis did not mention symptoms suggestive of cataplexy, sleep paralysis, or hypnagogic hallucinations, raising the question if, indeed, he was describing the disease. He followed the description of his work with a case report of a man: "(...) afterwards growing aged, being given to idleness and drunkenness, became dull and stupid, and also Dropsical, with a great paunch, and his thighs and legs swelled (...) at length, though he was freed from the Dropsie, he was oppressed with so heavy a sleepiness, and that almost perpetually, that in what place soever he was, or what ever he was doing he would sleep". Despite Willis' descriptions being aligned with the current understanding of narcolepsy, his case report includes clinical signs that suggest the patient might have had other diagnoses. These alternative diagnoses include hepatic encephalopathy and cardiac insufficiency, which could explain "dropsie" (a term historically used to describe edema or swelling), obstructive and/or central sleep apnea syndrome. These conditions could present symptoms like those described by Willis, such as excessive daytime sleepiness, although with the rare occurrence of the "sleep attacks" described. **Conclusion:** It is possible that some of Willis' patients might have had narcolepsy; however, among some of the patients that inspired his descriptions, alternative diagnoses were probably present. These could include systemic diseases and other causes of hypersomnia, especially given the absence of detailed descriptions of cataplexy, sleep paralysis, and hypnagogic hallucinations.

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Differences between triggers of migraine and tension-type headache in southern brazilian patients.

Luiz Carlos Porcello Marrone[1], Emanuela da Silva Foletto[1], Cassiana Konig de Lima[1], Rafaela Giulia Zatt[1], Andrei Leonardo Schuster[1], Marina Macedo dos Santos[1], Cássia Elisa Marin Dapper[2], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Universidade de Caxias do Sul, Caxias do Sul RS, Brazil.

Correspondence: Cássia Elisa Marin Dapper; Email: cassiaemarin@gmail.com.

Background: Headache is a very common neurological condition in clinical practice. Identifying triggering factors is part of the treatment strategy for patients with migraine and tension-type headache (TTH). **Objective:** To evaluate the frequency and association of triggering factors for migraine and tension-type headache in a population from Southern Brazil. **Methods:** We selected 309 patients seen at the Neurology Department of Hospital São Lucas da Pontifícia Universidade Católica do Rio Grande do Sul between January 2010 and December 2013. Patients were diagnosed with either migraine or tension-type headache according to the criteria of the International Classification of Headache Disorders-2 (ICHD-2). The presence of the following triggering factors was assessed: 1) chocolate; 2) cheese; 3) caffeine; 4) wine; 5) other alcoholic beverages; 6) aspartame; 7) processed foods; 8) unpleasant odors; 9) sleep deprivation; 10) menstrual period; and 11) stress. Statistical analysis was conducted using SPSS software, and the chi-square test was used for variable analysis. **Results:** Most patients (218 [70.6%]) were female, and 215 (69.6%) had a diagnosis of migraine. Among headache-triggering foods, there was a statistically significant difference in the presence of chocolate, 21.9% in migraine patients vs. 10.6% in TTH group (p=0.019), and in relation to cheese, 12.6% vs. 4.3%, respectively (p=0.025). Stress was also more common in migraine (54.9% vs. 41.5%; p=0.036). The only trigger more associated with TTH was sleep deprivation, although without statistical significance (29.8% vs. 20.5%; p=0.075). **Conclusion:** Our cohort in southern Brazil showed similar headache triggers compared to other countries. Headache triggers may be identified in 50-75% of migraine patients. These precipitants are diverse, and different studies have found varying results when comparing migraine and TTH patients. Geographical distribution, genetic, and cultural factors

may contribute to these differences. In our population, stress was the most common trigger for both headaches, followed by the menstrual period, sleep deprivation, alcoholic beverages, chocolate, and caffeine. Stress, a major factor in chronic pain, was more commonly associated with migraines than TTH. Other studies have found conflicting results. Both migraine and TTH may share a dysfunction of pain modulation, more likely to appear in stressful situations and making this trigger difficult to evaluate in individual patients. Female gender, chocolate, and cheese were more significantly associated with migraines. In summary, we found that migraine and TTH share many headache triggers, potentially facilitating internal pain modulation. Stress, chocolate, and cheese were significantly more common in migraineurs. Whether these differences are clinically significant is still debated, but awareness of potential triggers and avoidance strategies remain important for educating and supporting patients in their headache management.

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Differential diagnoses of inflammatory myopathies with positive biopsy for dysferlin

Ana Luísa Vilela Sá Fortes[1], Maria Clara Motta Barbosa Valente[1], Fernando Pierini Costa[2], Mariana Tonon Giovannone[2], Vanessa Moraes Rossette[2], Ana Paula de Almeida Campos[2], Edmar Zanoteli[3], André Macedo[3]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
[2]Santa Casa de Santos, Santos SP, Brazil.
[3]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Ana Luisa Vilela Sá Fortes; Email: dranaluisa@outlook.com.

Case presentation: A 24-year-old male presented with a four-year history of progressive proximal lower limb weakness and gait difficulty. He had previously been active and engaged in high-impact exercise, but he had discontinued exercise and gained weight over the past four years. His gait became increasingly impaired, and he reported worsening limb weakness Laboratory tests revealed elevated muscle enzymes, including CPK (16,907), aldolase (141), and DHL (878). EMG showed a myopathic pattern in all four limbs, with evidence of membrane instability in the paraspinal and proximal lower limb muscles. Magnetic resonance imaging of the thigh revealed mild fatty replacement of the posterior and medial compartments. Based on the clinical presentation, laboratory findings, and EMG results, a diagnosis of inflammatory myopathy was suspected. The patient was treated with a pulse of corticosteroids and immunoglobulin, but his symptoms did not improve significantly. Other immunosuppressive therapies were initiated, without any improvement in the condition. Due to the progressive worsening of his symptoms, the patient underwent a muscle biopsy which revealed a marked reduction in dysferlin staining, suggesting dysferlinopathy. There was no CD8 staining, moderate CD68 staining, and mild MHC-I staining in some fibers. The histologic pattern was myopathic with rare necrosis and dysferlin deficiency. The features of the biopsy suggest the diagnosis of dysferlinopathy. Patient maintains clinical stability and is being weaned off immunosuppressive drugs. **Discussion:** Dysferlinopathies are autosomal recessive neuromuscular disorders caused by mutations in the DYSF gene, which encodes the dysferlin protein. The dysferlin protein is involved in muscle membrane repair, and mutations in the DYSF gene lead to a spectrum of phenotypes, including Miyoshi myopathy and limb-girdle muscular dystrophy type 2B (LGMD2B). Inflammatory myopathies are a group of autoimmune disorders that also cause muscle weakness and inflammation. Both diseases can share many clinical features, including muscle weakness, elevated muscle enzymes, and a myopathic EMG pattern. However, there are also some key differences. In inflammatory myopathies, there is typically an inflammatory infiltrate present in the muscle biopsy. In dysferlinopathies, there is no inflammatory infiltrate, but there is a deficiency of the dysferlin protein. The initial diagnostic hypothesis in the reported case was inflammatory myopathy, however the lack of response to immunosuppression raised suspicion of an alternative diagnosis, with muscle biopsy being of extreme importance for diagnostic guidance. **Final comments:** This case highlights the importance of careful differential diagnosis in patients with myopathy. The combination of clinical history, laboratory findings, EMG results, muscle biopsy findings, and response to therapy can help to distinguish between inflammatory myopathies and other disorders, such as dysferlinopathies.

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Differential diagnosis between ischemic stroke and mitochondrial myopathy, encephalopathy with lactic acidosis and stroke-like episodes (MELAS): a case report

Lucas Antonissen Lima Verde[1], Jamine Yslailla Vasconcelos Rodrigues[1], Thiago Moita Fernandes[1], André Felipe Oliveira Fernandes[1], Francisco Jazon de Araújo Neto[1], Espartaco Moraes Lima Ribeiro[1], Marcondes Pimentel Cruz[1], Paulo Roberto Lacerda Leal[1], Jose Elmano Silva[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Jamine Yslailla Vasconcelos Rodrigues; Email: jamineysl@gmail.com.

Case presentation: A 26-year-old woman presented with a generalized tonic clonic seizure lasting one minute, sialorrhea and no sphincter loss. The patient developed right hemispasticity and her companion reported the occurrence of a similar episode a year ago, without medical supervision. A cranial magnetic resonance imaging (MRI) was performed showing multiple focal areas of diffusion restriction, with hypersignal on T2/FLAIR weightings, diffusely arranged in the cortex of the frontal, parietal, temporal, occipital and insula lobes, compatible with acute ischemic insult. Therefore, a craniocervical angiography was requested, which showed no signs of stenoses or vasculitis, being within the normal range. As a result, the patient was transferred to the hospital's subacute stroke unit, in order to conduct the etiological investigation of the condition, and a spectroscopy resonance was performed in order to evaluate the lactate peak, with MELAS being the main diagnostic hypothesis. The patient progressed well over the days, with an improvement in her previous condition, but, upon neurological examination, she had difficulty understanding more complex commands and lactic acidosis was observed on blood gas analysis. L-carnitine, coenzyme Q10 and citrulline were started with MELAS in mind. Eight days after hospitalization, the patient was discharge from hospital, the diagnosis of MELAS remained as the main hypothesis and instructions were given on outpatient return and treatment. **Discussion:** MELAS is a rare mitochondrial disease involving the central nervous system and its diagnosis is a challenge for neurologists and radiologists. The diagnostic criteria for this disease involve three main points: the presence of encephalopathy (often with dementia and seizure), stroke-like episodes in young people and biochemical evidence of mitochondrial dysfunction, as well as lactic acidosis on muscle biopsy. However, the clinical presentation of these criteria can be difficult to differentiate from cerebrovascular disorders. Technological advances in neuroimaging made it possible to differentiate between stroke and MELAS. MRI with spectroscopy is a non-invasive method used to acquire information about metabolic disorders in brain tissue, which can be used to clarify the diagnosis and monitor the patient's disease. Furthermore, cerebral angiography is used to differentiate stroke-like episodes in MELAS from an ischemic stroke caused by a vascular occlusion. The treatment of this disease is not specific, as is the case with other mitochondrial diseases, and there is still no option to delay the progression of the disease, focusing on options that reduce the symptoms, such as the medications used in the patient. **Final comments:** Although it is very rare, patients living with MELAS have a significant loss in their quality of life. Advances in imaging exams help diagnose MELAS. Treatment depends on the patient's symptoms.

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Differential diagnosis for sudden visual deficits: a case series and literature review on Anton-Babinski syndrome

Sephora Sabrina Candido de Almeida[1], Ana Luísa Rosas Sarmento[1], Sonia Maria Cesar de Azevedo Silva Mourão Magalhães Gomes[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Sephora Sabrina Candido de Almeida; Email: sephora_sabrina@yahoo.com.br.

Case presentation: Case 1 Female, 76 years old, admitted with a 7-day history of sudden visual deficit, initially partial, progressing 6 days after admission to complete bilateral visual deficit - noticed by family members as the patient frequently stumbled in addition to trauma against objects. The patient did not look at the examiner, responded incorrectly when questioned objectively, including compromising her perception of light, and denied any visual changes. Computed tomography of the skull with initial arterial computed tomography angiography of the skull and neck demonstrated: Right occipital corticosubcortical hypoattenuating area, suggestive of a subacute ischemic event. Areas of gliosis occipital and temporal encephalomalacia on the left and in the left centrum semiovale determining an atrophic effect with compensatory dilation of the corresponding lateral ventricle. Case 2 Female, 75 years old, admitted with a history of lipothymia witnessed by her family when she arrived at the emergency room in the morning. After admission, the patient presents difficulty in locating himself in the environment, apparently with bilateral visual deficits. The patient previously did not present any visual deficit. Upon neurological examination, the patient remained discreetly inattentive but collaborative. No somatic examination deficits. He actively denied any visual changes, but the patient did not keep his gaze fixed on the examiner, responded randomly during confrontation perimetry and responded incorrectly when questioned objectively. Skull tomography demonstrated: a small hemorrhagic focus with adjacent edema in the left occipital lobe, determining the effacement of local sulci. Hypoattenuating area affecting the corticosubcortical region of the right occipital lobe, causing erasure of local sulci, which may correspond to recent ischemia. **Discussion:** Anton-Babinski syndrome is characterized by blindness, which is the syndromic core, without critical deficit. Anterior visual tracts are intact and the lesion is located in the posterior cerebral cortex. In addition to visual deficit, confabulation is an important part of the syndrome. Its main characteristics are visual agnosia, anosognosia and confabulation. It is one of the most striking examples of anosognosia described in medicine. It is important to remember that for the diagnosis of the syndrome there must be no pre- existing memory impairment , such as dementia. This differentiates it from Dide-Botcazo syndrome, which is also known to include the presence of cortical blindness with visual anosognosia, but occurs involving the memory circuitry.The clinical diagnosis of Anton-Babinski syndrome is based on: absence of response to visual stimulation (blinking), preservation of pupillary reflexes, fundoscopy without changes and preserved extraocular movements (absence of damage to anterior visual pathways) and absence of cognitive deficit.

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Differential diagnosis of cluster headache: a systematic review

Gabriela Goveia de Sousa Castilho de Lima[1], Felipe Araújo Andrade de Oliveira[1], Ana Beatriz Alves[1], Sarah Ellen Alves Soares[1], Maria Kaylanne Pinheiro de Araújo[1], Luan Santana Martins da Costa[1], Daniel Rodrigues Lemos Neto[1], Filipe Alves da Costa Dias[1], Mariana Leandro Silva[1], Érycle Guilherme da Silva[1]

[1]Faculdade de Medicina de Olinda, Olinda PE, Brazil.

Correspondence: Gabriela Goveia de Sousa Castilho de Lima; Email: gabrielagscl@outlook.com.

Background: Cluster Headache belongs to the group of T rigeminal Autonomic Cephalalgias. It is a subtype of primary headache with an as-yet undetermined cause. It primarily affects men aged between 20 and 40 years, manifesting as intense, unilateral pain crises located in the cutaneous distribution territory of the trigeminal nerve in the orbital and periorbital region, associated with symptoms of autonomic dysfunction ipsilateral to the pain, with a characteristic temporal profile. **Objective:** To describe possible differential diagnoses of Cluster Headache. **Methods:** This is a systematic review, referencing scientific articles from MEDLINE, PubMed, and Scielo, a study of literature review from the 3rd Edition of the International Classification of Headaches, using articles from the last ten years in English and Portuguese. The following descriptors were used in the searches: "Cluster Headache", "Differential diagnosis of cluster headache" in English, and "Cefaleia em Salvas", "Diagnósticos Diferenciais da Cefaleia em Salvas" and "Cefaleias Primárias" in Portuguese. **Results:** Differential diagnosis is one of the most challenging aspects in headache cases, based on the duration, intensity, and topography of signs and symptoms. Several conditions can present as unilateral pain in the periorbital location. A literature review evaluated 63 cases that were compatible with the clinical presentation of cluster headaches. Among the differential diagnoses found in the study, two main pathologies stand out: closure-angle glaucoma and spontaneous or post-traumatic carotid dissection. Closure-angle glaucoma can present with intense, intermittent ocular pain and conjunctival hyperemia. Spontaneous or post-traumatic carotid dissection can lead to hemicranial headache with cervical radiation, accompanied by ipsilateral Horner's syndrome due to the involvement of sympathetic nerve fibers. Symptoms similar to a Cerebrovascular Accident (CVA) facilitate the diagnosis, as does a family history of dissection, connective tissue disorders, and trauma. **Conclusion:** A careful and precise evaluation of the signs and symptoms of cluster headache is needed, observing the diagnostic criteria of the International Classification of Headaches and concurrently maintaining vigilance for the aforementioned differential diagnoses to avoid diagnostic errors and consequent therapeutic errors.

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Differential diagnosis of epilepsy and the importance of the video-electroencephalography: a case report

Gláucia Monteiro Oliveira[1], Vivian Proença Lara[1], Luiza Barbosa de Oliveira[1], Vanessa Alves Lobato[1], Helena Barbosa Horta Santos[1], Aline Curcio de Moraes[1], Fernando Augusto Bruzzi[1], Igor Pereira Matos de Oliveira[1], Ana Luíza Xavier Drumond[1], Maria do Carmo de Vasconcellos[1]

[1]Santa Casa de Misericórdia de Belo Horizonte, Belo Horizonte MG, Brazil.

Correspondence: Gláucia Monteiro Oliveira; Email: glaucia.monoli@gmail.com.

Case presentation: A 2 years old patient, started at the age of 4 months with episodes of loss of consciousness associated with crying. She was referred to a video-electroencephalography (VEEG) in order to investigate breath-holding spells or epilepsy suspicion, as she was taking valproate with no response. This child had normal neuropsychomotor development and unknown comorbidities, but in her family record it was possible to identify a member with epilepsy. VEEG documented episodes that whenever the child was crying after some form of upset, it was followed by collapse with loss of consciousness associated with generalized hypertonia, upward gaze, lip cyanosis and pale skin. Electrocardiographic recording demonstrated bradycardia, followed by asystole lasting 10 seconds, heartbeat returned with irregular rhythm. Ictal EEG demonstrated hypersynchronous, high-amplitude slowing followed by flattening and later generalized spike-wave discharges. With this new diagnosis, the patient was referred for cardiac surgery at a reference center and she had a pacemaker fitted, progressing without recurrence of seizures without using any anticonvulsant drugs since then. **Discussion:** Epileptic seizures are paroxysmal events that must be distinguished from other disorders, specially those that mimic seizures. It is not always a simple task, because clinical presentation of paroxysmal spells attributable to cardiac diseases, syncope and functional seizure may resemble epileptic seizures. Anoxic seizures may present with decerebrate posturing with extensor stiffening that can mimic a tonic seizure and be followed by irregular tonic-clonic movements. In this case, the child presented an anoxic seizure as a consequence of cardiac etiology. When it is difficult to obtain clinical information that corroborates to a diagnosis, complementary exams such as VEEG are extremely important. **Final comments:** The VEEG is an important tool to investigate several differential diagnoses such as anoxic seizure. As we can see, the correct diagnosis allows us to treat our patient correctly. In this dramatic case, it was necessary surgical treatment based on findings on VEEG. This changed the life of the child, avoiding her to remain incorrectly diagnosed, taking inappropriate medications and live with a cardiomyopathy life threatening undiscovered.

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Differential diagnosis of neuromyelitis optica and brainstem glioma in a pediatric patient: a case report and how to differentiate between them

Valdecir Boeno Spenazato Júnior[1], Thales Pardini Fagundes[1], Isabela Rios Felix[2], Antonio Torrecilha Manfrinato[2]

[1]Fundação Pio XII, Barretos SP, Brazil.
[2]Faculdade de Ciências da Saúde de Barretos Dr. Paulo Prata, Barretos SP, Brazil.

Correspondence: Isabela Rios Felix; Email: isabelar.felix13@gmail.com.

A 12-year-old girl presented with severe vomiting, weight loss, ataxia, and nystagmus. MRI revealed a hyperdense lesion at the cervicomedullary junction, initially suspected to be a tumor. She began dexamethasone and was referred to our service. In April 2024, Anti-Aquaporin 4 IgG antibodies confirmed neuromyelitis optica spectrum disorder (NMOSD) with early-area postrema syndrome. Dexamethasone was discontinued, and prednisone at 1 mg/kg/day was initiated. She improved significantly, and Rituximab treatment is planned. Differentiating neuromyelitis optica (NMO) from brainstem gliomas is crucial due to different therapies and prognoses. NMO is marked by Anti-Aquaporin 4 IgG antibodies and responds to immunosuppressants. NMO lesions are extensive in the spinal cord and area postrema, typically non-infiltrative on MRI. Both conditions can present with cranial nerve palsies, ataxia, and long tract signs. Demyelination often has an acute onset, unlike tumors. MRI characteristics of demyelination include hyperintensity on T2-weighted and FLAIR images, with ill-defined margins and variable gadolinium enhancement. In contrast, brainstem gliomas exhibit more mass effect, heterogeneous appearance with necrosis or cystic changes, and pronounced, irregular enhancement patterns. In this case, the hyperdense lesion at the cervicomedullary junction initially suggested a tumor, leading to the administration of dexamethasone. However, the confirmation of Anti-Aquaporin 4 IgG antibodies redirected the diagnosis to NMOSD, highlighting the importance of specific biomarkers in differentiating these conditions. Advanced neuroimaging techniques, such as Amide Proton Transfer-weighted (APTw) imaging and 23Na MRI, can further aid in distinguishing these conditions by providing detailed tissue characterization. These techniques, although not used in this case, represent a valuable tool for future similar diagnostic challenges, potentially offering more precise differentiation between demyelinating lesions and neoplasms. Accurate differentiation between NMO and gliomas is essential for management and prognosis. Anti-Aquaporin 4 IgG antibodies, immunosuppressive response, and imaging are key. This case avoided invasive procedures and ensured effective treatment through precise diagnosis.

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Difficult-to-control epilepsy due to germ cell brain tumor and asymmetric hippocampal atrophy: case report

Bárbara Araújo Pereira Cabral[1], Ricardo Cézare Araújo[1], Vanessa C. S. Mautone Lima[1], Glenda Corrêa Borges de Lacerda[1]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Bárbara Araújo Pereira Cabral; Email: ba.araujo02@gmail.com.

Case presentation: AGJ, 27 years old, with a history of epilepsy characterized by tonic-clonic seizures since 7 months of age, underwent surgery for the removal of a brain tumor and for Ventriculoperitoneal Shunting at the age of 24, after which he began experiencing dysperceptive seizures. During the preoperative period for tumor excision, a cranial magnetic resonance imaging (MRI) was performed, revealing a heterogeneous expansive lesion with a solid-cystic component, slight non-homogeneous enhancement with venous contrast, and signs of diffusion restriction within the solid areas. The lesion was located in the midline, occupying the body of the lateral ventricles and involving part of the 3rd ventricle, as well as causing ectasia of the supratentorial ventricular system, notably on the right side. Surgery was performed to remove the mass, which was classified by biopsy as originating from germ cells, extending into the right fronto-parietal lobe, and also included the excision of part of the corpus callosum followed by Ventriculoperitoneal Shunting. A postoperative cranial MRI detected the possibility of residual lesion amid the excision of the tumor, with gliosis in the right fronto-parietal region extending to the 3rd ventricle, enlargement of the cerebrospinal fluid space in the posterior fossa, and asymmetric atrophy in the right hippocampus. **Discussion:** Germ cell tumors (GCT) of the central nervous system (CNS) are rare and primarily occur in the medial structures of the brain due to remnant germ cell lineage cells that have migrated to these locations. With rapid progression, these lesions can cause obstruction in the ventricular system due to their proximity, and patients often present with ventricular ectasia due to the obstruction of cerebrospinal fluid (CSF) flow. Dysperceptive seizures are frequently associated with hippocampal atrophy, the structure most affected in temporal lobe epilepsy, and show greater resistance to pharmacological treatment. Furthermore, since the hippocampus is associated with memory processes, memory deficit is the most significant alteration in patients with hippocampal atrophy.

Final comments: This is a rare case in which surgical intervention for tumor removal and Ventriculoperitoneal Shunting were important for managing hydrocephalus. However, postoperative examinations detected a residual lesion amid the excision, which, combined with right hippocampal atrophy, significantly contributed to complications such as dysperceptive seizures shortly after the shunting, and also proved to be a debilitating factor for memory processes. So, close monitoring is crucial to detect and manage complications early, emphasizing seizure control and reducing long-term cognitive and functional decline. This case highlights the difficulty in managing epilepsy in patients with double brain lesions who have already undergone surgery, due to the risk of a new interventional approach associated with resistance to drug therapy.

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Diffuse and extensive intracranial calcifications in a patient with systemic lupus erythematosus: an atypical case of Fahr's syndrome

Melissa dos Santos Matos[1], Daniel Mariani Favalessa[1], Ivo Gabriel Ribeiro Carvalho[1], Emanuelly Victoria Andrade Fonseca[1], Guilherme Valiate Picoli[1], Paula Zago Melo[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Melissa dos Santos Matos; Email: melissa.matos@edu.ufes.br.

Case presentation: A 47-year-old patient, diagnosed with Systemic Lupus Erythematosus (SLE) since 2016, was referred to the neurologist complaining of mild dysarthria, which began in 2017. The condition, however, progressively worsened over the years, accompanied by difficulty walking due to weakness in the lower limbs, insomnia and a mood disorder. Since 2019, she has also reported a frontal, throbbing, vertex headache with associated nausea, with episodes around 2 to 3 times a week and partial relief with dipyrone. He also had an episode of loss of consciousness preceded by tremors in the right eyelid in 2021. On physical examination, there was decreased tactile sensitivity in the left upper limb and left hemiface, as well as a gait with a broad base and a staggering gait in the left lower limb. Computed tomography (CT) of the skull showed gross calcifications affecting the frontal, occipital parietal and temporal lobes, as well as the thalamus, striatum, globus pallidus, cerebellum and midbrain. Brain MRI confirmed the CT scan findings. **Discussion:** Fahr's syndrome is a rare secondary condition of intracranial calcification, marked by its neurodegenerative nature, and is often underestimated and underdiagnosed. The patient in question had neurological symptoms, including motor deficits, headache, dysarthria, as well as psychiatric disorders such as mood disturbances. Among the main causes of this syndrome - unlike the primary form, which is hereditary - are hypoparathyroidism, as well as infections such as cytomegalovirus and HIV. The patient in this case had no similar family history, and all possible secondary causes were ruled out. It is therefore assumed that these calcifications were caused by SLE. CT is the preferred method for locating and assessing the extent of cerebral calcifications, which are identified as hyperdense lesions. The pathogenesis of these findings is unclear and there are no confirmed hypotheses, but possibly the calcifications result from repeated episodes of venous inflammation with focal immunological demyelination. A likely mechanism could be the extravasation of proteins from venous vessels with neurotoxic and pro-calcification properties. Treatment is based on symptoms and the underlying cause, and the prognosis is variable, with a tendency for symptoms and calcification to worsen. **Final comments:** We report this case considering the rarity of symptomatic Fahr's syndrome in a patient with SLE. The underlying mechanism of calcification in this context is not known, but it is suggested that it is due to ongoing venous inflammation. This report highlights the importance of considering Fahr's syndrome as a possible explanation for neurological manifestations in patients with SLE. This approach, in addition to broadening the diagnostic spectrum, improves understanding of the neuropsychiatric manifestations associated with SLE, allowing for more comprehensive and precise clinical management.

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Diffuse brain involvement in lymphomatoid granulomatosis

Marco Antônio Delazeri[1], Gustavo Henrique Tomasi[2], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil. [2]Hospital São Vicente de Paulo, Curitiba PR, Brazil.

Correspondence: Marco Antônio Delazeri; Email: marco.delazeri@outlook.com.

Case presentation: A 34-year-old immunocompetent woman had history at lung lymphomatoid granulomatosis (LYG) associated with Epstein-Barr virus (EBV), diagnosed in 2010 after a lung segmentectomy, treated with cyclophosphamide and methylprednisolone over a period of six months. At that time, she was carefully investigated with imaging tests in order to rule out LYG at other sites. Brain magnetic resonance imaging (MRI) findings showed infiltration of cranial nerves and meningeal enhancement, remitting after treatment. Five years later she seemed apathic and had suicidal ideation, also presenting disperceptive episodes. She was admitted in a psychiatric facility for a few days, after which she left home. For the next seven days, she progressed with slurred speech, ataxia, left hemiparesis, and motor ocular paralysis. Neurologycal exam showed left-side weakness, left cranial nerve paresis, horizontal nystagmus, left hypoesthesia and right internuclear paralysis. Cerebrospinal fluid was normal. MRI is described in Fig 1. It was started pulse therapy with methylprednisolone for seven days, with no clinical improvement. Hematologists considered this clinical condition as a progression of the LYG and patient received systemic chemotherapy (CHOP scheme from March 2015 at June 2015) and intratechal chemotheraphy. She showed clinical improvement of symptoms after chemotherapy with reduction of lesions in MRI and currently remains neurologically stable with no CNS recurrence. **Discussion:** Lymphomatoid granulomatosis (LYG) is a rare B-cell lymphoproliferative disorder, normally associated with Epstein-Barr Virus (EBV), that involves especially the lungs, but may affect skin and central nervous system (CNS) during its course. Nonetheless, primary CNS-LYG may represent a lymphoproliferative disorder with low-grade T cell malignancy, contrary to systemic LYG, which is composed of EBV-positive B cells. CNS-LYG has a variable spectrum of lesions at MR imaging. In a study, multiple focal lesions that involve the white matter, the deep gray matter, or the brainstem represent the most common abnormalities, but the hyperintensity on the T2-weighted MRI was not specific to this condition. The punctate and/or linear enhancement in these lesions appears to be a characteristic finding of lymphomatoid granulomatosis in the brain. This enhancement is related to the perivascular and vascular wall infiltration by pleomorphic and lymphoid cells, what was previously shown in biopsy specimens of the brains. Lesion morphology is a nodular infiltrate with polymorphous mononuclear cell, vascular infiltration and often necrosis permeated with CD20-positive B-lymphocytes in a background of CD3-positive small T lymphocytes and scattered admixed plasma cells and histiocytes. **Final comments:** There is scarce knowledge about LYG progressing to CNS and, therefore, the pathophysiology and phenotype are not well defined. Our case highlights its presentation and response to proper treatment.

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Diffuse cortical laminar necrosis in dengue encephalitis and status epilepticus: rare complications of a common disease in a climate crisis scenario

Leticia Bassani Devens[1], Giuseppe Dick Bonato[1], William Alves Martins[1]

[1]Hospital São Lucas, Departamento de Neurologia, Porto Alegre RS, Brazil.

Correspondence: Leticia Bassani Devens; Email: leticiabassanidevens@gmail.com.

Case presentation: We present a 72 year-old woman transferred to our tertiary hospital after 5 days of headache, diarrhea and high fever followed by 4 days of continuous conscious level impairment. Dengue infection was diagnosed after NS1 antigen positive test. A first episode of generalized seizure led to tracheal intubation and transference to ICU. Medical history included well controlled hypertension, diabetes and hypothyroidism. At arrival in our tertiary hospital, she had no arousal to stimuli and preserved brainstem reflexes. Brain CT scan was uneventful, and first CSF analysis showed 26 leucocytes/mm3 with neutrophilic predominance, 102mg/dl proteins, 3,2mg/dl lactate, and normal glucose ratio. Search for acid-fast bacilli and neoplastic cells, bacterial and fungal cultures and HSV PCR were negative. CSF results led to initiation of ampicillin, ceftriaxone and acyclovir. EEG showed severe cortical encephalopathy with frequent sharp waves in anterior quadrants compatible with clinical seizures. Despite levetiracetam and continuous IV midazolam introduction, continuous video-EEG revealed severe encephalopathy and frequent bilateral temporal focal non-clinical seizures. Propofol sedation was added and levetiracem increased. Subsequent EEG showed severe background suppression without further epileptic activity. Brain MRI had diffuse pachymeningeal enhancement and thickening, slight cortical signal increase, restricted diffusion and enhancement compatible with laminar necrosis. After 12 days of adjustments (levetiracetam 3g/day, clobazam 10mg/day, phenytoin 200mg/day, carbamazepine 1200mg/day), and no more epileptic events, propofol was tapered. Serum dengue IgM was positive, and CSF PCR negative. Minimal clinical improvement prompted empirical methylprednisolone 1g/day for 5 days without change in clinical status. New brain MRI after 16 days showed signal alteration and diffuse enhancement in cortex and temporal amygdalae. She was able to receive domiciliary care after traqueostomy and diet adjustment, being discharged with mRS 5/6. **Discussion:** Although patient showed negative CSF dengue PCR, this clinical case is compatible with dengue encephalitis. Dengue infection is highly prevalent in Brazil, and its Southern region was under a severe outbreak when this patient developed symptoms. Although not commonly recognized, dengue is regarded as the second most common cause of infectious encephalitis in countries where it is endemic. Pathogenesis is not fully understood, and dengue virus neurotropism is still questioned. **Final comments:** Dengue encephalitis is challenging: no specific treatment is available, and support measures are advised. Using video-EEG to determine epileptic control and safe sedation tapering prevents further continuous epileptic activity damaged. New treatments options are warranted, especially due to current environmental changes that lead to dengue vectors increase, turning a rare manifestation into a likely complication.

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Digital analysis of immunostaining and tissue distribution of the R132H mutant form of isocitrate dehydrogenase 1 in diffuse low or high-grade adult astrocytomas

João Vitor Gerdulli Tamanini[1], Arethusa de Souza[1], Guilherme Rossi Assis-Mendonça[1], Enrico Ghizoni[1], Helder Tedeschi[1], Mateus Dal Fabbro[1], Leandro Luiz Lopes de Freitas[1], Marcus Matsushita[2], Roger Frigério Castilho[1], Fabio Rogerio[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Faculdade de Medicina Franco Montoro, Cachoeira de Cima SP, Brazil

Correspondence: João Vitor Gerdulli Tamanini; Email: jvgtamanini@yahoo.com.br.

Background: Diffuse gliomas are common primary tumors of the central nervous system. The current diagnosis of such lesions, which include astrocytomas, depends on molecular features, including the presence of mutation in the enzyme isocitrate dehydrogenase(IDH). The wild isoforms of IDH-1(cytosolic) and 2(mitochondrial) convert isocitrate into α-ketoglutarate(αCG), promoting the reduction of NADP+ to NADPH. Both αCG and NADPH are antioxidants. The most common mutant form of IDH1(R132H) has a gain of function in which αCG is converted into D2-hydroxyglutarate, an oncogenic compound that consumes NADPH. Thus, a functional relationship is observed between IDH1, IDH2 and IDH1R132H. **Objective:** To perform neuropathological characterization of diffuse astrocytomas in adults as well as to investigate the immunostaining and possible alteration in tissue distribution of IDH1, IDH2 and IDH1R132H, considering the presence of the R132H mutation and histological grade. **Methods:** We evaluated 25 surgical specimens from adults with diffuse gliomas, which were molecularly and immunophenotypically characterized as astrocytomas of low- or high-grade (2 (n=10) or 4 (n=15), respectively), according to the World Health Organization current criteria. We also analyzed the immunostaining pattern of IDH1, IDH2 and IDH1R132H, as well as their tissue distribution (mean % of positivity assessed by using ImageJ® software), considering the R132H mutation and the lesion histological grade. **Results:** Regarding age at diagnosis(< or ≥55 years) and R132H immunopositivity, the lesions were classified as: astrocytoma, IDH-mutant, grade 2 (5/25); astrocytoma, not otherwise specified (NOS), grade 2 (5/25); glioblastoma, IDH-wildtype, grade 4 (9/25) or astrocytoma, NOS, grade 4 (6/25). IDH1, IDH2 and IDH1R132H showed a diffuse cytoplasmic staining pattern, regardless of grade. When comparing low- and high-grade lesions, the tissue distribution of IDH1 was lower in the former (14.20±1.70% and 16.40±1.60%, respectively; p=0.0051), no difference being observed for IDH2 (15.10±1.80% and 14.90±2.0%; p=0.6773) and R132H (5.20±5.60% and 1.80±4.80%; p=0.1234). Specifically, low-grade lesions with (5/10) or without (5/10) the mutation showed no differences regarding tissue distribution of IDH1 (14.40±2.30% and 14.10±0.90%, respectively; p=0.2963) or IDH2(15.60±1.0% and 14.70±2.40%; p=0.8345). Irrespective of the histological grade, there was correlation neither between tissue expressions of R132H and IDH1 nor between R132H and IDH2. **Conclusion:** In the present series, IDH1R132H was more frequent in grade 2 than in grade 4 lesions. Furthermore, our data suggest that (1) such mutation does not interfere with tissue expression of wild-type IDH1 and IDH2, and (2) the wild-type IDH1 allele would ensure protein synthesis at physiological levels.

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Digital tool validation for syndromic and topographic diagnosis in vascular neurology: LouiS

Thales Pardini Fagundes[1], João Brainer Clares de Andrade[2], Felipe Hideki Soga[2], Túlio Maranhão Neto[1], Renan Barros Lopes[1], Alice Marília Silva Abreu Mota[2], Victor Dyego Mena Romeiro Nishimura[3], Octávio Marques Pontes-Neto[1], Millene Rodrigues Camilo[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[3]Beneficência Portuguesa, São Paulo SP, Brazil.

Correspondence: Millene Rodrigues Camilo; Email: millene.camilo@gmail.com.

Background: Accurate syndromic and topographic diagnosis in Vascular Neurology is crucial for appropriate clinical management and therapeutic interventions. However, significant gaps in medical training hinder precise diagnoses. Educational digital tools may bridge these gaps, but few have undergone rigorous scientific validation. **Objective:** To validate the LouiS digital tool for syndromic and topographic diagnosis in Vascular Neurology. **Methods:** Vascular neurologists developed algorithms based on literature review and consensus. An independent programmer created the LouiS interface. A cross-sectional study was conducted where residents, fellows, professors, and neurology students, randomized into two groups, used the tool or not to diagnose 40 hypothetical cases. Diagnostic accuracy and user satisfaction were evaluated. Data were treated ethically and confidentially. User satisfaction was assessed in terms of time spent, quality of questions, ease of use, and platform friendliness. **Results:** LouiS tool is expected to increase diagnostic accuracy and user satisfaction, proving its educational efficacy in Vascular Neurology. The app has already been created and is available at https://louis-stroke.com/. The tool encompasses 171 symptoms, 98 syndromes, 170 images, and 4629 diagnostic possibilities, aiming to improve diagnostic precision and user experience. **Conclusion:** LouiS has the potential to significantly improve diagnostic accuracy in Vascular Neurology, addressing critical educational gaps. There are no direct risks or benefits to participants.

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Directional deep brain stimulation in the management of Parkinson's disease: efficacy and constraints - an analytical appraisal

Mayara Tszesnioski Maçaneiro[1], Ana Clara Azevedo[1], Bruna Maurício Poerner[1], Milena Dangui da Silva[1], Andrei Koerbel[1]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.

Correspondence: Mayara Tszesnioski Maçaneiro; Email: mayaratm.tsz@gmail.com.

Background: Deep brain stimulation (DBS) is a widely employed treatment for Parkinson's disease. However, conventional DBS utilizing ring-shaped leads can often result in undesirable side effects by stimulating nearby brain structures, thus limiting its effectiveness. To address this issue, a novel DBS electrode was developed to allow for directional stimulation, avoiding neighboring structures. **Objective:** This literature review aims to analyze the disparities between conventional and directional DBS and discuss the benefits and limitations associated with this innovative electrode design, focusing on the stimulation-induced side effects it can or cannot mitigate. **Methods:** A comprehensive search was conducted in MEDLINE/PubMed, ScienceDirect, and EBSCO databases using the Boolean search criteria: "Deep brain stimulation" AND "Parkinson" AND "Directional." Following the application of inclusion and exclusion criteria, the selected articles were downloaded for full-text reading. Subsequently, the results were organized and analyzed to compose this article. **Results:** Numerous studies have demonstrated that directional DBS effectively reduces side effects associated with brain stimulation, prevents the stimulation of non-targeted structures, and expands the therapeutic window, among other advantages. However, it has been observed that directional DBS may be more challenging to program and requires higher energy consumption. Furthermore, there is a lack of standardization among different manufacturers of directional DBS electrodes. Various stimulation-induced side effects, including dysarthria, dyskinesia, paresthesias, and symptoms of pyramidal tract activation, have been shown to be mitigated with the use of directional DBS. Moreover, directional electrodes offer a wider therapeutic window and a reduced incidence of undesired effects, requiring the same or lower minimum current for symptom relief compared to conventional DBS. **Conclusion:** The utilization of directional leads in DBS offers numerous advantages over conventional electrodes without significant drawbacks for patients undergoing directional DBS therapy.

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Discontinuation of disease-modifying drugs in older patients with multiple sclerosis: an observational study

Camila Lopes Figueiredo[1], Marcelle Sanjuan Ganem Prado[1], Maria Clara Motta Barbosa Valente[1], Leticia Machado Dumont[1], Rafael Chaves Claudino de Queiroga[1], Cicero Rodrigues Veloso[1], Filipe Brito Ferraz da Silveira[1], Eliana Tomomi Shimabukuro da Cunha[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Camila Lopes Figueiredo; Email: camila04_lopes@hotmail.com.

Objective: To evaluate the discontinuation of disease-modifying drugs (DMTs) in patients with multiple sclerosis (MS) aged 55 years or older. **Methods:** A retrospective observational study of 42 MS patients aged 55 years or older, followed at the demyelinating disease outpatient clinic of the Santa Casa de Misericórdia de São Paulo Hospital, Brazil, for at least 2 years, with clinical and radiological stability of the disease. Data was collected between March and April 2024. **Results:** Of the 42 patients, 33.3% (14) discontinued DMTs, 64.3% (9) due to disease stability, 7.1% (1) due to adverse effects, 7.1% (1) due to lack of medication, 14.3% (2) by their own decision, and 7.1% (1) due to other causes. The remaining 66.6% (28) remained on DMTs, the main ones being Beta-interferon, Dimethyl Fumarate, and Natalizumab. **Conclusion:** Patients who discontinued DMTs for any reason maintained stable disease, with no new radiological activity or worsening of EDSS. DMT discontinuation did not appear to influence clinical outcomes, corroborating the perception of the limited effect of immunomodulation in older patients. Keywords: Multiple sclerosis, disease-modifying drugs, discontinuation, older patients, clinical outcomes.

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Disease burden and outcomes in adult patients with Duchenne muscular dystrophy

Clara Gontijo Camelo[1], Raquel Diógenes Alencar[1], Mariana Cunha Artilheiro[1], Cristiane de Araujo Martins Moreno[1], Joemir Jábson da Conceição Brito[1], Karlla Danielle Ferreira Lima[1], Maria Bernadete Dutra de Resende[1], Marcos Antônio Veloso de Albuquerque[1], Umbertina Conti Reed[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Clara Gontijo Camelo; Email: claragc@gmail.com.

Background: Duchenne muscular dystrophy (DMD) is an inherited X-linked recessive neuromuscular disorder caused by variants in the DMD gene, resulting in the absence of the structural protein dystrophin. This condition leads to progressive limb-girdle muscle weakness, loss of ambulation (LOA) by the ages of 10 to 14 years, dilated cardiomyopathy, and respiratory insufficiency. Over the past 30 years, the natural history of individuals with DMD has evolved significantly due to implementing standards of care (SoC), including ventilatory support, glucocorticoid treatment, proactive cardiac intervention, and multidisciplinary care. However, the disease burden and outcomes for adult patients with DMD remain inadequately described. **Objective:** This study aims to describe the disease outcomes and burden in adult patients with DMD followed by a specialized care service. **Methods:** A retrospective cross-sectional study was conducted by reviewing medical records. Inclusion criteria were: (1) patients who lost ambulation before the age of 13 years (clinical diagnosis of Duchenne); (2) patients who lost ambulation by the age of 16 years at the latest (intermediate clinical presentation) but had loss-of-function variants (out-of-frame and nonsense); (3) patients older than 18 years old; (4) patients using steroids: prednisolone or Deflazacort, but with variable doses. **Results:** Thirty patients were included in the study. Eighteen out of thirty patients (60%) had deletions, six out of thirty had duplications (20%), and six out of thirty (20%) had point mutations in the DMD gene. The current average age of the patients is 22.5 years (SD: ±3.9; range: 18-33 years). On average, the patients used corticosteroid therapy for 12.8 years (SD: ±4.9; range: 5-24 years). The average age of LOA was 12 years (SD: ±2.2 years; range: 7-15 years). Twenty patients (66.6%) had cardiomyopathy with an ejection fraction (EF) of less than 50%, and sixteen out of thirty patients (53.3%) required more than one heart medication. Eleven patients (36.6%) used nocturnal non-invasive ventilation, with three out of thirty patients (10%) requiring daytime non-invasive ventilation. Five patients (16.6%) had been hospitalized for pneumonia at least once. Eight patients (26.6%) experienced dysphagia, necessitating adjustments to the consistency of their diet. Seventeen patients (56.6%) had osteoporosis, and nine (30%) had fractures, with three (10%) experiencing more than one fracture. **Conclusion:** DMD is a disease with a high burden, and adult care is complex, requiring a multidisciplinary team. Special attention should be given to the cardiac, respiratory, and nutritional aspects and the development of osteopenia due to the long-term use of corticosteroids and hypomotility.

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Disorders of nerves, nerve roots and plexuses in Brazil: an epidemiological analysis over the last 5 years

Matheus Prado dos Santos[1], Rafael Hendrick Freitas de Paula[1], Luan Nascimento Pereira de Amorim[1], Fernando Silva Galvão[2], Daniel Lucas Araujo Ramos[2], Paola Carolina Santos Foinkinos[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil. [2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Matheus Prado dos Santos; Email: imatheusprado@gmail.com.

Background: Disorders involving nerves, nerve roots, and nerve plexuses are significant in the context of public health due to their high prevalence in the Brazilian population and their impact on the quality of life of affected individuals. These conditions can range from difficult-to-control pain to loss or reduced functionality in the affected region. In this context, it is necessary to analyze and discuss hospitalizations and deaths related to nerve, root, and nerve plexus disorders in Brazil. **Objective:** To analyze the epidemiological profile of disorders of the nerves, nerve roots, and nerve plexuses in Brazil from 2019 to 2023. **Methods:** This is an ecological, cross-sectional, descriptive, and analytical study with a quantitative approach, carried out by collecting secondary data from the Department of Informatics of the Unified Health System (DATASUS) from 2019 to 2023. The following variables were analyzed: number of authorizations for hospital admissions, average cost per admission, mortality rate, nature of care, number of admissions, and number of deaths. **Results:** From 2019 to 2023, there were 148,553 hospitalizations for nerve and nerve root disorders in Brazil, with an approximate annual average of 29,710 hospitalizations. According to region, during this period, there were 4,176 hospitalizations in the North (2.81%), 19,917 in the Northeast (13.40%), 67,589 in the Southeast (45.49%), 45,777 in the South (30.81%), and 11,094 in the Midwest (7.46%). The number of hospitalizations per year was 30,858 in 2019 (20.77%), 18,422 in 2020 (12.40%), 21,245 in 2021 (14.30%), 35,901 in 2022 (24.16%), and 42,127 in 2023 (28.35%). The average cost per hospitalization was 1,078.21 reais in the North, 932.15 reais in the Northeast, 621.03 reais in the Southeast, 664.76 reais in the South, and 553.56 reais in the Midwest. The mortality rate was 1.72 in the North, 0.35 in the Northeast, 0.88 in the Southeast, 0.15 in the South, and 0.14 in the Midwest. Regarding the type of care, 115,171 hospitalizations were elective, and 33,382 were emergencies. In terms of admissions by gender, there were 36,833 male patients (24.8%) and 111,720 female patients (75.20%). Finally, the number of deaths by gender was 458 in males (56%) and 359 in females (44%), totaling 817 deaths. **Conclusion:** During the study period, the North region, despite having the lowest number of hospitalizations, had the highest average cost per hospitalization and the highest mortality rate. This highlights the need for improvements in health services in the North of Brazil, showing a significant contrast when compared to the Southeast. It is therefore essential that health professionals are well-informed about the epidemiological profile of individuals affected by disorders of the nerves, nerve roots, and nerve plexuses in Brazil.

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Dissection of the common carotid artery and anatomical variant of anterior cerebral artery leading to ischemia in multiple cerebral vascular territories

João Vitor Assumpção Silva[1], Giovanni Caniçali Simões[1], André Luiz de Rezende[1], Marcele Schettini de Almeida[1], Sônia Maria César Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: João Vitor Assumpção Silva; Email: joaosilva7890@hotmail.com.

Case presentation: A 67-year-old man, previously diabetic, presented to the emergency department with sudden onset of complete right hemiplegia and anesthesia, global aphasia, and a National Institute of Health and Stroke Scale (NIHSS) score of 18, within the time frame for potential chemical thrombolysis. Immediate CT scan of the head showed no signs of recent hemorrhage or ischemia. Cardiological investigations with ECG and TTE revealed no significant abnormalities. Further MRI imaging revealed ischemic areas in the left middle cerebral artery territory, correlating with the patient's deficits, along with small areas of recent ischemia in both medial frontal lobes. MRA identified kinking, and thread-like flow of the left internal carotid artery (ICA) (1.8 cm distal to the carotid bulb), consistent with vessel dissection, and an eccentric thrombus causing stenosis in the mid-third of the left internal carotid artery (2.7 cm above the kinking). Additionally, we found a hypoplasia of the A1 segment with both anterior cerebral arteries (ACA) originating from the left ICA, explaining the bilateral frontal ischemias. We decided for treatment with dual antiplatelet therapy based on the service's experience in treating cervical vessel dissection-associated strokes. After just 4 days of hospitalization, the patient's NIHSS score improved to 3, with mild right paresis and hypoesthesia, and complete recovery of language. **Discussion:** Cervical artery dissection (CeAD) represents an uncommon cause of stroke, estimated at 2.6 to 3 cases per year. This condition is predominantly spontaneous or related to minor trauma. CeAD is responsible for about 25% of strokes in patients under 50 years old. Typically, CeAD occurs due to rupture of the vessel's intimal layer and formation of a mural thrombus. Both dual antiplatelet therapy and anticoagulation can be used for secondary prevention of CeAD-related strokes, but consensus on the optimal approach is still needed. The absence and hypoplasia of the A1 segment are among the most characterized and studied variations of the ACA. These anomalies of the ACA have a prevalence between 9% and 10%. This alteration results in the medial frontal lobes of both hemispheres being primarily irrigated by the carotid circulation from only one side. Consequently, ischemic events in these cases can present with bilateral frontal infarcts, as observed in our patient, leading to disabling deficits. **Final comments:** Stroke caused by carotid artery dissection and those related to anatomical variants of ACA are unusual. The case presented demonstrates the occurrence of both in a single patient, which adds interest for study. It is imperative for neurologists to be vigilant for such variations, especially when encountering strokes with atypical presentations. There is still no definitive consensus for secondary prevention of CeAD-related strokes, and case reports like this one are crucial for ongoing discussions among specialists.

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Disseminated cryptococcosis in immunocompetent patients: a therapeutic challenge

Valeska Forza Julio[1], Waldemar Carlos Barros de Algemiro[1], Juliana Bruneli Secchin Algemiro[1], Yasmin Oliveira Gil de Almeida[1], Ramana Scardua Jonas[1], Thiago Caetano Valladão de Azeredo[1], Ricardo Coelho Miranda[1], Marcos Paulo Travaglia[1], Thais Oppenheimer Pitanga de Rezende[1]

[1]Hospital Evangélico de Cachoeiro do Itapemirim, Cachoeiro de Itapimirim ES, Brazil.

Correspondence: Valeska Forza Julio; Email: valeskafj@hotmail.com.

Cryptococcosis is a fungal disease caused by the inoculation of Cryptococcus sp, primarily affecting the lungs and brain of immunocompromised individuals. Although uncommon, it can also occur in immunocompetent individuals, as will be discussed in the following study. The case involves a young patient, 32 years old, with no comorbidities, who was referred to the emergency room due to a severe, medication-refractory holocranial headache, associated with intractable nausea and vomiting, and drowsiness. The patient reported the onset of pain about two months ago with progressive worsening. Upon neurological examination at admission, the patient presented with drowsiness, mental confusion, generalized weakness, hyperreflexia of the lower limbs with a bilateral triple flexion plantar response, Hoffmann's sign on the right, no alteration in superficial or deep sensitivity, and absence of meningeal signs. Cranial magnetic resonance imaging revealed countless supra and infratentorial cystic lesions. The diagnosis of disseminated cryptococcosis was made through cerebrospinal fluid puncture, skin biopsy, and lung biopsy. Specific treatment was initiated 10 days after admission (following fungal culture growth on the slide) with amphotericin B and fluconazole 1,200 mg/day. Currently, the patient remains hospitalized under the care of the medical team.

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Dissemination of leptomeningeal non-Hodgkin lymphoma in a patient with Sézary syndrome: a case report

Isabela da Costa Rodrigues[1], Leonardo Rafael Kayser Torres dos Reis[1], Ivo Marcos Darella Lorenzin Fernandes Neto[2], Eduarda Basso Badalotti[2], Angelo Marcelo Wosniacki Filho[2], Gabriel Angelo Garute Zenatti[2], Renato Enfler Iachinski[2], João Luis Corso Bandeira[2], Antonio Carlos de Andrade Soares[2]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Fundação Hospitalar São Lucas, Cascavel PR, Brazil.

Correspondence: Isabela da Costa Rodrigues; Email: isacostarod.2001@gmail.com.

Case presentation: Male, 41 years old, with Sézary syndrome (a variant of Non-Hodgkin Lymphoma), currently on Interferon, Prednisone, and Doxepin, manifesting retro-orbital headache and paravertebral lumbar pain, associated with anuria and global paresis predominantly in the lower limbs. Symptoms began 5 days prior, and presented reduction in the level of consciousness since the previous day. Physical examination revealed xerostomia, drowsiness and confusion, dysdiadochokinesia, intention tremor, proportional paraparesis in the lower limbs (4/5), and patellar hyporeflexia. Cranial computed tomography (CT) scan with any particularities, and cerebrospinal fluid (CSF) showed xanthochromia, high red blood cell concentration, and lymphocytic pleocytosis, leading to the hypothesis of meningitis and encephalitis. The patient was empirically treated with Ceftriaxone, later switched to Tazocin, and Acyclovir. Two days later, with progressive deterioration in the level of consciousness, the patient was transferred to the intensive care unit, remaining drowsy, without effective communication, and with global hyporeflexia, experiencing a single seizure. Developed respiratory failure 5 days after, requiring endotracheal intubation. A new CSF analysis revealed the presence of malignant cells, suggesting neoplastic dissemination, and a cranial magnetic resonance imaging (MRI) showed diffuse leptomeningeal enhancement and thickening of the cranium, cervical, and dorsal spine. In light of this suspicion, the oncology team was contacted to repatriate the patient to the institution where he was undergoing treatment. **Discussion:** Sézary syndrome (SS) is a chronic lymphoproliferative disorder, part of the group of cutaneous T-cell lymphomas. Its rare, most commonly characterized by skin lesions, scaly eruptions, and pruritus. Although uncommon, metastasis can occur, and particularly, the central nervous system is a rare site for such dissemination. Diagnosis and investigation of SS are primarily through blood tests and skin lesion biopsies. Staging is based on the assessment of skin involvement and whether there is lymph node or visceral involvement. SS can be treated with a wide range of chemotherapeutic, immunotherapeutic, and biological modifying agents, which may or may not be combined with skin-directed therapy, including topical agents, phototherapy, and radiotherapy. **Final comments:** The SS subtype itself is a rare manifestation of T-cell lymphomas, and its leptomeningeal dissemination is a peculiar finding. Therefore, recognizing symptoms caused by CNS metastasis can be challenging, making it essential to remain vigilant for possible lymphoma metastasis and maintain a multidisciplinary follow-up beyond treatment, aiming to prolong patient survival despite the unfavorable prognosis when neurological involvement is present. If necessary, therapy using agents that penetrate the CNS can be pursued for a more lasting remission.

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Distal hereditary motor neuropathy with HSJ1 chaperone mutation presenting with movement disorders

Helio Afonso Ghizoni Teive[1], Matheus Gomes Ferreira[1], Samia Talise El Horr de Moraes[1], Luis Eduardo Borges de Macedo Zubko[1], Fernando Kok[2], Louis Viollet[3]

[1]Universidade Federal do Paraná, Complexo Hospital de Clínicas, Curitiba PR, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[3]University of Utah, Salt Lake City, UT, USA.

Correspondence: Samia Talise El Horr de Moraes; Email: samiatalise1@gmail.com.

Distal hereditary motor neuropathies (dHMNs) form a clinically and genetically heterogeneous group of disorders, characterized by muscle weakness and atrophy predominantly in the distal parts of the limbs due to the progressive degeneration of motor neurons in the spinal cord. This disease is caused by mutations in the HSJ1 (DNAJB2) gene, which encodes a co-chaperone member of the heat shock protein family. To describe a new phenotype of a rare recessive distal hereditary motor neuropathy with HSJ1 chaperone mutation, we present a case of a 45-year-old male patient who developed a progressive gait disorder with lower limb weakness, bradykinesia, followed by gait ataxia and falls since the age of 30. The family history revealed three affected relatives: a sister with cerebellar ataxia and peripheral neuropathy, a maternal cousin with early-onset parkinsonism, and a maternal uncle diagnosed with spinal muscular atrophy. Neurological examination showed asymmetric tetraparesis with muscle atrophy, predominantly in the lower limbs, associated with global areflexia, bradykinesia, muscle rigidity with cogwheeling, resting tremor (right hand), postural instability, and cerebellar gait ataxia. Extensive work-up revealed increased CSF protein, EMG signs of peripheral axonal motor neuropathy, muscle biopsy findings of denervation, and peripheral nerve biopsy showing hypertrophic neuritis. Brain and spinal cord MRI were normal. Tests for FRDA (Friedreich's ataxia), EAOH, SCAR1 (ataxia and ocular apraxia types 1 and 2), POLG, SCA (types 1, 2, 3, 6, 7, 8, 10, 12, 14, and 17), PARK2, and SMN1 genes were negative. Whole-exome sequencing demonstrated the presence of a homozygous mutation in the DNAJB2 gene (DNAJ/HSP40 homolog, subfamily B, member) (Chr2: 220.146.784, Variant: c.352+1G>A (IVS5+1G>A)). The same homozygous DNAJB2 mutation has been associated with a rare lower motor neuron disease, characterized by distal hereditary motor neuropathy, previously described in a Jewish family from Morocco. Gess et al. have shown that other DNAJB2 mutations are the cause of Charcot-Marie-Tooth disease type 2 (CMT2). In this Brazilian case report, we describe a new phenotype of this disease, which includes movement disorders, parkinsonism, and cerebellar ataxia.

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Distinct patterns of CNS damage along the spectrum of ATXN2-related disorders

Paulo Schneider Wolmer[1], Fabricio Castro de Borba[1], Thiago Junqueira Ribeiro de Rezende[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Paulo Schneider Wolmer; Email: paulowolmer@gmail.com.

Background: ATXN2 is located on cromossome 12 and encodes the transcription factor ataxin-2. There is a polymorphic CAG-rich region within ATXN2 coding region that encodes a polyglutamine tract in ataxin-2. The normal (CAG) length is <27 repeats. When the CAG repeat falls between 27 - 34 repeats, it is a proven risk factor for ALS (ALS-ATXN2); and when it presents with >34 repeats, it causes spinocerebellar ataxia type 2 (SCA2). The huge phenotypic differences between these 2 allelic conditions related to ATXN2 is not yet understood. **Objective:** To characterize and compare central nervous system structural signature in ALS-ATXN2, SCA2, sporadic ALS (sALS) and matched controls. **Methods:** To accomplish that, we first selected four age-and-sex matched groups: controls (n=34), sALS (n=17), ALS-ATXN2 (n=16) and SCA2 (n=17). Then we used automated segmentation tools to calculate the volumes of different CNS structures from T1-weighted MRI images of the patients: FastSurfer for cerebral cortex, deep brain nuclei and brainstem; CerebNet for cerebellum; and SCT (SpinalCord Toolbox) for the cross-sectional area of the first 4 cervical spinal cord levels. And lastly, we compared the mean volumes and areas of the structures between the groups. **Results:** In comparison to healthy controls, we found that: (1) sALS group presented only bilateral precentral gyrus thinning; (2) ATXN2-ALS group presented cervical spinal cord, medulla and midbrain atrophy; and (3) SCA2 presented spinal cord, the whole brainstem (medulla, pons, midbrain and superior cerebellar peduncle), the whole cerebellum (both white and grey matter structures) and deep brain nuclei (thalamus, caudate, putamen, pallidum, hippocampus, amygdala, accumbens area and ventral diencephalon) atrophy as well as cerebral cortex thinning (precentral, bilaterally; paracentral and parsopercularis in the left hemisphere and superior temporal gyrus in the right hemisphere). Direct comparison between sALS and ATXN2-ALS groups revealed no significant difference. All the significant results presented an FDR-adjusted p-value < 0.05 and effect-size >0.6. **Conclusion:** We have shown that (1) sALS, ATXN2-ALS and SCA2 have different structural CNS signatures; (2) ATXN2-ALS is characterized by predominant brainstem and spinal cord damage with relative cortical sparing; (3) in ATXN2-related disorders, the length of the repeat expansion drives the pattern of CNS abnormalities. Additional studies should be made to understand the biological bases of such structural differences.

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Distribution of antiepileptic drugs by specialized component of pharmaceutical assistance (ceaf): a time series analysis

Lucas Abreu Dias[1], Pedro Henrique de Vilhena[1], Alisson Rafael de Oliveira Pereira[1], Gabriel Augusto Nava Taveira[1]

[1]Universidade Federal de Lavras, Lavras MG, Brazil.

Correspondence: Pedro Henrique de Vilhena; Email: phdvilhena@gmail.com.

Background: Most of the hard outcomes in epilepsy can be avoided by controlling seizures with the continued use of antiepileptic drugs (AEDs). Some of these medications are distributed through the Specialized Component of Pharmaceutical Assistance (CEAF). Most states currently offer 8 drugs for the treatment of epilepsy: Primidone (PRM), Clobazam (CLB), Vigabatrin (VGB), Ethosuximide (ESM), Gabapentin (GBP), Lamotrigine (LTG), Topiramate (TPM) and Levetiracetam (LEV). **Objective:** To analyze the distribution of AEDs made available by CEAF and identify drugs with the highest tendency to be prescribed for each type of crisis. **Methods:** A retrospective study was conducted using data recorded in the SIA available by DATASUS regarding AEDs distributed by the CEAF since its creation in 2009. The analysis was performed using regression for each drug according to the subdivision into groups for the best indication of each AED: Drugs with action on generalized crises (LGT and PMS), focal (GBP and LEV), absence (ESM), and refractory (PRM, CBZ, GBV). **Results:** Over the 14 years of CEAF activity, the most distributed AED was LTG (33%), followed by GBP (30%), and TPM (23%). Except for VGB (APC: -20.6; CI95%: -32.6 to -7.2; p<0.005), all drugs showed an increasing distribution trend at the end of the study period. Although LEV was identified as the AED with the highest trend, more samples are needed to achieve statistical significance (APC: 729%; p=0.15). Considering the groups, LTG was the main choice for generalized crises (60%), with a change in trend in 2017 (APC 14.2; 95%CI: 10.6 to 25.7; p<0.001). ESM was the only drug evaluated in the absence group, with a trend change in 2012 (APC:

15; 95%CI: 10.6 to 19.5; p<0.001). Although LTG also has an effect on this type of crisis, it was placed in the group of generalized crises because of its higher prevalence. In the focal seizure group, the most widely distributed drug was GBP, with two points of change: 2012 (APC: 51.7; 95%CI 42.7 to 66.5; p<0.001) and 2016 (APC 19.4; CI95%: 14.8 to 23; p<0.001). It is important to highlight that LEV was included in the program only in 2016, and that GBP has a wide distribution because it is also effective in the treatment of neuropathic pain. In the refractoriness group, VGB and PRM maintained their stationery trends, whereas CBZ was the most distributed drug, with trend modification in 2012 (APC: 15.8; 95%CI: 12.5 to 18.1; p<0.001), although it is important to note that CBZ is also prescribed as a pure anxiolytic. **Conclusion:** Although there is an overlap of drug indications in the context of type of epilepsy, when separating groups according to their main indication, we found that LTG is the most distributed AED. LTG was also the most frequent option for generalized seizures, while ESM was the most prescribed for absence seizures, GBP among AEDs for focal seizures, and CBZ in the refractoriness group. Except for VGB, all the drugs showed an increasing distribution trend.

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DNMT1 mutation cerebellar ataxia in rare expression in a Brazilian family: a report of 2 cases

Emanuelle Campos Amaral[1], Danilo Lemes Reis[1], David Franca Ferreira Cruz[1], Maria Esther Lima Rios Evangelista[2], Fernanda Almeida Carvalho[1], Manuela Brandao Ferreira Teles[1], Eduarda Kei-lan Tavares Pierre[1], Diogo Rabelo de Lima[1], Fernando Luiz Westphal Filho[2], Marcus Vinicius Della Coletta[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Emanuelle Campos Amaral; Email: eca.med22@uea.edu.br.

Case presentation: M.L.M.S., a 59-year-old female, Italian descendent, hypertensive and diabetic, has been under neurological care for 12 years. 5 years ago, A positive genetic test revealed a heterozygous mutation in exon 21 of the DNMT1 gene (NM_001130823.1:c.1709C>T [p.Ala570Val]). She clinically presents with cerebellar ataxia, deafness, and narcolepsy. Magnetic resonance imaging shows a T2 hypointense signal in the globus pallidus bilaterally, as well as in the gray matter nuclei of the midbrain, and diffuse brain atrophy. Polysomnography demonstrates a multiple sleep latency test with a sleep onset time of less than 8 minutes, associated with 3 sleep-onset REM periods. On physical examination, she exhibited Babinski sign and bradykinesia on the left side, postural and resting tremor (predominantly on the left), and hyperactive deep tendon reflexes (+++/4). N.M., a 63-year-old male, italian descendent, diabetic, with a positive genetic test confirming a heterozygous mutation in the DNMT1 gene (NM_001130823.3:c.1709C>T [p.Ala570Val]). He underwent testing following the diagnosis of his sister, M.L.M.S. He has a similar clinical presentation, with deafness, narcolepsy, and cognitive impairment. Magnetic resonance imaging revealed no abnormalities. **Discussion:** DNMT1 is a widely expressed DNA methyltransferase maintaining methylation patterns in development, and mediating transcriptional repression by direct binding to HDAC. Despite international reports of de novo mutations, in Brazil there is only one report on the mutation that occurs in this patient. Cerebellar ataxia associated with deafness and narcolepsy has an autosomal dominant genetic character and a hereditary inheritance pattern. When with the Ala570Val mutation, it has late symptomatic expression (between 30 and 40 years), with dementia associated with the final process of pathological development. Associated with the same clinical picture, there are the Val606Phe and Gly605Ala variants, where the first has earlier symptoms. Regarding the patient's genealogy, only the cases presented have a positive genetic test, the other family members, even with deafness or movement disorders, tested negative for the variant. Furthermore, one M.L.M.S patient was using carbamazepine 200mg, prolopa HBS 100/25 and fluoxetine 20mg with the aim of reducing symptoms. **Final comments:** Given the above, the impact of genetic variation on the development of the disease is clear, as well as the existence of its hereditary pattern and its impact on human development. Furthermore, the need to establish a correct diagnosis is evident to define an objective prognosis and ensure the individual's minimum well-being.

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Do not neglect sudden paresthesia: a case report of cheiro-oral syndrome

Samia Talise El Horr de Moraes[1], Camila Emi Fujiwara Murakami[1], Caroline Link[1], Valmir Vicente Filho[1], Carolina Mibielli de Souza[1], Catarina Dantas Corrêa[1], Isabelle Pastor Bandeira[1], Cesar Minoru Toita Koga[1], Alana Bacelar Limeira Sales[1], Caio Cesar Diniz Disserol[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Alana Bacelar Limeira Sales; Email: alanabls@hotmail.com.

A 59 years old man was admitted in a stroke protocol with a history of sudden paresthesia in the left upper and lower lips and ipsilateral upper limb, as well as dysarthria. He had a previous medical history of hypertension, diabetes, and dyslipidemia. At admission, his vital signs were stable, and no other abnormality in the physical exam was identified. Head CT was performed and showed a small hyperdensity of less than 1 milliliter in the right thalamus. He had not been taking his medications for 40 days before the admission. Thus, after head and neck CT angiography and brain MRI with no suspicion of another etiology, it was concluded that it was an intracerebral hemorrhage due to uncontrolled hypertension. This case excels in exemplifying the commonly neglected cheiro-oral syndrome (COS), which is considered a pure sensory thalamic lacunar syndrome with symptoms that affect the face, hand and/or foot. Although classically associated with thalamic ischemic lesions, there are descriptions of hemorrhagic strokes and multiple different affected regions, including the brainstem, internal capsule, operculum, cortex, corona radiata and thalamus. Whilst a mild clinical stroke presentation with potential misleading diagnosis, early recognition and diagnosis is essential to institute adequate early treatment and secondary prophylaxis. Physicians should be aware of COS and consider a low threshold for investigation, considering the potential severity of its etiologies.

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Do Parkinson's disease subtypes impact an individual's ability during the functional task of boarding a bus?

Renan Alexandre Mregalote[1], Eveline Gonçalves Maki[1], Andressa Leticia Miri[1], Patrícia Gonçalves Broto[1], Amanda Lima Nogueira dos Anjos[1], Guilherme Lopes Barbosa da Silva[1], Larissa Laskovski[1], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Eveline Gonçalves Maki; Email: eveline.goncalves@uel.br.

Background: Parkinson's disease (PD) is a neurodegenerative, chronic, and progressive condition. Its clinical spectrum is broad, with both motor and non-motor symptoms affecting the functional independence of the population that suffers from it. Due to the heterogeneous nature of its symptoms, classifying PD into subtypes based on predominant symptoms may help understand if (and how) these subtypes impact the performance of functional tasks and locomotion, such as boarding a bus. **Objective:** To compare and assess the correlation between upper limb strength and ground reaction force during the task of boarding a bus prototype, as well as to investigate the impact of cognition and the fear of falling during the task, according to the PD clinical subtypes. **Methods:** A cross-sectional study involving 30 individuals with idiopathic PD divided into the rigid-akinetic (RA: n=19) and tremor-dominant (TD: n=11) subgroups, based on the Unified Parkinson's Disease Rating Scale. These individuals underwent cognitive assessment using the Mini-Mental State Examination, fear of falling assessment using the Falls Efficacy Scale-International, grip and pull strength analysis using dynamometry, ground reaction force analysis using a force platform, and the time taken to board a bus, both in single-task (ST) and dual-task (DT) conditions. A comparative analysis between disease subtypes and a correlation analysis between variables were conducted, with significance set at p<0.05. The study was approved by the Research Ethics Committee (CAAE: 23272819.8.0000.5231). **Results:** There were no significant differences in the clinical characteristics and the variables analyzed for boarding the bus between the RA and TD subgroups. Correlation analysis revealed correlations between cognitive function and grip strength during ST (rho=0.759; p<0.001) and DT (rho=0.707; p=0.001), as well as pull strength during ST (rho=0.691; p=0.001) and DT (rho=0.703; p=0.001). Additionally, correlations were found between having the fear of falling and the time taken to board the bus during ST (rho=0.662; p=0.002) and DT (rho=0.606; p=0.006), as well as maximum grip strength (r=-0.475; p=0.04) and maximum pull strength (r=-0.479; p=0.038). **Conclusion:** There were no differences between PD clinical subtypes in performing the task of boarding a bus. The findings revealed that worse cognitive function was associated with poorer upper limb strength, particularly in the RA subtype. Furthermore, greater fear of falling was associated with lower maximum upper limb strength and longer boarding task execution time. These findings underscore the importance of considering not only motor manifestations but also cognitive factors in performing functional tasks, regardless of PD subtype.

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Do sleep distubances impact the qualrity of life and cognition similarly in men and women with Parkinson's disease?

Rogério José de Souza[1], Amanda dos Santos Siqueira[1], Luísa Vigiani Cassiano[1], Nathalia de Oliveira Franco[1], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Rogério José de Souza; Email: rogerio.jose.souza@uel.br.

Background: Much research has been conducted on the influence of sex-related differences on Parkinson's disease (PD). While men are approximately 1.5 to 2 times more likely to develop the disease and exhibit more tremors, rigidity, cognitive decline, and autonomic dysfunctions, women report more postural instability, depression, anxiety, and pain. Despite sleep disturbances being very prevalent in patients with PD, little is known about the differences when it comes to the quality of sleep experienced between men and women and the impact of the differences if any on their quality of life and cognition. **Objective:** To compare the quality of sleep between men and women with PD and to assess its correlation with the

quality of life and cognition according to sex-related differences. **Methods:** This is a cross-sectional study composed of 73 individuals (50 men and 23 women) diagnosed with idiopathic PD, at a mild to moderate stage, and without cognitive deficits. Quality of sleep was assessed using the Parkinson's Disease Sleep Scale (PDSS-2), the Pittsburgh Quality of sleep Index (PSQI), and the Epworth Sleepiness Scale (ESS). Individuals scoring above 5 on the PSQI were classified as having "poor sleep." Cognition and quality of life were evaluated using the Montreal Cognitive Assessment (MoCA) and the Parkinson's Disease Quality of Life Scale (PDQ-39), respectively. A significance level of 5% was adopted for the analyses. The study was approved by the Research Ethics Committee (CAAE: 54383021.8.0000.5231). **Results:** Regarding sleep classification, 72% of men and 52.2% of women were classified as having poor sleep quality. There was no difference in the total scores of the sleep scales between men and women. The only difference detected was that men had worse scores in the daytime dysfunction domain of the PSQI (p=0.015). In the correlation analysis between sleep and quality of life, there was a moderate correlation between the total PDSS-2 and PDQ-39 in both groups (Men: r=0.66; p< 0.001 - Women: r=0.61; p=0.002), and between the PSQI and PDQ-39 only in men (r=0.46; p=0.001). Concerning daytime sleepiness and the quality of life, there was a moderate correlation between the ESS and PDQ-39 in women (r=0.51; p=0.013) and a weak correlation in men (r=0.37; p=0.007). Regarding cognition, a moderate correlation was observed between the total PDSS-2 and the naming domain of the MoCA in men (r=-0.47; p<0.001) and the total PSQI with the abstraction domain in women (r=0.64; p=0.001). **Conclusion:** In this study, a difference was observed in the daytime dysfunction domain, with men scoring worse compared to women, indicating the impact of sleep on daily functionality. Additionally, men and women with better quality of sleep exhibited signs of a better quality of life. Finally, regarding cognition, men with better quality sleep performed better in the naming domain, while women with better quality sleep performed better in the abstraction domain.

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Does antidepressant use interfere with multiple sclerosis patient's outcome?

Mariana Martins Dantas Santos[1], Renata lameira Oliveira[2], Paulo Matheus Casal de Souza[2], Cintia Ramari[3], Carlos Bernardo Tauil[4]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Escola Superior de Ciências da Saúde, Brasília DF, Brazil.
[3]University of Hasselt, Faculty of Rehabilitation Sciences, REVAL, Belgium.
[4]Universidade de Brasília, Brasília DF, Brazil.

Correspondence: Mariana Martins Dantas Santos; Email: marianadantassantos@gmail.com.

Background: Depression is commonly associated with multiple sclerosis (MS), affecting between 15-50% of these patients. This co occurrence highlights a potential shared pathophysiology between these two conditions, characterized by inflammatory processes, structural abnormalities, and functional and genetic impairs. Recently, the correlation between depression and MS has gained increased significance, with emerging research indicating that antidepressant treatments may also potentially enhance disease-modifying effects in MS. Which is shown by a reduction of the frequency of MS flare-ups and the recurrence of symptoms, highlighting an important approach for the management of this disease. **Objective:** To assess the prevalence of MS patients using both disease-modifying therapies (DMTs) and antidepressants, and outcomes of MS patients who use antidepressants and those who do not. **Methods:** Data from 241 individuals using DMTs sourced from the Collaborative Latin American Database for Multiple Sclerosis (BRANDO) was analyzed. Outcomes included marital and employment status, sleep disturbances, sphincter control, sexual dysfunction, Expanded Disability Status Scale (EDSS), Nine Hole Peg Test - NHPT, walking capacity (Timed 25-Foot Walk Test - T25FWT), cognitive processing speed (Symbol Digit Modality Test - SDMT), and fatigue (Modified Fatigue Impact Scale - MFIS). Depression and anxiety were assessed by the Beck Depression and Anxiety Inventory (BDI and BAI) and Hospital Anxiety and Depression Scale (HADS-A and HADS-D). Chi-Square and Independent T-tests compared antidepressant users and non-users. **Results:** Out of the 241 patients, 129 (53%) used both antidepressants and DMTs. Data analysis revealed that these individuals had significantly higher EDSS scores (p=0.03), more prevalent sleep disturbances (p=0.013), and higher fatigue levels (MFIS) (p<0.001) compared to non-users of antidepressants. No significant differences were found in demographic outcomes or motor and cognitive functions. Depressive symptoms were significantly higher in the antidepressant user group. **Conclusion:** While recent studies suggest that antidepressants may have a beneficial effect on MS symptoms, our sample revealed that MS patients using antidepressants presented greater disability. However, this analysis could be biased by issues such as the characteristics of the population, potential side effects of these medications, and the timing of evaluations. Therefore, given the frequent co-occurrence of MS and depression, it is crucial to survey the long-term outcomes of these patients. Considering this, longitudinal studies should be performed.

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Does COVID-19 impact the modulation of the cardiac autonomic nervous system in patients with systemic arterial hypertension?

Pedro Igor Lustosa Roriz[1], Heitor Fernandes Silveira Cavalini[1], Ádrya Aryelle Ferreira[1], Camila Almeida Sá[1], Joice de Souza Batista[1], Matheus Sobral Silveira[1], Edelvita Fernanda Duarte Cunha[1], Leandro Campos de Brito[2] , Victor Ribeiro Neves[1]

[1]Universidade de Pernambuco, Petrolina PE, Brazil.
[2]Oregon Health & Science University, Portland, OR, USA.

Correspondence: Pedro Igor Lustosa Roriz; Email: pedroilroriz@gmail.com.

Background: Systemic arterial hypertension (SAH) when associated with COVID-19 increases the risk of morbidity and mortality. Heart rate variability (HRV) is considered an important predictor of clinical outcomes. An adequate HRV reflects good integrity of the autonomic nervous system. **Objective:** To evaluate the cardiac autonomic nervous system through HRV in hypertensive patients infected or not with COVID-19. **Methods:** This is an observational and cross-sectional study. Hypertensive patients, over 18 years of age, of both sexes, with or without a diagnosis of COVID-19 were included and grouped into: Group 1 (G1) - hypertensive patients diagnosed with COVID-19; and Group 2 (G2) - hypertensive patients without a diagnosis of COVID-19. All participants underwent a resting electrocardiogram. Recording of electrocardiogram RR intervals was performed in the supine position for 10 minutes. The time series of 256 points with greater stability was used to obtain HRV indices, acquired using linear (time and frequency domain) and non-linear (Symbolic and Entropy analysis) methods. Thirty-three patients were included in the study. **Results:** G1 was predominantly composed of white individuals (47.1%), male individuals (58.8%), sedentary (58.8%) and without associated diseases (52.9%). G2 was predominantly composed of black individuals (50%), male individuals (50%), sedentary individuals (62.5%) and patients with associated diseases (56.2%), especially diabetes and osteoarthritis. There was no statistical difference between the groups for these variables. Regarding the symptoms presented by hypertensive patients with COVID-19, we highlight headache, fever, cough, runny nose and dyspnea, which affected more than 30% of patients in G1. Regarding HRV indices in the supine position, there was no statistical difference between the groups. **Conclusion:** According to the data presented, it is suggested that COVID-19 does not affect the modulation of the cardiac autonomic neural system in hypertensive patients who presented mild symptoms of the disease.

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Does Ginkgo Biloba extracts really have any impact on patients with moderate to low acute ischemic stroke (NIHSS ≤15) ?: an updated meta-analysis of Chinese randomized clinical trials

Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Leonardo B. Oliveira[2] , David Abraham[3], Gustavo de Oliveira Almeida[4], Gabriel de Almeida Monteiro[5], Ana Beatriz Santos[6], Pedro Henrique Carvalho Leite Romeiro[7], Sávio Batista[8], Márcio Yuri Ferreira[9], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Universidade Estadual de Ponta Grossa, Ponta Grossa, PR, Brazil.
[3]Universidade Federal do Amazonas, Manaus AM, Brazil.
[4]Universidade Federal do Triângulo Mineiro, Departamento de Medicina, Uberaba MG, Brazil.
[5]Universidade Federal do Ceará, Departamento de Neurocirurgia, Sobral CE, Brazil.
[6]University of Costa Rica, School of Medicine, San Jose, Costa Rica.
[7]Centro Universitário de Maceió, Maceió AL, Brazil.
[8]Universidade Federal do Rio de Janeiro, Faculdade de Medicina, Rio de Janeiro RJ, Brazil.
[9]Lenox Hill Hospital/Northwell Health, Department of Neurosurgery, New York, NY, USA.

Correspondence: Ocílio de Deus da Rocha Ribeiro Gonçalves; Email: ocilio41@gmail.com.

Background: Ginkgo Biloba (EGb) leaf was used in treating cerebrovascular diseases due to its neuroprotective effects in Chinese Traditional Medicine. However, the potential benefits of Ginkgo Biloba extracts (GBE) in patients with moderate to low acute ischemic stroke (AIS) are still controversial. **Objective:** Therefore, we aimed to conduct an updated meta-analysis to compare GBE to other pharmacological interventions in stroke therapy. **Methods:** We systematically searched PubMed, Embase, and Cochrane for randomized clinical trials comparing GBE to placebo or standard treatment. The primary outcome of interest was functional outcome based on mRS 0-1 scale. Secondary outcomes were: functional outcome based on mRS 0-2 scale, change on MoCA total score and all-cause mortality. Comparative analysis with odds ratio (RR) and mean difference (MD) with 95% confidence intervals (CI) were used, and p-value <0.05 was considered significant. **Results:** Six randomized controlled trials (RCTs) comprising 8361 patients were included in this meta-analysis. Three RCTs, encompassing 4,840 patients, reported data for excellent functional outcome (mRS 0-1). GBE was associated with an improvement in functional outcomes compared to the control group, with a low heterogeneity in the results (RR: 1.17; 95% CI 1.11-1.24; p<0.001; I^2=0%). Regarding cognitive outcome, MoCA total score was reported by three studies involving 3,698 patients. MoCA total score was overall higher in the experimental group after GBE therapy (MD: 0.77; 95% CI 0.12-1.42; p=0.02; I^2=54%). All-cause mortality was reported by three studies, including 4,837 patients. GBE did not improve mortality rate compared to the control group (RR: 0.63; 95% CI 0.09-4.52; p>0.05; I^2=85%). **Conclusion:** GBE significantly improved post-AIS functional and cognitive outcomes based on mRS 0-1 scale and MoCA total

score, respectively, with no significant impact on overall mortality in these patients. In summary, our findings suggest that GBE alone has a positive impact in patients with AIS with low NIHSS. Larger randomized controlled trials outside of China are needed to adequately assess the efficacy and safety of GBE for AIS in different populations with bigger sample sizes.

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Does midbrain atrophy distinguish progressive supranuclear palsy from frontotemporal dementia? A multimodal neuroimaging investigation

Leonardo Cruz de Souza[1], Sarah Teixeira Camargos[1], Mauro César Quintão e Silva Cunningham[1], Vinícius Ribeiro Jeunon[1], Natália Pessoa Rocha[2], Antônio Lúcio Teixeira[2], Thiago de Oliveira Maciel[3], Elisa de Paula França Resende[1], Francisco Cardoso[1], Paulo Caramelli[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[2]UTHealth, Houston, USA.
[3]Quantum Research Center, Abu Dhabi, UAE.

Correspondence: Leonardo Cruz de Souza; Email: leocruzsouza@hotmail.com.

Background: The distinction between progressive supranuclear palsy (PSP) and behavioural variant frontotemporal dementia (bvFTD) is a clinical challenge, due to overlapping clinical, cognitive and behavioural features. Midbrain atrophy is observed in PSP, but the diagnostic value of patterns of midbrain atrophy to differentiate bvFTD from PSP has not been explored so far. **Objective:** To investigate whether quantitative and qualitative measures of midbrain atrophy contribute to the clinical differentiation between PSP and FTD. **Methods:** We included the following groups of participants: healthy controls (n=19; 6 male/13 female; mean age=63.1 years-old), PSP-Richardson' syndrome (n=20; 10 male/10 female; mean age=67.6 years-old), bvFTD (n=19; 11 male/8 female; mean age=66.5 years-old) and Parkinson's disease (PD; n=12; 7 male/5 female; mean age=61.9 years-old). Participants underwent 3T brain MRI. Quantitative and qualitative measures (Hummingbird sign rating scale [HBS-RS], global midbrain atrophy [GMA] and midbrain to pons ratio) were calculated with Radiante software. Brainstem measures were extracted with FreeSurfer. Raters were blind to the clinical diagnosis. **Results:** Groups did not differ in terms of age and sex distribution. Disease duration did not differ between PSP and bvFTD patients. Compared to controls, PSP and bvFTD had lower HBS-RS and GMA scores. Patients with PSP had significantly lower midbrain measures than PD. The HBS-RS, the GMA, the midbrain/pons ratio and the brainstem volumes distinguished PSP from FTD with 73%, 67%, 75% and 82% accuracies, respectively. Imaging measures did not correlate with clinical parameters within groups. **Conclusion:** In this multimodal neuroimaging investigation, both quantitative and qualitative measures of midbrain atrophy provided modest accuracy to distinguish PSP from FTD, thus reinforcing the complex interface between both diseases.

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Does posterior callosotomy reduces secondary bilateral synchrony? A proof-of-concept video-EEG study

William Alves Martins[1], Taiane Pigozzo[1], Eliseu Paglioli[1], Thomas More Frigeri[1], Rafael Paglioli[1], Fenanda Schuh Martins[1], Vicenzo Zarpellon[1], Andre Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: William Alves Martins; Email: walvesm.br@gmail.com.

Background: Drop attacks represent a severe expression of epilepsy, with frequent sudden falls, leading to significant injuries and morbidity. Such seizures are thought to arise from sudden bilateral motor cortex synchronization. One strategy for such seizures is to section the callosal fibers in an attempt to render synchronized discharges. A recently new surgical strategy is to section the posterior half of the corpus callosum, therefore sparing frontal fibers. However, no study has analyzed if section of the posterior corpus callosum indeed leads to diminished secondary synchronization. **Objective:** To study the rate of epileptic discharges and, especially, bilateral secondary synchronization. **Methods:** From 2014 to 2018, 12 patients with refractory epilepsy who underwent Posterior callosotomy at the Porto Alegre Epilepsy Surgery Program were analyzed. All had been previously discussed in a multidisciplinary conference and underwent high resolution MRI and prolonged Video-EEG (pre and post-callosotomy). Post-surgical Video-EEG lasted for at least 24 hours. We analyzed pre and post-callosotomy EEG according to the following variables: frequency of number/rate of synchronized discharges in 24 hours, regionalization of discharges, morphology of discharges, as well as frequency and morphology of electrographic and clinical seizures. In addition, we analyzed changes in the background rhythms. **Results:** Mean age at surgery was 18.8 ± 12.2 years and eight patients were female. Secondary bilateral synchrony (SBS) decreased from 2122±3.897.75 to 2±298.75 (p=0.002; Wilcoxon). Generalized EEG seizures significantly reduced post-surgically (62±177 x 1±7.5; p=0.002, Wilcoxon). Tonic seizures during sleep and rate of burst suppression also decreased (p=0.01 and 0.028, respectively). There was no significant change in relation to localization of discharges, morphology or background rhythms. **Conclusion:** Posterior callosotomy is a highly effective surgical treatment for drop attacks and greatly improves electroencephalographic patterns post-surgically. As demonstrated by our study, post-surgical EEG showed significantly less synchronized discharges and generalized EEG seizures. Further studies may analyze if such changes in epileptogenicity lead to improve cognition and a switch from pathogenic to healthy networks dynamics.

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Does postural control influence kinematic parameters of normal and dual-task gait in individuals with Parkinson's disease?

Sara Garcia Amaro dos Reis[1], Rogério José de Souza[1], Alessandra Cattaneo Estrada Melanda[1], Hayslenne Andressa Gonçalves de Oliveira Araújo[1], Amanda Lima Nogueira dos Anjos[1], Naiara Cândido Ferreira[1], Gabriela Casagrande Zago[1], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Sara Garcia Amaro dos Reis; Email: saragarcia.reis13@gmail.com.

Background: Postural instability and gait disturbances are among the most disabling symptoms in Parkinson's disease (PD), leading to a negative impact on quality of life and daily activities, with a significant increase in the risk and occurrence of falls in individuals with PD. **Objective:** To correlate postural control with kinematic parameters of gait in individuals with PD. **Methods:** This is a cross-sectional study, including 24 individuals diagnosed with idiopathic PD, in mild to moderate stages, without cognitive deficits, and attending a specialized neurofunctional physiotherapy outpatient clinic. Kinematic gait assessment was performed using a 3D motion analysis system (Optitrack), under conditions of single-task and dual-task gait. The parameters analyzed in one gait cycle were: step length and width, cadence, maximum and minimum ankle angles, vertical and medio-lateral center of mass amplitude, and gait speed. Postural control was assessed using the Mini Balance Evaluation Systems Test (Mini-BESTest). Correlations between the variables obtained in the kinematic assessment and the Mini-BESTest were analyzed. The significance level adopted was 5%. The study was approved by the Research Ethics Committee (CAAE: 72474923.3.0000.5231). **Results:** Moderate correlations were found between the total Mini-BESTest score and the variables of normal gait: step length 1 (r=0.41; p=0.04), step length 2 (r=0.42; p=0.03), and average speed (r=0.41; p=0.04), as well as the variables of dual-task gait: step length 1 (r=0.41; p=0.04) and maximum ankle amplitude (r=0.45; p=0.02). Detailed analysis by domains of the Mini-BESTest revealed a moderate correlation of the "anticipatory adjustments" domain with step width 1 in normal gait (r=-0.53; p<0.01). For the "reactive postural control" domain, there was a moderate correlation with maximum ankle amplitude in normal gait (r=0.54 and p<0.01) and with maximum ankle amplitude in dual-task gait (r=0.45; p=0.02). The "sensory orientation" domain showed a moderate correlation with step length 1 in normal gait (r=0.45; p=0.02) and step length 2 in dual-task gait (r=0.41; p=0.04). Finally, there was a moderate correlation between the "dynamic gait" domain and the variables in normal gait: step length 1 (r=0.47; p=0.02), step length 2 (r=0.42; p=0.03), and minimum ankle amplitude (r=-0.54; p=0.01). **Conclusion:** Individuals with PD with worse postural control presented worse kinematic parameters of gait, especially step length and speed in normal gait, as well as step length and maximum ankle amplitude in dual-task gait. Understanding that postural instability alters gait patterns in individuals with PD assists in the search for effective methods in the rehabilitation of these outcomes.

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Does the upper limb strength exerted to board a bus correlate to gait in individuals with Parkinson's disease?

Larissa Floripes de Souza Merética[1], Andressa Leticia Miri[1], Patrícia Gonçalves Broto[1], Hayslenne Andressa Gonçalves de Oliveira Araújo[1], Maria Eduarda Brandão Bueno[1], Guilherme Lopes Barbosa da Silva[1], Larissa Laskovski[1], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Larissa Laskovski Dal Molin; Email: larilask@gmail.com.

Background: Parkinson's disease (PD) is characterized by motor symptoms that affect mobility, transfers, balance, gait, and posture. Gait, in particular, is marked by increased cadence, reduced speed, shorter steps and stride length, a longer double support phase, and asymmetry of the upper and lower limbs. In patients with PD, the upper limb functionality is impaired by reduced muscle strength, altered manual dexterity, and deficits in motor coordination during bimanual activities. These dysfunctions limit functional mobility and result in restrictions when it comes to social participation as well as difficulties in performing daily tasks. Public transportation plays a significant role in urban mobility, and thus adequate infrastructure to ensure better accessibility and safety is crucial so as to avoid socially excluding this population. **Objective:** To correlate upper limb strength during the functional task of boarding a bus to gait in individuals with PD. **Methods:** This cross-sectional study included 24 participants. Data on disease staging and progression was collected using the Hoehn & Yahr and the Unified Parkinson's Disease

Rating Scales, respectively. Additionally, the following were administered: 1) a semi-structured questionnaire with clinical, sociodemographic, anthropometric questions and the frequency in which public transportation is used; 2) a cognitive state assessment using the Mini-Mental State Examination; 3) a fear of falling assessment using the Falls Efficacy Scale; 4) an upper limb grip and pull strength evaluated using two dynamometers attached to the handrails of a prototype bus similar to those used in the city of Londrina, Parana, measuring variables like maximum voluntary contraction, grip, and pull strength; and 5) a kinematic gait evaluation using a 3D motion analysis system, collecting variables such as step length, step width, cadence, and gait speed. Data analysis involved Pearson correlation testing with statistical significance set at $p<0.05$. The study was approved by the Research Ethics Committee (CAAE: 23272819.8.0000.5231). **Results:** A moderate and statistically significant correlation was observed between the maximum voluntary left-hand grip strength and the step length ($p\leq0.011$; $r=0.511$). In the functional task of boarding the bus, there was a moderate and significant correlation between the left-hand's grip strength and gait cadence ($p=0.009$; $r=0.523$) as well as between the right-hand's pull strength and the step width ($p=0.040$; $r=0.422$). **Conclusion:** The greater the maximum left-hand grip strength, the longer the step length and the higher the gait cadence. This result brings research closer to clinical practice, highlighting the importance of a thorough evaluation and upper limb training to ensure greater independence for individuals with PD.

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Don't close your eyes for this diagnosis: prevalence and characteristics of Jeavons' syndrome in an epilepsy monitoring unit in Southern Brazil

Leticia Bassani Devens[1], Glauco Kody Nagata[1], Pedro Simões dos Santos Pilau[2], Andressa de Oliveira Felício[2], Maria Paula de Carli Hanel[2], Stella Duarte Pinto[2], Natália Balbinot Zanin[2], Helena Pacheco Helms[2], William Alves Martins[1]

[1]Hospital São Lucas, Departamento de Neurologia, Porto Alegre RS, Brazil.
[2]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Leticia Bassani Devens; Email: leticiabassanidevens@gmail.com.

Background: Jeavons' syndrome (JS) or epilepsy with eyelid myoclonia is one of the most distinct generalized idiopathic epilepsies, corresponding to 7.9 to 12.3% of genetic epilepsies with predominant early age onset and affecting women more. Typically, this syndrome is manifested by a triad of myoclonia with or without absence, eye closure elicited paroxysms in the EEG and photosensitivity. Regarding semiological findings, the eyelid myoclonus occurs several times a day, mainly after eye closure. Patients also commonly experience generalized tonic-clonic seizures and myoclonus. Furthermore, intense blinking may elicit epileptic focus and can be useful for diagnostic ends. Finally, valproic acid and levetiracetam stand out among the medications for the seizure control. **Objective:** To analyze different presentations of patients diagnosed with JS in the epilepsy monitoring unit (EMU) in Hospital São Lucas da Pontifícia Universidade Católica do Rio Grande do Sul, as well as provide a brief literature review of the subject. **Results:** From 2021 to 2024 6 (1.63%) patients out of 367 patients admitted to the EMU of the Porto Alegre Epilepsy Surgery Program received a final diagnosis of JS. Four (67%) patients were younger than 20, predominantly female (67%). Furthermore, in 16% of cases the seizure onset was within the first 28 days of life, 33% with onset in infants and 50% had their first seizure throughout adolescence. None had a history of febrile seizures. Seizures occurred weekly for 4 (67%) patients and monthly for 2 (33%). Moreover, half the patients with weekly seizures progressed with daily absent seizures. Video-EEG showed generalized maximum posterior spike-wave complexes (33%) and polyspikes (67%) upon eye-closure and photosensitivity (83%), more prominently after medication tapering. Mild intellectual disability was present in 3 (50%) patients. Regarding semiology, 67% of them manifested as generalized bilateral tonic-clonic seizures, with 75% of thsese starting with eyelid myoclonus. The remaining was manifested by myoclonus and absent seizures. For treatment, 50% of patients were on valproic acid combined with lamotrigine and clobazam, while the other half used levetiracetam and clobazam. **Conclusion:** This case-series provides data on JS responsible for a significant share of genetic generalized seizure in the EMU. Understanding JS is crucial for accurate differential diagnosis.

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Dopa-responsive cervical dystonia: case report

Lavínia Damacena Perin[1], Lucas Grobério Moulim de Moraes[1], Vinícius Andreata Brandão[1], Jovana Gobbi Marchesi Ciríaco[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Lavínia Damacena Perin; Email: laviniaperin23@gmail.com.

Case presentation: A 21-year-old patient, with no complications related to childbirth, no previous comorbidities or family history of neurological diseases, started at around 11 years of age with involuntary movements of cephalic torsion to the right, usually associated with emotional stress, using musical instruments or physical exertion. She reported a clinical diagnosis of tics and myofascial pain triggered by anxiety for many years, without improvement with the use of fluoxetine 40mg/day, amitriptyline 25mg/day and carbamazepine 200mg/day. Electroneuromyography showed arrhythmic activity with irregular tonic alternation in the right splenius, levator scapulae, semispinalis and trapezius muscles and in the left sternocleidomastoid and semispinalis muscles. She reported a progressive worsening in the frequency and intensity of the crises and at the age of 20 she began to report intermittent pain and fixed posture, regardless of the triggering factor. Physical examination showed focal dystonia affecting the cervical muscles: right splenium, trapezius and scalene, and to a lesser extent the left semispinal, sternocleidomastoid and trapezius muscles. In view of the early onset of dystonia, a therapeutic trial was carried out with levodopa + benserazide 100/25mg twice a day, with a satisfactory response. Laboratory and neuroimaging tests ruled out iron deposition and copper metabolism diseases. The patient is currently being monitored on an outpatient basis, having already stopped antidepressant and anxiolytic medication. She reports complete improvement in pain and involuntary cervical movements. **Discussion:** In this report, we present the case of a patient with childhood onset cervical dystonia that was misdiagnosed as tics and myofascial pain triggered by anxiety, responsive to levodopa in a therapeutic trial. Cervical dystonia is a disorder characterized by involuntary contractions of the cervical musculature, resulting in abnormal positioning of the head, usually painful. When diagnosed and treated incorrectly, it becomes disabling and affects the person's quality of life. In most cases, the cause is unknown, and symptoms begin between the seventh and eighth decades of life, there is no cure and it requires long-term treatment. However, with the discovery of mutations in specific genes, more than 25 subtypes with distinct clinical manifestations have been mapped. Among them, a rare subtype, whose first symptoms appear in childhood and show a sustained response to levodopa, is DYT5, also known as Dopa-Responsive Cervical Dystonia (DRD). The clinical manifestations begin in childhood or adolescence with focal dystonia, with diurnal fluctuations and marked nocturnal worsening, which can impair sleep. **Final comments:** This case highlights the importance of levodopa testing for patients with childhood onset dystonia, due to its diagnostic and therapeutic potential, allowing patients to evolve with a good quality of life, without motor complications.

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Dorsal root ganglia and spinal cord imaging in genetic and acquired sensory neuronopathies

Rafaella do Rosario Tacla[1], Thiago Junqueira Ribeiro de Rezende[1], Paulo Schneider Wolmer[1], Gustavo Moreira Jarola[1], Alberto Rolim Muro Martinez[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Rafaella do Rosario Tacla; Email: taclarafaella@gmail.com.

Background: The etiology of sensory neuronopathies (SN) can be divided into two key groups: acquired and genetic. **Objective:** We designed an MRI-based study to compare volumetry of the dorsal root ganglia (DRG) and structural imaging of the spinal cord (SC) across healthy controls, patients with acquired SN and patients with Friedreich's ataxia (FRDA), the prototypical genetic SN. We also assessed clinical correlates of these imaging parameters. **Methods:** Sixty-two subjects (21 healthy controls, 20 acquired SN patients, and 21 FRDA patients) underwent 3T MRI acquisitions including: Diffusion-weighted imaging and T2WI of the cervical spine as well as PROSET isotropic T2WI of the lumbosacral spine to assess the DRG. Analyses correlating MRI metrics and clinical parameters were performed. Sex, age and body-mass index were used as covariates in between-group comparisons. **Results:** Patients with FRDA, but not acquired SN had widespread (C1-T1) SC atrophy relative to controls. Fractional anisotropy reduction in the dorsal columns was observed in both groups (effect size 1.42 SN group versus 1.58 FRDA group), but spinocerebellar and corticospinal tracts were affected only in FRDA patients. DRG volumes at L3-L5 were smaller in both groups compared to controls. Dorsal column fractional anisotropy at cervical SC correlated with disease severity in both groups (R: -0.6-0.7). A dedicated MRI protocol can capture SC and DRG abnormalities in both genetic and acquired SN. SC damage seems to be more widespread in genetic SN, which may be useful from a diagnostic perspective. The existing correlation between disease severity and SC diffusivity changes suggests MRI may be also relevant to track disease progression. **Conclusion:** Combined MRI-based evaluation of SC and DRG may be a potential (diagnostic and status) biomarker for sensory neuronopathies.

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Dosage of botulinum toxin in hemifacial spasm patients: a referral center experience

Fátima de Menezes Dantas[1], Felipe Olobardi Freire[1], Maria Weryca de Souza Belo Silva[1], Alberto Pereira Firmino Filho[1], Agabio Diogenes Pessoa Neto[2], Clécio de Oliveira Godeiro Junior[1], Rodrigo Alencar e Silva[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[2]Instituto de Ensino e Pesquisa Alberto Santos Dumont, Macaíba RN, Brazil.

Correspondence: Fátima de Menezes Dantas; Email: fatimamenezesdantas@gmail.com.

Background: Hemifacial spasm is a hyperkinetic movement disorder characterized by intermittent, involuntary contractions of muscles innervated by the facial nerve. The treatment of choice for this condition is regular administration of botulinum toxin. **Objective:** To evaluate the dose of botulinum toxin in patients who had been treated for hemifacial spasm over the last 14 years. **Methods:** A retrospective analysis of medical records from patients treated at a referral center from 2010 to 2024 was conducted. Doses (in units) and dates of botulinum toxin injections were collected and organized into 12-week intervals. An equivalence ratio of 5:1 U was adopted between abobotulinumtoxinA 500U and onabotulinumtoxinA 100U, the two formulations used in that service. The variables were subjected to a Mixed Linear Model test and Pearson correlation. The study was approved by the Local Ethics Committee (CAAE: 72821023.2.0000.5292) and all participants signed an informed consent form. **Results:** A total of 151 patients participated in the study. The average initial dose was 21.5 units. The analysis revealed that the dose of botulinum toxin increased 0.046 units per week ($p<0.05$; $r=0.1268$). During the first 3.46 years, this increase was more pronounced. In the long term, there was a slight increase in dose and a trend towards stabilization was observed. **Conclusion:** We concluded that the dose of botulinum toxin increases more pronouncedly during the initial years of administration, likely due to the need for dosage adjustments until symptom control is achieved with the lowest possible dose. Therefore, it is important to continue treatment even if the initial injections are not completely satisfactory. We believe that periodic and long-term consultations can contribute to achieving an optimized dose for the management of hemifacial spasm. Prospective studies with a larger number of participants may be able to validate these observations, allowing for a better understanding of long-term modifications in botulinum toxin dosage.

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Driver's license eligibility in adult patients with epilepsy

Gloria M. A. S. Tedrus[1], Pedro Augusto Silva Prado[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Gloria M. A. S. Tedrus; Email: gmtedrus@uol.com.br.

Background: Epilepsy can affect occupational and psychosocial aspects, including the eligibility to hold a driver's license. **Objective:** To assess the ownership of a National Driver's License (CNH) among adult patients with epilepsy (PWE) and explore its correlation with clinical variables. **Methods:** The possession of a driver's license was correlated with demographic and clinical data, as well as with the total scores on the Mini-Mental State Examination (MMSE) and the Quality of Life in Epilepsy Inventory-31 (QOLIE-31) among 305 PWE, with a significance level of $p<0.05$. **Results:** The mean age was 49.2 years, with 129 males and 176 females. The mean age at first seizure was 23.5 years, and with a mean of 5.7 years of schooling. Structural epilepsy was present in 203 cases, without an identified etiology in 75 cases, and genetic/generalized in 27 cases. Possession of an CNH (types AB in Brazil [classes M and D in the US] n=26, CDE in Brazil [classes B, C, and A in the US] n=9) was reported by 35 (11.5%) patients. Among those with an CNH, 14 had their first seizure before the age of 18 (Chi-square test, 14/21; $p=0.017$). More male subjects had an CNH (25 vs. 10, $p<0.001$), as well as PWE with no alterations in neurological examination (Fisher's exact test; 31 vs. 4, $p=0.01$) and in patients on a single antiseizure medication (29 vs. 7, $p=0.006$). There was no difference regarding CNH possession according to the type and frequency of seizures and epileptic syndrome. CNH possession was significantly associated with higher educational levels (years) (T-test, 7.5 vs. 5.5, $p=0.004$), better quality of life on the QOLIE-31 (67.0 vs. 59.9, $p=0.008$), and higher scores on the MMSE (25.6 vs. 22.7, $p<0.001$). In PWE with structural epilepsy (n=20), there was a significant difference in CNH type (AB vs. CDE), with fewer cases holding a CDE license (2 vs. 18, $p=0.047$). **Conclusion:** The rate of individuals with an CNH was low. Demographic and clinical aspects were associated with having an CNH, which was associated with better quality of life and better cognitive performance. This sample revealed aspects that are distinct from current legislation, suggesting a lack of information, denial of the disease, or the presence of various associated factors.

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Drug candidate prototypes for Parkinson's disease treatment: an in silico, in vitro, and in vivo study

Willian Alves Costa[1], Talita Alves Harrop[1], Gisele Rocha Lopes[1], Bianca Liffey Brito Marino[1], Lorane Izabel da Silva Hage Melim[1], Thiago da Costa Brito[2]

[1]Universidade Federal do Amapá, Macapá AP, Brazil.
[2]Universidade Estadual do Pará, Belém PA, Brazil.

Correspondence: Willian Alves Costa; Email: willianac2021@gmail.com.

Background: Parkinson's disease (PD) is the second most common degenerative disease in the world, especially in individuals over 60 years old. One of the mechanisms used in treatments for PD is monoamine oxidase B (MAO-B) inhibition, an enzyme that degrades dopamine. Recent studies have identified some substances of natural origin with that effect. **Objective:** To design in silico molecules with MAO-B inhibitory activity, based on natural products, carry out the synthesis and test them in vivo. **Methods:** A literature review was conducted to identify natural substances with proven antiparkinsonian activity in vitro and/or in vivo. Subsequently, an in silico study was performed with those substances, including molecular docking studies, prediction of pharmacokinetic and toxicological properties, structural modifications to obtain analogs, and prediction of activity and synthetic feasibility. After obtaining the analogs, synthesis was carried out (in vitro). Finally, for the in vivo study, an acute toxicity test was conducted using a Zebrafish animal model. **Results:** After a literature review, four natural substances were selected for the study: Amburosídeo A, Harman, Harmalina and Harmalol. The drug Selegiline was used as the standard molecule for comparison in the in silico study. In the prediction of pharmacokinetic activities, only the molecule Harman presented satisfactory results. In molecular docking, all substances interacted with more than three amino acids in the active site of MAO-B and some amino acids that Selegiline (the standard molecule) interacted with, suggesting probable inhibitory activity. After the analyses, Amburoside A was selected as the prototype for modifications, and three analogs (PMC1, PMC2, and PMC3) were obtained with results ranging from moderate to good synthetic feasibility. All three analogs showed predicted MAO inhibition activity. Subsequently, the prototypes were synthesized, and the PMC3's was selected for the acute toxicity assay in vivo using a Zebrafish model, due to its potential action on both the MAO-B and COMT enzymes. The test consists of administering the molecule studied at a certain dose to the fish and, if the fish survives for at least 7 days, this indicates a high probability that the substance is not toxic. In this analysis, the results were satisfactory. **Conclusion:** All the planned analogs showed possible MAO-B inhibitory activity in silico. Furthermore, the PMC3 analog also showed action on COMT, an easy synthesis and negative toxicity in the Zebrafish model, revealing a promising multitarget drug for Parkinson's disease treatment. However, more research is still needed to validate these findings and fully explore the safety and efficacy of this molecule.

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Drug-resistant epilepsy related to PPP2R5D

Lucas do Rego Barros Correa Martins[1], Laura A. M. Montouro[1], Maria da Penha Morita[1], Érica Nogueira Coelho[1], Lucia Helena Neves Marques[1], Charles Marques Lourenço[1], Humberto Ramos Crispim[1], Matheus Hisamitsu Facine[1], Caique Alberto Dosualdo[1], Eduardo de Paula Estephan[1,2]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Lucas do Rego Barros Correa Martins; Email: lucasrbcmartins@gmail.com.

Etiological diagnosis in cases of drug-resistant epilepsy and developmental delays is a daily challenge in clinical practice. This case report presents a 23-year-old male patient referred to the neurogenetics outpatient clinic due to drug-resistant epilepsy, cognitive delay, speech impairment and behavioral changes compatible of autism spectrum since childhood, that was diagnosed as PPP2R5D-related neurodevelopmental disorder. **Case presentation:** The patient has a history of epilepsy since the age of seven, with multiple seizure semiologies (being focal dysperceptive seizures the more prevalent). He was optimally managed with valproic acid and clobazam, maintaining an average frequency of one seizure per month. Additionally, cognitive delay and socialization difficulties since childhood were also reported. Neurological physical examination revealed dental malformations, hypertelorism, and low-set ears, without other alterations, but no ataxia signs were seen. Electroencephalogram revealed clustered sharp waves in the theta frequency in the left temporal region with occasional bilateralization. Brain magnetic resonance imaging showed no abnormalities. The patient underwent analysis through new generation sequencing, where a likely pathogenic variant (c.717T>G; p.Phe239Leu) was identified in the PPP2R5D gene. Other missense mutations in heterozygosity in this gene have been associated with Houge-Janssens Syndrome, with which the presented phenotype was consistent. **Discussion:** Houge-Janssens syndrome is a rare genetic disease caused by mutations in the PPP2R5D gene. This syndrome is characterized by mild to severe neurodevelopmental delay, craniofacial anomalies, hypotonia, speech impairment and autism spectrum disorder. Seizures are reported in fewer than half of individuals. Clinical Interpretation of the variant: The interpretation of variant as clinically relevant is based on the correlation between the clinical picture presented by the patient and the identified genetic alteration. Although not listed in international databases as pathogenic, the presence of specific clinical symptoms strengthens the association between the variant found in the PPP2R5D gene of the patient and Houge-Janssens syndrome. **Final comments:** The identification of the variant in the PPP2R5D gene in our patient provides important diagnostic insights and may guide future clinical management, including specific therapeutic options and genetic counseling for the family. It is crucial to continue investigating and understanding the genetic bases of neurological diseases to provide better care for patients and their families. Rational analysis of findings obtained in genetic tests is crucial for the diagnosis of these complex conditions. Although epilepsy is present in less than 50% of PPP2R5D-related cases, here it was the main clinical issue, which highlights the importance to include this condition in the work-up for drug-resistant epilepsy.

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Dual pathology in neurodegenerative dementias: when idiopathic normal pressure hydrocephalus diagnosis masks and modifies other dementias

César Minoru Toita Koga[1], Maria Clara Jorge Jardim[2], Gabriel Abrahão Stoliar[1], Sâmia Talise El Horr de Moraes[1], Alana Bacelar Limeira Sales[1], Alexandre Henrique Scheibe[1], Leonardo Ferreira Caixeta[3]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[2]Universidade Federal de Goiás, Goiânia GO, Brazil.
[3]Universidade Federal de Goiás, Hospital das Clínicas, Goiânia GO, Brazil.

Correspondence: César Minoru Toita Koga; Email: cesarmtkoga@gmail.com.

Case presentation: A 66-year-old right-handed woman with 8 years of education presented with progressive gait difficulty, cognitive symptoms, and impaired bladder control. She had a wide-based gait with shuffling steps and impaired working memory. In the next 2 years, her family noted progressive behavioral changes, ranging from irritability with physical aggression to apathy. She also developed preference for sweet foods and began eating compulsively. Cerebrospinal fluid (CSF) analysis and standard dementia laboratory screening were unremarkable. Brain magnetic resonance imaging (MRI) at the time showed extensive communicating hydrocephalus and bilateral frontal and temporal lobes atrophy, predominantly right-sided. At 69 years, she underwent a ventriculoperitoneal (VP) shunt, which resulted only in gait and urinary incontinence improvement. One year later, she exhibited behavioral disinhibition, loss of empathy and theory of mind (ToM), impulsivity, and social inappropriateness. Other symptoms included a persistent false sensation of fibers emerging from her teeth (Morgellons disease) and misidentification syndrome compatible with Frégoli syndrome. There were no visual hallucinations, and insight was absent. Neurological examination revealed the presence of palmomental reflex and inexhaustible glabellar reflex. Cognitive assessment showed predominantly verbal perseverance and frontal lobe dysfunction, with scores of 25/30 on Mini-Mental State Examination, 7 words/minute on Semantic Verbal Fluency test with animals, 12 words on FAS Verbal Fluency test and 5 points in the Clock Drawing Test according to Sunderland criteria. **Discussion:** This case highlights a diagnostic challenge in dementias. Properly diagnosed NPH can mask neurodegenerative dementias, such as behavioral variant frontotemporal dementia (bvFTD). NPH diagnose involves key symptoms (gait disturbance, urinary incontinence and cognitive impairment), typical neuroimaging findings including ventriculomegaly, and symptom improvement after CSF shunting. In our case, NPH presented with its classical triad and imaging findings, leading to VP shunting and motor symptons improvements. However, persistent and progressive behavioral and working memory symptoms post-shunting suggest an underlying bvFTD, or even a dysexecutive variant of Alzheimer's disease. Elements favoring bvFTD include complex neuropsychiatric symptoms (Frégoli and Morgellons syndromes) and significant right hemisphere cortical features such as disinhibition, compulsive behavior, and lack of insight and ToM. These features are atypical for NPH, which primarily causes subcortical dementia. Significant bilateral frontotemporal atrophy on MRI, especially with right-sided asymmetry, also suggests bvFTD. **Final comments:** This case illustrates how overlapping dementia pathologies can complicate each other's diagnosis and management. Careful clinical and neuroimaging assessment is crucial for accurate diagnosis and optimal treatment.

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Dural fistula and trigeminal autonomic headache

Cândida Driemeyer[1], Renato Dumbá Monteiro de Castro[1], Liselotte Menke Barea[1]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Cândida Driemeyer; Email: candidaplena@gmail.com.

Case presentation: A 62-year-old male patient received the diagnosis of dural fistula, fed by branches of the right external carotid artery and draining into the sigmoid sinus ten years ago when investigating a persistent headache that led him to seek the emergency department. In the last two years, he has been waiting to undergo elective embolization with Interventional Neuroradiology. He has undergone angiotomography which shows that the dural fistula is fed by branches of the right external carotid artery (middle meningeal artery) and a branch originating from the lateral wall of the ipsilateral cavernous segment of the internal carotid artery. The venous drainage occurs through the Labbé vein and the right transverse sinus. He has also undergone angiography, which did not demonstrate additional findings. He was then referred to the Neurology outpatient clinic for a unilateral right headache, pulsatile, which has been present over the last decade but severely worsened over the last two years, becoming disabling. The pain is continuous, but there are periods of exacerbation when moving the head forward. The pain is associated with conjunctival hyperemia and tearing, in addition to rhinorrhea and worsens when moving, and it is not triggered by touch. **Discussion:** Trigeminal Autonomic headaches are highly disabling, and their differential diagnosis is very important for correct management and improvement of the patient's quality of life. In our case, the patient underwent a therapeutic trial with indomethacin and he received a prescreption of valproic acid. He is still awaiting elective intervention on the dural fistula. **Final comments:** Trigeminal autonomic headaches still represent a diagnostic challenge in our setting and always require imaging tests to be performed, so that a secondary etiology can be correctly diagnosed.

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Dystonia caused by atp1a3 gene mutation: report of two cases

Vitor Santos de Souza[1], Débora Palma Maia[1], Ricardo Oliveira Horta Maciel[1], Lia Araújo Guabiraba[1], Francisco Eduardo Costa Cardoso[2]

[1]Universidade Federal de Minas Gerais, Hospital das Clínicas, Belo Horizonte MG, Brazil.
[2]Universidade Federal de Minas Gerais, Faculdade de Medicina, Belo Horizonte MG, Brazil.

Correspondence: Vitor Santos de Souza; Email: vitorsantosdesouza@outlook.com.

Case presentation: Case report of twin patients with suspected mutation in the ATP1A3 gene. Patient 1, 39 years old, daughter of non-consanguineous parents, with 2 sisters, one of them dizygotic twin. At the first evaluation in this service in July 2021, previously healthy patient developed sudden choreic movements in the right upper limb (RUL) and neck. Haloperidol and alprazolam were prescribed at a primary health unit. Despite improvement in choreic movements after 30 days of treatment, she developed stiffness in the affected limb, facial and cervical dystonia, gait difficulty, and behavioral changes (childlike behavior), excessive fear, social blunting, and impaired self-care. The neuroleptic was initially switched to risperidone and later to olanzapine, with no satisfactory motor response and worsening psychiatric symptoms and insomnia, leading to a switch to clozapine. On physical examination, there was cervical and trunk dystonia, and RUL dystonia associated with akathisia. The initial diagnosis was tardive dyskinesia (TD) with functional/psychogenic component. With a combination of trihexyphenidyl and clozapine, she remained psychiatrically stable but without improvement in dystonia. In April 2023, deutetrabenazine was prescribed. At this visit, it was reported that her twin sister started experiencing similar symptoms. Patient 2 reported right upper limb pain since 2019. Eight months ago, she started involuntary movements of raising her arm above her head, progressing to abnormal posture of shoulder elevation, internal rotation of the arm, flexion and internal rotation of the forearm, with functional limitation. On examination, there was RUL dystonia - ipsilateral shoulder elevation - with distal tremor, cervical dystonia - laterocollis to the right. The phenomenological pattern strongly suggests a mutation in the ATP1A3 gene. Genetic testing is ongoing, and flunarizine has been initiated. **Discussion:** Mutations in the ATP1A3 gene were initially associated with rapid-onset dystonia-parkinsonism (RDP), typically characterized by abrupt motor onset, sometimes accompanied by non-motor symptoms, mainly psychiatric. Since then, this mutation has been linked to various phenotypes including alternating hemiplegia of childhood and CAPOS (cerebellar ataxia, areflexia, pes cavus, optic atrophy, sensorineural hearing loss). The dystonic pattern in the twin sisters reported here is characteristic of ATP1A3 mutation, as described by Balint et al. There are reports suggesting individuals with this mutation may respond to flunarizine. **Final comments:** The etiological diagnosis of dystonias is often challenging. The patients described here demonstrate that clinical history, family history, and phenomenology remain fundamental tools for diagnosing rare conditions such as ATP1A3-related diseases.

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Early and long-term programming characteristics in Parkinson's disease patients submitted to subthalamic deep brain stimulation

Beatriz Azevedo dos Anjos Godke Veiga[1], Cristine Mella Cukiert[1], Rafael Basilio Guimarães[1], Arthur Cukiert[1]

[1]Clinica Cukiert, São Paulo SP, Brazil.

Correspondence: Beatriz Azevedo dos Anjos Godke Veiga; Email: bia-anjos@uol.com.br.

Objective: We aimed to describe the modifications of the subthalamic (STN) deep brain stimulation (DBS) programming parameters in Parkinson's disease (PD) patients in the early and long term. **Background:** PD is a neurodegenerative illness, and DBS has been a well-established efficacious symptomatic treatment for motor symptoms and motor complications. Assessing the best stimulation settings requires numerous clinic visits, and the stimulation intensity has the most significant effect on ameliorating Parkinsonian motor signs in the initial stages of STN-DBS programming. **Methods:** We performed a retrospective analysis of the parameter settings of fifteen patients who underwent STN-DBS with Medtronic devices. The patients were diagnosed with PD according to the UK Brain Bank criteria, and they had a positive result on the levodopa challenge test. We described continuous variables as means ± standard deviations (SD) and categorical variables as percentages. We used Repeated Measure ANOVA to continuous variables z-scores to analyze Levodopa Equivalent Daily Dose (LEDD) and DBS setting variations during six and twelve months after surgery. Scheffe post hoc test was applied, and the p-value considered statistically significant was <0.05. **Results:** Our sample was composed of 15 PD patients with a mean±SD age at PD onset of 48.7±9.0 years, and they took 11±3.7 years to undergo STN-DBS. We evaluated patients an average of 5.3±1.8 times during the first six months post-surgery and 8.6±3.3 times during the first year post-surgery. There was a significant LEDD reduction in six months compared to the pre-surgery (p=0.014) but not from the sixth to the twelfth month. Similarly, considering the variation period but inversely, the parameter amplitude increased only during the first six months (p=0.002). The other parameters, pulse width and frequency, did not statistically change after the first programming session. **Conclusion:** STN-DBS is a powerful symptomatic PD

treatment that requires skilled professionals and intense medical monitoring after neurosurgery to achieve the best results. Nonetheless, the arduous follow-up, including programming parameter variations, happens mainly in the early term.

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Early diagnosis of epidermoids cysts in the pontocerebellar angle up to age of 24 years: a challenge for managing neural tube defects - systematic review of the last 20 years

Evelyn Basilio da Silva[1], Rafael Augusto de Souza Santos[1], Anna Beatriz Popi e Souza[1], Ruan Gomez Carvalho Martins[1], Vitoria Rodrigues Carvalho da Silva[1], Milena Alexia Ferreira Cirino[1], Claudio Alberto Ioca Junior[1], Gabriel Borges Peres[1], Carolina Marques Kellner[1], Márcio de Souto Marques Filho[1]

[1]Universidade Nove de Julho, Bauru SP, Brazil.

Correspondence: Evelyn Basilio da Silva; Email: evelyn.basilio@uni9.edu.br.

Background: The neural tube is the origin from the embryo's notochord structure, between the third and fourth week of gestation. During this phase there may be anomalies which affect its proper closure, known as neural tube defects (NTDs). A possible NTD is the development of epidermoid cysts, which grow slowly, occupy space and, in most cases, have late symptoms, frequently occurring in the cerebellopontine angle. Although they are considered congenital defects, these cysts can be recognized at different ages. This singularity raises questions about the possibility of early diagnosis and treatment before changes in the affected individual's quality of life. **Objective:** Analyze the early diagnosis possibility and treatment of epidermoid cysts in the cerebellopontine angle, considering the age range up to 24 years. **Methods:** A systematic review was performed, following the PRISMA check-list, in the SciELO, PubMed and LILACS, using descriptors containing the keywords "epidermoid cyst" and "pontocerebellar angle". Considered those published between the year 2000 and January 2024, which addressed the presence of epidermoid cyst in the mentioned angle, in patients up to 24 years old. **Results:** 412 articles were collected, of which 95 met the criteria. Three main themes emerged from the explanatory content analysis: (1) the prevalence and incidence of epidermoid cysts in the cerebellopontine angle are more frequent in adults and with the potential for greater aggressiveness and worse prognosis in children, with 15 articles; (2) the most frequent symptoms were hearing loss, headache, dizziness and tinnitus, where patients reported that the symptoms persisted for months or even years before medical investigation, with 30 articles and (3) the diagnosis is made through exams imaging, mainly magnetic resonance imaging (MRI) and the most effective treatment continues to be cyst resection, with 40 articles. **Conclusion:** The analysis demonstrated that epidermoid cysts in the cerebellopontine angle can manifest at different ages, with greater aggressiveness in childhood and more frequently in adulthood. Regardless of the period in which they were discovered, these cysts usually affect the full lives of these people with different symptoms, bringing attention to adequate and early clinical investigation. Therefore, late diagnosis can increase the complications risk, including possible loss of neurological function due to its exponential growth around cranial nerves that emerge close to this location. To this end, MRI remains essential for accurate diagnosis, while surgery, with its nuances and risks, remains the most effective treatment of choice. Continuous research into development and progression factors, aiming to improve diagnostic and treatment techniques, is crucial to guarantee the best prognosis and quality of life for patients.

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Early diagnosis of laing myopathy with neuromuscular ultrasound: a case report and future directions

Antonio Edvan Camelo Filho[1], Pedro Lucas Grangeiro Sá Barreto Lima[1], Ellaine Dóris Fernandes Carvalho[2], Pedro Vitor Ferreira Rodrigues[1], Francisco Luciano Honório Barreto Cavalcante[1], Oliver Reiks Miyajima[3], Pedro Braga-Neto[1], Paulo Ribeiro Nóbrega[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Unichristus, Fortaleza CE, Brazil.
[3]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Antonio Edvan Camelo Filho; Email: edvan.camelo@gmail.com.

Case presentation: A 6-year-old boy was referred for evaluation for abnormal gait and frequent falls since age 3. The motor development was unremarkable until the third year of life, when the family reported the symptoms responsible for the referral. Upon neurological examination, the patient had isolated mild atrophy of the medial compartment of the legs with mild weakness of dorsiflexion. There were no sensitive, cerebellar or cranial nerve deficits. Patient creatine kinase (CK) was within normal limits (135 U/L). The patient performed an electromyographic evaluation that was normal. Due to high clinical suspicion, we evaluated with neuromuscular ultrasound (NMUS), which revealed mild bilateral sartorius and medial gastrocnemius muscle echogenicity changes (Heckmatt scale 2). These findings were suggestive of an early myopathic disorder. Leg MRI confirmed atrophy of the soleus and gastrocnemius muscles. Genetic testing of this patient revealed heterozygous pathogenic variants in MYH7 gene (p.Ala797Thr). Laing myopathy was diagnosed with these clinical, genetic, and radiological findings. **Discussion:** Laing myopathy is an inherited disorder caused by pathogenic variants in MYH7. This gene encodes the myosin heavy chain of type 1 fibers of skeletal muscle and cardiac ventricles, and its deficient expression produces a phenotype of distal myopathy, mostly affecting ankle dorsiflexors and toe extensors, associated with dilated cardiomyopathy. Age of onset is highly variable, reported from congenital to 50 years. However, most patients present signs and symptoms between the second and third year of life. This is a rare form of myopathy, being reported in 2 patients in a large Brazilian cohort of distal myopathies. CK levels are usually normal or only moderately elevated in these patients. Also, some LM patients may have nerve conduction studies reported as normal or even neurogenic. Neuromuscular ultrasound assessment is a trending tool in muscular disorders that may help in making an early diagnosis. In LM the ultrasound assessment may be a quicker and cheaper option to assess the characteristic atrophy of muscles, specially in children and early structural myopathies with normal CK values. **Final comments:** Integrating NMUS with traditional NCS studies enhances diagnostic accuracy, especially in the pediatric population, where EMG findings may initially be unremarkable for mild changes or non-cooperation. Our findings advocate for the wider adoption of NMUS in the diagnostic workup of suspected myopathies, facilitating prompt diagnosis and ultimately contributing to improved patient outcomes through earlier therapeutic intervention. Future research should investigate the potential utility of NMUS in standardized patterns of muscle and nerve assessment and the development of evidence-based protocols for its application in the clinical setting, ultimately improving the quality of life for affected individuals.

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Early fecal metabolomic alterations in pilocarpine-induced status epilepticus: insights into tle pathophysiology and resistance mechanisms

Gabriele Fernanda da Conceição Santos[1], Larissa da Silva Almeida[1], Manoela Alves Vieira de Souza[1], Karla Eduarda Silva Ferreira[1], Jeferson Santana Ursulino[1], Thiago Mendonça de Aquino[1], Daniel Leite Góes Gitaí[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.

Correspondence: Larissa da Silva Almeida; Email: larissa.almeida@famed.ufal.br.

Background: Epilepsy, a complex neurological disorder characterized by recurrent seizures, presents a significant health burden worldwide. Among the various types of epilepsy, temporal lobe epilepsy (TLE) stands out for its high prevalence in the adult population. TLE is often associated with alterations in the limbic system (epileptogenesis), typically triggered by epileptogenic insults such as status epilepticus (SE). Understanding its underlying mechanisms is crucial for effective treatment strategies. Recent advances in metabolomics offer a promising avenue for exploring the metabolic alterations associated with epilepsy. By analyzing metabolites, the byproducts of cellular processes, researchers can gain insights into the biochemical changes occurring in epileptic brains. **Objective:** To analyze fecal water metabolites in a Lithium-Pilocarpine-induced status epilepticus (SE) rat model to identify metabolic changes during early epileptogenesis. Specifically, metabolomic analysis will be conducted 24 hours post-SE induction, comparing with SE-resistant and naïve control groups. **Methods:** Male Wistar rats were induced into status epilepticus (SE) using Lithium-Pilocarpine administration and euthanized after 24 hours. Fecal samples were collected and processed for metabolomic analysis using Nuclear Magnetic Resonance (NMR). Three groups were defined: a 24-hour SE group (n=10), an SE-resistant group (n=11), and a naïve group (n=10). Statistical analyses, including T-tests, Principal Component Analysis (PCA), Partial Least Squares Discriminant Analysis (PLS-DA), and Analysis of Variance (ANOVA), were conducted to compare metabolite levels among groups. **Results:** Our findings reveal significant alterations in 24 metabolites, including upregulation of Phenylalanine, fumarate, and hypoxanthine, and downregulation of butyrate, threonine, and glutamate in the 24-hour post-SE group compared to naïve controls. Contrarily, the SE-resistant group exhibits distinct metabolic signatures, with upregulation of pyridoxine, propionate, and several amino acids, and downregulation of glucose and ethanol compared to naïve controls. Notably, ribose and succinate levels are higher in the SE-resistant group compared to the 24-hour post-SE group. Clustering analysis further highlights the unique metabolic profile of the SE-resistant group. **Conclusion:** The findings showed that specific metabolites exhibit distinct patterns in SE-induced and SE-resistant groups, indicating potential markers for disease progression and treatment response. The distinct metabolic signatures between SE-induced and SE-resistant groups underscore the complexity of TLE pathophysiology. Further research is necessary to clarify the underlying mechanisms and validate these findings for clinical application.

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Early reactivation of Chagas disease in the central nervous system after heart transplant: case report

Alexandra de Oliveira Lopes[1], Maria Luiza Nóbrega Lima Albuquerque[1], Gustavo Carvalho Oliveira Gonçalves Machado[1], Rafaela Pessoa Alves Luso[1], Bernardo Gomes Muffato[1], André Luís de Oliveira Silveira[1], Italo Pena Guimarães[1], Sabrina Gesteira Souza[1], Paulo Pereira Christo[1], Breno Franco Silveira Fernandes[1]

[1]Universidade Federal de Minas Gerais, Hospital das Clínicas, Belo Horizonte MG, Brazil.

Correspondence: Alexandra de Oliveira Lopes; Email: alexandramoraes11@gmail.com.

Case presentation: A 57-year-old female patient, previously diagnosed with Chagas cardiomyopathy, underwent heart transplantation 3 months ago. She was admitted to a quaternary hospital following her first-ever seizure episode. At the time of admission, she was on Cyclosporine and Mycophenolate Mofetil post-transplant and did not present with focal neurological deficits on initial neurological examination. Initially, a CT scan showed hypodensity in the right anterior opercular region with mild mass effect. Magnetic resonance imaging (MRI) confirmed a lesion with subcortical signal alteration in the right peri-rolandic region characterized by hypersignal on T2/FLAIR, hyposignal on T1, irregular central enhancement with contrast, foci of hyposignal on SWI, no diffusion restriction, measuring 27x27 mm. Cerebrospinal fluid analysis revealed: glucose 73 mg/dL, proteins 35.2 mg/dL, 3 cells (88% lymphocytes), negative cultures, serologies, and meningoencephalitis panel. Subsequently, Trypanosoma cruzi DNA detection in the cerebrospinal fluid via polymerase chain reaction was positive. Given the diagnosis of probable Chagas disease reactivation with an expansive lesion in the central nervous system (CNS), treatment with Benznidazole 5 mg/kg/day for 60 days was initiated. Additionally, she started levetiracetam for seizure control, with no recurrence of events. A follow-up MRI showed slight reduction in lesion dimensions post-treatment, confirming therapy efficacy. **Discussion:** Chagas disease is a complication of Trypanosoma cruzi infection, endemic in tropical countries and regions with low socioeconomic status. It rarely affects the CNS during acute disease or reactivation in the chronic phase, predominantly in immunocompromised individuals. About 89% of cases occur in HIV-infected individuals, with the remainder in post-transplant patients, undergoing chemotherapy, hematologic neoplasms, or immunosuppression due to autoimmune diseases. Acute disease presents as meningoencephalitis or cerebral abscess, while chronic forms manifest as mass-effect lesions with peripheral contrast enhancement, perilesional edema, and necrotic centers. Common symptoms include focal neurological deficits (47%) and seizures (32%). Diagnosis requires confirmation of the parasite in cerebrospinal fluid or lesion biopsy (gold standard). Treatment involves Benznidazole at 5-7 mg/kg/day for 60-90 days for acute and reactivated disease, with a guarded prognosis, especially in reactivation, where in-hospital mortality exceeds 70%. **Final comments:** Reactivation of Chagas disease in the CNS is rare but increasingly recognized due to the widespread use of immunosuppressants. Despite its rising relevance, the literature still lacks comprehensive evidence on epidemiology, diagnosis, and treatment, necessitating further research for better understanding and management of the disease.

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Early stroke as a primary manifestation of inborn error of immunity

Carolina Chong-Silva[1], Daniel Kazuo-Tomoto[1], Francisca Mirian Moura Lacerda[1], Samir Farhat Hach[2], Fernanda Manfroi-Guimarães[2], Lifkyn Remboski[1], Camila Andrade Neix[1], Gesmar Rodrigues Silva Segundo[3], Herberto José Chong-Neto[4]

[1]Universidade Paulista, Sorocaba SP, Brazil.
[2]Universidade Positivo, Curitiba PR, Brazil.
[3]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.
[4]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Carolina Chong e Silva; Email: carol.chong.silva@gmail.com.

Case presentation: A 6-year-old female patient presented with a history of multiple ischemic and hemorrhagic strokes with one year old, resulting in cerebellar ataxia, nystagmus, and right hemiparesis, associated with recurrent fever and livedo racemosa. She has no siblings, and her parents are not consanguineous. Magnetic Resonance Imaging showed an intraparenchymal hematoma in the left temporal lobe in the acute stage, signs of perilesional ischemia, and hemoventricle signs. Laboratory evaluation shows red blood cells=4.91 x 106/mL, hemoglobin=12.4g/dL, white blood cells=6.1 x 103/mL, neutrophils=4.3 x 103/mL, lymphocytes=1.2 x 103/mL, and platelets=217 x 103/mL. IgE<25IU/mL, IgG=467mg/dL, IgM=13.8mg/dL, and IgA=21.3mg/dL. Lymphocyte subtypes show CD3+=853 cells/microliter (81.2%), CD4/CD8=1.41, CD19+=113 cells/microliter (10%), and CD16/56=81 cells/microliter (8.8%). Due to the clinical history, genetic sequencing of the CECR1 gene was performed, showing a homozygous substitution at position -2 of the splice acceptor site in intron 6, c.847-2G>A. This is a highly conserved region with no alteration in phylogenetic studies and is predicted to be pathogenic. To confirm the functional alteration, the activity of Adenosine Deaminase-2 (ADA-2) was tested in dried blood spot, showing 0.0 mU/g of protein, confirming the loss of gene function and the pathogenicity of the mutation. **Discussion:** ADA-2 has partial structural homology with ADA-1. Both proteins convert adenosine to inosine and 2'-deoxyadenosine to 2'-deoxyinosine. ADA-2 deficiency is an immune dysregulation disease caused by an autosomal recessive mutation in the CECR1 gene, leading to mild immunodeficiency, more evident in B cells, accompanied by recurrent fevers and a spectrum of pathological vascular characteristics. **Final comments:** The authors presented a child with an inborn error of immunity with the primary manifestation of cerebrovascular disease due to a mutation in the CECR1 gene, causing loss of gene function and confirmed by the extremely reduced activity of ADA-2.

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Early vs classical facioscapulohumeral muscular dystrophy onset: a specialized muscular center profile

Filipe Tupinambá Di Pace[1], Cristiane Araújo Martins Moreno[1], Clara Gontijo Camelo[1], Gabriel Saboia de Araújo Torres[1], Raquel Diógenes Alencar Sindeaux[1], Gabriella Corrêa Dousseau[1], Mariana Hiromi Manoel Oku[1], Rodrigo de Holanda Mendonça[1], André Macedo Serafim Silva[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Filipe Tupinambá Di Pace; Email: filipedipace@gmail.com.

Background: Facioscapulohumeral muscular dystrophy (FSDH) is the second most common muscular dystrophy in adults, being termed early-onset (EO) when it starts until the age of 10. FSHD is divided into types 1 and 2 based on genetic markers, both caused by an abnormal expression of the DUX4 gene. This study presents a Brazilian FSHD cohort comparing EO and classical-onset (CO) aspects. **Objective:** To compare the demographic, clinical, and genetic profile of patients with EO versus CO in FSHD. **Methods:** Data were extracted from electronic medical records, between December 2016 and February 2024. Key data points included patient registration, birth date, sex, family history, age and site of the first symptom, genetic test results, muscular and extra-muscular involvement. **Results:** 46 patients were included, 14 (30.4%) in the EO and 32 (69.5%) in the CO group, in which females were affected in 57.1% vs 40.6%, and no disease family history was found in 53,8% vs 25%. The mean age at the first symptom was 5.8 (2-10) and 31.7 (12-70) years, and the mean time of disease duration was 24 (3-60) and 18 (3-49) years for EO and CO, respectively. Genetic tests were performed on 78% of patients. All CO patients had FSHD type 1, while two EO patients had FSHD type 2. The EO and CO groups started the disease in the upper limbs at 41.7 % vs 60%, in the lower limbs at 25% vs 40%, and in the face at 33.3% vs none. Facial involvement was present in 87.8% of patients, with no difference between groups at the last appointment. Wheelchair users were more frequent in the EO group (23.1% vs 11.5%), and the mean age at the first symptom was 3.3 vs 6 years compared to the ambulatory EO group. Among all patients, the time of disease duration of ambulatory and non-ambulatory was 22.9 and 21.8 years. 21 patients had the pulmonary functional tests available, 63,6% of them were normal, with no difference between groups. Regarding extra-muscular involvement, pain was a frequent symptom (86.3%), and cardiac test results, were available for 8 patients in the EO group and 17 in the CO group, showing abnormality in 20% of them (25% vs 17.6%). Ophthalmological and hearing exams were available for a few patients and were not evaluated. **Conclusion:** This FSHD cohort provides important insights into the disease profile among Brazilian patients. Key findings in the EO group include a higher frequency of affected females, less family history, more cases of FSHD type 2, the significance of age at first symptom over disease duration regarding wheelchair dependency, and more cardiac involvement. Notably, none of the CO patients presented with facial onset. Continued investigation of our patients' profiles is crucial for a better understanding of early symptoms and prognosis.

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Early-onset myopathy, areflexia, respiratory distress, and dysphagia (EMARDD): a case report and description of a novel variant

Laura Alonso Matheus Montouro[1], Janaina Moraes de Araújo[1], Regina Celia Ajeje Pires Albuquerque[1], Charles Marques Lourenço[1], Erica Nogueira Coelho[1], Eduardo de Paula Estephan[1], João Aris Kouyoumdjian[1], Carla Renata Graça[1], Maria da Penha Ananias Morita[1]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Laura Alonso Matheus Montouro; Email: lauramontouro@hotmail.com.

Case presentation: A 5-month-old female infant comes to the hospital with 1 week progressive respiratory distress. She was born at 41 weeks gestation by C-section without complications. There is no consanguinity, nor similar familial cases. On physical examination: absence of dysmorphisms, adequate visual/social contact, weak cry, decreased limb movement, global hypotonia and areflexia, intense weakness of the axial muscles. During hospitalization, she developed persistent respiratory failure and dysphagia. Creatine Phosphokinase: 60 U/L, whole body MRI shows diffuse muscle groups hypotrophy, no organomegaly and lungs hypoexpansion. Electroneuromyography: reveals a myopathic pattern on proximal limb muscles and on paravertebral muscles, in addition, these with spontaneous activity and complex repetitive discharge. Muscle biopsy: neonatal myosin and embryonic immunohistochemical studies reveal a large amount of positive fibers. Dried blood spot tests (Pompe/Gaucher/Niemann-Pick diseases, aminoacidopathies and disorders of urea cycle/organic/fatty acids/purine metabolism): negative. Serum/urinary carnitine and urinary organic acids: normal. MLPA for SMN1/SMN2: no deletions in the SMN1. Exome study: homozygous missense variant (c.229C>T) in MEGF10. **Discussion:** Infantile hypotonia with respiratory weakness can be related to the brain, anterior horn of the spinal cord or neuromuscular structures. Congenital myopathy is responsible for 14% of cases of neonatal hypotonia, occurs due to a structural defect in the components of the muscle fiber and is a genetically heterogeneous group with around 40 genes involved. The most prevalent genes are: RYR1, NEB, ACTA1, ADSSL1, SELENON, MTM1, DNM2, and TPM3. In 2011, the first study, with 7 patients from 5 families, of congenital myopathy associated with MEGF10 by Logan et al. This gene is related to two phenotypes: Early-onset Myopathy with Areflexia, Respiratory Distress & Dysphagia (EMARDD) (congenital myopathy 10A) and a late-onset syndrome, usually associated with minicore lesions on muscle biopsy (congenital

myopathy 10B). Our proband has a EMARDD phenotype and a homozygous variant that promotes the replacement of arginine by tryptophan in codon 77 (p.Arg77Trp), arginine in this position is conserved in different species and infrequent in heterozygous on population (3:141.000). However, it has never been previously described in the literature/variant repository. To date, there are 32 cases of EMARDD associated with MEGF10 in the literature. Only one reported in Brazil, presented at a national congress in 2022. An experimental study identified that selective serotonin reuptake inhibitors (SSRI) have a beneficial effect in vitro/in vivo animal models. **Final comments:** MEGF10 myopathy must be remembered as a cause of infantile hypotonia with respiratory failure due to prognosis, genetic counseling and, mainly, due to the prospect of SSRI as a treatment.

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Eaton Lambert myasthenic syndrome with cerebellar ataxia misdiagnosed as Guillain-Barré spectrum

Raíssa Josefa Pereira de Moura[1], Mayhara Rosany da Silva Santiago[1], Nereu Alves Lacerda[1], Ana Caroline Paiva Simeao[1], Renata Amaral Andrade de Mendonça[1], Natália Nunes Tenório[1], Heron Alves Vale[1], Maria Luisa Roberto Lima Lisboa[1], Ana Beatriz Roberto Lima Borba[1], Ana Carolina Martins Fragoso Carneiro[1],

[1]Hospital Pelópidas da Silveira, Recife PE, Brazil.

Correspondence: Raissa Josefa Pereira de Moura; Email: rayssamoura16@gmail.com.

Case presentation: A 57 years old woman, smoker (30 pack-years), presented with a 1 month lower limbs proximal weakness, evolving with diplopia and cerebellar ataxia. Physical examination revealed o ophthalmoparesis, a grade IV ileopsoas and quadriceps weakness, areflexia on lower limbs and cerebellar ataxia. Initially a Guillain Barré syndrome (Miller Fisher spectrum) was considered and intravenous human immunoglobulin was administered with significant clinical improvement. During diagnostic investigation, cerebrospinal fluid showed mild hyperproteinorrachia (72 mg/dl - VR 45 mg/dl) with no pleocytosis (02 cells). However, during the subsequent months, there was further clinical deterioration and was performed new clinical investigation. The electroneuromyography exam showed a presynaptic neuromuscular junction disorder (increment >100%) compatible with Eaton Lambert myasthenic syndrome (LEMS). All neoplastic screening was negative. **Discussion:** LEMS is a rare neuromuscular junction autoimmune disorder. In most patients serum antibodies against P/Q voltage-dependent calcium channels bind to the presynaptic motor nerve terminal and inhibit depolarization-dependent calcium influx, reducing the acetylcholine release at the neuromuscular junction. LEMS may occur as a paraneoplastic disorder, in association with an underlying cancer, or as part of a more generalized autoimmune state. The patient in this report underwent all accessible paraneoplastic screening, which was negative. The clinical findings of this pathology include gradual onset proximal muscle weakness and absent or reduced reflexes but we highlight the occurrence of cerebellar ataxia. This association is due to the high expression of autoantibodies in the cerebellar system and few case reports and small series of patients with simultaneous cerebellar ataxia and LEMS have been described, including one patient in the original report by Eaton and Lambert. Generally, electrophysiological studies have demonstrated decremental responses to low-frequency repetitive nerve stimulation, and may also show post-exercise facilitation greater than 100%. Effective, long-term symptomatic treatment options include agents that increase neuromuscular transmission and immunomodulators. **Final comments:** LEMS associated with ataxic syndrome is uncommon; a few reports in the literature demonstrate an association between these syndromes and autoantibodies against voltage-gated calcium channels, especially when the patient has some type of underlying neoplasm; our patient was screened and is being followed up, however, to date, there is no evidence of underlying malignancy. However, LEMS symptoms may appear before the discovery of an underlying malignancy, which demonstrates the need for follow-up and periodic paraneoplastic screening.

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Ecological study of hospitalization for dementia in Brazilian regions, 2014 - 2023

Danielle de Lima Pimentel[1], Verônica Guimarães Irias de Lima[1], Letícia Cavalcante da Costa Aragão[2], Danilo Pereira Fermino[3], Izabella de Campos Marinho[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.
[2]Universidade Federal do Delta do Parnaíba, Paraíba PI, Brazil.
[3]Unisalesiano, Araçatuba SP, Brazil.

Correspondence: Danielle de Lima Pimentel; Email: daniellepimentel@id.uff.br.

Background: According to the Ministry of Health, Brazil has about 2 million people with some form of dementia, which is expected to triple by 2050, mainly affecting the elderly. Alzheimer's disease and dementia with Lewy bodies are among the most worrisome clinical forms of neurodegenerative dementia. Studies which describing the pattern of onset and distribution of this disease in Brazil are essential due to its impact on public health with the increase in life expectancy in the country. **Objective:** To describe the epidemiological profile of patients hospitalized with dementia using different variables in the regions of Brazil. **Methods:** This is an ecological, descriptive study with a quantitative approach The information was obtained by collecting secondary data from the SUS Hospital Information System (SUS/HIS), provided by the Department of Informatics of the Unified Health System (DATASUS), from 2014 to 2023. The target population is made up of patients diagnosed with dementia, assessing the distribution of the number of visits by region and correlating with gender, age group, race and year. **Results:** There were 28,124 hospitalizations for dementia in Brazil during the period analyzed, with the South having the highest incidence of hospitalizations (2.12 per 100000 inhabitants), followed by the Southeast (1.86 per 100000 inhabitants). During this period, the highest incidence occurred in 2023, with a value of 1.46 per 100000 inhabitants, and the lowest incidences were in 2020, with 1.15 per 100000 inhabitants, and 2021, with 1.19 per 100000 inhabitants. In addition, there were 11,934 cases among white people, followed by brown people with 7,048 cases. There was no significant difference between women (13,983 cases) and men (14,141 cases), increasing linearly with age, especially over 60 years, which accounts for 62.33% of the cases (17,531 cases). **Conclusion:** It is evident that the population most at risk of dementia is mainly people over 60 years old, white and brown people, living in the south and southeast regions, which according to the 2022 IBGE census represent the majority of these populations justifing the figures. The low incidences in 2020 and 2021 may be related to the impact of the Covid-19 pandemic directly on the notifications. The number of cases of hospitalization for dementia is therefore worrying and on the rise, highlighting the need for further studies on the subject.

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Ecological study of meningococcal infection in the Southeast region of Brazil from 2015 to 2022

Danielle de Lima Pimentel[1], Gabrielle Gomes Garcia[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Danielle de Lima Pimentel; Email: daniellepimentel@id.uff.br.

Background: Meningococcal infection leads to diseases such as meningitis and meningococcemia, which may cause permanent sequelae and death. Neisseria meningitidis infection is related to contact with an asymptomatic, sick individual or through direct contact with respiratory secretions. Therefore, studies that analyze its incidence in recent years, including how it has been affected by the COVID-19 pandemic, are necessary in order to take action to control these diseases. **Objective:** To analyze the incidence of meningococcal infection in the Brazilian Southeast region. **Methods:** This is a descriptive ecological study with a quantitative approach, using data from secondary sources, through the Hospital Information System of the Brazilian Unified Health System (SIH/SUS) provided by the Ministry of Health, accessed through the Department of Informatics of the Unified Health System (DATASUS). Data covered the period from 2015 to 2022 for the population residing in the Southeast region of Brazil. Variables included year of hospitalization, age group, sex, and race/ethnicity, assessing the number of hospitalizations and mortality rates. **Results:** 2869 cases of hospitalizations were identified, of which 572 occurred in 2015, 442 in 2016, 435 in 2017, 410 in 2018, 380 in 2019, 220 in 2020, 181 in 2021 and 229 in 2022. Among hospitalizations in the Southeast region, the most affected groups were children up to 4 years old with 888 cases (31%), males with 1584 cases (55.2%) and individuals identifying as white with 1025 cases (35.7%). The mortality rate is higher in individuals aged 80 years or older (41.98 per 100 patients), in the female population (15.25 per 100 patients) and in the black population (19.86 per 100 patients) and yellow population (19.44 per 100 patients). **Conclusion:** Elderly, female, black and yellow population are at higher risk of death from meningococcal infection. In addition, there has been a significant decrease in the number of hospitalizations, especially from 2020 onwards, possibly correlated with the COVID-19 pandemic and the resulting social isolation reducing transmission. However, studies indicate a decline of meningococcal C immunization coverage in this period, which could lead to an increase in its incidence. Despite the limitations of the study, such as the inability to make a cause-and-effect association, the results highlight its importance for decision-making to face this disease.

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Ecological study of migraines and other headache syndromes in Southern Brazil, 2014 - 2023

Danielle de Lima Pimentel[1], Verônica Guimarães Irias de Lima[1], Letícia Cavalcante da Costa Aragão[2], Danilo Pereira Fermino[3], Izabella de Campos Marinho[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.
[2]Universidade Federal do Delta do Parnaíba, Paraíba PI, Brazil.
[3]Unisalesiano, Araçatuba SP, Brazil.

Correspondence: Danielle de Lima Pimentel; Email: daniellepimentel@id.uff.br.

Background: According to the World Health Organization, approximately 50% of the world's population suffers from acute headache at some point in their lives. Headache has potentially disabling features, negatively affecting well-being and causing economic and social harm. Some studies have already shown the major impact of headache on the health system in Brazil, but few studies have

investigated the socio-demographic aspects of the high incidence of cases in the southern region. **Objective:** To analyze the epidemiological profile of patients hospitalized with migraine and other headache syndromes due to different variables in southern Brazil. **Methods:** This is a descriptive ecological study with a quantitative approach. The information was obtained by collecting secondary data from the SUS Hospital Information System (SUS/HIS), provided by the Department of Informatics of the Unified Health System (DATASUS), from 2014 to 2023. The target population of this study is made up of patients hospitalized with a diagnosis of migraine and other headache syndromes in the Southern Region of Brazil, evaluating the distribution of the number of visits and correlating it with gender, age group, race and year. **Results:** During the period analyzed, there were 96,348 hospitalizations for migraine and other brain pain syndromes in Brazil, with the South accounting for 24,974 of these hospitalizations. The South had the highest incidence of hospitalizations (8.37 per 100000 inhabitants) in Brazil. The incidence of hospitalizations in the Brazilian population increased linearly from 2014 (3.87 per 100000 inhabitants) to 2019 (5.77 per 100000 inhabitants), and suffered a decrease in 2020 (4.1 per 100000 inhabitants) and 2021 (3.99 per 100000 inhabitants), which increased again in 2022 (4.84 per 100000 inhabitants) and 2023 (5.09 per 100000 inhabitants). The most affected age group was 40 to 44 years with 2212 cases, much more frequent in women (16125 cases) than in men (8723 cases) and in white people with 20131 cases. **Conclusion:** The identified population at risk for headache susceptibility corroborates data from other studies, with genetic factors, hormones, and the menstrual cycle as the main causes that could explain this prevalence. In addition, the COVID-19 pandemic may have influenced the number of cases of hospitalization for migraine, as a decrease was only observed in 2020 and 2021, the most significant period of the pandemic.

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Ecological study: analysis of morbidity and mortality of bacterial meningitis in preschool children in the Southeast region

Anna Gabriella Venâncio Neves de Moraes Tosta[1], David Cohen[2], Larissa Farah de Oliveira[3], Hadassa Lucena Sales Santos[4], Paulo Fernando Katsuo Ogatha Ito[5]

[1]São Leopoldo Mandic, Campinas SP, Brazil.
[2]Universidade Luterana do Brasil, Canoas RS, Brazil.
[3]Universidade Municipal de São Caetano do Sul, São Caetano SP, Brazil.
[4]Universidade Ceuma, Imperatriz MA, Brazil.
[5]Universidade para o Desenvolvimento do Estado e da Região do Pantanal, Campo Grande MS, Brazil.

Correspondence: Anna Gabriella Venâncio Neves de Moraes Tosta; Email: Annagabriellatosta@gmail.com.

Background: Bacterial meningitis (BM) is a severe infection that affects the central nervous system, predominantly in children, with high morbidity and mortality and rapid symptomatic progression. Early detection is essential for a better prognosis, as this disease involves severe complications and irreversible neurological sequelae. The main pathogenic agents associated with this infection are Streptococcus pneumoniae (S. pneumoniae), Neisseria meningitidis (N. meningitidis), and Haemophilus influenzae (H. influenzae). **Objective:** To analyze the morbidity and mortality of bacterial meningitis in the preschool population in the Southeast region from 2017 to 2023. **Methods:** This is an ecological, cross-sectional, descriptive study with a quantitative approach, conducted in May 2024, using data collected from the Hospital Information System of SUS (SIH/SUS - Sistema de Informações Hospitalares do Sistema Único de Saúde), available from the Department of Informatics of the Unified Health System (DATASUS - Departamento de Informática do Sistema Único de Saúde). The variables used were hospitalizations, total cost, deaths, and mortality rate. The collected data were organized in Microsoft Excel spreadsheets and analyzed using descriptive statistics. **Results:** The analysis revealed the mortality rates, deaths, hospitalizations, and total cost, respectively, for each state in the Southeast Region. In Minas Gerais, the mortality rate was 3.33% (18 deaths and 540 hospitalizations) with a total cost of R$1,585,164.03; in Espírito Santo, the mortality rate was 5.88% (5 deaths and 85 hospitalizations) with a total cost of R$387,698.65; in Rio de Janeiro, the mortality rate was 7.69% (23 deaths and 299 hospitalizations) with a total cost of R$653,258.55; in São Paulo, the mortality rate was 2.85% (54 deaths and 1,895 hospitalizations) with a total cost of R$5,779,650.62. In this context, it is noticeable that the state of São Paulo has the highest allocated funds but the lowest mortality rate compared to the other states. Hence, there is evidence of greater vulnerability in the distribution of funds in Brazil, contributing to the worsening situation in some more fragile states in regions such as the Southeast. **Conclusion:** The data evaluated from 2017 to 2023 show the disparity in the funds allocated among the states in the Southeast region. Bacterial meningitis is responsible for approximately 5 to 40% of deaths in children with this condition, emphasizing the need for significant attention to the care of this population, especially in hospitals with exposure to infections and in preschool children with pre-existing conditions, in order to assist in the prevention of bacterial meningitis.

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Effect of a 12-week protocol of strength and functional exercises on the treatment of motor symptoms in individuals with hereditary spastic paraparesis: adaptive crossover randomized clinical trial

Jonas Alex Morales Saute[1,2], Isabela Possebon Bevilacqua[1], Arthur Carpeggiani Weber[2], Barbara V. Motta[2], Cesar Augusto Weschenfelder[2], Bruna Faria Rosa[2], Julia Kersting Chadanowicz[2], Flávia Gomes Martinez[3], Diana Maria Cubillos-arcilla[1]

[1]Universidade Federal do Rio Grande do Sul, Programa de Pós-Graduaçao em Ciências Médicas, Porto Alegre RS, Brazil.
[2]Universidade Federal do Rio Grande do Sul, Faculdade de Medicina, Departamento de Medicina Interna, Porto Alegre RS, Brazil.
[3]Universidade Federal do Rio Grande do Sul, Escola de Educação Física, Fisioterapia e Dança, Porto Alegre RS, Brazil.

Correspondence: Jonas Alex Morales Saute; Email: jsaute@hcpa.edu.br.

Background: Hereditary spastic paraplegias (HSP) constitute a group of rare, neurodegenerative genetic disorders characterized by spastic hypertonia, hyperreflexia, muscle weakness in the lower limbs, and impaired vibratory sensation, resulting in the gradual loss of balance and walking ability. Due to clinical and genetic heterogeneity, as well as functional deficits, defining sensitive biomarkers for use in clinical trials and evaluating the disease course is challenging. **Objective:** To assess the effect of a 12-week strength and functional exercise protocol on the motor symptoms in individuals with hereditary spastic paraplegia and to analyze the sensitivity to change of the Clinician-Reported Outcome measures, performance outcomes, and Digital Health Technology. **Methods:** This is an adaptive cross-over randomized clinical trial. Initially, 20 patients diagnosed with HSP will be recruited. The study will be adapted based on an interim data analysis after 24 weeks, which will assess whether the effect is promising, favorable, or unfavorable through statistical analysis. Outcomes will be measured through the disease stage, Spastic Paraplegia Rating Scale, Ashworth Scale (Lower Limbs), muscle strength test (Medical Research Council), muscle strength test using manual dynamometry, Timed Up and Go, 10-meter walk test at maximum and self-selected speeds, kinematic gait evaluation, Clinical Global Impression of Change scale, Fall Efficacy Scale - Brazilian Version, Goal Attainment Scale, and the SF-36 Quality of Life Questionnaire. After randomization, the intervention group will have two online sessions per week, each lasting 60 minutes, for 12 weeks. Participants will be reassessed immediately at the end of the interventions, and group crossover will be conducted. Preliminary **Results:** To date, 10 participants have been included. The sample consists of 50% male participants. The mean age of participants is 46.9 years (±18.88), indicating a wide age range. The mean duration of the disease among participants is 19.0 years (±8.07). Regarding disease subtypes, 80% of participants were diagnosed with SPG4 and 20% with SPG10. Concerning the disease stage, 20% of participants are in stage 1, 40% in stage 3, and 40% in stage 4. We will have the results of the comparison between treatments only after the conclusion of the study. **Conclusion:** The findings of this study may contribute to the development of an exercise protocol aimed at treating motor symptoms associated with the disease. Furthermore, the results may assist in identifying the sensitivity to change and clinical meaningfulness of the changes in clinical outcome assessments and digital health technologies. This information is crucial for the design of symptomatic and disease-modifying therapy trials.

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Effect of organic purple grapes on behavioral tests of self-grooming and social interaction in an animal model of Valproic acid-induced autism

Ana Carolini Toporowicz Soares[1], Klaus Johann Jacques Schebek Teixeira[1], Kassielly Longo Brizola[1], Thayná Patachini Maia Becker[1], Alessandra Betina Gastaldi[1], Gabriela Borgmann[1], Katherine Plautz[1], Karine Louize Vincenzi Lüetke[1], Débora Delwing-dal Magro[2], Daniela Delwing-de Lima[1]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Fundação Universidade Regional de Blumenau. Blumenau SC, Brazil.

Correspondence: Ana Carolini Toporowicz Soares; Email: acstoporowicz@gmail.com.

Background: Autism spectrum disorder (ASD) is characterized by common patterns of social communication and restricted and repetitive sensorimotor behaviors of early onset, altered brain development and neural reorganization. ASD is influenced by the interaction of genetic and environmental factors, with possible prenatal exposure to toxic substances. In this sense, Valproic acid (VPA) significantly increased the incidence of autism in children of mothers who used this medication during the first trimester of pregnancy, with studies that show an increase in oxidative stress in ASD patients. Furthermore, the polyphenolic compounds present in organic purple grapes are known for their neuroprotective potential. **Objective:** To verify the effect of hydroalcoholic extract (HE) of organic purple grapes, at concentrations of 30, 100 and 150mg/kg, in modulating autistic-like behavior in male Wistar rats, induced by VPA. **Methods:** The project was approved by the Research Ethics Committee (CEUA 02/2022). The pregnant females received 800mg/kg of VPA diluted in 0.9% saline solution, intraperitoneally, in a single administration in the prenatal period (12th day of pregnancy). The male offspring were subjected to behavioral tests of self cleaning and social interaction to confirm the motor and psychotherapeutic responses typical of ASD. After, received treatment with the extract from organic purple Vitis vinifera at concentrations of 30mg/kg, 100mg/kg and 150mg/kg or saline as a control, via gavage, for 15 days and

underwent behavioral tests again to evaluate responses to treatment. In the end, they were euthanized by decapitation. **Results:** Behavioral tests showed stereotypy in the autistic group in the self-grooming test, and HE 150mg/kg decreased movement time, reducing repetitive behavior. In the social interaction test, the interaction time with the animal was shorter in the autistic group compared to the autistic control group. When evaluating interaction time with the object, the autistic group spent more time with the object than the autistic control group. HE at all concentrations reversed this time when compared to the autistic control group. **Conclusion:** This study demonstrated that the HE of organic purple grapes can modulate autistic-like behaviors in rats induced by AVP. The extract, in different concentrations, were effective in reducing repetitive behaviors and improving social interaction. These findings suggest a neuroprotective potential of polyphenolic compounds present in grapes, offering a promising perspective for future adjuvant therapies in the treatment of ASD.

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Effect of organic purple grapes on brain antioxidant enzyme activity in an animal model of autism induced by Valproic acid

Ana Carolini Toporowicz Soares[1], Klaus Johann Jacques Schebek Teixeira[1], Luana Gabriely de Almeida Campos[1], Mirela Parissa Bondan Misagh[1], Gustavo Schuetzler Gomes Fernandes[1], Thayná Patachini Maia Becker[1], Alessandra Betina Gastaldi[1], Natalia Cavichioli[1], Débora Delwing-dal Magro[2], Daniela Delwing-de Lima[1]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Fundação Universidade Regional de Blumenau. Blumenau SC, Brazil.

Correspondence: Ana Carolini Toporowicz Soares; Email: acstoporowicz@gmail.com.

Background: Developmental disorders have been the focus of neuroscience studies for many years and autism spectrum disorder (ASD) has received prominence in research over the last 30 years. The Valproic acid (VPA) significantly increased the incidence of autism in children of mothers who used this medication during the first trimester of pregnancy and studies demonstrate an increase in oxidative stress in individuals with ASD. The polyphenolic compounds present in grapes cause an increase in brain antioxidant activity. **Objective:** To reproduce autistic-like behavior in rats induced by VPA and evaluate the protective effects of hydroalcoholic extract (HE) of organic purple grapes at concentrations of 30, 100 and 150mg/kg on oxidative stress parameters in brain structures (cerebral cortex and cerebellum). **Methods:** The project was approved by the Research Ethics Committee (CEUA 02/2022). The pregnant females received 800mg/kg of VPA diluted in 0.9% saline solution, intraperitoneally, in a single administration in the prenatal period (12th day of pregnancy). The male offspring underwent behavioral tests to confirm the motor and psychotherapeutic response of autism. They were then administered HE (30mg/kg, 100mg/kg and 150mg/kg) or saline, via gavage, for 15 days, and were euthanized by decapitation to remove the cerebral cortex and cerebellum for analysis of the enzymatic antioxidant activity of superoxide dismutase (SOD), catalase (CAT) and glutathione peroxidase (GSH-Px). Data were analyzed by one-way ANOVA, followed by the Duncan post hoc test, when indicated ($p<0.05$). **Results:** Analysis demonstrated that ASD decreased SOD activity, while HE at a concentration of 30mg/kg partially reversed it and at concentrations of 100mg/kg and 150mg/kg completely reversed the enzymatic decrease in the cerebral cortex, while in the cerebellum there was no alteration in this enzyme. The model and administration of HE at different concentrations did not alter CAT activity in the cerebral cortex and cerebellum and GSH-Px activity in the cerebral cortex. On the other hand, GSH-Px activity in the cerebellum decreased in the group of animals with ASD. HE at a concentration of 30mg/kg partially reversed and at 100mg/kg and 150mg/kg, completely reversed the enzymatic decrease in rats with ASD. **Conclusion:** The animal model of autism-like behavior altered enzymatic antioxidant activity in the cerebral cortex and cerebellum of rats and the HE from organic purple grapes reversed the alterations in the brain, indicating its antioxidant capacity.

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Effect of theta burst transcranial magnetic stimulation associated with transcranial direct current stimulation to improve unilateral spatial neglect after stroke: a randomized controlled trial study protocol

Luana Aparecida Miranda[1], Fernanda Cristina Winckler[1], Gabriela Vieira de Paula[1], Tainá Gabriela Ricci[1], Natália Cristina Ferreira[1], Ricardo Silva Carvalho[1], Luiz Felipe Gomes Queiroz[1], Silméia Garcia Zanati Bazan[1], Gustavo José Luvizutto[2], Rodrigo Bazan[1]

[1]Universidade Estadual Paulista, Faculdade de Medicina de Botucatu, Botucatu SP, Brazil.
[2]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Ricardo Silva Carvalho; Email: carvalho.ricardo.med@gmail.com.

Background: Approximately 38% of patients who survive right hemisphere stroke present perception deficits, characterized by the syndrome of unilateral spatial neglect, with a likelihood of poor functional prognosis. Transcranial magnetic stimulation, or transcranial direct current stimulation, is employed in these patients to rebalance hemispheric activity. **Objective:** To assess the safety and efficacy of continuous theta burst transcranial magnetic stimulation in the left parietal cortex followed by anodic transcranial direct current stimulation in the right parietal cortex associated with rehabilitation to improve unilateral spatial neglect. **Methods:** This is a randomized, double-blind, multicenter clinical trial involving adult patients with unilateral spatial neglect following stroke, who will be randomized into three groups: Priming Group - 17 patients will receive continuous theta burst transcranial magnetic stimulation in the left parietal region followed by anodic transcranial direct current stimulation in the right parietal lobe; Conventional Group - 17 patients will receive anodic transcranial direct current stimulation in the right parietal region, and Sham Group - 17 patients will receive transcranial magnetic stimulation and sham transcranial direct current stimulation in the right parietal region, across 15 sessions distributed over 3 sessions per week lasting 30 minutes each. All groups will undergo physical therapy sessions, with assessments conducted at screening, after 8 sessions, and at the end of treatment. Follow-up of these patients will occur at 3, 6, and 12 months after the last session. **Results:** Based on the results of these studies, it will be possible to determine the effectiveness of the concomitant use of two transcranial stimulation techniques–continuous theta burst transcranial magnetic stimulation in the left parietal cortex followed by anodal transcranial direct current stimulation in the right parietal cortex–in improving unilateral spatial neglect, functional disability, quality of life, and self-efficacy in post-stroke patients. **Conclusion:** This is the first proof-of-concept randomized controlled trial to assess the effects of theta burst transcranial magnetic stimulation combined with anodic transcranial direct current stimulation on the perceptual performance post-stroke. Due to the combined clinical and functional measurements, this trial is intended to provide insights into the underlying post-stroke recovery processes and whether these can be influenced by combined therapies.

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Effect of transcranial direct current stimulation on the cognitive function of individuals with Alzheimer's disease: a systematic review with meta-analysis and meta-regression

Suellen Marinho Andrade[1], Clébya Candeia de Oliveira Marques[1], Larissa Coutinho de Lucena[2], Karina Vieira da Costa[1], Irinaldo Capitulino de Souza[1], Camila Beatriz da Silva Machado[1], Maria Eduarda Bezerra Sales Queiroz[1], Larissa Pereira Costa[1], Stephano Tomaz da Silva[1]

[1]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[2]Instituto Santos Dumont, Macaíba RN, Brazil.

Correspondence: Larissa Coutinho de Lucena; Email: larissa.lucena@isd.org.br.

Background: Alzheimer's disease (AD) leads to cognitive decline and loss of functional capacity. Transcranial direct current stimulation (tDCS) emerge as a potential therapy due to its ability to modulate cortical excitability. However, varying protocols and individual differences can hinder their effectiveness. Standardizing the most appropriate stimulation and effective parameters, such as intensity, polarity, frequency, targets, and duration, can help establish a reliable protocol for healthcare professionals, addressing current inconsistencies. **Objective:** To analyze the effects of transcranial direct current stimulation (tDCS) on the cognitive function and functional capacity of individuals with AD. **Methods:** This systematic review with meta-analysis and meta-regression included randomized clinical trials published until May 2022. The review protocol is registered in PROSPERO (CRD42020151189) and follows PRISMA guidelines. The studies involved individuals with AD, aged 55-85, treated with tDCS and compared with a placebo group. Subgroup analysis and meta-regression were conducted to address significant heterogeneity ($I2>75\%$) by considering moderating variables. **Results:** A total of 166 participants were subjected to active tDCS or sham stimulation, with an average age ranging from 64.22±(3.64) to 81.6±(8.0), and there were no participants with advanced-stage AD. In the studies included in the systematic review, there was a difference in the number of sessions, which ranged from a single session to 10 sessions; the stimulation time per session ranged from 15 minutes to 30 minutes; the total duration of the treatment ranged from one day to two weeks and all the studies investigated the anodal modality. A meta-analysis was conducted, which showed that tDCS did not have a greater effect than placebo on cognitive function in AD (standardized mean difference=0.69; 95% CI 0.00 to 1.38; $p=0.05$). The subgroup analysis conducted for the type of stimulation included studies that employed anodal tDCS and cathodal tDCS. From the results of this analysis, statistical significance was observed for the experimental group in the studies that investigated the application of anodal tDCS (standardized mean difference=0.76; 95% CI 0.20 to 1.31; $p=0.008$; $I^2=78\%$). Meta-regression suggested a significant influence of anodic tDCS with current intensity of 1.5 mA on cognitive function ($p=0.001$). **Conclusion:** Anodal tDCS have demonstrated the ability to improve cognitive function in AD by modulating neural activity. These therapies are safe and well-tolerated, being promising as adjuncts to available pharmacological treatments. Studies with greater methodological rigor and parameter standardization are warranted.

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Effect of vitamin D3 isolated or associated with donepezil on the spatial memory of adult and aged ovariectomized rat

Ana Paula Benedet[1], Laura Ceolin de Jesus[1], Eduarda Behenck Medeiros[1], Andrielly Vargas Lidio[1],

Gabriel Casagrande Zabot[1], Gabriela Piovesani Fenilli[1], Amanda Boaventura[1], Luisa Rosler Grings[1], Helder Robson de Oliveira Chaves Junior[1], Josiane Budni[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Ana Paula Benedet de Carvalho; Email: anapbenedet@hotmail.com.

Background: The prevalence of neurodegenerative diseases has been increasing due to longer life expectancy, with women being disproportionately affected 2 to 3 times more often following menopause. Research has shown an association between hypovitaminosis D and several age-related conditions, including cognitive decline. **Objective:** To assess the impact of vitamin D, either alone or in combination with donepezil, on spatial memory in adult and aging ovariectomized rats. **Methods:** Under protocol number 123/2019 and with approval from the Ethics Committee for Animal Use (CEUA) of the university, female Wistar rats aged 60 and 120 days underwent ovariectomy (OVX). They received treatment 1, 4, or 8 months post-surgery. They were administered chronic doses of vitamin D (42IU/kg or 420IU/kg) and donepezil (1mg/kg), independently or in combination, for 21 days. Spatial memory was evaluated using the octagonal maze from day 18 to day 22 of the experiment. **Results:** The results revealed that 60-day-old rats subjected to OVX for 1 or 4 months did not display impairment in spatial memory. However, those aged 60 days and subjected to OVX for eight months exhibited spatial memory impairment, which was mitigated over time by administration of vitamin D at 420IU/kg alone or in combination with donepezil. Similarly, 120-day-old rats subjected to OVX for 1 or 4 months did not exhibit spatial memory impairment, whereas those subjected to OVX for eight months did show impairment. Treatment with vitamin D at 420IU/kg and donepezil alone improved spatial memory performance. Overall, rats subjected to longer OVX durations performed poorer in spatial memory tasks than controls. **Conclusion:** Vitamin D supplementation at 420IU/kg and donepezil, either alone or in combination, demonstrated neuroprotective effects concerning spatial memory in OVX rats.

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Effective management of diabetic peripheral neuropathy associated with wound healing: a case report

Gustavo Gomboski[1], Jessica Vargas Lopes[2], Magda de Souza Reis[1], Renita Baldo Moraes[1], Suzane Beatriz Frantz Krug[1], Eduarda Salton Grando[1], Marcus Vinicius Schefer[1]

[1]Universidade de Santa Cruz do Sul, Santa Cruz do Sul RS, Brazil
[2]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil

Correspondence: Jessica Vargas Lopes; Email: jessicavargasl@outlook.com.

Case presentation: An elderly male patient with a 10-year history of Type 2 Diabetes Mellitus (T2DM) presented with poorly controlled blood glucose levels, an unbalanced diet, and continuous treatment with insulin and oral hypoglycemics. He also had hypertension and hypercholesterolemia. The patient had a recurring acute lesion on his left hallux, originating from a keratinocyte callus, showing signs of inflammation and local infection, including lymphadenopathy in the leg and groin. Treatment involved removing the keratinocyte debris, cleansing the wound, applying a local disinfectant dressing, and starting oral antibiotic therapy. Continuous monitoring and antimicrobial dressings led to complete healing within 10 days without requiring amputation. **Discussion:** Diabetic peripheral neuropathy is a common and significant complication of diabetes mellitus. This condition results in a loss of sensation, making patients highly prone to repeated injuries, especially in the feet. Two types of nerve fibers can be affected in neuropathy: thin and thick. They can be damaged due to demyelination linked to metabolic defects, which affect Schwann cells and myelin production in the peripheral neurons. If thin fibers are injured, the patient may experience a decrease in general sensitivity, as well as paresthesia and hyperesthesia. Conversely, a lesion to thick fibers will result in a reduction in tactile perception. Peripheral neuropathy severely impairs protective sensation in the feet, making minor injuries such as small cuts or blisters go unnoticed and untreated. Poor blood sugar control and inadequate preventive care can lead to chronic wounds and infections, complicating the patient's condition. The recurrence of lesions at the same site highlights the critical need for early and continuous interventions. Proper cleaning and the use of advanced wound dressings, along with prompt antibiotic therapy, are crucial in controlling the infection and promoting rapid wound healing. Patient education on self-care and injury prevention is vital, especially for patients suffering from peripheral neuropathy. This ongoing follow-up not only supports wound healing but also reduces the risk of new injuries and serious complications, such as amputations. **Final comments:** Early and consistent intervention, along with patient education, proved effective in managing lesions associated with diabetic peripheral neuropathy. A multidisciplinary approach is essential for minimizing adverse effects and promoting efficient wound healing. This case underscores the importance of specialized follow-up and the use of advanced dressing technologies for infection control and health promotion. Regular monitoring and guidance to prevent recurrences are vital for improving the quality of life for diabetic patients.

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Effective use of botulinum toxin for neuropathic pain management in a neuroma: a case report

Matheus Parra Barcelos de Mendonça[1], Camille Scabora de Melo[1], Mariana Almeida Lima[1], Bruna Andrade Vigo[1], Gabriel Taricani Kubota[2], Bruno Hojo Rebouças[2]

[1]Universidade de São Paulo, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Matheus Parra Barcelos de Mendonça; Email: matheus.parra.barcelos@gmail.com.

Case presentation: This case presents a 66-year-old woman who suffered a deep second-degree and circumferential third-degree electrical burn on the left wrist and hand in 2017. She underwent left upper limb amputation at the elbow level and developed a neuroma at the residual limb involving the median and ulnar nerves. She exhibited phantom limb pain, neuropathic pain, myofascial nociceptive pain with trigger points in shoulder girdle muscles, as well as allodynia and hyperalgesia in the residual limb. The patient went through several interventions for pain control, such as mirror therapy, acupuncture, and pharmacotherapy with gabapentin, methadone, amitriptyline, pregabalin, and duloxetine without a satisfactory result. In December 2021, the patient underwent botox injections and neurolysis for treatment of the lateral neuroma, which resulted in significant symptom improvement. Since then, she has undergone other interventions, such as electrostimulation and nerve block. Currently, she is taking methadone, amitriptyline, and gabapentin daily, with dipyrone as needed. **Discussion:** A neuroma results from unorganized axonal proliferation in a nerve injury site, forming a non-neoplastic tumor containing glial cells, myofibroblasts, fibroblasts, and connective tissue. Usually, neuromas are clinically presented as small and oval nodules, manifesting neuralgic pain and touch hypersensitivity. When symptomatic, a neuroma can severely impact the quality of life and adequate treatment is of the utmost importance for reestablishing the patient's functionality. Usually, the preferred treatment is surgical excision of the tumor, but it's neither available for nor wanted by all patients. An alternative is pharmacotherapy using membrane-stabilizing drugs, such as anticonvulsants, corticosteroids, local anesthetic drugs, and non-steroidal anti-inflammatories. Unfortunately, a low proportion of patients achieve adequate symptom control with conservative treatment. Despite the few studies evaluating its effectiveness in this specific scenario, botulinum toxin (Btx) seems a promising alternative treatment. Btx therapy already shows evidence of effectiveness in the management of neuropathic pain and signs of its probable value in neuroma symptomatology control. **Final comments:** The presented case demonstrates the difficulties in obtaining adequate pain control in patients with neuroma, which highlights the necessity of combined pharmacological therapy and other treatment modalities, such as electrostimulation, nerve blockage, and Btx application. In that scenario, the usage of Btx in managing a symptomatic neuroma reached a satisfactory result, contributing to neuralgia reduction. Therefore, considering the difficulties of achieving adequate management for the condition, Btx likely is a valuable addition to the arsenal of drugs used in cases like that, despite the incipient literature currently available.

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Effectiveness of botox (onabotulinumtoxin) in the treatment of migraines: a systematic review

Jonathan Vinicius da Silva Casarim[1], Anaclara Azalin Mattoso[1], Beatriz Cardoso Bálsamo[1], Carolina Pimentel Ladeira Gomes[1], Clara Rodrigues Oliveira[1], Lucas Vital Dias Gomide[1], Luiza Bianca Monteiro Castro[1], Nathalia Carolina Silva Moreira[1], Ana Paula Ferreira[1], Djalma Rabelo Ricardo[1]

[1]Faculdade de Ciências Médicas e da Saúde de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Nathalia Carolina Silva Moreira; Email: nathalia.zsmoreira@gmail.com.

Background: Excessive use of medications such as analgesics, triptans and opioids is one of the main risks for transforming an episodic migraine into a chronic one. In this context, OnabotulinumtoxinA emerges as a promising alternative to prevent migraine chronification, acting to reduce the release of inflammatory neurotransmitters. **Objective:** To investigate the effectiveness of migraine treatment through the use of Botox (OnabotulinumtoxinA) through a systematic review. **Methods:** Controlled and randomized clinical trials published originally in English, over the last ten years, on humans were analyzed, using the National Library of Medicine (MedLine) database as a reference. The search for descriptors and terms used was carried out by consulting the Medical Subject Headings (MeSH), using its variations: "migraine disorders", "botox", "treatment". Studies involving men and women between 19 and 65 years of age diagnosed with chronic migraine were included. Studies that did not specifically use the substance OnabotulinumtoxinA, those with unclear or poorly described methods, and publications available only in the abstract were excluded. The PRISMA scale was used to systematize the scope of this review. **Results:** Initially, 973 studies were found. After applying the inclusion and exclusion criteria, nine articles were selected for the final analysis. In total, 1310 participants participated, 78.7% of whom were female, with an average age of 41.5±2.8 - 18 to 65 years old. Of the nine studies analyzed, eight indicated that the application of Botox reduced headache intensity, migraine frequency, and the intake of pain relief medications ($p<0.05$). Evidence points to the onset of treatment effects after 4 weeks ($p<0.05$). Furthermore, there was no difference in treatment between men and women ($p<0.05$). One of the studies showed that there was no benefit between the experimental group and

the control group (p=0.19). **Conclusion:** The systematic review demonstrated that migraine treatment with botox (OnabotulinumtoxinA) is effective in the long term. Furthermore, the use of Botox has been associated with a reduction in the frequency of migraines, the intensity of pain, and the consumption of medications for acute pain. Therefore, Botox can be an interesting alternative for migraine management, with significant implications for clinical practice and future research to better analyze these correlations.

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Effectiveness of DBS as a treatment for Alzheimer's disease: meta-analysis

Julia Scaramal[1], Samuel Simis[2], Paulo Henrique Pires de Aguiar[2]

[1]Pontifícia Universidade Católica de São Paulo, Sorocaba SP, Brazil.
[2]Pontifícia Universidade Católica de São Paulo, São Paulo SP, Brazil.

Correspondence: Julia Scaramal Mello; Email: julia.scaramal71@gmail.com.

Background: Alzheimer's disease (AD) currently lacks a successful treatment despite being one of the most costly forms of dementia. Recently, deep brain stimulation (DBS) has emerged as a key treatment for diseases such as Parkinson's due to the neuromodulation achieved by targeting specific brain areas. Herrington's theory suggests that DBS can modify pathological pathways by modulating local neuronal activity. Consequently, some studies have investigated the effectiveness of DBS as a treatment for Alzheimer's disease. **Objective:** To perform a systematic review with meta-analyses to determine whether DBS is effective as a treatment for Alzheimer's disease based on the available literature. **Methods:** The search was conducted in PubMed and Cochrane using the terms "DBS" AND "Alzheimer". Articles were included if they met the eligibility criteria: published within the last 10 years, written in English, and discussing both DBS and Alzheimer's disease. Case reports and animal studies were excluded. **Results:** 17 articles were included after independent review by 2 reviewers. Of these, only 6 were clinical trials. Data extraction and analysis indicated that patients with mild AD showed less worsening on the ADAS-cog (Alzheimer's Disease Assessment Scale-cognitive subscale) following DBS. The most studied DBS targets were the fornix (66.67%), followed by the nucleus basalis of Meynert (33.34%) and the ventral capsule/ventral striatum (16.67%). Additionally, some studies reported an improvement in local metabolism as shown by PET imaging (66.67%). For the meta-analysis, the change from baseline for the ADAS-Cog scale shows no significant difference in correlation values (mean change of 5.8 points), and the studies exhibit low heterogeneity. Regarding the MMSE scale, the analysis revealed a confidence interval of -3 to +2 and a mean change from baseline of -0.54 points, indicating that DBS, over one year of stimulation, delayed the progression of Alzheimer's disease on that scale. **Conclusion:** Preliminary findings suggest that DBS may have some effectiveness in treating AD.

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Effectiveness of deep brain stimulation in Huntington's disease: a comprehensive literature review

Victória Stephane Tessaro[1], Larissa Paixão Batista[2], Laura Gabriela Ribeiro Reis[3], Emilly Pinheiro da Cruz[4], João Vitor da Silva Viana[5], Gean Gimenes Moura[6], Igor Joaquim Flores Barreto[7], Mariana Farias de Araujo Athayde[8], Verônica Oliveira Rodrigues[9], Karoline Schereck Kreitmeyer[10]

[1]Universidade de Fortaleza, Fortaleza CE, Brazil.
[2]Escola Superior de Ciências da Saúde do Distrito Federal, Brasília DF, Brazil.
[3]Centro Universitário FIPMoc, Montes Claros MG, Brazil.
[4]Universidade do Estado do Pará, Santarém PA, Brazil.
[5]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[6]Faculdade Multivix, Cachoeiro de Itapemirim ES, Brazil.
[7]Faculdade União Araruama de Ensino, Arurama RJ, Brazil.
[8]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[9]Universidade Federal do Pará, Altamira PA, Brazil.
[10]Santa Casa da Misericórdia de Limeira, Limeira SP, Brazil.

Correspondence: Victória Stephane Tessaro; Email: stephanetessaro.2020@gmail.com.

Background: Huntington's disease is an autosomal dominant neurological disorder that progressively affects cognition, motor function, and behavior. It is associated with a mutation in the HTT gene, responsible for encoding a protein involved in the development of the central nervous system. Neurodegeneration primarily affects striatal neurons, with characteristic symptoms including Huntington's chorea (involuntary muscle contractions), impaired attention and emotion recognition, apathy, irritability, and depression. Deep brain stimulation (DBS), for instance, has shown promise in treatment. This involves implanting electrodes that deliver electrical impulses to specific regions of the brain to modulate neural activity. **Objective:** To evaluate the efficacy and safety of DBS in treating Huntington's disease. **Methods:** A literature review was conducted focusing on studies published in the last 10 years investigating DBS application in Huntington's disease patients, using databases such as PubMed and the keywords "motor disorder," "Huntington's disease," and "deep stimulation." **Results:** Out of 27 studies, 4 were analyzed, indicating that DBS, particularly when targeted at the subthalamic nucleus and the internal globus pallidus, may lead to significant improvements in chorea, dystonia, and rigidity, which are debilitating motor symptoms in up to 80% of cases of Huntington's disease. Patients treated with DBS showed reduced severity scores of motor symptoms and improved daily functionality, suggesting that the intervention could enhance patients' quality of life. In addition to motor benefits, the review identified positive effects on psychiatric and cognitive symptoms associated with the disease. There were reports of improved emotional stability, reduced irritability and aggression, and in some cases, cognitive improvements. However, results are variable and depend on factors such as precise electrode placement, stimulation settings, and treatment duration. **Conclusion:** DBS appears to be a promising technique in treating Huntington's disease, potentially improving symptoms and restoring functional abilities in daily activities and routines of patients. Nonetheless, further studies are needed to analyze variability factors in the application of this aproach.

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Effectiveness of dordaviprone in H3 K27M-mutant diffuse glioma: a systematic review and proportional meta-analysis

Felipe Araujo Gouhie[1], João Pedro Alves Rodrigues[1], Lucas Figueira Vieira[1], Caio Luiz Nazar Cunha[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Felipe Araujo Gouhie; Email: felipeagouhie@gmail.com.

Background: Pediatric patients with Histone 3 (H3) K27M-mutant glioma have a poor prognosis despite current therapies. Dordaviprone is a first-in-class imipridone designed to treat resistant gliomas. However, there is a lack of literature studying the relationship between this new agent and this patient population. Therefore, the present study aims to explore the effects of Dordaviprone on the response rate in glioma-diagnosed patients. **Objective:** To systematically analyze existing literature to determine the efficacy of Dordaviprone (ONC201) in pediatric patients with H3 K27M-mutant glioma, comparing patients' states before and after treatment. **Methods:** We performed extensive literature searches using PubMed, Dimensions, and Cochrane databases. The assessed outcome was the overall response rate for patients undergoing Dordaviprone treatment. Statistical analysis was conducted using R Studio 4.3.1, and study heterogeneity was assessed using the I^2 index. **Results:** Five interventional studies were included in the analysis, comprising a total of 71 patients. The response rate with Dordaviprone in patients with H3 K27M-mutant diffuse glioma was 26.97% (95% CI 11.27% to 52.77%). **Conclusion:** The meta-analysis indicates a moderate overall response rate across the included studies, with significant heterogeneity observed among the results. This variability suggests differing study methodologies, population characteristics, or other contextual factors may influence the response rates.

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Effectiveness of dual task in a combined training on quality of life and concern about falling in individuals with Parkinson's disease: randomized controlled trial

Alessandra Cattaneo Estrada Melanda[1], Rogério José de Souza[1], Amanda Lima Nogueira dos Anjos[1], Patrícia Gonçalves Broto da Silva[1], Milena Marie Matias Martins[2], Samuel Lidione da Silva[2], Ana Raquel Lindquist[3], Adriana Costa-Ribeiro[4], Suellen Mary Marinho dos Santos Andrade[4], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.
[2]Centro Universitário Filadélfia, Londrina PR, Brazil.
[3]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[4]Universidade Federal da Paraíba, João Pessoa PB, Brazil.

Correspondence: Amanda Lima Nogueira dos Anjos; Email: amandalima.nd@gmail.com.

Background: Studies on quality of life (QOL) and concern about falling in Parkinson's disease (PD) generally seek to understand how motor and non-motor symptoms impact these outcomes. As these are complex and multifactorial outcomes, it is necessary that QoL and concerns about falling take center stage in clinical studies, as well as which therapeutic approaches are most effective and whether there is additional benefit in combined therapies for their management. **Objective:** To verify the effectiveness of dual-task training (DT) combined with treadmill gait training and anodal transcranial direct current electrical stimulation (tDCS) on QoL and concern about falling in individuals with PD. **Methods:** This is a randomized controlled, blind clinical trial, consisting of 37 individuals with mild to moderate PD and without cognitive impairment, allocated into two groups: 1) Experimental Group (EG) that performed DT+ gait training in treadmill + anodal tDCS) and 2) Control Group (CG) that performed gait training on a treadmill + anodal tDCS). 12 intervention sessions were carried out, three times a week. DT training was based on the application of a protocol consisting of 3 levels, with 6 tasks at each level and with gradual evolution of complexity, applied during 18 minutes of walking training on the treadmill. The current was applied to the left dorsolateral prefrontal cortex, with an intensity of 2 mA, concomitantly with gait training. To assess quality of life, the Parkinson's Disease Questionnaire (PDQ 39) was used and concern about falling was assessed using

the International Falls Efficacy Scale (FES-I). Assessments were carried out at three moments: pre-intervention, post-intervention and after a 4-week follow-up period. The ANOVA tow-way of repeated measures was used to compare groups according to intervention, time and group vs time interaction. CAAE:45459521.6.1001. 5188. **Results:** There was a significant improvement in QoL in the EG (time effect) between the pre- and post-intervention moments, in the domains: mobility, emotional well-being and total PDQ-39 score; and between the pre-intervention and follow-up moments in the cognition domain and in the PDQ-39 total score. For the CG, there was an improvement between the pre- and post-intervention moments in the areas of body discomfort and total PDQ-39 score. No differences were observed between groups. However, there was an interaction of time vs. group, only for the cognition domain (p=0.015), with improvement between the pre- and follow-up moments, in favor of the EG (d=-0.71). Regarding the concern about falling measured by FES-I, there were no significant differences when considering the factor time, group and the interaction time vs. group. **Conclusion:** The addition of DT to a combined protocol of treadmill gait training and anodal tDCS was not effective in improving QoL and fear of falling in individuals with PD, with the exception of the cognition domain of the PDQ-39.

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Effectiveness of GLP-1 receptor agonists in Parkinson's disease: a systematic review and meta-analysis

Yago Marcos Pessoa-Gonçalves[1], Victor Hugo Palhares Flávio Reis[1], Chamberttan Souza Desidério[1], Marlos Aureliano Dias-Sousa[2] , Carlo José Freire Oliveira[1], Rubens Gisbert Cury[3], Wellington Francisco Rodrigues[1]

[1]Universidade Federal do Triângulo Mineiro, Instituto de Ciências Biológicas e Naturais, Laboratório de Imunologia e Bioinformática, Uberaba MG, Brazil.
[2]Universidade Federal do Triângulo Mineiro, Hospital das Clínicas, Departamento de Neurologia, Uberaba MG, Brazil.
[3]Universidade de São Paulo, Departamento de Neurologia, Grupo de Distúrbios do Movimento, São Paulo SP, Brazil.

Correspondence: Yago Marcos Pessoa Gonçalves; Email: yagompg98@gmail.com.

Background: Animal studies have shown that Glucagon-like Peptide 1 (GLP-1) exerts neuroprotective effects on dopaminergic neurons in the substantia nigra and promotes functional recovery in animals. These preclinical findings suggest that GLP-1 receptor agonist drugs could be promising for treating Parkinson's disease (PD). **Objective:** To investigate the efficacy of GLP-1 agonists in the treatment of PD in randomized clinical trials. **Methods:** This study is a meta-analysis of randomized controlled trials that evaluated the efficacy of GLP-1 in PD. Embase, Pubmed and Cochrane were searched for trials published up to April 2024. Data were extracted from published trials and Cochrane tool for assessing risk of bias was employed. The extracted data were input into Review Manager (RevMan) 5.3 (Nordic Cochrane Centre, Denmark) to calculate the effect size. RevMan calculated as effect measure the mean difference with a 95% confidence interval (CI) using the inverse variance statistical method. **Results:** Regarding the risk of bias in each RCT, three were assessed to have an overall low risk of bias. The meta-analysis showed no significant overall improvement across all parts of the Movement Disorders Society Unified Parkinson's Disease Rating Scale (MDS-UPDRS). In Part I, which evaluates non-motor aspects of daily life, the MD between the intervention and control groups was -0.17; 95% CI -0.83, 0.50; p = 0.62. Part II assesses motor aspects of daily living and showed a MD of -1.67; 95% CI -3.42, 0.12; p=0.07. Part III, the motor scale, showed a MD of -1.75; 95% CI -3.54, 0.05; p=0.06 and finally the part IV, which assesses motor complications, showed a MD of -0.09; 95% CI -0.46, 0.28; p=0.64. However, subgroup analyses of studies with a minimum of 12 months of GLP-1 intervention revealed a significant improvement in motor symptoms (MDS-UPDRS part III) compared the control groups (MD -2.34; 95% CI -4.53, -0.14; p=0.04). In terms of levodopa equivalent daily dose, the MD between the intervention and control groups was not significantly different (MD, -42.11; 95% CI, -94.34, 10.11; p=0.11). Regarding cognitive functioning measured using the MATTIS-DRS2 scale, the MD between the intervention and control groups did not demonstrate a significant difference (MD, 1.60; 95% CI, -0.55, 3.75; p=0.14). In addition, no significant difference in quality of life (PDQ-39) was observed (MD -0.81; 95% CI -2.85, 1.23; p=0.44). **Conclusion:** GLP-1 interventions, particularly those lasting 12 months or longer, can effectively improve motor symptoms in PD patients as measured by MDS-UPDRS part III. These findings underscore the potential of GLP-1 in PD treatment and highlight the need for future clinical trials with larger numbers of participants and longer intervention periods. Such trials should focus on specific subgroups to better understand the full effect of GLP-1 agonists on PD management.

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Effectivity of tafamidis in the control of neurological deficits in patients with familial amyloid polyneuropathy: a systematic review and meta analysis

Emanuela Freire Caetano Davi[1], Caio Levi Mendes Costa[1], Mariana Borges Falcão Costa[1], Leonardo Elias Araujo dos Santos[1], Francisco Duque de Paiva Giudice Junior[1], Leonardo José Rodrigues de Araújo Melo[1], Mariana Alcântara Tavares[1], João Marcelo Albuquerque Beserra de Sousa[1], João Pedro de Oliveira Gouveia Marcotti[1], Kauane Emilly Ribeiro Aureliano[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Emanuela Freire Caetano Davi; Email: emanueladavi@alu.ufc.br.

Background: Familial Amyloid Polyneuropathy (FAP) is a progressive hereditary disease caused by the deposition of the protein transthyretin (TTR) in peripheral nerves and other tissues. Tafamidis meglumine, a stabilizer of TTR deposition, is the only medication for the treatment of FAP available on SUS. **Objective:** to evaluate the impacts of Tafamidis meglumine on the neurological deterioration in patients with FAP measured by NISS-LL and its statistical significance. **Methods:** This systematic review was executed according to the PRISMA 4 checklist, with search on the databases PUBMED, Scielo and EMBASE with the terms "transthyretin familial amyloid polyneuropathy" AND "Tafamidis" in portuguese or english for controlled trials with "neurology" filter, extracting 65 studies. Eligibility criteria were analysis of effectiveness of isolated Tafamidis 20 mg on humans diagnosed with PAF on neurological presentations.The effect measures analyzed for outcome were the population, baseline and mean difference of NISS-LL points, confidence intervals and p value. Certainty of results was evaluated with the GRADE tool and evaluation of bias with the RoB2 guidelines. The software Rayyan was used to automate exclusion, discarding 41 studies, and after manual screening, 12 studies were selected for inclusion. **Results:** 5 studies offered data of randomized controlled trials with comparison of tafamidis vs placebo, and 6 open-label studies, including extensions. Total population across studies was 282 patients, mostly caucasian, not accounting for 1 study due to possible repetition. Meta Analysis of tafamidis vs placebo was done using the mean difference analysis on Jamovi. The group's baseline NISS-LL differences weren't statistically significant, with the mean baseline NISS-LL of Tafamidis groups being 8.7 and 9.22 in control groups. Slight but statistically significant favorability (p<0.001) was found to the Tafamidis group, with the mean difference of NISS-LL from baseline across studies being - 0.39 NISS-LL points (CI, -0.547, -0.251). Open-label long term follow-ups varied between 12-66 months with overall the median NISS-LL change was 2.67 points (CI, 1.75, 3.58). Mean change of NISS-LL from baseline was 1.65 points in 12 months (CI, 0.549, 2.74), 2.21 in 24 months (CI, 0.60, 3.81) and 6.33 points in 48 months (CI, 2.89, 9.76). In all studies, the previous placebo-group presented a higher NISS-LL baseline and smaller rate of response to medication, indicating considerable impact of earlier start to treatment in prognosis. **Conclusion:** This systematic review showed modest benefit to the use of Tafamidis to control neuropathy in FAP and evidence of sustainability in long term treatment. The review limitations were the scarcity of present literature, with possible publication bias, as well the limitations in studies due to limited population and drop out rates. Due to this, the certainty of results with the GRADE tool was considered as class III.

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Effects of cannabigerol on neuroinflammation and spatial memory in mice with cerebral ischemia

José Guilherme Pinhatti Carrasco[1], Nathalia Akemi Kohara[1], Luis Fernando Fernandes Miranda[1], Rúbia Maria Monteiro Weffort de Oliveira[1], Cristiano Correia Bacarin[1]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil

Correspondence: José Guilherme Pinhatti Carrasco; Email: joseguicarrasco@gmail.com.

Background: Cerebral ischemia is an important morbidity factor worldwide. The abrupt deprivation of cerebral blood flow, as occurs in stroke and reversible cardiac arrest, triggers a series of pathophysiological events such as excitotoxicity, oxidative stress and neuroinflammation that culminate in neuronal death and white matter injury. Despite its significance, there are still few pharmacological alternatives available for cerebral ischemia, making it necessary the development of therapeutic strategies that demonstrate good efficacy and safety. In this sense, cannabigerol (CBG), a non-psychoactive phytocannabinoid, has been studied for its anti-inflammatory and neuroprotective properties demonstrated in in vitro and in vivo studies. **Objective:** Evaluate the anti-inflammatory and anti-amnesic effects of different doses of CBG through an animal model of transient global cerebral ischemia (tGCI). **Methods:** For the experiments, male C57BL/6 mice were used. All experimental protocols were approved by the ethics committee. The tGCI was induced by bilateral common carotid artery occlusion (BCCAO) for 20 minutes. Animals in the sham group underwent the same procedure, but without occlusion. CBG, at doses of 1, 5 and 10 mg/kg, or vehicle were administered intraperitoneally for 7 days, with the first dose given 1 hour after BCCAO. The anti-amnesic effect of CBG was evaluated through the object location task (OLT), which was conducted between the 10th and 14th day after BCCAO, with intervals of 1, 4, and 24 hours to calculate the discrimination index D2 and observe memory retention. On the 15th day after the ischemic event, the animals were euthanized and their brains were removed and processed for immunohistochemical analyses. The anti-inflammatory effect of CBG was analyzed by quantifying the integrated optical density of Iba-1 in the CA1 and CA3 subfields of the hippocampus. **Results:** In the OLT, only at the 24-hour interval and in the group treated with CBG 1mg/kg, there was an increase in discrimination index D2 scores compared to the vehicle-treated group (p=0.0314), indicating an anti-amnesic effect of this dose on long-term memory. Regarding neuroinflammation, compared with the vehicle, the treatment with 1mg/kg of CBG attenuated the consequence of OBACC in the CA1 (p=0.0089) and CA3 (p=0.0235) subfields of the hippocampus, possibly due

to an anti-inflammatory effect. However, doses of 5 and 10 mg/kg of CBG did not modify the immunoreactivity of Iba-1 in the CA1 and CA3 subfields when compared with the BCCAO + Vehicle group (p>0.05). **Conclusion:** CBG exhibited anti-amnesic and anti-inflammatory effects after BCCAO at the dose of 1mg/kg, but not at doses of 5 and 10mg/kg, indicating a dose-dependent neuroprotective effect of the compound.

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Effects of laser and ultrasound therapies on sciatic nerve axonotmesis: an experimental study

Natália Rebeca Alves de Araújo Karpejany[1], Danielle Cortellazzi Colonna Romano[2], Taianara Sampaio Reis[1], Juliana Do Nascimento Sousa[3], Vinícius Santana Vasconcelos[3], Allan Dellon da Silva[4], Antônio Luiz Martins Maia Filho[4], Wellington dos Santos Alves[4], Danniel Cabral Leão Ferreira[4], Ester Laignier Barroso[5]

[1]Universidade Federal do Piauí, Hospital Universitário, Teresina PI, Brazil.
[2]Universidade Nove de Julho, São Paulo SP, Brazil.
[3]Universidade Federal do Piauí, Teresina PI, Brazil.
[4]Universidade Estadual do Piauí, Teresina PI, Brazil.
[5]Centro Universitário Uninovafapi, Teresina PI, Brazil.

Correspondence: Natália Rebeca Alves de Araújo Karpejany; Email: nataliarebeca2607@gmail.com.

Background: Peripheral nerve injuries affect sensory and motor functions, with the sciatic nerve is commonly affected. Axonotmesis is a type of nerve injury in which the connective tissue surrounding the nerve's axons remains intact and represents the majority of cases of peripheral nerve injuries. Peripheral nerve recovery can be optimized by Low-Level Laser Therapy (LLLT), known for its bio-photomodulatory effect capable of accelerating the axonal regeneration process. Therapeutic ultrasound (UST) emits waves that improve blood flow and oxygen exchange, increasing cellular metabolism. This therapy can be applied to help reduce inflammatory processes and promote peripheral nerve regeneration by increasing nerve fiber density and stimulating axoplasm production. **Objective:** To analyze the effects of LLLT and UST on the regeneration process after axonotmetic injury of the sciatic nerve in Rattus norvegicus. **Methods:** This experimental, analytical, quantitative study investigated 30 healthy male rats (Rattus norvegicus). Rats were randomly divided into five groups: G1 (positive control, no lesion or treatment), G2 (negative control, with lesion but no treatment), G3 (lesion and treated with low-level laser therapy, LLLT), G4 (lesion and treated with ultrasound therapy, UST). All animals were anesthetized and a sciatic nerve injury was induced. G1 received no treatment, G2 received no treatment post-lesion, G3 received LLLT daily for 17 days, and G4 received UST daily for 17 days. Gait was assessed preoperatively, on postoperative days 1, 7, 14, and 21 using the Sciatic Functional Index (SFI) derived from footprint analysis. Data were analyzed using ANOVA and Tukey's test, with p<0.05 considered significant. **Results:** Preoperatively, all groups had Sciatic Functional Index (SFI) values near 0, indicating normal gait. Significant differences were observed between the lesioned groups and the positive control on postoperative days 1 and 7 (P<0.05). By day 14, only the ultrasound group differed significantly from the negative control (P=0.010271293). By day 21, no significant differences in gait were found between treated groups and controls. The laser group showed significant differences preoperatively and on all postoperative days, but did not fully recover by day 21. The ultrasound group showed significant recovery by day 14 (P=0.081191726). **Conclusion:** Intergroup analysis showed no practical difference between low-level laser therapy and therapeutic ultrasound treatment. But, when comparing the different treatment phases within the same experimental group (intragroup analysis), only therapeutic ultrasound showed better results in the recovery of gait and motor function of the crushed sciatic nerve on the 14th postoperative day. It is suggested that research on the subject continue to define analysis protocols and methodologies for future studies comparing the effects of low-level laser therapy and therapeutic ultrasound for nerve regeneration.

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Effects of melatonin use in children with sleep disorders: a systematic review

Ana Luiza Rossetto[1], Samyra Roberta Assis Souza[1], Maria Gabriella Vido Dutra[1], Maria Paula Vitalino Faria da Silva[1], Leila Maria Guissoni Campos[1]

[1]Universidade de Marília, Marília SP, Brazil.

Correspondence: Ana Luiza Rossetto; Email: rossettoanaluiza23@gmail.com.

Background: Sleep is a functional state that involves important neurological processes. It is known that prolonged sleep deprivation can result in serious physical and behavioral impairments, as well as metabolic and immunological changes. In children, the repercussion of sleep disturbances can be characterized by reduced performance and learning. When the biological causes are related to endocrine problems, such as abnormal production of the melatonin hormone, the use of exogenous melatonin becomes an option for treating sleep disorders. **Objective:** To analyse the effect of exogenous melatonin use in children as a form of treatment for sleep disorders. **Methods:** A systematic review was conducted by selecting studies from the BVS-PubMed database published between 2019 and 2024. A total of 238 studies were found, of which 42 were selected for analysis. Articles in English and/or Portuguese that addressed the effects of pharmacological use of melatonin in children with sleep disorders were used. The descriptors used were "Sleep" AND "Melatonin" AND "Children" AND "Sleep disorders". Articles that did not fit the inclusion criteria, such as animal studies, narrative reviews, and those unrelated to the review topic, were excluded. **Results:** The use of melatonin was indicated as a treatment method in different groups of children, such as those with neurodevelopmental disorders, intellectual disabilities, learning and motor disorders, communication disorders, and attention deficit disorder. A decreased sleep onset latency was observed, along with improvement in morning temperament, irritability, hyperactivity, and inappropriate speech. When melatonin use was combined with sleep hygiene (fixed bedtime routine, bedtime stories, avoiding light at night, calming music, among others), significant improvements were observed in sleep onset, anxiety, delayed sleep onset, sleep duration, night awakenings, and parasomnias, compared to the placebo group. Side effects were observed in a few cases, with mild symptoms characterized by headaches. **Conclusion:** The use of melatonin may represent an improvement for sleep disturbances; however, when combined with sleep hygiene, the results appear to be more promising. The quantity of melatonin - timing/dosage should be carefully evaluated to avoid possible side effects.

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Effects of mental practice on fog in people with Parkinson's disease: a randomized clinical trial

Paloma Rodrigues da Silva[1], Karina Yumi Takayama Honda[1], Isaira Almeida Pereira da Silva Nascimento[1], Katia Cirilo Costa Nobrega[1], Nathalia de Brito Pereira[1], Thayane Habache Barolli[1], Maria Elisa Pimentel Piemonte[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Paloma Rodrigues da Silva; Email: paloma.rsilva@usp.br.

Background: Freezing of gait (FOG) significantly impairs the quality of life in Parkinson's disease (PD). Mental practice (MP) is an emerging field in neurorehabilitation and Dynamic Neuro-Cognitive Imagery (NCI) is a new approach that combines MP with physical practice (PP). Despite limited research on NCI-based interventions, they are considered safe and effective for remote administration in individuals with PD. **Objective:** To compare the effects of an experimental intervention (EI) based on NCI with a control intervention (CI) of equal volume and intensity, differing only in mental practice components, on PD-related FOG. **Methods:** A randomized, controlled, double-blind trial with 23 participants, mean age 54, H&Y stages II/III, and about 17 years of schooling. Inclusion criteria included diagnosis of idiopathic PD, use of dopaminergic medication, at least one episode of FOG per day, ambulation without any orthosis or assistive device at home, internet access, and agreement to participate. Individuals with neurological, cardiovascular, or respiratory disorders, uncorrected visual or auditory impairments, severe cognitive impairments affecting verbal instruction comprehension, or inability to perform motor imagery during the Kinesthetic and Visual Imagery Questionnaire-20 (KVIQ-20) application were excluded. Assessments and interventions were remotely conducted via video conferencing, with each participant undergoing 10 training sessions lasting 45 minutes. All participants were assessed before (BT), after (AF), and 60 days post-training as a follow-up (FU). FOG frequency, intensity, and impact on functionality were assessed using the New Freezing of Gait Questionnaire (NFOG), Rapid Turns Test, and Percentage of Time with FOG (%FOG).The results were analyzed by ANOVA for repeated measures considering group and evaluation as factors. **Results:** Nineteen participants completed training and reassessment. The remote study protocol was feasible. Both groups exhibited reduced NFOG scores (p<0.0004), fewer FOG occurrences during the Rapid Turns Test, and a decrease in %FOG (p<0.036). **Conclusion:** The study found both interventions positively impacted FOG episodes, motor aspects, functionality, and quality of life, with no significant difference between groups. This suggests remote MP delivery could be beneficial in reducing FOG's impact on PD patients' daily lives.

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Effects of neuromodulation in patients with multiple sclerosis: a systematic review

Cicera Simplicio Machado Leandro, Maria Lucia Vellutini Pimentel[1], Letícia Fezer de Souza Mansur[1]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Santa Casa da Misericordia.

Correspondence: Cicera Simplicio Machado Leandro; Email: ciceramachado.medicina@hotmail.com.

Background: Multiple sclerosis (MS) is an immune-mediated and demyelinating disease that affects the young population and is more prevalent in women. Some studies show that due to neuroplasticity, neural reorganization can occur in the case of nerve injuries, such as multiple sclerosis. For this reason, therapeutic interventions that stimulate the neuroplastic process are increasingly indicated. It is known that transcranial neurostimulation is one of these interventions consisting of safe, well-tolerated and low-cost techniques, capable of stimulating the affected cortical region and promoting motor and functional benefits that can induce gains in rehabilitation with the advantages of being non-invasive, excellent tolerability and can be associated with other treatments. **Objective:** To analyze the evidence regarding the use of

neurostimulation in patients with sclerosis and its benefits in minimizing damage resulting from outbreaks, as well as in rehabilitation therapy. **Methods:** A literature review was carried out in three stages: elaboration of the research question, search for scientific articles in the Pubmed database, Google scholar and critical analysis of the included articles. The search was carried out between October 12, 2023 and December 5, 2023. 70 articles were identified, 11 of which were included in the qualitative synthesis. **Results:** Neuromodulation combined with physical therapies and conventional treatments is capable of promoting improvements in the quality of life and functionality of patients with MS, especially when started in the early stages of the disease. **Conclusion:** More studies are needed to better understand the relationship between neuromodulation and the genesis of MS. Furthermore, the limited number of articles found in the literature demonstrates the need for more studies to be carried out, with larger samples and specific protocols.

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Effects of non-invasive neuromodulation on food cravings and executive functions in health professionals

Marta Maria da Silva Lira Batista[1], Marcela Lima Silagi Siqueira[2], Maria Do Carmo de Carvalho e Martins[3], Lara Liane de Queiroz Pereira

[1]Universidade Federal do Piauí, Hospital Universitário, Teresina PI, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[3]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Marta Maria da Silva Lira Batista; Email: fonointensivista@gmail.com.

Background: Food cravings are common in individuals with and without eating disorders, especially for palatable and highly processed foods. Several factors, including blood sugar levels, hormonal variations and emotional state, influence eating decisions, mediated by neural networks associated with self-regulation, such as the dorsolateral prefrontal cortex (DLPFC). Changes in self-regulation can inhibit unwanted memories, facilitating eating goals. Studies in animal models show that deficiencies in memory inhibition increase food impulsivity and weight gain. Cortical direct current stimulation modulates neural activity and can reduce food cravings and inflammatory markers. This pioneering study also investigates chewing behavior as an analysis variable. **Objective:** To evaluate the effects of neuromodulation on food cravings, chewing patterns, brain executive functions and markers of inflammation/antioxidant activity before and after stimulation. **Methods:** A randomized, double-blind, parallel clinical trial was conducted with healthcare professionals from a University Hospital. Participants were excluded based on specific criteria and randomized to receive active or sham tDCS for 10 sessions. Assessments included food craving questionnaires, cognitive tests, biochemical and functional chewing analysis. tDCS was applied to the dorsolateral prefrontal cortex, with the aims of modulating neural activity and reducing food cravings. The study followed ethical guidelines and was registered on ClinicalTrials.com (NCT05046145), in line with CONSORT standards. **Results:** In this randomized, double-blind clinical trial, 36 participants were evaluated after receiving active or sham tDCS. They were predominantly women (63.2% in the tDCS group; 64.7% in the sham group), with an average age of 31 to 40 years, high educational level and overweight (52.6% in the tDCS group; 35.3% in the sham group). The intervention promoted a significant reduction in food cravings, especially for fatty foods and sweets, and improved executive functions such as working memory and cognitive inhibition. There was no impact on the biochemical parameters of inflammation and antioxidant activity. The tDCS may be a promising tool to modulate eating behaviors and executive functions in populations with specific characteristics, although a complete understanding of the biological, cognitive and sociodemographic interactions remains challenging. **Conclusion:** tDCS reduces food cravings and reaction time/execution of executive functions, without affecting brain biochemistry. More interdisciplinary studies are needed to understand impacts on the stomatoglossognathic system.

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Effects of trans-spinal direct current stimulation combined with motor training in traumatic spinal cord injuries: a case study

Caroline Rodrigues Osawa[1], Pedro Henrique Sousa de Andrade[1], Felipe Tumietto Domingues[1], Gustavo José Luvizutto[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Caroline Rodrigues Osawa; Email: fisiocarolineosawa@gmail.com.

Case presentation: A.L.B a 50-year-old married male from Uberaba, MG. In January 2020, he suffered a stab wound in the dorsal region, injuring the spinal cord at the T3 vertebra level (neurological level T1), resulting in a condition of sensory-motor paraplegia. The volunteer underwent a motor training (MT) protocol combined with cathodal trans-spinal direct current stimulation (tsDCS) at an intensity of 2mA, three times a week for 10 weeks (December 2023 to February 2024). The cathode was positioned at the lowest motor level, as determined by the ASIA International Standards for Neurological Classification of Spinal Cord Injury (ISNCSCI), and the anode was placed in the posterior deltoid region. The sessions lasted a total of 45 minutes, with the initial 20 minutes consisting of MT + tsDCS and the remaining 25 minutes of MT alone. **Discussion:** The evaluation considered the domains of the International Classification of Functioning, corresponding to electromyography (EMG) analysis for function, the Trunk Impairment Scale (TIS) for activity, and the Spinal Cord Independence Measure (SCIM) for participation. In the EMG, the volunteer showed no difference in peak contraction of the muscles evaluated (biceps brachii, triceps brachii, radial flexor and extensor of the wrist, rectus femoris, anterior tibialis, and lateral gastrocnemius). However, there was a 19% increase in TIS and a 5% increase in SCIM. **Final comments:** It can be inferred that functional changes are not observed due to the time since injury (4 years); however, even in this chronic phase, we can note benefits in the domains of activity and participation, highlighting the potential for improvement with trans-spinal stimulation in functionality and, consequently, the quality of life for these individuals.

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Efficacy and complications of lumboperitoneal shunt for idiopathic normal pressure hydrocephalus in elderly patients: a systematic-review and meta-analysis

Artur Menegaz de Almeida[1], Gabriel Marinheiro dos Santos Bezerra[2], Carlos Alberto Campello Jorge[3], Ítalo Barros Andrade[4], Lucca Lopes[5], Victória Morbach Siebel[6], Giovanna de Freitas Morais[7], Allan Polverini[8], Michele Kreuz[9]

[1]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[2]Universidade Federal do Ceará, Sobral CE, Brazil.
[3]Universidade Federal de Mato Grosso, Cuiabá MT, Brazil.
[4]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[5]Centro Universitário Lusíada, Santos SP, Brazil.
[6]Universidade Feevale, Novo Hamburgo RS, Brazil.
[7]Universidade Federal do Paraná, Curitiba PR, Brazil.
[8]HABP, Barretos SP, Brazil.
[9]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Giovanna de Freitas Morais; Email: giovanna.morais@ufpr.br.

Background: INPH, the most common form of hydrocephalus in adults, is a syndrome that can potentially be reversed by cerebrospinal fluid shunt. It is marked by a set of complex symptoms, with unexplained symmetric gait disturbance being the primary symptom. LPS is one of the recommended shunt types for iNPH. However, the benefits of LPS are conflicting in the literature. **Objective:** We sought to perform a Systematic Review and Meta-analysis to assess the functional outcomes of Lumboperitoneal shunt (LPS) in patients ≥60 years of age with Idiopathic Normal Pressure Hydrocephalus (iNPH). **Methods:** MEDLINE, Embase, and Cochrane were searched for RCTs and prospective studies that directly compared patients before and after LPS surgery. Studies were analyzed based on the following endpoints: Mini-Mental State Exam (MMSE), Evan's Index, subdural hematoma events, infection, and shunt malfunction. Data was examined using the Mantel-Haenszel method and 95% confidence intervals (CIs). Heterogeneity was assessed using I2 statistics. The Baujat plot was used to estimate the individual heterogeneity of each included study. **Results:** Seven studies and 809 patients were included. Evan's Index was significantly better in the post-shunt group (MD 0.0272; 95% CI 0.0012-0.0533; p=0.04; I2=75%). MMSE (MD −0.5656; 95% CI −1.3166-0.1854; p=0.1; I2=0%), subdural hematoma events (Proportion 0.05; 95% CI 0.03-0.07; p=0.39; I2=1%), shunt infection (Proportion 0.01; 95% CI 0.00-0.03; p=0.41; I2=0%), and shunt malfunction (Proportion 0.03; 95% CI 0.02-0.05; p=0.85; I2=0%) endpoints did not show any statistically significant difference between groups. **Conclusion:** This meta-analysis suggests that LPS may be an efficient intervention for iNPH in patients ≥60 years old, given the decrease in the Evan Index, a scale that measures the degree of ventricular distention.

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Efficacy and safety of argatroban in treatment of acute ischemic stroke in patients not receiving endovascular treatment: a systematic review and meta-analysis

Pedro Henrique Gorri Custódio de Melo Brites[1], João Marcelo Lorga Carnielli[1], Danilo Augusto Fernandes Moura[1], Guilherme Augusto Guimarães Resende[1], Julia Favaro Brenny[2], Maria Clara de Mattos Mira[1], Fernanda Peres Tomazoli[1], Camilla Stringuetta Gallo[3], João Felipe Hermann Costa Scheidt[3]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.
[2]Universidade Estadual de Londrina, Londrina PR, Brazil.
[3]Hospital Evangélico de Londrina, Londrina PR, Brazil.

Correspondence: Pedro Henrique Gorri Custódio de Melo Brites; Email: pedro@brites.srv.br.

Background: Globally, stroke is a major cause of death and disability. Argatroban, a direct thrombin inhibitor, is promising for the treatment and prevention of acute ischemic stroke. Known for its anticoagulant properties, especially in patients with heparin-induced thrombocytopenia, its role in managing acute ischemic stroke is of great interest. **Objective:** This meta-analysis assesses the effectiveness and safety of argatroban in the treatment of acute ischemic stroke. **Methods:** We conducted a comprehensive search of PubMed, Embase, and Cochrane Library for studies on argatroban in acute ischemic stroke treatment, covering literature up to April 12, 2024. Only randomized controlled trials and observational studies

were considered. Reviews, case reports, preclinical studies, and single-arm studies were excluded to focus on robust clinical data, as well articles that the control group had mechanical thrombolysis. Meta-analyses were performed using a random-effects model, pooling data as odds ratios (OR) with 95% confidence intervals (CI), considering p-values <0.05 as statistically significant. High heterogeneity was noted for I2 values over 50%. Safety outcomes included rates of intracranial hemorrhage (ICH), major bleeding (mB), and mortality. The main efficacy outcome measured was the modified Rankin Scale (mRS) score at 3 months. Another outcome investigated was early neurological deterioration (END), defined as a decrease in NIIHS score in a short period of time. **Results:** The systematic review included 17 studies, among them 7 randomized clinical trials. The meta-analysis revealed that argatroban does not significantly increase the risk of ICH (OR 1.10, 95% CI 0.76-1.59, p=0.60, I2=0%), major bleeding (OR 1.39, 95% CI 0.93-2.03, p=0.11, I2=0%), or mortality (OR 1.11, 95% CI 0.61-2.00, p=0.74, I2=3%). Furthermore, the argatroban-treated patients showed a significant improvement in achieving mRS scores of 0-1 (OR 1.58, 95% CI 1.08-2.33, p=0.02, I2=70%), 0-2 (OR 1.21, CI 1.04-1.42, p=0.02, I2=0%). In addition, the argatroban group has shown significant reduction in the rates of early neurological deterioration (OR 0.43, CI 0.29-0.64, p<0.0001, I2=23%). **Conclusion:** Argatroban treatment in acute ischemic stroke demonstrates safety comparable to standard treatments, with low rates of serious adverse effects like intracranial or major bleeding. It also significantly enhances functional outcomes as measured by mRS scores and decreased rates of END, indicating better prognosis. Further subgroup analyses may help reduce heterogeneity in mRS score outcomes, supporting the efficacy and safety of argatroban in this context.

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Efficacy and safety of atogepant for migraine prevention: a systematic review and meta-analysis of randomized controlled trials

Kaike Eduardo da Silva Lobo[1], Marcus Paulo Oliveira Lopes[1], João Lucas Lalor Tavares[1], Arthur Moraes dos Santos[1], Felipe Moraes Costa[1], Flavio Lobo Altieri[1], Arley Nikson Oliveira Barreto[1], Ruan Pablo da Silva Gomes[1], Leonardo Bastos Santos[1], Leonardo Aguiar dos Santos[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Marcus Paulo Oliveira Lopes; Email: marcus.polopes@aluno.uepa.br.

Background: Migraine is one of the most common neurological disorders, with an estimated prevalence of 15% of the population, affecting more than one billion people worldwide. Atogepant is an oral, small-molecule, calcitonin gene-related peptide receptor antagonist that was approved in 2021 by the US Food and Drug Administration for the preventive treatment of episodic migraine. However, its efficacy and safety are still uncertain. **Objective:** We performed a systematic review and meta-analysis to evaluate the efficacy and safety of atogepant for the preventive treatment of migraine. **Methods:** We searched MEDLINE, EMBASE, Cochrane Central Register of Controlled Trials and Web of Science from inception to June 2024 using "Atogepant", "MK-8031", "AGN-241689" and "migraine" search terms. Eligible articles were randomized clinical trials that showed efficacy and safety outcomes, such as changes from baseline in monthly migraine days and the incidence of adverse events (AEs), in patients who underwent migraine prevention with atogepant or placebo. Two independent reviewers extracted the data. Mean difference (MD) and risk ratio (RR) with 95% confidence intervals (95% CIs) were estimated. **Results:** Four studies were eligible for inclusion. The statistical analysis focused on the once-daily 60 mg dose of atogepant, as this dosage was the only one evaluated by at least three studies. In the patients who received atogepant, there was a significant decrease in monthly migraine days (MD -1.62; 95% CI -2.28, -0.96; p=0.00001; I'=100%), monthly headache days (MD -1.70; 95% CI -2.14, -1.26; p=0.00001; I'=100%) and monthly acute medication use days (MD -1.82; 95% CI -2.36, -1.29; p=0.00001; I'=100%) compared to the placebo group. The percentage of patients who achieved at least a 50% reduction from baseline in the 3-month average of monthly migraine days was significantly higher in the atogepant group (50.6% vs 28.4%; RR 1.82; 95% CI 1.34, 2.48; p=0.0001; I'=82%). In terms of safety endpoints, there was not a statistically significant difference between groups in treatment-emergent AEs (57.1% vs 52.1%; RR 1.09; 95% CI 0.93, 1.26; p=0.28; I'=65%), treatment-emergent serious AEs (1.55% vs 0.85%; RR 1.60; 95% CI 0.51, 5.08; p= 0.42; I'= 19%) and treatment discontinuation due to AEs (2.87% vs 2.8%; RR 1.02; 95% CI 0.58, 1.80; p=0.94; I'=0%). However, the treatment-related AEs (19.5% vs 11.9%; RR 1.64; 95% CI 1.25, 2.15; p=0.0004; I'=26%) were significantly increased in the atogepant group. **Conclusion:** Atogepant 60mg once a day was safe and showed significant reduction in mean monthly migraine days compared with placebo. However, further research is needed to evaluate different dosages and fully understand its safety profile.

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Efficacy and safety of botulinum toxin type a (bont-a) use in the treatment of chronic migraine: a systematic review and meta-analysis

Luana Izabela Azevedo de Carvalho[1], Ana Luiza Azevedo de Carvalho[2], Pedro Lucas Azevedo de Carvalho[2], Manuela Ribeiro de Queiroz[2], Kássia Violeta Rodrigues de Matos[3], ítalo Barros Andrade[4], Artur Menegaz de Almeida[5], Michele Kreuz[6], Lilianne Rodrigues Fernandes[7], Vitor Maia Arca[8]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Faculdade Metropolitana, Manaus AM, Brazil.
[4]Faculdade Santo Agostinho de Vitória da Conquista, Salvador BA, Brazil.
[5]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[6]Universidade do Brazil, Canoas RS, Brazil.
[7]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.
[8]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.

Correspondence: Pedro Lucas Azevedo de Carvalho; Email: pedrolucaslac@gmail.com.

Background: Chronic Migraine (CM) is a disabling neurological disorder classified as a complication of migraine. Patients with CM experience headaches on 15 or more days per month in at least three consecutive months. Despites the range of therapeutic treatments, in some cases current treatments often have limited efficacy and intolerable adverse events, highlighting gaps in prophylactic treatment options. OnabotulinumtoxinA (BoNT-A), a purified protein that blocks acetylcholine release from presynaptic neurons and inhibits muscle contractions, has shown pain relief in various conditions, including migraine. Clinical trials suggest that BoNT-A may be an effective and safe prophylactic medication for treating migraines and other headache disorders. **Objective:** To evaluate the efficacy and safety of BT-A in the treatment of chronic migraine through a systematic review and meta-analysis. **Methods:** The literature search was performed using PubMed, Scopus, and Web of Science databases. The inclusion criteria consisted of randomized controlled trials that evaluated the use of BoNT-A in patients diagnosed with chronic migraine, comparing it to a placebo group. Studies were screened and selected based on relevance, and data were extracted by independent reviewers. The primary outcomes were the reduction in the number of headache days and the incidence of adverse events. All statistical analyses were performed using the RStudio (version 4.3.2). **Results:** A total of 11 studies and 3.617 patients were included, with 2.092 (57.84%) receiving botulinum toxin type A (BoNT-A) and 1.525 (42.16%) in the placebo group. BT-A therapy was associated with a significant reduction in the number of headache days per month in the BoNT-A group (odds ratio: 0.89; 95% CI: 0.54 to 1.49; P<0.01; I^2=72%), suggesting that BoNT-A is effective in reducing the frequency of chronic migraine episodes. Adverse events were reported more frequently in the BT-A group compared to the placebo group (odds ratio: 0.96; 95% CI: 0.37 to 2.48; P<0.01; I^2=97%). Common adverse effects included neck pain, muscle weakness, and injection site pain. The high heterogeneity indicates variability in the reported adverse effects among different studies. **Conclusion:** The results of our study suggest that the therapy with BT-A is demonstrated a significant reduction in headache days per month compared to the placebo group. However, BT-A treatment was also associated with a higher incidence of adverse effects. Overall, BT-A appears to be a promising prophylactic treatment for chronic migraine, balancing its beneficial reduction in headache frequency against the consideration of its adverse effects.

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Efficacy and safety of galcanezumab for cluster headache preventive treatment: a systematic review and meta-analysis

Marina Barbosa da Silva[1], Domenica Baroni Coelho de Oliveira Ferreira[2], Cristian David Mendieta Barrera[3], Lívia Aguiar Nogueira Rosa[4], Nathalia Moraes Figueiredo[5]

[1]Centro Universitário Barão de Mauá, Ribeirão Preto SP, Brazil.
[2]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Universidad Mayor Real y Pontificia de San Francisco Xavier de Chuquisaca, Sucre, Bolivia.
[4]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[5]University of Rochester, New Yourk, USA.

Correspondence: Marina Barbosa da Silva; Email: marinabarbosa16@hotmail.com.

Background: Galcanezumab is a monoclonal antibody targeting the calcitonin gene-related peptide (CGRP) pathway and represents the latest disease-specific and mechanism-based therapeutic option for cluster headache (CH). **Objective:** We performed a systematic review and meta-analysis to evaluate the efficacy and safety of galcanezumab for CH. **Methods:** We searched PubMed, Embase, and Cochrane Library for studies implementing galcanezumab for episodic and chronic CH, with no language restrictions. We followed the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guideline. The primary outcome was efficacy, defined by a reduction from the baseline of at least 50% in the weekly frequency of cluster headache attacks and the Patient Global Impression of Improvement scale (PGI-I). Secondary outcomes included treatment-emergent adverse events (TEAEs) and serious adverse events (SAEs). **Results:** A total of 504 patients were included from 6 studies, of which 2 were RCTs. The follow-up period ranged from 3 weeks to 15 months. The mean age was 44.4±10.2 years, with 24.4% female patients. Overall efficacy was 76.0% (95% CI 0.67 - 0.85), TEAEs were observed in 48.0% of patients (95% CI 0.25 - 0.72), and the most common were nasopharyngitis, local injection pain, and local injection swelling. TEAEs were however considerably higher within the 300 mg dose group compared with the 240 mg dose group, 80.0% (95% CI 0.65 - 0.87) versus 28.0% (95% CI 0.12 - 0.47), respectively. **Conclusion:** This meta-analysis suggests that galcanezumab is effective in reducing the number of CH attacks and can be considered a safe medication.

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Efficacy and safety of hematopoietic stem cells transplantation in Krabbe disease: a systematic review

Leonardo José Rodrigues de Araújo Melo[1], Francisco Luciano Honório Barreto Cavalcante[1], Lucas Soares Radtke[1], Letícia da Costa Rios[1], Paulo Wendel Moura de Souza[1], Pedro Vitor Ferreira Rodrigues[1], Leonardo Elias Araujo dos Santos[1], Pedro Lucas Grangeiro de Sá Barreto Lima[1], Anderson Rodrigues Brandão de Paiva[2], Paulo Ribeiro Nóbrega[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Mendelics Análise Genômica, São Paulo SP, Brazil.

Correspondence: Leonardo José Rodrigues de Araújo Melo; Email: lleonardomello98@alu.ufc.br.

Background: Krabbe disease (KD) is a rare autosomal recessive leukodystrophy caused by deficiency of β-galactocerebrosidase (GALC) that leads to accumulation of toxic metabolites, causing progressive loss of myelin in the central nervous system and peripheral nerves, resulting in severe motor and cognitive impairment. Hematopoietic Stem Cells transplantation (HSCT) is one of the most promising therapies for infantile-onset KD (IKD). **Objective:** To investigate the efficacy and safety of HSCT for neurological improvement in IKD. **Methods:** A systematic literature search was conducted on PubMed and Web of Science platforms, following PRISMA protocol, using DeCS/MeSH descriptors "Krabbe disease" or "globoid cell leukodystrophy" and "Transplant" and "Hematopoietic Stem Cells". The inclusion criteria were cohorts or clinical trials published in English in which patients with IKD were submitted to HSCT therapy. Reviews, case reports or animal studies were excluded.The efficacy was measured by survival rates (SR) and neurological improvement after HSTC. Articles were analyzed by two blind observers using the Rayyan AI platform, with divergences analyzed by a third independent individual. Initially, 220 papers on the subject were obtained. **Results:** Twelve articles met the inclusion criteria, including 10 cohort studies and 2 clinical trials, totaling 181 patients, who were divided into two groups: symptomatic (SS) and asymptomatic (AS) patients at the moment of HSCT, with average follow-up time ranging from 6 months to 12 years. The SR at the end of follow-up varied from 69 % to 100% for AS and 42% to 100% for SS, higher rates than previously reported historical controls. Neurological improvement was assessed clinically and using scales; such as BAYLEY III, which indicated a greater proportion of neurodevelopmental milestones than historical controls in AS group compared to the SS group, with gross motor components showing little change after HSCT in both groups.It was reported that patients with early HSTC, especially before 1 month of age, had better neurological outcomes. GALC levels were restored after HSCT in most patients, but were not associated with clinical improvement in SS patients. The most common adverse events were graft-versus-host disease and urinary and pulmonary infections **Conclusion:** HSTC for IKD proves to be an effective therapy, with acceptable adverse effects, possibly better when performed earlier in patients with evidence of the disease but without symptoms. This may lead to longer survival times, greater chances of achieving neurodevelopmental milestones, although evidence is less clear in terms of improvement of gross motor skills. Study limitations include significant heterogeneity in the forms of neurological improvement assessment across the articles. Further studies with more standardized methodologies are needed to more accurately evaluate HSTC for IKD.

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Efficacy and safety of inotersen and eplontersen in hereditary transthyretin amyloidosis: a systematic review and meta-analysis

Fernanda Moraes Tamashiro[1], Artur Menegaz de Almeida[2], Thaís Luiza Oliveira de Holanda[3], Iasmin Isabelli Luiz Silvério[1], Rebeca Lima de Almeida Santos[4], ítalo Barros Andrade[5], Lilianne Rodrigues Fernandes[6], Michele Kreuz[7]

[1]Universidad de Buenos Aires, Buenos Aires, Argentina.
[2]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[3]Universidade de Pernambuco,Recife PE, Brazil.
[4]Universidade Estadual do Sudoeste da Bahia,Vitória da Conquista BA, Brazil.
[5]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[6]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.
[7]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Fernanda Moraes Tamashiro; Email: fermtamashiro@gmail.com.

Background: Hereditary transthyretin amyloidosis is a rare and progressive peripheral sensorimotor neuropathy. Inotersen, an antisense oligonucleotide inhibitor and Eplontersen, a transthyretin antisense oligonucleotide, are both approved by the Food and Drug Administration for the treatment of this condition. However, the efficacy of both has not yet been tested in a meta-analysis **Objective:** We sought to assess the safety and efficacy of Inotersen and Eplontersen among adult patients. **Methods:** We searched Pubmed, Scopus, and Web of Science databases for randomized controlled trials that investigated Inotersen and Eplontersen in comparison with a placebo group. A random-effects model was employed to compute the mean difference (MD) with a 95% confidence interval (CI). Statistical analysis was performed using R software 4.3.1. **Results:** In the two included studies, involving 308 patients, the follow-up period completed 66 weeks and the adjusted mean for the modified Neuropathy Impairment Score+7 composite score reached a statistically significant difference: (MRAW 2.58 [95% CI, −3.27 to 8.44]; P<0.01), trending towards the Eplonserten treatment. Moreover, Norfolk Quality of Life–Diabetic Neuropathy (MRAW −3.25 [95% CI, −11.28 to 4.77]; P<0.01) and Short Form 36 score (MRAW 0.73 [95% CI, 0.10 to 1.36]; P=0.26) also presented significantly better results for Eplonserten. **Conclusion:** In this meta-analysis of two studies, consistent results suggest that Inotersen and Eplontersen both associated with beneficial outcomes and a improved quality of life in patients with hereditary transthyretin amyloidosis

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Efficacy and safety of mechanical thrombectomy in posterior circulation ischemic strokes: a systematic review

Rayanne Pereira Mendes[1], Atilio Gomes Romani[1], Saulo Barros Teixeira[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Rayanne Pereira Mendes; Email: rayannepmendes@gmail.com.

Background: Posterior circulation ischemic strokes, caused by the occlusion of vertebral and basilar arteries, comprise 20% of all ischemic strokes and have higher morbidity and mortality than anterior circulation strokes. Mechanical thrombectomy (MT) is effective for large vessel occlusions, supported by guidelines based on randomized clinical trials. However, its benefits in posterior circulation were often not covered in most of these studies. **Objective:** To evaluate the efficacy and safety of mechanical thrombectomy in posterior circulation ischemic strokes. **Methods:** A systematic review synthesized results from 2021-2022 clinical trials on MT efficacy and safety in posterior circulation. Data was extracted from PubMed, with inclusion criteria of high-level evidence and recent randomized clinical trials, excluding studies that did not meet these standards. **Results:** The studies evaluate the efficacy of MT compared to the Best Medical Treatment (BMT), which consists of anticoagulants, antithrombotics, or thrombolysis in eligible cases. Primary outcomes analyzed included rates of successful recanalization, functional independence (modified Rankin scale score of 0-2 at 90 days), and safety outcomes, including symptomatic intracranial hemorrhage and mortality. In a trial conducted in 23 countries with 300 patients, 44.2% of patients undergoing MT within 6 hours of basilar artery ischemic stroke onset had a favorable functional outcome versus 37.7% in the BMT group at 90-day follow-up (CI: 0.92 to 1.50). Mortality was 38.3% in the endovascular group and 43.2% in the medical care group (P=0.29). Symptomatic intracranial hemorrhage was 4.5% in the MT group versus 0.7% in the BMT group. In another trial that included 217 randomized patients, 46% of patients undergoing MT within 6 to 24 hours of symptom onset achieved functional independence at 90 days versus 24% in the BMT group (P<0.00). Symptomatic intracranial hemorrhage was 6% in the MT group versus 1% in the BMT group, with 90-day mortality rates of 31% versus 42%, respectively. In a prospective multicenter study conducted with 2134 patients from 2017 to 2021, 82% of patients in the endovascular group achieved successful recanalization. There was a higher rate of functional independence at 90 days in the MT group (44% versus 25% in the BMT group) and a lower mortality rate in the MT group (36.9% versus 47.5%) with an absolute risk difference of −10.3%. However, there was a higher rate of symptomatic intracranial hemorrhage in the MT group (5.2% versus 0.9%) with an absolute risk difference of 4.5%. **Conclusion:** The studies show favorable evidence for using MT in posterior circulation ischemic strokes, demonstrating high recanalization rates, better 90-day functional outcomes, and reduced mortality compared to BMT. The safety profile is acceptable, with slightly higher rates of symptomatic intracranial hemorrhage but reduced mortality.

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Efficacy and safety of melatonin for migraine prevention in children and adolescents: a systematic review and meta-analysis

Isabella Christina Amaral de Lara[1], Mariana Rachas Reis[2], Matheus Ricardo Malveira Camacho[3], Amanda Gabriela Freitas Rodrigues[4], Ademar César Ignácio de Moraes[5]

[1]Universidade Cesumar, Maringá PR, Brazil.
[2]Santa Casa de Misericórdia de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[3]Universidade do Estado do Pará, Belém PA, Brazil.
[4]Universidade Federal do Pará, Belém PA, Brazil.
[5]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Isabella Christina Amaral de Lara; Email: isabellaadelara@gmail.com.

Background: Migraine is known to be the most prevalent primary headache in pediatric and adolescent patients, affecting around 28% of teenagers. Besides this, only a limited number of prophylactic treatments have proven effective for these age groups. Melatonin, a hormone primarily produced by the pineal gland, plays an important role in the modulation of the circadian rhythm and has shown effectiveness in preventing migraines in adults in previous studies. However, its efficacy in younger patients remains uncertain. **Objective:** To assess the safety and efficacy of melatonin for migraine prophylaxis among children and adolescents. **Methods:** Pubmed, Scopus, Web of Science, and Cochrane databases were systematically searched for randomized controlled trials (RCTs) testing melatonin for migraine prevention in pediatric or adolescent populations. A random-effects model was used to calculate the

mean differences (MDs) with 95% confidence intervals (CIs). Heterogeneity was determined using I^2 statistics. Efficacy was assessed based on headache frequency, while safety was evaluated by adverse events (AEs), including daily sleepiness, nausea, vomiting, worsened headache, worsened movement sensitivity, elevated heart rate, flu-like symptoms, constipation, fatigue, malaise, and fever. Statistical analysis was conducted using R, version 4.4.0. **Results:** We included 4 RCTs with 238 patients. The mean age ranged from 9.28 to 15.00 years, with a majority of female patients (55.9%). Compared with the control group, melatonin use was associated with a significant reduction in monthly headache days (MD 2.93 days; 95% CI 0.72-5.14; p=0.009310; I^2=65%). Additionally, the meta-analysis showed that the incidence of adverse events in the intervention group was comparable with control, hence, not associated with melatonin use (17.3% vs 24.24%; MD 0.69; 95% CI 0.22-2.12; p=0.518081; I^2=37%). **Conclusion:** This systematic review and meta-analysis suggest that the use of melatonin in younger patients with migraine leads to a reduction in monthly headache frequency. While AEs were reported, their incidence was comparable to the control group, indicating a favorable safety profile.

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Efficacy and safety of mycophenolate in AQP4-AB negative neuromyelitis optica spectrum syndrome associated with encephalopathy: report of a case

Marcella Turon Baran[1], Thais Lavinas Moura Baptista Barroso[1], Marina Papais Alvarenga[2], Marcos Papais Alvarenga[2], Regina Maria Papais Alvarenga[2]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.
[2]Hospital Federal da Lagoa, Rio de Janeiro RJ, Brazil.

Correspondence: Regina Maria Papais Alvarenga; Email: reginaunirio2016@gmail.com.

Case presentation: A 30-year-old white woman presented with a headache and left unilateral amaurosis and a normal cranial MRI (February 2007). One month later, she developed transverse myelitis syndrome with an extensive longitudinal spinal cord lesion identified on cervical MRI. Treated with methylprednisolone (1g/day /5days) led to gait improvement but not visual recovery. In August 2007, she developed right hemiplegia and aphasia. Brain MRI revealed an extensive left hemispheric lesion. Hospitalized, she received methylprednisolone and intravenous human immunoglobulin (cycles/IV) with partial symptom improvement. Three months later, still hospitalized, she experienced bilateral vision loss, worsening motor dysfunction, and new left cerebral lesion expansion with additional right occipital lesion. She underwent plasmapheresis followed by monthly pulse therapy with methylprednisolone and human immunoglobulin, leading to partial recovery of visual and cognitive manifestations. In 2008, she started off-label preventive treatment with mycophenolate (1g/day). Serial MRIs showed no new brain or spinal lesions. Testing for NMO-IgG antibodies in 2009 by indirect immunofluorescence and in 2017 by Cell Based Assay (CBA), considered the gold standard, were normal. At the last visit, she walked with support, had moderate motor dysfunction in her lower limbs and severe in the right upper limb, communicated verbally, and maintained left amaurosis. Mycophenolate treatment demonstrated complete efficacy and safety over a 10-year follow-up period. **Discussion:** Diagnostic criteria for Neuromyelitis Optica Spectrum Disorders (NMOSD) have evolved, with recognition of neurological syndromes beyond optic nerve and spinal cord involvement since 2006. This new criterion identified 6% of cases with extensive brain lesions in the NMO Mayo Clinic series. In 2015, NMOSD classification included AQP4-AB serological status. This patient, negative for AQP4-IgG by CBA, meets NMOSD AQP4-negative criteria and, despite experiencing four acute and severe events in the first year of disease, including encephalopathy with extensive brain lesions, achieved complete efficacy with off-label oral mycophenolate, endorsed in 2021 by a Latin American consensus of specialists as a first-line drug for NMO treatment. **Final comments:** Cases of NMO with extensive brain lesions are rare; we identified only six among 128 patients in our cohort. Highlighting that FDA-approved therapies are indicated only for AQP4-IgG-positive patients, we emphasize the indication of mycophenolate, which was a safe and effective treatment over ten years of follow-up.

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Efficacy and safety of rituximab in a Brazilian cohort of patients with refractory myasthenia gravis

Amanda Vitória Freire de Oliveira[1], Ana Flávia Andrade Lemos[1], Lucas Gondim Briand Vieira[1], Mariana Ribeiro Pereira[1], Mateus Gustavo Favaro[1], Rodrigo Siqueira Soares Frezatti[1], André Cleriston José dos Santos[1], Claudia Ferreira da Rosa Sobreira[1], Wilson Marques Junior[1], Pedro José Tomaselli[1]

[1]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Amanda Vitória Freire de Oliveira; Email: amandinha_ba@yahoo.com.br.

Background: Myasthenia gravis is an autoimmune disease characterized by muscle weakness and fatigability. The pathophysiological mechanism is mediated by B cells, with auto antibodies directed against proteins specific located in the postsynaptic membrane of the neuromuscular junction. **Objective:** To analyse the safety and efficacy of anti CD20 monoclonal antibody (Rituximab) in the treatment of patients with diagnosis of Myasthenia Gravis who are refractory to first-line medications, followed up at the referral centre of neuromuscular disorders at the Clinical Hospital of Ribeirão Preto, University of São Paulo. **Methods:** Patients with a definitive diagnosis of Myasthenia Gravis, defined by the presence of typical clinical features, along with at least one of the following criteria: repetitive nerve stimulation electromyography showing a decrement in Compound Muscle Action Potential, amplitude between the first and fourth responses greater than 10% in at least two muscle groups; presence of anti-AChR or anti-MuSK antibodies; or single-fiber EMG demonstrating altered jitter or the presence of blocking. Patients with a defined failure or absolute contraindication to the use of first-line medications, who received Rituximab as a rescue therapy and were followed up were recruited. The number of exacerbations, MGFA scale, and MG-ADL scale were observed before and after Rituximab administration. The MGFA and MG-ADL scales were assessed 4 weeks after induction. **Results:** A total of thirty-three patients were recruited. Of these, 5 patients were excluded due to overlap syndromes with other diseases that could confound the clinical picture. Among the recruited patients, 82.14% were women. The age distribution at onset was as follows: 57.14% had early-onset (19-50 years), 25% had late-onset (>50 years), 10.7% had juvenile-onset (≤18 years), and 7.16% had very late-onset (>65 years). Almost all patients (96.4%) exhibited generalized Myasthenia Gravis at the time of evaluation, with only 3.6% presenting ocular MG. Regarding serological status, 82.13% of the patients were seropositive: 60.71% had anti-AChR antibodies and 21.42% had anti-MuSK antibodies. Out of the 28 evaluated patients, 60.71% experienced a reduction in clinical exacerbation following Rituximab administration. Additionally, 71.4% and 42.8% showed improvement in the MG-ADL and MGFA scales, respectively. Twenty-five patients were on corticosteroid therapy prior to Rituximab treatment. Post-Rituximab, 80% of these patients were able to reduce their corticosteroid dose, of which 36% discontinued corticosteroid use entirely. All 28 patients were also using Pyridostigmine; following Rituximab administration, 35.71% of them reduced their Pyridostigmine dose. **Conclusion:** These findings suggest that Rituximab is effective in reducing clinical exacerbations and improving functional outcomes in patients with Myasthenia Gravis, as evidenced by the significant improvements in MG-ADL and MGFA scores.

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Efficacy and safety of tacrolimus for myasthenia gravis: a systematic review and meta-analysis

Lorhayne Kerley Capuchinho Scalioni Galvao[1], Nicole dos Santos Pimenta[2], Ana Clara Felix de Farias Santos[3], Deivyd Vieira Silva Cavalcante[4], Fernanda Valeriano Zamora[5], João Pedro Costa Esteves Almuinha Salles[6], Rodrigo Tomazini Martins[7]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Universidade Cidade de São Paulo, São Paulo SP, Brazil.
[4]Universidade Federal do Maranhão, São Luís MA, Brazil.
[5]Universidade Estadual do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[6]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[7]Mater Hospital, Neuroscience Department, Brisbane, QLD, Australia.

Correspondence: Ana Clara Felix de Farias Santos; Email: anafelix625@gmail.com.

Background: Myasthenia gravis (MG) is an autoimmune neuromuscular disorder characterised by muscle weakness and fatigability due to autoantibodies targeting the neuromuscular junction. While conventional treatments like acetylcholinesterase inhibitors and corticosteroids are effective for many, some patients face refractory disease or intolerable side effects, necessitating alternative therapies. Tacrolimus, a potent calcineurin inhibitor used in organ transplantation, has emerged as a promising option for such challenging cases. Tacrolimus modulates the immune response underlying MG by suppressing T-lymphocyte activation and antibody production against acetylcholine receptors, suggesting potential efficacy in symptom management and reducing reliance on other immunosuppressants. **Objective:** This systematic review and meta-analysis assesses current evidence on tacrolimus for MG, exploring its therapeutic benefits, safety profile, and the need for further research on optimal dosing and long-term management. **Methods:** A comprehensive search of PubMed, Embase, and Cochrane databases up to June was conducted to identify studies comparing tacrolimus with placebo or standard treatments in MG patients. We calculated odds ratios (ORs) for categorical outcomes and mean differences (MDs) for continuous measures, with 95% confidence intervals (CIs). A random-effects model was used with statistical significance set at p<0.05. Data were analysed using R software (version 4.4.0), and heterogeneity was assessed using I^2 statistics. **Results:** Eight studies involving 1083 patients were included, with 595 (54.94%) receiving tacrolimus. Tacrolimus treatment showed significant improvements in MG Activities of Daily Living (MG-ADL) scores (MD -0.91 points; 95% CI -1.50, -0.33; p<0.01; I^2=0%) and the proportion of patients achieving minimal myasthenic symptoms or better (OR 0.70; 95% CI 0.52, 0.92; p=0.011; I^2=0%). There were no significant differences between groups in prednisolone dose at endpoint (MD -3.70 mg; 95% CI -8.80, 1.40; p=0.15; I^2=92%), incidence of adverse events (OR 0.73; 95% CI 0.31, 1.69; p=0.462; I^2=60%), or Quantitative MG score (QMG score) (MD 0.20 points; 95% CI -2.31, 2.70; p=0.88; I^2=88%). **Conclusion:** Tacrolimus demonstrated significant efficacy in improving MG-ADL scores and achieving minimal myasthenic symptoms or better, with no significant differences in prednisolone dose compared to standard care. The incidence of side

effects was also similar between the groups. These findings support tacrolimus as a viable treatment option for MG, highlighting its potential to enhance patient outcomes in clinical practice.

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Efficacy and safety of tirofiban in acute ischemic stroke patients not receiving endovascular treatment: a systematic review and updated meta-analysis

João Marcelo Lorga Carnielli[1], Danilo Augusto Fernandes Moura[1], Guilherme Augusto Guimarães Resende[1], Pedro Henrique Gorri Custódio de Melo Brites[1], Maria Clara de Mattos Mira[1], Julia Favaro Brenny[2], Fernanda Peres Tomazoli[1], Camilla Stringuetta Galo[3], João Felipe Hermann Costa Scheidt[3]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.
[2]Universidade Estadual de Londrina, Londrina PR, Brazil.
[3]Hospital Evangélico de Londrina, Londrina PR, Brazil.

Correspondence: João Marcelo Lorga Carnielli; Email: joao_lorga@hotmail.com.

Background: Stroke is a leading cause of death and disability worldwide. Antiplatelet drugs are known to be effective in the treatment and prevention of acute ischemic stroke. However, the role of tirofiban, a nonpeptide selective inhibitor of the platelet glycoprotein IIb/IIIa receptor, in the treatment of acute ischemic stroke in patients not receiving endovascular treatment, remains under discussion. **Objective:** To determine the efficacy and safety of tirofiban, either as monotherapy or in combination with other standard therapies, in treating acute ischemic stroke in patients not undergoing endovascular treatment. Material and **Methods:** PubMed, Embase and Cochrane Library databases were searched for randomized controlled trials and observational studies on tirofiban therapy for acute stroke published up to April 22, 2024. Reviews, case reports, preclinical studies or single-arm studies were excluded. Studies in which patients underwent mechanical thrombectomy were also excluded. Meta-analysis were conducted using a random-effects model. The data were pooled as odds ratios (OR) with 95% confidence intervals (CI). A p-value<0.05 was considered statistically significant. Significant heterogeneity was defined as I2>50%. The safety outcomes included symptomatic intracranial hemorrhage (sICH), any intracranial hemorrhage (ICH), and mortality. The efficacy outcome was the modified Rankin Scale (mRS) score at the 3-month follow-up. **Results:** twenty-four studies, including 13 randomized clinical trials, were included in this systematic review. The meta-analysis indicates that treating ischemic stroke with tirofiban does not increase the incidence of any intracranial hemorrhage (OR 1.00, 95% CI 0.74-1.36, p=0.98, I2=0%), as well as symptomatic intracranial hemorrhage (OR 0.92, 95% CI 0.45-1.89, p=0.82, I2=0%) and mortality (OR 0.71, 95% CI 0.401.26, p=0.25, I2=47%). Additionally, the rates of mRS score of 0-2 (OR 1.89, 95% CI 1.49-2.40, p<0.00001, I2=46%) after 90 days significantly improved in the tirofiban group. The rates of mRS score of 0-1 (OR 1.87, 95% CI 1.33-2.64, p=0.0003, I2=65%) after 90 days were significantly higher in the tirofiban group; however, significant heterogeneity was observed in this specific analysis as well. **Conclusion:** treatment of acute ischemic stroke with tirofiban has shown to be safe compared to standard treatment alone, with a low incidence of adverse effects such as intracranial bleeding and mortality. Overall analysis has demonstrated a significant improvement in mRS score in the tirofiban group, indicating a better prognosis. Subgroup analysis could be conducted to decrease the levels of heterogeneity in the analysis of the rates of mRS score of 0-1. In conclusion, tirofiban is effective and safe in treating acute ischemic stroke when compared with standard treatment alone.

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Efficacy and safety of vamorolone in improvement of muscle function in Duchene muscle dystrophy: a systematic review and meta-analysis

Francisco Jazon de Araújo Neto[1], Jamine Yslailla Vasconcelos Rodrigues[1], Thomas Silva de Queiroz[1], Honohrária Bolivya Alencar de Oliveira[1], Gabriel de Almeida Monteiro[1], Espartaco Moraes Lima Ribeiro[1], Marcondes Pimentel Cruz[1], Paulo Roberto Lacerda Leal[1]

[1]Universidade Federal do Ceará, Sobral CE, Brazil.

Correspondence: Jamine Yslailla Vasconcelos Rodrigues; Email: jamineysl@gmail.com.

Background: Duchenne muscular dystrophy (DMD) is a rare and a progressive neuromuscular disorder caused by variants in the DMD gene. Treatment of corticosteroids slows the progression of weakness, but chronic glucocorticosteroid (GC) treatment causes significant morbidities. Vamorolone is a first-in-class, dissociative, synthetic steroidal anti-inflammatory drug that has been studied as an alternative for DMD treatment. A recent randomized controlled trial (RCT) provided new evidences regarding the effectiveness of vamorolone in DMD. Thus, we decided to perform an updated meta-analysis to assemble the current knowledge and reduce uncertainties regarding the use of the vamorolone in DMD treatment. **Objective:** To investigate whether vamorolone is an effective treatment for DMD. **Methods:** We systematically searched PubMed, Embase, and Cochrane Library, for randomized controlled trials (RCTs) and cohort studies comparing vamorolone versus placebo or glucocorticoids for Duchenne muscular dystrophy treatment. The outcomes of interest were Time To Stand From Supine (TTSTAND) and Time To Run/Walk 10 meters (TTRW). We pooled analysis of mean difference (MD) with 95% confidence intervals (CI) applying a random-effects model and using Review Manager 5.4.1 for all statistical analyses. **Results:** We included 2 RCTs and 3 non-randomized clinical trials comprising 391 patients, 181 in the vamorolone groups, 136 in the GC group, and 74 in the placebo group. The pooled analysis showed a statistically significant association between the vamorolone larger doses (≥2 mg/kg/d) group and an increase in the TTSTAND, compared to the placebo group (MD 0.11; 95% CI 0.10, 0.12; P<0.001), but non-significant results when compared to the GC group (MD -0.01; 95% CI -0.02, 0.00; P<0.001). Vamorolone in 2mg dosage also had a greater TTRW compared to the placebo group (MD 0.20; 95% CI 0.15, 0.25; P<0.001) and GC group (MD 0.14; 95% CI 0.11, 0.16; P<0.001). In addition, when comparing the vamorolone groups (vamorolone 2mg versus vamorolone 6mg), the vamorolone 2mg group had a significantly better results in the TTSTAND (MD -0.02; 95% CI -0.03, -0.02; P<0.001), but the vamorolone 6mg group had better results in the TTRW (MD 0.11; 95% CI=0.09, 0.14; P<0.001). **Conclusion:** Vamorolone improved TTSTAND in comparison with the placebo and the TTRW compared with placebo and glucocorticoids. Vamorolone 2mg group showed better results in TTSTAND compared to the 6mg group, while the 6mg group showed better results in TTRW. Larger, higher-quality randomized studies are needed to adequately evaluate the efficacy, safety and optimal dose of vamorolone in DMD.

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Efficacy of botulinum toxin in the management of trigeminal neuralgia: a systematic literature review

Geovanna Barros Rocha[1], Naely Nobre Costa[1], Stefanne de Cássia Pereira da Silva[1], Laura Lino Gonçalves[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Geovanna Barros Rocha; Email: geovannaroccha@outlook.com.

Background: Trigeminal neuralgia is characterized by unilateral, brief, shock-like paroxysmal pain, evoked by touch, which affects patients' daily activities, causing a direct impact on general status, sleep, mood and psychological health. The incidence of trigeminal neuralgia varies between 4.3 and 27 new cases per 100,000 people per year, being more common in women and increasing with age. The condition can lead to significant disability, with up to 45% of patients absent from their usual activities for 15 days or more. The initial treatment is pharmacological, with anticonvulsants, although many patients experience refractory pain. In this context, botulinum toxin type A has shown benefits in the treatment of neuropathic pain, reducing pro-inflammatory mediators and may be a safe and effective alternative in the management of trigeminal neuralgia refractory to conventional treatment. **Objective:** To analyze the variables associated with the treatment of trigeminal neuralgia using botulinum toxin. **Methods:** A systematic review was carried out, based on the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines, using the PECO strategy, to search the PubMed and Virtual Health Library (VHL) databases, including studies from 2014 to 2024, in English, Portuguese and Spanish. **Results:** 395 articles were found, of which 4 randomized clinical trials met the stipulated inclusion criteria, with only 3 being included in the study, due to the high risk of bias presented in one of them. Regarding participants, the average was 45 participants aged 57 years, the majority of whom were male. They were divided into a test group (GT) and control group (CG), which received, respectively, doses of botulinum toxin (25U to 100U) and placebo (doses of intramuscular saline solution). In the GT, a reduction of 66% was found at the 25U dose, 46% at the 50U dose and 78% at the 100U dose. In the CG, an average reduction of 20% was obtained. The main adverse effects found were: facial asymmetry and hematoma, ecchymosis and pain at the injection site. **Conclusion:** The use of botulinum toxin in trigeminal neuralgia was effective and safe in the articles analyzed, with mild and transient adverse effects and significant results in reducing pain compared to the placebo group, however, it was not possible to directly correlate doses higher doses of botulinum toxin with greater reductions in the pain scale. Therefore, it is important to carry out more clinical studies in this field, in order to elucidate the standardization of doses and the ideal stipulated therapy time.

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Efficacy of injectable sustained-release and autogel lanreotide for tumor reduction in pituitary adenomas: a systematic review and single-arm meta-analysis

Marianna Leite[1], Luana Violeta Rodrigues de Matos[2], Artur Menegaz de Almeida[3], Felipe Gouhie[4], Enzo Boccuto[1], ítalo Barros Andrade[5], Michele Kreuz[6], Rafael Batista João[7], Victor Domingos Lisita Rosa[8]

[1]Faculdade Santa Marcelina, São Paulo SP, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[4]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.
[5]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[6]Universidade Luterana do Brasil, Canoas RS, Brazil.
[7]Hospital de Caridade São Vicente de Paulo, Departamento de Medicina Interna, Divisão de Neurologia, Jundiaí SP, Brazil.
[8]Universidade Federal de Goiás, Faculdade de Medicina, Goiânia GO, Brazil.

Correspondence: Marianna Gerardo Hidalgo Santos Jorge Leite; Email: msmariannaleite@gmail.com.

Background: Numerous observational studies have investigated the use of lanreotide for pituitary tumors. However, it remains unclear whether there is a difference between injectable sustained-release (SR) or autogel formulations for tumor reduction. **Objective:** Therefore, we conducted a systematic review and meta-analysis to assess the efficacy of these drugs in mean tumor reduction. Additionally, we evaluated the proportion of patients experiencing any tumor shrinkage with different lanreotide formulations. **Methods:** We searched PubMed, Embase, and Cochrane Central for studies assessing injectable SR or autogel lanreotide for treating pituitary adenomas. We computed pooled mean difference (MD) or Risk Ratios (RR) for continuous and binary outcomes, respectively, with 95% confidence intervals (CI). A random effects model was used for the analysis. Heterogeneity was assessed with I2 statistics. Statistical analyses were performed with the R program (version 4.3.2) **Results:** We included eighteen studies comprising 892 patients, of whom 150 (41.7%) were treated with lanreotide autogel and 210 (58.3%) with injectable SR lanreotide. The mean age was 48.5 (12.3) SD, and 52.35% were female. Mean tumor reduction in patients using any formulation of lanreotide was 42% (95% CI=0.29 to 0.54; I2=94%). Overall, the proportion of patients experiencing any tumor reduction was 42% (95% CI=0.271 to 0.585; I2=88%). Such a proportion was 41% (95% CI=0.30 to 0.52; I2=82%) in those treated with injectable SR lanreotide, and 34% (95% CI=0.22 to 0.47; I2=82%) in those receiving autogel lanreotide. **Conclusion:** Lanreotide was associated with significant pituitary adenoma shrinkage. Moreover, subgroup analyses showed a higher proportion of patients with any tumor reduction in the injectable SR lanreotide group relative to the autogel lanreotide group.

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Efficacy of pimavanserin for patients with psychosis in Alzheimer's disease: a systematic review

Gabrielle Gomes Garcia[1], Amanda Rafaele da Silva[1], Ana Luiza Morgado Costa[1], João Carlos Amorelli Bohrer de Andrade Figueira[1], João Vitor Almeida e Silva[1], Leticia Kethelyn Bickel[1], Maryana Caetano da Silva de Oliveira[1], Pietro Pacheco Peregrini Cosentino[1], Sofia Gameiro de Souza[1], Izabela Jardim Rodrigues Pitta[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Gabrielle Gomes Garcia; Email: gabriellegarcia3g@gmail.com.

Background: Alzheimer's disease (AD) is the most prevalent type of dementia, characterized as a progressive neurodegenerative disease. About 25 to 50% of patients present with psychosis, featuring hallucinations and delusions. To this date, there are few safe and effective treatments available to treat this condition. New studies aim to explore the use of pimavanserin, an atypical antipsychotic that acts on the central nervous system, blocking the effects of the serotonin 5-HT2A receptor. It is being tested in individuals with psychosis in AD and has been approved by the FDA for the treatment of psychosis in Parkinson's disease. **Objective:** To evaluate the efficacy of pimavanserin for patients with psychosis in Alzheimer's disease. **Methods:** This literature review adhered to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. Studies searched four electronic databases: Cochrane, Pubmed, Lilacs and SciELO. The keywords used were: (alzheimer disease OR alzheimer dementia) AND (pimavanserin) AND (psychotic disorders). Randomized clinical trials published from 2014 to 2024, in English, and targeting the adult population were included. The exclusion criteria are as follows: trials not involving humans; no full text; non-randomized controlled studies. **Results:** 23 publications were found in the databases, and 9 of these were duplicates. Of the remaining 14, 6 studies were excluded as they did not fulfill the inclusion criteria. In the final analysis, only 3 original clinical trials were considered, with the remaining 5 publications referring to these same studies. In only one trial, all the individuals had AD (n=181), showing a 39.5% reduction in the psychosis score (NPI-NH) after 6 weeks of treatment with pimavanserin, 34 mg daily, compared to 19.3% in the placebo group. In the other trials, dementia and psychosis patients were analyzed, most of them suffering from AD (520 out of 1176). In these analyses, pimavanserin-responsive patients had fewer relapses (13%) of psychosis with the continuation of the drug than the placebo group, which discontinued treatment after 12 weeks (28%). Additionally, pimavanserin showed an improvement in the CGI-I neuropsychiatric scale compared to the placebo group, at the end of 8 weeks of treatment (mixed-effects model repeated measures analysis showed a least square means difference of -0.2 with a standard error of 0.07 (p=0.0140)). **Conclusion:** Primavanserin demonstrated positive results in treating psychosis in patients with AD. Also, the effects obtained indicate a potential efficacy in preventing psychotic relapses. However, the limited number of studies conducted so far investigating this therapeutic possibility, in addition to the limitations observed in the existing clinical trials, make this evidence questionable. Further studies are necessary in order to prove its benefits in these patients.

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Efficacy of subcutaneous apomorphine in Parkinson's disease: a meta-analysis of randomized controlled trials

Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Pedro Henrique Ximenes Ramalho Barros[1], Ester Vasconcelos Silva[1], Luciano Barbosa de Sousa Santos Filho[1], Mathias Williams Silva Cardoso Rabelo[1], Samuel de Castro Campos[1], Francisco Weslley Coimbra[1], Vitor de Deus da Rocha Ribeiro Gonçalves[2], Milene Vitória Sampaio Sobral[3], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Santa Casa de Misericórdia de Belo Horizonte, Belo Horizonte MG, Brazil.
[3]Universidade do Oeste Paulista, Presidente Prudente SP, Brazil.

Correspondence: Pedro Henrique Ximenes Ramalho Barros; Email: pedrohx587@gmail.com.

Background: Although the efficacy of apomorphine in the treatment of Parkinson's disease (PD) is subject to debate, it emerges as a promising option, especially for advanced cases. Not yet widely adopted in Brazil, its use stands out in recent studies, suggesting that this substance may offer significant benefits, challenging conventional perceptions and sparking interest in advanced disease management. **Objective:** To evaluate the efficacy of subcutaneous apomorphine in reducing motor symptoms in PD patients. **Methods:** Researchers systematically searched for randomized clinical trials comparing apomorphine and placebo in PD patients. The primary outcome of interest was the change in the UPDRS III scale up to 20 minutes after the intervention and after 90 minutes. Statistical analysis was performed using R Studio 4.3.2 software. **Results:** Three studies were included, totaling 163 patients. The average dose administered ranged from 4 to 5.4mg. The mean difference in the UPDRS III scale between the apomorphine and placebo groups was -14.12 points (95% CI -23.87 to -4.37; p<0.01; I^2=92%) up to 20 minutes after the intervention and -3.27 points (95% CI -6.12 to -0.43; p=0.02; I^2=0%) after 90 minutes. Meta-regression results applied to the outcome up to 20 minutes showed that the dosage accounted for 53.46% of the heterogeneity among studies and for 88.64% of the mean difference. The test for residual heterogeneity was not significant (p=0.171). The intercept of the regression plot was 38.01 points and the coefficient corresponding to the dosage was -11.24 points (p=0.0017). **Conclusion:** Evaluating the efficacy of subcutaneous apomorphine in PD, there was statistical significance for the reduction of motor symptoms of the disease.

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Eight-and-a-half syndrome due to pontine stroke: a case report

Laura Alonso Matheus Montouro[1], Luísa Bigatão Inácio[1], Júlia Gon Perez Nardoque[1], Giulia Rago Constâncio[1], João Renato Villas Bôas[1], Marina Morandin Rinaldi[1], Lívia Buqui Mazzoni[1], Rafaela de Freitas Valverde[1]

[1]Centro Universitário Padre Albino, Faculdade de Medicina de Catanduva, Catanduva SP, Brazil

Correspondence: Luísa Bigatão Inácio; Email: luisab.inacio@hotmail.com.

Case presentation: Female, 54 years-old, admitted in emergency with diplopia, eye movement abnormalities, vomiting and dizziness. On neurological examination, the patient scored 15 points on the Glasgow Coma Scale and 6 points on the National Institutes of Health Stroke Scale (NIHSS). The evaluation of cranial nerves: poor light reflex of left pupil, conjugate gaze deviation, divergente strabismus and nystagmus in the left eye and rhyme deviation to the left. About the muscle strength, the patient had a grade IV motor déficit according to the Medical Research Council in the left hemibody and no motor deficit in the right hemibody. The sensitivity analysis did not exhibit any changes. Brain magnetic resonance imaging (MRI) reveals lesions at dorsal pons and at small are of splenium of the corpus callosum and left occipital lobe characterized by high signal intensity on T2-weighted and Fluid-Attenuated Inversion Recovery (FLAIR) sequences, with real restricted diffusion on DWI/ADC map. **Discussion:** First reported by Eggenberger et al. in 1998, eight-and-a-half syndrome (EHS) is caused by a lesion of the abducens nucleus, pontine portion of the facial nerve and the medial longitudinal fasciculus (dorsal pons). Consequently, there'll be facial nerve palsy, horizontal gaze palsy and internuclear ophthalmoplegia (one-and-a-half syndrome plus VII nerve palsy). Rare neuro-ophthalmologic manifestation, 35 cases reported and two of them from brazilian origin. Cerebrovascular disease and demyelinating disease represents 90% of case etiologies. **Final comments:** Here, we report the third case of EHS in Brazil, caused by a pontine stroke. It is essential to know the complex anatomy of the pons and its pathways for the diagnosis of EHS and for the reasoning of the etiological investigation.

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Electroencephalography findings in a girl with Zellweger syndrome: case report

Bruno Eiji Nakano[1], Déborah Castro Ferreira de Oliveira[1], Paulo Emídio Lobão Cunha[1], Pedro Sudbrack Oliveira[1], Pedro Vicente Ferreira Naves[1], Isadora de Oliveira Cavalcante[1], Priscila Martins Câmara[1], Jennifer Jorge de Sales[1], Flavia Nunes da Silva Ferreira[1], Thaline Tiemi Nakano[2]

[1]Universidade de Brasília, Hospital Universitário de Brasília, Brasília DF, Brazil.
[2]Universidade Anhembi Morumbi, São Paulo SP, Brazil.

Correspondence: Bruno Eiji Nakano; Email:

Case presentation: A 2.5 year old girl, born full term, cesarean section with craniofacial dysmorphisms (large posterior fontanelle at birth), findings under neonatal hearing screening and abnormal liver enzyme tests. She also developed neonatal jaundice which required phototherapy. At 6 months of age, a magnetic resonance imaging of the brain and electroencephalogram (EEG) were carried out without any alterations. Generalized hypotonia, delayed neuropsychomotor development, severe cortical visual impairment, hearing impairment, hepatic dysfunction and severe oropharyngeal dysphagia

were identified. Exome sequencing identified two probably pathogenic variants, both in heterozygosity and in trans, in exon 1 and 8 of the PEX6 gene, associated with Peroxisome Biogenesis Disorder 4A (Zellweger) of autosomal recessive inheritance. She has 3 siblings with sickle cell trait and autism spectrum disorder, healthy parents and no history of consanguinity. At the age of 2.5, episodes of apnea lasting a few seconds onset. The EEG showed moderately disorganized background activity, occasional interictal epileptiform discharges (IEDs) of varying morphology (spike, sharp wave and polyspike-wave complexes) with multifocal distribution, predominating on the right side. **Discussion:** Zellweger syndrome is part of a group of genetic diseases related to the biogenesis of peroxisomes that, together with neonatal adrenoleukodystrophy and infantile Resfum's disease, constitute Zellweger spectrum disorders (ZSD). ZSD are autosomal recessive disorders with an incidence of 1:50,000 births and are related to mutations in the PEX gene, responsible for encoding the peroxin protein. The pathophysiology is related to a reduction in the number and dysfunction of peroxisomes. The clinical manifestations are varied, but include decreased muscle tone, facial dysmorphisms, growth delay, sensory and neurological dysfunction, renal and endocrine insufficiency, skeletal anormalies and developmental delays. Developmental brain malformations can be seen on MRI while electroencephalography may include diffuse cerebral dysfunction, continuous negative sharp waves and spikes at the vertex, and bilateral independent multifocal spikes, predominantly in the frontal motor cortex and its surrounding regions. Treatment consists of symptomatic therapy, multidisciplinary monitoring and genetic counseling. **Final comments:** The case is extremely relevant as it portrays a patient with no history of seizures, who started with apnea and whose EEG identified multifocal IEDs. These findings further reinforce the great clinical variability and complementary tests that can have a direct impact on the best management of the disease.

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Electroencephalography findings in Landau-Kleffner syndrome: case report

Bruno Eiji Nakano[1], Déborah Castro Ferreira de Oliveira[1], Paulo Emídio Lobão Cunha[1], Pedro Sudbrack Oliveira[1], Pedro Vicente Ferreira Naves[1], Isadora de Oliveira Cavalcante[1], Priscila Martins Câmara[1], Flavia Nunes da Silva Ferreira[1], Jennifer Jorge de Sales[1], Thaline Tiemi Nakano[1]

[1]Universidade de Brasília, Hospital Universitário de Brasília, Brasília DF, Brazil.
[2]Universidade Anhembi Morumbi,São Paulo SP, Brazil.

Correspondence: Bruno Eiji Nakano; Email: bruno.nakano1@gmail.com.

Case presentation: A 6-year-old male with expected neuropsychomotor development up to 2.5 years of age began to show language regression (echolalia and phonetic changes) with an impact on socializing. At the age of 3, the M-CHAT score was applied with a result of 8 and Autism Spectrum Disorder was considered. At the age of 5, verbal communication worsened. The cranial computed tomography scan showed no alterations and the electroencephalogram (EEG) showed occasional bilateral interictal epileptiform discharges (IEDs) in the centroparietal region, predominantly on the left. Valproic acid and periciazine were prescribed for behavioral control, with improvement but speech alterations remaining. The diagnostic hypothesis of Landau-Kleffner syndrome was considered and prednisolone (1.3mg/kg) was prescribed for 28 days. The patient's speech improved after taking the corticosteroid. **Discussion:** Laudau-Kleffner syndrome is a rare condition, with a common onset between 3-8 years of age and a predominance of 2 to 1 in males, characterized by language loss, IEDs on the electroencephalogram and seizures, although it is present in 80% of cases. Due to the outstanding behavioral alterations, the syndrome tends to have autism spectrum disorder and other cognitive disorders (neurodegenerative, traumatic, vascular) as a differential diagnosis and is therefore often diagnosed later, especially in cases where language dysfunction begins in younger children with a lower vocabulary. EEG findings show great variability, such as IEDs with polyspike, spike-wave, usually multifocal and bilateral, sometimes with a predominance in the temporal and parietal regions, activated mainly during non-rapid eye movement (NREM) sleep. In addition to these findings, it has been reported that a vertically oriented dipole can be found in the intraperisylvian region. **Final comments:** This case is extremely relevant, as it reinforces the variability in the morphology of IEDs which, associated with the wide range of clinical manifestations that make early diagnosis of Landau-Kleffner syndrome even harder, with a greater chance of cognitive sequelae.

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Elevated plasma levels of miR-206 in adult spinal muscular atrophy patients: a potential biomarker

Alexandre Hilário Berenguer de Matos[1], Felipe Franco da Graça[1], Cristina Iwabe[1], Marcondes Cavalcante França-Jr[1]

[1]Universidade Estadual de Campinas, Departamento de Neurologia, Campinas SP, Brazil.

Correspondence: Alexandre Hilário Berenguer de Matos; Email: alexandrehbm@gmail.com.

Background: Biomarkers are needed to assist in the management and follow-up of patients with spinal muscular atrophy (SMA) and circulating microRNAs (miRNAs) emerge as potential candidates. MiRNA-206 is involved in the denervation-reinnervation process and found to be clinically useful in amyotrophic lateral sclerosis. **Objective:** To analyze miRNA-206 expression in the plasma of adult patients with SMA and compare these results with healthy controls. **Methods:** Blood samples were collected from 28 adult patients with genetically confirmed spinal muscular atrophy (SMA) and 11 healthy controls. Blood was centrifuged, plasma was separated, and stored at -80°C. An RNA-enriched fraction was extracted from 200µl of plasma using the miRNeasy Serum/Plasma Advanced Kit (Qiagen®). Following this, the miRNA TaqMan® Advanced Kit was used for cDNA synthesis and specific assays to determine miR-206 expression levels in serum via RT-qPCR (Thermo®). Paired T-tests and Mann-Whitney U tests were used to assess significant differences in the expression of the selected gene. The significance level was set at $p \leq 0.05$. **Results:** The quantification revealed a significant ($p \leq 0.05$) increase in the relative expression of miR-206 in the plasma of SMA patients compared to healthy controls. This finding suggests a potential association between elevated miR-206 levels and the pathophysiology of SMA, further investigation is needed to elucidate the functional and clinical implications of this phenomenon. Interestingly, our results are consistent with previous findings demonstrating elevated miR-206 levels in adult patients with ALS, another motor neuron disease. **Conclusion:** Overall, our data suggest the potential of miR-206 as a stable and reproducible plasma biomarker for patients with spinal muscular atrophy (SMA). Further studies are necessary to investigate the correlation between miR-206 levels and clinical severity, as well as disease progression in SMA patients.

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Endovascular thrombectomy versus standard medical treatment alone in patients with acute ischemic stroke with large infarct (aspects ≤5): a systematic review and meta-analysis of randomized controlled trials

Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Matheus Felipe Henriques Brandão[2], Anthony Hong[3], Ana Beatriz Santos[3], Kairo Igor Freitas de Aquino[4], Pedro Barreto Pires Bezerra Filho[5], Gabriel Simoni[6], Gabriel de Almeida Monteiro[7], Victor Gonçalves Soares[8] , Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Faculdade de Medicina Nova Esperança, João Pessoa PB, Brazil.
[3]University of Costa Rica, San Pedro, Costa Rica.
[4]Centro Universitário Uninovafapi,Teresina PI, Brazil.
[5]Centro Universitário de João Pessoa, João Pessoa PB, Brazil.
[6]Centro Universitário de Várzea Grande, Várzea Grande MT, Brazil.
[7]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[8]Universidade Federal dos Vales do Jequitinhonha e Mucuri, Diamantina MG, Brazil.

Correspondence: Ocílio de Deus da Rocha Ribeiro Gonçalves; Email: ocilio41@gmail.com.

Background: Recent evidence suggests a beneficial effect of endovascular thrombectomy (EVT) in acute ischaemic stroke (AIS) with large infarct in comparison with standard medical treatment (SMT) alone. A previous meta-analysis evaluated the impact of EVT in AIS with extreme low ASPECTS (≤2). However there is no current comparative meta-analysis in the literature evaluating the effect of EVT in AIS with moderate to low ASPECTS (≤5). **Objective:** To assess the efficacy, the risk of bleeding and the risk of mortality in endovascular thrombectomy compared to standard medical treatment alone in patients with AIS with moderate to low ASPECTS (≤5). **Methods:** Medline, Embase, and Cochrane databases were systematically searched until June 6, 2024, assessing randomized controlled trials (RCTs) comparing EVT and SMT alone in patients with AIS with moderate to low ASPECTS (≤5). The primary outcomes of interest were: (1) proportion of successful reperfusion and (2) modified rankin scale (mRS) 0-2 and (3) 0-3. The secondary outcomes included: death from any cause, intracranial hemorrhage and EQ-5D-5L Utility Index. Data were examined using the Mantel-Haenszel method and 95% CIs for risk ratio (RR) and mean differences (MD). Heterogeneity was assessed using I^2 statistics. Software R Studio was used for statistical analysis. **Results:** Five RCTs were included in the meta-analysis, comprising 1586 patients. A total of 795 patients (50.13%) were assigned to receive EVT. The successful reperfusion was achieved in a proportion of 78.65% (95% CI 73.05% - 84.24%; I^2=61%) among the patients in the EVT group. Regarding mRS 0-2 (RR 2.71; 95% CI 2.04 - 3.61; I^2=0%) and mRS 0-3 (RR 2.04; 95% CI 1.52 - 2.74; I^2=62%), there was statistical significant difference favorable to EVT in neurological disability endpoints. Death from any cause was significantly lower in EVT group (RR 0.82; 95% CI 0.68 - 0.99; I^2=43%) and intracranial hemorrhage was significantly higher in the EVT group (RR 1.65; 95% CI 1.04 - 2.62; I^2=0%). Besides, the EQ-5D-5L Utility Index was significantly higher in the EVT group (MD 0.20; 95% CI 0.12 - 0.28; I^2=0%). **Conclusion:** EVT is an effective option to the treatment of large infarctions with moderate to low ASPECTS (≤5), and improves neurological functional outcomes and reduces mortality in comparison with SMT alone. However, it seems to show a higher risk of intracranial hemorrhage which is an important point to discuss for patients with considerable risk of bleeding.

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Enhanced RNA extraction and qPCR for microrna quantification in REM sleep behavioral disorder: potential biomarkers and methodology comparison

Sabrina Simplício de Araújo Romero Ferrari[1], Gabriel Ginani Ferreira[1], Deborah Almeida de Oliveira[1], Talita Guimarães Balieiro[2] , Danielle Macêdo Gaspar[3] ,

Manoel Sobreira-Neto[3], Raimundo Nonato Delgado Rodrigues[1], Pedro Renato de Paula Brandão[2], Simoneide Souza Titze-de-Almeida[1], Ricardo Titze-de-Almeida[1]

[1]Universidade de Brasília, Brasília DF, Brazil.
[2]Hospital Sírio-Libanês, Instituto de Ensino e Pesquisa, Brasília DF, Brazil.
[3]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Talita Guimarães Balieiro; Email: talitagbalieiro@gmail.com.

Background: The qPCR method for microRNA quantification in biological samples has been widely used to analyze clinical samples such as serum and blood plasma, in a quest for informative biomarkers, especially in neurodegenerative diseases. Previous studies suggest that microRNAs could potentially function as biomarkers for RBD, REM-sleep behavioral disease. Successful implementation of the methodology depends on both the microRNA extraction and target amplification protocols producing optimal results. **Objective:** To compare RNA purification and real-time qPCR methodologies for quantifying microRNAs as potential non-invasive biomarkers for RBD. **Methods:** Our cohort consisted of individuals with RBD and healthy controls. Data analysis in this study detected and excluded outliers, with values following a normal distribution. Pairwise group comparisons were carried out using the Student's t-test. RNA purification tests from serum samples of RBD patients and controls were conducted using two commercial kits from Qiagen: the miRNeasy/serum plasma kit and the miRNeasy serum/plasma Advanced kit (Qiagen, Hilden, Germany). Regarding qPCR methodologies, both utilized the Taqman system from Applied Biosystems (Applied Biosystems, Carlsbad, CA, USA). The conventional methodology, without target pre-amplification, was compared with the system based on RNA molecule pre-amplification followed by reverse transcription, known as the Advanced system. **Results:** The miRNeasy serum/plasma Advanced kit showed a significantly higher microRNA yield than the miRNeasy serum/plasma kit, measuring 1.45ng/µL versus 0.61ng/µL (P=0.006). Concerning qPCR methodologies, the Taqman Advanced system outperformed the conventional Taqman system, exhibiting earlier amplifications and more significant CT differences between the experimental groups. The study focused on the microRNA miR-19b-3p, known to have increased expression in RBD. Results revealed significant differences in CT values between the Advanced and conventional systems for both the control and RBD groups. The Advanced system consistently showed earlier amplification and larger CT differences between the groups, particularly for miR-19b-3p. Additional microRNAs, such as miR-7-5p, miR-21-5p, and miR-155-5p, were also examined. MiR-7 showed significant variations in CT values between the RBD and control groups. MiR-21 and miR-155 served as stable endogenous calibrators, along with cel-miR-39-3p as an exogenous spike-in calibrator. **Conclusion:** The current study highlights the importance of utilizing an enhanced RNA extraction system and a qPCR system with pre-amplification for accurate microRNA quantification in RBD. For calibration of the assays, our data recommend the exogenous calibrator spike-in cel-miR-39 and the endogenous miRNAs miR-21 and miR-155.

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Enhancing postoperative epilepsy assessment with Resectvol DL: a novel deep learning tool for brain lacuna segmentation

Raphael Fernandes Casseb[1], Brunno Machado de Campos[1], Wallace Souza Loos[3], Marcelo Eduardo Ramos Barbosa[2], Marina Koutsodontis Machado Alvim[1], Gabriel Chagas Lutfala Paulino[1], Enrico Ghizoni[1], Roberto Medeiros de Souza[3], Lara Jehi[4], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[3]University of Calgary, Calgary, Canada.
[4]Cleveland Clinic Foundation, Ohio, USA.

Correspondence: Raphael Fernandes Casseb; Email: rfcasseb2@gmail.com.

Epilepsy, a prevalent neurological disorder, often necessitates surgical intervention when pharmacological treatments fail. Accurate postoperative assessment is crucial for evaluating surgical outcomes and guiding future treatment strategies. In this study, we introduce ResectVol DL, a deep learning tool designed to automatically segment and characterize brain lacunas from postoperative MRI scans of epilepsy patients. Developed using the nnU-Net framework and leveraging the 3D U-Net architecture, ResectVol DL offers a fully automatic solution intended to avoid the traditional manual segmentation, which is tedious and time-consuming. Based on the Desikan-Killiany-Tourville segmentation atlas, our tool also identifies brain regions and estimates the volume of the parts removed. The dataset comprised post-operative T1-weighted MRI scans from 120 epilepsy patients who underwent resective surgery, with manual segmentations by experienced raters serving as the ground truth. We used five MRIs from healthy controls to test for false identification of brain lacunas. ResectVol DL demonstrated superior performance compared to two other freely available tools, achieving a median Dice similarity coefficient (DSC) of 0.92 (vs. 0.78 and 0.90), indicating high overlap with manual segmentations. When comparing lacuna volumes obtained manually with those obtained automatically, ResectVol DL presented the highest correlation coefficient (0.99 vs. 0.63 and 0.94) and the lowest median relative difference (9% vs. 44% and 12%). ResectVol showed no false positive results, while one of the other tools found false results in the five control images. The clinical implications of ResectVol DL include a rapid and precise volumetric analysis of resected brain tissue, which facilitates the identification of critical structures affected by surgery, enhancing our understanding of surgical impacts on cognitive and seizure outcomes. Furthermore, the tool's ability to process images without the need for preoperative scans simplifies its integration into clinical workflows. Beyond immediate postoperative evaluation, ResectVol DL holds promise for advancing predictive models estimating seizure freedom and cognitive recovery likelihood by providing volumetric information to feed the training of these models. In line with that, as a freely available tool, it can aid researchers and clinicians worldwide to improve epilepsy patient care through surgical planning and outcome prediction. ResectVol DL's accuracy, efficiency, and clinical relevance can be an asset for neurosurgeons and researchers, with potential to enhance patient care and prognostic capabilities.

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Epidemiologic analysis of the hospitalization of Parkinson's disease in Brazil from 2013-2023

Júlia Viana de Castro[1], Paulo Wendel Moura de Souza[1], Letícia da Costa Rios[1], Leslie Mateus de Sousa Nunes[1], Yasmin Dantas Babadopulos[1], Amanda Mendes Das Neves[1], Rodrigo Guimarães Bandeira[1], Mário Vinícyus Santos Alves[1], Matheus Henrique Brustolim Marcucci[1], Sofia Santana de Figueiredo[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Júlia Viana de Castro; Email: juliaviana2020@alu.ufc.br.

Background: Parkinson's disease (PD) is progressive, degenerative, chronic and irreversible, affects the central nervous system, being one of the neurological diseases that most affects individuals in the world, affecting between 1 and 3% of the world population over 60 years of age. **Objective:** To analyze the epidemiological profile of hospitalizations of Parkinson Disease in Brazil between the years 2013 and 2023. **Methods:** This study is an epidemiological investigation of hospitalizations of patients, between 30 and 79 years old, with PD between 2013 and 2023. Data were collected from the Sistema de Informação de Agravos de Notificação (Sinan Net) do Departamento de Informática do Sistema Único de Saúde (DATASUS), and data analysis was performed using Jamovi® software. **Results:** According to the data found in DATASUS, between 2013 and 2023, approximately 8421 hospitalizations of patients with PD in the established age group were recorded in Brazil. The age of hospitalized persons was predominant in the 60-79 age group, accounting for 65.96% of hospitalizations, although the number of persons under 60 years of age was significant (34.03%). There was also an increase in the number of hospitalizations in the years 2014 to 2016 compared to other years, with almost 3000 hospitalizations, and a considerable decrease in 2020, with a reduction of 32.74% compared to the previous year. In addition, making a percentage comparison between hospitalizations in the pre-pandemic period and the pandemic period, a percentage reduction of 24.12% is perceived. In addition, there was a considerable increase in the number of hospitalizations in 2023, with a percentage increase of 13.98% compared to the previous year, as well as an increase of 37.0% in the number of deaths compared to other years.Among these patients, it is also possible to observe a prevalence of males (61.38%), being more prevalent in all age groups, except in patients over 80 years of age. **Conclusion:** According to the data analysis, there is a decline in the number of hospitalizations from 2020 and in the following two years in all regions of the country, which is probably due to the COVID-19 pandemic, whose hospital care was concentrated on covid cases, generating a decrease in the number of hospital admissions for other diseases and in the medical demand by patients themselves. In addition, it is possible to observe an association between the increase in the number of hospitalizations and the number of deaths, especially in the age group of 60 to 79, since in this group it is common to have greater severity of symptoms. Regarding the age group and gender of the patients, there is a greater presence of men in all age groups, except over 80 years of age, which is explained by occupational risk factors, the fact that women have estrogen as a protective fact, and longer life expectancy.

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Epidemiologic analysis of the hospitalization of viral encephalitis in Brazil from 2013-2023

Júlia Viana de Castro[1], Matheus Henrique Brustolim Marcucci[1], Paulo Wendel Moura de Souza[1], Cynthia Paz Rodrigues[1], Ana Cecília Neves Rios Parente[1], Amanda Mendes Das Neves[1], Yasmin Dantas Babadopulos[1], Rodrigo Guimarães Bandeira[1], Sofia Santana de Figueirêdo[1], Mário Vinícyus Santos Alves[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Júlia Viana de Castro; Email: juliaviana2020@alu.ufc.br.

Background: Viral encephalitis is inflammation of the brain parenchyma caused by a virus. It is the most common type of encephalitis and often coexists with viral meningitis. Viruses invade the host outside the central nervous system and then reach the spinal cord and brain via hematogenous spread or in a retrograde manner from nerve endings. **Objective:** To analyze the epidemiological profile of confirmed cases of viral encephalitis in Brazil between the years 2013 and 2023. **Methods:** This study is an epidemiological investigation of the prevalence of viral encephalitis cases between 2013 and 2023 in Brazil.

Data were collected from the Sistema de Informação de Agravos de Notificação (Sinan Net) do Departamento de Informática do Sistema Único de Saúde (DATASUS), and data analysis was performed using Jamovi® software. **Results:** During the period of the SARS-CoV-2 pandemic from 2020 to 2022, there was a 24.87% reduction in the number of cases compared to the pre-pandemic period from 2013 to 2019. The reduction from 2019 to 2020, the first year of the pandemic, was 29.28% and from 2020 to 2021, the reduction was 10.08%. However, there was a gradual increase from 2022 onwards, with an increment of 16.56% by 2023. The age group from 40 to 49 years old was the one that suffered the most reduction in the number of cases in the pandemic period compared to the pre-pandemic period, with a percentage difference of 32.08%, and the one that was least affected were individuals from 50 to 59 years old, with a difference of 11.34%. Furthermore, the Northeast region of Brazil had the most considerable difference in the number of cases (16.12%). **Conclusion:** According to the data analysis, there has been a considerable decline in the number of cases during the pandemic period (24.37%) compared to the pre-pandemic period (2013 to 2023). This is due to social isolation, as well as the restrictive and preventive hygiene measures imposed during the pandemic period to control COVID-19, which may have contributed to lowering the rates of viral infections, directly influencing the drop in the number of cases of viral encephalitis. In addition, this difference can also be explained by a reduction in the notification of cases, as well as in hospital demand during the pandemic period, a scenario also seen in other diseases, such as meningitis. This can be verified by the gradual increase in the number of cases from 2022 onwards, whereas in 2023 the prevalence began to approach the values recorded before the pandemic.

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Epidemiologic profile of hospitalizations and deaths by brain neoplasms in the state of Bahia between 2013 and 2021

Cauã Araujo de Carvalho[1], Antônio Felipe Rocha Brandão[2], Bruna Rios Beserra[2], Bruno Levy de Souza Amaral Maia[1], Clara Carvalho Dantas Muniz[2], Ellen Santos Barreto[1], Fernanda Almeida da Silva de Sá Oliveira[2], Maria Clara Moysés de Siqueira[2], Thiago Costa Oliveira[2], Cleydson Santos[3]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[3]Hospital Mater Dei Salvador e Obras Sociais de Irmã Dulce, Salvador BA, Brazil.

Correspondence: Thiago Costa Oliveira; Email: thiagocoliveira23.1@bahiana.edu.br.

Background: Although rare, primary brain neoplasms consist of an relevant clinical entity with high organic impacts. It is known that the growing incidence of this cancer in Brasil is related to population ageing and to better access to diagnostic services, such as magnetic resonance. Therefore, it becomes necessary to identify its epidemiologic profile to implement health policies that go after the treatment and diagnosis of this illness. **Objective:** Describe the epidemiologic profile of hospitalizations and deaths by primary brain neoplasms in the state of Bahia between 2013 and 2021. **Methods:** Observational, descriptive and quantitative study that highlighted an ecological and retrospective approach of secondary data gathered in DATASUS/TABNET, without needing of approvation by the Ethics Committee. Data comprehend the period between 2013 and 2021 and were gathered in SIH/SUS, Oncology Panel of DATASUS and Mortality Atlas of INCA. There were investigated registers of hospitalizations and deaths by brain neoplasms in Bahia, taking into account the variables sex, age, race/color, therapeutical modality and treatment time. Exclusion criteria were other brazilian states and secondary brain neoplasms. **Results:** Between 2013 and 2021, there were registered 1.485 cases of primary brain neoplasms in Bahia, of which 47.8% (N=709) were in men and 52.2% (N=776) were in women. There was a progressive growth of 153% in notified cases between 2013 (N=97) and 2021 (N=246), mostly in the age range of 30 to 59 years old (51.5%; N=776). There were registered 7,051 hospitalizations at the analyzed period, of which 51.3% (N=3,622) were men and 48.7% (N=3,429) were women. About race/color, 45.5% (N=3,209) of the hospitalized were brown and 47.7% (N=3,364) did not have that information registered. In the analyzed period, radiotherapy was the preferential therapeutic modality, being used in 47.7% (N=704) of treatments, followed by surgery, with 18.2% (N=271), and chemotherapy, with 12.4% (N=185), while 20,4% (N=304) of treatments were not descripted. There were registered 3,800 deaths on the period, with a growth of 20.5% between 2013 (N=375) and 2021 (N=452) and more concentrated on the age range of 50 to 79 years old (N=2,199; 57.9%), mostly men (N=1.958; 51.5%). Mortality raised from 2.49 deaths by 100 thousand habitants (2013) to 3.01 deaths by 100 thousand habitants (2021). **Conclusion:** Brain neoplasms represent a relevant neurological entity which indicators are growing in Bahia. On the analyzed period, men, brown and adults were the most affected population, and radiotherapy was the preferential therapeutic modality, maybe because of inoperable lesions, reflecting late diagnosis and/or difficulty to access reference services. It is recommended the reformulation of health policies and specialized services of diagnosis and treatment of this cancer, in addition to new studies that follow the epidemiologic situation of the illness in Bahia.

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Epidemiological analysis of admissions, treatment, mortality and costs of cerebrovascular diseases in Brazil between 2017 and 2022

Thiago Oscar Goulart[1], Gleici da Silva Castro Perdoná[2], Rosane Aparecido Monteiro[3], Thire Baggio Machado Marazzi[3], Millene Rodrigues Camilo[3], Octávio Marques Pontes-Neto[3]

[1]Harvard University, Boston, MA, USA.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.
[3]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Thiago Oscar Goulart; Email: tgoulart@hsph.harvard.edu.

Background: Cerebrovascular diseases (CVD) represent a Global Health concern, being the first cause of disability worldwide and second cause of mortality (first in Brazil). Despite that, detailed data from Brazil was scarce and this study found alarming results. **Objective:** The study aims to analyze incidence, mortality, treatment, procedures and costs related to the three most common cerebrovascular diseases: Subarachnoid Hemorrhage (SAH), Intracerebral Hemorrhage (ICH) and Ischemic Stroke in Brazil. **Methods:** This is an epidemiological study. Data was obtained through DataSUS, a public database. For the search, we utilized the CID-10 notification system from I60 until I64, including admissions, deaths, treatments and costs related to each CID-10. **Results:** Between 2017 and 2022, 1,283,435 CVD cases were notified (SAH=4.7%; ICH= 8.8%; and ischemic stroke=86.5%). Admissions follow seasonality and there is evidence of subnotifications in 2020. 52.4% were Male and 47.6% were Female. Mortality percentages were: SAH=20.2%, ICH=25.3%, Ischemic Stroke=15.3%. Rates of Thrombolysis were alarming, but with an increasing tendency: 22969 were performed, from 1.6% in 2017 to 2.3% in 2022 of all ischemic strokes. 8290 aneurysms embolizations were performed (13.5% of all SAH cases). 4.9% of SAH cases were treated with neurosurgical clipping, demonstrating a decrease tendency after 2020, as well other procedures, such as Craniectomy and Angioplastia. Yearly costs related to these diseases increased steeply, from R$ 360,710,749.92 in 2017 to R$ 537,511,793.62 in 2022 (around US$107,502,358.00). **Conclusion:** This study has found increasing incidence and costs associated with low rates of thrombolysis and procedures in Brazil the results can direct Public Health policies in developing countries, and similar research in other low- and middle-income-countries are encouraged.

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Epidemiological analysis of deaths due to inflammatory polyneuropathies from 2018 to 2022

Paola Carolina Santos Foinkinos[1], Matheus Prado dos Santos[1], Maria Eduarda da Silva Santos[1], Bruna Martyres Gueiros[1], Zulma Catarina dos Santos Silva[1], Matheus de Carvalho Alves[1], Louise Araújo Corrêa[1], Hellen Jost Evaldt[2], Larissa Ádila Desireé Vieira de Andrade Duarte[1], Suzana Gomes de Oliveira[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.
[2]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Paola Carolina Santos Foinkinos; Email: paola.csfoinkinos@aluno.uepa.br.

Background: Inflammatory polyneuropathies are a group of diseases of the Peripheral Nervous System, which include Guillain-Barré Syndrome (GBS), Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) and others. They are acquired and related to immune response disorders, which affect components of the peripheral nervous system. As a result, these pathologies lead to physical and functional limitations, requiring early diagnosis and correct treatment to improve the clinical condition and prevent deaths. Therefore, a detailed investigation of deaths resulting from these diseases is necessary, as it can serve as a parameter for the clinical management of these pathologies in Brazil. **Objective:** To identify and analyze the epidemiological profile of deaths by residence caused by inflammatory polyneuropathies. **Methods:** This is an ecological, cross-sectional, descriptive and analytical study, with a quantitative approach, carried out by surveying secondary data available on the DATASUS electronic platform, about the epidemiological profile of deaths due to inflammatory polyneuropathies from 2018 to 2022. For the data analysis, the number of deaths by sex and color/race was considered. Then, the information was organized and tabulated in the Microsoft Excel 2021 software. **Results:** Between 2018 and 2022, 1,330 deaths were recorded due to inflammatory polyneuropathies, with the majority in 2022, with ¼ of the cases, followed by 20% in 2019, demonstrating an increase in occurrences. Deaths were predominant among males, with approximately 62% (824) of notifications, while only 38% were among women. Furthermore, the white color/race was the most affected among both sexes, accounting for 52.5% of the total, followed by 37% of browns and 7% of blacks. There were only 7 cases among yellow people and 4 among indigenous people. **Conclusion:** Therefore, based on the data made available by DATASUS, it is possible to conclude that there was an increase in the number of deaths due to inflammatory polyneuropathies throughout the analyzed period. Furthermore, it was found that males and white color/race had higher death rates. Therefore, the analysis of these data reinforces the importance of better targeting of treatment, since the evolution of this disease can cause physical and functional limitations, as well as early diagnosis, with the aim of reducing cases of death.

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Epidemiological analysis of deaths due to tuberculosis of the central nervous system in Brazil by geographic regions from 2013 to 2023

Vitor Hugo Bombarda Barros[1], Camila Vitória de Oliveira Zukovski[1], Maria Thereza Antoniolli Silva Sá Rosa[1], Bruna Mendes Ribeiro[1], Ana Beatriz Trentin de Moura Zago[1], Isabela Lopes Silva[1]

[1]Universidade de Araraquara, Araraquara SP, Brazil.

Correspondence: Vitor Hugo Bombarda Barros; Email: vitorhugobarros508@gmail.com.

Background: Tuberculosis, caused by Mycobacterium tuberculosis, constitutes a public health emergency in Brazil. Among the extrapulmonary manifestations of the disease, approximately 5-10% correspond to tuberculosis of the central nervous system (CNS-TB), which presents high morbidity and mortality rates. CNS-TB can develop from hematogenous dissemination or the rupture of subependymal or meningeal tuberculous foci. Its main clinical-pathological forms are tuberculous brain abscess/tuberculoma, spinal tuberculosis/tuberculous myelitis, and tuberculous meningitis. Some described complications of the condition include hydrocephalus, visual loss, focal neurological deficits, vasculitis, stroke, hemiplegias, cerebral edema, tuberculoma or abscess-induced seizures, diabetes insipidus, and syndrome of inappropriate antidiuretic hormone secretion. **Objective:** To gather, interpret, and discuss data regarding the number of deaths due to neurotuberculosis in the five regions of Brazil (North, Northeast, Central-West, South, and Southeast) from 2013 to 2023 and relate them to public health. **Methods:** A cross-sectional study was conducted. Data on deaths due to tuberculosis of the central nervous system from 2013 to 2023 were obtained from the "Sistema de Informações Hospitalares do SUS" (SIH/SUS) linked to DATASUS. In addition, a brief literature review on the pathology in question was performed. For this, the descriptors "Mycobacterium tuberculosis" and "Tuberculosis, Central Nervous System" were used, obtained with the help of the DeCS/MeSH platform. The database used was PubMed. Articles published between 2022 and 2024 were included. Articles that did not fall within this period and did not provide clear contributions to the study were excluded. Of the 161 articles found in the search, 9 articles were used to compose the brief review. Finally, a second search was conducted in the PubMed database using the descriptors "Brazil" and "Tuberculosis" for the period from 2013 to 2024 to enhance the discussion and findings. The data were presented according to regions of the Brazilian territory through tabular and graphical representation. **Results:** A total of 340 deaths due to tuberculosis of the central nervous system were recorded in Brazil over the 11 years studied. The Southeast region had the highest total number of deaths (170), followed by the Northeast (63), South (57), North (38), and Central-West (12). The year 2022 had the highest absolute number (48), while 2014 and 2015 were the least significant, with 19 deaths each year. The overall average number of deaths was approximately 31 per year. **Conclusion:** The distribution of deaths due to neurotuberculosis was heterogeneous across the territory. Factors such as the COVID-19 pandemic, socioeconomic situation, and underreporting of cases were identified as key influences on the data. The study provided a broad epidemiological view of the disease-related deaths in the country.

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Epidemiological analysis of deaths from neuromuscular junction and muscle diseases from 2018 to 2022

Matheus Prado dos Santos[1], Paola Carolina Santos Foinkinos[1], Suzana Gomes de Oliveira[1], Hellen Jost Evaldt[2], Bruna Martyres Gueiros[1], Maria Eduarda da Silva Santos[1], Zulma Catarina dos Santos Silva[1], Matheus de Carvalho Alves[1], Louise Araújo Corrêa[1], Larissa Ádila Desireé Vieira de Andrade Duarte[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil. [2]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Matheus Prado dos Santos; Email: imatheusprado@gmail.com.

Background: Neuromuscular diseases encompass several pathologies caused by involvement of the motor unit, which mainly encompasses the motor neuron, the neuromuscular junction and skeletal muscle tissue. Furthermore, the age range and mode of onset of these diseases are essential information for evaluation and differential diagnosis in adults and children, due to variable clinical manifestations, such as weakness, changes in reflex movements, spasticity, among others. Therefore, this study seeks to allow a better understanding of the epidemiological profile of patients affected and who died from neuromuscular junction diseases. **Objective:** To identify and analyze the epidemiological profile of deaths caused by neuromuscular junction diseases. **Methods:** This is an ecological, cross-sectional, descriptive and analytical study, with a quantitative approach, carried out by surveying secondary data from SIHSUS, available on the DATASUS electronic platform, referring to the number of deaths due to neuromuscular junction diseases in the period 2018 to 2022. To analyze the cases, the variables year, region, race/color and gender were considered, over the subsequent 5 years. Then, the data was organized and tabulated in the Microsoft Excel 2021 software. **Results:** Between 2018 and 2022, 9,543 deaths were recorded due to myoneural junction and muscle diseases, with a predominance of those in the Southeast (48%), Northeast (19.5%) and South (18%), and low occurrences in the Center -West (8%) and North (4.5%). Of these, 1,884 cases occurred in 2018, 2,040 in 2019, 1,614 in 2020, 1,939 in 2021 and 2,066 in 2022, showing a progressive increase in deaths, despite the drop in 2020. White color/race was predominant, representing 65% of notifications , followed by brown with approximately 27% and black with 4.5%; the yellows presented only 50 cases and the indigenous, 25. There was also a majority of males, with 64.5% of deaths in the period analyzed. **Conclusion:** Therefore, we observed a progressive increase in the number of deaths due to this disease, with a predominance in the southeast region compared to others. Additionally, there was a higher prevalence of deaths among white individuals and males. These results underscore the need for a more targeted approach in treating these conditions, considering their implications on patients' quality of life and mortality. The importance of early diagnosis is also highlighted to mitigate the adverse effects of these diseases and reduce the number of deaths.

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Epidemiological analysis of epilepsy cases in the state of Pará between 2018 and 2022: an ecological study

Walter Oliveira Verissimo[1], Leonardo Yuji Nihira Alencar[1], Dangilla Ribeiro dos Santos[2], Luiz Carlos Figueiredo Filho[2], Marcelle Rodrigues Carneiro de Souza Reis[3]

[1]Universidade Federal do Pará, Belém PA, Brazil. [2]Universidade do Estado do Pará, Belém PA, Brazil. [3]Centro Universitário do Planalto Central Apparecido dos Santos, Brasília DF, Brazil.

Correspondence: Walter Oliveira Verissimo; Email: walterverissimo17@gmail.com.

Background: Epilepsy affects 1% of the world's population and is characterized by epileptic seizures, the result of unbalanced neuronal discharges. There are two main types: focal and generalized, as well as seizures of unknown origin. Genetic predisposition is significant. Diagnosis is clinical and based on neurological tests. The WHO estimates that there are 65 million cases worldwide, with 5 million new cases every year, with a higher prevalence in low-income countries due to endemic diseases and lack of infrastructure. In Brazil, 2% of the population has epilepsy, with 150,000 new cases a year and an increase in mortality in the North and Northeast regions. **Objective:** To analyze the distribution of epilepsy cases in Pará between 2018 and 2022. **Methods:** This is a quantitative ecological study. We used data from the Notifiable Diseases Information System (SINAN) for the state of Pará between 2018 and 2022. The variables included were: sex, municipalities of hospitalization, race, age group, health regions and deaths. The data was tabulated using TABNET and analyzed using Microsoft Excel 2016. The non-parametric chi-square statistical test was applied, for equal expected proportions, with a significance value of 0.05%, with a 95% confidence interval (CI), as well as the relative risk value for comparing the risk of developing the outcome, both calculated using GraphPad Prism 8 software. **Results:** 5114 cases of epilepsy were analyzed, with 111 evolving to death and a 2.1% mortality rate. About gender, 55.73% of cases were in men (RR 1.16, 1.15 - 1.22, p<0.0001). Concerning race, brown people accounted for 62.59% of cases, followed by the group with no information, with 32.21%. Regarding age, individuals aged between 1 and 4 years had more cases reported, with 1002 cases or 19.59% of the total (RR 2.49, 2.22 - 2.80, p<0.0001). In addition, about the number of deaths compared between the sexes, 58.55% of the total was male (RR 1.26, 0.92 - 1.72, p=0.162). As for the analysis by health region, the Araguaia and Carajás regions stood out, with 17.44% and 16.41% of the cases. **Conclusion:** Epilepsy is more prevalent in the South, Southeast and Metropolitan regions of Belém, in men and children aged 1 to 4, groups most affected by the endemic aspects of the region. The data in this study can help reinforce health actions aimed at planning and implementing strategies for epidemiological surveillance and health policies directed at epilepsy, especially in the groups most affected by the disease. A limitation of this study is the possibility of underreporting of data. Actions are therefore needed to improve the updating of data, allowing for more work that can contribute to planning disease control and improving state indicators.

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Epidemiological analysis of Hansen's disease in the pediatric population in the state of Goiás: from 2010 to 2023

Victor Pedrosa Ferreira de Souza[1], Camila Pugliesi de Figueiredo[1], Amanda Mendes Santos[1], Michelle Oliveira Teixeira[1], Elzilaine Domingues Mendes[1], Priscila Ariede Petinuci Bardal[1]

[1]Universidade Federal de Catalão, Catalão GO, Brazil.

Correspondence: Victor Pedrosa Ferreira de Souza; Email: vvictor.pedrosa.vs@gmail.com.

Background: Hansen's disease or leprosy is an infection caused by the Mycobacterium leprae that can affect peripheral nerves, cause severe neural damage and lead to permanent grade-2 disability. Children are particularly vulnerable not only due to their immature immune systems, but also due to the possible psychological and emotional damage as a result of the stigma associated with this disease. **Objective:** To describe the profile of the pediatric population with leprosy in the state of Goiás in Brazil. **Methods:** A retrospective descriptive epidemiological study was carried out on the notified cases of leprosy in Goiás, between 2010 and 2023. The data was collected from the Brazilian Unified Health System (SUS) databases. The age group analyzed was from 0 to 14 years. The variables evaluated were: age, sex, skin color, operational class, number of multidrug therapy (MDT) doses and exit type. **Results:** Between 2010 and 2023, 978 cases were notified in the 0-14 age group. In 2015, there were 108 cases and in 2016, 64. In 2019, the number of cases was 53. In 2020, there was a significant drop to 34 cases. This data kept decreasing to 27 cases in 2021, 15 in 2022 and 6 in 2023. Regarding the age groups, in this period, 37 cases were reported in children aged 0-4, 292 in children aged 5-9 and 649 in people aged 10-14

years. More boys (500) were affected than girls (478). The majority of the individuals had brown skin color (62.37%). About the operational class, 573 individuals were diagnosed with multibacillary (MB) and 399 with paucibacillary (PB) leprosy. Of the filled data with number of MDT doses, 9.9% of PB and 23.4% of MB patients didn't complete the recommended treatment. About the type of exit, 779 were cured, 75 were transferred, 33 were misdiagnosed, 27 dropped out and 2 died. **Conclusion:** This study found that the number of notified cases has been decreasing since 2015, with an accentuated drop since the pandemic started in 2020. It's not possible to determine if this tendency is due to the reduction of the infection itself or if there has been an underreporting of new cases. However, the majority of cases are notified when the disease is already in an advanced stage, which is concerning. In addition, there is a noteworthy percentage of uncompleted treatment. Hence, prevention, early detection and treatment adherence are urgent to control this disease, especially considering Primary Care and the Family Health Strategy as a public policy.

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Epidemiological analysis of hospitalizations due to Alzheimer's disease in the Brazilian population between 2019 and 2023

Yasmim Melo Costa de Azevedo[1], Raul Pereira Bassotto Franco[2], João Pedro Rosa Barroncas[3], Marcelly Silva dos Santos Brito[4], Maria Regina Saddi Guerra[5], Cayara Mattos Costa[6]

[1]Universidade Nove de Julho , São Paulo SP, Brazil.
[2]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.
[3]Universidade Federal do Amazonas, Manaus AM, Brazil.
[4]Universidade Federal de Pernambuco , Caruaru PE, Brazil.
[5]Universidade Católica de Brasília, Brasília DF, Brazil.
[6]Universidade Federal do Maranhão, São Luís MA, Brazil.

Correspondence: Yasmim Melo Costa de Azevedo; Email: yaxmelo@outlook.com.

Background: Alzheimer's disease (AD) is a neurodegenerative condition that leads global dementia cases - accounting for 80% of the total. Its pathophysiology is closely associated with aging and it is considered a progressive, incurable disease. The memory, reasoning, and stimulus interpretation capacities, for example, of AD patients are compromised, requiring hospitalization due to complications that arise in their lives. According to the Ministry of Health, the number of hospitalizations due to AD in Brazil increased by 88% between 2010 and 2019. **Objective:** To analyze epidemiological data on hospitalizations for Alzheimer's Disease in Brazil between 2019 and 2023. **Methods:** Cross-sectional, quantitative, descriptive and observational research, with data from January 2019 to December 2023, collected in the SUS Hospital Information System, on the DATASUS platform. The total number of hospitalizations for Alzheimer's Disease were analyzed according to the International Classification of Diseases (ICD G30) in Brazil, considering the epidemiological variables: total hospitalizations per year, region, age group, sex, color/race, character of service and average length of stay in days. **Results:** In the period between 2019 and 2023, 7.866 hospitalizations for Alzheimer's were recorded in Brazil. The year 2023 had the highest number of hospitalizations, with 2.121 cases, while 2020 had the lowest number, with 1.212 cases. Between 2019 and 2020, there was a significant reduction of approximately 24%, possibly related to the COVID-19 pandemic and underreporting of cases. However, during most of the five-year period (2019-2023), an increase in the number of hospitalizations for Alzheime's was observed, with an average annual growth of 6%. In relation to the regions, the Southeast (47%) and the South (25%) were the most affected, while the North (3%) least. The majority of patients were women (66%), while men represented 34%. With regard to color, white people (n=3,977) and mixed-race people (n=2,079) were the most affected by the disease. Regarding the nature of these hospitalizations, emergency cases were the most significant, corresponding to 80% of the total, while elective cases represented 20%, with an average stay of 19.6 days. **Conclusion:** Hospitalizations for AD in Brazil showed a successive increase between 2020 and 2023, predominating in the Southeast region, on an urgent basis, and mainly affected white women aged 80 years or older. Therefore, in view of the growth in the incidence of AD and the number of elderly people in the country, it is urgent to invest in public policies focused on prevention and care strategies aimed at AD.

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Epidemiological analysis of hospitalizations due to multiple sclerosis in the Northeast of Brazil from 2018 to 2023

Rosa Carolina Santos de Oliveira[1], Álvaro Henrique de Moraes Santos[2], Daniela Meira dos Santos[1], Karolyne Vieira Mota[1], Mariana Mota Alves[2], Giovanna dos Reis Martins Barros[1], Helton Benevides Santana de Oliveira[2]

[1]Universidade Tiradentes, Aracaju SE, Brazil.
[2]Universidade Federal de Sergipe, Aracaju SE, Brazil.

Correspondence: Rosa Carolina Santos de Oliveira; Email: rosa.oliveira@souunit.com.br.

Background: Multiple sclerosis (MS) is an immune-mediated inflammatory demyelinating disease and is the most common demyelinating disease of the central nervous system (CNS). Among CNS pathologies, MS is one of the most recurrent cause of permanent disability in young adults, second only to traumatic brain injury. The incidence of MS varies due to environmental, geographical, population, gender, and age factors. Considering this, it is essential to understand the local panorama of this pathology to make advances in this field. **Objective:** Thus, the present study aims to identify the epidemiological profile of hospitalizations due to MS in the Northeast of Brazil, with the purpose of contributing to the development of effective therapeutic and preventive measures against this scourge. **Methods:** This is a multiple-group study with a quantitative, descriptive, and exploratory approach. Secondary data were collected through the hospital information system provided by DATASUS (SIH/SUS). The sample covers records between January 2018 and December 2023 in the Northeast region of Brazil. The analysis variables included age group, gender, and race. **Results:** During the analyzed period, between January 2018 and December 2023, there were 2,633 recorded hospitalizations for Multiple sclerosis in the Northeast region of the country. There was an increase in the number of hospitalizations per year, from 232 in 2018 to 858 in 2023, with the latter being the year with the highest number of cases. Regarding race, 184 (6.9%) hospitalized patients were white, 42 (1.59%) were black, 1,495 (56.77%) were brown, 63 (2.39%) were yellow, and 849 (32.24%) were not reported. Females accounted for 70.5% (1,857) of the hospitalizations, while males accounted for 29.5% (776). Considering the age group, 192 (7.29%) people aged 0 to 19 years were hospitalized, 704 (26.73%) were aged 20 to 29, 771 (29.28%) were aged 30 to 39, 550 (20.88%) were aged 40 to 49, 265 (10.06%) were aged 50 to 59, and 150 (5.69%) were aged 60 or older. **Conclusion:** In the present study, there was an increase in hospitalizations due to multiple sclerosis in the Northeast of Brazil. The profile of brown-skinned patients presented the highest numbers. The lowest numbers were among black individuals. Hospitalizations occurred mainly among females and in the age group between 20 and 39 years, with a slight predominance in the 30 to 39 years group. Patients at the extremes of age, such as children up to 4 years old and elderly individuals between 70 and 79 years old, accounted for few hospitalizations. Given these findings, further studies are needed to analyze the relationships of multiple sclerosis, its prevalence, and its influence on the conduct of the highlighted profiles.

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Epidemiological analysis of hospitalizations for facial paralysis and other paralytic syndromes across Brazilian regions

Aline Teixeira[1] , Ana Clara Lacerda Freitas[1], Laysa de Souza Maia[2] , Andressa Gonçalves Vicente[3], Ana Alice Lemos Lima[4] , Gabriela Hernandez Dumani[5], Ana Cristina Carvalho e Silva[3], Estefany Moreno de Souza[6], Geovana Franklim Gomes e Silva[7], Juliana Braga Rodrigies de Castro[8]

[1]Centro Universitário Funorte, Montes Claros MG, Brazil.
[2]Faculdades Integradas Aparício Carvalho, Jaru RO, Brazil.
[3]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[5]Universidade Franciscana, Santa Maria RS, Brazil.
[6]Universidade Aquino de Bolivia, Santa Cruz de La Sierra, Bolivia.
[7]Centro Universitário de Patos de Minas, Patos de Minas MG, Brazil.
[8]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Laysa de Souza Maia; Email: laysamaia1503@gmail.com.

Background: According to Walker et al. (2023), facial paralysis (FP) has an incidence of 10 to 40 cases per 100,000 inhabitants. Facial paralysis and paralytic syndromes are characterized by neurological conditions caused by viral infections or immunological diseases. However, FP occurs due to edema in the 7th cranial nerve. In contrast, Unspecified Paralytic Syndrome is characterized by paralysis in specific parts of the body without an identified cause. **Objective:** To analyze the distribution of hospital admission authorizations (AIH), average length of stay, and mortality rates among the five Brazilian macro-regions from 2019 to 2023 concerning Facial Paralysis and other paralytic syndromes. **Methods:** This is a descriptive and cross-sectional epidemiological study based on data verified from DATASUS from 2019 to 2023. The analyzed variables included hospitalizations, approved AIH, average length of stay, and mortality rate according to demographic regions, as well as sex and ethnicity. **Results:** Data analysis from 2019 to 2023 revealed a total of 117,315 AIH across Brazil, with the Southeast Region having the highest number of hospitalizations at 75.6% and the North Region at only 0.80%, suggesting that the distribution and quality of services offered are related to regional infrastructure. In 2019, Brazil recorded 8,677 hospitalizations and 25,017 approved AIHs, representing the highest number recorded during the studied period. However, in 2020, there was a significant reduction in these numbers, with only 6,321 hospitalizations and 22,292 approved AIHs; this decrease can possibly be explained by the underreporting of cases due to the COVID-19 pandemic. The total average length of stay was 71.5 days, with the Southeast Region showing data above this average at 133.5 days. The North Region had the lowest average length of stay at 10.0 days but had the second-highest mortality rate at 2.95%. The South Region had the highest mortality rate, while the Northeast Region had the lowest at 0.93%, possibly associated with better healthcare service conditions in this area. Males had the highest number of hospitalizations (24,231) and approved AIHs (65,406), while females had the highest average length of stay (85.5 days) and mortality rate (2.02%). The ethnicity with the most hospitalizations was Pardo (mixed race) with 9,934 cases but had the lowest average length

of stay at 65.3 days. **Conclusion:** This study highlights the importance of adequate and accessible healthcare infrastructure for the management of FP and paralytic syndromes in Brazil, fostering improvements in service quality and reducing disparities. Furthermore, it is evident that there is a lack of studies in Brazil specifically investigating the prevalence and incidence of facial paralysis and paralytic syndromes in the country.

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Epidemiological analysis of hospitalizations for microsurgery for intracranial tumors in Brazil between the years 2013 and 2022

Hálisson Ferreira Freitas Filho[1], Izabella Carolline Menescal Ferreira[1], Igor Moreira Miguez Godoy[2] , Antônio Marcos da Silva Henriques[1], Luan Víctor Silva Ribeiro[3] , João Victor Souza de Barros[1], Larissa Suzan Basilio e Silva[1], Kaliana Kennedy Oliveira Calixto[1], Felipe José Silva e Silva[1], Nathália Coimbra Coêlho[1]

[1]Universidade Federal do Pará, Belém PA, Brazil.
[2]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.
[3]Afya Faculdade de Ciências Médicas, João Pessoa PB, Brazil.

Correspondence: Hálisson Ferreira Freitas Filho; Email: halisson.filho@ics.ufpa.br.

Background: Microsurgery aims to obtain a histological diagnosis through the removal of intracranial tumors with the assistance of an intraoperative microscope, which provides illumination and magnification, allowing for the resection of tumors in critical areas in an attempt to restore neurological function. **Objective:** To perform a descriptive analysis of the epidemiological profile of hospitalizations for microsurgery for intracranial tumors in Brazil between 2013 and 2022. **Methods:** This is a descriptive, analytical, and retrospective study with a quantitative approach. Data were obtained from the Department of Informatics of the Unified Health System (DATASUS). Information was accessed through the Hospital Information System of the Unified Health System (SIH/SUS) and Hospital Production of the SUS. Procedures with the code "0403030145," corresponding to microsurgery for intracranial tumors, performed between January 2013 and December 2022, were included. The variables analyzed included the number of hospitalizations, length of stay, and total cost, analyzed over time and according to geographic region. **Results:** According to data from the Ministry of Health, there were 21,336 hospitalizations through SUS for intracranial tumor microsurgery in Brazil between 2013 and 2022. The number of hospitalizations increased from 2013 to 2015, decreased in 2016, remained relatively constant in 2017 and 2018, and increased again in 2019 and 2021, with a significant reduction in 2020. The Southeast and Northeast regions recorded the highest number of hospitalizations, with the Southeast region showing the greatest variation over the years. In 2013, the longest average length of stay was 16.3 days. The Northern region had the longest average length of stay at 17.9 days, while the Southeast region had the shortest at 13.8 days. The total resources allocated to the procedure were R$ 122,379,265.02 between 2013 and 2022. The Southeast region accounted for the majority of expenditures, corresponding to 42.60% (R$ 52,135,877.18), followed by the Northeast with 25.38% (R$ 31,067,048.82), South with 17.73% (R$ 21,702,877.70), North with 8.47% (R$ 10,373,860.39), and Midwest with 5.80% (R$ 7,099,600.93). **Conclusion:** It is concluded that the hospitalization rate varies significantly among the regions of the country, with a higher prevalence and monetary investment in the Southeast and Northeast regions, while the Midwest region presents the lowest rates in both aspects. The Southeast region records the shortest average length of stay, while the Northern region presents the longest. These data reveal an imbalance in the distribution of financial and structural resources in Brazil.

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Epidemiological analysis of hospitalizations from viral encephalitis in Brazil from 2019 to 2023: an ecological study

Leonardo Yuji Nihira Alencar[1], Janaína de Oliveira e Castro[2], Marcelle Rodrigues Carneiro de Souza Reis[3], Walter Oliveira Verissimo[1]

[1]Universidade Federal do Pará, Belém PA, Brazil.
[2] Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Centro Universitário do Planalto Central Apparecido dos Santos, Brasília DF, Brazil.

Correspondence: Leonardo Yuji Nihira Alencar; Email: leo1820.alencar@gmail.com.

Background: Viral encephalitis is inflammation of the brain that occurs when a virus directly infects the brain or when a virus, vaccine or other factor activates the inflammation. The mortality rate varies with the cause, but the severity of the epidemic resulting from the same virus varies over the years. Permanent neurological deficits are more likely to occur in patients who survive severe infection, which occurs mainly in the elderly, children and the immunodeficient. Therefore, given the considerable increase in cases, it is essential to analyze current data on the distribution of the disease to improve prevention actions and reduce mortality. **Objective:** To analyze the temporal distribution of hospitalizations and deaths from viral encephalitis between 2019 and 2023 in Brazil. **Methods:** This is an ecological, descriptive and quantitative study, which used data from the SUS Hospital Information System (SIH/SUS) tabulated by the Department of Informatics of the Unified Health System (DATASUS). Data was extracted between 2019 and 2023 from the Brazilian territory. The variables included were age group, gender, color/race, hospitalizations and deaths. The information was tabulated using TABNET and analyzed using Excel 2016. **Results:** In the period analyzed, there were 9818 hospitalizations and 497 deaths from viral encephalitis, both with peaks in 2019, corresponding to 24.8% of hospitalizations and 24.7% of deaths. In addition, 2023 had a considerable number of hospitalizations (2069), higher than previous years, with 1977 hospitalizations in 2022, 1647 in 2021 and 1689 in 2020. As for age group, throughout the period, children aged 1 to 4 had the highest number of hospitalizations, totaling 1784, with a peak in 2022 (433 cases), however, about the number of deaths, the number of occurrences was highest in the 30 to 34 age group, with 45 deaths in total and a peak in 2023 (11), followed by the 80 and over age group, with 39 deaths and a peak in 2019 (13). Males had the highest number of hospitalizations (58.3%) and deaths (53.4%), showing higher values than females in all years, with peaks in 2019, corresponding to 24.4% of deaths and 25.3% of hospitalizations for this group in the period. As for race, brown people had a higher number of hospitalizations (51.1%) and deaths (43.6%), with the peak in 2023 (26.1% of all hospitalizations for this group in the period). **Conclusion:** According to the data analyzed, the 1-4 age group had the highest number of hospitalizations, as well as males and brown people, groups that also had the highest number of deaths. A limitation of the study is the possibility of underreporting and patients being classified in incorrect groups. Further analysis of the distribution of viral encephalitis is therefore needed to integrate improvements in the control and prevention of the disease, to reduce the number of deaths resulting from it.

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Epidemiological analysis of intracranial hemorrhage cases in Goiás over the past five years

Gustavo Hayasaki Vieira[1], Giovana de Oliveira Araújo[1], Gustavo Cartaxo de Sousa Melo[1], Isabela Maciel Mesquita[1], Milena Almeida Barbosa[1], Paulo André Silva Miranda[1], Paulo Sérgio Machado Diniz[1], Rafaella Rodrigues de Freitas Vieira[1], Samira Dionísio Maia[1], Valentina Melo Rosa Centofanti[1]

[1]Pontifícia Universidade Católica de Goiás, Goiânia GO, Brazil.

Correspondence: Gustavo Hayasaki Vieira; Email: gushayasaki@gmail.com.

Background: Intracranial hemorrhage constitutes a serious clinical condition that can arise from trauma, hypertension, and vascular malformations. Due to its severity, the disease ranks among the leading causes of morbidity and mortality in Brazil, resulting in motor and cognitive dysfunctions that significantly impact the daily lives of affected patients. Thus, an epidemiological analysis in the state of Goiás over recent years is essential to better understand the factors contributing to this scenario and to enhance prevention, diagnosis, and treatment measures. **Objective:** To analyze the epidemiological profile of patients affected by intracranial hemorrhage from 2019 to 2023 in the state of Goiás. **Methods:** This is an ecological, descriptive, retrospective, and quantitative study using data from the Hospital Information System (SIH/SUS) provided by the Department of Information Technology of the Unified Health System (DATASUS), focusing on patients with intracranial hemorrhage in Goiás from 2019 to 2023. Filters applied included hospital morbidity within the SUS, geographical location, Central-West region, federal unit (Goiás), ICD-10 code (intracranial hemorrhage), processing year, age group, race/ethnicity, gender, number of hospitalizations, and deaths. Data were tabulated in Excel and descriptive statistics were conducted using R 4.40 with RStudio 2024.04.1 + 748. This study did not require ethics committee approval as it utilized publicly available secondary data. **Results:** During the study period, 868 deaths due to intracranial hemorrhage were confirmed, with an average annual incidence of 166.8 cases (±19.6). Individuals of mixed race were predominantly affected, comprising 78.34% of total deaths. The age group most vulnerable to the disease was 60-69 years, accounting for 198 deaths. Regarding gender, the average number of deaths was 87.8 (±19.7) for males and 79 (±2.54) for females. Additionally, the year 2021 recorded the highest number of hospitalizations, totaling 621 cases; whereas 2019 had the lowest with 462 hospitalizations within the analyzed period. Moreover, both hospitalizations and deaths showed an increasing trend until 2021, followed by a decline in subsequent years: 617 cases in 2022 and 567 in 2023. Furthermore, the age group most frequently hospitalized was 50-59 years (21%). **Conclusion:** The majority of intracranial hemorrhage deaths occur among individuals of mixed race. Additionally, males exhibit a higher mortality rate compared to females, and the disease has shown a declining trend in recent years. Individuals aged 60 to 69 years are the most vulnerable, resulting in higher mortality rates, while those aged 50 to 59 years experience the highest frequency of hospitalizations due to intracranial hemorrhage. Therefore, this study provides data that can be used as a basis for prevention strategies carried out by higher academic institutions and by the State Health Department.

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Epidemiological analysis of meningococcal disease in children up to 14 years old

Samara Brissi Benito da Silva[1], Gabriel Vergolino Eleres[2], Amábile Cirilo da Silva[3], Rafaella Ferreira dos Santos[4], Isabela Nishimura Megiani[5], Hévila Ferreira Gomes Medeiros Braga[6]

[1]Universidade de Mogi das Cruzes, Mogi das Cruzes SP, Brazil.
[2]Fundación Barceló, Buenos Aires, Argentina.
[3] Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[4]Universidade Santo Amaro, São Paulo SP, Brazil.
[5]União das Faculdades dos Grandes Lagos, São José do Rio Preto SP, Brazil.

[6]Universidade da Integração Internacional da Lusofonia Afro-Brasileira, Redenção CE, Brazil.

Correspondence: Samara Brissi Benito da Silva; Email: samarabrissi@gmail.com.

Background: Meningococcal disease (MD) is defined as a severe infection caused by the bacterium Neisseria meningitidis (meningococcus), which can lead to inflammation of the meninges (meningococcal meningitis) and bloodstream infection (meningococcemia), and is transmitted by the respiratory routes, such as secretions and droplets. Worldwide, an estimated 500,000 cases and 50,000 deaths due to MD occur annually, and in Brazil, it is considered endemic. In addition, the disease has 12 serogroups, with serogroups A, B, C, W, Y, and X being the most prevalent in reported cases. **Objective:** To analyze the epidemiological profile of Meningococcal Disease according to its serogroups between 2014 and 2023, among children up to 14 years of age. **Methods:** A descriptive ecological study conducted using data from the Notifiable Diseases Information System (SINAN). Confirmed cases and deaths from meningococcal meningitis (MM) with meningococcemia (MCC) in the population up to 14 years old were considered from 2014 and 2023. The variables used were: confirmed cases, deaths, age group, race/ethnicity, serogroup, and Brazilian region. The data was analyzed descriptively and organized in Microsoft Excel spreadsheets. **Results:** In the period analyzed, 1,266 cases of meningococcal disease were identified in Brazil. It was found that the disease predominated in 555 cases (43.8%) in individuals identified as white, aged 1 to 4 years (33%), located in the Southeast region (67%), with the majority of cases resulting in hospital discharge (77.6%). Among the analyzed serogroups, type B was the most prevalent with 338 cases (26.6%), and also resulted in the highest infant mortality among all serogroups with 66 deaths (52.1%). However, the vaccine against serogroup B is not provided by the Brazilian Unified Health System (SUS), accessible only through private healthcare service. This highlights a potential correlation between the increased number of cases and the higher infant mortality rate associated with this serogroup. **Conclusion:** The data shows the importance of recognizing Meningococcal Disease according to the epidemiological profile of the studied population and emphasize the need of developing strategies for disease control. The unavailability of the vaccine for serogroup B through SUS is concerning for public health in Brazil, particularly due to its prevalence in reported cases and deaths among children. Moreover, it is essential to raise public awareness about the disease and vaccination for prevention, which has been shown to be positive in the serogroups where vaccines are available through SUS, as well as early diagnosis and treatment, as indicated in the literature, have shown improved prognosis.

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Epidemiological analysis of meningoencephalic tuberculosis in vulnerable populations in Brazil: 2014-2023

Helmut Kennedy Azevedo do Patrocínio[1], Matheus Procópio Guimarães[1], Tiago Lameque de Sousa e Silva[1], Pedro Henrick Guimarães Carvalho[1], Cláudio de Souza Fernandes Júnior[1], Cláudio de Souza Fernandes[2], Tereza Cristina Batista Dias[1], Maria Eduarda Medeiros Martins[1], Matheus Araújo de Medeiros[1], Iury Hélder Santos Dantas[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[2]Universidade do Estado do Rio Grande do Norte, Mossoró RN, Brazil.

Correspondence: Helmut Kennedy Azevedo do Patrocinio; Email: hell.kenn@gmail.com.

Background: Meningoencephalic tuberculosis (MTB) is the most severe form of tuberculosis (TB), characterized by high mortality and morbidity rates. Risk factors for developing MTB include immunocompromised states, such as chronic steroid use, diabetes mellitus, alcoholism, and, primarily, HIV co-infection. To date, no studies have specifically addressed the risk of developing MTB in the vulnerable populations investigated in this study. **Objective:** Conducting an epidemiological analysis of the health issues associated with the development of tuberculosis in vulnerable populations. **Methods:** A retrospective epidemiological study using data from the Notifiable Diseases Information System to analyze the occurrence of MTB in vulnerable populations - health professionals, persons deprived of liberty (PDL), homeless people (HoP), and immigrants - in Brazil from 2014 to 2023. For each vulnerable population, both descriptive and inferential statistical analyses were performed to assess the occurrence of alcoholism, diabetes, smoking, mental illness, drug use and AIDS. To examine factors associated with TBM, binomial logistic regression analyses were conducted. Data analysis used the statistical software R. **Results:** Of the 5960 identified cases of MTB, 105 occurred in PDL, 102 in HoP, 62 in health professionals, and 31 in immigrants. When comparing MTB with exclusively pulmonary TB in all populations, the presence of AIDS was associated with a significantly higher odds ratio (OR): PDL (OR: 11.8, P<0.001), HoP (OR: 9.71, P<0.001), health professionals (OR: 9.01, P<0.001), and immigrants (OR: 8.53, P<0.001). Although other conditions had high odds ratios, such as mental illness in immigrants (OR: 4.7, P=0.064) and alcoholism in PDL (OR: 1.65, P=0.1) and HoP (OR: 1.4, P=0.2), no statistical significance was achieved. AIDS was the predominant factor in all four populations studied, with the highest proportion observed in the HoP group (75.25%) and the lowest in health professionals (31.15%). In addition, the HoP group showed a high frequency of alcoholism (56.82%), illicit drug use (60.64%) and smoking (41.11%). However, none of these variables was identified as a significant risk factor. **Conclusion:** The results of this study emphasize the importance of investigating the factors associated with the development of MTB in Brazil. AIDS was consistently associated with a higher risk of MTB in all the populations. Conditions such as alcoholism and mental illness also had odds ratios above 1, but did not reach statistical significance. A limitation of this study was the limited sample size, which restricted the development of a more accurate statistical model. Therefore, further research is needed to comprehensively investigate MTB, especially in vulnerable marginalized groups. Understanding the predisposing conditions for the occurrence of MTB is crucial for the implementation of targeted interventions tailored to each vulnerable population.

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Epidemiological analysis of migraine hospitalizations in the state of São Paulo from 2019 to 2023: an ecological study

Bruna Ravany Farias Marques[1] , Nicolas Inkei Chiba Frazao[2], Maria Clara Calixto Caetano[3], Giulia Chiavegato Locatelli[4], Beatriz Moraes Florenzano[5], Juliana Braga Rodrigues de Castro[6]

[1]Centro Universitário dos Guarapes, Guanambi BA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.
[3]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[4]Universidade Nove de Julho, São Paulo SP, Brazil.
[5]Faculdade Santa Marcelina São Paulo, São Paulo SP, Brazil.
[6]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Maria Clara Calixto Caetano; Email: maria.caetano@sou.unaerp.edu.br.

Background: Migraine is a chronic neurological disease with a prevalence of 15.2% in Brazil. Clinically, it manifests as recurrent episodes of headache associated with other symptoms, dependent on triggering factors. According to the World Health Organization (WHO), half of the global population experiences headaches at some point in life, with a higher prevalence among females, making it the fourth leading cause of disability in women worldwide. Therefore, migraine needs to be addressed as a public health problem since its occurrence in emergency settings is prevalent and potentially limiting, negatively impacting well-being, quality of life, and imposing a substantial burden on patients and Society. **Objective:** To analyze the epidemiological characteristics of migraine hospitalizations in the state of São Paulo from 2019 to 2023. **Methods:** An ecological, retrospective, and descriptive study was conducted using information on migraine hospitalizations in the state of São Paulo from 2019 to 2023. Information was obtained through consultation with DATASUS. The following variables were analyzed: hospitalizations, age group, sex, race/ethnicity, and year of admission. A descriptive analysis of the data was performed, calculating relative and absolute frequencies for the categorized variables. As this research involved secondary data, there was no need to submit the project for approval by an Ethics Committee. **Results:** A total of 9,316 hospitalizations were analyzed in the state of São Paulo during the period from 2019 to 2023. The results show a higher incidence of cases among females, accounting for 68.9% (n=6,424). The year with the highest number of hospitalizations was 2019, responsible for 22.5% (n=2,101). White individuals were the main group reporting migraine complaints during the analyzed period, representing 58.8% (n=5,476). The predominant age group was 30 to 39 years, accounting for 19.2% (n=1,792). **Conclusion:** The data reveal a significant number of hospitalizations due to migraines in the state of São Paulo in 2019, which decreased and tends to return to previous levels, suggesting impacts of the Covid-19 pandemic, such as underreporting. Additionally, there is a notable incidence of hospitalizations among adults aged 20 to 29 years and among white individuals, representing more than half of the cases. This study has limitations, such as the underreporting of hospitalizations due to the use of a secondary data system and the inability to associate the cause of migraines with the effect of hospitalization. Therefore, more studies are needed to investigate the high number of migraine hospitalizations to understand the real causality in the Brazilian context.

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Epidemiological analysis of mortality rate and treatment cost of uncontrolled epileptic seizures in the state of São Paulo

Marcela Cristina de Souza Lemus[1], Amanda Aparecida Aguiar Balduce[1], Nivea de Mattos Amarante Rodrigues[1], Victor Iovino Ferri[2], Henrique Rocha Mendonça[1]

[1]Universidade Federal do Rio de Janeiro, Macaé RJ, Brazil.
[2]Universidade de Mogi das Cruzes, Mogi das Cruzes SP, Brazil.

Correspondence: Marcela Cristina de Souza Lemus; Email: marcelalemus47190@gmail.com.

Background: Epilepsy is a chronic neurological disorder characterized by recurrent unprovoked epileptic seizures. Between 2020 and 2023, more than 229,420 individuals were hospitalized due to epilepsy, resulting in significant costs to the Brazilian Unified Health System (SUS) for the management of this condition. **Objective:** To analyze the epidemiological and economic aspects related to the hospitalization costs and mortality rates associated with uncontrolled epileptic seizures in the state of São Paulo. **Methods:** This observational ecological study utilized data obtained from the Hospital Information System (SIH/SUS) provided by the Department of Informatics of the Brazilian Unified Health System (DATASUS). We specifically focused on patients with uncontrolled epileptic

seizures in state of São Paulo from 2020 to 2023. Key variables analyzed included the number of hospitalizations, total costs incurred, mortality rates, and number of deaths per year of management. Ethical approval was not required as the study employed secondary data. **Results:** In the analyzed period, a total of 78,019 hospitalizations due to uncontrolled epileptic seizures were observed in the SUS of the Southeast region, with state of São Paulo accounting for 38,699 of these cases. The average cost per hospitalization in state of São Paulo during this period was R$1,255.06, resulting in a total expenditure of R$48,831,858.69. In 2023 (R$14,467,046.53), the highest total costs of hospitalizations for uncontrolled epilepsy were observed, followed by 2022 with costs amounting to R$13,582,040.96. The year 2022 represented an increase of approximately 30.7% compared to 2021, with a total cost of R$10,393,846.22. There was no significant difference in costs between 2020 and 2021. Over the study period, 1,419 deaths were reported, with mortality rates consistently above 3.0 per 100 hospitalizations: 3.69 in 2020, 3.57 in 2021, 3.58 in 2022, and 3.82 in 2023. Data underreporting is acknowledged as a potential limitation. **Conclusion:** There was an increase in costs, deaths, and hospitalizations due to uncontrolled epileptic seizures in the state of São Paulo between the first and last years analyzed. A significant highlight occurred during the period of the COVID-19 pandemic declared by the World Health Organization, which saw an increase in mortality rates from 2021 onwards and a rise in total annual hospitalization costs from 2020. These findings may be attributed to difficulties in healthcare access, utilization of telemedicine, and heightened fear of severe COVID-19 during the pandemic, exacerbating the epilepsy treatment gap with more hospitalizations, deaths, and costs related to epileptic seizures within the SUS. Furthermore, in 2023, an increase in mortality is observed, potentially linked to the worsening of epileptic seizures following COVID-19 infection.

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Epidemiological analysis of patients with epilepsy in Brazil

Luisa Maria Resende Morais[1], Gleison Carlos Arantes Filho[1], Leonardo Brandão Barreto[1]

[1]Universidade Federal de Ouro Preto, Ouro Preto MG, Brazil.

Correspondence: Luisa Maria Resende Morais; Email: luisamresendem@gmail.com.

Background: Epilepsy is a disease characterized by a chronic predisposition of the brain to generate epileptic seizures, whose underlying causes include structural lesions, genetic/metabolic alterations and infectious/autoimmune diseases, and is associated with higher mortality, psychiatric comorbidities and psychosocial problems. **Objective:** To analyze hospital characterization, clinical outcome and cost-effectiveness for patients with epilepsy in Brazil. **Methods:** An observational and retrospective study using data obtained from the Hospital Information System of the Unified Health System, between January 2014 and December 2023, with the following variables: hospitalizations, average length of stay, average cost, mortality rate, gender and age group. Descriptive and inferential statistical analysis was used. **Results:** The most affected profile was males (57.63%) and those aged between 1 and 4 years (16.27%) and 50 to 59 years (10.44%). In the last 10 years, 557,829 hospitalizations for epilepsy were recorded, most of them in the Southeast (42.77%). All regions of Brazil saw a significant increase in the number of hospitalizations, with the highest increase (59.81%) in the Northeast (p=0.001) and the lowest (10.59%) in the Southeast (p=0.012). The average hospital stay in the country was 6 days, with an increase in all regions, however, only in the Northeast, South and Midwest was the increase statistically significant (p<0.05). The biggest increase was in the Northeast, which went from 5.4 days in 2014 to 6.8 in 2023. The average cost to the Brazilian health system was R$848.41, and increased significantly over the period considered in all regions, especially in the Northeast, with an increase of 58.68% (p=0.025). Finally, the mortality rate from epilepsy in the country was 2.48 per 100 inhabitants, with the highest average in the Northeast (2.88) and the lowest in the South (1.82). There was an increase in the mortality rate in all regions of Brazil, with statistically significant increases in the North, Southeast and South, with increases of 69.28%, 47.14% and 41.29%, respectively. **Conclusion:** Epilepsy mainly affects children and the elderly, and is the main cause of hospitalization in children. The increase in average hospital costs is associated with treatment using new generations of antiepileptic drugs, the request for imaging tests and the use of specialized teams, which are costly resources. The increase in the mortality rate can be explained by the increased prevalence of the disease, the significant incidence of infectious diseases of the nervous system related to the etiology of epilepsy in the country and the still limited access to specialists and appropriate treatment. The findings of this study reveal regional disparities, highlighting the need for differentiated approaches to the management of epilepsy in different regions of the country, and underscore the importance of effective interventions to improve its prognosis in Brazil.

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Epidemiological analysis of pediatric hospitalizations for epilepsy in the state of São Paulo: an overview of the last 10 years

Amanda Fortes Cavalcante de Oliveira[1], Luiza Bisognin Marchesan[2], Celijane Almeida Silva[3], Isadora Carvalho de Sousa[4] , Mylena Cordeiro Aranha[4], Mariana Teixeira Mesquita[5], Luana Dutra Pinheiro da Silva[4], Lara Maia Pereira[4], Laiane Rodrigues Macêdo[6], Bárbara Pinheiro Gama de Andrade[1]

[1]Universidade Federal do Delta do Parnaíba, Parnaíba PI, Brazil.
[2]Universidade Franciscana, Santa Maria RS, Brazil.
[3]Centro Universitário Unifacisa, Campina Grande PB, Brazil.
[4]Universidade Potiguar, Natal RN, Brazil.
[5]Universidade Estadual de Campinas, Campinas SP, Brazil.
[6]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Mariana Teixeira Mesquita; Email: maritmesquita@gmail.com.

Background: Epilepsy is the main neurological condition that affects the pediatric age group. Its occurrence is significant in Brazil, with a total of 215,332 hospitalizations in the last 10 years. This condition is one of the most frequent complaints in pediatric emergency care, representing 1 to 5% of all visits. It is known that up to 10% of the population experiences at least one epileptic seizure by the age of 16. Understanding the epidemiology of pediatric epilepsy in São Paulo is limited. This analysis is essential to guide health policies and improve the treatment of this condition, aiming for a better quality of life for affected children. **Objective:** To describe the epidemiological profile of epilepsy among pediatric patients in the state of São Paulo from 2014 to 2023. **Methods:** A descriptive ecological study carried out through data collection from the SUS Hospital Information System (SIH/ SUS). The study population includes hospitalization for epilepsy in pediatric patients (from 0 to 14 years old) reported in the state of São Paulo between 2014 and 2023.The following variables were researched: age group,gender and color race. **Results:** Out of a total of 215,332 hospitalizations notified in Brazil during the period evaluated,the state of São Paulo accounted for 41,225 hospitalizations, representing approximately 20% of all cases. The most affected age group was from 1 to 4 years, with 17.209 hospitalizations, followed by 5 to 9 years, 1 year or younger and 10 to 14 years, with respectively 9.370, 7.738 and 6.908 hospitalizations. Regarding gender, there is a prevalence of males across all age groups,totaling 22,786 cases,with the 1 to 4 years age group being the most affected,representing 9736 cases. As for color/race, whites (n=22,304) showed the highest incidence, followed by brown (n=12,075). In all age groups, this pattern remained consistent. However, the numbers for black (n=1253), yellow (n=183) and native people (n=8) were significantly lower compared to the previous ethnicities. **Conclusion:** The study corroborates the existing literature by reinforcing the predominance of pediatric hospitalizations among white male children aged 1 to 4 years.Thus, considering the emerging state in pediatric hospitalizations and the detrimental implications to patients, it is crucial to observe inequalities, identify at-risk groups, and better prepare healthcare professionals to receive these patients. In this regard, the important role of primary care in preventing hospitalizations is highlighted.

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Epidemiological analysis of spina bifida in Brazil and of maternal factors

Amanda Mendes Santos[1], Camila Pugliesi de Figueiredo[1], Michelle Oliveira Teixeira[1], Victor Pedrosa Ferreira de Souza[1], Wânia Rezende Lima[1]

[1]Universidade Federal de Catalão, Catalão GO, Brazil.

Correspondence: Amanda Mendes Santos; Email: amanda.mendes@discente.ufcat.edu.br.

Background: Spina Bifida (SB) is a congenital defect resulting from the incomplete closure of the neural tube, between the third and fourth week of pregnancy, being one of the major causes of childhood motor disability. Although there are important genetic patterns that contribute to the prevalence of neural tube defects, it is known that factors, such as folic acid supplementation, are of the utmost importance in their etiology. Therefore, it is evident the need to elucidate what other external variables may contribute to its development. **Objective:** To evaluate infants with SB in Brazil and the maternal epidemiological profile. **Methods:** It's a retrospective descriptive epidemiological study of live births with SB in Brazil, based on data from the Congenital Malformations, Deformities and Chromosomal Anomalies Monitoring Panel. The data from 2014 to 2024 was analyzed, with the results from 2023 and 2024 being preliminary. The variables included were marital status, educational level, month of pregnancy in which prenatal care began and number of prenatal consultations. **Results:** In total, 7593 babies were born with SB. Specifically, there was a prevalence of lumbar SB (588), followed by cervical (425), sacral (370) and thoracic (123). Lumbar SB without hydrocephalus was the most prevalent (390), followed by cervical with hydrocephalus (312), sacral without hydrocephalus (247), lumbar with hydrocephalus (198), sacral with hydrocephalus (123), cervical without hydrocephalus (113), thoracic without hydrocephalus (71) and thoracic with hydrocephalus (52). 465 live births had unspecified SB with hydrocephalus and 5697 had unspecified SB. As for maternal characteristics, 45.6% were single; 32.9% married; 19.3% in stable union; 1.4% legally separated and 0.1% widowed. In 0.7% of cases, this information was not available. In addition, 32 had no formal education, 177 had from 1 to 3 years, 1172 had from 4 to 7 years, 4636 had from 8 to 11 years, 1528 had 12 years or more and in 48 cases the data was blank. Starting prenatal care early and, consequently, folic acid supplementation is essential in preventing SB. Thus, prenatal care was started in the first month of pregnancy in 19.8% of women, in the second in 36.9% and in the third in 19.4%. This data was not available for 442 cases. About the number of prenatal consultations, 32.1% of the women had fewer than 7 prenatal consultations. **Conclusion:** Lumbar SB was the most common in our analysis, despite the significant number of unspecified

SB. Among the maternal factors, live births with SB were more prevalent in single women and women between 8 and 11 years of schooling. Thus, pinpointing a possible association between SB and unplanned pregnancies. Furthermore, 18% of women did not start prenatal care in the first trimester and 32.1% had less than 7 prenatal consultations, that is, ideal prenatal care is still far from being universal in Brazil, which impacts the prevention of neural tube closure defects.

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Epidemiological analysis of strokes in the elderly in the southeast region of brazil: incidence and mortality

Dalva Damasceno Bispo dos Santos[1], Rebeca Leal da Cunha Torres[1], Maria Gabriela Lima Ribeiro Pires[2], Beatriz Claudio[3], Lívia Lucena Nunes[4], Marina Barbosa da Silva[5], Maria Eduarda Pires Albernaz[6], Bruna Gabrielle da Silva[7], Giovanna Carvalho Estefane da Hora[8]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil.
[2]Universidade de São Paulo, Bauru SP, Brazil.
[3]Faculdade Israelita de Ciências da Saúde Albert Einstein, São Paulo SP, Brazil.
[4]Universidade de Pernambuco, Garanhuns PE, Brazil.
[5]Universidade Barão de Mauá, Ribeirão Preto SP, Brazil.
[6]Universidade Federal dos Vales Jequitinhonha e Mucuri, Diamantina MG, Brazil
[7]Universidade Federal de Pernambuco, Caruaru PE, Brazil
[8]Fundação Hector Alejandro Barceló, Buenos Aires, Argentina.

Correspondence: Kayla Dalva Damasceno Bispo dos Santos; Email: kayla.santos@ufpe.br.

Background: A cerebrovascular accident (CVA), commonly known as a stroke, is a sudden event that disrupts cerebral blood flow, leading to brain cell damage. Strokes are classified as either ischemic or hemorrhagic. In Brazil, strokes are the second leading cause of death, with approximately 100,000 fatalities annually according to the Ministry of Health. In 2016, there were 188,223 hospitalizations for stroke treatment (Margarido, 2021). Given this significant health burden, research on stroke is vital for improving screening and health promotion. **Objective:** To describe the epidemiological profile of the elderly affected by strokes in the state of São Paulo (Brazil) between 2018 and 2023. **Methods:** This is a retrospective, quantitative, and descriptive study, with data obtained from the Hospital Information System (SIH/SUS) available on the DATASUS platform. The study covers the period from January 2018 to December 2023 and includes individuals aged 60 years or older. Descriptive statistics were applied to the study population, considering variables such as year of hospitalization, sex, color/race, hospitalizations by state, age group, and mortality rate. **Results:** Between 2018 and 2023, the Southeast region of Brazil reported 311,396 stroke hospitalizations. São Paulo had the highest incidence with 144,657 admissions (46.45%), followed by Minas Gerais with 97,583 (31.33%), Rio de Janeiro with 58,057 (18.64%), and Espírito Santo with 11,099 (3.56%). Stroke incidence by sex showed 52.18% were male and 47.81% female. In terms of color, white individuals accounted for 129,657 (41.64%) hospitalizations, pardos (multiracial) for 100,875 (32.39%), black individuals for 25,979 (8.34%), those of yellow ethnicity for 6,902 (2.22%), and indigenous people for 73 (0.23%). However, 47,910 (15.39%) did not specify their race, impacting the analysis. Regarding age groups, those aged 60-79 years were most affected: 113,566 (36.47%) were aged 60-69 years and 112,382 (36.02%) were aged 70-79 years. The 80-89 years group had 85,448 (27.44%) hospitalizations. The average mortality rate was 16.83 deaths per 1,000 inhabitants. Rio de Janeiro had the highest mortality rate at 21.66 per 1,000 people, followed by São Paulo at 16.68, Espírito Santo at 15.33, and Minas Gerais at 14.34. **Conclusion:** The results indicate that the incidence of stroke is higher in the state of São Paulo compared to other states in the Southeast region of Brazil. Additionally, the mortality rate is higher in men than in women. Based on color/race, white people have a higher stroke rate compared to other races. In terms of age, two groups (60-69 years and 70-79 years) show similar and higher likelihoods of experiencing a stroke compared to the 80-89 years age group. The mortality rate was considered high according to the data studied. Finally, it is worth noting that this work is a descriptive analysis of data, limiting the discussion about the variables studied.

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Epidemiological analysis of the prevalence and mortality of viral meningitis in the southeast region of Brazil

Amábile Cirilo da Silva[1], Samara Brissi Benito da Silva[2], Gabriel Vergolino Eleres[3], Rafaella Ferreira dos Santos[4], Sara Maria Soares Mcgill[5], Hévila Ferreira Gomes Medeiros Braga[6], Danielle de Souza Mometto[7]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[2]Universidade de Mogi das Cruzes, Mogi das Cruzes SP, Brazil.
[3]Fundación Barceló, Buenos Aires, Argentina.
[4]Universidade Santo Amaro, São Paulo SP, Brazil.
[5]Universidade Católica de Pernambuco, Recife PR, Brazil.
[6]Universidade da Integração Internacional da Lusofonia Afro-Brasileira, Redenção CE, Brazil.
[7]Universidade Potiguari, Natal RN, Brazil.

Correspondence: Samara Brissi Benito da Silva; Email: samarabrissi@gmail.com.

Background: Meningitis is defined as inflammation of the tissues surrounding the spinal cord and brain, with bacterial, viral, or aseptic etiologies. The disease is endemic in Brazil and is transmitted through respiratory droplets. Among the known etiologies, viral meningitis is the most common, accounting for approximately 46% of all cases, and showing higher prevalence in the fall, spring, and summer, reflecting the dominance of enterovirus infections such as Coxsackie and Echovirus. To date, there are no studies addressing the prevalence of viral meningitis in southeastern Brazil over the past 10 years, despite it being a significant public health concern. **Objective:** To analyze the epidemiological profile of the prevalence and mortality of viral meningitis in southeastern Brazil from 2013 to 2023. **Methods:** This is a descriptive ecological study of confirmed cases and deaths from viral meningitis in the southeast of Brazil, obtained from the Notifiable Diseases Information System. The study included users of the Unified Health System from January 2013 to May 2023. The following variables were analyzed: age group, deaths, notification region, and state. **Results:** In the period analyzed, the data showed that, in terms of the spatial distribution of the number of cases of viral meningitis, the Southeast region had the highest prevalence and the highest number of deaths among the regions of Brazil, representing 43,055 notified cases (61.8%), with the majority in the population in the aged group of 1 to 4 years, with 11,943 cases (27.7%) and 570 deaths (1.3% of cases) across all age groups. In the Southeast region, São Pulo state stood out with the highest prevalence and deaths, accounting for 37,549 cases (87.2%) and 406 deaths (71.2%). However, as described in the literature, sometimes, because it is a disease with fewer complications and a greater likelihood of progressing to a cure even without intervention, there are many unrecorded cases, especially in areas with difficult geographical access and limited health services coverage. In addition, cases that progress to death may also be underreported when recorded as having an unspecified cause. **Conclusion:** Viral meningitis is prevalent in the Southeast region of Brazil, especially in the state of São Paulo and among preschool aged children. These findings reinforce the need to improve actions to identify and prevent the disease, as well as the importance of including accurate notifications by healthcare. Moreover, it is plausible to make the population aware of the signs and symptoms to ensure that care is sought promptly. These efforts are essential for monitoring the disease´s epidemiological characteristics and implementing effective interventions.

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Epidemiological analysis of traumatic brain injury in the Brazilian unified health system, 2012 to 2022

Ana Clara de Lemos Guimarães[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Ana Clara de Lemos Guimarães; Email: anaguimaraes21.1@bahiana.edu.br.

Background: Traumatic brain injury (TBI) is the leading cause of death and disability in adults, primarily affecting men and individuals under 45 years of age. Its high cost to the Brazilian Unified Health System (SUS) and its high incidence and morbidity and mortality rates, coupled with a lack of epidemiological studies, make it urgent to develop demographic analyses to support the planning of interventions. **Objective:** To define the epidemiological profile of TBI hospitalizations in Brazil from January 2012 to December 2022. To determine the temporal trend of hospitalization and annual mortality rates; describe the spatial distribution by region and determine the lethality of TBI. **Methods:** This is an epidemiological, descriptive, retrospective, and quantitative study of TBI hospitalizations in Brazil. Data were obtained from the Hospital Information System (SIH) of the Department of Informatics of the Brazilian Unified Health System (DATASUS). The variables analyzed were: biological sex, age group, geographical region, total cost, and average length of hospital stay. Descriptive statistics were used to analyze the variables, which were presented as raw numbers, hospitalization, mortality, and lethality rates. **Results:** Between 2012 and 2022, there were 1,466,716 TBI-related hospitalizations in Brazil, with 77.31% being men and 22.68% women. The overall lethality of TBI was 7.57%; the total cost was R$2,514,910,263.00, and the average length of hospital stay was 5.9 days. The hospitalization rate showed a decreasing and statistically significant trend (R^2=0.357 β=-0.587 p=0.04). The South region had the highest hospitalization rates. Men had a hospitalization rate 3.53 times higher than women, and individuals aged 80 and over had the highest rates. The mortality rate was higher among men (average of 7.93 per 100,000 inhabitants) compared to women (average of 1.93 per 100,000 inhabitants) and higher in the age group of 80 and over (average of 32.37 per 100,000 inhabitants). Lethality was higher among men (7.9%) compared to women (6.49%), with a lethality ratio of 1.22 (95% CI: 1.20-1.23). Patients aged 80 and over had the highest lethality (19.23%), while the lowest lethality was among those under one year to nine years old. The South region had the lowest lethality rate (5.56%) and the Southeast region the highest (8.47%). **Conclusion:** The data indicate a high prevalence of TBI among men and increasing lethality with age, especially in individuals over 80 years old. There is a decreasing trend in hospitalization rates, but the financial and social impact of TBI remains significant. Regional analysis shows variations in lethality rates, with the Southeast region presenting greater challenges. These results highlight the need for health policies aimed at TBI prevention and improved care for more vulnerable groups, especially men and the elderly.

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Epidemiological analysis of traumatic brain injury in the Northern region of Brazil from 2019 to 2023

Barbara Zorzi Sanfins[1] , Lucas Charuri de Andrade Castello Branco[1], Lucas Pozza Gomes[2], Ana Carolina Bizetto[2], Lucas Valério Abrahim[2], Fernanda Araujo Kavlac[3], Lucas David de Souza Vital[3], Fábio de Melo Franzoni[2], Maria Fernanda Baptista Costa Monteiro[2], Francisca Isabelly Leite de Almeida[3]

[1]Faculdade São Leopoldo Mandic, Campinas SP, Brazil.
[2]Universidade Cesumar, Maringá PR, Brazil.
[3]Universidade Federal do Amazonas, Manaus AM, Brazil.
[4]Centro Universitário Unifacisa, Campina Grande PB, Brazil.

Correspondence: Barbara Zorzi Sanfins; Email: basanfins@gmail.com.

Background: Traumatic Brain Injury (TBI) is a leading cause of death and disability among young adults, accounting for approximately 2.5 million deaths and 60 million disability cases annually. Patients with TBI may experience cognitive, motor, behavioral, and social functionality impairments. The most common causes include traffic accidents, falls, and assaults. Consequently, TBI is a significant topic of discussion today, as this health issue primarily affects the active age group of the population. **Objective:** To identify the epidemiological profile of reported Traumatic Brain Injury cases in the Northern region of Brazil between 2019 and 2023. **Methods:** An exploratory, descriptive, epidemiological, time-series study was conducted on traumatic brain injury in the Northern region of Brazil between 2019 and 2023. Data were obtained from the Department of Information of the Brazilian Unified Health System (SUS) through the Hospital Information System of SUS, including cases of hospital admissions and deaths due to TBI. The variables studied included hospital admissions, deaths, age group, and gender. The information was tabulated in Microsoft Office Excel® and presented in charts and tables for better analysis. The database used is open for public access without the need for approval from the Research Ethics Committee, and accessible to the public and interested researchers. **Results:** The study identified a total of 41,129 hospitalizations due to TBI in the Northern region between 2019 and 2023, along with 3,367 deaths, this translates to an overall mortality rate of approximately 8.2%. Pará recorded the highest number of hospitalizations (55.5%) and deaths (51.9%). Conversely, Acre registered the fewest deaths, with a total of 89 cases. Additionally, three states showed a reduction in TBI-related deaths over the years: Acre, Amazonas, and Roraima, with decreases of 31.6%, 14%, and 26.5%, respectively. In terms of gender, there was a higher prevalence of males in both hospitalizations (76.5%) and deaths (80.7%). Thus, the mortality rate among men was 8.6%, while for women, it was 6.7%. The age group with the highest number of hospitalizations and deaths was 20 to 29 years, accounting for 21.3% of total hospitalizations and 18.7% of deaths. Moreover, the highest mortality rate was observed in the age group of 80 years or older, at 20.3%, while the lowest was in the 1 to 4 years age group, at 1.08%. **Conclusion:** As evidenced, TBI is a significant cause of hospitalizations and deaths in the Northern region, constituting a serious public health issue. Despite the increase in the number of traumatic brain injury cases in the region, there was a reduction in the number of deaths in three states, indicating the potential for improvement in the current alarming scenario.

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Epidemiological analysis of zika virus infections associated with cases of microcephaly in the Brazilian regions between 2015 and 2023

Mariana Teixeira Mesquita[1], Celijane Almeida Silva[2], Luiza Bisognin Marchesan[3], Mylena Cordeiro Aranha[4], Amanda Fortes Cavalcante de Oliveira[5], Isadora Carvalho de Sousa[4], Laiane Rodrigues Macêdo[6], Luana Dutra Pinheiro da Silva[4], Lara Maia Pereira[4], Bárbara Pinheiro Gama de Andrade[4]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Centro Universitário Unifacisa, Campina Grande PB, Brazil.
[3]Universidade Franciscana, Santa Maria RS, Brazil.
[4]Universidade Potiguar, Natal RN, Brazil.
[5]Universidade Federal do Delta do Parnaíba, PI, Brazil.
[6]Universidade Nove de Julho, São Paulo SP, Brazil

Correspondence: Mariana Teixeira Mesquita; Email: maritmesquita@gmail.com.

Background: Zika virus (ZIKV) represents a significant threat to public health due to its correlation with microcephaly in newborns when infecting pregnant women. Therefore, epidemiological studies of this association across diverse populations are crucial for effectively directing public resources and guiding the development of appropriate therapeutic and preventive measures. **Objective:** To identify and analyze the epidemiological profile of Zika virus infections associated with cases of microcephaly in regions of Brazil, from 2015 to 2023. **Methods:** This is an ecological observational epidemiological study with a descriptive quantitative approach, which relied on analyzed data from the Department of Informatics of the Unified Health System (DATASUS). The research was conducted using data from Notification of Suspected Cases of CZS (Congenital Syndrome associated with Zika virus infection) registered from 2015 to 2023. The parameters of "laboratory diagnosis of zika in pregnant women: yes" and "congenital abnormalities detected: microcephaly only, microcephaly with CNS alteration, and microcephaly with other congenital abnormalities" were selected. The variables considered were: year of notification and regions of Brazil. Finally, the collected data were organized and systematized in Microsoft Excel, and analyzed using descriptive statistics. **Results:** The years 2015 and 2016 saw the highest number of notifications of pregnant women diagnosed with Zika in association with newborns with congenital microcephaly, with 541 cases, representing 65.2% of the total. Subsequent years showed a decreasing trend in the association of cases across all regions of Brazil, with 2023 having the lowest frequency of records, with 20 cases (2.4%). The most affected regions were the Northeast with 396 cases (47.7%) and the Southeast with 278 cases (33.5%) in all analyzed years, with the peak of records in the Northeast occurring in 2015, with 189 cases, and in the Southeast occurring in 2016 with 149 cases. The Southern region was the least affected, with 13 cases (1.6%) during the period, followed by the Northern region with 57 cases (6.9%) and the Central-West region with 86 cases (10.4%). Despite significant regional variability, there was a general downward trend in reported cases over the years. **Conclusion:** Following the outbreak of Congenital Syndrome associated with Zika virus infection cases in 2015 - 2016, there was a significant decrease in notifications across all regions of Brazil, which may be related to increased awareness and disease prevention regarding Zika virus infection; development of herd immunity; advancements in epidemiological surveillance and case control; successful vector control measures, among others. Therefore , it is important to highlight the role of epidemiological data analysis in shaping public policies and improving the health condition of affected populations.

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Epidemiological and clinical characteristics of patients with Guillain-Barré syndrome in a hospital in the Northeast of Brazil after pandemia: preliminary results

Jesângeli de Sousa Dias[1,2], Leonardo Sousa de Jesus[2], Jessica Abreu de Azevedo Nascimento[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Secretaria de Saúde do Estado da Bahia, Instituto Couto Maia, Salvador BA, Brazil.

Correspondence: Jesângeli de Sousa Dias; Email: jesangelidias@gmail.com.

Background: The Guillain Barré syndrome (GBS) is the leading cause of flaccid paralysis in the world. In general, an infection precedes the disease in most cases. In Brazil, as in other countries, there was an increase in the number of GBS cases after the introduction of the Zika and Chikungunya viruses. Furthermore, cases associated with COVID-19 have been described and we do not know whether this virus may have led to increased GBS cases in our country. **Objective:** Investigate the influence of the pandemic on the prevalence and clinical-epidemiological characteristics of GBS cases admitted to a hospital in the Northeast of Brazil. **Methods:** This retrospective observational study is based on a review of medical records of individuals consecutively hospitalized from January 1, 2021, to December 31, 2023, with ICD 10 G61.0. We included cases defined according to the Brighton criteria and evaluated at least once by a neurologist. Records with insufficient data were excluded. Data were described as median and interquartile range or absolute and relative frequency as appropriate. **Results:** 33 medical records have been evaluated so far. Most participants came from the capital and metropolitan region (60.6%), 67% were male, and the median age was 28 (IQR: 21.5 - c 45) years. A history of previous infection was present in 60.6% of cases (dengue, 15.1%; diarrhea, 12.1%; respiratory infection with diarrhea, 12.1%; respiratory infection, 9.1%; undefined infection 9,1%). The time after infection and onset of symptoms was 15 (IQR: 7 - 25) days. History of vaccine use was present in 15.1%. All participants had some degree of muscle weakness with reduced or absent reflexes. The most frequent neurological manifestations were ascending muscle weakness (72.7%), mild to moderate sensory symptoms (66.6%), unilateral or bilateral facial paralysis (51.5%), dysautonomia (45.4%), dysphagia (39.4%) and need for mechanical ventilation in 15.1%. The length of stay was 18 (IQR: 10 - 40) days. CSF study showed 2 (IQR:1.25 - 2.0) cells/mm3 and 96 (IQR: 80.25 - 189.25) mg/100mL of proteins. Positive serology for arbovirus was found in 15 participants (IgM Zika, 3; IgM Chikungunya 7; IgM Dengue 5). The SARS-CoV-2 was identified in 3 participants. However, these infectious agents were not screened for in all participants. Twenty-six participants were treated with intravenous immunoglobulin. There were no deaths and at the time of discharge, 45.5% of individuals were confined to a bed or in a wheelchair. **Conclusion:** In our country, it seems that arboviruses continue to be the infectious agents most frequently associated with GBS cases. These results are in line with previous reports. These numbers may be an underestimate as not all participants were tested. We hope that the evaluation of the remaining cases will provide us with additional information. We must be aware that cases of Guillain Barre are expected to increase, following the increase in cases of arboviruses.

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Epidemiological and clinical features of patients admitted for stroke in an emergency department in the interior of Pernambuco

Joan Lins Serafim[1], Maria Vitória do Nascimento Silva[1], Maria Laura Lima da Silva[1], Jéssica dos Santos Coelho[1], Thiago Bezerra dos Anjos[1], Ana Luiza Sampaio Lopes[1], Américo Danúzio Pereira de Oliveira[1], Rita Di Cassia Oliveira Angelo[2]

[1]Universidade de Pernambuco, Serra Talhada PE, Brazil.
[2]Universidade de Pernambuco, Garanhuns PE, Brazil.

Correspondence: Maria Vitória do Nascimento Silva; Email: vitoria.nsilva@upe.br.

Background: Stroke is characterized by sudden neurological deficit with a national incidence of 108 cases per 100,000 persons and a crude mortality rate of 62 cases per 100,000 persons. As the leading cause of global mortality, it is necessary to establish primary prophylactic measures by altering modifiable risk factors and conducting etiological investigations to define secondary prophylaxis, especially after determining the epidemiological profile. **Objective:** To describe the epidemiological profile of patients admitted to an emergency service of the Pernambuco. **Methods:** This observational, descriptive, and cross-sectional study was conducted at the Professor Agamenon Magalhães Regional Hospital, a reference hospital for emergencies in the hinterlands of Pernambuco. The population included cases with a clinical-radiological diagnosis of stroke admitted to the service, and the sample was consecutive. After diagnostic confirmation, patients were clinically evaluated and subjected to the Self- Reported Health Conditions/Situations Questionnaire, TOAST Classification (Trial of ORG 10172 in Acute Stroke Treatment), and the Stroke Prophylactic Measures Questionnaire. **Results:** The sample consisted of 194 participants, of which 92.8% had ischemic stroke and 7.2% had hemorrhagic stroke. The majority (76.8%) were over 60 years old, predominantly male (54.6%) and mixed-race (49.5%). Regarding occupational status, most had previously worked as farmers (46.9%) and were currently retired (71.1%). In terms of education level, most of the sample was uneducated (38.7%). Due to the lack of care services in the region of origin, 35.6% of the patients came from 22 municipalities adjacent to the referral hospital. Concerning comorbidities, 81.4% had a diagnosis of systemic arterial hypertension, 50% of diabetes mellitus, 32.5% had a previous stroke, 10.3% heart failure, 8.2% acute myocardial infarction, 3.6% had a transient ischemic attack, 5.7% had chronic kidney disease, 4.1% malignant neoplasia, and 12.4% were bedridden. Regarding lifestyle habits, smoking and alcohol consumption were noted in 28.4% and 19.6% of patients, respectively. Furthermore, 87.6% were sedentary, and 79.3% did not follow a specific dietary guideline. In terms of prophylactic measures, only 39.7% of patients used some prior screening method for diabetes mellitus, only 30.4% underwent any screening test for dyslipidemia and 10.3% practiced at least 4 hours of physical activity per week. As for the etiology of stroke, about 58% were of indeterminate cause; 16% from large vessels; 10% from small vessels; and 8% cardioembolic. **Conclusion:** There is a need to implement an etiological investigation program for cases admitted to units in the northeastern interior, aiming at promoting secondary prophylaxis. Furthermore, this research advises investing in the control of modifiable risk factors, which are common in the evaluated population.

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Epidemiological and clinical profile of patients with Guillain-Barré syndrome at a university hospital in Mato Grosso do sul, from 2018 to 2023

Isabela de Carvalho Florencio[1], Thiago Dias Fernandes[1]

[1]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.

Correspondence: Isabela de Carvalho Florencio; Email: isabelacflorencio@hotmail.com.

Background: Guillain-Barré syndrome (GBS) stands as the predominant cause of acute flaccid paralysis on a global scale. It is characterized as an acute or subacute inflammatory polyradiculoneuropathy, resulting in progressive and symmetrical weakness, areflexia, often accompanied by sensory symptoms and autonomic manifestations. GBS is most associated with infections, vaccinations, malignancies, and even trauma. Diagnosis is clinical, but ancillary tests such as cerebrospinal fluid analysis and electromyography are useful for differential diagnosis. Early diagnosis and treatment significantly impact the disease's course, affecting morbidity and mortality. **Objective:** Describe the clinical and epidemiological profile of patients treated with GBS at a tertiary-level public hospital in Campo Grande from 2018 to 2023. **Methods:** Cross-sectional, descriptive study, based on data from medical records of patients with GBS, treated at the University Hospital in the capital of Mato Grosso do Sul, between January 2018 and February 2023. **Results:** During the study period, twenty-four patients were diagnosed with GBS. The mean age was 41.9 years. Most of the patients were male (69.6%). The incidence of cases was higher in summer (43.5%) and at its nadir was in spring (13.0%). Trigger events were identifiable in 68.2% patients, with 33.6% of arboviruses, 20% gastroenteritis, 40% influenza-like syndromes, and 6.3% trauma. The classic form of ascending tetraparesis was observed in 60.9% of patients, while 17.4% had the cervicobrachial pharyngeal form, and 8.7% presented an overlap of the cervicobrachial pharyngeal form and Bickerstaff's encephalitis. Albumin-cytological dissociation of CSF was detected in fourteen patients, with a mean protein value of 113.3 mg/dl. Twenty patients received immunoglobulin treatment, and two patients underwent two treatment cycles. The average hospital stay was 30.2 days, with 26.1% of patients requiring ICU and mechanical ventilation, with an average of 83.2 days in the hospital. The mean duration of hospitalization was 8 days for the classic form and 82 days for the cervicobrachial pharyngeal form, showing a statistically significant difference (p-value 0.004). Clinical complications were observed in 83.3% of patients with the cervicobrachial pharyngeal form compared to 7.1% of the classic form and 33% of other forms. **Conclusion:** The data from this study are in accordance with the literature and contributes to outlining an epidemiological profile of the GBS population in Mato Grosso do Sul.

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Epidemiological and healthcare indicators analysis of the stroke line of care within the emergency and urgency healthcare network

Gustavo Alves Cangussu[1], Giovana Andrade de Oliveira[1], Gustavo de Souza Campos[1], Matheus Nascimento Moura[2], Victor Santos Vilares[1], Maria Fernanda Aragão Andrade[1], Emmanuel Santos Trindade[1], Priscila Santos Oliveira[1], Philip George Glass Andrade[1], Davi Tanajura Costa[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.
[2]Universidade Federal da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Gustavo Alves Cangussú; Email: canguss.gustavo@gmail.com.

Background: Stroke is a multifactorial and complex disease that gains significance in the medical field due to its high incidence, mortality, and morbidity rates. In Brazil, Ordinance Nº. 665 was published on April 12, 2012, aiming to establish qualification criteria for establishments as Urgency Care Centers for Stroke Patients, to institute financial incentives and to approve the Stroke Line of Care within the Emergency and Urgency Healthcare Network (RUE) of the Unified Health System (SUS). **Objective:** To analyze the epidemiological and healthcare indicators of the Stroke Line of Care between 2012 and 2021. **Methods:** This is a cross-sectional study. Data from the Mortality Information System (SIM) were used to determine the mortality rate from cerebrovascular diseases. A joinpoint regression model was used to determine the trend in the mortality rate. Data from the National Registry of Healthcare Establishments (CNES) were used to determine the number of establishments qualified as Urgency Care Centers for Stroke Patients. Data from the Hospital Information System (SIH) were used to determine the percentage of patients who underwent thrombolytic treatment. **Results:** The average annual mortality rate from cerebrovascular diseases was 49.08 deaths/100,000 inhabitants in Brazil between 2012 and 2021. It was noticed a decreasing trend in mortality rate (AAPC: -0.68 [-1.03; -0.32]; p: 0.002). This trend was followed by the Northeast (AAPC: -0.52 [-0.99; -0.04]; p: 0.04), Southeast (AAPC: -0.78 [-1.19; -0.37]; p: 0.002) and South (AAPC: -1.27 [-2.03; -0.52]; p: 0.005) regions. The Midwest region, however, presented a stationary trend (AAPC: -0.05 [-0.37; 0.28]; p: 0.75). Only the North region showed an increasing trend (AAPC: 0.97 [0.26; 1.69]; p: 0.008). The North region was also the only one to present a joinpoint, dividing into a first period with an increasing trend between 2012 and 2014 (APC: 3.95 [0.07; 7.98]; p: 0.047) and a second with a stationary trend (APC: 0.14 [-0.37; 0.65]; p: 0.52). 110 Urgency Care Centers for Stroke Patients were identified. Among them, 16 were qualified as Type I, 30 as Type II, and 64 as Type III. 10 Federative Units did not have qualified establishments: Rondônia, Acre, Amazonas, Roraima, Amapá, Tocantins, Piauí, Rio Grande do Norte, Mato Grosso and Goiás. Only 1.14% of stroke cases underwent chemical thrombolysis. No Federative Unit reached the quality target of 20%. The best thrombolysis ratios were in Ceará (8.22%), Distrito Federal (4.46%) and Rio Grande do Sul (2.91%). **Conclusion:** A reduction in cerebrovascular diseases mortality has been observed, however a small proportion of patients treated are subjected to thrombolytic treatment. Despite a decade have passed since the institution of the Stroke Line of Care within the Emergency and Urgency Healthcare Network, there is still a deficiency in the distribution of Urgency Care Centers for Stroke Patients in Brazil.

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Epidemiological aspects of bacterial and viral meningitis in Brazil: a retrospective analysis from 2018 to 2023

Henrique Kallás Ribeiro do Valle[1], José Luiz Mendes Erthal Alves[2], Milene Alves de Souza Almeida[3], José Alexandre Bachur[4,5]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[2]Faculdade de Medicina de Petrópolis, Petrópolis RJ, Brazil.
[3]Universidade Estadual de Mato Grosso do Sul, Durados MS, Brazil.
[4]Universidade de Franca, Franca SP, Brazil.
[5]Instituto Projete Ciências.

Correspondence: Henrique Kallás Ribeiro do Valle; Email: henrique.krv@icloud.com.

Background: Meningitis is defined as the inflammation of the membranes surrounding the brain and the spinal cord. Its etiologies that demand the most attention are: viral (VM) and bacterial (BM). **Objective:** The study was carried out with the objectives of identifying data on hospitalizations and deaths due to BM and VM at national and regional levels, and evaluating the respective incidence, mortality and lethality rates. **Methods:** Quantitative and retrospective ecological study, based on public data collected from the System of Notifiable Diseases Information (SINAN), regarding hospitalizations and deaths due to BM and VM occurring in Brazil and the federative regions in the period from 2018 to 2023, available in the DATASUS system. The absolute and relative values were recorded in an electronic spreadsheet and the statistical significance between the means was evaluated based on the p value <0.05. **Results:** In the period from 2018 to 2023 in Brazil, The number of hospitalizations due to BM (21,190) was significantly higher (p=0.002) than those due to VM (12,768). Similarly, the number of deaths due to BM (2,052) was significantly higher (p=0.0002) than those due to VM (481). This quantitative superiority of BM in relation to VM is reflected

at national and regional levels in the rates of: incidence (BM - 10.43 / VM - 6.29), mortality (BM - 1.01 / VM - 6.29) and fatality (BM - 0.10 / VM - 0.04). There was no significant difference (p≥0.05) between the number of cases in men (57%) in relation to women (43%), regarding hospitalizations or deaths. Data on hospitalizations and deaths due to BM or VM reported in the Southeast region were significantly higher (p<0.05) in relation to other regions, followed initially by the Northeast or South regions, which presented values without significant differences between them, but significantly higher in relation to the North or Central-West regions, which presented statistically equal numbers of notifications. However, there is a variation in classification between regions in relation to the highest and lowest values relating to incidence (IR), mortality (MR) or fatality (FR) rates. The South presented the highest IR for both BM (14.46) and VM (8.66); and the Northeast, the lowest IR for BM (8.41); and the North, for VM (4.26). While the highest MR of BM (1.27) and VM (0.35) were also in the South, and the lowest in the Central-West (0.72 and 0.17). The Southeast, North and Northeast regions had the highest FR per BM (0.10) and the Central-West, the lowest (0.08). While in the North there was the highest FR per VM (0.05) and in the Northeast, the lowest (0.03). **Conclusion:** The orders of magnitude of occurrence of BM and VM associated with the respective risks to survival, both in men and women, highlight a situation of health problems and suggest the need to implement preventive public policies in all federative regions of the country, with greater emphasis on bacterial meningitis.

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Epidemiological aspects of hospital admissions for Alzheimer's in Brazil from 2015 to 2019

Ana Carolina Ribeiro de Araujo e Araujo[1], Aeriberto Carlos Lindoso de Souza[1], Consuelo Penha Castro Marques[1], José Carlos Gomes Patriota Neto[1], Tassya Jordana Coqueiro Batalha[1]

[1]Universidade Federal do Maranhão, Pinheiro MA, Brazil.

Correspondence: Ana Carolina Ribeiro de Araujo e Araujo; Email: anacarolinaraa@hotmail.com.

Background: Alzheimer's disease is a neurodegenerative dementia disease resulting from multiple pathophysiological factors, implying a progressive decline in the patient's logical and behavioral capabilities resulting from the manipulation of cholinergic neurons, causing fundamental losses that include behavioral and cognitive impairment, that is, the memory, language, the ability to understand and logical reasoning. It very commonly affects elderly people, resulting in several serious health complications, causing the entire society and the health system to be directly affected. **Objective:** To analyze the epidemiological profile of Alzheimer's disease hospitalizations in Brazil. **Methods:** An exploratory, descriptive, epidemiological, time series study was carried out from 2015 to 2019, with secondary data from DATASUS - Ministry of Health, Brazil. **Results:** During the period under study, there were n=7833 hospitalizations due to Alzheimer's in Brazil, from 2015 to 2019, with a highest peak in 2015, a lower number of hospitalizations in 2016 and an oscillating trend until 2019 where there was a large increase, making an average = 1566 .6 (±1823.93) and Coefficient of Variation (CV)=116.43%. The Southeast had the highest percentage of hospitalizations with 59% and the South 22%. Females accounted for 65% of hospitalizations, the most prevalent age group was 70 to 80 years old and + with 87%, white people were more frequent with 46.68%. The total amount spent was R$ 13,459,451.43, the Southeast was responsible for 77% of this amount spent. The average stay was 22.6 days. The number of deaths (n=1461) and the mortality rate of 18.65. **Conclusion:** Given this, considering the increase in the number of hospitalizations and deaths at a national level, corroborated by the prospect of an increase in the number of elderly people in the coming years, it is of fundamental importance that municipal, state and federal health managers turn their attention to the adoption of public policies that cover this population, guaranteeing, in addition to greater longevity, a better quality of life in terms of physical, mental and psychological aspects.

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Epidemiological aspects of patients affected by stroke in the Southeast region between Jan/2018 to Jan/2024

Lia Mayra Miranda Santos[1], Teofilo Dorneles Claro dos Santos Silva[2], Riadylla Pitzr Fonseca Guimarães[3], Ana Laura Fonteles Santos[4] , Letícia Gava[5], Danila Ferreira Silva[6]

[1]Universidade Anhembi Morumbi, São Paulo SP, Brazil.
[2]Universidade Federal do Maranhão, São Luís MA, Brazil.
[3]Centro Universitário de Ciências Gerenciais de Manhuaçu e Região, Manhuaçu MG, Brazil.
[4]Centro Universitário do Estado do Pará, Belém PA, Brazil.
[5]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.
[6]Centro Universitário de Excelência.

Correspondence: Lia Mayra Miranda Santos; Email: lia13mayra@gmail.com.

Background: The definition of stroke can be described as the presence of a neurological deficit of focal vascular etiology and abrupt onset, with a variable clinical picture. Stroke has a prominent role in the group due to its high incidence, given its position as the second leading cause of death in the world and the estimated increase in cases due to the advancement of population life expectancy. **Objective:** This study aims to analyze and describe the epidemiological profile of stroke in Southeast Brazil between 2018 and 2024. **Methods:** This is a cross-sectional, descriptive study with a quantitative approach, obtained by extracting data from DATASUS on the states of the Southeast region of Brazil between January 2018 and January 2024. **Results:** The study revealed a total of 443,529 hospitalizations for stroke in the Southeast region between 2018 and 2024, as well as a total of 67,425 deaths (15.2%). Minas Gerais recorded the highest number of hospitalizations, considering the total number of inhabitants, with a total of 137,087, while second place (Rio de Janeiro) recorded 82,808 hospitalizations. The state with the lowest record was Espírito Santo, which had 15,222 hospitalizations. As for the number of deaths, Rio de Janeiro recorded the highest number (16,305), followed by Minas Gerais (18,017) and São Paulo (30,976). In terms of gender, the majority of hospitalizations for strokes were of male victims (52.5%). Males also accounted for 51.2% of deaths in the period analyzed, with the highest mortality rate of all the states in the region. It can be seen that the number of hospitalizations increases sharply as the population ages. It can also be seen that strokes occur more frequently between the ages of 60 and 79 (52.5%). Furthermore, this disease is more lethal in the elderly aged 80 and over, given the higher mortality rate in this age group (29.3%). When color/race is analyzed, it is possible to observe a prevalence of cases in the white population in the Southeast region, which registered 176,464 cases of hospitalizations. The state of São Paulo led the way, both in terms of hospitalizations, which alone accounted for 114,703 cases (25.8%), and in terms of the number of deaths (16,982), which represents 25% of total deaths. The yellow population comes next with 150,995 hospitalizations and 22,326 deaths in all regions. It is important to note that of the total hospitalizations, 67,366 were not recorded according to color/ethnicity. **Conclusion:** As a result, it was concluded that, as the disease is one of the main causes of death in Brazil and its epidemiological profile affects a more debilitated population, characterized by elderly individuals, it is of the utmost importance to disseminate knowledge about stroke, with a view to prevention and early identification of cases through the development of preventive and structural strategies that can contribute to reducing the frequency of patients affected by the disease.

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Epidemiological aspects of patients affected by viral encephalitis in the Southeast between 2019 and 2023

Lia Mayra Miranda Santos[1], Sara Raquel Ribeiro Pires[2], Ivana Louise da Silva Martins[2], Larissa Andrade Ramos[3], Ana Camile de Freitas[4]

[1]Universidade Anhembi Morumbi, São Paulo SP, Brazil.
[2]Universidade Federal do Maranhão, São Luís MA, Brazil.
[3]Universidade Federal dos Vales do Jequitinhonha e Mucuri, Diamantina MG, Brazil.
[4]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Lia Mayra Miranda Santos; Email: lia13mayra@gmail.com.

Background: Encephalitis is defined as inflammation in the tissues of the nervous system and one of the most common etiologies is due to viral infectious agents. Bringing together a varied series of conditions, it most often presents with direct involvement of the brain parenchyma demonstrated by a lowered level of consciousness, seizures and/or focal neurological deficits. Being potentially fatal diseases, with associated sequelae and variable transmissibility, epidemiological understanding is of vital importance for effective management and reducing the impact on public health. **Objective:** To analyze the epidemiological profile of patients affected by viral encephalitis in the Southeast region between 2019 and 2023. **Methods:** This is a descriptive, cross-sectional epidemiological study that analyzed the epidemiological profile of patients affected by viral encephalitis in the Southeast Region of Brazil between 2019 and 2023. Data on hospitalizations, deaths and mortality rates were provided by the Department of Informatics of the Unified Health System (DATASUS) and were analyzed according to the variables: age group, gender, year and color/race. **Results:** There were 2,530 cases of people hospitalized for viral encephalitis in the Southeast between 2019 and 2023 and 145 deaths, resulting in a mortality rate of 0.03 per 100,000 inhabitants. Of the hospitalizations, 27.9% were children aged 0 to 4, with 8.4 deaths per 1,000 hospitalizations in this age group in the Southeast; however, there were more deaths in adults, with a peak in the 30 to 39 age group (22). With regard to the sex of the patients, there was a predominance of males in both the total number of hospitalizations and the total number of deaths. This trend was maintained in all states, with the exception of São Paulo, which had more female deaths in the period analyzed (41). As for the race/color variable, there was a higher number of hospitalizations and deaths among individuals identified as white and brown, totaling 76.2% of all hospitalizations and 73.8% of deaths. In relation to the number of total hospitalization records, the year 2019 showed the highest rate of hospitalizations in the years analyzed (586), however the highest number of deaths was seen in the year 2023 with 34 deaths. **Conclusion:** In this way, it can be inferred that viral encephalitis has low mortality associated with the number of cases, it is also understood the epidemiological profile of involvement that refers, above all, to hospitalizations for white and brown individuals, and males, it is noteworthy, however, the rate of hospitalizations for the child population, which denotes the need to evaluate this population in order to be able to screen for this condition.

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Epidemiological aspects of pediatric patients affected by viral meningitis in the Southeast region between 2019 and 2023

Lia Mayra Miranda Santos[1], Lorena Carvalho Plens[2], Aryane Agatha Rocha da Silva[3], Cleiton da Silva Santos[3], Victoria Verissimo Santos[4]

[1]Universidade Anhembi Morumbi, São Paulo SP, Brazil.
[2]Universidade Federal do Paraná, Toledo PR, Brazil.
[3]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.
[4]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Lia Mayra Miranda Santos; Email: lia13mayra@gmail.com.

Background: Meningitis is an infection caused by viruses, fungi, or bacteria, affecting the meninges region. The majority of meningitis infections are viral, with enterovirus being the most common in the pediatric population. Children usually present with fever, vomiting, anorexia, respiratory symptoms, and conditions similar to viral infections, with younger children often having a highly nonspecific presentation. Therefore, understanding the epidemiological profile of different regions in Brazil helps establish a more precise clinical approach in medical practice. **Objective:** Increase knowledge about the epidemiological profile of pediatric patients affected by viral meningitis in the Southeast region of Brazil between 2019 to 2023. **Methods:** This is a cross-sectional, descriptive study with a quantitative approach based on epidemiological data provided by the Ministry of Health in the "TABNET" data tabulation tool of the Department of Informatics of the Unified Health System (DATASUS). The data was collected from the SUS Hospital Information System (SIH/SUS), by place of hospitalization, in which viral meningitis was selected from the ICD-10 list of morbidities. For the collection, filters were applied for the period from January 2019 to December 2023, the age ranges up to 19 years and the location was restricted to the southeast region, which covers the states of Minas Gerais, Espírito Santo, Rio de Janeiro and São Paulo. In addition, the division by color/race was also inserted. **Results:** The study revealed a total of 2,723 hospitalizations for viral meningitis in the Southeast region between 2019 and 2023, as well as a total of 38 deaths among individuals up to the age of 19. Regarding the number of total hospitalization records, the year 2019 showed the highest rate of hospitalizations, reaching 31.3% of the 2723 recorded in the database during the period analyzed. Among the states, hospitalizations in São Paulo reached 71.9% of the total number of hospitalizations recorded. When comparing the records according to color/race and taking out the individuals not classified as black, white, brown, or yellow (2,304), we observed a greater number of records for white people, totaling 53.3% of the sample. There was a predominance of males among those hospitalized for viral meningitis in the total number of records both in the Southeast and in each state separately. The most affected age group was children under the age of 1, with a total of 922 records over the period analyzed. **Conclusion:** Pediatric meningitis is a public health concern due to its potentially serious nature. Epidemiological surveillance is crucial in contributing to accurate diagnoses and effective interventions, intending to minimize the sequelae and fatal outcomes associated with the disease. Thus, by producing up-to-date scientific information, medical practice can be based on the best scientific evidence available, and, as a result, public policies can become increasingly efficient.

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Epidemiological assessment of microcephaly in Brazil from 2015 to 2023

Roberta Leandrini Rossato[1] , Letícia Loreta Roque Stetter[2], Marcos Henryque Costa Soares[3] , André Oliveira Kiryu[4] , Evelyn Lorrany Oliveira Dantas[5] , Iuri Moraes Leite[6] , João Victor Zucolotto Scottá[7] , Thamires Coutinho Aguiar[8], Vitória Monteiro Marciano[9] , Catherine Yurie Minasse[10]

[1]Universidade de Mogi das Cruzes, Mogi das Cruzes SP, Brazil.
[2]Fundación Héctor Alejandro Barceló, Buenos Aires, Argentina.
[3]Centro Universitário FAMETRO, Manaus AM, Brazil.
[4]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[5]Centro Universitário Facisa, Campina Grande PB, Brazil.
[6]Universidade Estadual de Roraima, Boa Vista RO, Brazil.
[7]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[8]Universidade de Rio Verde, Formosa GO, Brazil.
[9]Centro Universitário de Valença, Valença RJ, Brazil.
[10]Universidade Cesumar, Maringá PR, Brazil.

Correspondence: Roberta Leandrini Rossato; Email: roberta.rossato@hotmail.com.

Background: Microcephaly is a congenital disease characterized by a head circumference less than two standard deviations from the mean for the gestational age and sex of the newborn. In severe cases, the head circumference is less than three standard deviations. Children with microcephaly may experience delays in neurodevelopment and sensory deficits in multiple systems, such as vision and hearing, depending on the severity of the condition. The causes of this condition are multifactorial, it has several causes, from genetic to infectious, such as Zika virus, STORCH, malnutrition, herpes, toxoplasmosis, cytomegalovirus, rubella, among others. Between 2025 and 2017, Brazil faced a Zika vírus epidemic, which resulted in a significant increase in microcephaly cases, followed by a sharp drop. Currently, little is discussed about microcephaly, which motivated this study. **Objective:** This work aims to provide a brief epidemiological analysis of microcephaly in Brazil from 2015 to 2023. **Methods:** This is a descriptive epidemiological study based on secondary data analysis. Epidemiological data obtained from the Brazilian Ministry of Health and DATASUS were used. The analysis covered the period from 2015 to 2023, focusing on the incidence of reported microcephaly cases. The data was organized and analyzed to identify trends and variations over the years. **Results:** Assessment of the incidence of microcephaly during the study period revealed an epidemic in 2015 (4.135 cases), with an increase of 107.69% in 2016 (8.588 cases), followed by a decrease of 69.04% in 2017 (2.658 cases). The downward trend continued in 2018 (1.733 cases, down 34.80%), 2019 (1.514 cases, down 12.63%), 2020 (1.029 cases, down 32.03%), 2021 (810 cases, drop of 21.28%) and 2022 (749 cases, drop of 7.53%). However, in 2023, there was an increase of 38.18% (1.035 cases). **Conclusion:** Data analysis indicates a strong reduction in cases of microcephaly after the Zika epidemic (2015-2016), but points to a new increase in 2023 with no apparent cause. This recent increase raises several questions: could it be due to the relaxation of prevention and control measures? Could it be associated with the recent Dengue and Covid-19 epidemics, with diverted attention from microcephaly? Could there be flaws in data collection and tracking methods? Or could it be a new pathology or condition that is still unknown? We conclude that it is necessary to turn attention to microcephaly and investigate what led to the recent increase in cases.

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Epidemiological characterization of polio vaccination in Brazil and in the state of São Paulo during the 21st century

Helena Rubini Nogueira[1], Helena Rubini Nogueira[1], Luma Falcon Maran[1], Bianca Stephani Ovando Pereira[1], Breno Nascimento Puertas[1], Giulianna Luzia Pereira[1], Geovanna Dhi Genaro[1], João Victor Salas Salles[1], Patrick Cristian Lima Orihuela[1], Lucas Valadão Ferreira de Lira[1]

[1]Universidade Municipal de São Caetano do Sul, São Caetano SP, Brazil.

Correspondence: Helena Rubini Nogueira; Email: helena.rnogueira@live.com.

Background: Poliomyelitis is an incurable viral infection that mainly affects children, with the last recorded case in Brazil being in 1989. Etiopathogenically, the virus replication leads to the damage of the neuronal cells and consequent impairment of the stimuli production responsible for muscle contraction, resulting in paralysis. The diagnosis involves clinical and laboratory investigation and the treatment is mainly to relieve the symptoms. In view of this, the eradication strategy was centered on mass vaccination campaigns, but as this coverage has decreased, the pathology has re-established itself as a global threat. **Objective:** To characterize polio vaccination in the 21st century in the Brazilian context and, more specifically, in the state of São Paulo. **Methods:** Descriptive analysis based on data obtained from DATASUS using the following variables: Vaccination Coverage (Poliomyelitis; Poliomyelitis 4 years); Brazilian Regions; Macro-regions of the state of São Paulo; Period (2001 to 2022). **Results:** Nationally, vaccination coverage rates showed variations from 2010 onwards, previously remaining above 100%. This reduction intensified from 2014 (maximum value 98.29%), with a second worsening in 2020 (average 75.01%). It should be noted that, since 2010, the South, followed by the Midwest, have had the highest vaccination rates, with 41.6% and 33.3% of the total, respectively. In comparison, this coverage fell by 22.22% over the century. When analyzing booster vaccination at the age of 4, there was a drop from 2019 (68.45%) to 2021 (54.61%), with a further increase in 2022 (67.56%). In this section, the South and Midwest once again stood out, with the highest average values. Within the 17 Regional Health Care Networks (RRAS) in the state of São Paulo, it can be seen that RRAS-6 (corresponding to the municipality of São Paulo) showed the lowest total percentage (88.71%) of coverage between 2000 and 2022, while RRAS-12 (which includes Santa Fé do Sul, Jales, Fernandópolis, Votuporanga, São José do Rio Preto, José Bonifácio, Catanduva, Lagos do DRS II, Central DRS II, Consórcios DRS II) showed the highest (100.63%). As for booster vaccination in São Paulo's health macro-regions, RRAS-6 and RRAS-12 showed the best results (averages between 81.83% and 91.31%), while the worst were recorded in RRAS-7 (Baixada Santista and Vale da Ribeira) and RRAS-3 (Franco da Rocha), with 59.12% and 60.91% respectively. **Conclusion:** There has been a sharp drop in vaccination coverage against Polio since 2014 at a national level, as well as in the state of São Paulo, illustrated by the decrease registered mainly in the regional health care network that includes the largest municipality in the country (São Paulo). The importance of vaccination to prevent new cases of the disease, which is considered incurable and highly infectious, is highlighted.

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Epidemiological disparities in hospitalizations for multiple sclerosis in the face of the COVID-19 pandemic

Giulianna Luzia Pereira[1], Helena Rubini Nogueira[1], Lucas Valadão Ferreira de Lira[1], Cássio Marques Ribeiro[1], João Victor Salas Salles[1], Patrick Cristian Lima Orihuela[1], Geovanna Dhi Genaro[1], Bianca Stephani Ovando Pereira[1], Breno Nascimento Puertas[1], Luma Falcon Maran[1]

[1]Universidade Municipal de São Caetano do Sul, São Caetano do Sul SP, Brazil.

Correspondence: Giulianna Luzia Pereira; Email: giuliannaluzia2013@gmail.com.

Background: Multiple sclerosis (MS) is an autoimmune disease that affects 2.3 million people worldwide and around 40,000 Brazilians. Its pathogenesis remains unknown, but there is a correlation between genetic susceptibility and environmental agents, such as lack of vitamin D, time of birth, tobacco use and exposure to the Epstein-Barr virus. The diagnosis of the disease is complex because no test or biomarker is specific. It consists of 2 symptomatic episodes lasting more than 24 hours, at least 1 month apart (different time and place). Its treatment consists of drug therapy (mainly with immunomodulators) and non-drug therapy (physiotherapy, physical exercise and psychotherapy). In addition, the advent of the COVID-19 pandemic has posed challenges for MS care, especially in terms of treatment and use of the vaccine, as well as access to health services, factors directly related to the prognosis of the condition. Few studies have managed to make direct correlations between MS and COVID-19 infection. **Objective:** To identify disparities in hospitalizations for multiple sclerosis before, during and after the COVID-19 pandemic according to epidemiological factors. **Methods:** Descriptive analysis based on data obtained from DATASUS using the following variables: ICD 10 morbidity list (multiple sclerosis); Age group; Brazilian regions; Sex; Hospitalizations; Death; Mortality rate; Period (Jan/2018 to Dec/2023). **Results:** There was a reduction in the number of hospitalizations for MS during the pandemic of approximately 1.5%, with an increase in the post-pandemic period of 11%. In addition, it was found that adults aged between 20 and 59 were the group of the population that suffered the most hospitalizations for the condition (n = 28.708; 88%), in contrast to 1.788 children (5.5%) and 1.993 elderly people (6.5%). More specifically regarding to the Brazilian regions, there was no significant variation in the data as a result of the pandemic, with the North remaining the place with the lowest number of hospitalizations recorded (n = 453; 1.4%) and the Southeast the highest (n=24,045; 73.9%), whose female population accounted for 69% of hospitalizations. Considering the number of deaths from MS, there was an increase until 2019 (n=51), led by the Southeast Region (50%). However, the regions showed a drop in this pattern during the pandemic (n=19 in 2020; n=26 in 2021), and a subsequent increase after its end: 2022 (n=34) and 2023 (n=33). In general, there was a reduction in the mortality rate during the pandemic period in all regions, except for the North (2019: 5.88 → 2020: 6.06), with the Southeast having the lowest rate (2020: 0.28 and 2021: 0.15). **Conclusion:** A decrease in the number of hospitalizations, deaths and mortality rates was observed in the vast majority of Brazilian regions during the COVID-19 pandemic, with the adult and female population suffering the most hospitalizations.

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Epidemiological factors implicated in Parkinson's disease: a preliminary study of patients from an outpatient clinic with a standardized routine questionnaire

Tatiane Thaís Santos Silva[1], Sophia Keiko Inuzuka[1], Júlio César Iaia[1], Maria Fernanda de Abreu Rodrigues[1], Rhanice Soares Huguenin[1], Iuri Sena Melo[1], Marcos Vinicius Souza Do Nascimento Júnior[1], Roberto Pereira Santos[1], Ana Lucia Zuma de Rosso[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Tatiane Thaís Santos Silva; Email: tatianethaisufrj@gmail.com.

Background: Parkinson's disease (PD) is a progressive neurodegenerative disease resulting from degeneration of dopaminergic neurons in substantia nigra. Although the exact cause of PD is not yet completely understood, it is fostered by advanced age and the interaction of a variety of environmental factors and genetic factors. **Objective:** To develop a standardized epidemiological questionnaire, quick to apply and easy to understand for patients, to be used in routine neurological consultations to assess socio-environmental factors described as predisposing to the development of PD. **Methods:** Cross-sectional study with patients with a >2y PD diagnosis attending the Movement Disorders outpatient clinic of a universitary hospital who agreed to participate. We developed a structured questionnaire with 19 items that covered medical history, environmental exposures, and lifestyle habits before PD diagnosis. The questionnaire was applied by one of the researchers immediately before the scheduled neurological consultation, noting its execution time and ease of understanding by the patients.The Hoehn & Yahr scale defined the stage of the disease. **Results:** We analyzed 22 patients (77.3% male), with a mean age of 70.1 + 8.2 y (range 51-91y) and an average diagnosis time of 10.1+5.9y. Most participants were white (73%), were in H&Y phase II-III (78.6%) and had <8 years of formal education (59.1%). The questionnaire proved to be relatively easy to understand and quick to complete, taking an average time of 17 min (range 10' to 22'). In 31.8% there was a family history of PD. In 45.5% there was a history of depression, 36.4% had an anxiety disorder and 54.5% referred REM sleep behavior disorder. Furthermore, 36.4% had chronic constipation and 22.7% complained about hyposmia. Only 18.2% reported lifetime pesticide exposure, while 22.7% had a history of traumatic brain injury. Regarding medication use, 13.6% used calcium channel blockers and/or beta-blockers. Behaviorally, 27.3% did not engage in physical activity, but 59.1% reported sun exposure >3 days/week. 50% of parkinsonians denied ever smoking. The average caffeine consumption was 130.7±110.2ml/day. **Conclusion:** The questionnaire developed in this work proved to be practical and should be incorporated into the care of parkinsonians in our hospital. Due to the limited size of the research sample so far, we found some discrepant data in relation to the literature, such as the low rate of pesticides exposure, depression and hyposmia. The importance of epidemiological factors and lifestyle habits for the development and progression of PD has been increasingly emphasized in recent global studies. In routine neurological consultations, however, this search is rarely carried out due to the lack of a quick standardized assessment instrument. In this way, we hope to contribute to future research by enabling the collection of large-scale behavioral, medical and environmental data from our patients.

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Epidemiological overview of migraine hospitalizations in Brazil: profile of hospitalized individuals and costs

Matheus Wendell de Almeida Pereira[1], Maria Alice Pereira Junqueira de Oliveira[1], Raissa Jaquelline dos Santos Dias[1], Éricka Pessôa de Azevedo[1], Gabriel da Conceição Veiga[1], Carla Caciana Leocadio Toledo[1], Luísa Lisbôa Machado[1], Camilla de Morais Lavadores[1]

[1]Universidade Federal do Rio de Janeiro, Macaé RJ, Brazil.

Correspondence: Matheus Wendell de Almeida Pereira; Email: mwap1313@gmail.com.

Background: Migraine stands as a pervasive condition worldwide, exerting a profound influence on individuals' quality of life and productivity. Despite its implications, the disorder remains neglected by both patients and healthcare providers. Therefore, understanding the current epidemiological situation regarding this issue is crucial for healthcare services. **Objective:** This study aims to conduct a quantitative analysis of hospitalizations in public healthcare institutions from January 2014 to January 2024, specifically focusing on cases attributed to migraine and other cephalic pain syndromes. The objective is to compare these findings across Brazil's five distinct regions, examining variables such as gender, ethnicity, age demographics, and the average cost of hospitalization. **Methods:** Using observational and cross-sectional approaches, this study utilizes data from Sistema de Informações Hospitalares do SUS (SIH-SUS) via DATASUS. Variables include hospitalizations for migraines and cephalic pain syndromes categorized by region, gender, ethnicity, age groups, and average costs. Data collection occurred from March 18 to 23, 2024. **Results:** During the specified period, Brazil documented 97,156 hospitalizations attributable to migraines and other cephalic pain syndromes, exhibiting a general inclination towards escalation over the years, albeit with declines observed in 2020 and 2021. The Southeast region registered the highest cumulative cases in the period, comprising 32.5% of the total, followed by the Northeast and South with 29.2% and 25.9%, respectively. Regarding gender distribution, females comprised 65.8% of hospitalizations, while males accounted for 34.1%. In terms of ethnicity, Caucasian individuals predominated, constituting 39.64% of cases, followed by "pardo" individuals (multiracial Brazilians), representing 39.62%. Notably, 15.4% of hospitalizations lacked ethnicity information. Moreover, the majority of hospitalizations occurred in individuals aged between 30 and 39 years, accounting for 18.5% of the total, followed by the age group of 20 to 29 years, accounting for 17.8%. It is pertinent to highlight that the average hospitalization cost surged from R$327.10 in January 2014 to R$673.80 in January 2024, peaking at a maximum of R$1,039.24 in the Northeast. **Conclusion:** It is noted that 15.4% of cases lacked information regarding the ethnicity of patients, thus compromising the accurate understanding of the individuals' profile. This deficiency hampers the formulation and execution of public health interventions aimed at preventing complications associated with the condition and ensuring the appropriate treatment of migraine. Furthermore, the two most affected groups represent a segment of the population with significant economic engagement. This underscores the need for effective disorder management, that could help mitigate complications and decrease the frequency cost of hospitalization cases.

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Epidemiological profile analysis of hospital admissions for viral encephalitis in Brazil from 2019 to 2023

Aline Teixeira[1], Ana Clara Lacerda Freitas[1], Laysa de Souza Maia[2], Andressa Gonçalves Vicente[3], Ana Alice Lemos Lima[4], Gabriela Hernandez Dumani[5], Ana Cristina Carvalho e Silva[3], Estefany Moreno de Souza[6], Geovana Franklim Gomes e Silva[7], Guilherme de Andrade Ruela[4]

[1]Centro Universitário Funorte, Montes Claros MG, Brazil.
[2]Faculdades Integradas Aparício Carvalho, Jaru RO, Brazil.
[3]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[5]Universidade Franciscana, Santa Maria RS, Brazil.
[6]Universidade Aquino de Bolivia, Santa Cruz de La Sierra, Bolivia.
[7]Centro Universitário de Patos de Minas, Patos de Minas MG, Brazil.

Correspondence: Laysa de Souza Maia; Email: laysamaia1503@gmail.com.

Background: Encephalitis is characterized by inflammation of the brain parenchyma due to an acute infection or a post-infectious immune reaction and is associated with altered brain function. Clinical manifestations include fever, headache, focal neurological signs, altered consciousness, and seizures. In 2018, the hospital admission rate for viral encephalitis in Brazil was approximately 1 per 100,000 inhabitants. Therefore, this study aims to provide

an updated epidemiological overview of the condition. **Objective:** To analyze the profile of hospital admissions through the SUS for viral encephalitis in Brazil over the past 5 years. **Methods:** This is an ecological, cross-sectional, and descriptive study using secondary data available from the Hospital Information System of the Unified Health System (SIH/SUS) from 2019 to 2023. The variables analyzed were: demographic region, hospital admissions, sex, age group, and mortality rate. Data were collected and analyzed in May 2024 using descriptive statistics. **Results:** A total of 9,816 hospital admissions for viral encephalitis were recorded in Brazil over the past five years, with the highest number of admissions in the Northeast region. In the first year analyzed, there were 2,436 hospitalizations for viral encephalitis. A 32.4% decrease in the number of hospitalizations was observed from 2019 to 2021. In 2022, there was a new increase, which remained constant until 2023, reaching 2,067 cases. The highest number of admissions was among males (53.5%) and in the age group of 1 to 9 years (30.1%). The highest mortality rate was in 2020 at 6.28%, but in subsequent years, it showed a downward trend despite the increase in the number of hospitalizations. **Conclusion:** There has been an overall increase in the total number of hospital admissions for encephalitis from 2019 to 2023. However, it is important to consider that the COVID-19 pandemic likely influenced the significant reduction in the number of admissions between 2020 and 2021. In this context, the current challenge for the country is to reduce the number of hospital admissions through the SUS, as although the mortality rate is decreasing, the increase in hospitalizations indicates that this condition remains a public health challenge in Brazil.

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Epidemiological profile and comorbidities of patients with Parkinson's disease at an university hospital

Ligia Aranha Puig Maldonado[1], Denise Hack Nicaretta[1], Maria Aparecida de Assis Patroclo[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Escola de Medicina e Cirurgia, Hospital Universitário Gaffrée e Guinle, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Instituto de Saúde Coletiva, Rio de Janeiro RJ, Brazil.

Correspondence: Denise Hack Nicaretta; Email: denisehn@terra.com.br.

Background: Parkinson's disease (PD) is a neurodegenerative disorder with onset prior to age 60 years, with unclear causes but linked to genetics, environment and aging. Risk factors include male gender, age over 80, pesticide exposure, head trauma, and genetics. It affects over 8.5 million people globally aged over 65, with an estimated 200,000 in Brazil and 7,121 in Rio de Janeiro. PD's prevalence is rising and expected to double by 2040 due to aging populations. **Objective:** To analyze the epidemiological profile and comorbidities in PD patients from the Parkinson's Disease Outpatient Clinic of a University Hospital in Rio de Janeiro city, over a five-year period from 2017 to 2022. The present study is justified due to the high prevalence of PD in the global population, and due to the many aspects to be elucidated both in its pathophysiology and in relation to epidemiology, risk factors, and associated comorbidities. **Methods:** Retrospective cross-sectional observational study. Target population: Adults diagnosed with PD according to the Movement Disorders Society (MDS) criteria from 2015, enrolled in the Parkinson's Disease Outpatient Clinic at a university hospital in Rio de Janeiro, from 2017 to 2022. Data were collected from patient records and self-declared. The project was submitted and approved by the Institutional Review Board for Human Research Ethics, and informed consent was obtained from all participants via telephone. **Results:** 53 patients met the criteria, agreed and gave consent. Among them, 20 (37.7%) were female, and 33 (62.2%) were male. The average age at diagnosis was 57.8 years, 28 (52.8%) identified as white, and a total of 47.9% identified as Black (Black + Mixed race), despite the majority of the population in Rio de Janeiro being black/mixed race. 49.2% had up to elementary education, and 51.8% had higher education levels. 71.6% (38 participants) did not have a history of smoking. 30 (56.6%) had systemic arterial hypertension; 14 (26.4%) had Type II Diabetes Mellitus; and 11 (20.7%) had other cardiovascular diseases. 23 (43.3%) patients had depression/depressed mood; 15 (28.3%) reported anxious mood; and 19 (35.8%) had other disorders. 28 (52.8%) patients reported insomnia; 22 (41.5%) had Rapid Eye Movement Sleep Behavior Disorder. 6 (11.3%) patients had a history of neoplasias. **Conclusion:** White males predominated, with an average age at diagnosis of 57.8 years. Comorbidities like cardiovascular diseases and depression were prevalent. The study suggests the need for standardized data collection and ongoing research to better understand and manage PD in Rio de Janeiro, especially with aging populations.

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Epidemiological profile and impact of hospitalizations for complicated migraine in Brazil (2019-2023)

Monyque Pinheiro Charchar[1], Marcos Lude da Silva Ferreira[2], Lívia Vitória Costa Dias[1], Alec Peixoto Rabelo[2], Edson Luis de Barros Siqueira[2], Luiz Carlos Figueiredo Filho[2], Luan Nascimento Pereira de Amorim[2]

[1]Centro Universitário do Pará, Belém PA, Brazil.
[2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Monyque Pinheiro Charchar; Email: mcharchar10@gmail.com.

Background: Most migraines are adequately treated on an outpatient basis. However, there is a form, known as migraine with aura, which involves more severe or prolonged symptoms, such as persistent auras and neurological symptoms, requiring more specific and immediate medical interventions. **Methods:** Through quantitative and temporal analysis, a study was conducted on data available in the Public Health Event Records (RESP) regarding the frequency of notification of complicated migraine treatment among the Brazilian Federal Units (UF), and variables such as type of hospitalization, death rate, mortality rate, average value, and annual average length of stay. Secondary information was obtained digitally using data tabulation tools TABNET and TABWIN, provided by the Brazilian Unified Health System Information System (SIH/SUS) and made available by the Department of Informatics of the Unified Health System (DATASUS). **Results:** Between 2019 and 2023, 38,908 hospital admission authorizations (AIH) were approved, with a peak in 2019. Additionally, 31.1% were recorded in the Southeast region, with São Paulo having the highest number of AIH (7,313). Pará (1,281), Maranhão (4,186), Paraná (6,185), and Goiás (2,343) also stood out in their regions. Urgent hospitalizations predominated in all states except Goiás, totaling 35,773, while elective admissions amounted to 3,133, all of medium complexity. During this period, the total death rate was 125 cases, with 24% occurring in 2019 and 35.2% in the Northeast region, mainly in Bahia. However, the highest mortality rates were observed in the North region, reaching 0.83 with a peak in 2021, especially in Amazonas (2.65), highlighting 2020 (7.69). The average expenditure was 312.66 Brazilian reais, with higher spending in 2023 (336.89), with the highest rates in the South region, mainly in Paraná (466.32). The average annual length of stay in all regions was 3.4 days, with the highest average in the Northeast (4.1), led by Sergipe (7.6). **Conclusion:** Between 2019 and 2023, 38,908 hospitalizations for complicated migraines were recorded in Brazil, peaking in 2019. The Southeast region had the highest number of hospitalizations, especially São Paulo. Most admissions were urgent, with higher mortality rates in the North, especially in Amazonas. Average costs increased, with the highest spending in Paraná. The average hospital stay was 3.4 days, higher in the Northeast, led by Sergipe. These data emphasize the need for regional public policies to optimize resources, reduce mortality and costs, and improve the treatment of complicated migraines

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Epidemiological profile and mortality of hospitalizations for epilepsy in the state of Pará from 2014 to 2023

Fernando Gabriel dos Santos Santiago[1], Danusa Neves Somensi[1], Mariangela Moreno Domingues[1], Emanuel de Jesus Soares de Sousa[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Fernando Gabriel dos Santos Santiago; Email: fernando_gsantiago@outlook.com.

Background: Epilepsy is defined as a predisposition to the development of paroxysmal events characterized by behavioral, sensory, psychic, autonomic, and cognitive changes with a strong neurobiological, psychological, and socioeconomic impact. It is estimated that it affects 50 million people worldwide, while nationally it has a prevalence of about 2%, being one of the most predominant neurological conditions. Therefore, due to its strong social, economic, and biological impact, it is essential to characterize the epidemiological profile and morbidity and mortality. **Objective:** To analyze the epidemiological profile of epilepsy and its morbidity and mortality in a hospital context in the state of Pará from 2014 to 2023. **Methods:** This is an ecological, exploratory and descriptive time series study, carried out through data extracted from the Hospital Information System, made available through the Department of Informatics of the Unified Health System (DATASUS), corresponding to hospitalizations for epilepsy in the state of Pará from 2014 to 2023. The variables collected were: age group, sex, color/race, number of hospitalizations, days of stay, total amount spent, and mortality rate. For comparison of the data published by DATASUS, searches were carried out for scientific articles that described such realities of the disease, being searched on the SciELO and LILACS platforms. **Results:** There was a total of 10,307 hospitalizations with a trend of increase over the evaluated period, in which the year 2023 corresponded to the highest number with 1,218 and 2015 with the lowest number with about 928 hospitalizations. Furthermore, the predominant color/race was brown, corresponding to 64% of the cases. In relation to sex, the prevalent was male with 56% of hospitalizations, while the most affected age group was 0-14 years with 47% of hospitalizations. The average length of stay was 4.7 days and the total amount spent during the period was 5,282,915.17, with 2023 being the year with the highest total expenditure of 971,704.56 and the lowest in 2015 with 323,891.69, representing a trend of increase in line with the increase in the number of hospitalizations. When analyzing the mortality rate, there was an average of 2.19, staying below the national and regional average of 2.48 and 2.2, respectively. In addition, there is a higher predominance of mortality in males over the age of 60 with an average mortality of 7.91. **Conclusion:** In summary, the epidemiological profile researched shows a higher prevalence in the age of 0-14 years of male sex and brown color/race with a trend of increasing number of hospitalizations and total amount spent. Likewise, a mortality rate was found below the national and regional average, but with a high risk in male patients over 60 years old. The characterization of this care profile favors the planning of actions that can provide early diagnosis and treatment of this so prevalent condition.

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Epidemiological profile and public expenditure of hospital admissions for migraines in Brazil

Joanna Sousa da Fonseca Santana[1], Laura Carolina Costa Andrade[1], Humberto de Castro Lima Filho[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Joanna Sousa da Fonseca Santana; Email: joannasantana20.2@bahiana.edu.br.

Background: Migraine is a neurovascular condition with a high global incidence and is prevalent in Brazil, ranking as the third most common complaint in medical consultations and the fourth leading cause of emergency room visits. Its socioeconomic impact is substantial, leading to healthcare costs, productivity loss, and standing out as the primary cause of disability in individuals under 50 worldwide. Therefore, an analysis of the epidemiological profile of migraine hospitalizations in Brazil is needed. **Objective:** To analyze the epidemiological profile and public expenditures on hospitalizations for migraine and other cephalalgias in Brazil. **Methods:** This is a cross-sectional, epidemiological study using secondary data from the Brazilian Unified Health System Hospital Information System (SIH/SUS). The study covered the Brazilian territory from 2012 to 2022, including data on patients hospitalized for migraine and other cephalalgias (ICD codes G43 and G44) from SIH/SUS. The variables studied were sex, race/ethnicity, age group, year of hospitalization, average length of stay, total days of stay, and total public expenditures on hospitalizations per year. Statistical analysis and data tabulation were performed using Excel for Windows. **Results:** A total of 97,093 hospitalizations were recorded, with 33,234 (34.23%) involving male patients and 63,859 (65.77%) involving female patients. The age group with the highest hospitalization rate was 30-39 years (18.68%), followed by 20-29 years (18.21%). The lowest rate among adults was 50-59 years (8.79%). Whites had the most hospitalizations (40.98%), and Indigenous the least (0.06%). The average annual hospitalizations over the decade were 8,789.63 ± 1,850.54. The total expenditure over the decade was R$ 42,923,587.81, with an average of R$ 3,887,132.70 ± 1,402,110.54 per year. Additionally, out of the 97,093 hospitalizations, 6,019 were in the North region (6.20%), 26,904 in the Northeast (27.71%), 33,501 in the Southeast (34.50%), 25,544 in the South (26.31%), and 5,125 in the Midwest (5.28%). There were a total of 377,687 days of stay across all years, with an average stay of 3.9 days. Hospitalizations peaked in 2019 with a 48.86% increase over three years, followed by a 28.32% decline until 2021, then a 14.22% rise in 2022. **Conclusion:** The epidemiological profile of hospital admissions is closely linked to the prevalence profile of migraines. In Brazil, the annual number of migraine-related hospitalizations is significantly high, as is the average length of stay, resulting in a substantial cost to the Brazilian Unified Health System. Improved access to prophylaxis, education of non-specialist healthcare providers in diagnosing and treating simple migraines, and prudent resource utilization are necessary to enhance quality of care and reduce the burden on public healthcare. Significant public health and economic impacts underscore the need for better preventive care and resource management.

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Epidemiological profile and temporal trends of meningococcemia in Brazil between 2010 and 2023

Ryan Lucas Barbosa Oliveira[1], Laís Souza da Silva Vitorino[1], Lorena Marques Ribeiro[1], Maria Danyelle Farias de Oliveira[1], William Correia da Silva Filho[1], Maryanne Maria Simplício Santos[1], Taíssa Kelly Novais Veiga[1], Mariana Araújo Oliveira Barbosa Rocha[1], Valdirene Maria de Barros Barbosa[2]

[1]Universidade Federal de Alagoas, Arapiraca AL, Brazil.
[2]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Ryan Lucas Barbosa Oliveira; Email: ryan.oliveira@arapiraca.ufal.br.

Background: Meningococcemia is one of the most severe and lethal clinical forms of meningitis, characterized by rapid progression to death due to shock, sepsis and disseminated intravascular coagulation. Therefore, it remains epidemiologically relevant. **Objective:** To describe the epidemiological profile of meningococcemia in Brazil from 2010 to 2023. **Methods:** This observational, retrospective, descriptive study utilized data from the Notifiable Diseases Information System (SINAN/SUS - DATASUS) covering 2010-2023, considering hospitalizations according to place of residence. The variables analyzed were sex, race, month, mortality rate, and year. The analysis was performed using Microsoft Office Excel® 2016 and Joinpoint Regression® 5.1 to determine the annual percentage change (APC) with a 95% confidence interval (CI). **Results:** Among the clinical forms of meningitis, meningococcemia was the most lethal (39.7%), followed by tuberculous meningitis (31.3%). From 2010 to 2023, hospitalizations were primarily among children aged 1-4 (21.2%), males (54.6%), and whites (41.7%). There were 5,757 hospitalizations, predominantly between May and September, peaking in 2010 (941 hospitalizations and 359 deaths). The years with the lowest number of hospitalizations occurred in 2021 (76 hospitalizations) and 2020 (108 hospitalizations). The 14-year analysis showed an annual reduction of -13.7% (APC:-13.7 | CI95%=-18.3,-11.7; p<0.001). By establishing an inflection point in 2021, there is a marked decrease from 2010 to 2021 (APC:-14.5 | CI95%= -23.8,-3.70; p=0.011), and an increase from 2021 to 2023 (APC:6.19 | CI95%= -37;30.1; p=0.697). São Paulo had the highest number of hospitalizations (45.1%), followed by Rio de Janeiro (16.8%), while Roraima and Amapá had the lowest (0.01% each). During the study, there were 2,287 deaths, yielding a national mortality rate of 0.07/100,000 inhabitants/year, higher in the 20-39 and 1-4 age groups. São Paulo and Rio de Janeiro had the highest mortality rates (0.397 and 0.395/100,000 inhabitants/year), while Roraima and Tocantins had the lowest (0.03 and 0.04/100,000 inhabitants/year). **Conclusion:** Hospitalizations and deaths declined significantly from 2011 onwards, likely due to the introduction of the meningococcal C vaccine in 2010 and its extension to adolescents aged 11-14 in 2018. The mortality rate was higher in states with more hospitalizations, such as São Paulo and Rio de Janeiro, suggesting a possible lack of notification in states with lower mortality and hospitalization rates, such as Roraima. The distribution of hospitalizations in the months mentioned shows a seasonal behavior, with a higher incidence in the autumn and winter periods. The lower hospitalization rates in 2020 and 2021 could be due to underreporting during the COVID-19 pandemic, warranting further analysis to confirm the increase from 2021 onwards.

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Epidemiological profile and treatment efficacy of cladribine in a single Brazilian multiple sclerosis center

Breno Sousa Paiva[1], Fabio Victor Vieira Rocha[1], Daniel José Siqueira Tosi[1], Guilherme Valiate Picoli[1], Paula Zago Melo[2] , Bruno Batitucci Castrillo[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.
[2]Hospital Universitário Cassiano Antonio Moraes, Vitória ES, Brazil.

Correspondence: Fabio Victor Vieira Rocha; Email: fabio.vvrocha@gmail.com.

Background: Multiple sclerosis (MS) is a demyelinating disease of the nervous system with an autoimmune inflammatory mechanism, affecting the white matter of the brain, spinal cord, and optic nerve. Oral cladribine is a therapeutic alternative for patients with Relapsing-Remitting Multiple Sclerosis (RRMS), as it offers a convenient dosing regimen from the perspective of both patients and the healthcare system, being an oral medication with spaced doses and the lowest monitoring burden among treatments. Oral cladribine was approved in Brazil in 2019, but there is limited local data on clinical practice. **Objective:** To analyze the efficacy and persistence of cladribine treatment among Brazilian patients with multiple sclerosis in a Single Center. **Methods:** This is an observational, retrospective, and longitudinal study involving 10 MS patients treated with cladribine. The inclusion criteria were: diagnosis of relapsing-onset MS according to current criteria, and initiation of oral cladribine treatment at least 6 months before analysis. The main clinical data included disease activity (relapses and MRI follow-up), worsening of EDSS (Expanded Disability Status Scale), and adverse effects. **Results:** 10 patients treated with cladribine were included in the initial analysis, 9 (90%) being females. Median age at disease onset was 33 years (19-49), and on starting cladribine was 33.5 years (25-51). Median initial EDSS score was 2 (1-2.5). The main indications for cladribine use were: gestational desire in 7 (70%) patients; risk of Progressive Multifocal Leukoencephalopathy (PML) in 2 (20%) patients; and convenience in 1 (10%) patient. 80% of patients were naïve. After 15.5 months (7-39) of follow-up, 9 (90%) patients remained relapse-free, 9 (90%) did not show EDSS progression, 8 (80%) did not show MRI activity, and 8 (80%) were in NEDA-3. There were no treatment discontinuations. The minimum lymphocyte count reached a median of 1000.5 (510-1390), with 5 (50%) patients experiencing lymphopenia. Among those with lymphopenia, 4 (40% of patients; 80% of lymphopenias) had grade 2 lymphopenia, while 1 (10% of patients; 20% of lymphopenias) had grade 1 lymphopenia. The median time to the worst lymphopenia occurrence after starting cladribine was 9.5 months (range: 1-15). Additionally, 9 patients experienced side effects, with only one considered severe. **Conclusion:** The data demonstrate therapeutic success, with high efficacy and persistence of treatment among Brazilian MS patients, evidenced by 80% of patients achieving NEDA-3. The result is probably attributed to the high number of naïve patients (80%) in the sample, although there is a temporal limitation of the study.

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Epidemiological profile of admissions for brain neoplasm in Northeast Brazil between 2008 and 2023

Vitor Fernando Bordin Miola[1], Luís Henrique dos Santos Sousa[2], Maria Fernanda de ávila Reis[3], Rafael Alvarenga de Oliveira Pereira[4], Nicolas Minavia Delgadillo[4]

[1]Universidade de Marília, Marília SP, Brazil.
[2]Universidade Federal de Pernambuco, Serra Talhada PE, Brazil.
[3]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[4]Universidade Metropolitana de Santos, Santos SP, Brazil.

Correspondence: Rafael Alvarenga de Oliveira Pereira; Email: rafael.oliva192@gmail.com.

Background: Brain neoplasms, although less frequent than other types of cancer, are notorious for their consumptive human impact, high mortality rates and the challenge they represent for the health system in view of the aging population. In the Brazilian context, the regional analysis of hospitalizations for brain neoplasms is essential to understand the epidemiological particularities and the specific challenges of each region. The Brazilian Northeast, a region marked by great socioeconomic disparities and difficulties in accessing specialized health care, represents a unique scenario in identifying epidemiological patterns and priority areas for interventions. **Objective:** To describe the epidemiological profile of hospitalizations for malignant neoplasia of the brain in the Northeast region of Brazil, analyzing data regarding age group, race/color, regime and type of care, length of hospital stay and geographic distribution in the period

from 2008 to 2023. **Methods:** It consists of a quantitative, observational and descriptive study based on secondary data from the Department of Informatics of the Unified Health System (DATASUS), of the Ministry of Health, on cases of hospitalizations for malignant neoplasia of the brain in the last 16 years (2008 to 2023). **Results:** During the 16 years observed, there were 43,727 hospitalizations for brain neoplasms in the Northeast of Brazil, corresponding to 20.3% of the total number of hospitalizations for this cause in the country. Of these, 81.0% were urgent, while at national level this figure was 76.5%. The average hospital stay in the region was 11.9 days, above the national average of 10.7 days; 26.9% occurred in the state of Pernambuco, 24.5% in Bahia and 17.5% in Ceará, 6.7% in Rio Grande do Norte, 6.3% in Paraíba, 6.3% in Piauí, 4, 1% in Maranhão, 4.0% in Alagoas and 3.6% in Sergipe. 57.2% of hospitalizations were mixed-race people; however, 35.0% did not have color/race information specified; 49.0% of hospitalizations were among individuals between 20 and 59 years old, 25.5% among individuals under 20 years old and 25.5% among individuals over 60 years old. **Conclusion:** There is a considerable incidence of brain neoplasia in the Brazilian Northeast when compared to the total number of Brazilian cases, representing a substantial part of the country's hospitalizations for this cause, highlighting the importance of this health problem in this region. Furthermore, the majority of hospitalizations occurred on an urgent basis in Pernambuco, Bahia and Ceará, indicating that some of these patients may already be in advanced stages of the neoplasia. The majority of hospitalizations were mixed-race people, with a considerable number of people without this specification and with an age range mainly between 20 and 59 years old. The importance of developing health policies aimed at screening and early diagnosis, as well as adequate treatment to reduce hospitalization rates for brain neoplasia, is highlighted.

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Epidemiological profile of central nervous system neoplasms in the last decade

Bruno Ale Bark[1], Enzo Figueiredo Martineli[1], Samir Ale Bark[2], Bruno Henrique Mota Segnori[2], Guilherme Dorabiallo Bark[3]

[1]Universidade de Marília, Marília SP, Brazil.
[2]Faculdade Evangélica Mackenzie do Paraná, Curitiba PR, Brazil.
[3]Hospital do Rocio, Curitiba PR, Brazil.

Correspondence: Bruno Ale Bark; Email: brunoale.bark12@gmail.com.

Background: The epidemiological profile is one of the fundamental pillars of public health as it offers a detailed view of the health condition of a population. It is observed that central nervous system neoplasms have gained prominence as an area of significant epidemiological studies in the field of neuro-oncology. These neoplasms include a wide range of tumors, from benign to highly malignant, and their differentiation is crucial for choosing the therapeutic approach and prognosis, considering that they can cause irreversible or even fatal neurological damage. The causes of these tumors are diverse, with age, sex, race, epidemiological factors, and lifestyle habits being the main modulators for the development of neoplasms. **Objective:** To analyze the epidemiological profile of patients diagnosed with central nervous system neoplasms. **Methods:** A cross-sectional, retrospective, observational, and quantitative study was conducted through the statistical analysis of data on malignant and benign neoplasms of the brain and other parts of the central nervous system obtained from DATASUS - Department of Informatics of the Unified Health System, with the variables "sex," "age group," and "race," from January 2014 to January 2024. **Results:** Patients in the age group of 30 to 59 years resulted in 70,736 hospitalizations for malignant neoplasms and 14,126 for benign neoplasms. In the age group of 1 to 29 years, there were 49,386 hospitalizations for malignant neoplasms and 5,500 for benign neoplasms. Additionally, in the age group over 60 years, malignant neoplasms accounted for 55,527 hospitalizations, while benign neoplasms totaled 7,528 hospitalizations. Analyzing the variable: sex, it was observed that malignant neoplasms of the central nervous system are more prevalent in men 92,045 (52.05%) than in women 84,794 (47.95%). Conversely, benign neoplasms were more prevalent in women 14,939 (54.55%) than in men 12,445 (45.45%). Regarding race, the results showed a predominance in whites 77,103 (51.94%), followed by browns 63,640 (42.87%) in malignant neoplasms, while in benign neoplasms, the values obtained were, respectively, 12,963 (58.43%) and 8,300 (37.41%) in the brown and white races. **Conclusion:** The epidemiological profile is not merely an expression of data but a facilitator for understanding, planning, and implementing effective public health policies. The present study showed that the age group of 30 to 59 years is the most affected by central nervous system neoplasms, while malignant neoplasms affect males more and benign neoplasms affect females more. Similarly, an inversion was observed concerning race, with whites having a predominance of malignant neoplasms, while in benign neoplasms, the predominance occurs in browns. In light of this scenario, attention should be paid to these variables for both early diagnosis and effective therapeutic implementation.

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Epidemiological profile of deaths due to brain cancer in Brazil between 2017 and 2021

Guilherme Rocha Cardoso[1], Arthur Mendes de Assis Martins Vieira[1], Matheus Nascimento Moura[2], Brenno Amaral da Silva Oliveira[2], Pedro Ferreira de Souza Evangelista[3], Emmanuel Santos Trindade[1], Priscila Santos Oliveira[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.
[2]Universidade Estadual do Sudoeste da Bahia, Instituto Multidisciplinar em Saúde, Vitória da Conquista BA, Brazil.
[3]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.

Correspondence: Emmanuel Santos Trindade; Email: emmanuelstrindade@gmail.com.

Background: Brain tumors are neoplasms of the Central Nervous System (CNS), characterized by abnormal cell proliferation in the region, which may include the cerebral hemispheres, hypothalamus, thalamus, basal ganglia, brainstem, cerebellum and meninges. According to data from the National Cancer Institute (INCA), 1.4 to 1.8% of all malignant tumors worldwide are in the CNS, with 70.9% of these in the cerebral hemispheres, brainstem and cerebellum. The annual incidence of CNS tumors is 4.85 per 100,000 in females and 5.61 per 100,000 in males. The highest occurrence is in patients over the fourth decade of life, with males being more affected in almost all types of CNS tumors. The symptoms of malignant brain neoplasms vary according to the location and size of the neoplasm, causing anything from headaches and seizures to permanent neurological deficits. **Objective:** To analyze the epidemiological profile of deaths due to brain cancer in Brazil between 2017 and 2021, according to the regions of the country. **Methods:** This is a descriptive study, examining the geographical distribution of deaths due to brain cancer in Brazil and its regions. A detailed analysis of the mortality from this disease throughout the Brazilian territory was presented using data from the Mortality Information System (SIM) in DATASUS for the period from 2017 to 2021, using the variables: processing year, age group, sex, ethnicity and region. **Results:** There were 40,983 deaths reported due to malignant brain neoplasms in Brazil from 2017 to 2021. The Southeast region accounted for the majority of deaths (44.3%), followed by the Northeast (22.3%), South (19.9%), Midwest (7.6%), and North (5.7%) regions. White individuals accounted for the majority of deaths (59.7%) overall. The predominance of white individuals was observed in the Southeast (67.1%), South (87.9%), and Midwest (53.3%) regions. This was not observed in the North and Northeast regions, where brown individuals were the majority, at 65.7% and 58.3%, respectively. There was a slight predominance in males, accounting for 52% of total deaths, which was evident in all regions. Regarding age groups, there was a lower number of deaths in the age group of 10 to 19 years (2.6%) compared to the age group of 0 to 9 years (3.3%). From the third decade onwards, the number of deaths increases until reaching a peak in the seventh decade of life (24.4%), after which the numbers start to decrease again. **Conclusion:** In view of the above, the epidemiological profile of deaths from brain cancer in Brazil between 2017 and 2021 was of male, white and elderly individuals, with the exception of the north and northeast regions, where the majority of deaths were of individuals brown. These findings are in agreement with the main population affected by malignant brain tumors and the advances in prevention, early detection, and treatment in younger patients.

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Epidemiological profile of deaths from hereditary ataxias in Brazil from 2013 to 2022

Larissa da Silva Almeida[1], Guilherme Carvalho de Souza[1], Antonia Cardoso Silva[1], Julio César Soares Barros[1], Alícia Eduarda Rios Soares[1], André de Oliveira Paiva[1], Gabriela Barbosa de Sá Rocha[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.

Correspondence: Larissa da Silva Almeida; Email: larissa.almeida@famed.ufal.br.

Background: Hereditary ataxias are neurological disorders characterized by progressive dysfunction of the cerebellum and its connections, leading to issues with coordination and balance. This group includes subtypes such as spinocerebellar ataxias and Friedreich's ataxia, which can be genetically transmitted in an autosomal dominant, recessive or X-linked inheritance. Besides cerebellar ataxia, patients often exhibit a variety of neurological symptoms, including peripheral neuropathy and visual deficits. **Objective:** This study aims to investigate the epidemiological profile of deaths due to hereditary ataxias in Brazil from 2013 to 2022. **Methods:** This is a retrospective, cross-sectional, quantitative, and descriptive epidemiological study. Data were obtained from the Mortality Information System, focusing on deaths from 2013 to 2022 categorized under Hereditary Ataxia (G11) according to the ICD-10. The variables examined included region, race/ethnicity, sex, age group, and place of occurrence. After collection, the data were tabulated in Microsoft Excel for statistical analysis. **Results:** A total of 1,256 deaths were recorded from 2013 to 2022. Regional distribution included 50 deaths in the North, 260 in the Northeast, 459 in the Southeast, 377 in the South, and 110 in the Central-West. Among the deceased, 848 (67.5%) were identified as white. The distribution between sexes was equal, with 628 deaths each for males and females. The most prevalent age group was 60-69 years, with 265 deaths (21%), followed by the 50-59 years group with 244 deaths (19.4%). Finally, 845 deaths (67.2%) occurred in hospitals or other healthcare facilities, while 383 (30.5%) occurred at home. **Conclusion:** The analysis of the epidemiological profile of hereditary ataxias in Brazil highlights the need for more detailed research, particularly in regions outside the South-Southeast axis. This gap is critical as it contributes to the underreporting of cases in these areas, complicating the diagnosis and therapeutic management of affected patients. Therefore, a detailed epidemiological investigation across all regions is essential to improve clinical support conditions for hereditary ataxias.

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Epidemiological profile of epilepsy hospitalizations among young people in Northeastern Brazil from 2013 to 2023

Italo Lafayette Silva[1], Gilberto de Paula Pinheiro Filho[1], Lucas Roque Evangelista[1], Nicolle Gabrielle Lima Silva[1], Henrique Katsumi Kawano[1], Dayanne Garcia de Freitas[1], Renan de Holanda Alves[1], Renato Vieira de Freitas Filho[1]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Italo Lafayette Silva; Email: italolafayette@gmail.com.

Background: Epilepsy is characterized by a permanent predisposition of the brain to cause epileptic seizures, with significant neurobiological, cognitive, psychological and social repercussions. Hospital admissions are an important public health indicator, reflecting not only the incidence of specific medical conditions, but also epidemiological dynamics over time. This study focuses on the hospitalizations of young people, aged between 15 and 29, in the Northeast region of Brazil, exploring aspects such as geographical distribution, temporal trends and demographic characteristics. **Objective:** This study aims to identify patterns and trends in hospital admissions of young people in the Northeast region of Brazil, offering insights for public health policies aimed at the prevention and treatment of conditions affecting this age group. **Methods:** A cross-sectional, descriptive and quantitative epidemiological approach was used. Data on epilepsy-related hospitalizations in the Northeast region of Brazil, from January 2013 to December 2023, were obtained from the SUS Hospital Information System (SIH/SUS), made available by the Department of Informatics of the Unified Health System (DATASUS). The variables analyzed included information on geographic region, gender, race and age group. **Results:** During the period analyzed, the Northeast region recorded a total of 18,500 hospitalizations for the 15-29 age group. The year 2023 stood out with the highest number of hospitalizations, totaling 2,189 cases, marking a significant increase over the decade. This represents a percentage increase of approximately 45.15% compared to 2013, when 1,508 hospitalizations were recorded. Bahia led the way with the highest number of cases, totaling 4,684 (25.32%) over the period studied, while Sergipe recorded the lowest number of hospitalizations, with 378 (2.04%). The annual average of hospitalizations was approximately 1,682 cases. Together, Bahia and Pernambuco accounted for around 50.10% of the total number of cases analyzed. It is noteworthy that males predominated, with 9,836 (53.15%) hospitalizations, compared to 8,664 (46.84%) for females. In addition, individuals of brown ethnicity were the most frequently hospitalized, with around 11,801 (63.71%) cases. **Conclusion:** This study showed a significant increase in the number of hospitalizations over the decade, with 2023 recording the highest peak. There was a predominance of patients from Bahia and of brown ethnicity; however, there was no significant differentiation by gender among the patients.

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Epidemiological profile of hospitalization for mental and behavioral disorders due to alcohol use in Brazil between the years 2019-2024

Vagner Costa da Cunha Júnior[1], Luana Cristina de Oliveira Lima[1], Maria Cecília Leal Menezes[1], Arthur Vinícius Feitosa Santos[1], Sabrina Sthéphanie Ataíde Martins[1], Thiago Pariz da Silva Dumas[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Vagner Costa da Cunha Júnior; Email: vccjr1@gmail.com.

Background: Alcohol is the legal drug most used by the world population, with repercussions related to symptoms of depression, anxiety and hypomania/mania during periods of intoxication and abstinence. Alcohol dependence has high rates among the user population, reaching a rate of 14.9%, followed by major depression (13%), dysthymia (4.9%), bipolar disorder (2%) and others. **Objective:** To analyze hospitalizations for mental and behavioral disorders due to alcohol use in Brazil and its regions from January 2019 to March 2024. **Methods:** Descriptive epidemiological study using secondary data from the SUS Hospital Information System (SIH/SUS) the Tabnet and data published in the Brazilian Institute of Geography and Statistics (IBGE), being tabulated in a spreadsheet in the Microsoft Excel program, according with data collected from hospitalizations during the period from January 2019 to March 2024. The variables analyzed were federative region, gender, ethnicity and age group. **Results:** In the period in question, there were a total of 161,021 hospitalizations for mental and behavioral disorders due to alcohol use, of which 139,631 (86.72%) were male patients. Furthermore, regarding the other parameters analyzed, it was possible to notice a higher prevalence of the condition under study in the white ethnic population (45.25% of cases), followed by mixed race (32.63%) and people without declared ethnicity (14, 29%), from the South region (2058.61 cases per one million inhabitants), which represents more than double the number of hospitalizations from the Central-West region (second most affected region, with 748.84 per one million inhabitants), and the age group that covers 30 to 59 years, with 124,439 cases (77.28%) in the period analyzed. **Conclusion:** After analyzing the results, it is concluded that the male population, mainly aged between 30 and 59 years old, white and resident in the southern region of the country, has a greater propensity to develop mental and behavioral disorders due to the use of alcohol. This trend can be explained by social factors, such as alcohol culture in the male population, as well as cultural and regional factors, such as German colonization in the South and the large production of alcoholic beverages in this region. All of these factors lead to an increase in alcohol consumption in the population, but end up having undesirable consequences in the long term. Therefore, we can see the need to create mechanisms to prevent alcohol abuse through the government, using public policies to raise popular awareness.

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Epidemiological profile of hospitalization for Parkinson's in Brazil between the years 2014-2024

Luana Cristina de Oliveira Lima[1], Arthur Vinícius Feitosa Santos[1], Vagner Costa da Cunha Júnior[1], Sabrina Sthéphanie Ataíde Martins[1], Maria Cecília Leal Menezes[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Luana Cristina de Oliveira Lima; Email: luana.cristina0410@gmail.com.

Background: Parkinson's disease (PD) is the second most common pathology in the world that causes neural degeneration, second only to Alzheimer's. It causes progressive loss of neurons and its main characteristics are tremors and bradykinesia, which can manifest itself in motor and non-motor forms. PD occurs due to the progressive loss of dopaminergic neurons, leading to a decrease in dopamine, an important neurotransmitter that is present in the substantia nigra in the midbrain and is associated with the basal ganglia and other neurotransmitters, generating system dysfunction due to its lack. Furthermore, this disease manifests itself in all ethnic groups and social classes and, despite being prevalent in elderly people, it can affect young individuals. **Objective:** To analyze hospitalizations for PD in Brazil and its regions during the period from January 2014 to February 2024, through analysis of data from the Department of Information and IT of the Unified Health System (DATASUS). **Methods:** Descriptive epidemiological study using secondary data from the SUS Hospital Information System (SIH/SUS), Tabnet, and data published in the Brazilian Institute of Geography and Statistics (IBGE), being tabulated in Microsoft Excel, according to data collected from hospitalizations during the period from January 2014 to February 2024. The variables analyzed were federative region, gender, ethnicity and age group. **Results:** In the period analyzed, cases of admissions due to PD were 48.66 per million inhabitants in Brazil, with the South region (with 72.6% of its population being white) having the highest rate of hospitalizations - 92.66 per million inhabitants - and the North region (with 20.7% of its population being white) the lowest - 19.94 per million inhabitants. The total number of hospital admissions in the period analyzed was 9881 cases, with a higher prevalence in men (58.76%) and those of white ethnicity (54.04%). It is noteworthy that the majority affected group is made up of whites, mixed race people and those with no information (95.68%) and the most prevalent age group is individuals over 50 years old (89.55%).**Conclusion:** According to the results, it was found that hospitalizations for PD are more prevalent in men of white ethnicity and over the age of 50, factors that may be associated with greater susceptibility of this population to toxic agents, head trauma, hormonal issues, as well as cellular degeneration arising from the most advanced stage of the physiological and natural aging process at this stage of life. Finally, population analysis is important for directing public policies to assist cases of this disease.

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Epidemiological profile of hospitalizations and deaths from multiple sclerosis in Brazil from 2019 to 2023

Caroline Patrícia Amaral Costa[1], Luan Daher Fernandes[2], Luiza da Paz de Souza[2], Suzana Gomes de Oliveira[2], Arley Nikson Oliveira Barreto[2], Moisés Cardoso Portilho[3], Luiz Carlos Figueiredo Filho[2], Luan Nascimento Pereira de Amorim[2]

[1]Universidade Federal do Pará, Belém PA, Brazil. [2]Universidade do Estado do Pará, Belém PA, Brazil. [3]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.

Correspondence: Caroline Patrícia Amaral Costa; Email: caroline.costa@ics.ufpa.br.

Background: Multiple sclerosis (MS) is a chronic autoimmune disease that affects the central nervous system, which includes the brain and spinal cord. In multiple sclerosis, the immune system attacks myelin, a structure formed by a lipid membrane rich in glycophospholipids and cholesterol, which covers axons and is found in the white matter of glial cells. MS can take different forms, the most common being the recurrent remitting form, where symptoms can disappear completely between outbreaks, and the progressive form, where symptoms get progressively worse over time. Despite the fact that this pathology has no cure, treatments are available through the health system in order to improve the quality of life of users of the Unified Health System. **Objective:** To describe the epidemiological profile of hospitalizations and deaths from multiple sclerosis in Brazil between 2019 and 2023. **Methods:** This is an observational, descriptive and retrospective epidemiological study, with a quantitative approach, carried out by collecting secondary data from the Hospital Admissions System (SIH), from DATASUS, referring to each region of Brazil, in the time interval from 2019 to 2023. To build this epidemiological profile, the categories of deaths and hospitalizations were evaluated, as well as average values and their relationship with deaths. In addition, the quantitative data that make up each contingent in the subcategories of sex, race/color and age group were also used. The data was then organized and tabulated in Microsoft Excel 2021 software. **Results:** From 2019 to 2023, 27,586 hospitalizations and 170 deaths

were reported in Brazil, with annual averages of 5,517 and 34 cases, respectively. Of this total, the region with the highest prevalence of cases was the Southeast, with 20,790 (75.36%) hospitalizations and 24 deaths (14.11%). In terms of epidemiological indicators, there were 8,52 hospitalizations (30.89%) and 79 deaths among males (46.47%), and 19,062 hospitalizations (69.11%) and 91 deaths among females (53.52%). As for age group, the highest prevalence was in the 30-39 age group (n=7,739; 28.05%) for hospitalizations, followed by the 60-69 age group (n=47; 27.64%) for deaths. The ethnic group most affected was white, with 15,612 (56.81%) hospitalizations and 85 (50%) deaths. Thus, the most affected population was found to be white females, of economically active age and middle-aged, from the Southeast region. **Conclusion:** The findings highlight that genetic and environmental factors, as possible precursors of MS, are prevalent unevenly across Brazil, highlighting the need for awareness campaigns to promote early diagnosis and appropriate treatment to mitigate the rate of hospitalization and mortality.

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Epidemiological profile of hospitalizations due to Alzheimer's disease between 2019 and 2023

Francisco Edvan Batista Filho[1], Eliezer Paulo Ferreira Júnior[1], Edinaldo dos Santos Ferreira[1], Vitor Gabriel Quaresma de Souza[1], Pedro Antônio Tavares Pinto[1], Mykaell Linhares Arruda de Sousa[1], Thiago da Costa Brito[2]

[1]Universidade Federal do Amapá, Macapá AP, Brazil. [2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Eliezer Paulo Ferreira Junior; Email: eliezerpfjunior7@gmail.com.

Background: According to Caixeta (2012), Alzheimer's disease (AD) is a progressive neurodegenerative disease and the most prevalent of dementias in population-based clinical studies, with a prevalence and incidence of 50 to 75% of cases. This clinical condition is characterized by a gradual decline in intellectual and cognitive abilities. Most cases arise after the age of 65 (late-onset), but some may appear in younger individuals (early-onset Alzheimer's disease). **Objective:** This study aims to analyze the epidemiological profile of patients with AD in the Brazilian population over the past five years (2019 to 2023), corroborating or not with the current literature. **Methods:** This is a cross-sectional, descriptive, quantitative study, using data from January 2019 to December 2023. Data were collected from individuals diagnosed with the condition throughout Brazil, regardless of age group. Data collection was conducted through the Informatics Department of the Unified Health System (DATASUS), analyzing parameters such as total number of cases by region, color/race, sex, age, and deaths. **Results:** In Brazil, there were 7,867 cases of hospitalizations during the period from 2019 to 2023. Regionally, the Southeast was the most affected with 46.70% (n=3,674), followed by the South with 25.55% (n=2,010), Northeast 17.74% (n=1,396), Central-West 6.53% (n=514), and North 3.47%. Analyzing by color/race, the most affected was white with 50.56% (n=3,978), brown 26.42% (n=2,079), and black 4.51% (n=355). Regarding sex, the majority were female with 65.84% (n=5,180) and male with 34.16% (n=2,687). The number of hospitalizations due to AD by age group included patients aged 80 and over, comprising 59.73% (n=4,699). The age group 70 to 79 years represented 27.90% (n=2,195), together comprising 87.63%. Chronologically, there was a gradual increase in the number of hospitalizations per year, with 2019 (n=1,584), 2020 (n=1,199), 2021 (n=1,353), 2022 (n=1,627), and 2023 (n=2,014). During the same period, 1,892 deaths were recorded, with 57.82% in the Southeast region, followed by the South (20.45%), Northeast (11.95%), Central-West (7.61%), and North (2.17%). **Conclusion:** Epidemiological research on "age-related" diseases is essential because early pathological changes occur before symptoms. As observed, AD has a high incidence in Brazil, especially in the Southeast, mainly affecting white women over 70 years old, with a gradual increase over time. The disease, which usually begins late, impairs the quality of life by affecting cognitive functions such as recent memory. Awareness campaigns are crucial for earlier diagnoses due to its progressive nature.

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Epidemiological profile of hospitalizations due to viral meningitis during the COVID-19 pandemic

Bianca Loureiro Maciel Gomes[1], Danilo Silva Nogueira[1], Juliana Alencar Isacksson Vieira[1], Francisco Edvan Batista Filho[1], Luan Henrique Portela de Aguiar[1], Pedro Antônio Tavares Pinto[1], Edinaldo dos Santos Ferreira[1], Thiago da Costa Brito[2]

[1]Universidade Federal do Amapá, Macapá AP, Brazil. [2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Danilo Silva Nogueira; Email: danilo-nogg@hotmail.com.

Background: Viral meningitis is an infectious disease that affects the meninges, membranes that cover the central nervous system. It is caused by viruses, mainly from the Enterovirus group. In 2019, however, the SARS-Cov-2 virus spread globally, showing itself as a new cause of this syndrome due to the presence of the receptors for this pathogen in the cerebral vascular endothelium and the breakdown of the blood-brain barrier, leading to the development of this pathology. **Objective:** The present study is an analysis of the epidemiological profile of patients hospitalized for viral meningitis in the period from 2019 to 2021, which is described as the COVID-19 pandemic. **Methods:** This is a cross-sectional, descriptive, quantitative study, using notification data from all over Brazil, regardless of age group, from January 2019 to December 2021. Data collection was done through DATASUS (Sistema de Departamento de Informática do Sistema Único de Saúde), where the parameters of: total number of hospitalization cases by region, color/race, sex, and age were analyzed. **Results:** In the period from 2019 to 2021, 5,750 cases of hospitalization were notified. Of these, 51.37% (n=2,954) were registered in 2019 only, while in 2020, 26.33% (n=1,514) and 2021, 18.61% (n=1,070). At a regional level, cases in the Southeast region represent 41.74% (n=2,400), in the South region, 25.44% (n=1,463), Northeast, 20.45% (n=1,176), Central-West, 6.40% (n=376) and North, 5.82% (n=335). When comparing the states with the highest and lowest numbers, respectively, São Paulo, 29.98% (n=1,724) and Roraima, 0.09% (n=5) stand out. Among hospitalizations by color/race, 40.31% (n=2,318) correspond to white ethnicity, 33.37% mixed race (n=1,919), black 2.97% (n=171), yellow 1.53% (n=88) and indigenous 0.24% (n=14). As for data by sex, men correspond to 56.56% (n=3,252) and women to 43.44% (n=2,498). The age groups with the highest index were under 1 year old, 20.71% (n=1,191), 1 to 4 years old, 17.29% (n=994) and 5 to 9 years old, 12.49% (n= 718). **Conclusion:** The data reveals that during the COVID-19 pandemic there was a reduction in the number of cases of viral meningitis compared to previous years, with 51.37% of the total reported in the first year of the period, showing a significant decrease in the following two years. Among the regions, Southeast has the highest values, 41.74% of cases, with predominant rates in the state of São Paulo. Conversely, the North region has the lowest index. As for sex, men are more affected than women. In relation to age group, the most affected are children under 1 year old. In terms of race/ethnicity, white, brown, black, yellow and indigenous cases stand out in decreasing order of number. Finally, it is concluded that this reduction in the number of people affected is possibly due to underreporting, affecting regions that lack health services, as well as the mitigation in incidence due to the social isolation regime in place during the period.

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Epidemiological profile of hospitalizations for central nervous system tuberculosis: a half-decade study across regions of Brazil

Luan Nascimento Pereira de Amorim[1], Bruno Kenji Hosoda Mineshita[1], Dangilla Ribeiro dos Santos[1], Fernando Lourenzo Rocha Vianna Oliveira[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Luan Nascimento Pereira de Amorim; Email: luanamorim010@gmail.com.

Background: Tuberculosis is an infectious disease caused by the bacterium Mycobacterium tuberculosis, which has tropism for the lungs, more precisely, the upper lobes. However, upon reaching the bloodstream, the extrapulmonary form can affect other organs, such as the Central Nervous System, and is then considered a severe form of the disease. **Objective:** To identify the epidemiological profile of hospitalizations for neurotuberculosis between regions of Brazil, from 2019 to 2023. **Methods:** This is a descriptive, cross-sectional and quantitative study, based on data from the Hospital Information System tabulated by the Information Technology Department of the Unified Health System (SIH/DATASUS), recorded from 2019 to 2023, in Brazil. Data were collected regarding ICD-10: tuberculosis of the nervous system. The variables for analysis were sex, age group and outcome, in addition to identifying the years and regions with the highest number of hospitalizations which, due to the lack of a population estimate for the year 2023, absolute data were used. Microsoft Excel 2021 software was subsequently used for calculations and analyses. **Results:** 1,629 hospitalizations for neurotuberculosis were recorded in Brazil from 2019 to 2023. Within the period studied, a reduction of approximately 13% in hospitalizations was noted from 2019 (n=325/19.95%) to 2021 (n=283 /17.37%), followed by an increase of approximately 27.62% until 2023 (n=391/24%). Almost half of the hospitalizations were concentrated in the Southeast region (n=770/47.27%), followed by the Northeast regions (n=292/17.93%) and the South (n=286/17.53%). Economically, the average value of hospitalizations for neurotuberculosis increased in the average value in the years 2022 and 2023, exceeding the amount of 3 thousand reais on average for each hospitalization. A predominance of males was identified (n=1068/ 65.56%) overall, it is worth highlighting that this proportion was present in all regions of Brazil. The age group between 30 and 39 years old concentrated the majority of cases (n=332/20.38%), followed by the age group between 40 and 49 years old (n=304/18.66%). In the outcome criterion, 193 deaths were recorded and a mortality rate of 9.21 in the year 2023, with its highest rate in the year 2022 (15.58) according to DATASUS. **Conclusion:** Therefore, the importance of screening for tuberculosis of the central nervous system is noted, given the increase in recent hospitalizations. Furthermore, it is important to understand the regions with the highest concentrations of cases so that it can assist in actions aimed at promoting health. Furthermore, despite the limitations found in this work, such as the lack of population estimates and data on treatment carried out, this does not nullify the research findings, such as the higher prevalence due to the profile: men aged 30 to 39 years, residing in the southeastern region of Brazil.

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Epidemiological profile of hospitalizations for epilepsy in childhood in Brazil from 2019 to 2023

Marcelly Silva dos Santos Brito[1], João Pedro Rosa Barroncaso[2], Yasmim Melo Costa de Azevedo[3], Raul Pereira Bassotto Franco[4], Maria Regina Saddi Guerra[5], Cayara Mattos Costa[6]

[1]Universidade Federal de Pernambuco, Caruaru PE, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade Nove de Julho, São Paulo SP, Brazil.
[4]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.
[5]Universidade Católica de Brasília, Brasília DF, Brazil.
[6]Universidade Federal do Maranhão, São Luís MA, Brazil.

Correspondence: Yasmim Melo Costa de Azevedo; Email: yaxmelo@outlook.com.

Background: Epilepsy is a neurological disease that occurs due to hyperactivity and dysregulation of neural activity, causing disordered electrical discharges. This pathology, which is characterized by recurrent seizures, is one of the most common neurological diseases in childhood and, worldwide, affects about 1 to 2% of the pediatric population. Therefore, the relevance of analyzing the epidemiological profile of epilepsy in children in Brazil is evident, given the prevalence of this pathology in childhood, as well as its negative impact on the quality of life of affected children. **Objective:** To analyze the epidemiological profile of childhood epilepsy in Brazil between 2019 and 2023. **Methods:** This is a cross-sectional, quantitative, descriptive and observational research, with data from January 2019 to December 2023, collected through the SUS Hospital Information System, obtained on the DATASUS platform. The total number of hospitalizations for epilepsy in the age group between 0 and 14 years of age in Brazil were analyzed, considering the variables: region, year, age group, sex, color/race, total hospital expenses, type of care, average stay and deaths. **Results:** Of the total of 110,891 hospitalizations for epilepsy in children aged 0 to 14 years in Brazil, the Southeast was the most affected region (33.6%), and the North the least affected (7.06%). In 2022, there was the highest number of hospitalizations (23,748) and, in 2020, the lowest number (19,604). Of the type of care, 93.5% were urgent. It was identified that brown individuals (50.4%), males (55.2%) and in the age group between 0 and 4 years (42.1%) were the most affected by the disease. The average length of hospital stay, in days, was 5.2, and 611 deaths were recorded in the period analyzed. **Conclusion:** The majority of hospitalizations for epilepsy in childhood occurred in the Southeast, in 2022, on an emergency basis and in brown boys aged between 0 and 4 years. Therefore, the high prevalence of epilepsy in children in Brazil highlights the importance of implementing public policies aimed at preventing situations that cause epileptic seizures and managing these seizures, in order to reduce the number of hospitalizations due to this cause.

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Epidemiological profile of hospitalizations for neurocysticercosis by regions of Brazil 2013-2022

Pedro Ferreira de Souza Evangelista[1], Emmanuel Santos Trindade[2], Indira da Silva Felício[1]

[1]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[2]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Pedro Ferreira de Souza Evangelista; Email: pedro.evvan@gmail.com.

Background: Neurocysticercosis is the most common parasitic infection of the central nervous system (CNS), caused by the Taenia solium larva, known as Cysticercus cellulosae. Man is the definitive host and pigs are the intermediate hosts. When it invades the nervous tissue, the cysticercus undergoes degeneration, accompanied by an intense inflammatory response, which can vary in degree and occur in different places, such as the subdural space, choroid plexus or ventricular wall. This degeneration process can result in intracranial hypertension, dementia, meningitis, hydrocephalus, stroke and permanent neurological damage. **Objective:** To estimate the number of reported cases of neurocysticercosis in Brazil from 2013 to 2022. **Methods:** This is a descriptive study to evaluate the geographical distribution of hospitalizations caused by neurocysticercosis, ICD B69.0, in different parts of the country. It presents a detailed analysis of hospitalizations resulting from this pathology distributed throughout the Brazilian federative units, which were stratified by sex, ethnicity and age group, using data from the DATASUS database, within the SIH/SUS Hospitalization Segment from 2013 to 2022. **Results:** Between 2013 and 2022, 2,534 hospitalizations for neurocysticercosis were recorded in Brazil. 55.92% of hospitalizations were of male individuals. This male predominance occurred in all years, with the exception of 2022, when 51.18% of hospitalizations were female. Regarding ethnicity, there was a higher proportion of hospitalizations of brown (39.82%) and white (28.57%) people at the expense of black (3.87%), yellow (1.74%) and indigenous (0.24%). However, 25.77% of hospitalizations did not present information regarding the patients' ethnicity. The age groups, in descending order of hospitalizations, were 40-49 years old (20.17%), 50-59 years old (18.27%), 30-39 years old (17.40%), 20-29 years old (12.23 %), 60-69 years (12.12%), 70-79 years (7.14%), 10-19 years (6.75%), 0-9 years (3.99%) and 80 years or over (1.93%). It is noticeable that hospitalizations are concentrated in middle age, with a reduction as they approach the extremes of age, which is in line with the natural history of the disease, as, although normally acquired in childhood, clinical manifestations take time to establish themselves, thus requiring hospitalization normally in adulthood. **Conclusion:** Based on the findings, it is possible to notice a predominance of brown, middle-aged male individuals among hospitalizations for neurocysticercosis in Brazil between 2013 and 2022. These findings reinforce those found in the literature, considering it is a predominant disease in rural areas, with precarious hygiene and basic sanitation conditions.

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Epidemiological profile of hospitalizations for Parkinson's disease in brazil from 2012 to 2022

Isabella Queiroz Barbosa[1], Pedro Ferreira de Souza Evangelista[1], Gabriel Lédo Pereira de Oliveira[1]

[1]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.

Correspondence: Isabella Queiroz Barbosa; Email: qbisabella@hotmail.com.

Background: Parkinson's disease (PD) is a neurodegenerative pathology that mainly affects the elderly and whose clinical manifestations involve the motor system with resting tremor and muscle rigidity. This condition has a multifactorial etiology that results in the degeneration of dopaminergic neurons in the substantia nigra. In Brazil, PD contributes significantly to hospital admissions, especially in relation to complications such as falls, severe motor crises and aspiration pneumonia. In addition, its high prevalence among the elderly population increases mortality rates. **Objective:** To analyze the epidemiological pattern of hospitalizations for PD in Brazil from 2012 to 2022. **Methods:** A descriptive analysis was carried out to assess the geographical distribution of hospitalizations caused by PD (ICD G20) in Brazil. The DATASUS database was used, which was segmented by age group, gender and ethnicity, within the Hospital Information System (SIH) segment, from 2012-2022. **Results:** From 2012 to 2022, the number of hospitalizations due to PD increased from 398 to 791 per year, an increase of 98.74%, totaling 7,427 cases in that period. The Southeast region had the highest number of hospitalizations (53.27%), while the Midwest had the lowest (3.54%). In terms of age group, those aged 60 and over accounted for the majority of hospitalizations (73.67%), which indicates a higher prevalence in older age groups and highlights the need for health care for the elderly. In terms of gender, men had more hospitalizations than women in all the years analyzed, with a total of 4,321 (58.16%) for men and 3,106 (41.84%) for women. As for color/race, white people accounted for the majority of admissions (64.94%), followed by brown (29.62%) and black (4%). **Conclusion:** Analysis of the results shows that the increase in hospitalizations during the period in question reflects the increase in the incidence of the disease, possibly due to better identification and recording of PD cases. The geographical distribution shows a more evident concentration in the southeast region, indicating regional disparities in access to health services or in the prevalence of the disease. The predominance of hospitalizations among individuals aged 60 and over highlights the vulnerability of the elderly population to PD. In addition, the differences observed between the sexes, with a higher proportion of hospitalizations among men, and between different racial groups, with a predominance of hospitalizations among white people, suggest that biological and socioeconomic factors may influence the incidence and severity of the disease.

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Epidemiological profile of hospitalizations for viral encephalitis in Brazil from 2014 to 2023

Fernando Gabriel dos Santos Santiago[1], Erick Antonio Rodrigues Mendes[1], Lucas Miquilini de Arrudas Farias[1], Savio Roberto Silva Costa[1], Danusa Neves Somensi[1], Mariangela Moreno Domingues[1], Emanuel de Jesus Soares de Souza[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Fernando Gabriel dos Santos Santiago; Email: fernando_gsantiago@outlook.com.

Background: Viral encephalitis is defined as inflammation of the brain parenchyma with neurological dysfunction of infectious or autoimmune origin, encephalitis has predominantly viral etiology, notably the Herpes virus (HSV-1 and HSV-2), followed by enteroviruses, varicella-zoster and other unknowns, whose frequencies vary in a multifactorial way. A large part of the population is seropositive for most encephalitis pathogens, which, via hematogenic or nervous routes, spread during periods of host immunosuppression, after a state of latency, as in the reactivation of HSV-1, latent in the trigeminal ganglia after primary infection. **Objective:** To determine the epidemiological profile of hospitalizations for viral encephalitis in Brazil between 2014 and 2023. **Methods:** This is a cross-sectional, descriptive and quantitative study, carried out from data extracted from the Hospital Information System of SUS (SIH/SUS) of the Department of Informatics of the Unified Health System of Brazil (DATASUS). The variables used were sex, age group, ethnicity, region, number of services per year, average length of stay and total amount spent. Data analysis was performed using the Biostat 5.3 software. **Results:** There were a total of 19,972 cases of viral encephalitis recorded in the period from 2014 to 2023. The most affected profile was male, in the age group of 1 to 4 years, in the brown cord. The most affected Brazilian region was the North, with 8,279 hospitalizations for viral encephalitis, about 41.45% of the total cases. With the exception of the year 2013, in which only 123 hospitalizations were recorded, this quantity maintained a heterogeneous pattern, with its maximum peak in 2019 with 2,452 records and minimum peak in 2020, with 1,644. The average length of stay fluctuated between 9.2 days, in the Midwest region, and 11.8 days, in the Southeast region, where hospitalizations also had higher mortality. In the analyzed period, about 40 million reais were spent due to these cases. **Conclusion:** This study showed that the profile most affected by hospitalizations for viral encephalitis in

the period from 2013 to 2024 was male, in the age group of 1 to 4 years, in the brown cord. In this 10-year period, cases of viral encephalitis totaled almost 20,000 hospitalizations. These cases were distributed heterogeneously over this period, showing little variation. There was no predominance of greater impact in a specific region, as all were impacted in different ways, such as the North region with the highest number of cases, the Northeast region with the longest stay and the Southeast region with the highest mortality, still showing little variation among themselves. This constant in the distribution of hospitalizations, whether in the temporal aspect or in the regional aspect, demonstrates the magnitude of the importance of this pathology, reinforcing the need to foster its research and treatment.

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Epidemiological profile of hospitalizations for viral meningitis in Brazil between the years 2019-2024.

Arthur Vinícius Feitosa Santos[1], Thiago Pariz da Silva Dumas[1], Vagner Costa da Cunha Júnior[1], Luana Cristina de Oliveira Lima[1], Maria Cecília Leal Menezes[1], Sabrina Sthéphanie Ataíde Martins[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Arthur Vinicius Feitosa Santos; Email: arthur.vinicius04@souunit.com.br.

Background: Viral meningitis is a neurological condition that generally evolves benignly. It usually presents with an acute onset of fever, headache, photophobia, neck stiffness and, frequently, nausea and vomiting, although younger children may not show signs of meningeal irritation. It can occur alone or in outbreaks and affects people of all ages, being more common in children under five years of age. Approximately 85% of cases are caused by Enteroviruses, including Poliovirus, Echovirus and Coxsackievirus. **Objective:** To analyze hospitalizations due to viral meningitis in Brazil during the period from January 2019 to March 2024. **Methods:** Descriptive epidemiological study using secondary data from the SUS Hospital Information System (SIH/SUS), Tabnet, and data published in the Brazilian Institute of Geography and Statistics (IBGE), being tabulated in Microsoft Excel, according to data collected on hospitalizations and deaths during the period from January 2019 to March 2024. The variables analyzed were federative region and age group. **Results:** In the period analyzed, the rate of hospitalizations for viral meningitis was 50.76 per million inhabitants in Brazil. The data demonstrated that children aged 0 to 9 years represent 51.98% of total cases of the disease, the prevalence of which decreases with increasing age of the population, whereas the elderly (people aged 60 or over) represented only 4.27 % of cases. Furthermore, it should be noted that cold weather is a factor that contributes to greater infectivity of the meningitis virus. That said, in the Northeast and North regions, 40.20 and 35.57 cases were recorded per million inhabitants, respectively, while in the South region 89 cases were recorded per million inhabitants. **Conclusion:** According to the analysis of the results, the younger population has a higher prevalence (51.68% of all cases) of hospitalizations for the disease. Also, it is possible to conclude that viral meningitis is related to an immature immune system, as the number of cases decreases drastically with increasing population age. Additionally, it is necessary to reinforce that this situation occurs equally in all brazilian states, showing that it is associated with viral meningitis. Furthermore, the literature demonstrates that there is a clear tendency for people to stay together at home, especially during colder periods. This behavior, along with the decrease in the effectiveness of the immune system due to the cold, corroborates the increased infectivity of viruses. This is evidenced by the fact that the number of hospitalizations in the Southern states more than doubles compared to those in the North and Northeast. Finally, it is important that public policies are created aimed at children in the fight against viral meningitis, focusing mainly on those who live in cold regions of the country.

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Epidemiological profile of meningitis in Brazil from 2014 to 2023

Daniel Rodrigues Lemos Neto[1], Mariana Leandro Silva[1], Maria Kaylanne Pinheiro de Araújo[1], Érycle Guilherme da Silva[1], Gabriela Goveia de Sousa Castilho de Lima[1], Luan Santana Martins da Costa[1], Filipe Alves da Costa Dias[1], Francisco Orlan de Souza Neto[1], Wagner Gonçalves Horta[2], Sarah Ellen Alves Soares[1]

[1]Faculdade de Medicina de Olinda, Olinda PE, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Daniel Rodrigues Lemos Neto; Email: danieldede3@hotmail.com.

Background: Meningitis is included in the National List of Compulsory Notifiable Diseases in Brazil and is defined as an inflammatory process affecting the membranes surrounding the brain and spinal cord. It presents with meningeal syndrome characterized by neck stiffness, fever, and headache, and may also show signs of meningeal irritation such as Brudzinski and Kernig signs. **Objective:** To outline the epidemiological profile of meningitis in Brazil from 2014 to 2023. **Methods:** This is an epidemiological, descriptive, and retrospective study conducted through the electronic platform from the Informatics Department of the Public Unified Health System (DATASUS). The population assessed included all patients affected by meningitis in Brazil over the past 9 years. To achieve the study objectives, the information from the patients was categorized according to age, sex, race, and location. The information used in this epidemiological study was obtained from a secondary database, and submission to an Ethics Committee was not required. **Results:** Various etiologies were evaluated and compared by age group, sex, and race over the past 9 years: meningococcemia (MCC), meningococcal meningitis (MM), meningococcal meningitis with meningococcemia (MM+MCC), tuberculous meningitis (MTBC), meningitis due to other bacteria (MB), unspecified meningitis (MNE), viral meningitis (MV), meningitis due to other etiologies (MOE), Haemophilus meningitis (MH), and pneumococcal meningitis (MP). During this period, a total of 144,001 cases of meningitis were diagnosed. Of note, viral meningitis (MV) predominated across all age groups from <1 year to 64 years, accounting for 63,590 cases (~44.16%), predominantly affecting males with 84,544 confirmed cases (~58.72%), and the black race with 67,466 confirmed cases (~46.9%). **Conclusion:** Regarding meningitis and its main subtypes, the epidemiological study revealed that viral meningitis was the most prevalent in Brazil over the past 9 years. Additionally, the disease was more common in males and among individuals of black race. These findings underscore the need for more active epidemiological surveillance and health promotion and prevention actions targeted at the most affected groups.

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Epidemiological profile of meningitis in the South region of Brazil from 2019 to 2023

Maria Regina Saddi Guerra[1], Raul Pereira Bassotto Franco[2], Yasmim Melo Costa de Azevedo[3], Marcelly Silva dos Santos Brito[4], João Pedro Rosa Barroncas[5], Cayara Mattos Costa[6]

[1]Universidade Católica de Brasília, Brasília DF, Brazil.
[2]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.
[3]Universidade Nove de Julho, São Paulo SP, Brazil.
[4]Universidade Federal de Pernambuco, Caruaru PE, Brazil.
[5]Universidade Federal do Amazonas, Manaus AM, Brazil.
[6]Universidade Federal do Maranhão, São Luís MA, Brazil.

Correspondence: Maria Regina Saddi Guerra; Email: mreginasaddi@gmail.com.

Background: Meningitis is an inflammation of the membranes covering the brain and spinal cord, mainly affecting the arachnoid space and occasionally impacting the cranial or spinal regions. Without treatment, meningitis has a fatality rate (TL) between 50% and 80%. With treatment, this rate is reduced to around 15%. Infectious meningitis represents an important challenge for Brazilian public health, and are diseases for which notification is mandatory. **Objective:** To detail and investigate the epidemiological profile of meningitis cases reported in southern Brazil between 2019 and 2023. **Methods:** This is an observational, descriptive and quantitative ecological study, with data collected from the Notifiable Diseases Information System (SINAN), on the DATASUS platform, in May 2024. The variables were investigated: total notifications, notifications from each federative unit, sex, age group, color or race, level of education, frequent etiology and evolution throughout the region (Rio Grande do Sul, Santa Catarina and Paraná) between January 2019 and December 2023. **Results:** Between 2019 and 2023, 13,686 cases of meningitis reported in the southern region were identified. Of these, 6,345 (46.3%) were in the state of Paraná, making it the most affected area, 3,400 (24.84%) in Santa Catarina and 3,941 (28.8%) in Rio Grande do Sul. The data indicate a higher incidence among men (58.32%), children under 1 year old (19.22%) and 40-59 years old (17.70%), and predominantly white (78.95%). Regarding education, 10% of those affected had completed high school. The most common etiology was viral (44.41%). Regarding the outcome, 78.94% were discharged from hospital. **Conclusion:** Public policies aimed at preventing meningitis must be widely implemented. In the South region, they should be mainly focused on the state of Paraná, on white children under 1 year of age. Early diagnosis is important for a hospital discharge prognosis.

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Epidemiological profile of multiple sclerosis in Brazil: an analysis of the last 5 years (2019-2023)

Anna Gabriella Venâncio Neves de Moraes Tosta[1], Aline Marques Barreto Scalioni[2], Giovana Ank Alves Ovídio[3], Isabella Pasqualotto[4], Clarice Arruda Villari[5], Beatriz Helena de Mattos Araújo Verri[6]

[1]São Leopoldo Mandic, Campinas SP, Brazil.
[2]Universidade Nove de Julho, Guarulhos SP, Brazil.
[3]Afya Faculdade de Ciências Médicas de Ipatinga, Ipatinga MG, Brazil.
[4]Centro Universitário de Várzea Grande, Várzea Grande MT, Brazil.
[5]Universidade de São Paulo, São Paulo SP, Brazil.
[6]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Anna Gabriella Venâncio Neves de Moraes Tosta; Email: annagabriellatosta@gmail.com.

Background: Multiple sclerosis (MS) is a chronic disease that causes the demyelination of the myelin sheath, it is an autoimmune disease, with its etiology being diverse and still heavily researched. MS causes neurological disabilities and affects young patients, most of them in the age range of 20 to 40 years. It causes a variety of symptoms,

such as fatigue, muscle weakness and paresthesias, as well as visual, cognitive, and mental alterations, and presents in three forms: relapsing-remitting, secondary progressive, and primary progressive. The first type being present in 85% of cases. For MS diagnosis, it is used the McDonald's 2017 criteria involving clinical aspects, imaging, and analysis of cerebrospinal fluid with specific markers. **Objective:** To analyze the epidemiological characteristics of the patients affected by multiple sclerosis between 2019 and 2023. **Methods:** Ecological and descriptive study, using data collected from the Brazilian Unified Health System Hospital Information. Were collected informations on sex, race, region of origin, type of care (elective or emergency), hospitalization rate, and mortality for the years 2019 to 2023. **Results:** There were 27,577 hospitalizations for Multiple Sclerosis (MS) in Brazil, with the year 2022 having the highest number of cases (26.46%, n=7,299). These hospitalized patients were mostly in emergency care (52.31%, n=14,427). The Southeast region presented the highest frequency (75%, n=20,786) of cases, while the North region was the least frequent (1.33%, n=367). The analysis revealed a higher occurrence in females (69.10%, n=19,057), with the most affected age group being between 30 and 39 years (28.04%, n=7,735). Whites accounted for 57% of hospitalizations (n=15,609), while those of indigenous origin accounted for only 0.01% (n=4). In addition, it is worth noting that 10% of cases (n=2,767) did not contain information on race. The average mortality rate of MS was 0.61%, peaking in 2018 with a representation of 1.13%, and the lowest number in 2022 with 0.45%. Regarding the mortality rate by age group, there is a gradual and direct proportional increase with age, the highest percentage is in patients who are over 80 years old at 16% and the lowest percentage is between 15 and 29 years old at 0.13%. **Conclusion:** Between 2019 and 2023, there are significant differences in the mortality rate and hospitalizations due to Multiple Sclerosis depending on the year of registration. Age is an aggravating factor and there is a big discrepancy depending on region. Regarding race, the possibility of information bias related to databases should be considered, with possible underreporting and/or differences in access to healthcare services among the studied populations. However, further studies are needed to understand the relationship between epidemiological data and patient characteristics in order to advance treatment, diagnosis, and patient support.

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Epidemiological profile of multiple sclerosis inpatients in Brazil in the last 10 years

Tainá Reis Martins[1], Wívian Maria Pires Figueredo[1], Mariana Guimarães Mundim[1], Karina Andrade de Prince[1]

[1]Centro Universitário FIPMoc, Montes Claros MG, Brazil.

Correspondence: Tainá Reis Martins; Email: taina2002@hotmail.com.

Background: Multiple sclerosis (MS) is an immunomediated, demyelinating and neurodegenerative disease of the central nervous system. The first symptoms of MS tend to appear between 20 and 40 years of age, representing the main cause of non-traumatic neurological disability in young adults. In Brazil, it is estimated that about 40,000 individuals live with the disease, but there are weaknesses and obstacles in the Brazilian epidemiological data due to late diagnosis, the difficulty of classifying the disease and the limited access to centers specialized in the diagnosis and treatment of MS. **Objective:** To analyze the epidemiological profile of patients who required hospitalization for MS in the last 10 years in Brazil. **Methods:** This is a descriptive, retrospective, ecological time series study. It had as a research universe the database of the Hospital Information System of SUS (SIH/ SUS), made available by the Department of Informatics of SUS (DATASUS) number of hospitalizations and hospital expenses per MS in Brazil from January 2014 to December 2023. The variables analyzed were gender, hospital cost, nature of care, average hospitalization rate and deaths. **Results:** In the period analyzed, a total of 43,950 hospitalizations were reported, with a higher number recorded in 2022 (16.6%), and a significant increase in the number in 220% (4,464) in the last 10 years. Hospitalizations predominated in females, representing 69.6% (479,643), that is, 2.2 times more than men, being the common age group between both sexes more affected from 30 to 39 years, with 28.7% (12,645). The region with the most hospitalizations was the southeast with 70.5% (31,006). The mortality rate over these years decreased 11.9% (9.4 to 8.3), in contrast, it is noteworthy that the highest mortality rate occurred in males, with index 1.1 and rate approximately 103% higher than for women. In addition, the group with the highest mortality was 80 years or more, with an index of 11, and the northern region with the highest mortality rate, with an index of 2.6. Regarding the nature of the visits, 53.7% (23,617) of the visits were urgent. The highest total number of hospitalizations was in 2023 (R$ 3,649,310.85), with an increase from 2014 to 2023 of 118% (R$ 1,980,724.00). The female gender comprised 66.5% (R$ 16,498,959.12) of the expenses of the total value. The average value of hospitalization decreased 31.8% (R$ 262.89). **Conclusion:** The epidemiological profile more incident were women from 30 to 39 years old. Currently the number of hospitalizations and the total value cost is increasing, while the average values per hospitalization decrease significantly. Although they are not the most affected population, residents of the North Region and males have higher mortality, and the reasons for this should be analyzed, either by precarious care, availability of resources, and biological susceptibility or self-neglect.

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Epidemiological profile of muscle disorders: insights from a specialized service in São Paulo

Filipe Tupinambá Di Pace[1], Cristiane Araújo Martins Moreno[1], Clara Gontijo Camelo[1], Raquel Diógenes Alencar Sindeaux[1], Karlla Danielle Ferreira Lima[1], Marco Antônio Veloso Albuquerque[1], Alullin Fonseca[1], Rodrigo de Holanda Mendonça[1], André Macedo Serafim Silva[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Filipe Tupinambá Di Pace; Email: filipedipace@gmail.com.

Background: Neuromuscular diseases (NMDs) are conditions characterized by primary dysfunction of muscles, neuromuscular junction, nerves or motor neurons. Understanding the distribution and frequency of these conditions is important for improving diagnostic and therapeutic approaches. **Objective:** To provide an epidemiological profile of patients attending a specialized muscle service. **Methods:** Data obtained between November 2020 and February 2024 were automatically extracted from electronic medical records and revised by an examiner. Key data points included patient registration, birth date, sex, genetic test results, disease topography, and diagnosis. **Results:** There were 5160 appointments and a total of 1526 patients, of them, 55% were male, the mean age was 31(0-88) years, and 62.6% were adults. The muscle was the most frequent topography (63%), and the top diagnosis was dystrophinopathy (24.8%). Regarding other muscle diseases, the most common diagnoses were congenital muscular dystrophy (11.3%), mitochondrial myopathy (8.9%), congenital myopathy (8.4%), and limb-girdle muscular dystrophy (7.9%), and when a genetic test was conclusive mostly of them were respectively related to LAMA2, TK2, RYR1, and DYSF. Myotonic dystrophy accounted for 7.4% of cases, all of them due to DMPK expansion. Facioscapulohumeral muscular dystrophy (3.3%) included one patient with type 2 disease. Metabolic myopathies (2.3%) and channelopathies (2.2%) were mainly caused by glycogenosis and congenital myotonia, respectively. Other inherited myopathies made up 2,8% of the cases, mostly myofibrillar myopathy secondary to DES pathogenic variant, and 6 patients with hereditary inclusion body myositis, including 4 with the pathogenic variant in ANXA11. Inflammatory myopathy was the most frequent non-genetic diagnosis (3.8%), and an atypical presentation and/or family history motivated a genetic test in 15.5% before the final diagnosis. Congenital myasthenia (2.9%) was mostly related to CHRNE pathogenic variants. Although the clinic is aimed at myopathies, other neuromuscular conditions were also seen during this period: Inherited neuropathies (4.2%) mostly included Charcot-Marie-Tooth disease (54%) and acute hepatic porphyria (28%). For motor neuron diseases (13%), spinal muscular atrophy was the predominant diagnosis (72.3%), followed by amyotrophic lateral sclerosis and non-5q spinal muscular atrophy. An undefined diagnosis was present in 14.1% of cases, in which 43.5% had at least one inconclusive genetic test. **Conclusion:** The data offers valuable insights into the epidemiology of muscle diseases in São Paulo. The high rate of genetic diseases and the significant proportion of undefined diagnoses underscores the need for advanced genetic testing with availability to the population, and research to identify novel mutations. Improved diagnostic tools and therapeutic strategies tailored to the specific profiles of the local population could enhance patient outcomes.

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Epidemiological profile of Parkinson's disease in Brazil: an analysis of the last 10 years

Victória Stephane Tessaro[1], Larissa Paixão Batista[2], Sofia Volpi[3], Laura Gabriela Ribeiro Reis[4], Julia Cavalcanti Brito[5], Karoline Schereck Kreitmeyer[6]

[1]Universidade de Fortaleza, Fortaleza CE, Brazil.
[2]Escola Superior de Ciências da Saúde, Brasília DF, Brazil.
[3]Universidade Federal do Rio Grande, Rio Grande RS, Brazil.
[4]Centro Universitário FIPMoc, Montes Claros MG, Brazil.
[5]Universidade Federal do Vale do São Francisco, Petrolina PE, Brazil.
[6]Santa Casa de Misericórdia de Limeira, Limeira SP, Brazil.

Correspondence: Victória Stephane Tessaro; Email: stephanetessaro.2020@gmail.com.

Background: Parkinson's disease (PD) is a degenerative, chronic, and progressive disorder of unknown etiology that affects the central nervous system. Its primary symptoms include bradykinesia, tremors, muscular rigidity, and postural instability. The main cause of the disease is the death of cells in the substantia nigra, which produces dopamine, a neurotransmitter that controls movement. Studies indicate that the incidence of PD increases with age. **Objective:** To characterize the epidemiological profile of PD in Brazil over the past 10 years. **Methods:** A retrospective, descriptive, and cross-sectional epidemiological analysis was conducted using data provided by the Department of Informatics of the Unified Health System of the Ministry of Health (DATASUS/MS). **Results:** The results showed that PD accounted for a total of 629 deaths and 9,991 hospitalizations in the last 10 years, with a notable concentration in the southern and southeastern regions, and its peak incidence occurred in 2023. The age group with the highest number of deaths was 80 years and older, with a significant mortality rate of 14.65%, while the age group of 70 to 79 years surpassed others in hospitalizations (2,645). There was a higher prevalence of mortality among females (6.82%), particularly in the northern region, whereas males predominated in hospital admissions (5,876), especially

in the southeastern region. The Black (13.06%) and Yellow (7.83%) races also exhibited higher rates of morbidity and mortality compared to other racial groups. **Conclusion:** It can be concluded that mortality rates were higher among elderly individuals aged 80 years and above, particularly in females and in the northern region. Conversely, hospitalization rates were higher among males in the 70 to 79 age group, especially in the southeastern region. Additionally, morbidity and mortality were higher in non-White races. These findings are important for informing public health policy decisions regarding PD based on the observed trends during this period.

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Epidemiological profile of patients hospitalized for tetanus in Brazil over the last 10 years

Pedro Henrique de Vilhena[1], Júlia de Oliveira Bessegatto[1], Alisson Rafael de Oliveira Pereira[1], Rodrigo Ferreira de Moura[1]

[1]Universidade Federal de Lavras, Lavras MG, Brazil.

Correspondence: Pedro Henrique de Vilhena; Email: phdvilhena@gmail.com.

Background: Tetanus is an acute poisoning caused by the neurotoxins produced by Clostridium tetani. Infection occurs by the inoculation of spores into the skin or mucous membranes, mainly through wounds. Although the treatment of the disease can be carried out with support, antibiotic therapy, and neutralization of the toxin, only the complete vaccination schedule followed by reinforcement with adult bacterial duo (adult dT) every 10 years confers lasting immunity. Thus, knowledge of the epidemiological panorama of patients hospitalized for tetanus may indicate the population profile most vulnerable to infection. **Objective:** To analyze the epidemiological profile of patients hospitalized for tetanus in Brazil. **Methods:** A retrospective study was conducted using secondary data recorded in SINAN, SI-PNI, and SIH available in DATASUS from 2013 to 2022, analyzing notifications, hospitalizations, sex, color/race, age group, type of care, deaths, average hospitalization costs, and adult dT vaccination coverage. **Results:** A total of 2291 cases of accidental tetanus were confirmed in the study interval, 1825 (79%) hospitalizations and 335 (15%) deaths. The average number of cases per year was 230, with 90% of the cases being urgent. The total cost of hospitalization was R$11,321,436, about R$6,203 per hospitalization. Spending on males represents 85% of this amount, which is explained by the proportion of cases by gender, with 4.4 times more cases occurring in men. The average expenditure/hospitalization rate maintained greater proximity between the sexes. In terms of race/color, there was a change in trend with a reduction in hospitalizations by whites in 2020 (APC: -32.04, 95%CI: -45 to -11, p<0.001). A reduction in the number of cases was observed in the "no information" group, with an increase in the brown group and a small peak in 2019 in the black group. Thus, of the total set, 26% of the cases did not have identification information, whereas the most affected identified themselves as brown (42%) and white (27%). As for the regional analysis, Minas Gerais (MG), Rio Grande do Sul (RS) and Paraná (PR) together represent 27% of the country's cases, while Roraima (RR) and Tocantins (TO) have the lowest number (<1%). By extension, MG and RS had the highest number of deaths, although when analyzed proportionally, RR, Amapá (AP) and RO reached lethality rates above 50%. These data also reflect adult dT vaccination coverage. The number of confirmed cases was negatively correlated with adult dT vaccination coverage (Kendall= -0.23; p<0.08). The age groups 50-59 years (24%) and 60-69 years (20%) groups were more hospitalized and received the lowest adult dT booster doses. **Conclusion:** Brown men aged between 50 and 69 years living in MG and RS in emergency care represent the social group with the lowest vaccination coverage and the greatest vulnerability to tetanus.

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Epidemiological profile of patients who death from Alzheimer's disease in Salvador – Bahia: 2013 to 2023

Ana Beatriz de Oliveira Andrade[1], Juarez Pereira Dias[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Ana Beatriz de Oliveira Andrade; Email: anabeandrade@hotmail.com.

Background: With the aging of populations, chronic non-communicable diseases gain notoriety, especially dementia - syndromes with a progressive and generalized decline in cognitive functions, the most recurrent being Alzheimer's disease (AD), an irreversible degenerative neurological disease, with behavioral and affective disorders. **Objective:** To analyze the epidemiological profile of patients who died from AD in Salvador - Bahia in the period from 2013 to 2023. In addition to describing deaths by sex, age group, annual distribution, temporal trend, spatial distribution by health district and analyzing characteristics. **Methods:** A descriptive study was conducted with secondary data obtained from the Mortality Information System on the website of the SUS Department of Information Technology. The variables used were: number of cases per year of notification and health district of residence, sex, age group, race/skin color and education. The variables were analyzed according to their absolute and percentage distribution. Pearson's Chi-Square test and Simple Linear Regression were used, with statistical significance set at p<0.05. Statistical storage and analysis were performed using the Statistical Package for Social Sciences. **Results:** During the study period, 3,087 deaths from AD were recorded in the population over 60 years of age in Salvador. Most deaths occurred in females, 2,248 (72.56%), and in the age group over 80 years, 2,471 (79.76%). The mortality rate increased by 286.01% during the study. The Health District with the highest average rate was the Historic Center (38.35/100,000 inhabitants). There was an increase as the age groups advanced, with the majority of those affected being over 80 years of age (99.13/100,000 inhabitants). There was an increasing trend between the sexes, with a prevalence of males among the youngest and females over 80 years of age. The highest frequency of deaths was in the white race/skin color, 1,481 (47.80%). In addition to the prevalence among those who had eight to eleven years of schooling, 733 (23.66%). **Conclusion:** AD is a major public health problem. Its increasing incidence in recent years is a reflection of aging, better access to resources and information, and improvements in diagnostic methods. In this scenario, early diagnosis, training of health professionals, clinics and hospitals, and technological advances in the scientific field are imperative.

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Epidemiological profile of patients with Guillain-Barré syndrome from 2013 to 2023 in the Federal District, Brazil

Maria Regina Saddi Guerra[1], Fernanda Massue Komatsu Rabelo[1], Fernanda Carneiro e Cardoso[1], Maria Fernanda Alves Barros[1], Mariana Santos Vogel[1], Ana Clara Monte Varandas[1], Cayara Mattos Costa[2]

[1]Universidade Católica de Brasília, Brasília DF, Brazil.
[2]Universidade Federal do Maranhão, São Luís MA, Brazil.

Correspondence: Maria Regina Saddi Guerra; Email: mreginasaddi@gmail.com.

Background: Guillain-Barré syndrome (GBS) is an autoimmune disease characterized by demyelination or axonal degeneration of peripheral nerves, resulting in muscle weakness or loss of sensation. In Brazil, from 2008 to 2014, were recorded 15,512 hospitalizations, showing the relevance that this syndrome has on the health of the Brazilian population, as it not only compromises patients' quality of life, but also has a considerable impact on the health system due to the need for prolonged hospitalizations and intensive care. **Objective:** Describe the epidemiological situation of GBS in the Federal District (DF) between the years 2013-2023. **Methods:** Ecological time series study using data from the "Sala de Situação-Infosaúde" platform of the Health Department of the Federal District in the Hospital Information System database, CID G61.1 (Guillain-Barré syndrome) from 2013 to 2023. The variables investigated in the Federal District (DF) were: total hospitalizations, year of prevalence, nature of the intervention, gender, age group, length of stay, prevalence among hospitals and procedures. **Results:** In the period analyzed, there were 460 hospitalizations, with an average of 48.2 hospitalizations per year. From 2013 to 2017, the average number of hospitalizations was 32 per year, however there was a sudden increase in hospitalizations in 2018, a pattern that remained constant until 2023, a period in which the average was 50 per year. Regarding the nature of the intervention, there is a predominance of urgency (90.65%) over elective (9.35%). In the gender variable, there was a higher rate of male patients (58.7%) compared to female patients (41.3%). In terms of hospitalization authorizations by age group, there was a higher prevalence in the 15 to 19 year old range (47). The Ceilândia Regional Hospital has the longest average length of stay (27.5 days), followed by the Guará Regional Hospital with (18.07 days). The highest number of hospitalization authorizations per health establishment was at Hospital de Base do Distrito Federal (267), followed by Hospital Regional da Asa Norte (34). In relation to hospitalization authorizations by administrative region of residence, there was a predominance of those from the Ceilândia region (52) and in relation to those residing outside the DF, there was a predominance of inhabitants of the municipality of Luziânia (13). The most commonly performed procedures were the treatment of polyneuropathies (347) and Acute Demyelinating Polyradiculoneuritis (81). **Conclusion:** The study showed the predominance of hospitalizations for GBS in the Federal District from 2013 to 2023: men, between 15 and 19 years old, with elective hospitalization, at the Hospital de Base of the Federal District and with the treatment procedure of polyneuropathies. This study highlights the need for new actions aimed at intervention and treatment of this disease, both acute and late management with sequelae.

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Epidemiological profile of patients with headache in neurology outpatient clinics in the Southern region of Santa Catarina

Camila Pezente Dal Toe[1], Debora Dagostin Casagrande[1], Tatiana Pizzolotto Bruch[1], Natalli Studnicka[1], Maria Laura Zanette Naspolini[1], Natália Effting Crema[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Camila Pezente Dal Toé; Email: camilapezente@hotmail.com.

This study aimed to evaluate the clinical and epidemiological profile of patients with headache treated at neurology outpatient clinics of the Brazilian Unified Health System (SUS) in the cities of Criciúma, Forquilhinha, and Içara, Santa Catarina. A total of 212 medical records were analyzed, predominantly from female patients (77.4%) with a mean age of 47.01 years. The majority of cases (87.7%) were classified as primary headaches, with migraine being the most common form (31.1%), followed

by Tension-Type Headache (5.7%). The research did not find significant abnormalities in imaging studies related to headache. Regarding the association with COVID-19, only 32.4% of patients reported worsening or onset of headache after contact with the virus. These findings suggest that primary headaches, particularly migraine, are prevalent in trained neurology outpatient clinics, with little evidence of significant impact. This study contributes to understanding the profile and clinical characteristics of headache in the population served by SUS in the southern region of Santa Catarina, emphasizing the importance of epidemiological studies to guide public health policies.

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Epidemiological profile of patients with multiple sclerosis from 2013 to 2023 in the Federal District, Brazil

Maria Regina Saddi Guerra[1], Marcela Pontes Paulo[1], Sarah dos Reis Alves de Paiva[1], Guilherme Lacerda Oliveira[1], Vinícius Turial Assis dos Santos[1], Gabriel Ferreira de Jesus Malta[1], Cayara Mattos Costa[2]

[1]Universidade Católica de Brasília, Brasília DF, Brazil.
[2]Universidade Federal do Maranhão, São Luís MA, Brazil.

Correspondence: Maria Regina Saddi Guerra; Email: mreginasaddi@gmail.com.

Background: Multiple sclerosis (MS) is an autoimmune neurological condition characterized by an abnormal immune response that causes inflammation and damage to the myelin sheaths of the central nervous system. Today, the disease affects 2.8 million people worldwide and it is estimated that 40 thousand people have this disease in Brazil. Due to the multiplicity of symptoms and long-term debilitating consequences, the study of MS is essential for the technical and theoretical improvement of the disease. **Objective:** To outline the epidemiological profile of patients diagnosed with MS in the Federal District from 2013 to 2023. **Methods:** This was an ecological time-series study using data from the public platform "Sala de Situação-Infosaúde" of the Health Department of the Federal District in the database of the Hospital Information System (SIH) of ICD G35 (multiple sclerosis) between the years 2013-2023. The total number of hospitalizations, age group, sex, prevalence among administrative regions, the nature of the intervention, the average length of stay and the cost were analyzed. **Results:** There were 2437 hospital admissions due to MS in the observed period, of which 74.39% were female and 25.61% male. The age distribution prevails in the range of 20 to 49 years of age (80.18%), with emphasis on the group of 40 to 44 years of age (17.23%). The origin of MS patients is concentrated in four administrative regions of the Federal District (35.82%): Ceilândia, Taguatinga, Samambaia and Plano Piloto, respectively, in order of prevalence. From 2013 to 2018, the number of hospitalizations increased progressively, with 782 in 2018 (32.08%). In 2019, there was a sudden drop in the number of hospitalizations (27), which continued throughout 2020 (25) and only recovered between 2021 (241) and 2022 (506), most likely as an effect of the spread of the SARS-CoV-2 virus. In 2023, the values decrease again with 31 hospitalizations, possibly arising from administrative and operational changes at SES-DF. Of the total number of hospitalizations, 58.6% were urgent and 41.4% were elective, with a single hegemonic prevalence in clinical beds (99.17%) and an average length of stay of 1.56 days. Costs totaled 615,587.48 reais during the period. **Conclusion:** The epidemiological profile of the patients was, for the most part, between the ages of 40 and 44, female, residents of administrative regions with a high population, with elective care and length of stay of 1.56 days. The importance of public policies aimed at caring for patients with MS, as well as specialized support centers, is highlighted.

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Epidemiological profile of patients with neurotuberculosis in Brazil over the last 5 years

Marcela dos Santos de Deus[1], Mariana Aires Silva[1], Pedro Keemper Rocha Alpino[1], Jéssika Yumi Taba Ono[1], Maria Clara Caparroz Cassioti[1], Maria Luiza Leme Camargo[1], Danielli Casonato da Silva[1], Luah da Silva Ishikawa Manhani[1], Luana Fornetti Castilho Cazani[1], Maiara Silva Tramonte[1]

[1]Universidade Nove de Julho, Bauru SP, Brazil.

Correspondence: Marcela dos Santos de Deus; Email: marcela.dsdd@uni9.edu.br.

Background: Neurotuberculosis is a bacterial infection caused by Mycobacterium tuberculosis, affecting the Central Nervous System (CNS). The dissemination occurs when the bacteria enter the bloodstream and cross the blood-brain barrier. CNS TB commonly occurs in individuals previously treated for pulmonary TB years earlier, due to reactivation of latent infection. The primary infection, usually acquired by inhaling droplets containing bacilli, can lead to dissemination to the meninges and brain surface. Bacilli can remain latent in tubercles, which may rupture into the subarachnoid space, triggering tuberculous meningitis, the main clinical presentation of neurotuberculosis. **Objective:** Analysis of the epidemiological profile of patients with neurotuberculosis in Brazil over the past 5 years. **Methods:** This is a descriptive and retrospective epidemiological study with quantitative analysis between 2019-2023. Data were obtained from the Department of Health Informatics of the Unified Health System (DATASUS) and articles available in the National Library of Medicine (NIH) PubMed. The analysis considered variables such as region, sex, race/ethnicity, age group, number of deaths, and mortality rate. Data were collected in April 2024. **Results:** During the analyzed period and location, 1,629 hospitalizations of patients diagnosed with neurotuberculosis were recorded. There was a variable trend over the years, with a progressive decrease in cases in 2019 (325), 2020 (322), and 2021 (283), followed by an increase in 2022 (308) and 2023 (391). The southeast region registered the highest number of hospitalizations (770), followed by the northeast (292) and south (286) regions. Among ethnic groups, individuals of mixed race (755) were the most affected, followed by white (497) and black individuals (113). Regarding sex, men accounted for the majority of hospitalizations, totaling approximately 65.6%. In terms of age group, the most affected groups were those aged 20-29 years (233), 30-39 years (332), 40-49 years (304), and 50-59 years (259). The mortality rate per hospitalization was 11.8%. Furthermore, it is noteworthy that the mortality rate remains higher in the southeast region (13.51), among individuals of mixed race (11.39), although it is higher in females (12.48), and increases with advancing age group. **Conclusion:** The results show that neurotuberculosis is a serious public health problem, with variations in the number of hospitalizations over time. Despite fluctuations, there is a general upward trend, highlighting the importance of continuous surveillance and intervention. The Southeast region has the highest number of cases and a higher mortality rate. Vulnerable groups include men, people of mixed race, and certain age groups. These findings underscore the need for specific prevention and control strategies, especially in these areas and populations most affected, to reduce both the incidence and mortality of neurotuberculosis.

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Epidemiological profile of pediatric patients hospitalized for multiple sclerosis in Brazil: 2014-2023

Tayenne Nélly de Lucena Viana[1], Vitor Cortês Cabral Magalhães[2], Nathanael Willian Florencio de Freitas[1], Oliver Reiks Miyajima[1]

[1]Universidade de Fortaleza, Fortaleza CE, Brazil.
[2]Instituto de Ensino e Pesquisa Insper, São Paulo SP, Brazil.
[3]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Oliver Reiks Miyajima; Email: oliver.miyajima@aluno.uece.br.

Background: Multiple sclerosis (MS) is an autoimmune disease causing demyelination of central nervous system (CNS) that affects mainly young adults, with strong female preponderance. This disease is characterized by recurrent episodes of neurological dysfunction, such as weakness in the limbs, impaired vision and poor coordination, and causes significant disability after onset. A small percentage of MS diagnoses occur before the age of 18 years, with slower progression than in adults due to neuroplasticity, but capable of generating moderate-to-severe disability at a younger age. **Objective:** To analyze the data available on the DATASUS platform about the hospitalizations by MS in children aged 0-19 years in Brazil over the ten years 2014-2023 and provide epidemiological relevant information. **Methods:** This is a cross-sectional, descriptive and retrospective study. Data were extracted from the Hospital Morbidity System (SIH/SUS – DATASUS) from the years 2014 to 2023 considering the number of hospitalizations due to MS in children aged 0-19 years in Brazil according to place of residence. The variables analyzed were region of the country, sex, age, race, deaths and year of hospitalization. **Results:** Between 2014-2023, there were a total of 2,628 hospitalizations, with the largest share occurring in the southeast, 1,728 (65.75%), and the smallest share in the north, 61 (2.32%). In the south, between 2021 and 2023, cases increased by 56.1%. In addition, in the northeast region, in the total period analyzed, they increased by 275%. Among the states, the ones with the most hospitalizations were São Paulo 1,140 (43.37%) and Minas Gerais 452 (17.19%). Of all the cases, there was only one death (0.0038%), and this was a male in the 15-19 age group. The predominant gender in hospitalizations was female, with 1,689 cases (64.26%), while males accounted for 939 cases (35.73%). The most affected pediatric age group was 15-19 years old with 2060 cases (78.38%), followed by 10-14 years old with 478 cases (18.18%), 5-9 years old 81 cases (3.08%), 1-4 years old 7 cases (0.26%) and under 1 year old 2 cases (0.07%). Among the races, whites accounted for the majority of hospitalizations with 1,399 cases (53.23%), while the smallest race with recorded hospitalizations was yellow with 24 (0.91%). There were no hospitalizations recorded for the indigenous population. The year with the most hospitalizations was 2022 with 367 cases (13.96%), and the lowest was 2014 with 137 cases (5.2%). **Conclusion:** Although the pediatric age group represents a small number of MS diagnoses, there has been an increase in the number of hospitalizations over the years, with a predominance of females and a later age in adolescence. In addition, even with the predominance of cases in the southernmost regions of the country, the large increase in the northeast region is noteworthy. Further studies on the subject are needed in order to provide a better understanding.

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Epidemiological profile of transient ischemic stroke (CVA) and related syndromes in adults and elderly people in Northeast Brazil from 2018 to 2023

Luana Cristina de Oliveira Lima[1], Arthur Vinícius Feitosa Santos[1], Vagner Costa da Cunha Júnior[1], Sabrina Sthéphanie Ataíde Martins[1], Maria Cecília Leal Menezes[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Luana Cristina de Oliveira Lima; Email: luana.cristina0410@gmail.com.

Background: Stroke is one of the main causes of death and disability in the world, especially in low and middle income countries. It is characterized by interruption of blood flow to an area of the brain, resulting in tissue damage. Ischemic stroke, responsible for approximately 85% of cases, occurs due to the occlusion of a cerebral artery, usually by a thrombus or embolus. Its risk factors include high blood pressure, diabetes mellitus, smoking, hypercholesterolemia and atrial fibrillation. **Objective:** To analyze cases of transient ischemic stroke and other related syndromes in adults and elderly people in Northeast Brazil between January 2018 and December 2023 through analysis of data from the Department of Information and Informatics of the Unified Health System (DATASUS). **Methods:** Descriptive epidemiological study using secondary data from the SUS Hospital Information System (SIH/SUS), Tabnet, and data published in the Brazilian Institute of Geography and Statistics (IBGE), being tabulated in Microsoft Excel, according to data collected from cases registered during the period from January 2018 to December 2023. The variables analyzed were state, sex, race/ethnicity and age group over 20 years old. **Results:** In the period analyzed, registered cases of transient ischemic stroke were 615.76 per million inhabitants in the Northeast. The states with the highest prevalence per 100,000 inhabitants are Alagoas (191.81), Maranhão (88.13) and Pernambuco (80.45); and the lowest are Sergipe (16.2), Paraíba (23.7) and Rio Grande do Norte (30.15). Furthermore, there was a higher prevalence in males (53.14%), mixed people (51.07%) and those aged over 50 years (60.88%). Finally, the highest number of deaths occurred in the states of Sergipe (21.42%), Paraíba (12.74%) and Maranhão (12.23%) and, in relation to age group, individuals over 80 years old are the most affected (29.68%), followed by those aged 70 to 79 (28.19%) and aged 60 to 69 (20.56%). **Conclusion:** Based on the results, mixed men over 50 years of age have a higher prevalence of transient ischemic stroke, which can be correlated with a sedentary lifestyle, high blood pressure, diabetes and family history, which can lead to arterial clogging, representing risk factors. Furthermore, the states with the highest number of deaths are those with the lowest number of hospitalizations, in the case of the states of Sergipe and Paraíba, suggesting disparity in access to health services, especially in highly complex ones. Finally, it is necessary to implement public policies, highlighting the importance of recognizing clinical signs and implementing prevention strategies in order to reduce sequelae and improve prognosis.

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Epidemiological profile of traumatic cervical spinal cord injury in Northern Brazil: a 10-year retrospective analysis.

Maria Cândida Barros Arantes Romano[1], Júlia Oliveira e Freitas[1], João Lazaro da Silva Rodriguez Oquendo[1]

[1]Centro Universitário Presidente Antônio Carlos, Araguaína TO, Brazil.

Correspondence: Maria Cândida Barros Arantes Romano; Email: candinha_barros-ar@hotmail.com.

Background: The traumatic spinal cord injury (SCI) consists in a break of the spinal medulla, being ocasioned by a traumatic event that might cause the loss of function, be it, motor, sensible ou autonomic in a way which may be transitory or irreversible. The condition of a medullar injury varies according to the affected portion of the spine, being the main manifestations, the paralysis or paresys of the limbs, alteration in the muscle tone, alteration in the superficial and profound reflexes, among others. Spinal cord injury is not subject to notification, with little epidemiological data and published work. The worldwide incidence of SCI is estimated between 9 and 50 cases/million inhabitants. **Objective:** The study aimed to analyze patients hospitalized with spinal cord injury of the cervical region in the north of Brazil, using data from the SIH/SUS to understand the profile and associated factors. **Methods:** This is a descriptive, quantitative and retrospective study with 918 victims of cervical spinal injury, between the years 2012 and 2022, in the states of Pará, Tocantins, Rondônia, Amazonas, Acre, Roraima and Amapá. Data from the SUS Hospital Information System (HIS/SUS) was used as a source. The variables sex, age, treatment and condition were studied. **Results:** The data consists of 82.4% male patients and 17.5% female patients, with 20 to 35 years being the age group with the highest prevalence of cervical SCI. Regarding the incidence by state, there is a higher occurrence in the state of Pará (48%), followed by Rondônia (19%) and Tocantins (12%). Furthermore, there is also a preference for conservative treatment in 69% of cases. **Conclusion:** The group at highest risk for cervical spinal cord trauma are young men, with a higher prevalence in the state of Pará and the most adherent treatment is conservative. Therefore, the occurrence of cervical SCI in the northern region is significant, requiring improvement in the quality of pre-hospital and hospital care for these patients to reduce the risks and consequences of emergency surgeries, which can lead to irreversible sequelae. Furthermore, there is a need for epidemiological studies and notifications about spinal cord injury and its causes, especially in the northern portion of the country, for subsequent prevention planning.

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Epidemiological study of cases of ischemic stroke in a hospital of Minas Gerais: an observational study

Letícia Aparecida de Figueiredo[1], Isabela Pinto Braga[1], Mauro Eduardo Jurno[1], André Brasil Tollendal[1], Denise Suemy Rocha Hashimoto[1]

[1]Centro Hospitalar de Barbacena, Barbacena MG, Brazil.

Correspondence: Isabela Pinto Braga; Email: isabela.braga7@gmail.com.

Stroke is an important cause of morbidity and mortality worldwide, being the second most common cause of death in Brazil and worldwide. Considering the impact of the disease related to failures in hospital care, a Stroke Unit was implemented in an institution at the state of Minas Gerais in June 2022 with expectation of improvements in management of those cases. The goal of this study was to outline the epidemiological profile of stroke victims in a Hospital at the state of Minas Gerais from October 2022 to December 2023. This is a cross-sectional observational study based on the evaluation of medical records of patients treated during this period. The total number of patients with suspected stroke/TIA was 277 patients. The average age was 68.5 years and the predominant sex was 58% male. Regarding the presence of comorbidities, the most prevalent diseases present in admitted patients were hypertension, diabetes mellitus, dyslipidemia and congestive heart failure. Regarding admission, it was observed that the NIH score was applied to 44.2% of admitted patients, with the average score on this scale being 9. The average time between admission and imaging was 79.5 minutes. In this service, the Stroke Unit was created in June 2022, and since then it has gradually brought improvements to provide complete and effective care for patients in the region. The analysis shows demographic data with similar numbers compared to the numbers encountered in Brazil. Data on comorbidities prior to admission slightly differed from the rest of the Country. Data such as door-to-image and door-to-needle times and data related to rehabilitation and mortality were discrepant between the service studied and the rest of Brazil, which may reflect, among other reasons, prolonged tomography maintenance time, delay in dispensing medications and reduced number of professionals in the multidisciplinary team.

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Epidemiological study of live births with spina bifida in Brazil

Camila Pugliesi de Figueiredo[1], Victor Pedrosa Ferreira de Souza[1], Amanda Mendes Santos[1], Michelle Oliveira Teixeira[1], Wânia Rezende Lima[1]

[1]Universidade Federal de Catalão, Catalão GO, Brazil.

Correspondence: Camila Pugliesi de Figueiredo; Email: camila.figueiredo@discente.ufcat.edu.br.

Background: Spina bifida (SB) is a congenital defect, resulting from the incomplete closure of the neural tube between the 3° and 4° week of pregnancy, impacts life quality in the baby, causing, for example, motor disabilities. Although there are important genetic patterns that contribute to the prevalence of neural tube defects, it is known that factors, such as folic acid supplementation, are of the utmost importance in their etiology. **Objective:** To evaluate the epidemiological scenario of live births with spina bifida and fetal factors in Brazil. **Methods:** It's a retrospective descriptive epidemiological study of live births with spina bifida, based on data from the Congenital Malformations, Deformities and Chromosomal Anomalies Monitoring Panel. The data from 2014 to 2024 was analyzed, with the results from 2023 and 2024 being preliminary. The data used is from public domain sources, thus there is no need to submit the work to the National Research Ethics Commission (CONEP) and analysis by the Research Ethics Committee (CEP) system. The variables included were gestational duration, delivery type, Apgar Score from the first and fifth minute, birthweight, sex and skin color. **Results:** Of the live births with SB, 722 were diagnosed in 2014, 689 in 2015, 821 in 2016, 787 in 2017, 795 in 2018, 743 in 2019, 671 in 2020, 718 in 2021, 704 in 2022, 718 in 2023 and 225 in 2024 (up to the most recent June update). On the gestational duration, 2265 babies were premature, with 7 being extremely premature and 5280 were born at term. This information was ignored in 48 cases. Regarding the delivery type, the most prevalent was c-section (6,306), corresponding to 83% of the total, while only 1,276 were vaginal (16.8%). The data was marked as blank or ignored in 11 births. As for the APGAR, in the first minute, 485 babies had an APGAR of 0 to 2, 601 of 3 to 4, 1787 of 5 to 7, and 4633 of 8 to 10, with 87 marked as blank or ignored. In the fifth minute, 244 live births had an Apgar score of 0 to 2, 124 of 3 to 4, 660 of 5 to 7, 6471 of 8 to 10, with 94 as blank or ignored. Concerning birthweight, 71.5% of live births weighed 2500g or more, 21.5% weighed 1500 to 2499g, and 6.9% weighed less than 1500g; Only 2 births had this data ignored or left blank. Considering the sex, 3838 were male, 3549 were female and, in 106, this information was absent. Furthermore, the skin color was marked as brown in 51.7%, white in 37%, black in 6.7%, yellow in 0.4% and 0.4% were indigenous. This information was blank in 3.7% of live births. **Conclusion:** The year with the highest number of SB live births was 2016. Most births were cesarean sections, although around 17% were vaginal deliveries. It is crucial to emphasize that a baby born with spina bifida requires greater care. Therefore, a reasonable percentage of live births were premature, had low birth weight or had a low APGAR at the 5th minute, indicating asphyxia. Finally, the majority of those born were brown and male.

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Epidemiological study on multiple sclerosis hospitalization in Brazil comparing sex, age and region between January 2008 to December 2023

Maria Eduarda Medeiros Martins[1], Lucca Ferdinando Queiroz Fernandes[1], Diógenes Emanuel Dantas da Silva[1], Matheus Araújo de Medeiros[1], Arthur Rebouças de Souza[1], Tiago Lameque de

Sousa e Silva[1], Amanda Lima Sampaio[1], Claúdio de Souza Fernandes Júnior[1], Pedro Henrick Guimarães Carvalho[1], Iury Hélder Santos Dantas[1]

[1]Universidade Federal do Rio Grande do Norte, Caicó RN, Brazil.

Correspondence: Maria Eduarda Medeiros Martins; Email: mariaeduardammartins1209@gmail.com.

Background: Multiple sclerosis (MS) is a chronic autoimmune disease of the central nervous system, characterized by inflammation, demyelination, and neurodegeneration. MS is known to present with diverse clinical manifestations and can lead to significant disabilities in affected individuals. Despite its relatively low prevalence compared to other neurological diseases, there is a growing recognition of MS as a significant health problem, which highlights the need for comprehensive epidemiological studies to better understand its impact. **Objective:** To conduct an epidemiological analysis of hospitalizations due to MS in Brazil between 2008 and 2023. **Methods:** Data were obtained from the Notifiable Diseases Information System (SINAN) of the Ministry of Health, and analyzed to identify the number of hospitalizations for multiple sclerosis in Brazil between 2008 and 2023, including analyses by year, region, sex, and age group. **Results:** Regionally, the Southeast Region had the highest concentration of hospitalizations, representing 67.0% of the total, followed by the South (15.6%), Northeast (8.4%), Central-West (7.1%), and North (1.9%) regions. There was a significant variation over the years, with hospitalizations gradually increasing from 2008, when 1,682 cases were recorded, reaching a peak in 2022 with a total of 7,299 cases. Regarding gender, women accounted for a higher number of hospitalizations, representing 69.0% of the total, while men accounted for 30.0%. In terms of age group, the majority of hospitalizations occurred in young adults, especially in the 20 to 49-year age range, which represented 73% of the total. There was a gradual decrease in hospitalizations with increasing age, with only 437 cases recorded in individuals aged 80 and over. Among children under 5 years old, there were only 13 hospitalizations, equally distributed between boys and girls, highlighting the low incidence of MS in this age group. **Conclusion:** This study highlights the complexity and diversity of the impact of MS in Brazil, emphasizing the need for more effective and targeted public health policies for early diagnosis, appropriate treatment, and continuous support for patients, especially in the most affected regions and groups. However, there is a need for additional studies on the subject, particularly to improve the organization of resources dedicated to the treatment and prevention of hospitalizations due to multiple sclerosis.

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Epidemiological study on the prevalence of Parkinson's disease, parkinsonism, and rem sleep behavior disorder in the community of Candangolândia - DF

Simoneide Souza Titze de Almeida[1], Isabel Oliveira Araujo Moraes[2], Milena Vieira Ramos[3], Talita Guimarães Balieiro[4], Gustavo Henrique Campos de Sousa[2], Juliana Soares Guimarães[5], Marcelo Evangelista Lobo[2], Pedro Manzke de Carvalho[2], Pedro Renato de Paula Brandão[4]

[1]Universidade de Brasília, Brasília DF, Brazil.
[2]Hospital de Base do Distrito Federal, Brasília DF, Brazil.
[3]Universidade de Rio Verde, Faculdade de Medicina, Rio Verde GO, Brazil.
[4]Hospital Sírio Libanês, Brasília DF, Brazil.
[5]Instituto Brasileiro de Neuropsicologia e Ciências Cognitivas, Brasília DF, Brazil.

Correspondence: Isabel Oliveira Araujo Moraes; Email: isaoraujo@outlook.com.

Background: This study is part of the project: Prodromal and Overt Parkinson's Disease Epidemiological Study in Brazil (PROBE-PD), which aims to determine the prevalence of Parkinson's disease (PD) and other parkinsonian syndromes in individuals aged 60 and over within a population sample from the North, Northeast, South, and Midwest regions. **Objective:** To present partial results regarding the prevalence of PD, parkinsonism, and REM sleep behavior disorder (RBD) in the community of Candangolândia-DF, Midwest region. **Methods:** This study, approved by the Research Ethics Committee of the Beneficent Society of Ladies Hospital Sírio-Libanês (CEPesq/HSL-5.482.147), is conducted in two phases. In Phase I, screening is carried out using the Tanner Questionnaire, wherein volunteers respond to nine yes-or-no questions related to motor symptoms, and the Single-Question Questionnaire for RBD (RBD1Q). A positive screening is indicated by either two affirmative responses on the Tanner Questionnaire or one on the RBD1Q, prompting an invitation to Phase II, where participants undergo evaluation by a neurologist specializing in Movement Disorders. PD diagnosis follows the MDS Diagnostic Criteria, supported by the Unified Parkinson's Disease Rating Scale (MDS-UPDRS). The evaluation further includes the RBD Screening Questionnaire (RBDSQ), Montreal Cognitive Assessment (MoCA), and Beck Depression Inventory (BDI-II). Data are entered into RedCap for subsequent statistical analysis. Results are presented as frequencies, percentages, means, and standard deviations. **Results:** To date, 1253 individuals out of a target of 1500, with a mean age of 70.5±7.95 years have been screened between July 2022 and June 2024. Among these screened individuals, 63.6% were female. The prior diagnosis of PD was 1.8%, with 11.7% screened positive for suspected RBD and 34.6% for the Tanner questionnaire. Individuals screened positive for suspected RBD and Tanner, or previously diagnosed with PD, were invited to participate in Phase II. Following the clinical evaluation of 307 patients, preliminary prevalence was estimated at 1.67% for PD, 2.87% for parkinsonism of any cause, and 3.27% for probable/possible RBD. **Conclusion:** Of the individuals diagnosed with PD, 53.85% were male. The result reveal that although the relative proportion of males was lower than that of females, the prevalence of PD diagnoses was higher among males, corroborating existing literature reports. Continued data analysis and detailed clinical evaluations will provide a more precise understanding of the results found in the present study. These findings are crucial for informing and potentially adjusting public health policies in Brazil, aiming to improve early diagnosis and management of these neurodegenerative diseases. On behalf of PROBE-PD

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Epidemiology of Alzheimer's disease in Ceará: an analysis of hospitalized patients (2008-2022)

Guilherme Nobre Nogueira[1], Rafaela Fernandes Gonçalves[2], Gustavo Rassier Isolan[2], Gustavo Henrique Tomasi[3], Matheus Roriz-Cruz[4]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Faculdade Evangélica Mackenzie do Paraná, Curitiba PR, Brazil.
[3]Universidade do centro oeste do Paraná, Guarapuava PR, Brazil.
[4]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Guilherme Nobre Nogueira; Email: guiermenobre@gmail.com.

Background: Alzheimer's disease (AD), as the predominant cause of dementia, constitutes a growing concern in global health. Epidemiology plays a critical role in assessing the burden of AD, providing essential data on its prevalence, incidence, and societal impact. There are well-founded projections indicating that by 2050, the prevalence of dementia will double in Europe and triple worldwide. **Objective:** This study aims to highlight the crucial importance of epidemiology in understanding AD, focusing on the population of the state of Ceará during the period from January 2008 to December 2022, using data from hospitalized patients. **Methods:** This is a retrospective descriptive epidemiological study based on secondary data, using the Epidemiological Information System available in the Notification of Diseases Information System (SINAN), researching AD in the territory of Ceará, evaluating age groups and gender with the number of hospitalizations, deaths, and patient mortality rate from January 2008 to December 2022. **Results:** After a comprehensive analysis of the data related to patients hospitalized for AD in the state of Ceará we obtained the following results: a total of 298 patients were diagnosed with Alzheimer's Disease in the state of Ceará. Of these, 180 (64%) were female, while 118 (36%) were male. Regarding age groups, we observed the following distribution: Age 15-44 years: 28 cases, with 15 (54%) female and 13 (46%) male. No deaths were recorded in this age group. Age 45-59 years: 21 cases, with 12 (57%) male and 9 (43%) female, resulting in one death. Age 60-79 years: 105 cases, with 57 (54%) female and 48 (46%) male, accompanied by 5 deaths. Age 80 years or older: 144 cases, with 45 (31%) male and 99 (69%) female, and 18 deaths recorded in this age group. The chi-square analysis showed a p-value of 0.028 and a Cramer's v-value of 0.175. **Conclusion:** These results indicate a complex distribution of AD in the state of Ceará, with notable variations by gender and age group. They also emphasize the importance of epidemiology in understanding AD and formulating more personalized public healthcare strategies, considering gender and age differences. Furthermore, they underscore the need for further research to investigate risk factors in different demographic groups and the importance of multidisciplinary medical teams in the comprehensive care of AD patients.

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Epidemiology of Alzheimer's disease hospitalizations in Brazil: analysis regarding color/race, age group and sex

Maria Alice Pereira Junqueira de Oliveira[1], Matheus Wendell de Almeida Pereira[1], Raissa Jaquelline dos Santos Dias[1], Josué Natã Sampaio Monteiro[1], Peri Bucken Gobbi[1], Ellen dos Santos Brito[1], Gabriel Balbi Fernandes Silva[1], Orlando Dias Canichio[1], Camila de Souza Ferreira[1], Laura Rodrigues Melo[2]

[1]Universidade Federal do Rio de Janeiro, Macaé RJ, Brazil.
[2]Faculdade de Medicina de Petrópolis, Petrópolis RJ, Brazil.

Correspondence: Maria Alice Pereira Junqueira de Oliveira; Email: mariaalice353@gmail.com.

Background: Demographic transition and population aging represent prevalent phenomena in contemporary Brazil. Consequently, there is a need for tailored public health interventions to address the evolving needs of this population group, which is expected to become the majority in the coming years. In light of this, the prevalence of people living with dementia follows this increasing trend, which means that its most common entity, Alzheimer's disease (AD), significantly impacts the daily lives of several individuals as well as the dynamics of health services. **Objective:** To conduct a quantitative analysis of hospitalizations due to AD spanning from January 2014 to January 2024. This analysis aims to juxtapose findings across Brazil's five distinct regions, considering variables such as sex, ethnicity, and age group. **Methods:** This study is an observational, cross-sectional, descriptive, and quantitative analysis conducted utilizing data extracted from the Sistema de Informações Hospitalares do SUS (SIH-SUS), integrated with DATASUS. The investigation

explores variables associated with hospital admissions for AD across the five geographical regions of Brazil, categorized by sex, ethnicity, and age group. The data was collected between March 11th and 22nd, 2024. **Results:** During the analyzed period, a total of 15,445 hospitalizations for AD were documented in Brazil, reflecting an upward trend all over the years. Among the country's five regions, the Southeast region reported the highest caseload, comprising approximately 52.9% of hospitalizations, followed by the South and Northeast regions with 24% and 13.9%, respectively. Women represented the majority of hospitalizations, constituting 65.4% of the total, whereas men accounted for 34.5%. Regarding ethnicity, individuals of Caucasian descent exhibited a predominant presence, representing 48.7% of the total, followed by records with lacking information regarding the variable in question at 23.3% and the "pardo" (brown-skinned) Brazilian race at 22.5%. Regarding age distribution, individuals aged 80 and above constituted the majority of hospitalizations, comprising 58.5% of the total. **Conclusion:** It is noted that 23.3% of recorded hospitalizations lack information regarding the patient's ethnicity, which compromises the development of epidemiological studies as well as the formulation and implementation of public health measures aimed at the effective management of AD. Hence, it is imperative to underscore to various healthcare professionals and administrators the significance of correctly completing records. Furthermore, the prevalence of hospitalizations among individuals aged over 80 underscores the necessity of adequately preparing hospital teams for geriatric care, a facet often overlooked in clinical practice.

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Epidemiology of encephalocele in newborns in Brazil: an analysis of the last decade

Michelle Oliveira Teixeira[1], Amanda Mendes Santos[1], Camila Pugliesi de Figueiredo[1], Victor Pedrosa Ferreira de Souza[1], Guaraciara de Andrade Picanço[1]

[1]Universidade Federal de Catalão, Catalão GO, Brazil.

Correspondence: Amanda Mendes Santos; Email: amanda.mendes@discente.ufcat.edu.br.

Background: Encephalocele is defined as the herniation of intracranial contents through an opening in the skullcap due to defects in the closure of the neural tube. Patients may present a variety of symptoms, such as seizures, ataxia, visual deficits, microcephaly and developmental delays. Also noteworthy is the high mortality during pregnancy and after birth. Therefore, it is important to delineate the population with this condition and promote the adoption of prevention measures, such as early folic acid supplementation. **Objective:** To analyze the epidemiological profile of live births with encephalocele in Brazil. **Methods:** This quantitative, descriptive and retrospective study collected data from the Congenital Malformations, Deformities, and Chromosomal Anomalies Monitoring Panel. The variables used were indicator, location, month of pregnancy that started prenatal care, weeks of gestation, type of delivery, APGAR in the 1st and 5th minute and birth weight. The period from 2014 to April 2024 was analyzed, with preliminary data from the last two years. **Results:** During the period studied, a total of 1979 live births with encephalocele were reported, with unspecified encephalocele being the most prevalent (1207). Of the specified encephaloceles, 498 babies were born with occipital encephalocele, 158 with encephalocele of other locations, 107 with frontal encephalocele, and 23 with nasofrontal encephalocele. Dividing by each Brazilian region, 769 cases occurred in the Southeast, 604 in the Northeast, 267 in the South, 181 in the North, and 149 in the Central-West. Regarding prenatal care, the earlier it starts, the better the chances of prevention with folic acid supplementation. That said, 18.2% of pregnant women began in the 1st month, 37.5% in the 2nd, 19.5% in the 3rd, and 17.8% from the 4th month onwards. On the gestational duration, 37.4% were born prematurely. Regarding the delivery type, 21.1% were born vaginally. Regarding APGAR, 59.5% of live births had an APGAR score below 8 in the 1st minute, with 19.5% having severe asphyxia, 13.9% being moderate, and 26.1% having mild. While for the APGAR, at the 5th minute, 38% had a score lower than 8, with 13.9% having severe asphyxia, 6.5% being moderate and 17.7% having mild. About birth weight, 12.7% were born with meager weight, 25.7% were underweight, and 61.5% weighed more than 2500g. **Conclusion:** Although rare, encephaloceles are relevant diagnostic possibilities. A significant percentage of live births with this condition presented perinatal asphyxia, with an APGAR score of less than 8 in the 1st and 5th minute. Prematurity and low weight were also characteristic, although they did not correspond to most cases. As for the late start of prenatal care, its rate was notable, showing that early and universal use of this service is not a reality in Brazil. Therefore, the prevention of neural tube closure defects with folic acid supplementation is still inaccessible for many women.

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Epidemiology of multiple sclerosis mortality in Brazil over the last 20 years

émile de Carvalho Morais Fraga[1], érica Otoni Pereira Miranda[1], Guilherme Rocha Cardoso[1], Luídi Neves Nascimento[1], Naila Maria Magalhães da Silva[1], Rebeca Gabrielle Almeida Maciel[2], Emmanuel Santos Trindade[1], Priscila Santos Oliveira[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.
[2]Universidade Federal da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Érica Otoni Pereira Miranda; Email: ericaotoni15@gmail.com.

Background: Multiple sclerosis (MS) is the most prevalent demyelinating disease of the central nervous system (CNS). It is estimated to affect between 2 and 2.5 million people globally, with a higher prevalence in females, white individuals and have a peak incidence at the age of 30. Current knowledge of MS mortality in Brazil is limited. **Objective:** To analyze the time trend in MS mortality rate in Brazil between 2001 and 2022, stratified by sex, ethnicity and age group. **Methods:** This ecological time series study assessed the MS mortality rate in Brazil, stratified by sex, ethnicity and age group. Death data was obtained from the Mortality Information System of the Ministry of Health's Department of Informatics of the Unified Health System. Demographic data was obtained from the National Household Sample Surveys and the Continuous National Household Sample Surveys of the Brazilian Institute of Geography and Statistics. Mortality rate trends were analyzed by joinpoint regression. **Results:** 6,848 MS deaths were reported in Brazil between 2001 and 2022. Females accounted for 60.48% of deaths. 66.41% of deaths were of white individuals. 28.8% of deaths occurred in those aged 70 or over. The average annual mortality rate was 0.16 deaths/100,000 inhabitants. Females presented a higher average annual rate (0.19 deaths/100,000 inhabitants) than males (0.13 deaths/100,000 inhabitants). When analyzing the average annual mortality rate by ethnicity, white individuals led the ranking (0.23 deaths/100,000 inhabitants), followed by blacks (0.11 deaths/100,000 inhabitants) and, lastly, browns (0.08 deaths/100,000 inhabitants). The mortality rate is directly proportional to age group, with the population aged 70 years or over having the highest average annual mortality rate (0.93 deaths/100,000 inhabitants), followed by those with 60 to 69 years (0.49 deaths/100,000 inhabitants), 50 to 59 years (0.35 deaths/100,000 inhabitants), 40 to 49 years (0.21 deaths/100,000 inhabitants), 30 to 39 years (0.08 deaths/100,000 inhabitants), 20 to 29 years (0.03 deaths/100,000 inhabitants), and, finally, those under 20 years (0.004 deaths/100,000 inhabitants). It was noticed an overall stationary trend in mortality rate (AAPC: -0.54 [-1.37; 0.29]; p:0.20). Black individuals (AAPC: -1.48 [-2.76; -0.19]; p:0.03), those aged 20 to 29 (AAPC: -3.19 [-6.07; -0.21]; p:0.04) and 70 or over (AAPC: -4.72 [-5.38; -4.05]; p:<0.0001) demonstrated a decreasing trend. Brown individuals (AAPC: 2.26 [0.64; 3.91]; p:0.006) presented an increasing trend. **Conclusion:** This study showed a low mortality rate from MS when compared to countries with a majority Caucasian population. In addition, the data presented corroborates other studies, showing a predominance of Multiple Sclerosis mortality in females, white individuals and elderly individuals in Brazil. The mortality rate trend was stationary.

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Epidemiology of patients followed by the post-stroke palliative care team in a reference hospital for stroke treatment in Santa Catarina

Maria Eduarda Angelo de Mendonça Fileti[1], Patrícia Vera[1], Lila Jerusa Nascimento Pereira Abreu[1], Marta Gabriele Pires da Rosa Schmen[1], Camilla Ramos Mohr[1], Jane Rossi[1], Brenner Henrique de Oliveira Marques da Silva[1], Joselma Siqueira de Aquino[1], Carla Heloísa Cabral Moro[1]

[1]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Maria Eduarda Angelo de Mendonça Fileti; Email: neuro.mef@gmail.com.

The word palliate derives from the Latin pallium, which means to protect, a form of care, that aims to alleviate pain and suffering. According to the World Health Organization (WHO), in the concept defined in 1990 and updated in 2002, Palliative Care is an "approach that improves the quality of life of patients and their families, who face problems arising from a life-threatening illness, through prevention and relief of suffering, through early identification, impeccable assessment and treatment of pain and other physical, psychosocial and spiritual problems". Studies identified high case fatality after severe stroke, numerous long-term physical problems, substantial emotional distress, prognostic uncertainty, and a sense of abandonment after hospital discharge. Our experience in a reference center for stroke treatment has shown the need of a Post-Stroke Palliative Care Protocol establishment, to support a holistic approach, improving quality of life and ensuring treatment in accordance with the wishes and values of our patients and their families. After the Palliative Care for Post-Stroke Patients Protocol was implemented, a retrospective analysis of the clinical database collected by the electronic medical record was fulfilled. Patients included were those diagnosed with stroke from June 2023 to June 2024 and who received palliative care during the study period. Data from clinical information, medical diagnosis and outcome were collected. Analisis was carried out to obtain the epidemiological profile of patients receiving post-stroke palliative care. 81 patients were followed from June 2023 to June 2024, 31 male patients and 50 female patients, predominantly in the age group of 70 to 89 years. 81% had ischemic stroke and 19% had hemorrhagic stroke. Among the main comorbidities analyzed, approximately: 76% had systemic arterial hypertension, 39% had diabetes mellitus, 7% had dyslipidemia, 3% had a previous stroke and 23% had atrial fibrillation. More than 95% of individuals had multimorbidities. Of these, approximately 33% were previously classified as mRankin level 1 and 32% as mRankin level 3. After the cerebrovascular event, mRankin level 5 predominated, in 75 patients. From the total of 81 patients: 33 patients died, 38 returned home, 9 went to nursing home and one patient still remains in hospital. It is very important to notice that the majority of our patients were discharged from the hospital, counting on our home monitoring program and on the care guidelines provided during hospitalization. Understanding the epidemiology of palliative care after stroke is essential to inform health policies, service planning and clinical practice, aiming to improve the quality of life and health outcomes of these patients.

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Epilepsy during the COVID-19 pandemic: an analysis of trends in hospitalizations and deaths in Brazil

Alisson Rafael de Oliveira Pereira[1], José Sidinei Rodrigues Dias[1], Julia de Oliveira Bessegatto[1], Rodrigo Ferreira de Moura[1]

[1]Universidade Federal de Lavras, Lavras MG, Brazil.

Correspondence: Alisson Rafael de Oliveira Pereira; Email: alissonropereira@gmail.com.

Background: From the perspective of public health management, the COVID-19 pandemic between Mar/2020 and May/2023, highlighted the need to overcome operational challenges and implied complications in hospital dynamics, such as instability in bed availability. While the focus was centered on COVID-19, care for other chronic noncommunicable diseases, such as epilepsy, may have been impacted. Thus, to assess the existence of this correlation and create other measures aimed at mitigating these impacts, it is important to understand the behavior of epidemiological data regarding this syndrome. **Objective:** To analyze trends in deaths and hospitalizations due to epilepsy during the COVID-19 pandemic in Brazil. **Methods:** This retrospective study used secondary data recorded at SIH using DATASUS. Data on hospitalizations and deaths due to epilepsy were collected refering to the period May/2019-Mar/2024. The analysis of the time series was performed with joinpoint regression and an ARIMA model to evaluate the influence of the COVID Pandemic as a covariate. **Results:** The regression showed four points of change during the period: Jan/2020, Apr/2020, Aug/2022 and Jan/2023. The initial stationary trend was interrupted by a drop in hospitalizations in Jan/2020, generating a brief downward trend (APC: -8; p=0.1), which was replaced in Apr/2020 by a longer-lasting, linear, and significant upward trend (APC: 1.3; p<0.05). As for Aug/22, a non-significant slight fall regime was established (APC: -0.19; p=0.55), which remained until Jan/23, when there was a new important drop (APC: -27; 95%CI: -32.6 to -18.6; p<0.01). Summarizing the entire period, there was a significant drop in hospitalization for epilepsy (AAPC: -1; 95%CI: -1.2 to -0.7; p<0.01). From deaths point of view, the series presented seasonal characteristics that remained similar to the pattern before the beginning of the COVID pandemic. Although it is not possible to pinpoint this causality to the changes, some of the joinpoints in hospitalizations are related to milestones in the pandemic period, and ARIMA models (0, 0, 0) illustrate the influence of the COVID parameter as a covariate in deaths (B=-0.34; SE=0.1; t=-2.6; p<0.05). The first confirmed case of COVID was registered in Brazil in Feb/2020, although it is likely that the virus was already circulating throughout the country. A decrease in hospitalization may indicate an initial occupancy of beds due to flu-like syndromes. The next elevation may represent the presence of epilepsy-COVID comorbidity. **Conclusion:** Although the ARIMA model demonstrated the influence of the pandemic variable on epilepsy deaths, the previously presented seasonal pattern was maintained. Hospitalization trends initially varied in accordance with milestones and progression of the COVID pandemic in Brazil. Further research is needed to explain the seasonal death pattern and recent trend of a sharp drop in hospitalization.

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Epilepsy in the Brazilian pediatric population: an ecological study between 2018 and 2023

Samyra Roberta Assis Souza[1], Letícia Hanna Moura da Silva Gattas Graciolli[2] , Ana Carolina Ventura de Santana de Jesus[3], Yasmin da Silva Moura[4], Lauany évellin Pires da Silva[5] , Daniely Alves Dalla Zana Frazão[4], Maria Gabriella Pereira dos Santos[6], André Valério da Silva[7]

[1]Universidade de Marília, Marília SP, Brazil.
[2]Faculdade de Medicina de Jundiaí, Jundiaí SP, Brazil.
[3]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[4]Universidade Salvador, Salvador BA, Brazil.
[5]Faculdade Zarns, Salvador BA, Brazil.
[6]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.
[7]Universidade Federal de Mato Grosso, Cuiabá MT, Brazil

Correspondence: Samyra Roberta Assis Souza; Email: samyraassis@hotmail.com.

Background: Epilepsy is one of the most common neurological disorders in childhood, with early implementation of treatment being fundamental for optimizing quality of life and reducing long-term consequences, such as behavioral and learning problems. Therefore, the epidemiological analysis of pediatric epilepsy in the national territory is imperative to guide public health policies that meet the specific demands of each region, thus ensuring adequate access to treatment for all children. **Objective:** To analyze the epidemiology of epilepsy in the pediatric population between 2018 and 2023 in Brazil. **Methods:** This is a descriptive ecological study conducted through data extraction from the Hospital Information System (SIH/SUS), available on DATASUS, in the time series from 2018 to 2023. The variables used were: hospitalizations, total average length of stay, deaths, and epilepsy mortality rate. The covariates used were: sex, age group (0 to 14 years), and region. **Results:** From 2018 to 2013, there were 133,526 pediatric hospitalizations for epilepsy in Brazil, of which 45,018 (33.7%) were recorded in the Southeast, thus being the state with the highest number of hospitalizations. The most affected age group was 5 to 9 years (31,239 cases), corresponding to 23.3% of the total. Regarding sex, there was a predominance of males with 73,901 (55.3%), 24,839 only from the Southeast region. Regarding the total average length of stay, 5.2 days were obtained. However, individually, infants under 1 year old had the highest average (7.3 days), as well as the Northeast region of this same age group had 8.4 days. In 2023, there were 21,256 hospitalizations, equivalent to 15.9% of the total, with a decrease of 2% compared to 2018 (23,942), the year with the highest number of hospitalizations. The total number of deaths was 743, and the highest mortality rate was in the North region, occurring in infants under 1 year old with 1.28%. **Conclusion:** The data collected show that the highest number of epilepsy cases occurred in the Southeast region, in the age group of 5 to 9 years, mainly in males. However, infants under 1 year of age spend more time hospitalized. And although the highest number of hospitalizations is in the Southeast region, the highest epilepsy death rate is in the North region. In view of such data, it is concluded that the formulation of specific public health policies for each region is necessary in order to prevent and minimize the impact and effects of epilepsy on the pediatric population.

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Epilepsy mortality trends in Brazil (2001-2022): a joinpoint analysis

Emmanuel Santos Trindade[1], Gustavo Alves Cangussú[1], Priscila Santos Oliveira[1], Émile de Carvalho Morais Fraga[1], Maria Fernanda Aragão Andrade[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Emmanuel Santos Trindade; Email: emmanuelstrindade@gmail.com.

Background: Epilepsy is a condition that causes long-term predisposition to seizures. It is a diverse condition with multiple types and possible syndromes, resulting in variable etiologies and prognoses. Seizures affect approximately 10% of the population, with epilepsy affecting 1 to 2% of the world's population. Individuals with epilepsy have mortality rates 2 to 3 times higher than the general population. Analyzing epilepsy mortality rates is a crucial indicator of the quality of healthcare services. **Objective:** To analyze the mortality rate of epilepsy in Brazil from 2001 to 2022. **Methods:** This is an ecological time-series study of the epilepsy mortality rate in Brazil, stratified by sex, ethnicity and age group. For that, data on deaths from epilepsy were obtained from the Mortality Information System (SIM) and demographic data from the Brazilian Institute of Geography and Statistics (IBGE). The annual mortality rate due to epilepsy was calculated and, subsequently, a temporal analysis was carried out using a joinpoint regression model, through which the Annual Percent Change (APC) and Average Annual Percent Change (AAPC) were obtained with 95% Confidence Interval (95% CI). A value of p<0.05 was considered statistically significant. **Results:** Between 2001 and 2022, 47,437 deaths from epilepsy were reported in Brazil, characterizing an average annual mortality rate of 1.09 deaths per 100,000 inhabitants. The epilepsy mortality showed an increasing trend for the general population (AAPC: 4.67 [4.20; 5.13]; p:<0.01), as well as for males (AAPC: 4.49 [4.02; 4.97]; p:<0.01) and females (AAPC: 5.30 [3.22;7.42]; p:<0.01). The increasing trend was more meaningful in brown (AAPC: 6.52 [5.32; 7.73]; p:<0.01) and white people (AAPC: 5.17 [4.53; 5.81]; p:<0.01) when compared to black individuals (AAPC: 3.36 [1.63; 5.12]; p:<0.01). The age groups with the greatest increasing trend in mortality rate were, in descending order, ≥60 years (AAPC: 6.58 [5.73; 7.44]; p:<0.01), 50-59 years (AAPC: 5.11 [4.23; 6.00]; p:<0.01), < 20 years (3.77 [3.12; 4.42]; p:<0.01); 40-49 years (AAPC: 2.16 [1.50; 2.83]; p:<0.01) and 20-29 years (AAPC: 1.52 [0.88; 2.17]; p:<0.01). The 30-39 age group was the only one to show a stationary trend over the period analyzed (AAPC: 0.64 [-0.27; 1.56]; p:0.17), with an initial decreasing trend from 2001-2016 (APC: -0.89 [-1.60; -0.16]; p:0.02) and subsequent increasing trend from 2016-2022 (APC: 4.56 [1.57; 7.65]; p:<0.01). **Conclusion:** The epilepsy mortality rate in Brazil from 2001 to 2022 presented an increasing trend, regardless of sex. Besides, this trend was higher in brown and white people, as well as in older age groups. This differs from the decreasing trend related in previous national studies. An increasing trend was related in the USA from 1999 to 2017, which may be associated with an increase in all-neurological mortality and in epilepsy prevalence and a change in underlying causes of death.

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Epilepsy prevalence as a cause of hospitalization for central nervous system diseases in Brazil: an epidemiological study

Samyra Roberta Assis Souza[1], Ana Luiza Rossetto[1], Luigy Matheus de Freitas Santos[2], João Pedro Diniz Freire[1]

[1]Universidade de Marília, Marília SP, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Samyra Roberta Assis Souza; Email: samyraassis@hotmail.com.

Background: The central nervous system, composed of the brain and spinal cord, bears the primary responsibility for controlling and coordinating human bodily activities. However, it is susceptible to various pathologies and conditions that can hinder its functioning. Among these diseases, epilepsy stands out as one of the most widespread and well-known, representing approximately one-fifth (22.2%) of hospitalizations for Nervous System Diseases in Brazil. This chronic neurological disorder manifests as abnormal brain activity, leading to recurrent seizures; despite available treatments, epilepsy presents significant challenges for both patients and healthcare professionals. This summary will briefly discuss some of the disorders

affecting the CNS. Additionally, epidemiological findings related to CNS diseases, as well as epilepsy, will be compared. **Objective:** To compare epidemiological data of diseases affecting the Central Nervous System and Epilepsy. **Methods:** This is a descriptive ecological study concerning the hospitalization rates of patients aged 20 to 59 with epilepsy from February 2022 to February 2024, in relation to all hospitalizations due to central nervous system causes, divided by regions of Brazil. Data extraction was performed from the Hospital Morbidity System of the Unified Health System (SUS), available at the Department of Health Informatics of the Unified Health System (DATASUS). **Results:** In Brazil, 50,001 hospitalizations due to Epilepsy were recorded, accounting for approximately 22.2% of the 224,917 hospitalizations for Diseases of the nervous system. The majority of those hospitalized for CNS Disease were women, representing 56.3%. In contrast, 61.6% of those hospitalized for epilepsy were men. It was also noted that the Southeast region recorded the highest number of cases, with 96,472 (42.8%) for CNS Disease and 21,459 (42.9%) for Epilepsy. However, the second region with the most hospitalizations for CNS disease was the South, with 50,295 (22.3%) of the total, whereas for Epilepsy it was the Northeast region with 11,873 (23.7%) records. **Conclusion:** The study results reveal a gap between findings from different regions, representing social inequalities as well as differences in population size among them. The focus of prevalence lies on the most populous states, such as the Southeast, with the highest number of cases in both comparisons, and the Northeast ranking second in epilepsy cases, although it ranks third when surpassed by the South in nervous system diseases. In light of this information, it is imperative for healthcare professionals to address these conditions, deepening their understanding of risk factors and approaches to epilepsy and nervous system diseases, as well as implementing public policies to prevent and minimize these complications, aiming to ensure a quality life.

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Epilepsy: inference of quantitative data on hospitalizations in Brazil between 2019 and 2023

Deborah Araujo Silva[1], Rynele Almeida de Fonseca[1], Annayza de Oliveira Bessa[1], Larissa Bianca Borsone[1], Amanda Oliva Spaziani[1], João Carlos Bizinotto Leal de Lima[1], Gustavo Henrique da Silva[2], Vanessa Maria Gonçalves de Souza[1], Luis Carlos Spaziani[3], Francieli Gomes Lima[4]

[1]Universidade Brasil, Fernandópolis SP, Brazil.
[2]UPA de Santa Fé, Santa Fé do Sul SP, Brazil.
[3]Fundação Educacional de Fernandópolis, Fernandópolis SP, Brazil.
[4]AME Fernandópolis, Fernandópolis SP, Brazil.

Correspondence: Deborah Araujo Silva; Email: araujodeborah@outlook.com.

Background: Epilepsy is a common disorder of the central nervous system, affecting approximately 1% of the global population. It is a reversible and temporary condition that alters brain function, characterized by motor, sensory, autonomic phenomena, and varying degrees of altered consciousness. Due to its complexity, epilepsy has numerous etiological causes, including metabolic, immunological, infectious, genetic, and unknown factors. **Objective:** This study aims to evaluate the profile of hospitalizations due to epilepsy in Brazil between the years 2019 and 2023. **Methods:** Data collection was conducted using publicly available information from the Hospital Information System of the Brazilian Unified Health System (SIH/SUS) in the Tabnet/DATASUS database between January 15 and May 15, 2024. This study employed a retrospective longitudinal design with a quantitative descriptive approach. Data were grouped by Brazilian macro-regions, type of care, sex, age group, and race/ethnicity. Statistical inference was performed using BioEstat 5.3 software, utilizing paired T-tests for related samples to compare groups, and descriptive statistics, measures of central tendency, and dispersion were used to present the data. **Results:** Between 2019 and 2023, there were 285,241 hospitalizations for epilepsy in Brazil. The majority, 57.90% (n=165,164), were male (p<0.0001), and 45.39% (n=129,468) were of mixed race (p=0.0148). The age group most affected was 1-4 years, accounting for 16.38% (n=46,729; p<0.0001) of the cases. The Southeast region had the highest concentration of cases (n=114,768), corresponding to 40.24% (p=0.0005), with an average annual treatment cost of R$ 42,773,521.85. Mortality was most frequent in the Southeast, with 46.82% (n=3,561) of deaths (p=0.0005) and an average mortality rate of 5.03 (p=0.0013). The number of hospitalizations peaked in 2023, accounting for 22.67% (n=64,650) of the total, compared to only 17.25% (n=49,204) in 2020. The majority of hospitalizations, 93.22% (n=265,915; p<0.0001), were classified as urgent. **Conclusion:** Hospitalizations due to epilepsy are predominantly concentrated in the Southeast region of Brazil, with higher proportions of deaths and treatment costs during the study period. Most cases occurred among male children aged 1-4 years who were of mixed race, with urgent care being the most common type of hospitalization.

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Epileptic and developmental encephalopathy secondary to a mutation on gene STXBP1: a case report

Gabrielle Miranda Magalhães Pinto[1], Lourdes Maria Silva de Assis[1], Isabelle Diniz Melo[1], Maria de Fátima Miranda de Oliveira[2], José Lucivan Miranda[3], Fabiane Elpídio de Sá Pinheiro[1], João Joaquim Freitas do Amaral[1], Norma Martins de Menezes Morais[3]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade de Fortaleza, Fortaleza CE, Brazil.
[3]Núcleo de Tratamento e Estimulação Precoce, Fortaleza CE, Brazil.

Correspondence: Gabrielle Miranda Magalhães Pinto; Email: gabriellemmp1@hotmail.com.

Case presentation: A 7 year old female patient, born at full-term via vaginal birth with APGAR 7/10. At 4 months, the patient started presenting epileptic spasms, using vigabatrine. She displayed hypotonia and developmental delay (DD). A brain magnetic ressonance, requested for etiological investigation, was normal for her age. The patient evolved persisting with hypotonia and important DD, with poor social interaction, motor stereotypies, hypertonia in the lower limbs, agitared behavior and refractary epileptic seizures. Therefore, an epilepsy genetic panel was requested, resulting in developmental and epileptic encephalopathy-4 (DEE-4) secondary to the pathogenic variant c.364C> T: p.(Arg122Ter), found in heterozygosis on gene STXBP1. She utilized Levetiracetam and Topiramate, but had to interrupt use due to adverse reactions (diskynesias and weight loss). She arrived at our service when she was 4 years old, using valproic acid and clobazam, still presenting daily generalized tonic clonic and atonic seizures. Her eletroencephalogram resulted in polypoint trains and acute waves on central regions durong sleep, eletroclynic compatible to Lennox-Gasteaut Syndrome. Cannabidiol was, in that context, prescribed, resulting in lesaebibg of seizures and clinical improvement, reduce of drowsiness, improvement on development (the patient started sitting without support). She is still unable to speak, make eye contact, comprehend commands, persisting with motor stereotypies, but showing good containment of epileptic seizures. **Discussion:** This case describes a characteristic clinical manifestation of DEE-4, an autossomic dominant disorder caused by a pathogenic de novo variant c.364C> T: p.(Arg122Ter), which generates a premature mRNA stop codon, resulting in loss of function of gene STXBP1. This condition may cause developmental delay (DD), Intelectual disability, movement disorders, autistic characteristics and hyperactive behavior. Furthermore, this encephalopathy is commonly associated with electroclinical syndrones, such as infantile spasms and Lennox-Gastaut, and it's crisis can alter the patient's prognosis according to how it began. Besides, it was reported that 25% of patients have epileptic seizures refractary to anticonvulsivant therapy, making the use of cannabidiol and other alternative therapies. **Final comments:** Therefore, it is necessary for DEE-4 to be more studied by the scientific community, once early diagnosis allows personalized multidisciplinary interventions, contributing for the neuropsychomotor development of the carriers of this mutation. In addition, innovation of therapeutic options is fundamental for advance against the adversities provoked by this condition.

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Erdheim-Chester manifesting purely as a neurological disease: case report

Iago Resende Carvalho[1], Iasmim Resende Carvalho[2], Anna Maria de Senna Migueletto[1], Arthur Pereira Resende[3], João Paulo Sanches Zana[1], Laura Caroline Ferreira Borba[1], Victor Pereira Resende[3], Bruno de Carvalho Dornelas[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.
[2]Centro Universitário de Goiatuba, Goiatuba GO, Brazil.
[3]Instituto Master de Ensino Presidente Antônio Carlos, Araguari MG, Brazil.

Correspondence: Iago Resende Carvalho; Email: iagorcarvalho42@gmail.com.

Case presentation: A 60-year-old male, with a history of hypertension and deep vein thrombosis, presented with vertigo, paraparesis, and dysarthria for three months. On physical examination he had dysarthria, grade 3 hip flexion (bilaterally) and grade 4 plantar flexion and dorsiflexion of the feet, no sensory deficits, truncal cerebellar ataxia, and more pronounced left-side appendicular dysmetria. Magnetic resonance imaging (MRI) showed nodular masses involving the pons, both peduncles and cerebellar hemispheres, and an expansive lesion in the left cerebellar hemisphere, suggesting metastatic neoplasm. A month later, he was submitted to a craniectomy. Pathological findings showed a dense proliferation of histiocytes with abundant cytoplasm, giant Touton cells, and a moderate lymphocytic infiltrate consistent with Erdheim-Chester disease (ECD). On cytogenetic analysis, BRAF V600 mutation-positive tumor status confirmed. Then, vemurafenib therapy was initiated. On the 6-month follow-up, still in immunotherapy, he maintained dyslalia and left hemiparesis. He eats without assistance but does not deambulate. **Discussion:** We report a case of ECD in a 60-year-old patient with pure neurological signs. ECD is a rare multi-system non-Langerhan cell clonal histiocytosis, with less than 1000 reported cases, that may affect the CNS. The mean age of diagnosis is 57 years with a male prevalence. Its etiology involves activation of the mitogen-activated kinase pathway, leading to tissular accumulation of histiocytes. It is clinically heterogeneous, with bone pain in distal extremities in almost all cases. The most common neurological manifestations are cerebellar and pyramidal syndromes and even though those present in almost 41% of the patients, pure neurological disease is rare, with 39 cases reported. At MRI, the lesions are T2-hyperintense masses affecting mostly the pons, and cerebellum. Classic histological features are foamy histiocytes, multinucleated giant cells, and Touton giant cells. Immunohistochemical staining is positive for CD68 and negative for CD1a. The gold standard for diagnosis is the detection of any MAPK pathway alterations, especially BRAF mutation, that was present in our patient, the treatment for which is BRAF-inhibitors such as vemurafenib. It is important to consider other histiocytic disorders, solid

metastatic tumors, or primary central nervous system tumors in the differential diagnosis. In our case, the MRI suggested metastatic lesions, which were later proven a manifestation of the ECD. The prognosis is poor, as there is no known cure, and neurological involvement worsens its outcome. **Final comments:** ECD is a rare disease that may have only CNS symptoms, which greatly increases mortality. In patients with new-onset cerebellar and pyramidal syndromes with masses in imaging with unclear etiology, ECD should be considered as a differential diagnosis. Correlation of clinical features, histology, and radiological findings is essential.

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Erenumab for migraine: a systematic review and meta-analysis

Celso de Almeida Souza Miranda[1], Pedro Henrique Gunha Basilio[2], Ádem Ferreira do Nascimento[3], Agnes Zanotto Manoel[4], Artur Menegaz de Almeida[5], ítalo Barros Andrade[6], Lilianne Rodrigues Fernandes[7], Michele Kreuz[8]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Faculdade Pequeno Príncipe, Curitiba PR, Brazil.
[3]UniNorte, Manaus AM, Brazil.
[4]Universidade Federal do Rio Grande, Rio Grande RS, Brazil.
[5]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[6]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[7]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.
[8]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Celso de Almeida Souza Miranda; Email:

Background: Erenumab is a fully human monoclonal antibody that targets the calcitonin gene-related peptide (CGRP) that blocks selectively the CGRP receptor, a neuropeptide that plays a critical role in the pathophysiology of migraine. CGRP levels contribute to the transmission of pain signals, being elevated during migraine attacks. Targeting this pathway, erenumab reduces the frequency and severity of migraine episodes. However, as a novel treatment, erenumab safety and efficacy is still unclear. **Objective:** Assess the safety and efficacy of Erenumab in different dosages for patients with migraine through the analysis of reduction in Monthly Migraine Days and the intensity of symptoms by the analysis of the Headache Impact Test (HIT-6) and Migraine Physical function impact diary(MPFID) tests. **Methods:** A thorough literature search was conducted across PubMed, Scopus, and the Cochrane databases, focusing on randomized controlled trials comparing the efficacy and safety of Erenumab with a placebo. Mean differences were employed to evaluate the endpoints with 95% confidence intervals (CI) estimated through a random-effects model. We used I^2 statistics to evaluate heterogeneity. All statistical analyses were performed using the R statistical software (version 4.3.2). **Results:** We included 11 studies involving 4784 patients, of whom 2743 (57.33%) received Erenumab. The follow-up period ranged from 4 to 26 weeks. Compared with the placebo, there was a statistically significant difference between groups tending for erenumab in reducing Monthly Migraine Days (MD -1.64; 95% CI -1.90;-1.39; p<0.000001; I^2=25%), which was also demonstrated with the HIT-6 (MD -2.18; 95% CI (-2.61;-1.75); p<0.000001; I^2=0%). In the analysis of MPFID, there was an also statistical significance favoring Erenumab therapy, associated with the reduction of Migraine Physical Function Impact Diary- Everyday Activities (MPFID-EA) (MD -2.4; 95% CI (-3.59;-1.22); p=0.000071; I^2=90%) outcomes. **Conclusion:** This meta-analysis underscores the effectiveness of Erenumab in reducing Monthly migraine days and decreasing the intensity by reducing the HIT-6 and MPFID scores. Furthermore, the safety of the medication has been proven by analyzing the Relative Risk (RR) of the side effects. Some mentioned effects were Constipation with 95 from 2534 (3.74%) in the intervention and 38 from 2842 (1.33%) in the placebo group (RR 2.58; 95% CI (1.76; 3.77); p=0.000001; I^2=0%) favoring the placebo. But also, Fatigue with 31 from 1519 (2.02%) in the intervention and 27 from 1768 (1.52%) in the placebo group (RR 1.22; 95% CI (0.73; 2.03); p=0.454686; I^2=0%) and Nasopharyngitis with 205 from 2440 (8.4%) in the intervention and 243 from 4000 (6.07%) in the placebo group (RR 0.99; 95% CI (0.55; 1.77); p=0.975082; I^2=92%) but without statistical significance.

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Erenumab for treatment of episodic migraine with previous treatment failures: a systematic review and meta-analysis

Caio Luiz Nazar Cunha[1], Lucas Kenzo Ozera[1], Ana Clara Nogueira Cezar[1], Eduarda Lemos Gonzaga[1], Laura Caroline Felipe de Freitas[1], Lucas Figueira Vieira[1], Felipe Araujo Gouhie[1], Diogo Fernandes dos Santos[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Caio Luiz Nazar Cunha; Email: caiolnc@hotmail.com.

Background: Enumab is a fully human monoclonal antibody, whose use has emerged as an alternative for the preventive treatment of episodic migraine. However, the effects and outcomes of Erenumab use for this purpose still need to be clearly understood. **Objective:** To evaluate the efficacy of Erenumab treatment compared to placebo in adult patients with episodic migraine who have previously failed preventive treatment by analyzing randomized clinical trials. **Methods:** A comprehensive systematic review using the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) was conducted to evaluate the efficacy of Erenumab in adults with episodic migraine and a history of treatment failures. The analyzed outcome was the percentage of patients with at least > 50% reduction in Monthly Migraine Days (MMD). The search was conducted in the following databases: PubMed, Embase, and Cochrane. Statistical analysis was performed using R studio 4.3.1, and study heterogeneity was assessed using the I^2 index. **Results:** Four randomized clinical trials were analyzed, encompassing 2,693 patients with episodic migraine who had failed previous treatments. Erenumab was the treatment strategy for 1445 patients. Dosage of 70mg (OR 2.12; 95% CI; 1.16 - 3.87; p=0.03; I^2=80%) and 140mg (OR 3.57; 95% CI; 2.00 - 6.36; p=0.12; I^2=58%) was analyzed. A statistically significant symptomatic improvement was detected, favoring patients in the Erenumab group (OR 3.20; 95% CI; 1.96 - 5.24; p<0.01; I^2=87%). **Conclusion:** The analysis of four randomized clinical trials involving 2,693 patients revealed a statistically significant improvement in migraine symptoms among those treated with Erenumab compared to placebo. These findings suggest that Erenumab offers a promising option for patients with episodic migraine, opening up new possibilities for those who have not benefited from conventional preventive therapies.

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Escobar syndrome associated with arthrogryposis multiplex congenita: a rare case presentation

Gabriela Moraes Azevedo[1], Laís Brandão Mota[1], Cleo Alcantara Fernandes Silva[1], Camilla Rocha Medeiros Pinho[1], Rafael Amaral de Matos Albuquerque[1], Guíniver Couto Almeida[1], Gabriel Negreiros de Camargo Martins Moreira[1], Luna Souza Silva Vilanova[1], Marilaine de Matos Menezes Ferreira[1], Marcela Câmara Machado Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Laís Brandão Mota; Email: bmota.lais@gmail.com.

Case presentation: A 17-year-old female patient diagnosed with Escobar syndrome associated with arthrogryposis multiplex congenita (AMC) at birth attended a neurological clinic presenting pterygium and myopathy in both upper and lower limbs, associated with hypotrophy and muscle strength graded at +/IV. She uses long orthoses and specific shoes, since a scanometry identified a 12 cm asymmetry in the lower limbs, with the left femur being longer. Her osteotendinous reflexes are grade 2 in the upper limbs, but patellar reflexes were not obtained due to knee arthrodesis. The patient has undergone previous surgeries for clubfoot correction and an unspecified knee surgery, resulting in a lack of flexibility at 180°. Additionally, she experiences difficulty extending the second distal quadrant bilaterally, with ulnar deviation of the fingers and a single transverse palmar crease. The patient presents an S-shaped scoliosis and rapidly progressing lumbar hyperlordosis, scheduled for surgical correction. Tongue atrophy and fasciculations, along with alterations in the lips, buccinator, soft palate, and a short, winged neck, were observed. She reports a hypernasal and hoarse voice, reduced loudness, pneumophonoarticulatory incoordination and dysphagia, requiring her to crush pills before ingestion. Patient is underweight (BMI: 13.88 kg/m^2). She undergoes irregular lung expansion therapy with a bag valve mask and uses Bilevel Positive Airway Pressure (BiPAP) at night, but shows progressive deterioration of respiratory function, with reduced vital capacity (950mL) and forced vital capacity (750mL). Laboratory tests reveal decreased creatinine (0.4mg/dL), elevated mean corpuscular hemoglobin concentration (33.7g/dL), and increased red cell distribution width (13.5%). Patient is being monitored by an orthopedist, pulmonologist, and motor/respiratory physiotherapist. She also had a first-degree cousin who died with a similar condition. **Discussion:** This case describes a patient with rare diseases: Escobar syndrome, a genetic and attenuated form of multiple pterygium syndrome, and AMC, characterized by joint contractures at birth. The diagnosis was confirmed by medical history, physical examination and additional tests. The negative progression of respiratory function and spinal deformities is a classic manifestation of the diseases, impairing mobility and quality of life, especially with irregular adherence to physiotherapeutic treatment. The rarity of these conditions underscores the importance of family genetic counseling. **Final comments:** Managing rare diseases such as Escobar syndrome and AMC emphasizes the crucial need of a multidisciplinary approach to ensure comprehensive care for the patient. The progression of complications is the primary challenge, as there is no definitive cure, only palliative treatments. Further research is essential to expand knowledge about these diseases, aiming to improve the prognosis and quality of life for the patient.

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Etiological profile and outcome of people with optic neuritis treated at a neurological clinic between 2005 and 2022 in the interior of the Brazilian Amazon

Marcos Manoel Honorato[1], Savio Fernandes Soares[1], Lucas Silva Maia[1], Fernanda Lira Duarte[1], Frank Josué Costa Do Nascimento[1], Ana Karoline Pereira Camarão[1], Kelson Vinicius Dallabrida Padilha[1], Samuel Oliveira de Amorim[1], Bernardo Barbosa Penalber[1], Átila Barros Magalhães[1]

[1]Universidade do Estado do Pará, Santarém PA, Brazil.

Correspondence: Marcos Manoel Honorato; Email: marcos.honorato.dr@gmail.com.

Background: Optic Neuritis (ON) includes any inflammation involving the optic nerve and it is closely related to nerve demyelinating processes, typically idiopathic, when not associated with Multiple Sclerosis (MS) or Neuromyelitis Optica (NMO). Viral infections can also be implicated. Estimated annual incidence of new-onset ON ranges from 0.56 to 5.1 cases per 100,000 person-years. The presentation of ON can be typical (unilateral visual loss, subacutely, with ocular pain and mild disc edema) or atypical. Severe visual loss (loss of light perception), optic disc or retinal hemorrhages, significant edema, uveitis or exudates and bilateral involvement is considered atypical. **Objective:** Characterize the etiological profile and treatment response of individuals with Optic Neuritis treated at a private clinic in the Amazon from 2005 to 2022. **Methods:** This retrospective, descriptive, cross-sectional study employed a quantitative approach to analyze data from 32 individuals diagnosed with Optic Neuritis and treated at a neurological clinic from 2005 to 2022. It examined sociodemographic and clinical characteristics, complementary tests, definitive diagnoses, treatments, and therapeutic responses. Fisher's exact test was used to assess the correlation between categorical variables and clinical improvement of Optic Neuritis. **Results:** Females accounted for 71.88% of the sample. Among the patients treated, the mean age at diagnosis was 30.62 years, ranging from ± 11.23 years. As to etiologies, idiopathic cases accounted for 56.25%, followed by multiple sclerosis (18.75%), neuromyelitis optica (12.50%) and collagenosis (12.50%). Among the clinical aspects, bilateral visual symptoms were identified in 46.88% of the cases, ocular pain in 37.50%, in addition to motor and/or sensory alterations related to the underlying disease (78.14%), with a predominance of hemiparesis (21.88%). Neuroimaging showed abnormalities in 71.88%, with optic nerve abnormalities in 21.88%. Abnormal cerebrospinal fluid corresponded to 34.88%, the mean number of cells found was 8.37±33.23 and the mean number of proteins was 49.18±57.24, with an increase in gamma globulin of 21.88%. Anti-aquaporin 4 antibody was positive in 3.13%. Therapy occurred in 78.13%, interventions such as pulse therapy within one month of symptom onset (56.25%), isolated use of oral corticosteroids (34.38%) and use of immunoglobulin were adopted. The outcome revealed partial improvement after medication (81.25%) and, to a lesser extent, complete recovery after its use (12.5%). Pulse therapy was effective in 88% of cases, while 80% of those who used oral corticosteroids alone improved. **Conclusion:** Although no statistical correlation or predictors of a more favorable prognosis were identified, a significant improvement in optic neuritis was observed in patients undergoing some type of treatment. This discrepancy can be attributed to the small sample size.

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Evaluating the impact of mechanical thrombectomy availability on ischemic stroke outcomes in Brazil's public health system

Victoria Cavalcanti de Souza[1], Alice Volpato Rocha[1], Ester Namie Hanai[1], Laura Louise Marques Nobrega Vieira[1], Gabriel Volpato Rocha[1], Viviane de Hiroki Flumignan Zétola[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Victoria Cavalcanti de Souza; Email: victoriacavalcantisouza@gmail.com.

Background: Mechanical thrombectomy increases by three times the likelihood of a patient with acute ischemic stroke, caused by the occlusion of a large cerebral artery, achieving functional independence and avoiding major sequelae if performed within 8 hours after the onset of symptoms. Additionally, the recanalization rate after thrombolysis is 30% in patients with proximal middle cerebral artery occlusion and less than 10% in those with carotid artery occlusion. However, in many public healthcare services in Brazil (SUS), thrombolytic therapy is still more frequently used over thrombectomy, and in most cases, thrombectomy is not an available option. **Objective:** To assess the number of patients who could benefit from mechanical thrombectomy if it were widely available in the Brazilian public healthcare system (SUS). **Methods:** This is an observational and retrospective study, in which patients referred to the Stroke Protocol via local emergency medical services (SAMU) to a referral hospital in Curitiba from January 2023 to May 2024 were analyzed. Data regarding the time of stroke onset, arrival at the hospital, and adherence to DEFUSE-3 criteria were collected through active search in the medical records. **Results:** The medical records of 176 patients who arrived at the referral hospital through the Stroke Protocol via (SAMU) and were diagnosed with ischemic stroke were reviewed. Of these, only 56 (31.8%) underwent thrombolysis, and 44 (25%) patients had a time interval of six to twenty-four hours from the event onset to hospital arrival, making them potentially eligible for mechanical thrombectomy. **Conclusion:** Although mechanical thrombectomy is now available in the Brazilian Unified Health System (SUS), only 12 hospitals are equipped to perform it, with only one in the Northeast region (Hospital Geral de Fortaleza (CE)). Ischemic stroke patients within a six to twenty-four-hour time interval from stroke onset who underwent mechanical thrombectomy in addition to clinical treatment had greater functional independence compared to those who received only clinical treatment. Moreover, there is evidence that mechanical thrombectomy significantly increases the success rate of vessel recanalization and reduces the risk of neurological injury relative to clinical treatment alone.

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Evaluation along a comparison within Brazil's regions of factors linked to hospital internment from cerebral infarction and number of chemical thrombolysis used to treat them over the period of 2014-2023

Pedro Eduardo de Moura Souza[1], Juliana Cerqueira Gomes[2], Ana Carolina Campos Carvalho[2], Antônio de Souza Andrade Filho[3], Bianca Oliveira Tavares[4], Cecília Leite Dias[2], Frederico Luiz da Silva Figueirôa[3]

[1]Universidade do Estado da Bahia, Salvador BA, Brazil.
[2]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[3]Fundação de Neurologia e Neurocirurgia, Instituto do Cérebro.
[4]Faculdade Zarns, Salvador BA, Brazil.

Correspondence: Juliana Cerqueira Gomes; Email: juligomesc5@gmail.com.

Cerebral infarction is a condition that interrupts the blood flux from the brain generated by thrombus, emboli or other elements. Chemical thrombolysis is one of the therapeutics used in the acute phase of the disease when the patient has indication and is on the time window. Evaluate and compare Brazil's regions with factors linked to hospitalizations from cerebral infarction and number of chemical thrombolysis done to treat them in 2014-2023. It's a temporal series ecological study with data from the Hospitalar Information System linked to the Informatics Department of the Unified Health System along an analysis of the number of Hospitalar internment authorizations, mean of the length of stay, deaths and in-hospital mortality rate for cerebral infarction and the number of thrombolysis used to treat this condition, ICD10 code 0303040300, in the period of 2014 and 2023. The data was tabulated in Excel 2023 with calculation of dispersion and central tendency measures. The statistical analysis happened with the BioEstat v5.3 software, adopting p<0.05 as meaningful. In the period, there were 209,377 (20,938±2,780) hospitalizations in Brazil, with the Southeast presenting 45% of them and possessing the biggest mean (9,473±1,943) (p<0.05). Furthermore, there were differences between the mean of all regions except amidst the North and the Central-West (p<0.05). The mean of the length of stay in days had a lower median in the South (7.9±0.3) being similar to the Northeast (8.1±0.5) with statistical difference with all other clusters (p<0.05). There were a total of 31,865 deaths in Brazil (Mean: 3,186±407) with 50% of them in the Southeast (Mean: 1,599±363) that presented the biggest mean of all regions (p<0.05). The mean of the in-hospital mortality rate was higher in the North (17.5±3.2) with statistical difference with the Northeast (14.5±1.7) and the Central-West (14.1±2.2) (p<0.05). The South had the lowest median of the in-hospital mortality rate (12.6±0.8) with a difference happening when paired with the North (17.57±4.88) and Southeast (16.7±1.45) (p<0.05). There were a total of 31,405 thrombolysis done in Brazil (Mean: 3,140±1,572) with the Northeast responsible for 35% (1,013±611), the South for 30% (946±285) and the Southeast for 29% (922±578) with a difference of means happening with these regions and the Central-West (168±140) (p<005), but without differences between them. The North had 18 procedures done (Median: 0±0.25) with statistical difference with all the regions except the Central-West (168±140) (p<0.05). The majority of deaths and hospitalizations happened in the Southeast. The North had the highest in-hospital mortality rate and the lowest number of thrombolysis done meanwhile the Northeast and South had the majority of thrombolysis done and had the lower mortality rates. This could show regional disparities in the follow-up and investment destined to this condition.

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Evaluation and differentiation of edema and tumor infiltration in patients with high-grade glioma: initial experience using T2 relaxometry tests

Letícia Mansano de Souza[1], Sérgio Lourenço Polo Filho[1], Brunno Machado de Campos[1], Thiago Magalhães de Souza[1], Cleiton Formentin[1], Marcus Vinicius Giglio[1], Fernando Cendes[1], Enrico Ghizoni[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Sérgio Lourenço Polo Filho; Email: s205772@dac.unicamp.br.

The differentiation between edema and tumor infiltration in high-grade gliomas is fundamental for surgical planning. However, this distinction is challenging using conventional magnetic resonance imaging (MRI). New advanced magnetic resonance imaging (MRI) techniques are capable of inferring tissue composition, potentially contributing to such differentiation. In this study, we used magnetic resonance images in multi-echoT2 sequence (RMT2m) processed using a quantitative technique known as relaxometry to differentiate between edema and tumor infiltration in high-grade gliomas. In this study, we report our initial experience using multi-echoT2 relaxometry to differentiate perilesional edema from tumor infiltration in patients with high-grade gliomas. Patients who underwent RMT2m and with an anatomopathological diagnosis of brain tumor were retrospectively selected, divided into two groups: metastasis/meningiomas, in which it is assumed from the literature that hyperintensity in RMT2m corresponds to vasogenic edema, and high-grade gliomas, where there are present both edema and tissue infiltrated by tumor cells. The areas of hyperintensity in RMT2m were manually delimited in both groups and analyzed using the Relaxo-LNI software, developed in-house. In the group with high-grade gliomas, this analysis was subdivided into regions of interest in the center (close to the capturing lesion) and in the periphery of the tumors (far from the capturing lesion). Ten patients were

included in the group with metastases/meningiomas and twenty patients in the group with high-grade gliomas. The mean relaxation times in RMT2m were higher for the high-grade glioma group when compared to the control group (p<0.05). The relaxometry times were also longer means of relaxation of central areas of high-grade gliomas (mean value of 498.68), where tumor infiltration is the most common component, in relation to peripheral areas (mean value of 136.13), p<0.05, where edema tends to be predominant. The mean T2 relaxometry value in the periphery of the glioma was significantly closer to the T2 values observed in patients with meningiomas and metastases, which have a mean value of 196.8, p< 0.05. This proximity in values suggests that areas peripheral to the glioma exhibit characteristics that are more similar to edema than to tumor infiltration difference was statistically significant (p<0.05). Statistical results suggest that the RMT2m relaxation times' analysis - in this study using the Relaxo-LNI software - could be helpful to differentiate edema and tumor infiltration in patients with high-grade gliomas, allowing for a more efficient preoperative planning and postoperative assessment of tumor infiltration resection rate. Next steps include determining cut-off values for relaxation time where it is safe to assert the absence of tissue infiltrated by tumor cells, contributing to pre-operative planning and post-operative assessment of the extent of resection of these lesions.

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Evaluation of atypical festinating gait in a patient with Wilson's disease

Pedro Henrique Ximenes Ramalho Barros[1], Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Celli Veloso Cavalcanti[1], Kayron Rodrigues Lima[1], Jonas Leite de Souza Filho[1], Gustavo Viana Castro[1], Luma Rodrigues da Silva[1], Dário Correia Negreiros[1], Tiago José Do Nascimento Teixeira Leite[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Pedro Henrique Ximenes Ramalho Barros; Email: pedrohx587@gmail.com.

Case presentation: A 35-year-old man, diagnosed with Wilson's disease (WD) for 9 years, was admitted with reports of insomnia, affective lability with pseudobulbar humor and irritability, hypotimia, negativism, anticipatory suffering, self-referential and death thoughts, low self-esteem, persecutory delusions, and challenging behavior, which started 8 months ago with progressive worsening. On physical examination, he presented choreiform facial movements, bilateral resting tremor, generalized muscular dystonia, hardness in gait, inability to stand without support, and limited to the wheelchair. During the dynamic gait, the patient had a festinating gait with Bobbing characteristics and was unstable with predominate freezing of gait in all the states of the gait. The patient's speech and swallowing are impaired. Escitalopram 10mg/day was started without improvement, which was then replaced by Mirtazapine 30mg/day and Quetiapine 25mg/day, with slight mood improvement, reduced irritability, and delusions after 20 days of use. He discontinued the chelation therapy for WD 5 years ago. During the same period, he began continuous abuse of alcoholic beverages. The use of other psychotropic drugs was not reported. Currently, he refuses psychotropic medication, using only the chelator, thus maintaining agitation, impulsivity, and challenging behavior. **Discussion:** WD is a genetic condition affecting the ATP7B gene, essential for the transport and subsequent biliary excretion of copper. A defect in this pathway leads to a progressive accumulation of copper in the body, especially in the liver. Additionally, the metal can deposit in other tissues, causing a variety of symptoms, such as dysarthria, ataxia, dystonia, tremors, parkinsonism, and Kayser-Fleischer. The patient's physical evaluation presented a rare finding: the presence of a festinating, exhausting, and bobbing gait (more related to severe atypical parkinsonism). It was characterized by the progressive shortening of step length, a compensatory increase in cadence, and difficulty in postural control. **Final comments:** Early diagnosis is common, making treatment essential in controlling the progression of the disease. Thus, this report expresses atypical characteristics of WD, especially festinating gait, and the implications of discontinuing specific therapy. That WD can exhibit different patterns of presentation, reflecting the challenge of establishing an early diagnosis. Is probable that gait findings are due to chronic ineffective that prevents the onset of the first symptoms.

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Evaluation of benign multiple sclerosis beyond the EDSS: cognitive, fatigue and mood disorders

Josué de Almeida David[1], Thomas Vieira de Paula[1], Alfredo Damasceno[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Josué de Almeida David; Email: j219131@dac.unicamp.br.

Objective: This study aims to analyze cognitive and clinical aspects of patients with benign MS (BMS) to gain a deeper understanding of this condition. **Methods:** Sixty patients from a tertiary MS service were included in the study, with 30 individuals exhibiting an Expanded Disability Status Scale (EDSS) score below 3.0 and a minimum of 10 years since disease onset. Participants underwent cognitive evaluations as well as assessments for mood disorders, fatigue, and motor dysfunction. More stringent criteria for BMS were also tested. **Results:** Most patients identified as having BMS in this cohort were female, with a mean EDSS score of 1.9 and good performance on both the Timed 25-Foot Walk (5.2 sec) and Nine-Hole Peg Test (23.2 sec) motor assessments. Cognitive deficits were more prevalent in traditional MS patients compared to those classified as having BMS. The frequency of depression was similar between the two groups. Moreover, employing stricter criteria for classification did not yield superior clinical or cognitive outcomes. **Conclusion:** This study reinforces previous epidemiological findings observed in other countries within the context of the Brazilian population. Additionally, it highlights the prevalence of non-motor symptoms and cognitive impairment in both traditional MS and BMS. These findings underscore the need for a distinct approach when evaluating and classifying BMS patients.

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Evaluation of brain morphometry by magnetic resonance imaging in patients with psychogenic nonepileptic seizures

Fernando Cendes[1], Lucas Gabriel Alves Figueiredo[1], Luciana Ramalho Pimentel[1], Clarissa Lin Yasuda[1], Brunno Machado de Campos[1], Marina Koutsodontis Machado Alvim[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Lucas Gabriel Alves Figueiredo; Email: l201917@dac.unicamp.br.

Background: Psychogenic nonepileptic seizures (PNES) are episodes of alterations in consciousness and subjective experience accompanied by self-control impairment. Due to their similarity, patients with this condition are often misdiagnosed and treated as if they had epilepsy. A possible tool for better understanding PNES is neuroimaging. However, there are few such studies, and the information in the literature is inconclusive. **Objective:** Evaluate magnetic resonance imaging (MRI) morphometry - cortical thickness, cortical surface area, curvature, and volume of subcortical structures - in patients with PNES, mesial temporal lobe epilepsy (MTLE), and controls. **Methods:** 163 participants were divided into three groups: PNES (with or without epilepsy, n=40), MTLE only (n=54), and a control group (n=69). Image processing was performed using Freesurfer 6.0 software. Differences between groups means were evaluated with generalized linear models, including age as a covariate. Correction for multiple comparisons was performed using FDR and Sidak. **Results:** Regarding volume, 8 structures in the PNES group were larger (P<0.046) than in MTLE, including the corpus callosum, left hippocampus and left cerebellar cortex. The volume of 7 structures was smaller (P<0.041) in PNES compared to controls (bilateral thalami, nucleus accumbens, and total gray matter), and 30 structures were different (P<0.01) between MTLE x control, with the vast majority being smaller in MTLE as expected. Regarding curvature, the left entorhinal cortex was smaller (P=0.019) in PNES than in MTLE. However, curvature was increased in 13 structures (P<0.03) in PNES and 15 structures (P<0.03) in MTLE compared to controls. Additionally, the right rostral anterior cingulate cortex was thinner (P=0.011) in the PNES group than in MTLE, but there was no significant difference between PNES versus controls and MTLE versus controls. Thickness was also smaller in 17 structures (P<0.03) in PNES and 12 structures (P<0.035) in MTLE compared to controls. **Conclusion:** The regions with the differences between the groups identified are fundamental for cognitive, emotional, behavioral, and executive control. These morphometric differences may contribute to a better understanding of PNES and to defining biomarkers for differentiating PNES and epilepsy. However, further studies are needed to draw better conclusions.

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Evaluation of cerebello-cortical pathways using the virtual brain in a patient with cerebellar mutism: study methodology

Thalles Olivi de Almeida[1], Jose Reynaldo Walther de Almeida[2] , Brigida S Oliveira[1], Ana Carolina Campos[1], Marcos Vinicius Calfat Maldaun[1]

[1]Hospital Sírio-Libanes, São Paulo SP, Brazil. [2]Intituto de Neurociencias de Araraquara e de Barretos.

Correspondence: Thalles Olivi de Almeida; Email: almeida.thalles@outlook.com.

Background: Cerebellar mutism or posterior fossa syndrome (PFS) is a rare occurrence in adult patients undergoing surgery for cerebellar tumors. Brigida S Oliveira recently showed that tractography has limited utility in predicting PFS in children. Palesi et al. used The Virtual Brain (TVB) to evaluate whether the inclusion of cerebellar connectivities is important for simulating brain dynamics in healthy individuals. We evaluated an adult patient who underwent resection of a left cerebellar metastasis and developed severe posterior fossa syndrome immediately postoperatively. She underwent MRI with DTI and EEG with nonlinear analysis of the time series. **Objective:** To study the pathways: 1- cerebellum-thalamus-basal nuclei-cortex: primary motor cortex (1-M1), premotor cortex (1-PMC), supplementary motor area (SMA) and 2- cerebellum-thalamus-basal nuclei-cortex: dorsolateral prefrontal cortex (2-PFC), orbitofrontal cortex (2-OFC), using TVB and nonlinear metrics. **Methods:** After preprocessing the T1 and DTI data, the TVB avatar was generated using the Glasser atlas with 180 cortical regions and 17 subcortical regions. Empirical EEG from 19 channels generated the functional connectivities. We used the Jansen-Rit model in TVB. The

model parameters were adjusted according to the literature. The described networks were isolated and compared between hemispheres using mean mutual entropy and the relationship MPR-complexity x permutation entropy, global and local efficiency. **Results:** GLOBAL/LOCAL EFFICIENCY: 1-M1 right = 0.36 / 0.31 and left = 0.21 / 0.15. 1-PMC right = 0.35 / 0.36 and left = 0.23 / 0.21. 1-SMA right = 0.36 / 0.30 and left = 0.20 / 0.18. 2-PFC right = 0.39 / 0.32 and left = 0.19 / 0.12. 2-OFC right = 0.33 / 0.29 and left = 0.19 / 0.19. MEAN MUTUAL ENTROPY: 1-M1 right = 0.25 and left = 0.15. 1-PMC right = 0.28 and left = 0.09. 1-SMA right = 0.45 and left = 0.14. 2-PFC right = 0.39 and left = 0.09. 2-OFC right = 0.33 and left = 0.16. MPR COMPLEXITY / PERMUTATION ENTROPY MEAN: 1-M1 right = 0.254 and left = 0.219. 1-PMC right = 0.247 and left = 0.217. 1-SMA right = 0.244 and left = 0.225. 2-PFC right = 0.247 and left = 0.226. 2-OFC right = 0.234 and left = 0.225. **Conclusion:** The analyses showed interesting differences, with the left networks severely compromised, and suggest that TVB can be a useful tool in evaluating cerebello-cortical networks, with potential for clinical use in the future.

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Evaluation of cognitive impairment in long-severe COVID patients

Leidys Marina Pedrozo García[1], Marciéli Gerhardt[2], Fabio Jean Varella de Oliveira[1], William Alves Martins[2], Eduardo Leal Conceição[2], Helena Scartassini Erwig[1], Larissa Frozza Fernandes[1], Helena Morsch Marques[1], Francine Nunes Pereira[1], Jaderson Costa da Costa[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Marciéli Gerhardt; Email: marcieligerhardt@yahoo.com.br.

Background: "Long Covid" is an expression for describing the syndrome with signs and symptoms that emerge or continue after a Covid-19 acute infection. These symptoms must persist for over 12 weeks and are not explained by an alternative diagnosis. Persistent symptoms include breathing difficulty, fatigue, mental health disorders and cognitive impairment. **Objective:** To measure and characterize the cognitive impairment and psychiatric disorders related to Covid-19 infection. **Methods:** This is a cross-sectional study, approved by our institution's ethics committee (CEP:6.123.125), in a population-based cohort. Two hundred patients admitted to the ICU of a hospital in Porto Alegre due to severe Covid-19 infection and who were discharged from 01/15/2021 to 03/30/2021 were assessed for eligibility. Data regarding cognitive function (Montreal Cognitive Assessment [MoCA] and Addenbrooke's Cognitive Examination [ACE-III]), psychiatric comorbidities (Hamilton scales) and quality of life (Short Form Health Survey [SF-36]) were collected. **Results:** A total of 46 patients were enrolled, regardless of their cognitive status, with an average age of 52.2 years (DP 12.1). Twenty-four (54,3%) were male. The vast majority (95.6%) was not experiencing social isolation and had at least eleven years of education (91.3%). We found that many patients suffered from executive and visuospatial dysfunction, performing similarly in MoCA and ACE-III, with a median score of 3.87 (maximum score of 5; DP 1.29; P=0.099) and 14 (maximum score of 16; DP 1.08), respectively. Language was also impaired in our patients, with average scores of 2.52 (maximum score of 3; DP 0.72; P=0.380) and 24.82 (maximum score of 26; DP 1.82) in MoCa and ACE-III, respectively. Regarding memory, MoCA had an average score of 2.7 (maximum score of 5; DP 1.48), while ACE-III showed a score of 18.7 (maximum score of 26; DP 1.5). Unexpectedly, memory performance in both tests was significantly worse even in patients with more than 14 years of formal education. Of all patients, 30,5% (n=14) had normal cognition, 13.04% (n=6) had physical deficit without cognitive impairment, 34.78% (n=16) had mild cognitive impairment (MCI) and 21.73% (n=10) had dementia. The median scores in MoCA were 27.36 (DP±1,216), 27.67 (DP±0,816), 22.75 (DP±2,543) and 22.30 (DP±3,129) in each group, respectively. The median scores in ACE-III were 87.79 (DP±5,820), 92.33 (DP±5,502), 83.81 (DP±8,101) and 80.2 (DP±7,642), respectively. **Conclusion:** This study provides evidence for a protracted cognitive impairment years after severe Covid-19 infection, showing a combination of executive, visuospatial, language and memory impairments. Although other factors, such as prolonged ICU stay, may have had a role, our findings support that even relatively heathy patients suffer from Covid-19 cognitive dysfunction, which was displayed even more prominently in highly educated patients.

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Evaluation of epidemiological factors in patients diagnosed with meningitis in 2023 in Mato Grosso do Sul, Brazil

Emanuela Corrêa da Costa[1], Gabriela Corrêa da Costa[2]

[1]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.
[2]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Emanuela Corrêa da Costa; Email: emanuela.correa@ufms.br.

Meningitis, an inflammation of the meninges surrounding the Central Nervous System (CNS), can be of viral, bacterial, fungal, or parasitic origin, with the bacterial form being the most severe, potentially leading to death or leaving sequelae in survivors. The epidemiological expression of the disease is influenced by various factors, including the etiological agent, population clusters, socioeconomic characteristics, and climatic conditions. In Brazil, meningitis is a notifiable disease due to its severity, high mortality rate, and irreversible sequelae, especially in children. This study analyzed meningitis cases in Mato Grosso do Sul in 2023, using data from the Notifiable Diseases Information System (SINAN). There were 129 suspected cases recorded, with 80% in Campo Grande. Most patients were discharged from the hospital, especially men aged 20 to 59 years and of mixed race/color. Unspecified meningitis was the most common etiology, followed by bacterial. The chemocytological exam was the most used diagnostic method. Underreporting is a significant challenge, reflecting the need for continuous epidemiological surveillance and preventive measures. It is essential to direct special attention to children and the school environment, where preventive interventions can be particularly effective.

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Evaluation of facial expressions for the recognition of parkinsonian fasciations based on artificial intelligence learning

Pedro Henrique Ximenes Ramalho Barros[1], Luma Rodrigues da Silva[1], José Nazareno Alves Rodrigues[1], Rodrigo de Melo Souza Veras[1], Lara Beatriz Alves Batista[1], Samuel de Castro Campos[1], Jonatas Paulino da Cunha Monteiro Ribeiro[1], Dário Correia Negreiros[1], Tiago José Do Nascimento Teixeira Leite[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Pedro Henrique Ximenes Ramalho Barros; Email: pedrohx587@gmail.com.

Background: Facial recognition technologies are important in genetic, endocrinological, neuromuscular and neurodegenerative diseases. Non-invasive diagnostic tools and strategies based on videos and photos can generate automated facial analysis software whose integration with artificial intelligence learning fosters the development of new products. Facial hypomimia, often present in patients with Parkinson's disease (PD), is characterized by impaired facial expressiveness, with decreased orofacial and eye movements. Thus, based on patterns found in parkinsonian facies, the use of Machine Learning is promising, becoming an auxiliary technological mechanism for identifying facial expressions. **Objective:** To develop a database and an algorithm for recognizing facial image parameters in subjects with idiopathic parkinsonian syndrome, to evaluate its accuracy and to identify the best facial expression for recognizing facial hypomimia. **Methods:** This was an experimental, cross-sectional, non-interventional study that included patients diagnosed with parkinsonian syndrome, assessed by a neurologist using the London Brain Bank criteria, who underwent photo capture of facial expressions (surprise, happiness, fear, disgust, anger and sadness). With the database generated and also using the "FER 2013" database, an algorithm was formulated using technology based on training from examples through supervised learning by inputs and outputs calculated by an unknown function y = f(x) where the pairs (Xn, Yn) of a new function (h), approximate a true function, using a Convolutional Neural Network (CNN). Performance was evaluated using a Binary Classification Confusion Matrix and the information obtained generated total accuracy, precision and recall variables. **Results:** Accuracy for PD patients was 0.18, with recall rates for anger, disgust, fear, happiness, sadness and surprise of 0.00; 0.00; 0.10; 0.57; 0.24 and 0.00, respectively. Patients without PD had an accuracy of 0.17, with recall of anger, disgust, fear, happiness, sadness and surprise of 0.07; 0.00; 0.11; 0.23; 0.24 and 0.00. When a new model using only the emotions of sadness and happiness was analyzed, the accuracy of the method rose to 0.57. **Conclusion:** A database and an algorithm for recognizing facial image parameters in idiopathic parkinsonian syndromes have been developed. The initial accuracy when evaluating 6 emotions was low. The best facial expression for recognizing facial hypomimia was sadness. The artificial intelligence learning method proved to be a potentially promising tool for identifying subjects with parkinsonism.

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Evaluation of gait and balance in patients with spinocerebellar ataxia type 1 using wearable inertial sensors

Nadson Bruno Serra Santos[1], Camila Caroso Lobo[1], Rachel Paes Guimarães[1], Fabricio Diniz de Lima[1], Alberto Rolim Muro Martinez[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Nadson Bruno Serra Santos; Email: nadson_bruno@hotmail.com.

Background: Spinocerebellar ataxias (SCA) is a subtype of hereditary cerebellar ataxia, with an autosomal dominant pattern, which presents with cerebellar degeneration, involving multiple pathways that connect to this structure and the brainstem. The natural history of SCA1 involves a long pre-symptomatic period, and the first symptoms starts at the 4th and 5th decade of life, the most common of which are gait ataxia and incoordination, nystagmus and dysarthria. **Objective:** To evaluate gait and balance in patients with spinocerebellar ataxia type 1 (SCA1) using Wearable Inertial Sensors. Materials and **Methods:** It's an open study, cross-sectional, for clinical evaluation of patients with SCA1, comparing to controls. It was recruited 10 patients treated at the Neurogenetics outpatient clinic at the Tertiary Hospital and 10 healthy subjects. The

participants used six inertial sensors Mobility Lab TM (APDM, Inc.) while executing a 3-step protocol (Sitting down and getting up from a chair 5 times; Standing in orthostasis for 30 seconds and Walking in a 10-meter range for 2 minutes). **Results:** The SCA1 and Control groups were composed by 7 women and 3 men, with a mean age of 43.8 years and SARA 9.2 in SCA1 group, and 49.7 years and SARA 0.05 in controls group. The measures that were most related, according to Pearson's correlation were Centroidal Frequency (r=0.689; p=0.010); Step Length (r=0.512; p=0.003) and Axis Area (r=0.206; p=0.022), presenting directly correlation to the severity of the SARA scale. Step Length stands out (d=9.628; p=0.003); Axis area (d=4.520; p=0.022); Turning Time (d=0.366; p=0.002) and Number of Steps (d=0.358; p=0.013) were markers of disease progression with the largest effect size in Cohen's correlation (d). **Conclusion:** This is the pilot study in Brazil with the SCA1 population, revealing that the use of Wearable Inertial Sensors is useful for the evaluation and clinical follow-up of patients with SCA1. Incorporating the data from the Wearable Inertial Sensors into clinical practice can optimize care regarding the balance of our patients, with a great impact on evaluation in clinical trials, given its potential to measure silent progression.

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Evaluation of micrornas as biomarkers of malignant evolution in patients with diagnosis of glioma

Caroline Souza dos Anjos[1], Victor Cunha Tirapelli[1], Ana Laura Ribeiro e Silva Stevanato[1], Lucas Cunha Tirapelli[1], José Guilherme Marques Farinha[1], Andressa Romualdo Rodrigues[1], Fernanda Maris Peria[1], Carlos Gilberto Carlotti Jr[1], Mucio Luiz de Assis Cirino[1], Daniela Pretti da Cunha Tirapelli[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: José Guilherme Marques Farinha; Email: zeguifarinha@hotmail.com.

Background: Gliomas are neuroepithelial tumors and correspond to approximately 24.6% of all primary brain tumors. In the last decade, great effort has been made in academic circles to characterize gliomas in an attempt to describe clinical behavior, define prognosis and predict therapeutic response. A current problem in clinical practice is the absence of molecular tools to predict the evolution of gliomas classified as low grade, since during postoperative radiological follow-up these patients may have tumor recurrence with histopathological confirmation of high glioma degree. Tumor transformation in high-grade glioma has a worsening of prognosis and reduction of therapeutic possibilities. **Objective:** To analyze the expression of miR- 124a, miR-138, miR-155, miR-1275 in human primary and peripheral blood samples from patients diagnosed with low and high grade gliomas. **Methods:** MicroRNA expression analyzes were performed using the real-time PCR technique. Sixty-five adult patients (18-65 years of age) with confirmed histopathological diagnosis of glioma with cryopreserved tumor biological material, were analyzed. Information collected in the medical records regarding clinical, epidemiological, clinical and radiological characteristics and time for recurrence and death were collected. Considering a level of significance of 5%, the association of categorical qualitative variables and microRNA expression were performed by the Mann-Whitney test. Survival analysis was performed using the Kaplan-Meier non-parametric method. **Results:** The microRNAs under study did not present hyperexpression in tumor tissue of patients diagnosed with glioblastoma. Only miR-1275 showed increased expression in patients diagnosed with pilocytic astrocytoma and grade II oligodendroglioma. In addition, tumors of low and high grade oligodendroglial lineage demonstrated overexpression of miR-1275. In peripheral blood, significant hypoexpression of miR-1275, miR-124 and miR-138 were observed in a glioblastoma sample. There was also a marked hypoexpression of miR-138 in samples of grade III oligodendroglioma. The microRNAs miR-124a, miR-138, miR- 155 and miR-1275 did not show hyperexpression in tumor tissue of patients diagnosed with GBM. MiR-1275 showed increased expression in patients diagnosed with pilocytic astrocytoma and miR-155 was hyperexpressed only in samples of grade II oligodendroglioma. In addition, tumors of oligodendroglial lineage, of low and high grades, demonstrated hyperexpression of miR-1275 and miR-124a. In peripheral blood, significant hypoexpression of miR-1275, miR-124 and miR-138 were observed in the GBM sample as well as marked hypoexpression of miR-138 in samples of grade III oligodendroglioma.

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Evaluation of motor results in patients with Parkinson's disease undergoing deep brain stimulation implantation at the neurological institute of Curitiba

Luiza Gonçalves Fraga[1], Joel Fernando Sanabria Duarte[1], Daniel Benzecry de Almeida[1], Murilo Sousa de Meneses[1], Marcela Ferreira Cordellini[1]

[1]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.

Correspondence: Luiza Gonçalves Fraga; Email: luizaag.fraga@gmail.com.

Background: Parkinson's disease (PD) is one of the most common neurological diseases and an important cause of disability in individuals of working age. Treatment is aimed at controlling symptoms and there is no therapy capable of curing or stopping the progression of the disease. One of the approved therapies for treatment is DBS. This study evaluated the effectiveness of DBS on motor symptoms in patients who underwent the procedure and who underwent preoperative evaluation with the levodopa overload test. **Objective:** To evaluate the effects of deep brain stimulation (DBS) on motor symptoms in PD in patients undergoing the procedure at the Neurological Institute of Curitiba (INC), a Brazilian tertiary hospital. Compare motor symptoms pre and post-DBS implantation through part III of the MDS-UPDRS, evaluate whether there was a reduction in medication after DBS implantation and assess the impact on quality of life after DBS implantation through the questionnaire on DBS Parkinson's disease (PDQ 39). **Methods:** 8 patients were evaluated before and after DBS implantation. For this purpose, the MDS-UPDRS was used. In addition to motor symptoms, medication reduction and quality of life were assessed using the Parkinson's disease questionnaire (PDQ39). The data were placed in an Excel spreadsheet and then statistically analyzed using the JAMOVI statistical software version 2.5.0. **Results:** The average age at DBS implantation was 62.5 years and average disease duration was 9.89 years. In 87.5% the target was the subthalamic nucleus. There was a reduction in medication after the procedure with a mean difference in levodopa equivalent dose of 337mg (p 0.024). It was shown with statistical significance that DBS improved motor symptoms, either alone (p 0.008) or in conjunction with medication (p 0.032). **Conclusion:** After the implantation of DBS, there was a reduction in symptoms and medication used to treat PD after surgery. The PDQ39 assessment suggests an improvement in quality of life, as seen in another Brazilian study.

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Evaluation of MRI findings associated with benign multiple sclerosis

Josué de Almeida David[1], Thomas Vieira de Paula[1], Alfredo Damasceno[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Josué de Almeida David; Email: j219131@dac.unicamp.br.

Objective: This study aims to analyze neuroimaging aspects of benign multiple sclerosis (BMS) and traditional multiple sclerosis patients. **Methods:** Sixty patients from a tertiary MS service were included in the study, with 30 individuals exhibiting an Expanded Disability Status Scale (EDSS) score below 3.0 and a minimum of 10 years since disease onset. They underwent clinical evaluation and analysis of the number of cerebral, infratentorial, and spinal cord T2-hyperintense lesions on Magnetic Resonance Imaging (MRI). More stringent criteria for BMS were also tested. **Results:** Patients with worse clinical prognoses exhibited higher numbers of infratentorial and spinal cord lesions. The traditional MS patients had a mean medullary number of lesions of 4.16, while the BMS patients had a mean of 2,33 lesions (p=0.01). We also found out that 11 patients had no medullary lesions, while there were no traditional MS patients without medullary lesions in the MRI. Additionally, patients with a more stringent criteria for BMS definition did not show different neuroimaging parameters. **Conclusion:** This study shows that spinal cord and infratentorial lesions are associated with a worse clinical prognosis and that low/absence of spinal cord lesions is a hallmark of BMS.

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Evaluation of prognosis from admission data in patients diagnosed with stroke accident in a reference hospital in Bahia

Larissa Nunes Almeida dos Santos[1], Joanna Sousa da Fonseca Santana[1], Amanda Hellen Sousa Leal[1], Ana Clara de Lemos Guimarães[1], Mariana Rodrigues dos Santos[1], Gracchо Cardoso da Silva Neto[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Amanda Hellen Sousa Leal; Email: amanda.h.f1@gmail.com.

Background: Stroke encompasses a group of pathologies associated with impaired cerebral blood flow and its vessels, with diverse clinical presentations of high epidemiological relevance worldwide. The management of this urgent clinical condition, from admission, requires complementary tests to monitor conditions that influence therapeutic decisions, such as hyponatremia, hypertension, and hyperglycemia. Therefore, it is essential to understand the impact of these conditions on the patient's prognosis and the association between clinical interventions and mortality. **Objective:** Analyze and describe admission data, prognosis, and mortality outcomes in stroke patients. **Methods:** This is a descriptive, observational, retrospective clinical study based on secondary data, as a preliminary result from a major stroke study with 6,993 patients, collection from the electronic medical records of patients previously admitted to a reference hospital in Salvador, Bahia. The study population comprises patients diagnosed with stroke, aged over 18 years, who were admitted between January 2021 and December 2022. The study sample is non-probabilistic by convenience. The variables of interest are: Age, Sex, Type of Stroke, Capillary Blood Glucose, Diastolic and Systolic Blood Pressure, Serum Sodium on Admission, and Death. **Results:** Data from 221 patients were collected. 122 patients (55%) were female. 128 patients (58%) were aged 65 or older. Ischemic stroke was diagnosed in 82% and hemorrhagic stroke in 13.5%. 59 patients (26.6%) died. Analyzing serum sodium, the highest incidence of deaths, 39.13%, was in patients with values below 135mmol/L. Among patients between 136-145mmol/L, the mortality rate was 25.79%. Regarding capillary blood glucose, patients with values between 126-200mg/dL, 33.3% died; between 200-400mg/dL, the mortality was 37.5%. In patients with blood glucose up to 126mg/dL,

the death rate was 9.62%. Analyzing systolic blood pressure, in patients greater than or equal to 180mmHg, 13.64% died. Those with SBP between 90-130mmHg, 50% died. Regarding diastolic blood pressure, patients greater than or equal to 110mmHg, 29.55% died. Those with DBP between 60-85mmHg, 31.25% died. **Conclusion:** The analysis revealed relevant findings about the clinical outcomes of stroke patients, with a predominance of females and patients aged 65 or older. Ischemic stroke was the most prevalent. The overall mortality rate was 26.6%, with significant variations associated with different levels of serum sodium, capillary blood glucose, and blood pressure. Patients with serum sodium below 135mmol/L had the highest incidence of deaths, 39.13%, those with capillary blood glucose between 200-400mg/dL had a mortality rate of 37.5%. Blood pressure was a crucial factor, with a mortality rate of 50% in patients between 90-130mmHg. These findings highlight the importance of rigorous monitoring and appropriate management of clinical parameters to improve outcomes in stroke patients.

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Evaluation of pulse therapy with intravenous methylprednisolone on leprosy neuritis: an observational study on a Brazilian referral center

Eduardo William Farinha Carvalho[1], Ana Clara Nogueira Cezar[1], Ariane Aparecida Correa de Miranda[1], Caio Luiz Nazar Cunha[1], Laura Caroline Felipe de Freitas[1], Lucas Figueira Vieira[1], Lucas Kenzo Ozera[1], Leonardo Peixoto Garcia[1], Isabela Maria Bernardes Goulart[1], Diogo Fernandes dos Santos[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Eduardo William Farinha Carvalho; Email: eduardowilliam0603@gmail.com.

Background: During the natural history of leprosy, several patients present immune hyperactivity, known as leprosy neuritis (LN), that can occur before, during, or after multidrug therapy. LN is responsible for the deformities and sequelae present in this disease, making this neuropathy a potentially disabling condition. Therefore, as in other inflammatory neuropathies, intravenous corticosteroids should be considered in the treatment of LN, especially in severe and refractory cases. **Objective:** To evaluate the use of intravenous methylprednisolone in refractory LN in a national reference center of leprosy in Brazil. **Methods:** This observational study included leprosy patients who underwent intravenous methylprednisolone for refractory LN treatment. Epidemiological and clinical data were collected. The following Instruments were used in the evaluation: visual analog pain scale (VAS), Douler Neuropathic 4 questionnaire (DN4), Screening of Activity Limitation and Safety Awareness (SALSA), and the simplified neurological assessment protocol used by the Ministry of Health in Brazil. Data obtained was compared with the results of the initial and end of pulse therapy with methylprednisolone. **Results:** We evaluated 18 patients with a male predominance. The majority (66.7%, 12/18) presented the LR after the end of treatment. After pulse therapy, there was no difference regarding sensory and motor scores in all nerves evaluated, confirming the maintenance of the previously observed deficits. At the beginning of the treatment, 66.7% (12/18) of patients were using oral prednisone at 35.0mg (±12.43). After using methylprednisolone, 38.9% (7/18) of the individuals maintained oral prednisone at a lower dose (16.4mg ±6.3; p=0.002). Regarding the SALSA scale, the score significantly improved at the end of treatment (p=0.020). Regarding neuropathic pain, there was a substantial drop in the DN4 scores from 6.7 (±1.7) to 5.2 (±2.1) (p<0.0001). **Conclusion:** This study showed that pulse therapy with endovenous methylprednisolone in LN was effective in decreasing pain intensity, preventing worsening of sensory-motor function, and reducing the dose of corticosteroids, reflecting the patient's improvement of functionality.

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Evaluation of quantitative muscle ultrasound in monitoring the progression of Duchenne muscular dystrophy

Karlla Danielle Ferreira Lima[1], Pedro Henrique Marte Arruda Sampaio[1], Marco Antônio Veloso Albuquerque[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Karlla Danielle Ferreira Lima; Email: karlla.dfl@gmail.com.

Background: Duchenne muscular dystrophy (DMD) is a severe, progressive, muscle-wasting with an X-linked recessive inheritance caused by mutation in DMD gene. It is cause progressive muscle weakness that begins in early childhood in boys. Progressive weakness results in loss of ambulation early, and death from respiratory impairment or cardiac dysfunction. Quantitative muscle ultrasound (QUS) in boys with DMD shows increased echo intensity as muscle is replaced by fat and connective tissue. There are few studies evaluating the patterns of muscle involvement using ultrasound in patients with DMD. **Objective:** To determine the pattern of involvement and its relationship with the evolution of the disease. **Methods:** This is a prospective observational study from a single center, carried out at the Muscle Clinic of a reference center, from January 2022 to July 2023, in 42 patients with DMD between 6-18 years of age (average of 11 years). QUS of 12 muscle groups (axial, appendicular and respiratory), the mobility and thickness of the diaphragm (TE) [thickness during inspiration (Tinsp) and expiration (Texp)] were assessed in all patients using ultrasound and the Image J program. **Results:** In the upper limbs, the most affected muscle was the biceps brachii, while in the lower limbs it was the rectus femoris. Whilst for the respiratory muscles, the intercostal muscle had a higher average involvement than the diaphragm. The correlation between muscle histograms and the age variable was statistically significant only with the 1st interosseous, biceps brachii, medial gastrocnemius and digastric muscles. When evaluating diaphragm mobility using thickness measurement, only one patient had diaphragm paralysis (thickening fraction <20%), at 12 years of age. The average TE was 89%, ranging from 13 to 182%. There was no statistical significance between TE and age, while there was a positive statistical correlation with Tinsp and Texp (p<0.06). **Conclusion:** Our study demonstrated that ultrasound can be used as a method to help monitor disease progression with age, but not all muscles correlate with statistical significance. In the age range studied, there was no reduction in diaphragmatic thickness in patients with increasing age. However, our sample was composed of very young patients with an average of 11 years of age. Interestingly, our results demonstrated a positive correlation between age and Tinsp and Texp, suggesting that in a younger age group, patients may have diaphragmatic pseudohypertrophy. Studies with a larger sample are necessary to better understand diaphragmatic dynamics in DMD patients.

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Evaluation of rAAVrh74 antibodies seroprevalence: report from the first tested cohort of Duchenne patients in Brazil

Felipe Franco da Graça[1], Clara Gontijo Camelo[2] , Mariana Rabelo de Brito[1], Marcondes Cavalcante França Jr.[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Felipe Franco da Graça; Email: felipefgraca@hotmail.com.

With the increasing utilization of viral vector-based gene therapies, knowing the seroprevalence of the virus- specific antibodies is crucial for estimating the pool of eligible patients within a population. Recombinant AAV serotype rh74 (rAAVrh74) is a vector used in Delandistrogene Moxeparvovec (DM), the first gene therapy approved for the treatment of Duchenne muscular dystrophy. Its seroprevalence in the north-american population is acknowledged to be low (13.9%). However, these data remain unknown for other demographic groups. In this study, we have assembled, for the first time, a cohort of tested Brazilian patients. Eight patients aged over 4 and under 6 years with a confirmed diagnosis of Duchenne muscular dystrophy were included. Each participant had a single blood sample collected, which was analyzed for total binding antibodies against rAAVrh74 using an enzyme-linked immunosorbent assay (ELISA). Antibody levels below 1:400 were classified as not elevated or seronegative. All eight evaluated patients (100%) exhibited negative titers for the antibody. This data, albeit derived from a limited sample size, corroborates the low prevalence observed in the north-american cohort. Nevertheless, larger studies are required to determine a more representative seroprevalence in the Brazilian population.

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Evaluation of sleep perception and presence of daytime sleepiness and its impact on the quality of life of patients with Fabry disease: a case series

Rafaella Resplande Xavier[1], Matheus Gabriel Castro Bezerra[1], Priscila de Paiva Nazareth[1], Sophia Oliva Schommer[1], Vitória Batista e Matricardi[1], Beatriz Fulador[1], Victor Hugo de Souza Silva Gomes[1], Heloise Helena Siqueira[1], Julia Mayumi Yakabe Nelson[1], Manuela Oliveira Alves Victor[1]

[1]Hospital Geral, Cuiabá MT, Brazil.

Correspondence: Beatriz Fulador; Email: beatriz_fulador@hotmail.com.

Background: Fabry disease is a genetic disorder caused by an inborn error of metabolism resulting from a mutation in the X chromosome. This mutation leads to a coding error of the enzyme alpha-galactosidase A, preventing its synthesis and thereby inhibiting the degradation of its specific substrate, globotriaosylceramide (Gb3). Consequently, there is an accumulation of Gb3 in the lysosomes of various organs, leading to a wide variability of symptoms due to its pro-inflammatory and cytotoxic effects, manifesting in nervous, integumentary, renal, and cardiovascular systems. Thus, the disease generates functional limitations both physically, such as pain, general health, and vitality, and socially and emotionally, significantly impacting patients' quality of life and sleep. **Objective:** To evaluate the general aspects of sleep and quality of life impairment in a series of patients with Fabry disease regularly undergoing enzyme replacement therapy. **Methods:** This is an observational, prospective study of a case series of patients with Fabry disease followed by the Neurology Service of HGU in Cuiabá/MT, evaluated between April and May 2024. Structured questionnaires with the following scales were used: Short-Form Health Survey 36 (SF36), Pittsburgh Sleep Quality Index (PSQI), and Epworth Sleepiness Scale (ESS), which were applied to all individuals in this case series. **Results:** Six individuals undergoing Enzyme Replacement Therapy (ERT) were included in this study, with a male-to-female ratio of 1:1 and an average age of 51.6 years (41-69 years). The PSQI evaluated sleep quality, analyzing 7 components with a

final score ranging from 0-21 points. It was concluded that 5 patients had good sleep quality and only 1 evidenced a sleep disorder. Furthermore, daytime sleepiness was assessed using the ESS, which considers the likelihood of the patient dozing or sleeping in different situations. According to the results obtained, no patient showed excessive sleepiness, with 4 exhibiting normal sleepiness and 2 showing moderate sleepiness. Regarding quality of life assessed by the SF-36 questionnaire, 8 domains were observed, each varying from 0-100%. The outcome identified that almost all categories are compromised, with the worst outcomes in domains involving emotional aspects (83.3%), social aspects (75%), functional capacity (74%), and limitations due to physical aspects (66%). **Conclusion:** After analyzing the results, it is concluded that the evaluations of the validated scales are essential in the follow-up of these patients and should be performed periodically, aiming to identify disabilities and limitations and thus propose the best pharmacological and non-pharmacological treatment for these patients, along with a multidisciplinary team, aiming for a better quality of life

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Evaluation of the functional capacity of patients with CADASIL in the state of Rio de Janeiro

Ana Luiza Sandoval Góes[1], Barbara T. Mothe[1,2], Ralph Lettieri[1], Cristiane Cardoso[1], Danielle Vianna[1], Diana Goes de Souza[1], Carolina P. Pandino[1,2], Ana C. Carvalho[1,2], Thayna Gomes[1,2], Vanderson Carvalho Neri[1,3,4]

[1]CADASIL Study Group, Brazil.
[2]Faculdade de Medicina de Campos, Campos dos Goytacazes RJ, Brazil.
[3]Hospital Federal da Lagoa, Departamento de Neurologia, Rio de Janeiro RJ, Brazil.
[4]Faculdade de Medicina de Campos, Departamento de Neurologia, Campos dos Goytacazes RJ, Brazil.

Correspondence: Ana Luiza Sandoval Goes; Email: analuizagoes@gmail.com.

Background: Autosomal dominant cerebral arteriopathy with subcortical infarctions and leukoencephalopathy (CADASIL) resulting from mutations in the NOTCH 3 gene on chromosome 19 is considered a rare condition and is the most prevalent hereditary small vessel vascular disease. Course with ischemic stroke, migraine and vascular dementia. It has a broad neuropsychiatric spectrum including mood changes, apathy and depression. The progression of the disease leads to the emergence of neurological sequelae and loss of functional capacity, physical and mental disability. **Objective:** This work aims to evaluate functional capacity, the occurrence of disability and cognitive decline in 28 patients from 11 families with CADASIL. **Methods:** Standardized functionality scales were applied, such as the modified Rankim scale (IRM), Barthel index (BI) and cognitive screening with the Montreal Cognitive Assessment (MoCA). The SPSS tool was used for statistical analysis of the results. **Results:** 28 cases from 11 families with lesions detected in the NOTCH3 gene were evaluated. 16 (57.1%) were women, 12 (42.9%) men; the average age was 46.36 years, the youngest age 22, the oldest age 67, the sample standard deviation was 12.53 years, coefficient of variation of 27.02%. The MoCA partnered 13 (46.4%) with cognitive impairment and 15 (53.6%) preserved cognitive impairment; MRI without symptoms in 19 (67.9%), without disability in 3 (10.7%), moderate disability in 5 (17.9%) and severe disability in 1 (3.6%); BI with a degree of severe functional dependence in 1 (3.6%), a degree of mild dependence in 8 (28.6%) and functional independence in 19 (67.9%). **Conclusion:** The application of the functionality and cognitive screening scales evaluated the sequelae of the disease, which were expressed in varying degrees of functional disability, physical disability and cognitive impairment. The sample had a predominance of women, the age variability between the second and sixth decade of life was compatible with literature data, with age being a homogeneous set. The IRM detected that the majority were able to carry out all their activities and take care of their own interests, showing no symptoms or moderate disability. The IB showed that the majority were functionally independent, although cases of severe disability for ADLs were identified. The high prevalence of cognitive impairment shows that neuropsychiatric symptoms can be present even in asymptomatic patients without functional disability. This study seeks to collect unprecedented epidemiological data on functionality in Brazil and be useful for adapting and collaborating in the development of health strategies.

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Evaluation of the hospitalization landscape for multiple sclerosis in children and adolescents aged 1 to 19 across Brazilian regions over the past 5 years

Marina Ottmann Boff[1], Ingrid Lizier Couto Pereira[1], Luísa Castro Gomes[1], Taís Michele Werle[1], Eduarda Gandolfi Horst[1], Gabriella Sityá Moojen da Silveira[1], Paola Saraiva Marinho[1], Marina Maestri Denardin[1], Frederico Orlando Friedrich[1], Magda Lahorgue Nunes[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Marina Ottmann Boff; Email: mottmannboff@gmail.com.

Background: Multiple sclerosis (MS) is an autoimmune condition affecting the myelin sheath of neurons, leading to degenerative inflammation. It's more common in women but can occur in both sexes. Typically diagnosed between ages 20 and 40, sometimes even in childhood. Its cause involves genetic predisposition and environmental factors. While there's no cure, current treatments aim to manage symptoms and slow disease progression, enhancing patients' quality of life. **Objective:** To evaluate the landscape of MS hospitalizations in children and adolescents under 1 to 19 years old in Brazil over the past 5 years. **Methods:** This is an ecological, cross-sectional, descriptive, and quantitative study using data from the Hospital Information System available on DATASUS. The period analyzed was from January 2019 to December 2023, considering the place of residence. The variables investigated included hospitalizations, average length of stay, deaths, mortality rate, and sex by region. The data were tabulated in Microsoft Excel and analyzed using descriptive statistics. **Results:** The total number of hospitalizations in children and adolescents was 1,589, with an average length of stay of 4.5 days, and there was only 1 death in all of Brazil (mortality rate of 0.06), which occurred in the 15 to 19 age group in the North region. There was 1 hospitalization in children under 1 year old, with an average stay of 8 days and no deaths; 4 hospitalizations among children aged 1 to 4 years, with an average stay of 10.8 days and no deaths; 45 hospitalizations among children aged 5 to 9 years, with an average stay of 8.6 days and no deaths; 289 hospitalizations among children aged 10 to 14 years, with an average stay of 6.9 days and no deaths; 1,250 hospitalizations among adolescents aged 15 to 19 years, with an average stay of 3.7 days and 1 death. Hospitalizations were more prevalent in females (62.2% of the total hospitalizations) across all regions and most age groups, except in the Central-West region for the 5 to 9 years (66.67%) and 10 to 14 years (70.0%) age groups, where hospitalizations were more frequent in males. The average length of stay varied among regions: 8.1 days in the North, 10.6 days in the Northeast, 4.5 days in the Central-West, 3 days in the Southeast, and 5.8 days in the South. **Conclusion:** the data reveal an increase in hospitalizations with age, peaking between 15 and 19 years. The average duration of hospitalizations decreases with age. Hospitalizations were more common among females in all regions, suggesting gender disparities in exposure or response to the disease. The average lengths of hospital stay varied among regions, possibly due to differences in healthcare systems and socioeconomic factors. The observed mortality rate in this study was low, n=1, corresponding to 0.06% of the total hospitalizations. These findings can guide prevention strategies and resource allocation for hospitalizations in children and adolescents.

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Evaluation of the prevalence of functional neurological symptom disorder in an outpatient epilepsy clinic of a tertiary hospital in Cuiabá and the importance of its recognition

Luis Felipe da Cruz Macedo[1], Tiago Ferreira Portela[2], Thyago dos Santos Amorim[2], Bruno Reginato Gumiero[1], Bianca Barbosa Araldi[1], Luisa Franciscatto[1], Luiz Henrique Granja de S. Vieira Miller[1], Marlon Figueiredo Alves Correa[1], Nathalia Carvalho Braga[1], Larissa Kozow Westin[1]

[1]Empresa Cuiabana de Saúde Pública, Cuiabá MT, Brazil.
[2]Universidade Federal do Mato Grosso, Cuiabá MT, Brazil.

Correspondence: Luis Felipe da Cruz Macedo; Email: luisfcmacedo@gmail.com.

Background: Functional neurological symptom disorders or psychogenic non-epileptic seizures (PNES) are paroxysmal events that resemble clinical episodes of epileptic seizures, due to motor, sensory or cognitive events that are not associated with electrical brain discharges or epileptiform activity in electroencephalographic recordings. There are no specific seizure presentation in PNES, requiring an experienced epilepsy specialist to suspect functional seizures. The definitive diagnosis is made using a video electroencephalogram (VEEG) and treatment is transdisciplinary, with psychological and psychiatric follow-up in collaboration with neurology. **Methods:** We carried out a cross-sectional study at an epilepsy treatment center (ETC) created in February 2024 at a tertiary hospital in Mato Grosso, Hospital Municipal de Cuiabá - HMC. We reviewed medical records from the beginning of the ETC's activity until June 24, 2024. The frequency of diagnoses, age, gender, use of antiepileptic drugs, multidisciplinary referral and VEEG (7-day protocol) were evaluated descriptively and the graphs were made using the R Studio program. **Results:** The HMC epilepsy treatment center (ETC) started on February 21, 2024 with 109 patients diagnosed with epilepsy and 14 with suspected PNES (12%), 7 men and 7 women (1:1). Under investigation from PNES included 6 patients (42%) with confirmed diagnoses with VEEG, 8 patients (57%) with suspicion still had no VEEG, and 3 patients (21%) were being evaluated previously for refractory epilepsy. Concerning antiepileptic drugs, 6 patients remained on the medication for the time being (42%) and 8 (57%) were completely withdrawn, and these same patients are undergoing multidisciplinary follow-up with psychiatry. Overall, 12 VEEGs have been completed at the CTE to date, with the PNES therefore representing 50% of the VEEGs completed in the 3 months since the ETC was set up. **Conclusion:** PNES are a major challenge for diagnosis and appropriate treatment, since their definitive diagnosis requires VEEG, a long-term exam that is difficult to access outside of large centers in order to conclude the diagnosis, withdrawal of antiepileptic medication and multidisciplinary follow-up, improving the quality of life of these patients. Likewise, PNES are great imitators of refractory epilepsies, and it is imperative to epilepsy treatment centers provides access to VEEG to confirm PNES as a important imitators of inadequate seizure control.

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Evaluation of the results of transcranial magnetic stimulation protocol in the rehabilitation of patients post-stroke

Camila Pezente Dal Toe[1], Tatiana Pizzolotto Bruch[1], Paula Abatti Moretto[1], Laura Fretta[1], Carlos Fernando dos Santos Moreira[1], Rafaela Silveira Passamani[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Camila Pezente Dal Toé; Email: camilapezente@hotmail.com.

Background: After conventional physiotherapy-based treatment, some sequelae of Stroke (AVE) persist. Transcranial Magnetic Stimulation (TMS), which modulates neuroplasticity, shows promise in restoring balance and motor function in upper and lower limbs. **Objective:** To evaluate the effectiveness of the TMS protocol in rehabilitating post-stroke patients in Criciúma, Santa Catarina. **Methods:** Eight electronic records were analyzed using Tinetti, Dash, and Berg functional scales to assess balance and motor function recovery. Scales were administered at the beginning and end of the TMS protocol, with scores analyzed statistically. **Results:** Results demonstrated significant improvements in balance rehabilitation as measured by the Tinetti scale (mean 7.80±5.97 before and 18.60±7.30 after the protocol, p=0.002) and Berg scale (from 43.00 to 54.00). Gait, also assessed by the Tinetti scale, showed increased scores (7.80±5.97 to 18.60±7.30, p=0.002). Regarding upper limbs, the DASH scale revealed a reduction in motor deficit (89.33±22.50 to 68.33±30.02, p=0.181) following TMS. **Conclusion:** These findings suggest that Transcranial Magnetic Stimulation may be an effective approach in rehabilitating post-stroke patients, particularly in improving balance and motor function.

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Evaluation of the select score as a predictor of risk for epilepsy development in patients with ischemic stroke

Darah Fontes da Silva Assunção[1], Marcelle Sanjuan Ganem Prado[2], Gabriel Chung[2], Filipe Brito Ferraz da Silveira[2], Marina Buldrini Filogonio Seraidarian[1], Vanessa Cristina Colares Lessa[2], Fábio Barreto Cabral[2], Cristine Mella Cukiert[2]

[1]Universidade de São Paulo, São Paulo SP, Brazil. [2]Santa Casa de Misericórdia de São Pauilo, São Paulo SP, Brazil.

Correspondence: Marcelle Sanjuan Ganem Prado; Email: cellesanjuan@yahoo.com.br.

Background: In patients with ischemic stroke, the risk of recurrence after a first late-onset seizure is estimated at seventy per cent when diagnosis can be established. Several factors may predict the risk of epilepsy development, including stroke severity, etiology, affected arterial territory, presence of cortical involvement, and occurrence of acute symptomatic seizures, variables assessed by the SeLECT score. **Objective:** To analyze whether the variables selected in the SeLECT score were predictive of risk in the studied population. **Methods:** Retrospective observational study, reviewing medical records of patients followed in a cerebrovascular disease outpatient clinic over a 2-year period, aged 18 years and older, diagnosed with epilepsy and ischemic stroke. **Results:** Thirty-seven patients were evaluated, with a mean age of 50.32 years. The NIHSS frequency was 62.2% for moderate (4-10). Regarding etiology classified by TOAST criteria, 16.2% were attributed to large artery atherosclerosis and 18.9% to cardioembolism. Cortical involvement was observed in 81.1% of patients. Most cases, 83.8%, involved the ACM territory. The occurrence of acute symptomatic seizures, a variable with higher scores in the SeLECT score, was observed in 13.5% of the population. Among the patients, 16.2% underwent acute reperfusion therapy. No statistical significance was found when comparing the total SeLECT score and TOAST classification. However, significant association (p<0.05) was demonstrated when analyzing higher SeLECT scores with increased risk of epilepsy development, higher NIHSS scores, presence of cortical involvement, ACM territory affected, and occurrence of acute symptomatic seizures. Age at stroke onset, patient sex, reperfusion therapy, occurrence of hemorrhagic transformation, and assessed comorbidities showed no significant association with SeLECT scores and likely did not increase seizure occurrence risk. **Conclusion:** The SeLECT score showed applicability in predicting late seizures after stroke. No other predictors among the analyzed variables were observed. The importance of acute reperfusion therapy as a seizure-inducing factor remains uncertain in this scenario. Clearer insights may be gained from prospective multicenter studies, guiding follow-up of high-risk patients.

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Evidence of cortical thinning in patients with hereditary transthyretin amyloidosis

Iara Senem[1], Maria Paula Foss[1], Carolina Lavigne-Moreira[1], Antonio Carlos dos Santos[1], Pedro J Tomaselli[1], Jan Axelsson[2], Jonas Wixner[2], Renan Flávio de França Nunes[3], Marcondes Cavalcante França Júnior[3], Wilson Marques Jr.[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil. [2]Umeå University, Umea, Swenden. [3]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Iara Senem; Email: iara.senem@usp.br.

Background: Hereditary Transthyretin Amyloidosis (ATTRv) is a systemic disease that primarily affects the peripheral and autonomic nervous systems. Recently, the Central Nervous System has been investigated. Besides cognitive dysfunction, amyloid deposition was identified in the brain, cerebellum, leptomeningeal and subarachnoid meningeal vessels, and the brainstem. In vivo brain MRI studies showed diverse results, such as leptomeningeal involvement, microbleedings, cortical atrophy, and, in some cases, no abnormalities were observed. To date, studies have not investigated cortical and subcortical areas using software for automatic brain segmentation. **Objective:** We aimed to investigate the brain volume, and cortical thickness in patients with ATTRv amyloidosis. **Methods:** 3DT1-weighted images (T1) of brain MRI of twenty ATTRv patients (mean age=41.7±9.2) and 23 sex/age-matched healthy individuals (41.7±9.3) were analyzed. Cortical measures of volume and thickness were obtained for both groups through FreeSurfer software ("crecon-all" script). SPSS software was used for the statistical analyses. **Results:** Compared to controls, ATTRv patients displayed smaller volumes of cerebellum. No difference between the groups was found in the hypothalamus volume. The study indicated a thinner cortical thickness in ATTRv amyloidosis in the following regions: banks of the superior temporal sulcus, fusiform gyrus, inferior, superior, and middle temporal gyri, orbitofrontal cortex, pars opercularis, pars orbitalis, lateral orbitofrontal cortex, precentral gyrus, inferior parietal cortex, supramarginal and postcentral gyri, occipital lateral cortex, pericalcarine cortex, cuneus, and insula. Disease duration was negatively correlated with cortical thickness of insula, pars opercularis and inferior and superior temporal gyri. No correlation was found between disease duration and the cerebellum volume. **Conclusion:** The study showed a thinner cortical thickness in specific brain regions in ATTRv amyloidosis. The reasons for this finding and its relationship with amyloid deposition are not fully understood. Further research involving a larger number of patients is necessary to confirm these findings and identify their clinical manifestations.

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Evolutionary profile of the Walking test and Rotterdam scale in a group of patients with late-onset Pompe disease undergoing enzyme replacement therapy

Beatriz Fulador[1], Rafaella Resplande Xavier[1], Albérico Rocha Lima Neto[1], Rhaymysom Jasmy Gomes Abreu[1], Helen Luiza Silvestrini[1], Carine Calegaro[1], Letícia Lage Silva[1], Hingrid Cristine Arruda de Oliveira[1], Victor Hugo de Souza Silva Gomes[1], Heloise Helena Siqueira[1]

[1]Hospital Geral, Cuiabá MT, Brazil.

Correspondence: Beatriz Fulador; Email: beatriz_fulador@hotmail.com.

Background: Pompe disease (PD), also known as Glycogen Storage Disease II, is a genetic disorder caused by pathogenic variants in the gene encoding acid alpha-glucosidase (GAA), resulting in GAA deficiency and consequent glycogen accumulation in the lysosomes of tissues. Due to muscle involvement, patients experience proximal muscle weakness, often progressive, which is directly related to their motor functionality. Motor impairment symptoms are usually the first manifestation of the disease. Several tests and scales are proposed for its evaluation, notably the Rotterdam Handicap Scale (RHS) and the 25-Foot Walk Test (T25-FW). According to major studies, the motor functionality of patients diagnosed with PD has a median 25% lower than that of healthy adults based on the Rotterdam scale analysis. In addiction, the motor system responds significantly better than the respiratory system to enzyme replacement therapy (ERT). **Objective:** To comparatively evaluate the results of motor functionality assessment tests in a group of patients with PD who are regularly using enzyme replacement therapy. **Methods:** This is a comparative study conducted through the application of validated scales and tests to assess the motor function of patients with Pompe Disease: the Rotterdam Handicap Scale (RHS) and the 25-Foot Walk Test (T25-FW), in the years 2017 and 2024, in a group of five patients regularly using enzyme replacement therapy (alpha-glucosidase) at a rare diseases center. **Results:** Five individuals were included in the study, of which 3 were men and 2 were women, with ages ranging between 35 and 66. All have the late-onset form of the disease. In terms of motor functionality assessment, the average RHS score increased from 27.2 in 2017 to 27.6 in 2024, with 9 being the minimum score and 36 the maximum score for the ability to perform activities. Similarly, the average time for the T25-FW decreased from 15 seconds (ranging from 8 to 20) in 2017 to 11 seconds (ranging from 5 to 18) in 2024. Of the 5 patients, only one showed an increased time to complete the test over the 7 years, from 8 to 18 seconds, representing a 125% increase. However, it is noteworthy that this patient suffered a stroke between assessments, resulting in significant gait impairment. The other patients had no additional comorbidities. **Conclusion:** Enzyme replacement therapy (alpha-glucosidase) proved effective and had a positive impact on the motor functionality of the patients in this study, although one patient showed less improvement due to the presence of another pathology. Therefore, it is concluded that the routine and periodic use of validated assessment scales is essential for monitoring the limitations and disabilities of these patients, in order to evaluate the need for therapeutic changes, the association of adjunct therapies such as motor physiotherapy, and for screening for potential comorbidities that may cause unexpected and sudden clinical deterioration.

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Evolutive course of recovery of transcortical motor aphasia following focal resections in the frontal lobe for refractory epilepsy

Eliseu Paglioli[1], Rafael Paglioli[1], William Martins[1], Fernanda Schuh Martins[1], Vicenzo Zarpellon[1], Thomas Frigeri[1], Victoria Schvartzman[1], Natalie Donida[1], Bibiana Thome[1], André Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Andre Luis Fernandes Palmini; Email: alpalmini@gmail.com.

Background: Rationale and objective: Since the description by Norman Geschwind of the disconnection syndromes, neurologists began to understand deficits in the so-called higher cortical functions as also resulting from subcortical lesions in white matter tracts connecting specific cortical regions. Perhaps few neurological syndromes illustrate this concept so well as the aphasias. From a simplistic view of "Broca's versus Wernicke's" neurology has evolved to the description of several other types of aphasia, resulting from the disconnection between these and other cortical regions. When aphasias result from stroke, lesions in the affected territories often involve a number of cortical and subcortical structures that compound a complex clinical picture, with variable perspectives of functional recovery. On the other hand, in well delineated, limited resections performed to resect epileptic foci it is possible to analyze both symptomatology and evolution in "pure" syndromes. **Objective:** Here, we present the clinical picture and the evolution toward complete recovery over days and weeks of two patients with focal cortical dysplasia (FCD) in the middle frontal gyrus of the dominant (left) frontal lobe (dMFG) who had post-operative transcortical motor aphasia. **Methods:** Two women (#1: 17yo; #2: 40 yo) with refractory seizures due to FCD in the dMFG underwent resections of the epileptogenic zone following presurgical evaluation. They had normal neurological examination preoperatively and objective motor and language evaluations were performed daily in the first week after surgery, documented in video. Following discharge, evaluations were weekly, through simple tests applied and documented by relatives. Post-op MRIs were obtained in both. **Results:** Both became seizure free, except for brief seizures in patient #1 in the immediate post-op period. In the first few days, neither patient could start spontaneous speech, although comprehension and repetition were intact. Answers to simple questions were only possible, albeit incompletely, following induction by the examiner, ie, options were given by the examiner and selected by the patient. Similar difficulties with writing (only after priming) were also observed in patient #1. Post-op MRIs showed resection of the dMFG and discrete edema of the underlying fibers. Following 1 week, both could hesitantly already begin sentences and produce spontaneous answers. Complete recovery was seen between 30 and 60 days after operation and both are now fully functional. **Conclusion:** Transcortical motor aphasia is a disconnection syndrome due to involvement of fibers connecting the supplementary motor and Broca's areas. Resections of the dMFG can lead to transcortical motor aphasia, yet with excellent functional recovery, providing resection carefully respects the underlying white matter.

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Excessive daytime sleepiness in Parkinson's disease: an observational analysis

Luma Santos Arruda[1], Diego Monteiro de Carvalho[2], Marcello Facundo do Valle Filho[2], Marcus Vinicius Della Coletta[3], Alvaro Siqueira da Silva[3], Raissa Costa Said[4], Débora de Oliveira Ferreira[1], João Victor da Costa Nunes[4], Anelisa Campana Itinose[4], Carlos Mauricio Oliveira de Almeida[3]

[1]Centro Universitário Fametro, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Fundação Hospital Adriano Jorge, Manaus AM, Brazil.
[4]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Luma Santos Arruda; Email: lumaarruda10@hotmail.com.

Background: Parkinson's disease (PD) is the second most common neurodegenerative disease in the elderly, second only to Alzheimer's disease, representing about 1% of the population over 60 years old. In PD sleep disorders are observed in up to 90% of patients. Literature data demonstrate that Excessive Daytime Sleepiness (EDS) occurs in 50 to 80% of PD patients and has multifactorial causes, such as medications, sleep disorders, psychiatric disorders, and the neurodegenerative process itself. **Objective:** To analyse the association between the Epworth Sleepiness Scale (ESS) in patients with PD at early/intermediate stages according to the Hoehn-Yahr (H&Y) scale. **Methods:** This is a cross-sectional observational study, with a sample composed of PD patients from the neurology clinic at the reference hospital in Manaus-AM. The study was approved by the local Ethics Committee under Certificate of Presentation of Ethical Consideration No. 58684622.2.0000.0007. Inclusion criteria were: PD (aged over 18) diagnosed according to the London Brain Bank criteria, with staging 0-3 on the H&Y scale without a diagnosis of obstructive sleep apnea (OSA). Patients were excluded if they had a diagnosis of dementia, OSA (AHI >15/h by polysomnography), atypical parkinsonism, neoplastic disease, systemic diseases, alcohol abuse or use of illicit drugs, or language or speech articulation disorders that prevented the application of the questionnaires. After acceptance into the study, patients were subjected to the application of clinical questionnaires, ESS and the disease staging (H&Y). EDS was defined by a score greater than 10 points on the ESS. **Results:** Thirty-three patients were evaluated, with 75% presenting with EDS. No significant association was observed between those who presented with or without EDS and the stage of the disease (H&Y) (p=0.94), nor with anxiety ($\chi2$=4.8; p=0.11) or depression ($\chi2$=3.29; p=0.19). **Conclusion:** Our study confirms the literature data that PD patients frequently present with EDS (50-70%). We did not observe an association between this sleep disorder and mood disorders or the stage of the disease. As we know, the cause of EDS is multifactorial and sometimes involves the neurodegenerative process affecting the regulatory centers of wakefulness and sleep.

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Excruciating pain during exercise reveals colloid cyst: a case of potential life-threatening risk

Vanessa Gil Humberto dos Santos[1], Amanda Melo Leite Leão[1], Laís Borneo Moreira[1], Thais Pereira Albernaz de Souza[1], Jorge Luiz Alves[1], Pollyana Marcela Silva de Farias[1], Lais Felix Teixeira da Cunha[1], Laís Borneo Silva de Farias[1]

[1]Universidade Unigranrio Afya, Duque de Caxias RJ, Brazil.

Correspondence: Vanessa Gil Humberto dos Santos; Email: vanessaterapia@hotmail.com.

Case presentation: A 42-year-old Italian man, a regular practitioner of physical activities, experienced sudden and intense head pain during a weightlifting session while performing a high-load isometric exercise on the leg press machine. The pain, described as excruciating headache, occurred without associated autonomic signs such as nausea, vomiting, or visual changes. Due to the persistence of the pain, he sought medical attention a week after the episode. Cranial magnetic resonance imaging (MRI) revealed the presence of a colloid cyst in the third ventricle. A more conservative approach was chosen, with regular monitoring and analgesic measures for pain relief. **Discussion:** Effort-induced headache is a classic symptom in patients with colloid cysts, occurring due to increased intracranial pressure during activities that elevate intrathoracic pressure and, consequently, intracranial pressure. High-intensity isometric exercises, such as the leg press, can exacerbate this condition, leading to episodes of intense pain. Additionally, the headache can be positional, worsening when the patient is lying down or inclined.The diagnosis of a colloid cyst is typically confirmed through MRI. In this patient's case, the MRI revealed a colloid cyst in the third ventricle, corroborating the etiology of the headache.Colloid cysts pose a risk of growth, especially in young men, and can obstruct the foramen of Monro, resulting in hydrocephalus and increased intracranial pressure, which can lead to sudden death, if not properly treated, causing na emergency situation that requires immediate intervention. The management of colloid cysts can range from conservative to surgical, depending on symptoms and the risk of complications. In the presented case, a conservative approach was chosen. **Final comments:** This case illustrates the importance of considering colloid cysts as a potential cause of sudden and intense headache in patients undergoing intense physical exercise. Although rare, colloid cysts can present significant clinical symptoms that impact the patient's quality of life. Early diagnosis and appropriate management are crucial to avoid serious complications, including hydrocephalus and the risk of sudden death. Conservative management can be a viable option for asymptomatic or mildly symptomatic patients, while surgical intervention is reserved for cases with severe symptoms or risk of complications. This case report contributes to the clinical understanding of colloid cysts and highlights the need for a comprehensive diagnostic approach in patients with sudden headache following physical exercises. Awareness of this condition can lead to quicker diagnoses and more effective treatments, improving clinical outcomes for patients.

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Experience in the switch from alglucosidase to avalglucosidase in real-life late-onset Pompe disease treatment

André Macedo Serafim Silva[1], Michele Abdo Paiva[1], Clara Gontijo Camelo[1], Ana Lucila Moreira[1], Edmar Zanoteli [1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Andre Macedo Serafim da Silva; Email: macedosera@gmail.com.

Background: Late-Onset Pompe disease (LOPD) is an inherited condition caused by glucogen accumulation due to deficiency of acid alfa- glucosidase and consequent muscle weakness and respiratory insufficiency. Enzyme replacement therapy (ERT) with alglucosidase alfa has been considered useful to treat LOPD in clinical trials, case series and real-life studies. Recently, a new ERT with avalglucosidase has been demonstrated as non-inferior to alglucosidase, with the promise of better outcomes in the clearance of glycogen in the muscle. **Objective:** To describe the real-life experience in the switch from alglucosidase to avalglucosidase in LOPD patients treated with ERT. **Methods**: We retrospectively reviewed all medical records from LOPD patients followed in a tertiary neuromuscular center from Jan 2019 to May 2024. Patients who changed the ERT therapy were analyzed according to timed tests, forced vital capacity (FVC) before and after the ERT switch. **Results:** Fourteen LOPD patients with regular treatment with ERT were identified from the neuromuscular clinic records. Two of them switched from

alglucosidase to avalglucosidase. One male patient, diagnosed at 41 years of age presented progressive worsening of respiratory symptoms and FVC, needing continuous non-invasive ventilation, despite alglucosidase treatment for four consecutive years. After two years of changing the treatment to avalglucosidase his FVC increased from 64% to 75% and 10m walk test improved from 25 seconds to 15 seconds. The other male patient, diagnosed at 46 years of age were using doble dosis of alglucosidase (20mg/kg weakly) over a period of two years, with stable respiratory and motor condition. After six months of switch to avalglucosidase, his FVC improved from 52% to 55% and his 6-minute walk-test increased from 390m to 460m. **Conclusion:** Two out 14 LOPD patients in our center switched ERT from alglucosidase to avalglucosidase, and this Brazilian single-center experience shows a trend of better motor e respiratory outcomes with the new generation ERT, like other real-life reported studies. Further studies are still needed to demonstrated the superiority of avalglucosidase over alglucosidade in treating Pompe disease.

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Experience with the use of cladribine in patients with multiple sclerosis at the neuroimmunology outpatient clinic of Faculdade de Medicina ABC

Victor Ezaki Alves Silva[1], Ana Claudia Piccolo[1], Eduardo Macedo de Souza Tieppo[1], Renato Morais Bueno[1], Rayanne Pereira Mendes[1], Guilherme Bitencourt Barbosa[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Victor Ezaki Alves Silva; Email: ezaki_victor@hotmail.com.

Background: Multiple sclerosis (MS) is a chronic autoimmune disease affecting the central nervous system (CNS), characterized by inflammation and neurodegeneration. Diagnosis relies on clinical examination, laboratory tests and radiological findings. Long-term treatment involves disease-modifying therapies (DMTs), among which Cladribine is an option. It is an oral administered medication with moderate to high efficacy, exerting cytotoxic action on B and T lymphocytes by inhibiting DNA synthesis. Food and Drug Administration (FDA) in United States approved Cladribine in 2019. In 2023 it was incorporated by Brazil SUS, directed for patients who are unresponsive to other DMTs. **Objective:** To report the experience made at a Neuroimmunology reference center in Santo André - SP, Brazil, with the usage of Cladribine regarding the clinical and radiological efficacy in MS patients. **Methods:** It is a retrospective observational study, including patients with regular follow up at our NeuroImmunology outpatient clinic of ABC Medical School, from August 2020 to December 2023. Patients that were using Cladribine during this period were included, while patients not treated with Cladribine were excluded. Seven participants were included, and each one received Cladribine at a dose of 1.75mg/kg per year for two years. Epidemiological data, clinical history, neurological examination (including Expanded Disability Status Scale - EDSS), and contrast-enhanced magnetic resonance imaging (MRI) of the brain and cervical and thoracic spine were analyzed. Assessments were made before treatment initiation until two years post-second dose of Cladribine. Therapeutic failure was considered a relapse of the disease and/or radiological activity, such as lesions with contrast enhancement or an increase in the number or volume of lesions on MRI. Brain atrophy on MRI was not evaluated. **Results:** Results showed therapeutic failure in 3 patients (42.8%) and effectiveness in 4 patients (57.2%). All patients had highly active disease, however the 3 patients with therapeutic failure using Cladribine had previous unsuccessful experiences with multiple DMTs and aggressive MS disease. Treatment of MS offers diverse therapeutic strategies for disease phenotypes and stages. Cladribine, has already been proven as an effective option for the treatment of MS and is an avaluable DMT for this therapeutic spectrum. Its inclusion in SUS provides an option for the management of patients with MS, like those who have not previously used DMTs, as those have experienced relapses despite prior DMT use. Cladribine may present therapeutic failure in patients who already present an aggressive, highly active disease, with a high number of lesions on MRI and functional incapacity

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Experience with use of rituximab in a patient with Susac syndrome: a case report

Allefy Beltrão Albano[1], Ana Rosa Santana[2], Eduardo Sousa de Melo[1], Paulo Felipe Bezerra Silva dos Santos[3], Evelliny Gomes da Silva[1], Fabrício Martins Veras[1], João Pedro Matos de Santana[1], Letícia Klabinske Marques Monteiro[1], Amanda Sampaio de Sá[3]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[3]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Allefy Beltrão Albano; Email: allefymed@gmail.com.

Case presentation: A 34-year-old male patient presented to our service in 2021 for investigation of a condition characterized by progressive crural spastic paraparesis with evolution since 2019, low visual acuity of recent onset, and subacute cognitive-behavioral condition. She had been diagnosed with Multiple Sclerosis in another service, but no follow-up. On physical examination, he presented spastic paraparesis and global hyperreflexia. Sensitivity preserved. On examination of cranial nerves, fundoscopy did not show visible alterations, Rinne's test denoting decreased auditory and bone conductions, with the former prevailing. Magnetic resonance imaging of the brain and spinal cord showed central injuries in the corpus callosum ("snowballs"), with preservation of the most peripheral fibers, which were better evidenced in T2/FLAIR sagittal sequences. Lumbar puncture showed an increase protein (86mg/dL) and white cells(9 with 88% of neutrophils). He also performed ophthalmological investigation with Optical Coherence Tomography, which showed previous retinal ischemic alterations without major clinical repercussions, and audiometry showing bilateral sensorineural loss for high frequencies (6000-8000 Hz). In view of the main diagnostic hypothesis being Susac syndrome, the patient received pulse therapy with methylprednisolone for five days followed by immunosuppressive therapy with oral azathioprine, showing no clinical response over the following months, being hospitalized again in 2023 due to the progression of weakness and the appearance of urinary and fecal incontinence. In this context, the patient received a new pulse therapy and started modifying therapy with Rituximab, with semiannual doses of 1g. Since then, he has been showing progressive recovery of functionality, with a decrease in spasticity and pattern of weakness in the lower limbs, in addition to progressive sphincter control, denoting improvement in quality of life. **Discussion:** Susac syndrome is a rare, fluctuating and progressive disease caused by autoimmune-mediated occlusion of microvessels in the brain, retina and inner ear which are responsible for the clinical triad characterized by encephalopathy, visual disturbances caused by retinal artery ischemia and sensorineural hearing loss. Its treatment consists of initial administration of pulse therapy with methylprednisonolonone and maintenance therapy with disease modifying immunosuppressive drugs, as Rituximab, an anti-CD20 monoclonal antibody, being a first- line option for this pahtology which is still rarely mentioned in the literature. **Final comments:** The patient in this case presents clinical and radiological criteria for the diagnosis of Susac syndrome. The use of Rituximab associated with progressive clinical improvement, without progression of deficits and apparent improvement of existing ones evidenced in the last outpatient visits, make this drug a good option for the disease-modifying treatment of the syndrome.

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Exploratory study of cognitive dysfunction and brain mri correlates in hereditary transthyretin amyloidosis

Iara Senem[1], Maria Paula Foss[1], Carolina Lavigne-Moreira[1], Antonio Carlos dos Santos[1], Pedro J. Tomaselli[1], Jan Axelsson[2], Jonas Wixner[2], Renan Flávio de França Nunes[3], Marcondes Cavalcante França Júnior[3], Wilson Marques Jr.[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Umeå University, Umea, Sweden.
[3]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Iara Senem; Email: iara.senem@usp.br.

Background: Hereditary Transthyretin Amyloidosis (ATTRv) is a systemic disease that mainly impacts the heart and the peripheral nervous systems. The most common mutations (ATTRV30M (p.V50M) and ATTRV122I (p.V142I)) result in two main phenotypes: polyneuropathy and cardiomyopathy, respectively. Recently, it was recognized the involvement of the central nervous system in ATTRV30M amyloidosis. The amyloid deposition was identified in the leptomeningeal and subarachnoid meningeal vessels, brainstem, and cerebellum. The cognition also was more extensively studied in ATTRV30M. The cognitive impairment was described in liver transplanted patients, in patients with long-standing disease, and in carriers of ATTRV30M mutation in the asymptomatic phase. Despite the description of cognitive dysfunction in ATTRv amyloidosis, there is no previous study associating cognition and brain MRI findings in this population. **Objective:** To examine brain characteristics (volume and cortical thickness) and their association with neuropsychological measures in ATTRv patients. **Methods:** An observational study of a sample of 21 patients (mean age: 42.5±9.7 years) with genetic confirmation of TTR mutation was conducted. All patients were submitted to an extensive neuropsychological assessment and brain MRI. T1-weighted images were automatically segmented in the FreeSurfer software. Patients were sorted into two groups according to cognitive dysfunction: (1) ATTRv normal cognition and (2) ATTRv cognitive dysfunction. **Results:** Fourteen ATTRV30M (p.V50M) and seven ATTRV122I (p.V142I) patients participated in the study. Thirteen ATTRv patients (43.6±8.9 years) presented neurocognitive dysfunction. Neuropsychological evaluation showed memory dysfunction in ten patients. Seven patients presented dysfunction in visuospatial ability. Executive functions were impaired in four patients. Cognitively dysfunctional ATTRv patients presented lower cortical thickness compared with cognitively normal patients in the following regions: lingual gyrus, left pericalcarine cortex, right precentral gyrus, right supramarginal, and right transverse temporal. The lingual gyrus was strongly correlated with memory (RAVLT, delayed recall) and executive function task (TMT B). Memory was also correlated with the left pericalcarine and right precentral gyrus. The right supramarginal gyrus correlated with attention (TMT A). **Conclusion:** This study suggests that cortical thickness can be associated with cognitive dysfunction in ATTRv amyloidosis. Cortical regions such as the lingual gyrus, right precentral gyrus, and left pericalcarine cortex were correlated with cognition in individuals with cognitively dysfunctional ATTRv amyloidosis.

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Exploring differentiation of common demyelinating CMT subtypes: a pilot study using supervised machine learning

Thales Pardini Fagundes[1], Manoella Guerra de Albuquerque Bueno[2], Rodrigo Siqueira Soares Frezatti[2], Gustavo Maximiano Alves[2], Lucas Gondim Briand Vieira[2], Claudia Ferreira da Rosa Sobreira[2], Wilson Marques Junior[2], Pedro José Tomaselli[2]

[1]Hospital de Amor, Barretos SP, Brazil. [2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Lucas Gondim Briand Vieira; Email: lucasbriand@hotmail.com.

Background: Charcot-Marie-Tooth (CMT) disease and related neuropathies pose diagnostic challenges due to genetic heterogeneity and overlapping clinical and electrophysiological features. **Objective:** We used a machine learning model, RandomForestClassifier, to classify CMT subtypes based on clinical and electrophysiological data. **Methods:** Data were obtained from 54 patients with genetic confirmed CMT, including CMT1A, CMT1B, males with CMTX1 and CMT4C subtypes. The dataset included: sex, age of onset, and electrophysiological measurements. Missing values were imputed with the median of the corresponding class. The target variable, diagnosis_class, was mapped to four categories: CMT1A, MPZ, GJB1, and SH3TC2. The dataset was split into training (80%) and test (20%) sets, stratified by diagnosis_class. SMOTE was used to address class imbalance. A RandomForestClassifier was trained on the resampled training set. Model performance was evaluated using accuracy, precision, recall, F1-score, and ROC-AUC. Optimizations included setting the number of trees to 75, maximum depth to 10, minimum samples to split to 5, and minimum samples at a leaf to 2, aiming to enhance performance and reduce overfitting. **Results:** The optimized model achieved an accuracy of 95% on the test set. Precision, recall, and F1- scores were: CMT1A (precision 1.00, recall 0.80, F1-score 0.89), MPZ (1.00), GJB1 (1.00), and SH3TC2 (precision 0.83, recall 1.00, F1-score 0.91). The confusion matrix showed minimal misclassification, primarily in CMT1A. Feature importance analysis revealed that ulnar conduction velocity, age of onset, and median nerve latency were the most significant predictors. **Conclusion:** The optimized RandomForestClassifier demonstrates high accuracy in classifying demyelinating CMT subtypes, with excellent performance for MPZ and GJB1. The model shows limitations in distinguishing between CMT1A and SH3TC2 subtypes, suggesting further refinement with additional data and feature engineering. This study highlights the potential of machine learning models in aiding the diagnosis of genetically and clinically heterogeneous neuropathies.

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Exploring how cognitive components influence social cognition in patients with dementia

Lucas de Andrade Saraiva[1], Dreicy Helen de Souza Rodrigues[1], Ana Laura Daldegan[1], Viviane Amaral Carvalho[1], Larissa de Souza Salvador[1], Luciano Inácio Mariano[1], Vinícius Tinarelli Silva[1], Paulo Caramelli[1], Elisa de Paula França Resende[1], Leonardo Cruz de Souza[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Lucas de Andrade Saraiva; Email: lucasdeandradesaraiva@gmail.com.

Background: Social cognition involves a complex set of mental abilities related to perceiving, processing, interpreting, and responding to social cues. These skills enable people to behave appropriately in various social settings. Social cognition includes essential skills such as recognising facial expressions, understanding others' perspectives (theory of mind), social reasoning, moral judgment, and decision-making. **Objective:** This study aims to investigate how different elements of cognition may influence the social cognition of patients with dementia, specifically those with AD and FTD. **Methods:** We included two groups of participants: 1) FTD (n=19, 11M/8F; mean age=63.7±9.8 years; mean schooling=11.7±5.4; Disease Duration:4.1±2.4; MMSE=23.0±6.2); and 2) AD (n=25; 9M/16F; mean age=69.8±10.4 years; mean schooling=9.2±5.6; Disease Duration:6.31±5.1; MMSE=22.1±4.0). All participants underwent a comprehensive cognitive assessment using the following tests: Craft Story 21, The Beson Complex Figure Test, Digit Span test, Semantic Verbal Fluency, Trail Making Test, The Multilingual Naming Test (MINT) and Phonemic Verbal Fluency (P and M). The neuropsychiatric symptoms were evaluated using the Neuropsychiatric Inventory (NPI). Social cognition was assessed with Mini-Sea (Faux-Pas and Ekman test). **Results:** Patients with FTD performed statistically better than the AD group in cognitive tests assessing immediate memory, visuoconstructive function, processing speed, and executive functions. When assessing the cognition elements that could affect the social cognition of patients with dementia, it was found that both patients with FTD and those with AD showed a positive correlation between social cognition and various cognitive functions. These included working memory and attention (Digit Span test), semantic memory (MINT test), and language skills (animal category fluency and phonemic verbal fluency). Additionally, both groups of dementia patients displayed a negative relationship between social cognition and the time it took to complete the Trail Making Test, which is linked to processing speed and executive functions. On the other hand, when evaluating the differences between both groups, it was possible to identify that the social cognition of patients with AD, unlike patients with FTD, showed a positive correlation with visuoconstructive functions (Beson Complex Figure) and a stronger correlation with language (not only animal category but also vegetable category fluency). Additionally, it is worth noting the negative correlation between the neuropsychiatric symptom agitation (NPI) and social cognition in the AD group. **Conclusion:** The study highlights the complex relationship between cognitive functions and social cognition in patients with dementia, emphasising the need to consider specific cognitive profiles and neuropsychiatric symptoms when assessing social cognition for tailored interventions and support strategies.

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Exploring ITPR3 mutations in Charcot-Marie-Tooth neuropathy: a case report from Brazil

Gabriel Thomaz Candido[1], Carolina Braga Moura[1], Leonardo Sena de Souza[1], Ana Luiza Morgado[1], Maria Clara de Oliveira Lemes[1], Helena do Amaral Barreto[1], Camila Castelo Branco Pupe[1], Osvaldo Jose Moreira do Nascimento[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Gabriel Thomaz Candido; Email: gabriel_candido@id.uff.br.

Case presentation: A 25-year-old patient presents with a history of scoliosis diagnosed at 19 and torsion of the thoracic vertebra following a car accident at 25. Since then, he has experienced chronic lumbar pain radiating to the left leg, worsening during the pandemic. Currently, he reports acute pain on the left side and had difficulty walking due to lower limb weakness. Medications for pain management include tramadol, Amytril, Pregabalin, Reconter, Restiva, and Frontal. Physical examination reveals symmetrical muscle strength, reduced sensitivity in distal limbs, and normal coordination. According to the MRC scale, he presents grade 4 for wrist extension, finger extension, finger abduction, thumb opposition, and hip flexion; grade 3 for knee flexion and knee extension; grade 2 for ankle dorsiflexion. Bilateral peroneal atrophy, flat feet, and hammer toes are observed. Hyporeflexia in biceps, triceps, and brachioradialis tendons, and areflexia in patellar and Achilles tendons. Reduced sensitivity to pain, temperature, touch, and vibration in the distal portions of both upper and lower limbs. Nerve conduction study shows sensory-motor axonal and distal predominant polyneuropathy affecting all four limbs with predominance of the lower limbs. There is a mild reduction in conduction velocities of the median, ulnar, tibial, and sural nerves, with reduction in the amplitudes of the sural nerve potentials. Study of F waves shows borderline values of minimum latency of the median and tibial nerves. Electromyography reveals increased motor unit amplitudes and neurogenic recruitment in the distal muscles of the lower limbs. Genetic testing reveals a variant of uncertain significance (VUS) was found in heterozygosity in the ITPR3 gene, revealing a rare mutation. **Discussion:** The pathogenesis of ITPR3 mutations involves dysregulation of Ca2+ signaling pathways, disrupting mitochondrial function, ER-mitochondrial Ca2+ transfer, and cellular homeostasis. These molecular mechanisms contribute to the neuropathic phenotype observed in CMT patients with ITPR3 mutations. This case highlights the significance of genetic testing in diagnosing rare neuropathic conditions and underscores the emerging role of ITPR3 in CMT neuropathy. Understanding the molecular mechanisms underlying CMT pathogenesis may pave the way for novel therapeutic interventions targeting Ca2+ flux dysregulation. Further research is warranted to elucidate the functional consequences of ITPR3 variants and explore potential therapeutic targets for CMT. **Final comments:** Although the ITPR3 mutation appeared as VUS, the clinical phenotype is classical for CMT. Further research is needed to elucidate the functional consequences of ITPR3 variants and explore its pathophysiology for CMT as a new form of the disease.

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Exploring the medication use: a comparative study of HTLV-1 patients facing HAM/TSP and those without

Ana Flávia Novaes[1], Davi Tanajura Costa[1], Fernanda Khouri Barreto[1], Qesya Rodrigues Ferreira[1], Carolina Souza Santana[1], Jéssica Oliveira de Souza Nascimento[1], Ariana Leal Borges da Cruz[1], Alice Palmeira Nascimento Cerqueira[1], Felipe de Oliveira Andrade[1]

[1]Universidade Federal da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Ana Flávia Novaes; Email: anaflavia635@gmail.com.

The human T-cell lymphotropic virus type 1 (HTLV-1) is a retrovirus capable of infecting human cells and causing severe diseases such HTLV-1-associated myelopathy/tropical spastic paraparesis (HAM/TSP) that is an inflammatory disease of the central nervous system, with symptoms typically appearing in the fourth decade of life. The symptoms manifest as spasticity, neurogenic bladder, and chronic pain. Due to its late onset, it can be associated with other comorbidities in the individual. Thus, the use of different classes of medications may be common among patients with neurological symptoms. This study aims to describe the medication use of patients infected by HTLV-1 with or without HAM/TSP. This cross-sectional study was carried out with patients living with HTLV-1 who are assisted at an HTLV-1 reference center in Bahia, Brazil, from March 2023 to May 2024. Data were obtained by analyzing medical records and during neurological examination. The statistical significance analyses were

performed using Fisher's exact test with SPSS software v. 27. A total of 110 patients were evaluated, 82 (84.5%) of whom were classified as non-HAM/TSP and 28 (25.4%) as HAM/TSP. The mean age of the patients classified as non-HAM/TSP was 48.66±16.78 years, of which 65 (80%) were female. The mean age of the HAM/TSP patients was 56.46±19.75 years, of which 21 (75%) were female. The use of lipid-lowering therapy was higher in the HAM/TSP group (7 or 25%), thyroid drugs (3 or 10%), and antihypertensives (11 or 39%). On the other hand, antidiabetics were more prevalent in non-HAM/TSP group (9 or 10%). No statistical difference between the groups was observed. It is important to note that among the most commonly used medications by the study group, antidepressants were used by the majority of them (11 out of 28 patients), ($p<0.01$). Depressive disorder in people living with HTLV-1 or HTLV-2 can be a primary or secondary consequence of the infection, so it should be evaluated individually, as it is known that the psychological profile of patients can influence their therapeutic process. The HAM/TSP group also reported higher use of analgesics (5 or 17%) with statistical differences between groups ($p<0.01$). This may lead to reflection on untreated neuropathic pain, self-medication, and, primarily, pain as a cardinal symptom in patients living with myelopathy. The data reveal that patients with neurological involvement associated with HTLV-1 infection have a diverse medication profile, offering insights into associated comorbidities like the use of lipid-lowering agents. The increased use of antidepressants highlights the often overlooked psychological impact faced by these patients. The notable difference in analgesic usage underscores the crucial need for effective pain management, especially for the neuropathic pain they experience.

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Exploring transient global amnesia: insights from demographic, clinical, cognitive and neuroimaging profiles

Ana Luiza Figueiredo Campos, Marina Isoni Martins[1], Paolla Giovanna Rossito de Magalhães[2], Antonio Jaeger[1], Rachel Laguardia Rego[2], Bárbara Cristina Silveira de Oliveira[2], Aretha Miranda Silva[2], Vinícius Canhestro Roedel[2], Augusto Antunes[3] , Leonardo Cruz de Souza[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[2]Hospital Madre Teresa, Belo Horizonte MG, Brazil.
[3]Axial Metódos de Imagem, Belo Horizonte MG, Brazil.

Correspondence: Ana Luiza Figueiredo Campos; Email: aninhaalfc8@hotmail.com.

Background: Transient global amnesia (TGA) manifests as a sudden onset of anterograde amnesia without other neurological deficits, typically lasting 2 to 24 hours, and resolving spontaneously. Its estimated incidence in the general population is 5-10 cases per 100,000 people per year. However, there is a scarcity of studies investigating TGA in the Brazilian population **Objective:** To characterize the demographical, clinical, cognitive and neuroimaging features in a series of consecutive TGA patients. **Methods:** This longitudinal and translational study focuses on patients presenting with TGA symptoms seeking emergency neurological care. Inclusion criteria encompass patients exhibiting cognitive impairments limited to episodic memory, validated by an external observer. All patients underwent the Brief Cognitive Battery (including the Mini-Mental State Examination [MMSE] and the Figure Memory Test for episodic memory), and the Facial Emotion Recognition Test (FERT), which assesses seven emotions: happiness, sadness, fear, disgust, surprise, anger, and neutral. Results from the TGA group were compared with data from cognitively health controls (n=20) matched for age, sex and education. **Results:** A total of 28 patients were included in the study, comprising 18 women and 10 men, with a mean age of 59.2 years. Medical history revealed that 45.83% had systemic hypertension, 25% had obstructive sleep apnea, and 16.7% had migraine. None of the participants had a prior history of stroke, intracranial hemorrhage, or epilepsy. Regarding potential triggering events, 37.5% of patients cited emotional stress, 16.7% Valsalva maneuvers, 8.3% sexual activity, while 16.7% denied any specific triggering events. Compared to controls, TGA patients had lower scores on the MMSE ($p<0.0001$), on the immediate memory ($p<0.0001$), delayed recall ($p<0.0001$) and in the recognition scores ($p<0.0001$). Patients did not differ from controls in the recognition of facial emotions (total score) or in the subscores of each emotion. Brain MRI was conducted on 24 patients, revealing bilateral high-signal abnormalities in the hippocampus on DWI in 20% patients, high-signal abnormalities in the right hippocampus in 30% patients, and in the left hippocampus in 15% patients. No signal abnormalities were observed in 35% patients. **Conclusion:** While clinical and cognitive data are homogeneous among patients, neuroimaging findings are heterogeneous, with different patterns of hypersignal. Patients have spared recognition of facial emotions. These findings highlight the strict memory involvement that characterizes TGA. Longitudinal follow-up studies are essential to deepen our understanding of the underlying pathophysiological mechanisms and the long-term cognitive outcomes in TGA patients.

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Explosive onset of non-epileptic seizures following levetiracetam introduction – is there a possible association

Cássia Elisa Marin Dapper[1], Gislaine Baroni[2], Rudá Alessi[2], Eliseu Paglioli[2], André Palmini[2], William Alves Martins[2]

[1]Universidade de Caxias do Sul, Caxias do Sul RS, Brazil.
[2]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Cássia Elisa Marin Dapper; Email: cassiaemarin@gmail.com.

Case presentation: A 14-year-old female high school freshman began neurological evaluation at our institution in 2019 following episodes of unconsciousness that began at age 7, characterized by unilateral head deviation, occasional generalized tonic-clonic movements, and tongue biting. She was diagnosed with epilepsy and effectively managed with oxcarbazepine, reducing seizures to less than one per year. In October 2018, a breakthrough seizure prompted a switch from oxcarbazepine to levetiracetam (LEV), considering her age and reproductive stage. Subsequently, her seizures evolved into episodes marked by disconnection, excessive salivation, ocular deviation, and rotational head and hip movements lasting up to five minutes. These episodes increased in frequency, often triggered by anxiety-provoking situations. Her parents noted new behavioral changes including irritability, impatience, inattention, and forgetfulness affecting both home and school life, leading to conflicts with her father. Her medical history included a term cesarean birth with nonconsanguineous parents, neonatal hypoglycemia, and a 14-day hospitalization at four months due to suspected metabolic issues and poor weight gain. Until age six, she experienced recurrent unexplained vomiting episodes that resolved without a definitive diagnosis. Childhood was generally peaceful with good academic performance, though occasional disagreements occurred with her mother and sister; she had a closer relationship with her father. Family history included only a paternal aunt with childhood febrile seizures. A 72-hour video-EEG captured three typical events during wakefulness characterized by vertical ocular deviation, blinking, ocular flutter, and irregular head and limb movements progressing to spitting movements lasting about six minutes. No corresponding EEG abnormalities were observed. Brain MRI findings were normal. A diagnosis of psychogenic nonepileptic seizures (PNES) was established, leading to referral for psychiatric assessment. LEV was discontinued, and oxcarbazepine was reintroduced gradually up to 1200mg daily, resulting in progressive improvement of PNES over subsequent months. **Discussion:** PNES and epileptic seizures (ES) can coexist in up to 5-60% and be a diagnostic and therapeutic challenge. In addition, patients with epilepsy have a close association with behavioral, psychiatric and developmental disorders, which may be influenced by anti-epileptic drugs effects. Behavioral side effects of LEV have been frequently reported, including aggression as well as change mood states such as depression, agitation, irritability and hyperexcitability. Furthermore, some recent published papers showed a possible association of LEV use and occurrence of PNES. **Final comments:** Our case highlights the possibility of explosive PNES onset after treatment with LEV in patients with no previous functional disorders. Such condition must be kept in mind with severe worsening of seizures upon introduction of LEV.

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Expression of cellular pluripotency regulated micrornas associated with tumor recurrence in meningiomas

Liliane Cristina da Silva[1], Victor Cunha Tirapelli[1], Ana Laura Ribeiro e Silva Stevanato[1], Janaina Regina Léllis[1], José Guilherme Marques Farinha[1], Mariana Matheus Pupin[1], Fernanda Maris Peria[1], Carlos Gilberto Carlotti Jr[1], Mucio Luiz de Assis Cirino[1], Daniela Pretti da Cunha Tirapelli[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: José Guilherme Marques Farinha; Email: zeguifarinha@hotmail.com.

Background: The expression of miRNAs is associated with a variety of diseases, including neoplasms. In recent years, a large number of abnormally expressed miRNAs have been shown to be effective in understanding the oncogenesis, development, progression and prognosis of meningiomas. Furthermore, it is known that miRNAs act as oncogenes or tumor suppressors and that they regulate essential molecular pathways such as transcription factors involved in the pluripotency phenotype of stem cells. Overexpression of these transcription factors is associated with metastatic formation and tumor recurrence in many types of tumors, including meningiomas. **Objective:** Therefore, our aim was to analyze the expression of microRNAs miR- 34a, miR-145 and miR-221 that regulate the pluripotency pathway of stem cells and correlate with tumor recurrence in grade I meningiomas. **Methods:** We used 30 samples, belonging to 15 patients who presented relapse of grade I meningiomas. **Results:** We observed low expression levels of miR-34a in the group of tumor relapses when compared to the groups of control individuals and primary tumors, which may be associated with the role of tumor suppressor of this miR. The miR-145 also showed a decrease in expression levels observed between the control group and the group of tumor recurrences. We also observed a decrease in miR-145 between the groups of the control group and the group of primary tumors. miR-221 did not differ between the groups studied.

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Expression of micrornas: miR-27A, miR-636, miR-1229 associated with memory in tonsils, hippocampus and blood of patients diagnosed with epilepsy

Adriana Lis Pereira[1], Victor Cunha Tirapelli[1], Lucas Cunha Tirapelli[1], José Guilherme Marques Farinha[1], Janaína Regina Lellis[1], Maria Julia Nunes[1], Jairo Pinheiro da Silva[1], Carlos Gilberto Carlotti Junior[1], Mucio Luiz de Assis Cirino[1], Daniela Pretti da Cunha Tirapelli[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: José Guilherme Marques Farinha; Email: zeguifarinha@hotmail.com.

Background: Epilepsy is a chronic disease of the central nervous system (CNS), which generates neurobiological, cognitive and psychosocial consequences. The most common type of focal epilepsy is temporal lobe epilepsy with hippocampal sclerosis, characterized by cell loss and gliosis in the hippocampus and amygdala. Several studies have demonstrated the presence of memory deficits in patients with difficult-to-control epilepsy. However, little is known about the molecular aspects involved in the process of changes in the cognitive function of patients with epilepsy. In this context, current literature has demonstrated that small RNA molecules called microRNAs can regulate molecular pathways important in epileptogenesis. Therefore, identifying possible modulation of molecular targets associated with memory by microRNAs can contribute to the development of new therapeutic approaches as well as tools that help in the diagnosis and prognosis of the disease. Temporal lobe epilepsy has been associated with cognitive dysfunctions, especially short- and long-term memory disorders. Many studies have demonstrated the presence of memory deficits in difficult-to-control epileptic patients. Our hypothesis is that regulatory microRNAs of molecular targets associated with memory may be involved in the prognosis of drug-resistant patients after surgical treatment. **Objective:** To analyze the expression of microRNAs: miR- 27a, miR-636, miR-1229 that control targets associated with memory in tonsils, hippocampus and blood of patients diagnosed with epilepsy. **Methods:** Hippocampus and blood samples from 20 patients with TLE were collected, 10 with good postoperative evolution (Engel I) and 10 with unsatisfactory postoperative evolution (Engel III and IV). Hippocampal tissue from deceased individuals and blood from healthy individuals were used as controls. The expression analysis of the miRNAs was performed using the RQ-PCR technique. **Results:** The microRNA-27a was differentially expressed between the control group and the epilepsy group both in the hippocampus and in the blood. However, the same was not observed in the tonsils. It was also differentially expressed between Engel I and III and IV both in the blood and in the tonsils and hippocampus. However, the expression pattern was inverse, that is, in the tonsils, miR-27a showed higher expression levels in the control group when compared to the epilepsy group and in the hippocampus and in the blood, the expression levels of this microRNA were higher in the epilepsy group. MicroRNA-636 was differentially expressed by observing the Engel classification in hippocampus. microRNA-1229 was not differentially expressed between the groups studied in tonsils.

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Expression of miR-19B and miR-7 in serum samples from patients with probable rbd as potential biomarkers

Sabrina Simplício de Araújo Romero Ferrari[1], Talita Guimarães Balieiro[2], Deborah Almeida de Oliveira[3], Simoneide Souza Titze-de-Almeida[3], Ricardo Titze-de-Almeida[1], Pedro Renato de Paula Brandão[2]

[1]Universidade de Brasília, Laboratório de Tecnologias para Terapia Gênica, Centro de Pesquisa de Grandes Temas, Divisão de Doença de Parkinson, Brasília DF, Brazil.
[2]Hospital Sírio-Libanês, Instituto de Ensino e Pesquisa, Brasília DF, Brazil.
[3]PROBE-PD Prodromal and overt Parkinson's Disease Epidemiological Study in Brazil.

Correspondence: Talita Guimarães Balieiro; Email: talitagbalieiro@gmail.com.

Background: MicroRNAs (miRNA) are small, double-stranded RNAs and post-transcriptional modulators of gene expression, which control the expression of messenger RNAs (mRNAs) by binding to complementary sequences, causing gene silencing. Aberrant miRNAs have been extensively studied regarding their diagnostic and prognostic value for various diseases. In this study, we sought to assess the expression patterns of miR-19b and miR-7 in serum samples from patients with REM sleep behavior disorder (RBD), a condition characterized by dream enactment during REM sleep due to loss of muscle atonia. The progression of idiopathic RBD to Parkinson's disease (PD) and Dementia with Lewy Bodies underscores its significance as a clinical indicator of the early stages of these synucleinopathies. Studies reveal that miR-19b levels are altered in patients with RBD. The altered expression of miR-19b occurs years before the onset of motor symptoms, suggesting its potential as an early biomarker for the prodromal phase of these disorders. The miR-7 has been implicated in the underlying mechanisms of PD, including the regulation of alpha-synuclein, dopaminergic cell death and neuroinflammation. **Methods:** The samples were collected from patients participating in the PROBE-PD - Epidemiological Study on Established and Prodromal Parkinson's Disease in Brazil Project (Candangolândia, Distrito Federal) and processed in the Technology for Gene Therapy Laboratory - Research Center for Major Themes (University of Brasilia). Peripheral venous blood from 6 controls and 6 patients with RBD was collected by a standardized procedure with serum collection tubes. After 30 minutes were centrifuged at 2000 xg for 10 min and maintained at 4°C. Following, in a safety cabin the serum was dispensed in aliquots into 2 mL cryovials and stored at -80°C. The extraction was performed using a commercial kit (miRNeasy Serum/Plasma Advanced) and quantified using Qubit (Invitrogen). The RT-qPCR reaction was performed with commercial kits for microRNA (TaqMan Advanced miRNA cDNA Synthesis and TaqMan Advanced miRNA Assays) and calibrated with miR-39-3p. Data analysis was performed based on the relative expression of delta delta Ct (2^-ΔΔCT). The GraphPAD Prism 9 software was used for statistical analysis. **Results:** The qPCR analysis revealed an increased expression of miR-19b and miR-7 in patients with probable RBD compared to the control group. Specifically, miR-19b and miR-7 exhibited fold increases of 7.206 (P=0.1647; t-test) and 8.890 (P=0.1694; t-test), respectively, in RBD patients. **Conclusion:** The observed trend towards increased expression of these microRNAs suggests that miR-19b and miR-7 are potential biomarker candidates for patients with RBD. The study is ongoing, with additional samples being analyzed to further assess the accuracy of miR-19b and miR-7 as biomarkers for PD.

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Eye movement patterns and decision-making in soccer: an eye-tracking study of professional athletes

Luís Eduardo D'Almeida Manfrinati[1], Maria Tereza Martinez[2], Diogo Haddad Santos[3], Ana Carolina Gomes[3], Isadora Wasserstein Anghinah[3], Renato Anghinah[4] , Alex Kinderman[5], Sephora Sabrina Candido de Almeida[3]

[1]Sindicato dos Atletas Profissionais do Estado de São Paulo, São Paulo SP, Brazil.
[2]HP Neurologia, São Paulo SP, Brazil.
[3]Santa Casa de Misericórdia de São Paulo, Centro de Cognição, São Paulo SP, Brazil.
[3]Faculdade de Medicina Nove de Julho, São Paulo SP, Brazil.
[4]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, Ambulatorio de Reabilitação Cognitiva Pós, TCE São Paulo SP, Brazil.
[5]Neurolign USA LLC, Pittsburgh, Pensilvânia, USA.

Correspondence: Diogo Haddad Santos; Email: dhaddadsantos@gmail.com.

Background: The importance of visual perception in sports performance is crucial, especially in high-speed sports like soccer. Professional athletes develop specific visual skills enabling rapid and accurate processing of information. Effective visual perception is pivotal for good performance in soccer. The eye-tracking (ET) technique offers insights into how athletes visually explore the playing field, providing a deeper understanding of the specific visual skills necessary for success. **Objective:** To compare the pattern of eye movement in soccer athletes, assessing the speed and efficiency with which athletes process visual stimuli and how variations in movement patterns influence decision-making during a soccer match. **Methods:** In a randomized double-blind study, professional soccer players from three teams (A, B, and C), N=63, aged 18 to 21 and older, underwent ET tests using the Dx100 system. Vergence Pursuit Test: Each athlete followed a target moving towards and away from them. Saccade Test: Participants fixed their gaze on a point while a visual stimulus appeared in different positions. Smooth Pursuit Test: Players followed a point moving in specific directions (horizontal or vertical). Optokinetic Test: The eye movements were recorded and analyzed to assess coordination in response to the optokinetic stimulus. **Results:** 1. Team's performance Saccade Horizontal and Vertical: Teams A, B, and C showed latency values below the normal limit (<220msec) and precision within the normal range. 2. Team's performance Saccade and Reaction Time: Team A had a saccade latency time of 216 msec, while Team B had 203 msec, both within normal limits for motor reaction time. 3. Smooth Pursuit Horizontal and Vertical: All teams showed gains within normal limits for low frequencies. Teams A and B also showed gains at higher frequencies. 4. Teams' performance, Optokinetic, 20 and 60 deg/sec: Teams showed visual tracking gains above normal at 20 deg/sec, but below normal at 60 deg/sec. Goalkeepers showed better performance than defenders. 5. Teams' performance, Subjective Visual Vertical: Teams A and B showed negative deviations, while Team C showed a positive deviation. Good visual response speed is essential for decision-making, with no significant differences between teams and the control group. Difficulties with high-speed stimuli were noted, highlighting a moderate capacity for visual tracking. Competitive athletes may show better results, indicating the need for further research. **Conclusion:** Eye-tracking provides a comprehensive analysis of eye movements in soccer athletes. Tests revealed positive results compared to normal limits, with goalkeepers showing better performance than defenders. Differentiated training benefits visual skills, and further studies are necessary to confirm results.

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Fabry disease, a diagnostic challenge: case report

Yasmin Ramalho Mandarino[1], Luana Cristina Stefanes[1], Mariana Moreira Alves[1], Pietra Cani Linzmeier[2], Natália Cardoso Darabas[2], Hannah Bang[2], Maria Eduarda Angelo de Mendonça Fileti[2], Thábata Emanuelle Martins Nunes[2], Carla Heloísa Cabral Moro[2], Alexandre Luiz Longo[2]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Yasmin Ramalho Mandarino; Email: yasminramalhoo@gmail.com.

Case presentation: A 17-year-old male patient visited the neurology emergency room due to intermittent loss of vision in his left eye. No abnormalities were found in the head CT and in the initial lab work, but an intraconal infiltrative lesion was detected on the MRI. He had a history of orbital pseudotumor in the right eye, diagnosed the

previous year. The patient reported ongoing follow-up with a rheumatologist for chronic pain in the axial joints since the age of 12, without a defined diagnosis, and abdominal pain and diarrhea after consuming dairy products. The family medical history didn't present relevant findings. Physical examination revealed optic disc atrophy in the right eye and optic disc stasis in the left eye, with visual acuity of 20/20 in the left eye and blindness in the right eye, with no other abnormalities. The orbital MRI revealed signal alteration and atrophy of the posterior intra-orbital, prechiasmatic, and chiasmatic segments of the right optic nerve, appearing as sequelae, with no changes in the left orbit. Laboratory tests showed alpha-galactosidase activity of 0.32µmol/L/h and lyso-Gb3 biomarker levels of 46.4ng/mL, with no other alterations. Based on these findings, a diagnosis of Fabry disease was made. Genetic sequencing of the GLA gene revealed the presence of a pathogenic variant in homozygosis described as cys142Arg, associated with alpha-galactosidase A deficiency. During the hospital stay, the patient was prescribed pulse therapy and presented complete resolution of the new symptoms. Optic nerve biopsy showed no abnormalities. The final diagnosis was orbital pseudotumor due to Fabry disease. Enzyme replacement therapy was indicated. The patient continues to be monitored with cranial and orbital MRI. The clinical condition has remained stable since. **Discussion:** Fabry disease is an X-linked disorder caused by a mutation in the gene encoding the lysosomal enzyme Alpha-Galactosidase-A, responsible for breaking down neutral glycosphingolipids and globotriaosylceramide in lysosomes. It progressively damages vital systems such as the heart, kidneys, and nervous system, due to systemic accumulation of these substances in tissues. This progressive buildup causes cellular dysfunction and can result in secondary inflammation or fibrosis. Fabry disease is rare, with a frequency of 1 in 40,000 in men and 1 in 117,000 in women. The pathophysiological link with the abnormal metabolism of glycosphingolipids is unclear and requires further studies to be elucidated. **Final comments:** Fabry disease is a rare genetic disorder that challenges diagnosis due to its heterogeneous presentation. In the typical form, symptoms generally begin around the age of 10, being the average age of diagnosis usually around 28. Early diagnosis is essential.

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Facial diplegia, sixth nerve palsy and hyperreflexia: rare variant of Guillian-Barré syndrome and response to plasmapheresis

Cássia Elisa Marin Dapper[1], William Alves Martins[2]

[1]Universidade de Caxias do Sul, Caxias do Sul RS, Brazil.
[2]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Cássia Elisa Marin Dapper; Email: cassiaemarin@gmail.com.

Case presentation: A 55-year-old man presented with a 2-week history of diplopia, facial paresis, and gustatory alterations. An upper airway infection had occurred two days before the onset of symptoms but resolved before admission. His previous medical history was unremarkable. Neurological examination revealed partial left sixth nerve palsy along with complete facial diplegia (House-Brackmann grade VI), so severe that it prevented him from closing his eyes, resulting in a bilateral Bell's sign. He was also unable to purse his lips. Deep tendon reflexes were brisk and symmetric in all limbs, but the plantar response was flexor. There were no weakness or any sensory symptoms, despite gustatory dysfunction. Cerebrospinal fluid (CSF) examination showed slightly elevated protein (48mg/dl) and a normal cell count (1 leukocyte/mm^3). Brain MRI revealed bilateral seventh nerve enhancement without any other lesions. Rheumatologic tests, cancer screening, and other studies were normal. Serologic tests showed no signs of Campylobacter jejuni or Borrelia burgdorferi infection. EMG demonstrated bilateral seventh nerve injury with severely reduced motor conduction amplitude (0.03mV on the left and 0.04mV on the right) and normal latency (3.85ms). Administration of methylprednisolone (1g/day) for 5 days did not alleviate his symptoms. Antibodies against gangliosides (anti-GM1 IgM and anti-DT1a) were positive, leading to a diagnosis of a Guillain-Barré syndrome (GBS) variant. Treatment with plasmapheresis was initiated, and after five sessions, his ocular movement returned to normal and facial diplegia began to recover. Eight months later, he remains asymptomatic, with facial movements almost completely returned to normal (House-Brackmann grade II). **Discussion:** GBS encompasses a spectrum of autoimmune neuropathies, with variants such as Miller Fisher syndrome and acute motor axonal neuropathy presenting distinctive clinical features. Facial diplegia, observed in some GBS variants, manifests as bilateral facial weakness, potentially accompanied by other cranial nerve involvement and areflexia, complicating the presentation and requiring prompt differentiation from other etiologies to initiate immunomodulatory treatments like IVIG or plasmapheresis, like in our case. Hyperreflexia is uncommon, nonetheless brisk reflexes might occasionally be observed early in the disease course or in specific variants. Pathophysiology involves molecular mimicry where antecedent infections trigger an immune response that cross-reacts with peripheral nerve components, notably gangliosides, leading to demyelination or axonal damage. Comprehensive neurological examination, CSF analysis showing albuminocytologic dissociation, and electrodiagnostic studies, alongside serological tests for specific antibodies, are pivotal for accurate diagnosis and management, with recovery potential varying widely among patients based on the subtype and severity of nerve involvement.

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Factors associated with intensive care unit admission for patients with acute stroke: a cohort study in a reference hospital in Bahia

Luiz Paulo Oliveira Martins[1], Gabriel Souza Medrado Nunes[1], Maria Eduarda Messias Vassoler[1], Milena Fernandes de Oliveira[1], Gabriel Teles de Oliveira Piñeiro[1], Thaís Barbosa de Oliveira[1], João Pedro Fernandes Gonçalves[1], Pedro Antonio de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Luiz Paulo Oliveira Martins; Email: luizpom2000@gmail.com.

Background: Intensive care unit (ICU) admission is common for patients in need of continuous monitorization. Physicians often need to assess whether patients with acute neurological injuries require ICU admission. However, the factors associated with ICU admission are not well explored in hospitalized stroke patients. **Objective:** To investigate clinical and laboratory factors associated with ICU admission in acute stroke patients. **Methods:** This study enrolled patients with acute stroke admitted to a referral stroke unit in Bahia, between July/2023 and March/2024. Clinical and epidemiological characteristics were extracted from patients' medical records or obtained through personal interviews. The initial National Institutes of Health Stroke Scale (NIHSS) was obtained through physician assessment during admission. Thrombolysis was defined as the administration of alteplase noted in medical records. Uremia and thrombocytopenia at admission were defined by levels of urea above 50mg/dl and platelets count below 150x109/L, respectively. The Mann-Whitney and Chi-Square tests were employed to evaluate associations. **Results:** The study included 213 patients, 51.2% of whom were female and the median age was 66.3 years (IQR: 59-74.3). During the hospitalization period, 20 patients (9.4%) were admitted to the ICU. Regarding comorbidities, 78.95% had hypertension, 36.2% had diabetes and 27.7% had dyslipidemia. Among all patients, the median NIHSS at admission was 7 (IQR: 4-12). In general, higher NIHSS values were associated with ICU admission ($p<0.05$). Thrombolysis was performed in 87 patients (40.8%) and its occurrence was not significantly associated with ICU admission ($p>0.05$). The mean urea value and platelets count were 35.8 ± 16.5 mg/d and 215x109±101x109/L, respectively. Uremia was diagnosed in 14 patients (6.6%), showing a significant risk of ICU admission ($p<0.05$; OR 4.00; 95% CI: 1.11 – 14.43), while thrombocytopenia was diagnosed in 28 patients (13.1%) and also presented a significant risk of ICU admission ($p<0.05$; OR 3.94; 95% CI: 1.37 – 11.30). **Conclusion:** Our cohort study identified no significant association between thrombolysis and ICU admission in acute stroke patients. However, higher NIHSS scores, uremia, and thrombocytopenia were all significantly linked to increased ICU admission rates. This identification may serve as early warning signs for adverse outcomes. By recognizing these clinical and laboratory factors, healthcare providers can promptly intervene, potentially preventing the progression to severe complications and the need for ICU admission. However, more comprehensive research is needed to better understand these associations.

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Fahr's syndrome: a case report

Maria Carolina Marçon Barroso[1], Laura Alonso Matheus Montouro[1], Matheus Vieira Santili[1], Letícia Raiane Fogari[1], Ana Luíza Duran Casseb[1], Marina de Carvalho Bofo[1], Carolina Mendes Rahal[1], Cibele Alexandra Ferro[1]

[1]Faculdade de Medicina de Catanduva, Catanduva SP, Brazil.

Correspondence: Maria Carolina Marçon Barroso; Email: barrosomariacarolina@gmail.com.

Case presentation: Male patient, 75 years old, was admitted due to the first bilateral clonic tonic seizure associated with sialorrhea. On the previous day, reported epigastric discomfort, excessive sweating and lipothymia – followed by postictal confusion. Brain CT with calcifications bilaterally and symmetrically affecting the semioval centers, basal ganglia, thalamus, and dentate nuclei. Laboratory tests were normal. He was discharged a few days later with a prescription for valproic acid 500mg, without adherence. One year later, he returned to the emergency room with a new clinical condition of tonic-clonic seizure and Lamotrigine 100mg/day was initiated. He denies consanguinity. Deceased brother with unknown neurological disease. He denies chronic kidney disease, known previous neurological diseases and addictions. **Discussion:** Fahr syndrome is a rare neurodegenerative condition characterized by bilateral and symmetrical calcification of the basal ganglia, secondary to another pathology or familial association, an autosomal dominant inheritance, or idiopathic. Its main causes involve calcium metabolism, such as hypoparathyroidism, genetic diseases, and infections. The clinical condition is variable, there are behavioral and mood changes, movement disorders, balance disorders, headache, parkinsonism and seizures. The diagnosis is made through neuroimaging tests with a characteristic pattern and the investigation should include research for secondary causes. In this case, the patient presented with episodes of tonic-clonic seizures, one of the possible and nonspecific clinical manifestations of the disease. In the investigation, cranial tomography identified bilateral and symmetrical calcifications of the centrum semiovale, basal ganglia, thalamus and dentate nuclei, a characteristic finding of the syndrome. This diagnostic hypothesis was raised, proceeding with etiological investigation, due to its importance in the prognosis of the disease, because, in some cases, the calcifications have a good response to the treatment of the underlying cause. The main disorder to be investigated is hypothyroidism, diagnosed by reduced serum parathyroid hormone and calcium, and elevated phosphorus. In addition, other tests should be performed, such as TSH, T4, renal and hepatic function, HSR, CPK, ANA, VDRL, copper, ceruloplasmin, in order to rule out infectious and organic causes. After the investigation,

which proved to be normal, raising the diagnostic hypothesis of idiopathic or familial Fahr syndrome, suggesting research of other cases in the family. Treatment should be aimed at controlling symptoms or the underlying disease, if present, improving quality of life and preventing disease progression. **Final comments:** Its investigation begins after adequate anamnesis of neurological and clinical symptoms associated with characteristic neuroimaging. It is a syndrome with multiple etiologies associated with different clinical management and prognosis.

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False negative genetic panel for spinocerebellar ataxias: a case report

Maiza de Oliveira Tenório[1], Lucas de Andrade Alexandre[1], Edvaldo Lucas da Costa Silva[1], Marcelo Palmares Oliveira e Silva[1], Paulo Felipe Bezerra Silva dos Santos[2], Amanda Sampaio de Sá[2], João Pedro Matos de Santana[1], Evelliny Gomes da Silva[1], Miriam Carvalho Soares[1], Marcos Eugênio Ramalho Bezerra[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Centro de Ciências Médicas, Recife PE, Brazil.

Correspondence: Paulo Felipe Bezerra Silva dos Santos; Email: pfbsilva2@gmail.com.

Case presentation: Three sisters, aged 59, 62 and 65 years, were followed in an outpatient clinic of a public university hospital, due to a clinical presentation of slowly progressive imbalance and incoordination over a 10 years period. They reported similar cases in the family (father, paternal grandmother and paternal uncles). On physical examination, they presented dysmetria, dysdiadochokinesia and impaired tandem gait. Due to clinical suspicion, a panel for repeat expansions genes of spinocerebellar ataxias (SCA 1, 2, 3, 6 and 7) was performed in one of the sisters, with a negative result. Next gene sequencing was performed on the other two sister, with the following result: heterozygosity in the EEF2 gene (SCA26) present in two sisters and absent in the other, and heterozygosity in the CCDC88C gene (SCA40) present in two sisters and unknown in the other. As the diagnosis remained undefined, it was decided to perform, on the same patient with a previous negative panel, a molecular diagnostic test for SCA3, with a positive result this time: an allele with 20 repeats and another with 62 CAG repeats in the ATXN3 gene. Therefore, a presumed diagnosis was performed on the other sisters and a new genetic panel was requested for them. **Discussion:** The diagnosis of spinocerebellar ataxias is made based on clinical signs of progressive ataxia, family history, and positive genetic testing. Spinocerebellar ataxia type 3, known as Machado-Joseph disease, is the most common autosomal dominant subtype and results from the ATXN3 gene mutation, caused by a CAG repeat expansion on the long arm of chromosome 14. It presents a broad spectrum of manifestation, encompassing pyramidal symptoms, peripheral neuropathies, sleep disorders and oculomotor abnormalities. Currently, the definitive diagnosis is through a genetic panel capable of detecting the mutation causing the disease, which has an accuracy of around 99%. Rare diagnostic errors can occur due to rare mutations or polymorphisms. Other studies have already shown false negative results in cases of SCA, especially when there is a very high number of repetitions, in which the PCR technique shows 2 alleles that appear to have the same number of repetitions, in a normal quantity. In this case, the number of repetitions was not that high, but a false negative was also possible. Further investigations were compatible with SCA26 in two sisters and SCA40 in two other sisters, which are extremely rare subtypes of genetic ataxias. SCA 26 was described in the literature in a Norwegian family and SCA 40 in a Chinese family. As the result was divergent between patients from the same family and there was a similar clinical condition between them, it became important to review the investigation. **Final comments:** Even if the genetic panel yields negative results, due to the suggestive clinical condition, it should be considered for repetition, taking into account the possibility of a false negative result.

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Familial amyloidosis with compound heterozygosity of the V122I and V30M: a case report

Matheus da Silva de Oliveira[1], Antônio Marcos Reissureição[1], Felipe Bento Michelato[1], Luís Fernando Fernandes Miranda[1], Yves Henrique Ramos Mansano[1], Pamella Francisquini Gardin[1], Ana Letícia Manso Assakawa[1], Isabela Kirach Gaio[1], Maria Júlia Costa Rocha[1], Gabriel Bortoli Ramos[2]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Matheus da Silva de Oliveira; Email: ra125466@uem.br.

Case presentation: The first case of Familial amyloidosis identified was a V122I/V30M compound heterozygosity. The first symptoms began at age of 50 with congestive heart failure and recurrent episodes of asthenia. After four years without a final diagnosis, the clinical condition progressed with neuropathic pain and proprioceptive dysfunction. The cardiac and neurological symptoms, associated with concentric hypertrophy of the heart identified by magnetic resonance imaging, raised the diagnostic hypothesis of amyloidosis, which was confirmed by pyrophosphate scintigraphy. Subsequently, a hereditary study was initiated through material extracted via oral SWAB for sequencing the TTR gene for Hereditary Transthyretin Amyloidosis. As a result of this study, 17 tests came back positive: nine individuals with heterozygosity for V122I variant, six for V30M and two with compound heterozygosity for V122I/V30M. **Discussion:** Transthyretin-associated amyloidosis is a genetic disease with autosomal dominant inheritance, caused by mutations in the TTR gene. Consequently, it commonly manifests in multiple individuals from the same family. The most significant aspect of the case is the identification of familial amyloidosis with 2 cases of compound heterozygosity for the V122I and V30M variants. Furthermore, the description of the first identified case allows the inference of which variant manifested first in the patient. The current literature describes the V122L variant as predominantly related to cardiac amyloidosis and reports the following main findings: cardiomegaly, conduction block, arrhythmia, anginal pain, congestive heart failure and sudden death. On the other hand, V30M is associated with amyloid neuropathy, whose main findings are: sensorimotor polyneuropathy, carpal tunnel syndrome, autonomic dysfunctions, possible evolution with cardiomyopathy, nephropathy and CNS symptoms. Although the V30M variant may progress to cardiomyopathy, this is not considered an initial finding. Therefore, it can be inferred that the V122I variant preceded its manifestations compared to the V30M variant. **Final comments:** Familial amyloidosis is a rare disease. The fact that the disease presents itself as a compound heterozygosity of V122I/V30M variants makes it even rarer. For this reason, the medical understanding of the different presentations of the disease is crucial to ensure an accurate diagnosis and to promote a treatment that guarantees the best prognosis for the clinical condition.

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Familial amyloidotic polyneuropathy: an epidemiological study at a university hospital

Camila Oliveira dos Santos[1], Larissa Bittencourt de Carvalho Chaffin[1], Ligia Rocha Andrade[1], Julia Chaves Cordovil[1], Márcia Maria Jardim Rodrigues[1]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Camila Oliveira dos Santos; Email: camilaodsmedicina@gmail.com.

Background: Familial transthyretin amyloidotic polyneuropathy is a severe sensory, motor and autonomic neuropathy that comes from autosomal dominant inheritance. It happens because of the deposition of amyloid material in peripheral nerves and other organs. It can progress because of a severe functional incapacity and, when not treated properly, a person can die in 10 years, approximately. Diagnosis is based on clinical criteria and the confirmation comes by the identification of the mutation in the TTR gene. **Objective:** To identify the epidemiological profile of patients treated by the Amyloidosis team in the neuromuscular diseases clinic. **Methods:** It is a quantitative, observational, and descriptive study that reviewed the medical records of patients seen by the neurology department between August 2023 and July 2024. **Results:** The sample consisted of 46 patients, most of them part of the male gender, with 60.86%. The percentage of deaths in the sample was 4.34%. For the consanguinity parameter, 78.26% had this information in their medical records, of which 30.55% reported consanguinity and 63.88% denied cases in the family. In the family index evaluation, 82.60% of the patients provided the information: the most frequent mentions were the father - with 34.42% - and the patient himself - with 23.68%. 68.88% of patients were symptomatic, of whom 69.56% reported the age of their first symptoms:12.5% had an early onset – that means before the age of 30 - and 46.87% had a late onset - after the age of 50. The average age was approximately 47.97 years, with a standard deviation of around 18.64 years. Among the 34 patients who reported first symptoms, the analysis revealed that paresthesia was the most common symptom, with 18 occurrences. 100% of the patients underwent genetic testing: the Val 50 Met and Val142 Ile variants were the most frequent, with 36.95% and 23.91%, respectively. **Conclusion:** The evaluation of this group of patients revealed that, although familial amyloidotic polyneuropathy is a rare disease, it should be considered in patients that revealed sensory-motor polyneuropathy with dysautonomia, even without a family history. Evaluating the epidemiological profile of this group of patients can provide information on patterns that can guide future research, improve clinical care and reduce underdiagnosed cases.

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Familial distal amyotrophy sord related

Nadson Bruno Serra Santos[1], Izadora Vieira Bolzani Lopes Lima[1], Diego Silva Figueredo[1], Aldrin Pedroza Martins[1], Anamarli Nucci[1], Marcondes Cavalcante França Jr.[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Nadson Bruno Serra Santos; Email: nadson_bruno@hotmail.com.

Case presentation: A 63-year-old man was referred to our outpatient clinic and reported frequent stumbling, difficulty in running and jumping, progressing to loss of strength and atrophy in the lower limbs since he was 12 years. At the beginning his symptoms were predominantly distal, with later involvement of the proximal muscles in the lower limbs. He developed bilateral foot drop without complains in sensitivity, and now he is walking with unilateral support. On physical examination he had muscle strength graded IV+ (MRC scale) in iliopsoas, quadriceps and hamstrings. His anterior tibialis and soleus were graded zero and extensor hallucis longus graded I.

Reflexes were all reduced with bilateral abolished Achilles tendon response. His younger brother was 55-years-old by the time of consultation, with past medical history of hypertension and diabetes, and reported a similar clinical picture, with a later onset of symptoms at 17 years. He presented significant loss of strength and muscular atrophy of distal predominance in the lower limbs. On physical examination he had predominant distal weakness with MRC with grade zero strength in the anterior tibialis and triceps surae and abolished reflexes in lower limbs. Both siblings had electroneuromyography with evidence of a chronic diffuse, symmetrical, motor, axonal process, without signs of current denervative activity, of moderate intensity in the upper limbs and severe intensity in the lower limbs. Molecular genetic analysis was performed and confirmed mutation of the SORD gene: c.757del p.(Ala253Glnfs*27) // c.778G> C p.(Gly260Arg). **Discussion:** Despite advances in molecular genetics, Neuropathies "Charcot-Marie-Tooth like" disease of the axonal type (CMT2) and distal hereditary motor neuropathies (dHNM) remain without molecular diagnosis in almost 70% of cases, especially when they are sporadic. In 2020, description of biallelic variants of SORD gene opened a new diagnostic range, highlighting the great potential to solve part of this gap. SORD gene encodes sorbitol dehydrogenase, a key enzyme in the polyol pathway and possibly involved in the development of diabetic neuropathy. CMT2/dHMN first symptoms start 2nd-3rd decade of life and are characterized by cavus feet, a slowly progressive length-dependent motor deficit of lower limbs, often abolished reflexes in lower limbs and preserved in upper limbs. EMG usually reveals distal axonal motor neuropathy, with moderate reduction in motor conduction velocities and/or reduction in the amplitudes of sensory potentials. **Final comments:** Several studies have increasingly shown that biallelic variants of SORD are a frequent cause of autosomal recessive CMT2/dHMN. Screening for biallelic variants of SORD has been recommended in patients with axonal peripheral neuropathy and predominant distal myopathy. Recent studies highlight the potential utility of fasting serum sorbitol and CK levels as sensitive biomarkers of SORD-diseases.

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FDG-PET changes in LGI1-encephalitis

Giuseppe Dick Bonato[1], Carolina Heck Haubrich[1], Leticia Bassani Devens[1], Rafael Do Amaral Cristovam[1], Camila Lima Ribeiro[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Carolina Heck Haubrich; Email: carolhhaubrich@gmail.com.

Case presentation: We present the case of a 63-year-old female who was brought to the emergency department due to seizures and progressive memory deficits. She had a prior hospitalization 3 months earlier after presenting with a new-onset generalized tonic-clonic seizure, following a 1-month period of insomnia and forgetfulness. Initially, she was diagnosed with a symptomatic seizure due to severe hyponatremia (116mEq/L), thought to be secondary to multiple oral nootropic drugs. She had no comorbidities, and both brain MRI and prolonged EEG monitoring were normal. She was discharged with normal blood sodium levels and minor improvement in cognitive status, with the recommendation to withdraw oral drugs. During the following 3 months, however, she experienced progressive cognitive deterioration. An FDG-PET was ordered and showed hypermetabolism in the right temporal lobe, and her MoCA score was 18. At the second admission, she had three new short and self-limited episodes of generalized tonic-clonic seizures. Her sodium level was again 116mEq/L. After an extensive search for the cause of hyponatremia, a diagnosis of probable SIADH was made without an evident clinical cause. A new MRI showed no significant findings, and continuous EEG suggested cortical dysfunction with occasional right TIRDAs and FIRDAs. Lumbar puncture results were normal, without pleocytosis and oligoclonal bands. Oral levetiracetam was started, and hyponatremia was slowly corrected; the patient remained seizure-free. Serum anti-LGI1 was ordered, and the patient was discharged with close outpatient follow-up. A month later, results came back positive for anti-LGI1, and the patient underwent corticotherapy with IV 1g methylprednisolone for 5 days followed by oral 80mg prednisone. She showed marked cognitive improvement in the first week after therapy, with MoCA score improving to 26, with minor visuospatial memory deficits. **Discussion:** We presented a case of anti-LGI1 encephalitis with significant improvement following corticotherapy, as consistently shown in previously reported cases. Clinical criteria for limbic encephalitis were all met except for the absence of MRI changes. Most cases present with typical bi-temporal changes on MRI; however, they can show multiple patterns of impairment. Metabolic imaging could reveal more subtle temporal changes and act as a more precise tool for diagnosing limbic encephalitis. There is, however, a lack of definite criteria for analyzing FDG-PET imaging, as anti-LGI1 encephalitis can present as either hypo or hypermetabolism in the temporal lobes or basal ganglia. The presence of anti-LGI1 antibodies in such cases is indispensable. **Final comments:** Anti-LGI1 encephalitis is considered the second most common autoimmune encephalitis and tends to show good outcomes. It may be beneficial to have a low threshold for considering it even when not all criteria for limbic encephalitis are met, especially if paraclinical signs such as hyponatremia are present.

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Feasibility analysis of using non-invasive monitoring technology to assess brain compliance during sleep

Karla Carlos[1], Carla de Oliveira Carletti[1], Lívia Jane Galvão Farias[1], Tainã Paulo Zanata Trombetta[1], Douglas Sousa Soares[1], Ana Flavia Machado Pitz[1], Letícia Lambert[1], Lucas Martins Teixeira[1], Gilmar Fernandes do Prado[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Karla Carlos; Email: ka.carlos1@hotmail.com.

Background: Accurately monitoring intracranial pressure waveforms in a noninvasive, cost-effective, and safer manner could assist in managing patients with imbalances in cerebral homeostasis and autoregulation challenged by some sleep disorders. The Brain4care (B4C) technology can detect small cranial deformations caused by changes in intracranial pressure linked to the cardiac cycle. **Objective:** To analyze the feasibility of using the B4C device with a Type I sleep study (PSG) all night. **Methods:** After completing the PSG setup, we placed the B4C headband around the patient's head, letting the sensor just above the ear at the frontotemporal transition. We verified the register time, signal continuity, sensor displacement, device tolerability, and quality of derived variables like wave morphology relation P2/P1, and entropy. A reliable signal should be 5 minutes long and free of any artifacts for analysis. **Results:** We included 51 participants (30 females), with a mean age of 39±12 years. The sensor was positioned on the left side in all patients due to the location of the right oculogram electrode of the PSG. The B4C recording's duration mean was 315±153 min (5 hs and 15 min) of continuous monitoring (maximum: 468; minimum:162). The average morphology of the acquired pulse was analyzed minute by minute, and the analysis did not include minutes with insufficient sample sizes. The average entropy was 2.0 ± 0.44 (maximum value of 2.2; minimum value of 0.5), representing the degree of data dispersion. Discussion. This is the first study showing that a B4C headband can be tolerated by most patients during a Type I sleep study, which requires a lot of wiring, without causing additional discomfort to the patient. The proper and precise placement of the device allows it to remain in stable contact with the patient's scalp, ensuring good acquisition of pulse morphology determined by small cranial deformations associated with ICP. Most studies documenting the use of this device involve an average usage time of 7.5 minutes for data recording. There is no data about the behavior of cerebral compliance during a night sleep monitored by PSG. Once the brain needs to precisely control the cerebral flow, and sleep is a neurophysiological and physiological variable state that includes the brain, it is of interest to monitor how brain compliance (indirectly associated with ICP) behaviors in normal and abnormal sleep. **Conclusion:** Noninvasive intracranial compliance monitoring technology through the Brain4care® device during sleep is safe and feasible. Our maximum recording time was 25 times longer than in other neurological research protocols. No patients experienced decoupling or loss of the Brain4care® device during the night of sleep. Patients reported that using the device did not cause any painful discomfort. The sensor should be positioned on the left side when used in conjunction with PSG.

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Femoral neuropathy secondary to compression by retroperitoneal hematoma: a rare differential diagnosis for neuroschistosomiasis

Mariana Holanda Gameleira[1], Maria Iris de Morais Machado[1], Rosevaldo Alves Junior[1], Lucas Chaves Lima[1], Brenda Letícia Lopes Batista[1], Yanne Almeida Aguiar[1], Brenda Albuquerque de Araújo Marinho[1], Bárbara Borges de Oliveira[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Mariana Holanda Gameleira; Email: marianagameleira_@hotmail.com.

Case presentation: A 53-year-old female patient came to hospital with sudden onset of low back pain radiating to the lower limbs, worse on the left, accompanied by ipsilateral motor and sensory deficits. She has advanced chronic liver disease due to schistosomiasis, and was in the ICU about two weeks before the neurological episode due to acute pulmonary edema. She developed severe low back pain and motor and sensory deficits in the left lower limb, and hematocrit drop requiring blood transfusion. No sphincter disorders. The examination showed weakness in hip flexion and knee extension, patellar hyporeflexia on the left and hypoesthesia and pain in the anterior aspect of the thigh and medial aspect of the leg. As the patient had schistosomiasis, cerebrospinal fluid analysis, which was normal, and imaging tests were performed for differential diagnosis. Magnetic resonance imaging spine did not show compressive disturbance compatible with the clinical case. However, a large infrarenal retroperitoneal hematoma was observed, compressing the left psoas muscle. Abdominal arteriography was performed to define the bleeding origin, showing splenorenal shunt and mass effect of the hematoma on the left kidney and psoas muscle. Electroneuromyography of the lower limbs was also performed, showing acute left lumbar axonal plexopathy (femoral nerve mostly affected) related to compression by retroperitoneal hematoma. Discussed case with surgery team, conservative treatment was chosen because of the risk of hemodynamic instability. **Discussion:** In patients with liver disease, there is an increased risk of complications such as digestive bleeding and coagulopathies, which can cause hematomas in the retroperitoneal region. Hemorrhage in iliopsoas muscle is a rarely described etiology of neuropathy. Its presentation is usually unilateral, with symptoms related to compression of the nervous plexus of the lower limb and with a wide differential diagnosis, one of which is neuroschistosomiasis. Symptoms depend on the presence of the parasite in the brain, cerebellum, subarachnoid space or spinal cord; the last one is most commonly affected. For

understanding the symptoms, anatomical knowledge is essential. The femoral nerve begins at the union of L2-L4 roots, which run through the iliopsoas muscle tendon and the femoral canal, providing motor and sensory innervation to the lower limb. Due to the compressive action of the hematoma, the clinical is wide, initially with sudden low back pain, up to paresthesia or paresis, and massive bleeding and shock may occur. The diagnosis is based on clinical and imaging examination showing increase volume on affected muscles, in addition to compatible electroneuromyography. Treatment involves hematoma drainage and control the underlying etiology. **Final comments:** Ileopsoas retroperitoneal hematoma should be considered in cases of compressive axonal injury in the lower limbs as a cause of significant femoral neuropathy.

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Finding the spot: results of a noninvasive strategy in MRI negative patients submitted to epilepsy surgery guided by ECoG

William Alves Martins[1], Fernanda Schuh Martins[1], André Luis Fernandes Palmini[1], Eliseu Paglioli[1], Thomas Frigeri[1], Rafael Paglioli[1], Ricardo Paganin[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Fernanda Schuh Martins; Email: fschuhmartins@gmail.com.

Background: Epilepsy surgery is crucial for treating patients with pharmacoresistant epilepsy. Seizure freedom is closely linked to the complete resection of the lesion, but patients with normal or negative MRI results have poorer outcomes. For these patients, extensive presurgical investigations often fail to provide sufficient data for a surgical plan, leading to the use of chronic extra-operative monitoring with subdural grids or SEEG. However, this approach is often too costly for many epilepsy patients, especially in developing countries. **Objective:** To describe the surgical outcomes of MRI-negative patients who underwent non-invasive investigations followed by ECoG-guided cortical resection. **Methods:** From 2012 to 2019, 62 MRI-negative patients underwent epilepsy surgery at the Porto Alegre Epilepsy Program (PAESP) and had at least 12 months of follow-up. The cohort included 36 males (58.1%) and 4 patients (6.5%) with a history of febrile seizures, with a mean age at surgery of 29±15 years. The median follow-up period was 43 months (12-146 months), showing a significant reduction in seizure frequency (58.5 to 7.2; p=0.001). Resections were temporal in 34 patients (54.8%), frontal in 17 (27.4%), multi-lobar in 4 (6.5%), occipital in 5 (8.1%), and parietal in 2 (3.2%). At 12 months, 29 patients (47%) were seizure-free, with 20 (32.3%) remaining completely seizure-free (Engel Ia) at last follow-up. Engel class distribution was I: 25 (40.3%), II: 13 (21%), III: 14 (22.6%), and IV: 10 (16.1%). There was a small trend toward better seizure freedom for temporal resections at last follow-up (41.2% vs. 21.4%, p=0.112). **Results:** Our long-term results, showing 32% seizure freedom in MRI-negative patients, align with literature that typically involves intracranial EEG. A meta-analysis indicated seizure-freedom rates of 34-45% for nonlesional MRI patients, compared to higher rates for lesional patients. Late recurrence occurred after a year in a third of those initially seizure-free, yet Engel I was seen in 40.3% of patients in a median follow of 43 months. This pattern of recurrence could be due to more widespread underlying conditions like FCD type I, the most common pathological finding in nonlesional epilepsies, leading to reactivation of epileptic foci following tapering of antiseizure drugs. Consistent with our findings, Wang et al. also identified FCD I as the predominant pathological substrate in 40% of MRI-negative patients. **Conclusion:** Combining strong electroclinical hypotheses and noninvasive tests with ECoG-guided resection can be an effective strategy. Novel noninvasive evaluation methods are needed to expand access to epilepsy surgery in low-resource countries.

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Findings from the illuminate prospective natural history study (NHS) in individuals with adult-onset leukoencephalopathy with axonal spheroids and pigmented glia (ALSP)

Paulo Victor Sgobbi de Souza[1], Wolfgang Koehler[2], David D. Lynch[3], Jeffrey M. Gelfand[4], Charles Wade[3], Nicole Wolf[5], Caroline Bergner[2], Ludger Schols[6], Zbigniew K. Wszolek[7,8]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]University of Leipzig Medical Center, Leipzig, Germany.
[3]University College London, London, England.
[4]University of California San Francisco, San Francisco, United States of America.
[5]Amsterdam University Medical Centers, Amsterdam, Netherlands.
[6]Tübingen University Hospital, Tubigen, Germany.
[7]Mayo Clinic, Jacksonville, States of America.
[8]Illuminate Study

Correspondence: Paulo Victor Sgobbi de Souza; Email: pvsgobbi@gmail.com.

Background: ALSP is a rare, autosomal-dominant, neurodegenerative disorder caused by a colony-stimulating factor1 gene (CSF1R) mutation and characterized by brain white matter (WM) demyelination, swollen axons, and pigmented glial cells and clinical symptoms of cognitive, neuropsychiatric, and motor dysfunction, typically developing around 40 years of age. No approved therapies exist. **Objective:** Report clinical, biomarker, and MRI findings from ILLUMINATE, the first NHS in individuals with ALSP (NCT05020743). **Methods:** ILLUMINATE is a prospective, multicenter NHS of individuals with definitive or prodromal (satisfies genetic and radiologic but not full clinical symptom criteria) ALSP. Up to 50 enrollees will be followed for 24 months, with clinical assessments and MRI collected at screening and every 6 months. MRI ALSP severity score and WM lesion and regional brain volumes will be derived; blood and cerebrospinal fluid (CSF) disease biomarkers will be measured at specific visits. **Results:** As of September 2023, of 41 enrollees with a confirmed CSF1R mutation, 23 were symptomatic (mean±SD age, 45.2±9.5 years; 52.2% female) and 18 were prodromal (42.9±17.0 years; 47.8% female). Baseline WM lesion and ventricle volumes were higher in symptomatic vs prodromal participants, while total gray matter volume was lower. Longitudinally, greater expansion in WM lesion volume and ventricle volume and gray matter volume reduction were observed in symptomatic vs prodromal participants. Baseline soluble CSF1R was substantially reduced in prodromal and symptomatic participants vs healthy volunteers, indicating reduced microglial activity. CSF and serum levels of neurofilament light chain (NfL), a marker of neuroaxonal injury, were elevated multifold in symptomatic participants vs prodromal and healthy individuals. Longitudinal fluid biomarker and clinical data will be disclosed at the meeting. **Conclusion:** These ILLUMINATE findings demonstrate that MRI measures and NfL levels are sensitive markers of ALSP pathophysiology, with potential to inform optimal clinical endpoints and therapeutic intervention study design in ALSP and improve overall understanding of disease progression.

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Findings in optical coherence tomography in patients with Wilson's disease: a cross-sectional study.

Hélio Afonso Ghizoni Teive[1], Aline Shiokawa[2], Brendha David Pinto[1], Léo Coutinho[1], Luiz Fernando Pabis[1], Mario Teruo Sato[1], Max Victor Peter Schramm[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Retina e Vitreo Consultoria, Curitiba PR, Brazil.

Correspondence: Brendha David Pinto; Email: brends3003@gmail.com.

Background: Wilson's disease (WD) is an autosomal recessive disorder resulting in a defect in copper metabolism. This defect leads to decreased excretion of this metal and its accumulation in organs such as the liver, brain, and eyes. Clinical manifestations are often nonspecific and variable, including extrapyramidal symptoms and the presence of Kayser-Fleischer rings, indicating extrahepatic copper accumulation. Diagnosis commonly relies on the triad: serum levels of ceruloplasmin and copper, urinary copper excretion and, when necessary, liver biopsy. However, the early detection of chronic neurodegeneration remains challenging, as neurological symptoms usually appear after hepatic symptoms, thereby complicating both monitoring and comprehension of disease progression. Therefore, it is crucial to have methods that allow the tracking of neuronal degeneration for maintaining the health of affected individuals. The literature suggests the use of Optical Coherence Tomography (OCT), a non-invasive imaging exam already employed in the monitoring of patients with multiple sclerosis. This method is based on measuring the thickness of axons that compose the retinal nerve fiber layer, assessing its neuronal integrity. However, due to the limited number of patients with the disease undergoing the exam, it is still not possible to use OCT as a marker of neurodegeneration for WD. **Objective:** To investigate whether there is a significant difference in the retinal changes detected by OCT between patients with neurological manifestations of WD and those with exclusively hepatic symptoms. **Methods:** Ten patients with neurological manifestations and ten with exclusively hepatic manifestations of the disease were selected, and their clinical and epidemiological information was recorded. Subsequently, they will undergo neurological examination guided by the Global Assessment Scale for Wilson's disease (GAS-WD), ophthalmological evaluation, and OCT examination to measure the retina's thickness in six ocular sectors. After data collection, normality analysis will be performed using the Shapiro-Wilk test. For normally distributed data, the t-student test will be used, and for non-normally distributed data, the Mann-Whitney test will be employed. **Results:** The study is currently ongoing, with patient data being collected and analyzed. **Conclusion:** The research results are expected to show a high incidence of retinal changes in patients with neurological symptoms of WD. Such findings could support the use of OCT as a method for monitoring neurodegeneration progression in these individuals.

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Fine motor dexterity impairment in a large group of people with epilepsy without lesions in the motor areas

Gabriela Ribeiro Prada[1], Marina Alvim[1], Rafael Batista João[1], Lucas Scárdua Silva[1], Thais Maria Bezerra[1], Fernando Cendes[1], Mateus Henrique Nogueira[1], Clarissa Lin Yasuda[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Gabriela Ribeiro Prada; Email: g236806@dac.unicamp.br.

Background: While several neuroimaging studies have demonstrated gray matter atrophy in the precentral gyri, basal ganglia and alterations of corticospinal tracts in focal and generalized epilepsies, little is known about the fine motor alterations in people with epilepsy (PWE). Besides, fewer studies have investigated the impact of symptoms of anxiety and depression on motor performance. **Objective:** To investigate gross and fine motor dexterity in PWE and possible associated factors such as

depression and anxiety. **Methods:** We interviewed and tested 195 people with epilepsy (PWE, median of 35 years of age and 11 years of education) at a Tertiary Centre (before their outpatient appointment) and 252 healthy volunteers (HV, median of 35 years of age and 15 years of education) at the University Campus. All individuals performed the 9-hole peg test (9-HOLE), Box and Block Test (BBT), Tower of Hanoi (TH), the Block design subtest of the Wechsler Adults Intelligence Scale (BD-WAIS), Generalized Anxiety Disorder Questionnaire (GAD-7) and Neurological Disorders Depression Inventory for Epilepsy (NDDI-E). PWE was separated into seizure-free (76 subjects), lower-frequency (13 subjects) and pharmacoresistant (85 subjects). We used SPSS23 for statistical analyses with non-parametric tests for group comparisons (Mann-Whitney and Spearman correlation). We excluded subjects with intellectual or motor impairment. **Results:** Although the groups of PWE and HV were balanced for age (p=0.7), scores of anxiety (GAD-7, p=0.4) and of depression (NDDI, p=0.2), we detected poor performance of 9-HOLE (bimanual, p<0.001), BBT (bimanual, p<0.001), TH (bimanual, p<0.001), and of BD-WAIS (bimanual, p<0.001). Overall, the PWE performance was 15-22% worse than their age-paired controls; patients were twice slower than controls to finish the first movement of the Tower of Hanoi. We did not observe significant differences in motor performance related to seizure control. **Conclusion:** Although groups of patients and controls presented similar levels of anxiety and depression, our results consistently showed poor motor performance for PWE, related to both fine dexterity (9-hole peg test) and gross tests (Box and Block Test). Besides, PWE also presented worse scores for more complex tests (Block design subtest of the Wechsler Adults Intelligence Scale and the Tower of Hanoi) that require integration of different cognitive functions. These findings add new information related to epilepsy comorbidities, as little is known about fine motor impairment in patients with generalized and focal epilepsies (without lesions in the motor cortex, corticospinal tracts or basal ganglia). Future analyses with subgroups of different types of epilepsies and the effects of different types of medications are necessary to understand the dysfunction and create specific strategies for rehabilitation.

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FIREFISH parts 1 and 2: 5-year efficacy and safety of risdiplam in type 1 SMA

Edmar Zanoteli[1], Maria Mazurkiewicz-Bełdzińska[2], Giovanni Baranello[3], Odile Boespflugtanguy[4], John W. Day[5], Nicolas Deconinck[6], Andrea Klein[7], Riccardo Masson[8], Eugenio Mercuri[9], Kristy Rose[10]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[2]Medical University of Gdańsk, Gdańsk, Poland.
[3]The Dubowitz Neuromuscular Centre, London, UK.
[4]Université Paris Cité, Paris, France.
[5]Stanford University, Palo Alto, USA.
[6]Université Libre de Bruxelles, Brussels, Belgium.
[7]University of Bern, Bern, Switzerland.
[8]Istituto Neurologico Carlo Besta, Milan, Italy.
[9]Catholic University and Nemo Pediatrico, Rome, Italy.
[10]University of Sydney and Sydney Children's Hospital Network, Sydney, Australia.

Correspondence: Edmar Zanoteli; Email: edmar.zanoteli@usp.br.

Background: Risdiplam (EVRYSDI®) is a centrally and peripherally distributed, oral survival of motor neuron 2 (SMN2) pre-mRNA splicing modifier, approved in over 100 countries worldwide. **Methods:** FIREFISH (NCT02913482) is a multicenter, open-label, two-part study of risdiplam in children with Type 1 spinal muscular atrophy (SMA) and two SMN2 gene copies (inclusion criteria: 1-7 months old at enrollment). In the dose-finding Part 1 of FIREFISH the safety, tolerability, and pharmacokinetics/pharmacodynamics of different risdiplam doses were assessed. The pivotal Part 2 assessed the safety and efficacy of risdiplam over 24 months at the dose selected from Part 1. Thereafter, children entered a 3-year open-label extension phase and continued to receive risdiplam at the pivotal dose. **Results:** The FIREFISH results after 4 years of risdiplam treatment showed that 91% of children were alive and 81% did not require permanent ventilation. Additionally, children either maintained or improved their motor skills in terms of developmental milestones and motor function, which is not observed in the natural history of Type 1 SMA. Pooled safety and efficacy data were available from 58 enrolled infants who completed risdiplam treatment over a period of 5 years (Part 1 high-dose cohort, n=17; Part 2, N=41). The last FIREFISH patient completed their final assessment after 5 years of risdiplam treatment at the pivotal dose on December 22, 2023. Here we present the final pooled safety and efficacy results from the FIREFISH study after 5 years of risdiplam treatment. **Conclusion:** FIREFISH Parts 1 and 2 provide long-term clinical trial safety and efficacy data on risdiplam in the treatment of Type 1 SMA. Acknowledgments: The authors would like to thank all the patients who participated in this study, their families, as well as the investigators and trial staff. This study was sponsored by F. Hoffmann-La Roche Ltd, Basel, Switzerland.

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First Brazilian study in central disorders of hypersomnolence using precision medicine: high-resolution hla sequencing and hypocretin levels in patients with narcolepsy and idiopathic hypersomnia

Christianne Martins Correa da Silva Bahia[1,2], Luis Cristovao Porto[1], Victoria Mendes Oliveira[1], Danielle Angst Secco[1], Romulo Vianna[1], Gilson Costa dos Santos Junior[1], Carlos Otavio Brandão[3], Emmanuel Mignot[4], Bruna de Freitas Dias[4], Soniza Vieira Alves Leon[2,5,6]

[1]Universidade do Estado do Rio de Janeiro, Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal Fluminense, Niterói RJ, Brazil.
[3]Neurolife, Rio de Janeiro RJ, Brazil.
[4]Standford University, California, USA.
[5]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[6]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Christianne Martins Corrêa da Silva Bahia; Email: christiannemartins@gmail.com.

Background: Narcolepsy and idiopathic hypersomnia (IH) are prominent central disorders of hypersomnolence. HLA-DQB1*06:02 is the primary genetic risk factor for narcolepsy, but other alleles have also been identified. However, data on these conditions in Brazil is limited, as previous studies have focused solely on this allele without exploring hypocretin measurement, now considered a biomarker for NT1. Ongoing research aims to find a biomarker for diagnosing narcolepsy type 2 (NT2) and IH. Therefore, genetic studies in diverse populations like Brazil are essential to provide valuable information on primary hypersomnias. **Objective:** To investigate the genetic characteristics associated with susceptibility or protection for narcolepsy and IH in the Brazilian population using precision medicine for diagnosing NT1 and NT2. **Methods:** Diagnosis of narcolepsy and IH was based on the third version of the International Classification of Sleep Disorders, with sub-classification into NT1 and NT2 determined by CSF hypocretin 1 concentration using radioimmunoassay. High- resolution HLA sequencing for class I (A, B, and C) and class II (DPB1, DRB1, and DQB1) alleles was conducted using massive parallel sequencing. The study compared data obtained with the National Registry of Bone Marrow Donors (REDOME) database, matching by sex, self-declared ethnicity, and geographic location in a ratio of 1 case to 5 controls. **Results:** The study included 45 narcolepsy patients and 225 controls, as well as 11 IH patients and 55 controls. Genetic differences, indicated by the Fixation Index, were observed between NT1 patients (n=17) and controls (n=85), as well as between NT1 and NT2 (n=16) groups, with significant p-values (p<0.001). NT1 patients exhibited a higher frequency of the HLA-DQB1*06:02 allele compared to controls and NT2 patients (88.24% vs. 23.53% vs. 18.75%, respectively; p<0.001). Following DQB1*06:02, DRB1*15:03 was identified as the most common allele associated with NT1, forming a haplotype with an odds ratio of 9.5 compared to controls (95% CI: 2.93-30.81, p<0.005). Rare haplotypes were also found in three NT1 cases. IH did not show significant genetic risk alleles compared to controls or narcolepsy groups after Bonferroni correction. There were no differences in hypocretin values between NT2 and IH groups. **Conclusion:** This first Brazilian study on primary hypersomnias using precision medicine revealed that HLA-DQB1*06:02 is the main genetic risk factor for NT1 but not for NT2 and IH. Notably, DRB1*15:03, rather than DRB1*15:01, was more prevalent among NT1 Brazilian patients, and this allele has been described in several studies as associated with African heritage. Rare haplotypes were found in some cases, suggesting they may be more frequent among Brazilians. In contrast to NT1, NT2 and IH patients did not exhibit significant genetic differences in HLA sequencing compared to their controls.

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Five-year experience in spinal muscular atrophy treatment with nusinersen: a single-center experience

Rodrigo Holanda Mendonça[1], Graziela Jorge Polido[1], Ciro Matsui Jr[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Graziela Jorge Polido; Email: grazielapolido@gmail.com.

Background: Spinal muscular atrophy (SMA) is a genetic motor neuron disease caused by mutations in the SMN1 (Survival Motor Neuron) gene, which leads to childhood-onset muscle weakness. Nusinersen, the first approved treatment for SMA, is an antisense oligonucleotide for intrathecal use, which leads to greater survival and gain in motor acquisitions. Studies on the safety and efficacy of long-term treatment are still scarce. **Objective:** To present long-term results (5 years of follow-up) in SMA patients under treatment with Nusinersen. **Methods:** We followed a total of 24 patients with SMA type 1 and 30 patients with SMA type 2 and 3. The patients were evaluated by the functional scales CHOP-INTEND (The Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders) and Hammersmith Functional Motor Scale Expanded (HFMSE), in relation to gain of motor milestones (head control, sitting with or without support, standing and walking) in addition to assessment of survival and use of continuous ventilation (CV) and feeding route. **Results:** Among SMA type 1 patients, after five years of follow-up, 22 (91.6%) patients are alive, two deaths occurred: one after gene therapy and the other after respiratory failure. Two patients received gene therapy but continued to use Nusinersen - combined therapy. Eight patients gained some motor milestone, all of them started treatment before 12 months disease. The greatest gains in CHOP-INTEND occurred up to 24 months of treatment, and after this period, the scores tended to stabilize, without further gains. Thirty patients with SMA types 2 and 3 were followed for five years. In these patients, five developed severe scoliosis, defined by a Cobb angle greater than 45°, and six underwent spinal arthrodesis, totaling eleven patients (36.6%) with clinically significant spinal deformity. These patients began to demand greater

complexity in the intrathecal application, requiring the use of imaging (Fluoroscopy or computed tomography) in subsequent applications. From a motor point of view, the HFMSE assessment showed an average gain of +2.1 (±6.9) and +1.3 (±7.1) points at 36 and 48 months of follow-up, respectively. In 48 months of treatment, 21 patients (70%) remained stable or continued to gain points in long-term follow-up, and nine patients (30%) lost 2 or more points on the scale, these developed severe scoliosis or underwent arthrodesis of column. **Conclusion:** Nusinersen showed continuous benefit over five years of treatment, bringing motor improvement mainly within the first 2 years of treatment and maintaining motor function acquired at 4 years. Among SMA type 1, only patients who started treatment before 12 months of illness gained some motor milestone. No new adverse events were reported in this long-term follow-up.

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Fluctuating scoliosis due paravertebral dopa-responsive dystonia

Caio César Alves Lins de Oliveira[1] , Lara Candido Pena[1], Alex de Novais Batista[1], Lisa Ananda Rodrigues Soares[1], Laura Luiza Barbosa Menezes da Mota[1], Leonardo Monteiro Lauria[1], Virgílio Sátyro da Nóbrega Segundo[1], Carlos Frederico Leite de Souza Lima[1]

[1]Hospital Universitário Oswaldo Cruz, Recife PE, Brazil.

Correspondence: Caio César Alves Lins de Oliveira; Email: caiolins97@gmail.com.

Case presentation: A 22-year-old male patient presented in the moviment disorders outpatients clinic of the Neurology service in March 2021 complaining of paravertebral muscles involuntary and sustained contractions throughout the day for approximately 7 months. He stated that this symptomatology resulted in the formation of scoliosis with a fluctuating pattern, being described as worse in the morning and lessening at the end of the day. On physical examination, was observed paravertebral muscles involuntary and sustained contraction, culminating in thoracolumbar scoliosis, other examination parameters was normal. Therefore, considering the age range and fluctuating nature of postural changes due paravertebral dystonia, Levodopa 100mg in association with benserazide 25mg (Prolopa BD) was prescribed 3 times a day, as a therapeutic test based on the main diagnostic hypotheses of the case. The patient returned to the consultation approximately 3months after starting the medication, reporting and showing substantial improvement of contractures in the dorsal region, in addition to a significant reduction in the degree of scoliosis with resolution of its fluctuating nature. He denied adverse effects from the use of medication. **Discussion:** Dopa-responsive dystonia (DRD) encompasses a clinically and genetically heterogeneous group of disorders that generally manifest as dystonia of appendicular onset, but which can also begin in the axial muscles, which fluctuates throughout the day and improves with the use of low-dose levodopa. Its origin is related to disorders in the dopamine synthesis pathway, mainly due to dysfunction or deficiency of the enzymes responsible for its biosynthesis, such as Guanosine Triphosphate Cyclo-Hydrolase 1 (GTP-CH-1), Tyrosine Hydroxylase (TH), Sepiapterin Reductase and Pyruvate Tetrahydrobiopterin Synthase. The diagnosis of DRD is generally based on the presentation of symptoms and therapeutic tests with levodopa, however the definitive diagnosis, with specific etiological determination, is made through genetic tests such as whole exome sequencing. Although the patient had a typical history and met the DRD diagnostic criteria, he was not subjected to genetic testing due to unavailability in the Brazilian Unified Health System (SUS). **Final comments:** DRD is a condition that, despite being rare, should be always be considered, given the age group affected, the degree of negative impact on patients' quality of life when left untreated and also due to the great symptomatological response to the use of Levodopa, which is a medication easily accessible through public government programs. Furthermore, a greater accessibility to genetic tests that enable definitive diagnosis is extremely important for genetic counseling of patients and their families, and also for a better understanding of the different clinical-epidemiological manifestations of DRD.

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Focal impaired awareness seizures in patient with choroidal fissure cyst

Wesley Alfredo Gawlinski de Arruda[1], Thaise Fellini Dal Moro[1], Estevão Daniel Wohlenberg[2]

[1]Hospital São Vicente de Paulo, Passo Fundo RS, Brazil.
[2]Universidade Federal da Fronteira Sul, Passo Fundo RS, Brazil.

Correspondence: Wesley Alfredo Gawlinski de Arruda; Email: wesley.alfredo@hotmail.com.

Case presentation: A 71-year-old female patient with no previous history of neurological events began experiencing mental confusion and cognitive decline a few months ago, with episodes of "blackouts" starting to occur approximately 8 days ago. According to family members, she would become apathetic, unresponsive, and unable to communicate, not improving when called during a regular conversation or while performing daily activities, lasting for a few seconds but sometimes up to a minute. Neurological examination showed no abnormalities. Cognitive screening revealed significant impairment (Semantic Verbal Fluency Test (SVFT) with 5 animals in 1 minute, Sunderland Clock Test (SCT) 2/10, Mini-Mental State Examination (MMSE) score of 21 points), but no functional loss according to family members (Pfeffer = 0 points). Brain Magnetic Resonance Imaging (MRI) identified a right-sided choroidal fissure cyst in the temporal lobe measuring 1.3 x 0.9 cm. Electroencephalogram (EEG) with normal baseline rhythm showed interictal spike discharges in the right temporal region, with an electrographic seizure (with clinical manifestation) beginning in the ipsilateral temporal region, lasting approximately 2 minutes. Therefore, due to the hypothesis of epilepsy with focal impaired awareness seizures secondary to choroidal fissure cyst, treatment with an antiepileptic drug (Levetiracetam 250 mg BID) was initiated. About two months later, the patient returned for reassessment, showing substantial cognitive improvement (SVFT = 13 animals in 1 minute, SCT 9/10, MMSE 24 points), with the patient and family denying any new episodes of "blackouts" or seizures, and being well-adapted to the medication. **Discussion:** The choroidal (or choroid) fissure is a cleft in the lateral ventricle between the fornix and thalamus, which can develop cysts due to errors during embryonic choroid plexus formation. Although small cysts are common and often asymptomatic, some studies suggest a connection between these cysts and epilepsy, but their causal relationship is still controversial. Neuroimaging (especially magnetic resonance imaging) is extremely useful for differentiating choroidal fissure cysts from other differential diagnoses, including cystic neoplasms, dermoid/epidermoid cysts, and choroid fissure enlargement due to focal temporal lobe atrophy. If a patient presents with clinical symptoms suggestive of epilepsy, with electroencephalogram (EEG) findings compatible with a focal disorder, seizures can be controlled with the use of antiepileptic drugs. **Final comments:** Choroidal fissure cyst is a brain lesion known to be predominantly asymptomatic. However, as in the case of our patient, secondary epilepsy due to a temporal lobe lesion may occur. The correlation between focal epilepsy and cysts in the choroidal fissure is present, but a causal relationship is not yet well established, highlighting the need for further studies on the topic.

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Foot drop as the predominant symptom in facial onset sensory and motor neuronopathy (FOSMN)

Izadora Vieira Bolzani Lopes Lima[1], Aldrin Pedroza Martins[1], Nadson Bruno Serra Santos[1], Diego Silva Figueiredo[1], Fabricio Diniz de Lima[1], Alberto Rolim Muro Martinez[1], Anamarli Nucci[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Izadora Vieira Bolzani Lopes Lima; Email: izadoralima7@gmail.com.

Case presentation: A 59-year-old woman with no past medical history, presented in 2019 signs of right-side tongue paresthesia. Later in the same year, she developed difficulties using high heels because her shoes slipped off her feet. By 2021, she started using a unilateral stick on the right side due to frequent falls. In 2022 the patient noted dysarthria and dysphagia, searching for medical assistance. Later in 2023, she was referred to our hospital for further investigations. At the first consultation her neurological examination revealed bilateral loss of touch and pinprick sensation affecting all trigeminal branches and bilateral facial paresis with a prominent wrinkle between the mouth and the chin, bilaterally. There was also mild dysarthria and tongue atrophy specially on the left side with fasciculations and marked atrophy of both sternocleidomastoids, with difficult to hold the head in the supine position. There was weakness graded IV(according MRC) in her deltoids, triceps, iliopsoas, quadriceps femoris and hamstrings muscles. Both tibialis anterior and triceps sural were grade III. She could not walk on tip toes or on the heels. The sensory examination revealed numbness of the upper limbs. Deep tendon reflexes were all absent. There was no auditory disfunction. Brain magnetic resonance imaging (MRI) revealed mild nonspecific gliosis. The CSF was normal. Neurophysiology examination exposed a preganglionic, chronic and asymmetrical motor neuronopathy of bulbar, cervical, thoracic and lumbar segments, with signs of acute denervation. There was also multifocal, not length-dependent neuropathic sensory involvement of moderate intensity, compatible with a sensory neuronopathy. These concomitant patterns in association with prolonged R2 latency on blink reflex were highly suggestive of FOSMN. The patient was then started on Riluzole 50mg twice a day therapy with relative slowdown of progression of her disease. **Discussion:** FOSMN is a rare disease caracterized by asymmetrical facial and perioral numbness and paresthesia with reduced touch and pain sensation in the trigeminal distribution and progression of sensory symptoms to the upper trunk. Lower motor neuron signs are noted by muscle weakness and wasting, especially in the face, neck and upper limbs. Most references consider FOSMN a TDP-43 proteinopathy within the amyotrophic lateral sclerosis-FTD spectrum. Diagnosis is established based on clinical signs and symptoms with exclusion of similar conditions and the absence of the blink reflex on EMG in addition to denervation. **Final comments:** Clinicians must be aware of of the doble neuronopathy of FOSMN and, that the interval between the onset of facial sensory signs and weakness can range from years to simultaneous onset and patients tend to neglect pure sensory symptoms, delaying diagnosis. Treatment is still controversial as some sources report little or no benefit from immunotherapies or riluzole and more research is needed for this purpose.

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Foramen ovale electrodes can identify a focal seizure onset when surface EEG fails in mesial temporal lobe epilepsy?

Izabela Dayany França Feitosa[1], Marina Teixeira Ramalho Pereira Dalio[1], Frederico Nakane Nakano[1], Alexandre Veriano Junior[1], Americo Ceiki Sakamoto[1], Tonicarlo Rodrigues Velasco[1]

[1]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Izabela Dayany França Feitosa; Email: draizabelafeitosa@gmail.com.

Temporal lobe epilepsy (TLE) is the most common form of focal epilepsy in adults, with mesial temporal lobe epilepsy associated with hippocampal sclerosis (MTLE-HS) is the commonest subtype. Approximately 30% of MTLE patients are pharmacoresistant (CHOI, 2008), half of them are candidates for temporal lobe resection (ROSENOW, 2001). For indication of surgery, is necessary the accurate delineation of the epileptogenic zone, which can be defined from the analysis of the seizure, interictal and ictal electroencephalogram activity, analysis of highresolution MRI (KILPATRICK, 1997). However, the surface EEG fail to demonstrate an unambiguous unilateral ictal onset, semi-invasive studies, with foramen ovale electrodes (FOE), and/or invasive studies with depth electrodes (stereotactic depth electrode implantation or subdural grids/strips) is necessary (DIEHL, 2000). In the present study, we analyze the data of 161 patients with MTLE undergoing evaluation with foramen ovale electrodes. The evaluation with FOE was performed when the surface EEG presented: (a) bitemporal independent seizures, (b) non lateralized ictal onsets, (c) side contralateral to the side of hippocampal sclerosis. When the FOE study was also unable to define an unequivocal ictal onset, an evaluation with invasive electrodes was proposed. EFO was effective in 91 cases (56.5%). Of these 78 patients undergo a temporal lobectomy surgery with 73.1% of patients who are free of crises. Twenty-one patients were evaluated with invasive electrodes after FOE study. The invasive electrode confirmed the results of FOE in 16 (76.2%) cases. The study with invasive electrodes guided a surgical procedure in 7 patients and temporal lobectomy was performed, making 04 patients (57.1%) free of seizures. The evaluation with foramen ovale electrodes is cost-effective, well tolerated by patients and has a lower rate of complications when compared to other methods. Therefore, we conclude that EFO evaluation, after nonconcordant surface EEG, can be a safe alternative, more economical when compared to the implantation of invasive electrodes and with an excellent postoperative outcome in a patient with MTLE.

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Foreign accent syndrome in a patient with stroke at pons: case report

Danielli Regina da Silva[1], Airton Sena Oliveira Sales[1], Laisy Bárbara Souza Silva[1], Amanda Soares Pimenta[1], Daniela Aparecida Lima Viana[1], João Henrique da Matta Clementino[1], Ana Luisa Rufino de Sousa[1], Elisangela Ferraz Pazinato[1], Giovani Zago Borges[1], Danielle Patrícia Borges Margato[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Amanda Soares Pimenta; Email: amandaspiment@gmail.com.

Case presentation: Female, 51 years old, Brazilian, previously type 1 diabetic, insulin-requiring, in addition to dyslipidemia, heart disease and hypothyroidism, admitted to a teaching hospital in a city located in the Triângulo Mineiro region, in 2015, with a history of speech and language alterations for a week, associated with hypoglycemia on admission. Dysarthria and/or aphasia were not identified, with writing and language comprehension preserved, however the patient had divergent fluency in relation to the native language. At the time, the patient denied changes in behavior, strength, sensitivity, balance, sleep and wakefulness, headache and/or other neurological signs and symptoms. An investigation was carried out for a possible cerebrovascular event during hospitalization, and a lesion with an ischemic appearance on the leftside of the pons was then identified in an imaging study, identified on a T2-weighted sequence. Reevaluated in May 2024, with the speech alteration maintained. **Discussion:** Foreign accent syndrome (FAS), considered a rare speech disorder, is characterized by the emergence of an accent perceived as "foreign" due to the combination of segmental deficits, such as phonetic distortions and phonemic paraphasias, associated with suprasegmental changes with accentuation, variation of tone and rhythm. Stroke is the most common etiology, generally with left frontal brain lesions, which differs from the reported case. However, other etiologies are found, such as trauma, metastatic breast carcinoma, multiple sclerosis and migraines. The pathological lesions have a wide and heterogeneous anatomical location, which include the left basal ganglia, left corona radiata and internal capsule, right precentral and postcentral gyri, right middle cerebral artery region, right frontal area, brainstem and cerebellum. Thus, it is believed that in FAS there is interruption of widely distributed speech networks and there is not just one anatomical location affected. However, it is still a poorly understood and studied syndrome, given its rarity and complexity. **Final comments:** This report reinforces the complexity of FAS, highlighting the interruption of speech networks widely distributed in the brain, which makes it difficult to identify a precise anatomical location as the sole cause. Current understanding is limited due to its rarity and etiological diversity, requiring further studies to elucidate its underlying mechanisms and optimize therapeutic approaches. In short, this patient's case highlights the importance of meticulous investigation in cases of unusual speech disorders, especially in complex clinical contexts such as the one presented, in order to guide clinical management and provide better results for patients affected by this unusual syndrome.

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Frequency and characterization of facial pain in individuals with hemifacial spasm in an outpatient clinic of a public tertiary center

Mariana de Almeida Cruz Silva[1], Míriam Carvalho Soares[2], João Augusto de Macedo Cavalcanti de Albuquerque[1], Clélia Maria Ribeiro Franco[2], Pedro Augusto Sampaio Rocha Filho[1]

[1]Universidade Federal de Pernambuco, Centro de Ciências Médicas, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Hospital das Clínicas, Serviço de Neurologia, Recife PE, Brazil.

Correspondence: Mariana de Almeida Cruz Silva; Email: mariana.almeida20021@gmail.com.

Background: Hemifacial spasm (HFS) is a movement disorder marked by involuntary facial muscle contractions, often due to a nearby blood vessel pressing on the facial nerve. Typically, HFS onset occurs between the fourth and sixth decades of life and has an annual incidence of 0.78 per 100,000, being considered a rare disorder. Botulinum toxin type A (BoNT-A) stands as the gold-standard therapy in the treatment of HFS. The presence of facial pain associated with HFS is commonly observed in clinical practice, although there is still little research on the subject. **Objective:** To characterize facial pain patterns in patients with HFS by evaluating the location, frequency, intensity, and duration of pain, as well as its relation to BoNT-A injections. **Methods:** A cross-sectional study was conducted with 53 HFS patients (F: 35, M: 18) at a public tertiary hospital over 6 months. The average age was 66.1 years (SD 12.2). Patients with cognitive impairment, BoNT-A use in the last three months, and other movement disorders were excluded. Data were collected using sociodemographic and structured facial pain questionnaires, the McGill Pain Questionnaire, and the HFS-Score for quality of life. Statistical analyses were performed using Epi Info software. **Results:** Among the 53 patients, 23 (43%) reported facial pain. The most common location was the periorbital region in 17 cases (65%), followed by maxillary region in 4 patients (15.3%), mandibular in 2 patients (7.6%), and auricular, whole hemiface and bilateral palpebra in 1 case (3.8%) each. All patients described pain ipsilaterally to the spasm side, and 1 patient described pain involving palpebral areas bilaterally. Spasms were present on the left side of the face in 33 (62%) patients. The average pain intensity was 6.1 on the Visual Analog Scale (VAS) (SD 2.1), with a mean frequency of 8.6 (SD 7.7) days/month and 2.3 (SD 2.3) episodes/day, lasting an average of 9.6 hours (SD 10). The average time to pain onset after HFS motor symptoms was 9.9 years (SD 9). Twenty patients reported that previous BoNT-A applications improved facial pain, while the remaining three reported no change. No patient reported a perception of worsening pain after BoNT-A injections. Although facial pain was frequently reported by the patients, no statistically significant difference was found between the severity of facial pain and the quality of life among individuals with HFS (p value 0,1993). The most common descriptors for the facial pain were: pulling (11 patients), itchy (10 patients), annoying (10 patients), miserable (9 patients), pinching (9 patients) **Conclusion:** This study highlights the need to investigate and treat facial pain in patients with hemifacial spasm, a rare and debilitating condition. Analyzing pain patterns and the efficacy of botulinum toxin treatment underscores the importance of tailored therapeutic approaches and guides future research.

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Frequency of GAA-FGF14 ataxia in a large cohort of brazilian patients with unsolved adult-onset cerebellar ataxia

Luiz Eduardo Novis[1], Rodrigo Siqueira Soares Frezatti[2], David Pellerin[3], Marcondes Cavalcante França Jr.[4] , Shahryar Alavi[5], Mariana Spitz[6], José Luiz Pedroso[7], Orlando Graziani Povoa Barsotini[7], Wilson Marques Jr.[2], Helio Teive[1]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Programa de Pós-Graduação em Medicina Interna e Ciências da Saúde, Curitiba PR, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Departamento de Neurologia, Ribeirão Preto SP, Brazil.
[3]McGill University, Montreal Neurological Hospital and Institute, Departments of Neurology and Neurosurgery, Montreal, Canada.
[4]Universidade Estadual de Campinas, Campinas SP, Brazil.
[5]University College London, Queen Square Institute of Neurology and the National Hospital for Neurology and Neurosurgery, Department of Neuromuscular Diseases, London, UK.
[6]Universidade Estadual do Rio de Janeiro, Departamento de Especialidades Médicas, Serviço de Neurologia, Rio de Janeiro RJ, Brazil.
[7]Universidade Federal de São Paulo, Departamento de Neurologia, Unidade de Ataxias, São Paulo, SP, Brazil.

Correspondence: Luiz Eduardo Novis de Farias ; Email: luizeduardonovis@hotmail.com

Background: Dominantly inherited GAA repeat expansions in intron 1 of the fibroblast growth factor 14 (FGF14) gene have recently been found to be a common cause of hereditary adult-onset cerebellar ataxia (GAA-FGF14 ataxia; spinocerebellar ataxia 27B). The global epidemiology and regional prevalence of this newly reported disorder remain to be established. **Objective:** We here investigated the frequency of GAA-FGF14 ataxia in a large

cohort of Brazilian patients with unsolved adult-onset cerebellar ataxia. **Methods:** We recruited 93 index patients with genetically unsolved adult-onset cerebellar ataxia despite extensive genetic investigation and genotyped the FGF14 repeat locus. Patients were recruited across 4 different regions of Brazil. **Results:** Of the 93 index patients, 8 (9%) carried an FGF14 (GAA)≥250 expansion. The expansion was also identified in one affected relative. Seven patients were of European descent, one was of admixed American ancestry, and one was of African descent. One patient carrying a (GAA)376 expansion developed ataxia at age 28 years, confirming that GAA-FGF14 ataxia can occur before the age of 30 years. One patient displayed episodic symptoms, while none had downbeat nystagmus. Cerebellar atrophy was observed on brain MRI in 7 of 8 patients (87%). **Conclusion:** Our results suggest that GAA-FGF14 ataxia may be a common genetic cause of adult-onset cerebellar ataxia in the Brazilian population, although larger studies are needed to fully define its epidemiology.

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From independent to incapacitated: a case of antiepileptic drug-induced rapidly progressive dementia

Guilherme Lago Miranda[1], Amanda Freitas Alves[1], Ingrid Lorena da Silva Gomes[1], Matheus Reis Rosa[1], Giovanni Caniçali Simões[1], Henrique de Moraes Bernal[1], Roberta Arb Saba Rodrigues Pinto[1], Marcele Schettini de Almeida[1], Herval Ribeiro Soares Neto[1], Camila Hobi Moreira[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Guilherme Lago Miranda; Email: lagomiranda.g@gmail.com.

Case presentation: A 76-year-old woman, previously independent in instrumental activities of daily living, diagnosed with epilepsy at age 12, was taking irregularly phenytoin 200mg/day and phenobarbital 200mg/day. Approximately 4 months prior, her family assumed the task of medication intake, so she started to use it as prescribed. In two months, she developed progressive cognitive decline, difficulty walking and insomnia. She was admitted to the emergency room due to a significant worsening of her condition over the past 2 weeks, with a reduced level of consciousness, verbalizing only incomprehensible sounds and unable to eat. On neurological examination, the patient was torporous, showed no motor response to pain, and showed neither pyramidal signs nor involuntary movements. Faced with the hypothesis of rapidly progressive dementia (RPD), extensive further investigation took place. Her cerebrospinal fluid showed the presence of 1 cell, 53 proteins and 67 glucose. Brain MRI had no abnormalities, and her EEG showed an excess of diffuse beta activity. Neoplastic investigation and a serum onconeural panel were unremarkable. Phenobarbital serum concentration was above 80 (reference value 15-40). Therefore, the medication was withdrawn without tapering. Phenytoin was on normal range. A short course of high-dose steroid therapy was also performed for the possibility of immune-mediated encephalitis, despite non-suggestive complementary tests. The patient started to gradually improve, being discharged able to walk with assistance and on oral feeding, and without immunosuppressants. At the most recent outpatient follow-up, 8 months later, her status was restored, with good seizure control with lamotrigine, and her phenobarbital serum concentration was zero. **Discussion:** Drug-induced reversible dementia can occur in the context of excessive use, polypharmacy and side effects. The antiepileptic drugs most commonly associated with this condition are phenytoin and valproic acid, but there have been reports in the literature of RPD in patients using phenobarbital in combination with lithium and levomepromazine. Phenobarbital is a barbiturate that enhances the GABAergic effect by prolonging the opening of chloride channels. Serum levels above 50 to 60ug/ml can lead to progressive neurological dysfunction and decreased level of consciousness leading up to coma. The interaction between the two first-generation antiepileptics she was on might have caused the intoxication. As suggested in the literature, we also initiated empirical treatment with corticosteroids due to the severity of her condition, but no other immunosuppressive treatment was maintained, and the patient continued to show progressive improvement with the discontinuation of the anticonvulsant and gradual decrease in serum medication levels. **Final comments:** Toxic-metabolic and iatrogenic RPD etiologies should always be thoroughly investigated and ruled out, given their potential reversibility.

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From reversible cerebral vasoconstriction syndrome to subarachnoid hemorrhage, a continuum presentation of the same pathology: a case report associated analgesic abuse in kidney transplant recipient

Renato Dumbá Monteiro de Castro[1], Amanda Fernandes Klajn[1], Sacha Allebrandt da Silva Ries[1], Taís Luise Denicol[1], Alexandre Laisson Prado Taschetto[1], Carolina Matté Dagostini[1], Juan Sebastián Sánchez León[1], Pedro Hall Ruschel[1], Cândida Driemeyer[1], Liselotte Menke Barea[1]

[1]Santa Casa de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Renato Dumbá Monteiro de Castro; Email: renato.castro@ufcspa.edu.br.

Case presentation: Female patient, 61 years old, admitted to the emergency department reporting the onset of new pulsating headache with severe intensidade (visual analog scale 9/10) 5 days ago, bilateral, parietooccipital region, partial relief after use of analgesics, lasts more than 24 hours with recurrence and increasing intensity when taking a shower, accompanied by nausea and dizziness at the peak of pain without visual changes. Previously diabetic and kidney transplant 17 years ago by polycystic kidneys. In regular use of metformin, 1000mg/day, sirolimus 2mg/dia (macrocyclic lactone class immunosuppressant), predinisone 10mg/day, Gabapentina 300mg/day for facial postherpetic neuralgia and intermittent tramadol 100mg 4 to 6 times a week for 4 months due to ankle tendinopathy. Secondary headaches were investigated and the initial tomography angiography showed focal areas of luminal narrowing in bilateral middle cerebral arteries, M2/M3, and posterior cerebral arteries, P2/P3, suggestive of vasoconstriction. No aneurysmal formations or hematological content were identified. Laboratory tests were normal and liquor analysis with hematic macroscopic appearance identified an increasing red blood cells (RBC) in two consecutive samples, ruling out puncture accident. The first collection showed bottles 1, 2 e 3 with erythrocyte count of 4480, 5800 and 6400 , respectively. Second confirmatory sample in bottles 1 and 2 with RBC of 4800 and 11200. Given the suspicion, potential drug causes were suspended and replacement of the immunosuppressive class. Administered vasospasm preventive drugs , such as nimodipine 360mg/day. Complementary investigation magnetic resonance of the vessel wall without parietal enhancement reinforced the suspected reversible cerebral vasoconstriction syndrome (RCVS). After 10 days followed by neurological surveillance in the intensive care unit and transcranial echodoppler monitoring, she presented a focal neurological deficit with paresis of the right upper limb. A new control with digital subtraction angiography ruled out microaneurysms and intracranial milrinone was administered to endovascular hoping to control vasospasms, in addition to clinical drug support and euvolemia maintained for 21 days, according to the best recommendations in current medical literature still scarce. **Discussion:** RCVS is widely described in lists of association with specific drug induction. However, complications such as SAH, vasospasm and pathophysiological progression still require details so that prevention and therapeutic procedures can be protocoled with quality scientific evidence. **Final comments:** Cases of headache suggestive of a secondary cause and clinical presentations with their red flags are frequent complaints in neurological care. Recognizing the main etiologies can prevent complications. Therefore, it is important to disseminate presentations and less common pathological progress among health care professionals and scientists.

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Frontotemporal dementia (FTD) and amyotrophic lateral sclerosis (ALS): is clinical diagnosis sufficient? A case report

Wellington Costa da Silva Filho[1], Helton Benevides Santana de Oliveira[1], Renê Vasconcelos Silva[1], Gabriel Ribeiro Andrade Neto[1], Myllena Maria Santos Santana[1], Jorge Dornellys da Silva Lapa[1], Flávia Barreto Garcez Carvalho[1], Roberto César Pereira Prado[1]

[1]Universidade Federal de Sergipe, Aracaju SE, Brazil.

Correspondence: Helton Benevides Santana de Oliveira; Email: heltonbenevides@vidaampla.com.br.

Case presentation: An 84-year-old hypertensive patient began experiencing dysarthria and progressive dysphagia in 2020. One year later, notable behavioral changes emerged, characterized by aggressiveness, agitation, and disinhibition. These behaviors included repetitive folding and unfolding of clothes, public lifting of her skirt, and attempts to attack family members with a weapon. Notably, these symptoms manifested without significant memory impairment. Neurological examination revealed motor aphasia, paralysis of the orbicularis oris and right eye muscles, jaw drop, atrophic tongue with fasciculations and immobility, and reduced muscle strength (grade 4+) in the right hemibody and left lower limb, and grade 4 in the left upper limb. Fasciculations were observed in the left deltoid and right quadriceps. Deep tendon reflexes were hyperactive, and there was a questionable plantar response on the right. Electromyography indicated axonal degeneration with chronic reinnervation signs in the C5, C6, C7, C8, T1, L4, L5, and S1 myotomes bilaterally, active denervation in the tongue muscles and left C8/T1 myotomes, and no sensory abnormalities. **Discussion:** Motor neuron disease may precede or follow the onset of FTD, often presenting as the behavioral variant. In patients with classical amyotrophic lateral sclerosis (ALS), characterized by neuromuscular symptoms, 15 to 20% also exhibit cognitive and behavioral symptoms consistent with FTD, particularly the behavioral subtype. These patients frequently present with bulbar symptoms more prominently than those with classical ALS. A hexanucleotide repeat expansion in the C9orf72 gene is the most commonly identified genetic cause of the combined FTD and ALS presentation, accounting for approximately 5-10% of sporadic ALS cases. Other implicated genes include the autosomal dominant FUS gene, responsible for about 2% of sporadic cases, and the rarely recessive TARDBP gene. In this case, the diagnosis of ALS is confirmed by electromyography according to the El Escorial criteria. The 2011 criteria of the International Behavioral Variant FTD Criteria Consortium (FTDC) suggest possible frontotemporal dementia. **Final comments:** While most patients with FTD do not initially exhibit significant motor symptoms, these can precede the onset of dementia and may result from frontotemporal lobar degeneration. Patients with concurrent FTD and ALS have a shorter survival than those with isolated FTD. Given the strong association of FTD-ALS with the C9orf72 gene expansion, alongside other known genetic factors, genetic testing is recommended but not mandatory for diagnosis.

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Functional connectivity of brain networks in patients with mild cognitive impairment after 24 weeks of resistance training

Gabriela Barbosa Rodrigues[1], Isadora Cristina Ribeiro[1], Brunno Machado de Campos[2], Thamires Naela Cardoso Magalhães[3], Rafael Brandes Lourenço[2], Camila Vieira de Ligo Teixeira[4], Italo Karmann Aventurato[1], Marjorie Cristina Rocha da Silva[1], Fernando Cendes[2], Marcio Luiz Figueredo Balthazar[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Laboratório de Imagens e Biomarcadores em Doenças Cognitivas, Campinas SP, Brazil.
[2]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Laboratório de Neuroimagem, Campinas SP, Brazil.
[3]Texas A&M University, Brain Sciences Institute for Neuroscience, Department of Psychological, Texas, USA.
[4]West Virginia University, Neuroscience Institute, West Virginia, USA.

Correspondence: Gabriela Barbosa Rodrigues; Email: g242237@dac.unicamp.br.

Background: Mild cognitive impairment (MCI) is characterized by a decline in multiple cognitive domains without significant impairment in daily living activities. The pathophysiology of MCI involves abnormal brain network connectivity patterns, particularly in fronto-medial regions responsible for memory processing, attention, and linguistic and visuospatial comprehension. These include the auditory, language, sensorimotor, vision, visuospatial, and basal ganglia networks. While physical exercise has been shown to slow the progression of MCI by enhancing brain connectivity, the specific impact of resistance training (RT) on MCI remains a relatively unexplored area of research. **Objective:** To investigate the impact of 24 weeks of RT in patients diagnosed with MCI on the functional brain connectivity (FC) of the auditory, language, sensorimotor, vision, visuospatial, and basal ganglia networks. **Methods:** 37 older adults diagnosed with MCI (NIA and Alzheimer's Association 2018 criteria) were selected and divided into a training group (N=19) and a control group (N=18). The training group performed a 24-week RT protocol based on exercises for large muscle groups. All subjects underwent Magnetic Resonance in an Achieva 3T PHILIPS. FC data from the auditory, language, sensorimotor, vision, visuospatial, and basal ganglia networks were evaluated before and after the intervention using seed-based resting-state functional MRI (fMRI) methods (UF2C toolbox running on MATLAB software). Statistical analysis was conducted using a paired t-test, considering all the ROIs evaluated. Significance was set at $p<0.01$, uncorrected for multiple comparisons. **Results:** Our analysis of the training and control groups revealed increased connectivity in the visuospatial network (T-score> 2.7195, $p<0.01$, uncorrected) in the training group. No differences in FC were found in the control group. These findings indicate a potential positive effect of RT on the functional connectivity of the visuospatial network in MCI patients. **Conclusion:** Our findings suggest that RT might have improved the functional connectivity of the visuospatial network, which may help enhance visuospatial skills in patients at risk for dementia. More clinical studies of longer duration are required to determine its role in controlling symptoms and progression to the dementia stage and to search for other affected areas.

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Functional recovery following resections of focal cortical dysplasia for epilepsy in the primary motor cortex (M1)

Eliseu Paglioli[1], Rafael Paglioli[1], William Martins[1], Fernanda Schuh Martins[1], Vicenzo Zarpellon[1], Thomas Frigeri[1], Victoria Bizzi Schvartzman[1], Natalie Donida[1], Bibiana Thome[1], André Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Victoria Bizzi Schvartzman; Email: bizzivictoria@gmail.com.

Background: Focal cortical dysplasias (FCD) occasionaly localize to functionally relevant regions in the primary motor cortex (M1), and resections inevitably lead to motor deficits, with variable degrees of recovery. **Objective:** The available literature reports functional outcomes in groups of patients, but lacks precision when these data must be applied to counselling individual patients. **Methods:** In this study we present the motor evolution following focal resections in the hand (#1) and foot (#2) areas of M1 over several weeks in 2 patients with FCD and daily (often prolonged or recurring) seizures. Patients and methods: A 30yo woman (#1) and a 13 yo boy (#2) with small FCDs in the M1 hand and foot area, respectively, underwent lesional resection, preserving the vascular supply of the underlying white matter. Examinations of sensori-motor functions were performed daily in the first post-operative week. Subsequently, patients were evaluated through simple tests at home with video documentation. Both are on intensive programs of physical therapy. **Results:** Preoperatively, both had some degree of weakness, that fluctuated according to seizure frequency. Following resections, both were fully plegic of the hand or foot for 3 days, yet after one week patient #1 could performed discrete global finger flexion, followed by exponential recovery in the following weeks. Six weeks later she could perform flexion of the wrist and is quickly improving on fine finger movements and extension of the wrist. The boy (#2) began to walk without assistance on post-op day 7-, and 3-weeks following surgery was with better functionality than previous to surgery, as seizures stopped and there were no further functional worsening thus related. Both had complete seizure control. **Conclusion:** Focal resections of FCD in M1 may control intensely refractory seizures with expressive functional recovery over weeks, following a transitory period of focal plegia. Crucially, the underlying white matter must be carefully preserved.

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Functional/dissociative seizures in the pediatric/hebiatric population: videoelectroencephalography results from Santa Casa de Belo Horizonte

Vivian Proença Lara[1], Glaucia Monteiro Oliveira[1], Igor Pereira Matos de Oliveira[1], Caio Cobucci Esteves[1], Ana Luíza Xavier Drumond[1], Luiza Barbosa de Oliveira[1], Vanessa Alves Lobato[1], Aline Curcio de Moraes[1], Helena Barbosa Horta Santos[1], Maria Do Carmo de Vasconcellos Santos[1]

[1]Santa Casa de Belo Horizonte, Belo Horizonte MG, Brazil.

Correspondence: Vivian Proença Lara; Email: vi.prlara@gmail.com.

Background: Functional/dissociative seizures (FDS) are episodes that may resemble epileptic seizures but lack abnormal brain electrical activity. They have been described mainly in adults and less frequently in the pediatric/hebiatric population. Clinical diagnosis may be difficult, even for specialists. The gold standard is videoelectroencephalography (VEEG), not avaliable in all centers. This exam is able to distinguish between FDS and epileptic events and target proper treatment. **Objective:** To provide a descriptive analysis of clinical characteristics (age, sex, concomitant epilepsy, use of antiseizure/psychotropic medication and semiology) of patients under 20 years old with FDS from our VEEG unit and to compare the results with literature. **Methods:** Medical and VEEG records of patients aged 0-19 years (defined as child and adolescent by World Health Organization) were analyzed. All of them had at least one functional event recorded during monitoring between january/2020 and may/2024 at Santa Casa de Belo Horizonte. Epilepsy was defined by abnormal eletroencephalography or structural lesions in brain image exams. **Results:** 296 VEEG were performed on children and adolescents during the period mentioned in our service. Of these 19 presented FDS during the monitoring (6.4% of the total) which is in accordance with prevalence of 3.5% to 20% already described. The average age was 16 years (higher than in the population without FDS, whose average was 10). Furthermore 78.9% were female (compared to 44.4% in those without functional events). Both information are in agreement with the literature that shows higher frequencies of FDS in adolescents (compared to children) and in females. 63.2% didn't have concomitant documented epilepsy; in this subgroup 83.3% was using antiseizure medication due to the fact that they were diagnosed on an outpatient basis as epileptic before the VEEG investigation, showing frequent diagnostic difficulties. 58.3% of the group without epilepsy was using psychotropic drugs. We didn't know if they were following a psychological treatment. In relation to the semiology of FDS, 78.9% showed non-synchronized movements in the video, 57.9% forced eye closure, 52.6% immediate return to the baseline state, 42.1% behavioral arrest, 42.1% side-to-side movement of the head, 15.8% arched lumbar position and 10.5% seizures lasting more than 2 minutes. These signs are usually described as having high specifity for FDS. One of the children presented the "teddy bear sign" which is controversial in literature. **Conclusion:** This work is in line with previous studies that shows FDS are relatively common events, even in the pediatric/hebiatric population and that they are many times misdiagnosed. The accuracy of assessing these events early is essential for adequate clinical management of patients. Also expertise for physicians who diagnose and manage these disorders in children and adolescents is needed.

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Functionality and disability according to whodas 2.0 in people with stroke: a cross-sectional study

Paloma Rodrigues da Silva[1], Daniel de Sá Horácio Pereira[1], Rebeka Amanda Dias[1], Júlia Teixeira Magalhães[1], Leonardo Barcellos[1], Maria Elisa Pimentel Piemonte[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Paloma Rodrigues da Silva; Email: paloma.rsilva@usp.br.

Background: The International Classification of Functioning, Disability, and Health (ICF) describes functionality as the positive aspects of the interaction between an individual (with a health condition) and their contextual factors (environmental and personal). It considers functionality as the third health indicator, complementing previous indicators of morbidity and mortality. The World Health Organization Disability Assessment Schedule 2.0 (WHODAS 2.0) is a universal assessment tool created by WHO, based on the ICF, to provide a standardized method for measuring the impact of any health condition on functionality and disability across different cultures. A stroke (cerebrovascular accident) can cause numerous changes across various dimensions of an individual's functionality. **Objective:** To characterize the changes in functionality according to WHODAS 2.0 in individuals who experienced a stroke. **Methods:** A cross-sectional study conducted according to STROBE guidelines, with 37 participants (20 men)

with a mean age of 67.67±13.7, diagnosed with ischemic or hemorrhagic stroke. Inclusion criteria were: (1) age 18 years or older; (2) diagnosed with ischemic or hemorrhagic stroke based on imaging; and (3) patient or guardian capable of providing informed consent and completing the questionnaire. Exclusion criteria were: (1) history of stroke more than one year ago or less than six months ago; and (2) presence of other neurological conditions such as Parkinson's or Alzheimer's disease. Participants were assessed in person by trained physiotherapists using WHODAS 2.0 to evaluate functionality and disability levels. **Results:** The WHODAS 2.0 total scores were 71.62±27.2. Total scores were not correlated with age, schooling, or type of stroke. The most affected functionality dimensions were life activities related to household (51.75%), mobility (48.75%), and participation (47.97%). Only 8 participants returned to work activities and obtained scores for this dimension (44.79%). **Conclusion:** These findings have important implications for patient care and management. Characterizing the different affected dimensions of functionality should be considered when conducting and adapting pharmacological and nonpharmacological interventions for people with chronic sequelae of stroke accordingly.

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Fungal encephalitis by Kluyveromyces marxianus (Candida kefyr) associated with pyelonephritis in an immunocompetent patient

Kauan Alves Sousa Madruga[1], João Felipe Guimarães Santos[1], Luis Paulo Machado Scalise[1], Fernanda Alcântara Nascimento Aguiar[1], Valdecir Boeno Spenazato Júnior[1]

[1]Santa Casa de Misericórdia de Barretos, Barretos SP, Brazil.

Correspondence: Kauan Alves Sousa Madruga; Email: kauanmbr@gmail.com.

Case presentation: This case report details a 37-year-old immunocompetent woman from Barretos, SP, previously controlled for hypertension, who presented with refractory right hip pain, fever, right flank pain, confusion, disorganized speech, fluctuating episodes of myoclonus, migrainous headache, occasional unexplained laughter, and a single visual hallucination. Hospitalized on May 3, 2024, at Santa Casa de Barretos, initial treatment targeted urinary tract infection with ceftriaxone. Neurological evaluation revealed intentional tremor worse on the right, frontal release signs, recent anterograde amnesia, and anomia. Diagnostic hypotheses included encephalitis associated with pyelonephritis/sepsis, prompting cerebrospinal fluid (CSF) puncture, cranial CT, and MRI. Empirical treatment with acyclovir, ceftriaxone (CNS dose), and dexamethasone was initiated but showed no clinical improvement. CSF analysis revealed hyperproteinorrachia, hypoglycorrhachia, and lymphocytic pleocytosis, suggestive of fungal encephalitis despite negative fungal serologies and culture. Infectious disease consultation advised repeat CSF sampling, broadening the investigation to include acid-fast bacilli, MTB-RIF (molecular tuberculosis rapid test), viral panel, fungal culture, and escalated antibiotic therapy (ampicillin and amikacin), alongside continued acyclovir for 21 days. Persistent clinical decline led to a repeat MRI showing findings indicative of fungal encephalitis, confirmed by positive urine culture for kluyveromyces marxianus. Empirical treatment with amphotericin B for CNS was commenced, resulting in significant improvement in consciousness level and CSF parameters after one week. Discharge planning included fluconazole therapy with scheduled serial imaging and CSF exams to guide future medication discontinuation. Ongoing CSF fungal culture is pending. Fungal CNS infections, including fungal encephalitis, are particularly challenging due to their dependence on host immune status and the virulence of the fungal strain involved. These infections often lead to substantial morbidity in immunocompromised individuals and can result in severe, potentially fatal neurological outcomes. This case highlights the unusual occurrence and diagnostic hurdles of fungal encephalitis in immunocompetent patients, originating from a primary urinary tract infection. The timely initiation of empirical antifungal therapy with amphotericin B, despite initial negative cultures, played a critical role in achieving clinical improvement. Given the intricate diagnostic and therapeutic landscape, further investigation into the diverse etiologies of encephalitis, including fungal infections in immunocompetent individuals, is crucial for optimizing patient outcomes.

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GABA-B encephalitis as a paraneoplastic manifestation of malignant neoplasm

Andre Felipe Candêas Amorim[1], Victor Cardoso de Faria[1], Gabriela Rodrigues Tomaz[1], Isabella Vargas Baldon[2], Marianna Pinheiro Moraes de Moraes[1], Enedina Maria Lobato de Oliveira[1], Orlando Graziani Povoas Barsottini[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]Universidade Estadual Paulista, São Paulo SP, Brazil

Correspondence: Andre Felipe Candêas Amorim; Email: andfcamorim@gmail.com.

Case presentation: A 68-year-old man began to experience significant apathy and after approximately 15 days evolved with episodes of focal to bilateral tonic-clonic epileptic seizures. The patient was previously hypertensive, diabetic and an active smoker with a smoking history of 50 pack-years. He was initially seen at an external service and, after MRI and cerebrospinal fluid collection, the hypothesis of herpetic encephalitis was suggested. He was treated with 14 days of acyclovir and anti-seizure medication was started, which improved the epileptic seizures. Nevertheless, the patient remained apathetic and started to experience delirium and visual hallucinations. In our service, the hypothesis of autoimmune encephalitis was raised, and the patient underwent a head MRI which showed hypersignal on T2/FLAIR in the hippocampus and right uncus associated with slight edema. The patient's CSF showed pleocytosis with 42 cells/mm^3 with lymphomonocyte predominance, protein 69 mg/dL, glucose 55mg/dL, negative infectious tests and a negative viral panel. The antibody panel for autoimmune encephalitis revealed positivity for GABA-B The patient underwent treatment with pulse therapy with methylprednisolone 1g/day for 5 days and then received human immunoglobulin 2g/kilo distributed over 5 days. The patient evolved with complete improvement of psychiatric symptoms, however he remained dependent on anti-seizure drugs. During neoplasia investigation, small cell lung carcinoma was diagnosed, and the patient began treatment with chemotherapy while still hospitalized. **Discussion:** GABA-B encephalitis is associated with the phenotype of limbic encephalitis, epilepsy, rapidly progressive dementia, and ataxia. Half of the cases are usually paraneoplastic manifestations and generally associated with small cell lung carcinoma.. The main differential diagnosis of the case mentioned is herpetic encephalitis, which should always be considered due to its severity. In these cases, head MRI (herpes usually shows more significant edema, generally unilateral, more significant white matter involvement and a greater chance of changes on DWI) and CSF with a viral panel can help in managing the cases. **Final comments:** It is extremely important for neurologists to know about autoimmune encephalitis, its clinical manifestations and its treatment. It is worth mentioning that such diseases are potentially treatable causes of encephalitis, and may even be the first manifestation of an undiagnosed neoplasm.

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GAD-linked cerebellar ataxia and chronic use of phenytoin

Lucas Medeiros Dunga[1], Matheus Procópio Guimarães[1], Tereza Cristina Batista Dias[1], Fabricia dos Santos Almeida[1], Amanda Lima Sampaio[1], Lucca Ferdinando Queiroz Fernandes[1], Cláudio de Souza Fernandes Júnior[1], Pedro Henrick Guimarães Carvalho[1], Iury Hélder Santos Dantas[1], Helmut Kennedy Azevedo Do Patrocínio[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Lucas Medeiros Dunga; Email: lucasdunga2000@gmail.com.

Case presentation: Patient MFBA, female, 45 years old, with a history of epileptic seizures that began in 2018, on chronic use of phenytoin (300 mg/day) since diagnosis. In 2022, she started experiencing imbalances associated with falls. At the first consultation, the patient was unable to walk without support, needing assistance. She had already been evaluated by otolaryngologists and psychiatrists. She denied diarrhea, recent infections, and had never had COVID-19 (received the Oxford and Pfizer vaccines). Comorbidity: hypothyroidism, using Puran 25 mcg. Muscle strength was G5 globally, except in the proximal muscles of the lower limbs (G4). Tendon reflexes were brisk in the upper limbs and absent in the lower limbs, with a bilateral extensor plantar response, without clonus. Motor coordination without dysmetria and diadochokinesia. Sensitivity without sensory level, with anesthesia in the lower limbs and preserved pressure sensation, hypopalesthesia in the right lower limb and apalesthesia in the left lower limb, proprioception absent on the left and reduced on the right. Balance with positive Romberg and ataxic gait. With the withdrawal of phenytoin and replacement with levetiracetam, there was a partial improvement in imbalance and falls. Cranial MRI revealed cerebellar folia atrophy. Antibody test showed anti-GAD greater than 2000IU/ml, normal vitamin E (5.9), elevated anti-thyroperoxidase antibodies (>1300, NR <60), anti-thyroglobulin 27.3 (NR <6.5) and positive ANA with a dense fine speckled pattern 1/320. Serologies for HIV, VDRL, hepatitis B and C were negative. The patient presented a medical picture that suggested a cytokine storm due to alterations in various autoimmune tests. The patient showed significant clinical improvement, being able to walk more independently, although with a cautious gait. The patient was referred for evaluation with a rheumatologist and suggested to undergo electromyography (EMG) and cerebrospinal fluid (CSF) examination, in addition to being directed to the ataxia clinic at HUOL for continuous follow-up. **Discussion:** The patient presented a condition that was compatible with an cytokine storm, manifested by several autoimmune changes and cerebellar ataxia, probably exacerbated by the chronic use of phenytoin. Withdrawing the drug and replacing it with levetiracetam brought a significant improvement in the clinical picture, suggesting that phenytoin toxicity could be contributing to the ataxia. **Final comments:** This case highlights the importance of considering the long-term adverse effects of antiepileptic medications, such as phenytoin, in patients presenting new neurological symptoms. However, it indicates how symptoms of autoimmunity can be masked with the use of inappropriate medications in the treatment of epilepsy.

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Gamma-delta T cells suppress microbial metabolites that activate striatal neurons and induce repetitive/compulsive behavior in mice

Laura M. Cox[1], Bruna K. Tatematsu[1], Danielle S. Leserve[1], Thais G. Moreira[1], Marília G. de Oliveira[1], Ana C. Rosa[1], Roberta C. Fonseca[1], Andre G Oliveira[2] , Howard L. Weiner[1], Rafael M. Rezende[1]

[1]Harvard Medical School, Brigham and Women's Hospital, Ann Romney Center for Neurologic Diseases, Boston, USA.
[2]Department of Biophysics and Physiology, Biologic Institutes of Sciences.

Correspondence: Ana Clara Rosa Coelho Guimarães; Email: anaclararcg@gmail.com.

Intestinal γδ T cells are crucial for maintaining the gut microbiota homeostasis, which significantly influences brain function and behavior. Abnormalities in gut microbiota have been associated with conditions such as autism and depression, yet the specific mechanisms remain unclear. Here, we show that mice lacking γδ T cells (γδ -/-) developed a pattern of repetitive/compulsive (R/C) behavior that was characterized by repeated jumping and increased number of buried marbles as compared to wild type (WT) littermate controls. R/C behavior was associated with the activation of the striatum, but not with other brain areas, as measured by increased number of cells positive for the transcription factor FosB/ΔFosB. Moreover, co-expression of FosB/ΔFosB and NeuN, a specific neuronal marker, indicated that neurons, rather than non-neuronal cells were critical for γδ -/- mouse repetitive behavior. We then investigated the mechanisms by which γδ -/- mice developed R/C behavior and found that the gut microbiota played a critical role, since γδ -/- mice colonized with WT microbiota no longer had repetitive behavior and WT mice colonized with γδ -/- microbiota developed repetitive behavior. Importantly, by performing a large-scale metabolomic analysis, we identified the microbial metabolite hippurate (HIP), a glycine conjugate of benzoic acid formed through gut bacterial metabolism of aromatic amino acids, to be highly increased in the stool, serum and cerebrospinal fluid (CSF) of γδ -/- mice. To investigate whether HIP induced R/C behavior in WT mice, we treated mice with HIP and found that it induced R/C behavior similarly to what we found in γδ -/- mice. Moreover, WT mice treated with HIP showed increased activation of striatal neurons. To investigate which striatal neurons responded to HIP by upregulating FosB/ΔFosB, we performed RNAscope in situ hybridizatio of WT mouse striatum after treatment with HIP. We used Fosb, Drd1a (dopamine type 1 receptor; D1R) and Drd2 (dopamine type 2 receptor; D2R) probes to investigate whether medium spiny neurons, the most abundant neuronal type in the striatum, were involved in this process. We found that the majority of FosB+ cells in the striatum co-expressed D1R after HIP treatment, suggesting that HIP dysregulates striatal circuits associated with D1R+ MSNs. Notably, culture of striatal slices in the presence of HIP or the D1R selective agonist dihydrexidine hydrochloride (DIH) led to similar c-Fos expression in striatal neurons. Accordingly, the use of a D1R selective antagonist abolished this effect, indicating that HIP induces R/C behavior by activating D1R expressed on D1R+ MSNs and thus by dysregulating the direct pathway of the cortical-striatal-basal ganglia-thalamic circuit. Altogether, these data suggest that intestinal γδ -/- T cells shape the gut microbiota and their metabolites and prevent dysfunctions of the striatum associated with behavior modulation.

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Ganglionopathy and leprosy: a rare clinical presentation or overlapping pathologies?

Stefano Machado[1], Iago Resende Carvalho[1], Arthur Costa Nascimento[1], Isabela Maria Bernardes Goulart[1], Diogo Fernandes dos Santos[1]

[1]Universidade Federal de Uberlândia, Hospital de Clínicas, Uberlândia MG, Brazil.

Correspondence: Stefano Machado; Email: stefanomachadomed@gmail.com.

Case presentation: A 52-year-old man with a progressive asymmetrical sensory deficit for several years was diagnosed in 2016 with borderline tuberculoid leprosy. He was treated with the multibacillary polychemotherapy regimen, achieving cure criteria in 2018. However, presented some episodes of leprosy neuritis after the end of treatment. In 2023, subacutely (over a period of 4-8 weeks), he experienced sensory worsening and postural instability and subsequently developed a decrease in visual acuity, both progressive. The clinical evaluation revealed xerophthalmia, xerostomia, mydriatic and hyporeactive pupils, thermal and pain hypoesthesia in distal segments (with an asymmetrical pattern, also with greater impairment of cold areas), apalesthesia, global areflexia, ataxic gait and presence of Romberg sign. Faced with a new condition of sensory ataxia, electroneuromyography was performed, demonstrating absences of all sensory nerve action potentials evaluated (median, ulnar, radial, superficial peroneal, and sural) with an asymmetric axonal motor impairment (already observed in previous evaluation). An extensive laboratory evaluation was carried out, including metabolic, hormonal, serological and deficiency screening - serum and CSF - which showed no pathological changes. Furthermore, given the possibility of a paraneoplastic condition, an investigation for neoplasia was carried out, but there were no changes. Investigation of inflammatory diseases showed no changes. However, the Schimmer test and salivary flow assessment confirmed the presence of xerophthalmia and xerostomia. Based on the results of the investigation, a diagnosis of ganglionopathy secondary to Sjogren's Syndrome was made and immunosuppressive treatment was started with an excellent response and significant improvement after the Cyclophosphamide infusion. **Discussion:** Leprosy neuropathy is extremely common and prevalent, especially in underdeveloped countries. It commonly presents as a multiple mononeuropathy, classically with sensory predominance (truncal and/or intradermal), rarely with signs of sensory ataxia, even in cases of long evolution. Proximal involvement of the peripheral nervous system (dorsal root ganglion, brachial plexus, and motor roots) and even the central nervous system (medullary) have already been described in some cases in the literature. However, there are still doubts regarding the mechanism of injury, which may be related to direct aggression by M. leprae to these structures or be related to an immune-mediated process. **Final comments:** Ganglionopathy is usually caused by paraneoplastic, autoimmune, genetic, toxic, deficiency, and infectious conditions. Despite being a rare presentation, it may be present in the context of leprosy, especially in multibacillary cases, during episodes of leprosy reaction. Otherwise, leprosy is a systemic inflammatory disease and can trigger the development of other rheumatological diseases.

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Gene expression analysis of the WNT and mtor pathways from brain tissue of patients with focal cortical dysplasia type II

Douglas Bottega Pazzin[1], Thales Thor Ramos Previato[1], Marina Ottmann Boff[1], Fernando Xavier[1], Daniel Rodrigo Marinowic[1]

[1]Instituto do Cérebro, Porto Alegre RS, Brazil.

Correspondence: Marina Ottmann Boff; Email: mottmannboff@gmail.com.

Background: Focal cortical dysplasia (FCD) is the most common malformation of cortical development to cause drug-resistant epilepsy (DRE) in children and adults. The origin of the malformation in FCD remains unknown. Changes in the gene expression profile may be responsible for alterations in cell responses and tissue malformations. The PI3K/AKT/mTOR pathway is the main one related to type II FCD and is modulated by the WNT pathway. The WNT and mTOR pathways are responsible for signaling related to cell differentiation, apoptosis, and migration in different tissues, fundamental for corticogenesis. The main treatment for ERDs is resection of the epileptogenic zone, with an efficacy of 60-80% for DCF II (complete seizure control with anti-epileptogenic drugs (AED)). Seizure control has been reported in patients with ERD using AED adjunct to mTOR inhibitors. It is therefore of interest to investigate potential new targets related to the formation of these epileptogenic zones in FCDs. **Objective:** Evaluation of the expression of WNT and mTOR pathway genes in patients with Focal Cortical Dysplasia type II. **Methods:** The groups were divided into control (healthy cortex tissue), acquired from surgery for hippocampal resection; focal cortical dysplasia IIa; and focal cortical dysplasia type IIb. From these samples, genetic material was extracted and amplified to observe expression levels of 16 genes involved in the WNT pathway and 3 in the mTOR pathway through qPCR. Amplification values were analyzed by relative expression using the formula: 2(-ΔΔCt). The values found were distributed in a heat map following the Pearson correlation. **Results:** Using relative expression values, a greater change in hyperexpression was identified for WNT genes in patients with FCD IIb to group IIa. The type IIa FCD group showed a decrease in expression for 7 WNT genes and 1 mTOR gene, while the type IIb FCD group showed increased expression for all 16 WNT genes and 2 mTOR genes. The increase in expression in the mTOR and RHEB genes is consistent with patterns in the literature for increased mTOR activity in patients with type II FCD, however, it was possible to identify a clear difference in expression profile between these diseases when analyzing WNT. The evaluation of Peason's correlation presented a strong positive correlation among all WNT pathway genes for FCD IIa, compared to 14 genes with a strong positive correlation and 2 with a strong negative correlation for FCD IIb. **Conclusion:** These data showed a specific relationship between type II FCD and changes in the WNT pathway, in addition to a distinct expression profile between the two types of the disease, with different expression correlations, enhancing assistance in post-surgical diagnosis.

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Gene therapy for Duchenne muscular dystrophy in Brazil: a case report

Clara Gontijo Camelo[1], Felipe Franco da Graça[2], Mariana Cunha Artilheiro[3], Edmar Zanoteli[3]

[1]Instituto Neurolum, São Paulo SP, Brazil.
[2]Universidade Estadual de Campinas, Campinas SP, Brazil.
[3]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Clara Gontijo Camelo; Email: claragc@gmail.com.

Case presentation: A boy, the first child of non-consanguineous parents, had an uneventful pregnancy and delivery, reaching developmental milestones on time. He showed elevated liver enzymes, prompting further investigation that revealed a CPK level of 19,700U/L. During a pediatric neurology evaluation, he exhibited difficulty running, an inability to jump, and required assistance to climb stairs. Further investigation identified a duplication of exon 2 in the Duchenne muscular dystrophy (DMD) gene. He was diagnosed with DMD at 4 years and 11 months old and began corticosteroid therapy at 5 years old (prednisolone 0.75mg/kg/day, daily use). An anti-AAVrh74 antibody test was below the reference value, and the patient received gene therapy with Microdystrophin (Elevidys) at 5 years and 8 months old. The medication infusion lasted 1 hour and 30 minutes and was uneventful. Per the infusion protocol, the prednisolone dose was increased to 1.75mg/kg/day (an increase of 1mg/kg/day) one day before the infusion and will be maintained at this dose for at least 60 days post-infusion. For five days post-infusion, the patient experienced nausea and vomiting, which were manageable with medication. There were no reports of fever or fatigue. One week post-infusion, the patient showed no abnormal laboratory results (platelets, liver function, or troponin-I). **Discussion:** This was the first DMD patient receiving gene therapy in Brazil. In June 2023, the FDA approved Microdystrophin therapy

(Elevidys) under accelerated approval for ambulatory boys aged 4-5 years with DMD, provided they do not have variants affecting exons 8-9. The therapy involves a single dose, and post-infusion monitoring is required for at least three months, including weekly liver function tests and clinical exams for three months, weekly blood counts for one month, and weekly troponin-I levels for one month, with more frequent monitoring as needed. Clinical studies suggest that patients receiving this medication show disease stabilization with maintenance of time to rise. However, studies have yet to show positive results for the primary outcome of the North Star Ambulatory Assessment. The most serious side effects reported to date include immune-mediated myositis and myocarditis. Common side effects, though not severe, include nausea, vomiting, fever, and malaise. **Final comments:** A new era of treatment for patients with DMD has begun, highlighting the need to understand the long-term efficacy and safety of this medication.

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Gene therapy for spinal muscular atrophy amid the dengue outbreak in Brazil: a case report

Felipe Franco da Graça[1], Marcondes Cavalcante França Jr.[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Felipe Franco da Graça; Email: felipefgraca@hotmail.com.

Dengue fever is a mosquito-borne viral disease prevalent in tropical and subtropical regions globally, primarily spread by the Aedes aegypti mosquito. It manifests in a spectrum from mild flu-like symptoms to severe, life-threatening conditions. Among the classic laboratory manifestations, we observe thrombocytopenia and alterations in liver enzymes, which are mostly asymptomatic but can potentially evolve into more severe conditions. Given that these same laboratory changes can also be observed following therapy with onasemnogene abeparvovec (OA) for spinal muscular atrophy (SMA), the occurrence of dengue in patients after infusion may lead to potential complications. We describe the case of a patient with SMA Type I, diagnosed at 6 months of age (with 2 SMN2 copies) due to severe respiratory complications from an infectious episode. After diagnosis, the patient began treatment with Risdiplam with good response until the age of 9 months when gene therapy was infused. The patient was put into prednisolone at a dose of 1mg/kg for one month. Only mild thrombocytopenia (120,000/mm^3) and an elevation in transaminases less than 2 times the upper limit of normal were observed. One month after OA infusion, as per protocol, a gradual reduction of corticosteroid therapy was started, and by the eighth week post-infusion, the patient was on a 0.25mg/kg dose of prednisolone. At this point, the patient began to exhibit symptoms of fever, lethargy, red spots on skin, thrombocytopenia of 80,000/mm^3 and transaminase levels elevated to three times the upper limit of normality. Considering the concurrent dengue outbreak in Brazil at the time, serology was ordered and turned out to be positive. Since there were no similar cases described in the literature, we decided to keep prednisolone dose, and to recollect samples every 2 to 3 days. Eight days after the onset of symptoms, there was an improvement in laboratory findings with normalization of all values. Thereafter we decided to discontinue corticosteroid therapy.

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General evaluation of drugs used by post-stroke patients in the city of Marabá/Pará

Francielly de Fátima Araújo da Silva[1], Adalberto Alves Moreira Neto[1], Maria Joana da Silva Pinto[2], Thawanny Gomes Varão[1], Ruth Carvalho Machado de Mendonça[1], Samuel Nunes Silva[1], Alexia Adriane Santiago Abdon[1], Vinicius Dirlan Rodrigues Pinto[1], Jorge Luiz Dutra Junior[1], Alexsandro Julio Rodrigues da Silva Júnior[1]

[1]Universidade do Estado do Pará, Marabá PA, Brazil. [2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Francielly de Fátima Araújo da Silva; Email: franciellyamrr@gmail.com.

Background: A stroke is defined as an abrupt interruption of blood flow to the brain parenchyma, causing significant neurological deficits in the affected region, lasting more than 24 hours. Physiological mechanisms classify it as ischemic (iS) and hemorrhagic (hS) and its main risk factors, also called disease-modifying factors, are comorbidities such as high blood pressure, diabetes mellitus, cardiovascular arrhythmias, dyslipidemia, smoking, alcoholism and obesity. In Brazil, the medication list available for stroke's treatment is based on the classes of non-steroidal anti-inflammatory drugs, antihypertensives, antithrombotics and thrombolytics. **Objective:** Outline the main classes of medication used after a stroke episode by patients treated at a healthcare unit in the state of Pará, Brazil. **Methods:** This is a cross-sectional, unicentric study with quantitative criteria, with a sample of 20 patients with cerebrovascular disease (α=0.05), selected after a routine consultation at the neurology outpatient clinic at the São Padre Pio dos Capuchinhos Healthcare unit, in the city of Marabá, in the State of Pará. The data acquisition took place in October and November of 2023, using a general clinical-epidemiological assessment instrument created by the authors, which questioned the medications used by patients. **Results:** In total, 46 different drugs were found based on the responses of the 20 patients in the sample, with 1 being the minimum amount of medications taken daily by a patient and 8 concomitant medications being the maximum amount. Antihypertensives were the medications most frequently taken by patients (rf=0.40), followed by hypoglycemic drugs (rf=0.14), non-steroidal anti-inflammatory drugs (rf=0.12) and anticonvulsants (rf=0.06). The presence of anti-vertigo medications such as betahistine dihydrochloride in the routine of these patients got our attention (rf=0.03). These results are related to the risk factors for stroke, especially the high number of antihypertensive drugs, in addition to the fact that the registered medications demonstrated the presence of polycomorbidities that accompanied cerebrovascular accident (CVA) episodes, such as inflammatory diseases, Alzheimer's disease, Parkinson's and diseases that affect the vestibular system, such as Ménière's disease (endolymphatic hydrops). **Conclusion:** In the panorama of the studied population, the use of post-stroke medications was associated with risk factors and polycomorbidities. The highest use of antihypertensive drugs was highlighted, followed by hypoglycemic drugs, NSAID's and anticonvulsants.

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Genetic migration dynamics: unraveling the influence of internal migration on Huntington's disease prevalence in the Amazonas state

Marcus Vinicius Della Coletta[1], Maria Esther Lima Rios Evangelista[2], Emanuelle Campos Amara[3], Fernanda Almeida Carvalho[3], Danilo Lemes Reis[3], David França Ferreira Cruz[3], Manuela Brandão Ferreira Teles[3], Diana Vieira Brito[3], Helio Afonso Ghizoni Teive[1], Carlos Henrique Ferreira Camargo[1]

[1]UFP, Curitiba PR, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Maria Esther Lima Rios Evangelista; Email: mariaesther.rios@hotmail.com.

Background: The state of Amazonas, located in the northern region of Brazil, exhibits a demographic profile characterized by indigenous and African influence alongside European admixture. Historically, the region has attracted migrants from various parts of Brazil, mainly from Pará (north of Brazil) and northeastern states. These migrations, driven by economic opportunities and social factors, have led to a complex intermingling of genetic backgrounds within the Amazonian population. **Objective:** In this study, we investigate the influence of internal migration within Brazil on the prevalence of Huntington's disease in the state of Amazonas. **Methods:** We studied 22 families affected by HD in the state of Amazonas and identified a total of 51 genetically confirmed cases. We trace the origin of the oldest affected member of each family, within Brazil or abroad. We analized the genetic characteristics of families according to their geographic origin. **Results:** We identified 22 patients (43.1%) originated from families with roots in northeastern states. Another 22 cases (43.1%) were from the North of Brazil, 1 (1.9%) patient from Venezuela and 6 patients (11.7%) of unknown origin. The paternal transmission is predominant among patients from the Amazonas state, with 8 out of 13 cases exhibiting this mode of inheritance. Similarly, patients from Ceará show a higher prevalence of paternal transmission, with 10 out of 15 cases following this pattern. The other states display a more balanced distribution between paternal and maternal transmission. Regarding the size of CAG expansions, patients from the Amazonas state exhibit the largest average expansion size at 50.9 repeats, followed by patients from Pará (47.6 repeats), Ceará (46.4 repeats), and Pernambuco (46.2 repeats). **Conclusion:** This study highlights the impact of internal migration on the distribution of Huntington's disease within Brazil with a significant contribution from other states to the prevalence of Huntington's disease in Amazonas. The influx of migrants from northeastern states, where HD prevalence may be higher due to genetic and environmental factors, has likely contributed to the increased occurrence of the disease in Amazonas. This underscores the importance of considering local demographic factors and historical migration patterns in understanding disease epidemiology.

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Genetic search: techniques that resulted in the discovery of the Huntington's disease gene

Gustavo L. Franklin[1], Hélio A. G. Teive[2], Fernando Spina Tensini[2], Carlos Henrique Ferreira Camargo[2], Nayra de Souza Carvalho de Lima[3], Diego de Castro dos Santos[4], Sarah J. Tabrizi[5,6], Camilla Vanessa Araújo Soares[7], Tiago César Costa da Silva[7], Alex Tiburtino Meira[8]

[1]Pontifícia Universidade Católica do Paraná, Faculdade de Medicina, Departamento de Medicina Interna, Curitiba PR, Brazil.
[2]Universidade Federal do Paraná, Hospital de Clínicas, Departamento de Medicina Interna, Serviço de Neurologia, Unidade de Distúrbios do Movimento, Curitiba PR, Brazil.
[3]Universidade Vila Velha, Faculdade de Medicina, Vila Velha ES, Brazil.
[4]Universidade de São Paulo, São Paulo SP, Brazil.
[5]University College London, Queen Square Institute of Neurology, Huntington's Disease Centre, Department of Neurodegenerative Diseases, London, UK.
[6]University College London, Dementia Research Institute, London, UK.
[7]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[8]Universidade Federal da Paraíba, Departamento de Medicina Interna, João Pessoa PB, Brazil.

Correspondence: Camilla Vanessa Araújo Soares; Email: camilla.vanessa46@gmail.com.

Background: The identification of the Huntington's disease (HD) gene was a major historical milestone that enhanced understanding of the pathology and paved the way for future therapeutic approaches. **Objective:** Celebrating the last three decades since this event, the aim of this study is to revisit the techniques that led to the discovery of the HD gene. **Methods:** This is a review using the PubMed, Web of Science, LILACS, Embase, and SciELO databases, with the search terms "Huntington Disease", "Genetic Markers", "Chromosome Mapping" and "Genetic Linkage" combined with boolean operators. **Results:** In 1979, prominent geneticists gathered with the aim of exploring the potential of Restriction Fragment Length Polymorphism (RFLP) technology, which had recently been described, to map the location of the HD gene. It was essential to evaluate large families, as genetic linkage required observing multiple transmissions within the family to determine the proximity of the markers on the chromosomes. The main method to identify the HD gene was positional cloning via linkage analysis, mapping genes based on their co-segregation with specific genetic markers. Using linkage analysis, researchers mapped the HD gene to chromosome 4, observing marker co-inheritance with the disease in large families. Researchers used linkage analysis with extensive HD family pedigrees and a limited set of polymorphic markers detected via Southern blot. The G8 probe showed a distinctive RFLP pattern linked to HD in two large affected families. They identified HindIII palindromic sites (H1 and H2) within the chromosomal region using this probe. Fragment lengths varied among different HD lineages. Despite identifying G8's proximity to the HD gene, they used human-mouse somatic cell hybrids to determine its specific human chromosome origin. By performing Southern blot with the G8 probe and chromosomal DNA from hybrids, they identified cell lines with human DNA homologous to G8. This mapped the G8 probe to a specific human chromosome, as they knew the chromosomal composition of each cell line. They concluded the G8 probe only hybridized with DNA from human chromosome 4. Consequently, in 1983, they concluded that the G8 DNA probe was located on human chromosome 4 and inferred that the HD gene resided in the region corresponding to the G8 DNA probe. **Conclusion:** The main techniques that enabled the discovery of the HD gene were RFLP, positional cloning, linkage analysis, and hybridization of human and mouse somatic cells. These techniques allowed for the identification of the HD gene, driving studies and publications on the disease. Now, we understand HD is autosomal dominant with CAG expansion in HTT gene on chromosome 4p16.3. Although disease-modifying treatment is still emerging, the search for the HD gene has illuminated the scientific community and introduced technologies used in other diseases. Today, genetic therapies are a closer possibility, inspiring future generations of researchers.

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Genetic testing in patients with oligo: or asymptomatic elevated CK

Ana Luisa de Carvalho Cardozo Hernández[1], Weverton Carlos da Silva Teixeira[1], Andre Macedo Serafim da Silva[1], Edmar Zanoteli[1], Cristiane de Araújo Martins Moreno[1], Lucas Marenga de Arruda Buarque[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Ana Luisa de Carvalho Cardozo Hernández; Email: ana_luisa_1997@hotmail.com.

Background: Elevated creatine kinase (CK) levels are the most sensitive indicator of muscle injury. An increased serum level of CK in pauci- or asymptomatic individuals is a diagnostic challenge, as it may be associated with either physiological conditions, such as exercise, or even signal an ominous neuromuscular disease. **Objective:** To assess the percentage of patients diagnosed with a neuromuscular disorder in the setting of elevated CK with an oligo- or asymptomatic presentation within a specialized neuromuscular service. **Methods:** Retrospective transversal study performed in the outpatient clinic of neuromuscular disorders from HCFMUSP. Medical records from all patients who underwent genetic testing in our Neuromuscular Clinic from the period of 2015-2023 were obtained and the inclusion criteria were (1) CK dosage equal to or higher than 5 times the upper limit reference value and (2) asymptomatic or oligosymptomatic patients, with clinical complaints compatible with the hypothesis of a metabolic disorder (we searched for the words myalgia, fatigue, mild weakness' without objective clinical muscle weakness - and/or history of rhabdomyolysis). Exclusion criteria: Occasional causes of elevated CK, such as malignancies, drug and alcohol abuse, rheumatic, thyroid, and parathyroid disorders, infections, hematologic diseases, or use of drugs such as statin. **Results:** 34 out of 724 patients were selected. 23 patients were male and 11 female, the mean age at onset was 24 years, and the mean disease duration was 9 years. Mean serum CK was 2692, and levels varied between 978 to 3x106 (Reference value <192). 26 (76.4%) patients had no family history of neuromuscular disorders. 22 (75.9 %) patients complained of myalgia, 20 (69%) had fatigue, 16 (55.1%) complained of some degree of muscle weakness and 5 (17.2%) were asymptomatic. In additional exams, 4 (26.7%) patients presented with a myogenic pattern on EMG, 7 (53.8%) presented with myopathic changes in MRI, and 13 (56.5%) had alterations in muscle biopsy. 31 patients were tested with a genetic panel for metabolic muscle disorders (12 from INVITAE and 19 from Mendelics), and 3 individuals underwent full exome analysis. Genetic testing revealed pathological variants in 11 patients, variants of unknown significance in 13 individuals, and the absence of variants in 7 subjects. The most frequent mutated genes were ETFDH (n=3), followed by ANO5 (n=2) and GAA (n=2). Other mutated genes included B4GALT1 (n=1), CAP3 (n=1), PYGM (n=1), and RYR1 (n=1). **Conclusion:** Genetic testing in oligo- or asymptomatic patients with elevated CPK was associated with pathological variants in about a third of patients. ETFDH, ANO5, and GAA were the most common mutated genes found. This highlights the importance of this finding in metabolic disorders and certain forms of myopathy.

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Genotype/phenotype correlation and peripheral neuropathy in Wilson's disease (WD) in the state of Ceará, Brazil

Caio Levi Mendes Costa[1], Jéssica Queiroz de Sousa Sobrinho[1], Francisco de Assis Aquino Gondim[1], Alan Duarte Braz[1], Leonardo José Rodrigues de Araújo Melo[1], Francisco Duque de Paiva Giudice Junior[1], Leonardo Elias Araujo dos Santos[1], Mariana Alcântara Tavares[1], Rodrigo Fagundes da Rosa[1], Emanuela Freire Caetano Davi[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Caio Levi Mendes Costa; Email: caiogrebr@gmail.com.

Background: Wilson's disease (WD) is a rare autosomal recessive disorder caused by mutations in the ATP7B gene located on chromosome 13, affecting copper metabolism. Patients may be homozygous for a single mutation or compound heterozygous with two different mutations. More than 700 pathogenic variants of the ATP7B gene have been identified, and most WD patients exhibit compound heterozygosity. We have previously reported small fiber neuropathy in 4 patients with WD (Gondim et al. 2014). **Objective:** To analyze the ATP7B mutations and evidence of neuromuscular involvement (especially neuropathy) in patients followed up at Universidade Federal do Ceará. **Methods:** This summary describes the genetic test results of 13 patients of both sexes with WD, evaluated for neuromuscular complications at the Neurology Service of the Walter Cantídio University Hospital – Fortaleza/CE (CAAE: 05954818.9.0000.5045). Almost all (n = 12) of the patients were part of the cohort from Sobrinho, MSc dissertation, 2021 (n = 14). **Results:** Thirteen patients were analyzed, 7 men and 6 women, with an average age of 44 (30 to 58 years). Of these, 6 (46.15%) are homozygous, and 7 (53.85%) are compound heterozygous. Of the homozygous patients, 4 out of 6 (66.67%) have point mutations, and 2 (33.33%) have deletions. Both deletions correspond to the c.3402del (p.Ala1135Glnfs*13) mutation. Among the point mutations, there are 2 (33.33%) cases of c.2123T>C (p.Leu708Pro), 1 (16.67%) case of c.3809A>G (p.Asn1270Ser), and 1 (16.67%) case of c.1708-1G>A. Of the compound heterozygous WD patients, 2 out of 7 (28.57%) have combinations of Point Mutation + Deletion: c.3517G>A (p.Glu1173Lys) + c.3402del (p.Ala1135Glnfs13) and c.1708-1G>A + c.3402del (p.Ala1135Glnfs13). One patient (14.29%) has the combination Deletion + Point Mutation: c.3402del (p.Ala1135Glnfs13) + c.1934T>G (p.Met645Arg). There is 1 (14.29%) case of Point Mutation + Splice Donor: c.3517G>A (p.Glu1173Lys) + c.1543+1G>C, and 1 (14.29%) case of Intronic + Splice Donor: c.3061-12T>A + c.1543+1G>C. There is 1 (14.29%) case of Duplication + Point Mutation: c.2304dup (p.Met769Hisfs26) + c.3772A>T (p.Lys1258Ter), and 1 (14.29%) case of Point Mutation + Point Mutation: c.3694A>C (p.Thr1232Pro) + c.3061-12T>A. The most frequent allelic mutation was the deletion c.3402del (p.Ala1135Glnfs*13), present in 7 alleles (29.92%). Related to the neuromuscular changes in Sobrinho (2021), 8 (57%) patients had peripheral small fiber neuropathy, identified by skin wrinkling test, and 2 (14%) had involvement of large fibers: one with axonal sensorimotor polyneuropathy and myopathy; the other exhibiting loss of ankle jerks and carpal tunnel syndrome. **Conclusion:** The genetic alterations of the 13 WD patients reinforces the wide variety of possible mutations in the disease and the high heterogeneity of mutations in the Brazilian population due to diverse ethnic backgrounds. Neuromuscular involvement is common in WD patients.

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Genotype-phenotype correlations of FGFR2/3 variations: beyond craniosynostosis - a systematic review

Gisele Lucia Canto Gomes[1], Vinicius Bacelar Ferreira[1], Bruno Machado da Silva[1], David Abraham Batista da Hora[1], Patrícia Ferreira Borges Peixoto[1], Paulo Victor Zattar Ribeiro[2]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Gisele Lúcia Canto Gomes; Email: glc.canto@gmail.com.

Background: The FGFR genes provide instructions for the construction of a protein called fibroblast growth factor receptor. Variations of FGFR2 gene are commonly linked to syndromes causing craniosynostosis and FGFR3 is generally related to achondroplasiahypochondroplasia. However, various conditions have been linked to pathogenic variants within that gene group. Understanding genotype-phenotype correlations can contribute to more accurate diagnoses, enabling personalized treatments and genetic counseling strategies. **Objective:** To explore and synthesize the genotype-phenotype correlations of FGFR 2 and 3 gene variations in addition to craniosynostosis, to provide a more comprehensive understanding of their clinical and biological impact. **Methods:** A systematic review was conducted according to PRISMA guidelines. PubMed, Embase, Web of Science and Cochrane Central databases were searched for studies. The words used for research were: "FGFR 3 variants", "FGFR2 variants", "phenotype", "clinical features" and "manifestations". All studies that cited variations in FGFR 2 and 3 genes and phenotypes were included regardless of the study design. Animal models, in vitro studies, and reviews were excluded. **Results:** Nine studies were included, three of

which referred to cases of craniosynostosis phenotype associated with FGFR2/3 mutations. Celie et al. associated the c.1024T>A;p.C342 S variation in FGFR2 with Jackson-Weiss Syndrome. Kant et al. associated the p.M5281 and p.F384L variations with short stature. . Paur et al. associated FGFR3 with hepatocellular carcinoma and Sturla et al. reported FGFR3 variations with neuroblastoma, Ewing's sarcoma, neurofibrosarcoma, rhabdomyosarcoma, bladder carcinoma, melanoma, prostate carcinoma, colorectal carcinoma, breast cancer and myeloma. And Wang et al. reported two cases of c.1024T>C, p.Cys342Arg variations in FGFR2 associated with Crouzon syndrome. **Conclusion:** FGFR are critical for various developmental processes, and pathogenics variations in FGFR2, and FGFR3 genes are well-documented for their role in craniosynostosis syndromes. However, nowadays, evidence has been suggesting the influence in a broad spectrum of clinical phenotypes, such as Jackson-Weiss syndrome and neoplasia. These insights underline the importance of comprehensive genetic testing and tailored clinical management for individuals with FGFR gene variations, extending beyond the conventional focus on craniosynostosis. Future research should further explore these genotype-phenotype relationships in order to achieve these therapeutic goals.

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Gerstmann syndrome as a rare presentation of cerebral venous thrombosis

Guilherme Antonio Silva Ribeiro[1], Sofia Mônaco Gama[1], Eva Carolina Andrade Rocha[1], João Vitor Gerdulli Tamanini[1], Luccas Almeida Andrade[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Guilherme Antonio Silva Ribeiro; Email: guiribeiro1018@gmail.com.

Case presentation: A 36 year old woman presented to the emergency department with a thunderclap headache that started one day before. She also complained of blurred vision and denied fever, seizures or focal deficits. The past medical history was notable for tobacco smoking and she was not taking anticoagulants. Physical exam was remarkable for severe arterial hypertension (245x126 mmHg), papilledema and right homonymous hemianopsia. The patient also had left-right confusion, finger agnosia, dyscalculia and dysgraphia. Laboratory tests and cerebrospinal fluid analysis were normal. Brain computerized tomography (CT) and venogram revealed a left parieto-occipital lobar hemorrhage (volume: 57mL, ICH score 1) and thrombosis of the superior sagittal sinus, respectively. The diagnosis of intracerebral hemorrhage secondary to cerebral venous thrombosis was made. Intravenous anticoagulation was initiated, resulting in a complete resolution of the headache within 5 days. **Discussion:** In 1924, the neuroscientist Josef Gerstmann first described the Gerstmann syndrome (GS), which is characterized by a tetrad of symptoms: left-right confusion, dysgraphia, dyscalculia and finger agnosia . This condition likely results from disconnection of the parietal subcortical white matter due to injury of the left angular gyrus (parietal lobe) and adjacent structures. GS is a rare neurological condition, with no clear epidemiology. The most common etiology is ischemic stroke of the left parietal lobe due to occlusion of the left posterior cerebral artery. It can also be secondary to trauma, neoplasms, degenerative diseases, vascular malformations, infections and other less common etiologies. This case describes a rare presentation of GS due to intracerebral hemorrhage secondary to superior sagittal sinus thrombosis. **Final comments:** GS is characterized by left-right disorientation, acalculia, finger agnosia and agraphia. Usually, this condition is caused by ischemic stroke, trauma, neoplasms, degenerative diseases, vascular malformations and infections GS can be a rare presentation of intracerebral hemorrhage secondary to superior sagittal sinus thrombosis

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Giant axonal neuropathy: case report of an intermediate form with late manifestations

Amanda Leme da Silva Campinas[1], Lucas Do Rego Barros Correa Martins[1], Laura Alonso Matheus Montouro[1], Maria da Penha Ananias Morita[1], Érica Nogueira Coelho[1], Eduardo de Paula Estephan[1]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Amanda Leme da Silva Campinas; Email: amandalsc12@hotmail.com.

GAN-related neurodegeneration (GAN-RD) comprises a phenotypic spectrum ranging from severe to milder pure early-onset sensorimotor neuropathies Oversized axons are the pathological hallmark of the disease. In this case report we depicted a GAN-RD with intermediate phenotype, illustrating tardy manifestations of this ultra-rare condition. **Case presentation:** A Brazilian female, with no familial history of neurological disorders, born to non-consanguineous parents, started with difficulty to jump, to run and to keep slippers on feet (due to weakness on toes), at the age of 3. At 6-years-old, weakness progressed and bilateral foot drop was present. At 15-year-old upper limbs were affected, with weakness and atrophy. At 22, a wheelchair was needed to move for long distance, and at 34 patient were restricted to wheelchair and with urinary incontinence. Nocturnal ventilatory assistance, was initiated at age of 42. At the last attendance, with 43 years, she presented with areflexic paraplegia and predominantly distal upper limbs weakness, with intense atrophy on feet and hands. Were also noted: appendicular ataxia, dysphagia, dysarthria, facial weakness, hypoesthesia and impaired vibration sensibility on 4 limbs, more intense distally. Neither curly/kinky hair nor intellectual disability were present. Electrophysiology revealed severe diffuse sensory-motor neuropathy with axonal predominance. Brain magnetic resonance imaging (MRI) showed, cerebellar atrophy and bilateral and symmetrical hyperintense lesions on T2-weighted sequences compromising the middle cerebellar peduncles and brainstem in medulla region. On New Generation Sequencing panel for neuropathies biallelic variants were seen on GAN gene two variants in heterozygosity: Chr16:81.388.038AGTT>A (pVal106del ENST00000648994 - c.315_317delGTT - ENST00000648994) and Chr16:81.388.010 A>C (p.Ile95Leu ENST00000648994 - c.283A>C - ENST00000648994). **Discussion:** Giant axonal neuropathy is an inherited autosomal recessive neurodegenerative disease that affects both central and peripheral nervous systems. For differential diagnosis must be considered Charcot-Marie-Tooth (CMT) hereditary neuropathy, especially for intermediate and mild GAN-phenotypes. Other diagnosis to differentiate from include classic infantile neuroaxonal dystrophy and leukodystrophies. There is no genotype-phenotype correlation for GAN-RD, and family members with the same pathogenic variant can present different clinical phenotypes. Actually, our case had been followed as a hereditary neuropathy until the possibility of genetic test. The severity, and the presence of cerebellar ataxia were red flags against CMT hypothesis. Moreover, in the last years, she began to have cranial nerve involvement, which is also uncommon for most hereditary neuropathies. **Final comments:** This case report brings attention to this ultra-rare disease, illustrating a late clinical picture of an intermediate form of GAN-RD.

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Girdle muscular dystrophy, severe LGMD 2F subtype in a pediatric patient: case report

Cláudio de Souza Fernandes Júnior[1], Cláudio de Souza Fernandes[1], Matheus Araújo de Medeiros[2], Lucas Medeiros Dunga[2], Lucca Ferdinando Queiroz Fernandes[2], Artur Rebouças de Souza[2], Maria Eduarda Medeiros Martins[2], Diógenes Emanuel Dantas da Silva[2], Tereza Cristina Batista Dias[2], Iury Hélder Santos Dantas[2]

[1]Universidade Estadual do Rio Grande do Norte, Caicó RN, Brazil.
[2]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Cláudio de Souza Fernandes Júnior; Email: claudio.junior.711@ufrn.edu.br.

Case presentation: A 12-year-old male patient developed progressive muscle weakness of the pelvic and scapular girdle in a short period of time during early childhood, with significant involvement of the lower limbs, compromising walking and sitting. The progression of the disease began at the age of 6, with proximal and symmetrical weakness of the hip muscles. At the age of 8, the patient's pelvic girdle was affected, with a significant loss of strength in both upper limbs. In addition, fractures in the right upper limb caused by low-energy trauma demonstrate a significant correlation between the development of dystrophy and bone fragility. The condition evolved into weakness of the breathing muscles which culminated in obstructive sleep apnea (OSA), especially in the REM phase, due to hyper-relaxation of the pharyngeal muscles. The clinical suspicion was muscular dystrophy of the waist, characterized as delta-sarcoglycanopathy. A genetic evaluation confirmed the diagnosis of LGMD 2F, with a homozygous pathogenic variant in the SGCD gene. In addition, he was diagnosed with severe gastroesophageal reflux, with daily episodes of vomiting and regurgitation, requiring maximum doses of prokinetics and control medications. The condition reinforces the picture of intestinal hypomotility due to the inability to walk. In 2024, the patient had a BMI of 14.3 (below the 3rd percentile), maintaining inadequate nutrition due to dyspepsia and gastroparesis. Intake is limited to 150 ml per meal, insufficient to meet nutritional needs, resulting in a loss of 3.85 kg between February and May (13.3% of previous body weight). A diet was proposed and he was referred to physiotherapy and speech therapy. **Discussion:** Girdle muscular dystrophy type 2F is a rare form of muscular dystrophy, characterized by progressive weakness of skeletal muscles, predominantly of the scapular and pelvic girdles. The SGCD gene is implicated in this condition, with homozygous pathogenic variants frequent in some populations, including Brazilian families. The case highlights the complexity and severity of LGMD 2F, with rapid progression leading to a significant loss of function and mobility. Management of the disease is multidisciplinary, involving physiotherapy to maintain muscle function, nutritional support to combat malnutrition and dyspepsia, as well as respiratory care to prevent complications associated with obstructive sleep apnea. Bone fragility increases the risk of fractures, requiring constant vigilance and interventions to prevent new injuries. **Final comments:** Severe LGMD 2F subtype girdle muscular dystrophy is rare, especially with progression in such a short time. Several factors contribute to the severity of the disease, especially additional comorbidities. The multidisciplinary team plays a decisive role in the evolution and outcome of the condition.

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Globus pallidus internus deep brain stimulation for severe Tourette's syndrome: a case report

Rodrigo Fernandes Nitsch[1], Patrícia Áurea Andreucci Martins Bonilha[1], Mariana de Oliveira Trintinalha[1], Nathalia Cabral Bergamasco Torato[1], Mateus Coltro Dornelles[1], Leonardo Almeida Frizon[1], Alexandre Novicki Francisco[1], Marina Farah[1]

[1]Pontifícia Universidade Católica do Paraná, Hospital Universitário Cajuru, Curitiba PR, Brazil.

Correspondence: Mateus Coltro Dornelles; Email: mateuscdornelles@gmail.com.

Case presentation: G.D.N 30 years old, female was referred by her psychiatrist to a specialized deep brain stimulation (DBS) clinic due to a lifelong severe and refractory Guilles de La Tourrete Syndrome. Symptoms began at the age of 5 and slowly progressed with important vocal and motor tics associated with aggressiveness, coprolalia and aggression both self-inflicted and towards others. Pharmacological treatment, at time of presentation was Clozapine 100mg bd, |Risperidone 1mg bd, Fluoxetine 20mg bd, Biperiden 2 mg bd, Levomepromazine 5 drops at night. The patient was referred for a DBS approach. Initial Yale Global Tics Severity Scale (YGTSS) was 86 and both DBS devices were gradually adjusted (initially every 2 weeks; later, every 3 months) with significant clinical improvement: motor tics and aggressiveness lessened (current YGTSS after 1 year: 63), and both the patient and her mother referred gains in independence and life quality, after one year of DBS. **Discussion:** Tourette syndrome is a rare psychiatric disorder with estimated prevalence of approximately 0.66% of school age children with only a small fraction of those cases lasting until adulthood with debilitating symptoms. Tics brief movements or sounds that resemble voluntary actions, but appear suddenly, without regularity, often exaggerated in intensity and are repetitive and inopportune to social context.- (more than 1 motor and 1 vocal) beginning before 18 years of age and lasting for over a year define the syndrome. Even though the pathophysiology involving these symptoms is not completely identified, functional neuroimaging studies have already brought light upon the involvement of important structures from the dopaminergic inhibitory circuit (such as the basal ganglia), the motor cortex and complex cortico-cortical neural networks. Managing these cases demands a multidisciplinary approach and often turns out to be a challenging task. The development of new treatment strategies, especially regarding severe and pharmacologically resistant cases - such as the one presented above -, is a necessity and DBS poses as a viable, potentially effective, and relatively safe invasive option. **Final comments:** We present a case of an adult patient struggling with severe Tourette syndrome, impairing daily life activities, with life long pharmacological resistance, who presented significant improvement following bilateral GPi DBS implantation - a novel approach for a rare debilitating disease.

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Glossopharyngeal neuralgia secondary to bone fibrodisplasia: a case report

Ahmad Kassem El Zein[1], Rogério Tuma[1], Mauricio Silva Teixeira[1], Paula Fiuza Rodrigues de Medeiros Lacerda[1], Lucas Henrique de Moura Rogerio Garcia[1], Matheus Rodrigues Nogueira[1], Joana Laura Aihara Barbosa[1], Josenice Gomes Trindade Seidel[1], Julia Martins Brunelli[1], Lucas D'Andrea Pereira Sousa[1]

[1]Hospital Sírio-Libanês, São Paulo SP, Brazil.

Correspondence: Ahmad Kassem El Zein; Email: ahd-95@hotmail.com.

Case presentation: A 38-year-old previously healthy woman presented with multiple episodes of sharp pain on the right side of her face and ear, radiating to her jaw and malar region, each episode lasting about a minute. The pain, described as stabbing, improved with cold stimuli. She reported increased discomfort while chewing and swallowing but denied fever or other symptoms. Despite heightened sensitivity on the right side while chewing, she did not identify other triggers. A neurological examination revealed no deficits. She was hospitalized for further investigation and better pain management, receiving intravenous phenytoin empirically due to the neuropathic nature of her pain.Laboratory evaluations, including metabolic, rheumatologic, and hematologic tests, showed no abnormalities. Magnetic resonance imaging (MRI) of the temporal and cranial bones with arterial studies revealed T2/FLAIR hyperintensity and post-contrast enhancement, suggesting a fibro-osseous developmental condition in the right temporo-occipital region, compressing the jugular foramen. These findings supported a diagnosis of glossopharyngeal neuralgia secondary to jugular foramen compression caused by osteofibrous dysplasia. She was initially treated with carbamazepine, 200mg twice daily, gradually increasing to 800mg/day, and pregabalin, 75mg once daily, which provided satisfactory pain control. Rhizotomy was considered if pharmacological therapy failed. **Discussion:** Glossopharyngeal neuralgia is a rare condition characterized by shock-like pain in the jaw and face, often triggered by coughing or swallowing. In rare cases, it can lead to cardiovascular issues due to involvement of the vagus nerve. Classic glossopharyngeal neuralgia is typically caused by vascular contact, often from the posterior inferior cerebellar artery, while secondary forms result from structural changes such as fibromuscular dysplasia or tumors. Differential diagnoses, including oral cavity tumors and trigeminal neuralgia, must be ruled out when an idiopathic condition is suspected. Diagnosis relies on patient history, clinical examination, and MRI of the temporal and cranial bones.For treatment, carbamazepine is often used at a starting dose of 200 mg twice daily, potentially increasing to 1200 mg if tolerated. In refractory cases, additional medications and surgical options like rhizotomy or Stereotactic Radiosurgery (SRS) may provide better pain control. **Final comments:** This case study highlights a rare instance of glossopharyngeal neuralgia where the clinical presentation could easily be confused with other conditions. Imaging is crucial for accurate diagnosis.

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Glossopharyngeal neuralgia: a case report

Bibiana Mayer[1], Chadi Emil Adamo[2]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Chadi Emil Adamo; Email: neurochadi@outlook.com.

A.P.T, woman, 27 years old. At the age of 13, she experienced pain in the lower region of the right hemiface. This complaint was concentrated in the masseter and pre-auricular region, radiating to the angle of the jaw, with paroxysmal (multiple pulsating) quality, and a duration of approximately 1 minute and ocurred several times in a day. There was also burning pain in the right half of the tongue. The pain was aggravated by touch, swallowing, washing her face, laughing, and brushing her teeth. A few weeks ago, she underwent a dental procedure and experienced a significant exacerbation of the condition, making her unable to work. In this context, she underwent treatment with medications such as codeine, tramadol, and morphine infusions, but with disappointing results. During a neurological consultation, a diagnosis of trigeminal neuralgia was suggested with red flags for other diagnoses (age <50 years and pain in the hemilanguage) and glossopharyngeal neuralgia due to the topography of her pain. Therefore, a cranial MRI with FIESTA sequence was requested. This showed an extra-axial formation with intensity equal to cerebrospinal fluid posterior to the emergence of the IX, X, and XI cranial nerves on the right, with slight anterior displacement of these nerves, measuring approximately 1.9x0.9x0.7 cm at its largest diameters. The trigeminal pathway was intact. This image was compatible with a probable arachnoid cyst with slight anterior compression of the emergence of the IX cranial nerve on the right. In this context, a diagnosis of glossopharyngeal neuralgia was made, and treatment with carbamazepine was started, with a dose progression up to 800mg/day. The patient achieved complete remission later with the combination of low-dose lamotrigine (100mg/day). She is currently undergoing pharmacological management, and the surgical option for arachnoid cyst removal will be considered in case of refractory response. Glossopharyngeal neuralgia is a severe transient stabbing pain experienced in the ear, base of the tongue, tonsillar fossa, or beneath the angle of the jaw. The pain is therefore felt in the distributions of the auricular and pharyngeal branches of the glossopharyngeal nerve. It is commonly provoked by swallowing, talking, or coughing and may remit and relapse in the fashion of TN. Stimulation of the vagus can result in syncope. In most cases, this condition is idiopathic, but some instances might be due to symptomatic causes, again compression of the nerve by tumours, in this case, an arachnoid cyst. Patients will suffer from episodes of pain lasting for weeks or months, and then have periods of remission.

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Glossopharyngeal neuralgia: case report

Marco Antonnio dos Santos Rocha[1], Leticia Suppi de Oliveira[1], Roberta Derossi de Souza[1], Maiara Leticia Geiss[1]

[1]Universidade do Planalto Catarinense, Lages SC, Brazil.

Correspondence: Leticia Suppi de Oliveira; Email: suppilele@gmail.com.

Case presentation: A 75-year-old female presented to the emergency department of a major hospital situated in the Serra Catarinense region with left-sided cervicalgia radiating to the jaw, tongue, palate and left auricular region, dizziness and lipothymia. Her symptoms started abruptly the day before, were severe recurrent and associated with talking and swallowing. Her neurological exam was unremarkable, with the exception of an absent gag reflex bilaterally. Lidocaine spray application to the patient's posterior oropharynx resulted in localized paresthesia and symptom amelioration. A hypothesis of Glossopharyngeal Neuralgia (GN) was made. Past medical history included well-controlled arterial hypertension managed with Losartan and diffuse thyroid gland enlargement with a total volume of 22.62 cm^3, devoid of clinical consequences. Brain Magnetic Resonance Imaging and Magnetic Resonance Angiography confirmed a contact between the left posterior inferior cerebellar artery and glossopharyngeal nerve. This finding, with the whole clinical picture, confirmed the given hypothesis. She began on carbamazepine with dose increasing on the following days, as well as codeine for pain control and more applications of topical lidocaine. She was discharged with minor pain and no need for opioids and anesthetics. **Discussion:** GN clinically manifests as abrupt, intense unilateral pain episodes localized to sensory territories innervated by the glossopharyngeal nerve, including the mastoid bone, posterior pharynx, posterior third of the tongue, Eustachian tube, middle ear, oropharyngeal tonsils, and vocal folds, and may be accompanied by vagal symptoms. These episodes typically range from seconds to minutes and may occur multiple times daily, often triggered by head movements, coughing, speaking, sneezing, laughter, or mastication. The identification of concurrent diagnostic indicators is crucial for topographic diagnosis, which may also delineate etiological considerations. Notable diagnostic features include motor manifestations and the presence or absence of the gag reflex. Evaluation of swallowing and phonation functions involving the glossopharyngeal and vagus nerves is particularly pertinent in dysphagic and/or dysphonic patients. Accurate localization of compression is imperative for therapeutic stratagem. For patients without contraindications, contrast-enhanced MRI of the brain and soft tissues of the neck stands as the diagnostic modality of choice. Pharmacotherapy typically involves neuropathic agents that modulate action potentials and attenuate synaptic transmission, with Carbamazepine and Oxcarbazepine being foremost among them. In cases of refractory disease, stereotactic radiosurgery is a preferred minimally invasive alternative preceding consideration of neurovascular decompression surgery as a final resort. **Final comments:** GN constitutes a seldom-diagnosed entity that can be mistaken for other severe causes of head and cervical pain.

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Glycemic imbalance, but not diabetes mellitus (DM), increases the risk of infection in patients with acute stroke admitted to a stroke unit: a cohort study in a reference hospital in Bahia

Gabriel Souza Medrado Nunes[1], Maria Eduarda Messias Vassoler[1], Milena Fernandes de Oliveira[1], Luiz Paulo Oliveira Martins[1], Gabriel Teles de Oliveira Piñeiro[1], Thaís Barbosa de Oliveira[1], João Pedro Fernandes Gonçalves[1], Pedro Antonio Pereira de Jesus[2] , Luana Araujo Passos[1], Gabriel Souza Suzart[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil. [2]Hospital Geral Roberto Santos, Salvador BA, Brazil.

Correspondence: Maria Eduarda Messias Vassoler; Email: maria.vassoler@ufba.br.

Background: Diabetes Mellitus (DM) is known to be associated with a higher risk of infection, particularly during hospitalization. However, the correlation between glycemic imbalance during hospitalization and infection rates in patients with and without DM is not well explored in many populations, including acute neurological patients. **Objective:** To understand the relationship between glycemic imbalance, diabetes, and the risk of infection during the hospitalization period in stroke patients at a stroke unit. **Methods:** This study enrolled patients with acute stroke admitted to a referral hospital's stroke unit in Bahia, between July/2023 and March/2024. Clinical and epidemiological characteristics were extracted from patients' medical records or obtained through personal interviews. Infection was defined as the administration of antibiotics or documented infection noted in medical records. The glycemic imbalance was defined by a hemoglucotest (HGT) reading exceeding 180mg/dL. The Mann-Whitney and Chi-Square tests were employed to examine associations. **Results:** The study included 213 patients, 51.2% of whom were female, with a median age of 66.3 years (IQR: 59-74.3). Regarding comorbidities, 36.2% had diabetes, 78.95% had hypertension, and 27.7% had dyslipidemia. Infection was diagnosed in 95 patients (44.6%). Among patients with diabetes, 54.4% had an infection, compared to 38.3% of the patients without diabetes, showing a significant risk of infection for diabetic patients ($p<0.05$) in the univariate analysis. Across all patients, 52.2% experienced at least one episode of glycemic imbalance (GI), with a median of 3 (IQR: 0-3) episodes. In general, more episodes of GI were associated with infection occurrence ($p<0.05$). Diabetic patients had an 83.1% frequency of GI, a median of 3 (IQR: 1-7) episodes, while non-diabetics had a 30.8% frequency and a median of 0 (IQR: 0-1) episodes. Subgroup analysis showed that the higher GI occurrence in diabetics was linked to a higher infection risk ($p<0.01$), but this was not observed in non-diabetics ($p>0.05$). In the multivariate analysis, GI remained significantly associated with infection ($p<0.01$; OR 1.27; CI 1.1-1.4), while diabetes was not independently associated with infection ($p>0.05$). **Conclusion:** In the present study, the risk of infection was found to be associated with a prior diagnosis of diabetes mellitus and a higher rate of GI. GI was also identified as a predictor of infection in multivariate analysis, while diabetes was not. However, further research is necessary to gain a deeper understanding of these associations.

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Gradenigo syndrome secondary to mucormycosis in an elderly patient: a case report

Nathalye Fernanda Pedroso Dircksen[1], Marcelo Simplicio Carvalho[1], Letícia de Barros Dias[1], Lorena Fernandes Kronbauer[1], Felipe Leonardo Semensati[1], Fábio Gonçalves[1], Walton Luiz Del Tedesco Junior[1], Damacio Ramon Kaimen Maciel[1]

[1]Irmandade da Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Nathalye Fernanda Pedroso Dircksen; Email: n.athalye@hotmail.com.

Case presentation: A 63-year-old man with diabetes mellitus was admitted with headache holocranial, paresis and pain in left face of strong intensity (facial and trigeminal nerves). After 3 weeks he developed binocular diplopy horizontal (paralysis in left abducent nerve), in addition to losing 8kg during the period and brief episodes of confusion mental. The magnetic resonance imaging (MRI) in skull showed sinusopathy inflammatory and veiling in cells from the mastoid bilateral, more sharp on the left , suggesting otitis mild media/mastoiditis. The liquor was light proteinorrhaquia. He used clindamycin and ceftriaxone for 10 days, without improvement in the condition. One new skull MRI showed an area of signal alteration compromising the wall posterior sphenoid sinus on the left, sella turcica and cranial portion of the clivus, with peripheral contrast enhancement (abscess), in addition to thickening and enhancement of the plans meningeal from middle cranial fossa left. Based on the above findings, the diagnosis of Gradenigo syndrome (GS) was made and extended the antibiotic therapy to clindamycin and cefepime for 4 weeks, as well as surgical drainage due to the presence of the abscess, with culture what posteriorly pointed mucormycosis. Like this, started treatment with amphotericin liposomal per 14 days, with resolution of the condition. The patient maintained pain in the trigeminal path, with improvement after introduction in carbamazepine and gabapentin. **Discussion:** GS is composed of the triad of facial pain (trigeminal nerve), palsy abducent nerve and acute suppurateve otitis media. It is a rare complication of otitis media chronic what occurs due to petrous apicitis. The abducent and trigeminal nerves are affected due to the proximity to the petrous apex of the bone temporal. Difficultly if manifests at population elderly, mainly at it was post-antibiotic. If not treated he can cause serious complications, as paralysis of the cranial nerves, abscess intracranial, meningitis, venous sinus thrombosis, empyema subdural and death. Computed tomography is the initial diagnostic method, as it detects opacification of petromastoid air cells and bone erosions. MRI provides more details, as involvement nervous, osteomyelitis adjacent and involvement meningeal, as at the case introduced. The isolation of agent causative It is fundamental for guide the treatment, being, at majority of the cases, accomplished treatment conservative with intravenous antibiotics, if necessary tympanoplasty or myringotomy, and in refractory cases, aggressive surgical interventions such as mastoidectomy and apicectomy petrous. **Final comments:** The classic GS triad was present in less than half of the patients from the series original by Giuseppe Gradenigo. Therefore, physicians should be aware of presentations atypical, being essential the assessment radiological immediate andearly use of antibiotics for avoid complications potentially fatal.

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Graves' ophthalmopathy from the neurologist's perspective: a case report

Olívia de Mendonça e Morais[1], Danyel Augusto Sousa Castro Oliveira[1], Bárbara Ferreira Nascimento[1], Amanda Soares Pimenta[1], Luis Maximo de Araujo Junior[1], João Henrique da Matta Clementino[1], Daniela Aparecida Lima Viana[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Olívia de Mendonça e Morais; Email: d201911308@uftm.edu.br.

Case presentation: A 38-year-old woman, without previous comorbidities, presented to the Emergency Department complaining of sudden vertical deviation of the right eye concurrent with asymmetry of the upper third of the face for about 3 days. Additionally, she had right periorbital edema, ipsilateral ocular pain without visual changes for 1 week. On physical examination, she exhibited saccadic eye movements on the right, convergent strabismus to the right, and asymmetry of the midface with left eyelid ptosis. A lumbar puncture showed no abnormalities in cerebrospinal fluid, and cranial MRI revealed normal brain findings. Laboratory tests indicated suppressed TSH, elevated free T4, positive anti-thyroid peroxidase antibodies (Anti-TPO), anti-thyroglobulin antibodies (Anti-TG), and Thyrotropin Receptor Antibody (TRAb), suggesting a diagnosis of Graves' ophthalmopathy. The patient showed partial improvement with nearly imperceptible saccadic movements at discharge. She continues to be followed up by neurology and endocrinology specialists on an outpatient basis. **Discussion:** Graves' ophthalmopathy is an autoimmune disease that typically develops in cases of thyrotoxicosis or in patients with a history of hyperthyroidism; however, these ocular changes can precede or follow thyroid dysfunction by up to 18 months. It can occur at any age but is most commonly observed between 20 and 50 years old, with a predominance in women (8 times more frequent than in men). Risk factors include female sex, smoking, hypercholesterolemia, older age at onset, and radioiodine therapy. In 80% of cases, the manifestation involves 2 or more extraocular muscles without tendon involvement. The disease is associated with exophthalmos, staring gaze, diplopia, and rarely, visual loss due to compressive dysthyroid optic neuropathy. Its pathogenesis involves inflammation of periorbital soft tissues, overproduction of glycosaminoglycans by orbital fibroblasts, and adipose tissue hyperplasia. Treatment depends on disease activity (active or inactive) and may include corticosteroids, radiotherapy, or surgical therapy. **Final comments:** Based on the reported case, acute manifestations of Graves' ophthalmopathy were observed, including symptoms of pain on eye movement, conjunctival hyperemia, and unilateral periorbital edema. The described clinical picture is not typically associated with initial manifestations of hyperthyroidism caused by Graves' disease, and combined with other neurological findings, may lead to various differential diagnoses. Therefore, in the presence of evident neurological abnormalities and innocuous laboratory and imaging results, investigating toxic-metabolic causes becomes essential for diagnostic clarification.

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Guillain-Barré syndrome after dengue infection: case report

Mayara Tszesnioski Maçaneiro[1], Caio de Lima Ferreira[1], Giuliana Moro[1], Klaus Johann Jacques Schebek Teixeira[1], Felipe Ferreira de Almeida[1], Arthur Stedile Ribeiro Pacheco[1], Giulianna Luzia Pereira[1], Marcos Silvio Gomes[2], Felipe Ibiapina dos Reis[1]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Mayara Tszesnioski Maçaneiro; Email: mayaratm.tsz@gmail.com.

Case presentation: An 83-year-old female presented to the Emergency Department with speech alteration, sphincter release, and a fall within the last 24 hours. Examination revealed left-sided hemiparesis, hypoesthesia, and disorientation. Her medical history included systemic arterial hypertension, hypothyroidism, and dyslipidemia, for which she was taking levothyroxine, simvastatin, and losartan. A preliminary diagnosis of stroke in the anterior cerebral region was made, along with a suspicion of Dengue B. Cranial computed tomography (CT) and angio-CT showed no acute lesions or cerebral artery occlusions. She was placed on antiplatelet therapy with clopidogrel and monitored in an acute stroke unit.

Laboratory confirmed positive NS1 and a platelet count of 81,000; hence, Clopidogrel was discontinued due to thrombocytopenia. On the fifth day of hospitalization, her neurological deficits worsened, and she developed dyspnea, dysarthria, dysphagia, and tetraparesis. Guillain-Barré syndrome (GBS) was suspected, and further investigation was initiated. Cerebrospinal fluid (CSF) analysis confirmed the diagnosis of GBS by showing albuminocytologic dissociation (1 cell, 72 proteins). Treatment with immunoglobulin was initiated. Despite immunoglobulin treatment, the patient remained globally weak, necessitating plasmapheresis as an additional treatment. During the treatment, the patient required mechanical ventilation and received Meropenem for ventilator-associated pneumonia. After 48 days post-GBS diagnosis, the patient achieved clinical stability and discharged to a rehabilitation facility. **Discussion:** Dengue, an acute febrile infectious disease endemic in tropical regions, had over 5.2 million cases in Brazil in 2024 transmitted by the mosquito Aedes aegypti, with symptoms ranging from mild (fever, headache, myalgia, arthralgia, rash, and retro-orbital pain) to severe, with 1-25% potential neurological complications like GBS. GBS, an acute polyradiculoneuropathy, often follows infections or vaccination, with a mortality rate of 3-10%. It involves autoimmune mechanisms damaging nerve structures, presenting as ascending muscle weakness and hyporeflexia. Diagnosis includes clinical evaluation, CSF analysis, electromyography, and imaging. Treatment comprises immunoglobulins and plasmapheresis. The association between GBS and dengue is rarely documented in the literature. Nevertheless, GBS can occur as a neurological complication of dengue fever, especially with DENV-2 and DENV-3 serotypes. Inflammatory cytokines from the immune response to dengue may increase blood-brain barrier permeability, contributing to GBS pathogenesis. Moreover, cross-reactive IgM or IgG antibodies against flavivirus antigens are more common in GBS patients. **Final comments:** GBS, characterized by muscle weakness and immune system involvement, can occur post-infection, with dengue being one of the triggering viruses.

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Guillain-Barré syndrome after dengue virus infection: an unusual association

Luíza Toledo de Castro[1], Henrique Bernal[1], Ana Flávia Pincerno Pouza[1], Herval Ribeiro Soares Neto[1], Sônia Maria Cesar de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Luiza Toledo de Castro; Email: luizavga@hotmail.com.

Case presentation: A 60-year-old man presented walking difficulty five days after dengue fever due to imbalance sensation. He also referred to lower limbs ascending numbness. Three days later, symptoms progressed to the upper limbs.Within one week after symptoms onset, he could not walk without assistance. Neurological exam revealed four limbs proximal weakness (grade 4/5), global areflexia and lower limbs dysesthesia. Regarding deep sensation, he showed a significant impairment: distal hypopalesthesia, sensory ataxia, pseudoathetosis and sensory ataxic gait. Complementary exams showed protein-CSF dissociation (2 cells and 61 proteins). Electrophysiological study showed primarily demyelinating sensory-motor polyneuropathy. Intravenous immunoglobulin (2g/kg) was started and a significant improvement was obtained: the patient was able to walk again. (...) At the outpatient follow-up visit, the patient reported improvement in muscle strength, in addition to progressive improvement in gait. Keeps discreet tingling and hands and feet. It shows the result of a new electroneuromyography performed 40 days after the onset of symptoms, which reveals: non-uniform sensory and motor polyradiculopathy, primarily demyelinating, subacute, with no signs of recent activity. Neurophysiological improvement compared to previous examination. **Discussion:** Guillain-Barré syndrome (GBS) is an acute immune-mediated polyradiculoneuropathy that typically occurs 2 to 8 weeks after a viral or bacterial infection. Dengue virus is an uncommon trigger to SGB. Regarding neurological manifestations due to dengue infection, DENV-2 and DENV-3 are the most associated. Motor function is usually most affected, in an ascending pattern over a period of up to 4 weeks. Areflexia, sensory disturbances, and cranial nerve involvement may also occur. Symptoms onset usually occurs after two weeks from the infection, but may also start concomitantly with dengue fever. Neurological manifestations are due to direct invasion, immune-mediated reactions or metabolic changes. Diagnosis and treatment adhere to the same principles as those for SGB in other contexts**. Final comments:** Although rare, arboviral infections can precede SGB as well as other viruses. This case highlights the importance of topographical diagnosis, leading to recognition of acute sensory-motor demyelinating polyneuropathy triggered by an uncommon cause. Early treatment enabled an almost complete improvement, thereby reducing disease morbidity.

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Guillain-Barré syndrome in children: a 10-year retrospective analysis of a Brazilian electrophysiology diagnostic center archive

Rafael Gomes Costa[1], Fernando Cavalcanti de Sá e Benevides Falcão[1], Renata Barbosa Paolilo[1], José Pedro Soares Baima[1], Carlos Otto Heise[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Rafael Gomes Costa; Email: rgc0711@gmail.com.

Background: Guillain-Barré syndrome (GBS) is the leading cause of acute flaccid paralysis in children globally following the eradication of poliomyelitis. Recent studies indicate a potential bimodal age distribution among infants, with the first peak occurring in preschoolers around 2 years old. In young infants, the syndrome may present as hypotonia or motor delay. While lumbar puncture and nerve conduction studies are crucial diagnostic tools, challenges include the child's cooperation and the technical expertise required for performing and interpreting these tests. Mortality rates in children can range from 7-12% in African, Asian, and Latin American populations, compared to zero in European countries. **Methods:** This study was conducted at the Electrophysiology Department affiliated with the Neurology Unit of a major reference center in Brazil. We reviewed all electrophysiological studies conducted between 2013 and 2023 to identify cases suspected of GBS. Patients under 18 years old were included, and their medical records were analyzed to confirm GBS diagnosis according to Brighton's criteria. Electrophysiological studies were conducted for all patients. We also examined the correlation between clinical severity, pathogenic subtypes, and other clinical features. **Results:** Out of 51 exams initially suspected of GBS, 35 were confirmed as GBS cases. The mean age was 8.6 years, with a slight male predominance (51%). A potential triggering event preceding GBS was identified in 82% of patients. Most patients required hospitalization, and half of them were admitted to the ICU. The mean EGRIS score at admission was 2.34, and approximately 68% of patients experienced loss of walking ability before or during their hospital stay. Electrophysiological classification revealed that 60% had the demyelinating variant, while 34% had the axonal form. Three patients showed reversible conduction failure. No statistical difference was found between severity variables and electrophysiological variants. **Conclusion:** In low-income settings, nerve conduction studies are not always necessary for diagnosing and treating GBS. Our study, based on data from an electrophysiology department database, found over a quarter of cases with at least one clinical atypical feature, differing from studies based on broader inpatient databases, certainly representing one of the reasons why the exam is requested by the clinician at first place. The pathogenic subtype does not appear to significantly influence the disease's clinical course, aligning with global literature. Reversible conduction failure may provide new insights into prognosis estimation.

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Guillain-Barré syndrome succeeding dengue infection: a case series of three patients

Mayko Nascimento Merscher[1], Gustavo Freitas Alves de Arruda[1], Lucas Vieira Pinto[1], Gabriel Costa dos Reis[1], Alice Jardim Zaccariotti[1], Laura Araújo Paulino[1], Luiz Fernando de Oliveira Urzeda[1], Camila Caroso Lobo[1], Marcio Luiz Figueredo Balthazar[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Mayko Nascimento Merscher; Email: maykomerscher@gmail.com.

Case presentation: CASE 1: A 55-year-old female presented with a 4-day history of headache, myalgia and fever, with a positive IgM serological test for dengue. Nine days after the symptoms onset, she reported numbness, weakness, inability to walk and slurred speech. The physical exam highlighted proximal weakness of upper and lower limbs, global areflexia, bilateral peripheral facial palsy, hypoesthesia, and Laségue sign. Cerebrospinal fluid (CSF), collected in the first week of clinical course, had no abnormalities. CASE 2: A 52-year-old female started a clinical condition of headache, low back pain and lower limbs pain associated with a diagnosis of dengue through a positive IgM test. After 5 days, she developed weakness, inability to walk, burning sensation in feet, and feeding difficulty. On examination, it was found global weakness and areflexia. CSF evidenced albuminocytological dissociation. CASE 3: A 39-year-old female with a history of headache, myalgia, fever and retro-orbital pain, along with positive IgM for dengue, presented after a few days with weakness, sensitivity symptoms, and urinary retention. The neurological exam demonstrated global weakness, areflexia, hypoesthesia, bilateral peripheral facial palsy, dysphonia, and dysphagia. Soon after, she developed the need for mechanical ventilation due to the rapid evolution of the disease with respiratory failure. CSF confirmed albuminocytological dissociation. **Discussion:** Dengue is a viral disease transmitted by infected Aedes aegypti mosquitoes in Brazil, which are also vectors for the transmission of other arboviruses such as zika and chikungunya. Neurological manifestations due to dengue vary between 0.5-21% and represent both direct and indirect complications. Among the possible manifestations, one of the most common is Guillain-Barré syndrome (GBS), which may also occur after oligosymptomatic cases of dengue. In this case series, 3 female patients with a previous dengue infection developed different clinical manifestations, but all of them presented cranial nerves involvement somehow. Only one didn't present protein-cytological dissociation. All were treated with human immunoglobulin and improved during and after the end of treatment. **Final comments:** Through greater serological test availability, it is possible not only to identify dengue cases, but also to better characterize associated neurological complications and its frequency, guiding strategies for mapping and preventing new cases. As it is an endemic disease in Brazil, the description of GBS in oligosymptomatic cases of dengue should draw attention, as the increase of GBS cases without a clear history of previous infection or vaccination might be related to undiagnosed dengue. Therefore, it is essential that risk areas for dengue have the availability of confirmatory exams and also public policies to act in transmission foci of its vector to interrupt the transmission chain, and consequently its complications.

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Guillain-Barré syndrome: an uncertain etiology between influenza and acute contagious gastroenteritis - a case report

Isabela da Costa Rodrigues[1], Lucas Victoy Guimarães Zengo[1], Ivo Marcos Darella Lorenzin Fernandes Neto[2], Eduarda Basso Badalotti[2], Angelo Marcelo Wosniacki Filho[2], Gabriel Angelo Garute Zenatti[2], Renato Endler Iachinski[2], Antonio Carlos de Andrade Soares[2]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Fundação Hospitalar São Lucas, Cascavel PR, Brazil.

Correspondence: Isabela da Costa Rodrigues; Email: isacostarod.2001@gmail.com.

Case presentation: Male, 71 years old, with a bitter taste in his mouth 16 days ago, which progressed to dysarthria and dysphagia for solids, as well as cruralis tetraparesis predominantly, leading to loss of the ability to walk and difficulty holding objects. History of self-limited diarrhea 45 days ago, influenza vaccination 27 days ago, constipation 8 days ago, and significant eye tearing 1 day ago. He have controlled hypertension and social alcohol use. On physical examination: symmetrical facial diplegia with bilateral Bell's sign, global paresis (3+/5 in the lower limbs and 4+/5 in the upper limbs), areflexia in the lower limbs, and hyporeflexia in the upper limbs. MRI of the cranio-caudal axis with no abnormalities. After cerebrospinal fluid collection, due to the high protein concentration observed in the result, the CSF appeared visibly frothy, clear, and slightly xanthochromic, with albumin-cytological dissociation (total protein: 174mg/dL with 0.5cells/μL), indicating the possibility of Guillain-Barré syndrome (GBS), and treatment with immunoglobulin for 5 days was initiated. During the hospitalization, the patient did not show improvement or progression of symptoms. The etiology is still not clearly defined, suggesting that it could have been caused either by recent diarrhea or by a previous vaccination history. Ophthalmologic evaluation revealed blepharitis in both eyes and a nasal pterygium in the right eye. Additionally, due to dysphagia, a nasoenteral tube feeding diet was chosen. **Discussion:** GBS is an acute polyneuropathy whose symptoms include progressive, ascending motor weakness and, in most cases, is secondary to an infectious pathology. The most feared complication of GBS is respiratory failure, caused by bulbar weakness. Treatment with intravenous immunoglobulin (IVIG) is one of the main therapeutic approaches for GBS. IVIG therapy is preferred due to its rapid action and lower incidence of serious side effects compared to plasmapheresis. In many cases, the pathological agent remains unidentified. In patients who had gastroenteritis, infection by Campylobacter jejuni are especially common. An increased risk of GBS following influenza infection, and it´s vacination. **Final comments:** This case illustrates a presentation of GBS and the challenges in its management. Despite timely IVIG treatment, the patient showed no improvement, highlighting the need for close monitoring and possible additional interventions. Albumin-cytological dissociation in the CFS supported the GBS diagnosis, underscoring the importance of comprehensive diagnostic evaluation. Determining the exact cause of GBS is often challenging. Cases following respiratory infections tend to have better outcomes compared to those following gastrointestinal infections like Campylobacter jejuni. It's crucial to monitor GBS cases post-influenza vaccination closely due to potentially worse prognoses and more severe clinical conditions at admission.

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GYG1 gene mutation causing a mild phenotype of polyglucosan body myopathy type 2

Ana Carolina Leite Ribeiro[1], Bárbara Pina Aiello[1], Roberto Pereira Santos[1], Caio Faria Tardin[1], Matheus Cecatto[1], Rhanice Soares Huguenin Câmara[1], Amanda de Morais Affonso[1], Marcos Raimundo Gomes de Freitas[1], Victor Evangelista Rodrigues Pereira[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Ana Carolina Leite Ribeiro; Email: anacarolinaneuro@gmail.com.

A 30-year-old man presented to the neuromuscular disease clinic reporting post-exercise myalgias and hyperCKnemia. His condition was first noted at the age of 20 when he experienced myalgia and muscle weakness after swimming. The patient was, on occasion, referred to the emergency department, where laboratory tests revealed a creatine kinase (CK) level of 70.000, and he was diagnosed with rhabdomyolysis. He denied having had an acute kidney injury at that moment and reported that the CK eventually returned to basal levels with rest. The patient is a doctor and kept track of his CK levels through the following years. He noticed that he often experiences hyperCKemia, frequently asymptomatic, following physical exercise. The patient reported that his brother had three episodes of rhabdomyolysis during his life. Neurological examination was normal during the consult, and a genetic panel for rhabdomyolysis was requested for diagnosis, which revealed a potentially pathogenic heterozygous variant in the GYG1 gene - NM_004130.4(GYG1):c.304G>C (p.Asp102His), associated with myopathy with polyglucosan bodies type II (PGB2). The GYG1 gene encodes the isoform glycogenin-1, a protein involved in glycogen biosynthesis. Pathogenic variations in the GYG1 gene may result in reduced, absent, or functionally abnormal glycogenin-1 protein, leading to defective glycogen synthesis, glycogen depletion in skeletal muscle, and abnormal glycogen accumulation in cardiac muscle. Homozygous or compound heterozygous variants in GYG1 are associated with PGB2, a genetic disease characterized by different clinical phenotypes, ranging from exercise intolerance to progressive proximal weakness with cardiac involvement. The clinicopathological spectrum of GYG1 disease is rapidly expanding. Most cases illustrate a slowly progressive proximal myopathy with or without cardiac involvement starting in adulthood. However, distal, asymmetric, or isolated facial weakness, fluctuating weakness, exertional myalgia, and exercise intolerance are also described. Our patient experienced recurrent episodes of exercise-induced rhabdomyolysis associated with a probably pathogenic heterozygous variant in the GYG1 gene, probably representing a milder clinical phenotype of PGB2. He was instructed to avoid intense physical exercises, and genetic counseling was advised. Heterozygous variants of the GYG1 gene may present with a mild clinical phenotype of PGB2 disease. This case highlights the need to correlate clinical phenotypes and gene variants while investigating possible genetic diagnosis because mutations in the same gene may present with a broad range of clinical spectra.

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HACD1-related congenital myopathy in four unrelated patients

Alexandre Motta Mecê[1], Cristina Iwabe[1], Alberto Rolim Muro Martinez[1], Felipe Augusto Moreschi[2], Luis Fernando Glossklauss[3], Anamarli Nucci[1], Marcondes Cavalcante França Júnior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Hospital Pequeno Príncipe, Curitiba PR, Brazil.
[3]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Alexandre Motta Mece; Email: amottamece@gmail.com.

Case presentation: Case 1. A 54-years-old male underwent to investigation of a congenital myopathy. A delayed motor milestones was informed, with unsupported walk at age 7. Progressively he noted improvement of motor skill, never up to normal. Myalgia was the prominent complain. Examination revealed upper and lower limb girdle weakness (MRC=4), mild hypotonia and hyporeflexia. Motor function measure (MFM) scale total score=83.33%; Dimension (D) D1=69.23%; D2=91.66%; D3=95.23%. CK and cardiac evaluation was normal. EMG showed myopathic pattern and biceps brachii biopsy diagnosed fiber type disproportion. His parents were consanguineous; he had a brother dead in infancy due to muscle disease. Genetic panel showed a biallelic pathogenic variant in HACD1 gene c.373_375+2delsplicesite. Case 2. A 31-years-old female was investigated because weakness and diffuse myalgia. She had to stay in an intensive care unit (ICU) for several days since birth as hypotonic child and walked after age 5. On examination: predominant proximal weakness, myopathic gate, mild diffuse hypotonia, moderate atrophy and hyporeflexia. MFM scale total score=69,79%, D1=33.33%; D2=91.66%; D3=100%. Genetic panel showed homozygotic pathogenic variant in HACD1 gene (c.375_377+2delsplicesite). Patient has consanguineous parents, no similar case in the family. Case 3. A 15-years-old male born after a normal gestation and delivery required an ICU for survival due to hypotonic syndrome. Motor milestones were mild delayed, and several pulmonary infections occurred in infancy. Under motor and pulmonary physiotherapy, he had a good evolution. CK exam was normal. A muscle biopsy showed fiber type disproportion and genetic panel detected c.373_375+2delGAGGT homozygous pathogenic variant in HACD1 gene. Case 4. A 5-year-old male born with Apgar 5 and 6, after normal gestation and delivery required an ICU stay. Evolution resulted in mild motor delay but normal psychological milestones. Examination showed: myopathic gate, global hypotonia and hypotrophy and, normal stretch reflex. MRC was 4+ in arms and 4- in proximodistal muscles of legs. CK and brain MRI were normal. A genetic panel for detected a biallelic variant c.373_375+2delGAGGT in HACD1 gene. His parents have no consanguinity and there is no similar case in the family. **Discussion:** We present four unrelated patients with congenital myopathy caused by homozygous pathogenic variants in HACD1 gene. Clinical main feature was severe hypotonia forcing early life stay in ICU in 3/4 patients. Evolution was benign in all cases. Myalgia was prominent in oldest patients. CK was unremarkable; 3/4 patients' fiber type disproportion was the morphological hallmark. **Final comments:** HACD1-congenital myopathy is a rare disease, described since May/2024 in 13 patients of Asian ancestry. Our presentation highlights the necessity of think in this causative gene in Caucasian patients, whereas none of our patients had oriental origin.

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Haloperidol versus atypical antipsychotics in the management of delirium: a systematic review and meta-analysis of randomized controlled trials

Murilo Marmori Cruccioli[1], Jorge Ferreira Jasmineiro Pitanga[2], Milena Lima Loures[3], Pedro Fraiman[4], Regina Carneiro Leão[5], Caio Rezende Lima[6], Camila Carneiro Ferreira[7], Filipe Sarmento[8]

[1]Universidade Evangélica de Goiás, Anápolis GO, Brazil.
[2]Universidad de Buenos Aires, Buenos Aires, Argentina.
[3]Faculdade Ciências Médicas de Minas Gerais, Belo Horizonte MG, Brazil.
[4]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[5]Universidade Federal de Pernambuco, Recife PE, Brazil.
[6]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

[7]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[8]University of Florida, FL, US.

Correspondence: Murilo Cruccioli; Email: murilocruccioli@gmail.com.

Background: Delirium is a common neurocognitive disorder in medically ill patients, affecting at least one in seven hospitalized patients overall and around half of Intensive Care Unit (ICU) patients. It is characterized by an acute disturbance in attention, awareness, and cognition that tends to fluctuate in severity during the course of the day and is not better accounted for by a pre-existing neurocognitive disorder. Delirium treatment is complex. There is frequent use of antipsychotics in clinical practice despite insufficient evidence. Haloperidol, a first-generation antipsychotic, is often the first-line pharmacological treatment for delirium, particularly for agitation-related events, despite being commonly associated with a higher rate of adverse effects, such as extrapyramidal symptoms (EPS). Second-generation antipsychotics, or atypical, generally have a better side effect profile and have been evaluated for their usefulness in treating delirium. However, studies comparing haloperidol and atypical are limited. **Objective:** To perform a systematic review and meta-analysis of randomized controlled trials (RCTs) comparing haloperidol and atypical antipsychotics in the management of patients with delirium. Our primary objective was to compare their effectiveness, while the secondary objective was to assess the occurrence of extrapyramidal symptoms. **Methods:** We conducted a systematic search of PubMed, Scopus, and the Cochrane Central Register of Controlled Trials for RCTs. The outcomes of interest included overall mortality, changes in delirium scale scores, and the incidence of extrapyramidal symptoms. Due to the use of different delirium scales across studies (DRS-R-98, MDAS, and CGI-S), we used the standard mean difference (SMD) to evaluate this outcome. This approach allowed us to compare effect sizes across studies that used different scales. Statistical analysis was performed using RevMan 5.4, and heterogeneity was assessed using I^2 statistics. **Results:** Our analysis included 12 RCTs with a total of 1,510 patients (57% male), of whom 41.8% received haloperidol. Delirium severity (SMD, -0.03 [95% CI, -0.20 - 0.15]; p=0.78) and overall mortality (OR, 1.26 [95% CI, 0.88 - 1.80]; p=0.20) did not differ significantly between groups. However, patients treated with haloperidol had a significantly higher incidence of extrapyramidal symptoms compared to those treated with atypical antipsychotics (OR 2.72, 95% CI 1.26-5.87; p=0.01). **Conclusion:** The results of our meta-analysis suggest that overall mortality rates and changes in delirium assessment scales do not differ significantly between patients treated with haloperidol and those treated with atypical antipsychotics. However, haloperidol is associated with significantly higher odds of experiencing extrapyramidal symptoms, indicating a potential advantage of atypical antipsychotics in minimizing adverse effects.

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Has Creutzfeldt-Jakob disease become more frequent in Brazil? A case report and mini literature review

Priscila Januário de Oliveira[1], Bruna Grando Kroeff[1], Marcelo Savio Paiva do Amaral[1], Giorgio Fabiani[2], Giuliana Carpentieri Pipolo[1], Silvia Escarlate Andersen[1], Monica Jaques Spinosa[1], Natália Paschoal Divino[1], João Guilherme de Toledo Justo[1], Thabata Baechtold da Silveira[1]

[1]Hospital Nossa Senhora do Rosário, Serafina Corrêa RS, Brazil.
[2]HAC.

Correspondence: Priscila Januario de Oliveira; Email: joliveira.pri@gmail.com.

We want to report the case of a 61-year-old man from the city of Siqueira Campos, State of Parana - Brazil, with no significant medical history, presented with a 3-month history of generalized weakness, a prominent gait disturbance - "looking like he was drunk," as his daughter said -, and memory loss. He went to see a neurologist and started taking medication for labyrinthitis and antidepressants as well, without any symptomatic improvement. His mental and cognitive state decreased one month into his symptomatic course, and he progressed to temporospatial disorientation, hallucinations, and incoherent speech. He had also developed diffuse myoclonic jerks. He was admitted to the Neurological Department of the tertiary referral hospital, and the investigation included a Brain MRI that demonstrated symmetric T2 and fluid-attenuated inversion recovery (FLAIR) caudate, putamen, and cortical hyperintensities. An EEG showed a diffuse slowing (7Hz) with a normal posterior dominant rhythm and a generalized pattern in frontal regions with periodic sharp-wave complexes that represented 5% of the total activity. The CSF analyses excluded CNS inflammation, and a sample was sent to the specialized clinical laboratory out of the state for research of protein 14-3-3, that was elevated. During the hospitalization, the mental state deterioration continued until he lapsed into a coma, and ventilatory support was needed. He died approximately 8 months after the onset of the symptoms. The Creutzfeldt-Jakob disease is a prion disease and the most common type of spongiform encephalopathy. This protein, cellular prion protein (PrPc) and its misfolded form, prion protein scrapie-type (PrPSc), lead to neurodegeneration. It was described in 1920 by Hans Gerhard Creutzfeldt and later, Alfons Maria Jakob. In humans, the sporadic form (sCJD) is seen in most cases, but other forms are described such as: sporadic fatal insomnia (sFI) and variably protease-sensitive proteinopathy, genetic forms due to mutations in PRNP (familial CJD) or genetic CJD, Gerstmann-Straussler-Scheinker syndrome and fatal familiar insomnia, acquired forms (as iatrogenic CJD), Kuru and variant CJD. The annual incidence is one to two cases per million people. The diagnostic criteria for probable sCJD include a rapidly progressive cognitive impairment and two of the neurological signs: visual or cerebellar disturbance, pyramidal or extrapyramidal signs, myoclonus or akinetic mutism, plus at least one biomarker criterion: elevated levels of protein 14-3-3 in CSF, or MRI or EEG. Or a progressive neuropsychiatric syndrome and a positive PsPSc seeding activity (RT-QuIC positive in CSF or Other fluids or tissues). CJD is always fatal, and most patients die in one year or less. Between 2005 to 2021, 1,576 notifications of CJD suspected cases were registered in Brazil. The states with the highest number of cases reported in the period were São Paulo with 32.5%, Paraná with 12.0% and Minas Gerais with 10%.

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Headache and epilepsy in a female young patient: MELAS

Ruan Gambardella Rosalina de Azevedo[1], Gabriel Pinheiro Martins de Almeida e Souza[1], João Victor Cabral Correia Férrer[1], Márcio Pinheiro Lima[1], Victor Cardoso de Faria[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Ruan Gambardella Rosalina de Azevedo; Email: ruangambardella@gmail.com.

This case reports a 21-year-old female with stroke-like events and seizures. At eighteen and nineteen years-old she experienced episodes of migranous headache followed by impared awareness motor onset focal seizure to bilateral tonic-clonic. After the last (fifth) event, she complained of left temporal visual loss and anomic aphasia. Physical examination was also marked by: incomplete, disproportionate, left hemiparesis, Babinski and hypoesthesia on left lower limb and bilateral hearing loss. There were no signs of: ocular motricity impairment, dystonia, parkinsonism, myoclonus. Laboratory tests resulted normal for renal, thyroid and liver function. There were normal blood levels of inflammatory markers and ANA test was negative. Ammonia arterial blood level was high two months after the last seizure. Cerebral spinal fluid and blood arterial lactate were high at the follow up. Brain magnetic resonance image described water restriction signs in the cortical aspects of left: inferior parietal lobe, posterior insula and superior temporal gyrus. There were also minor bilateral occipitotemporoparietal subcortical T2/FLAIR alteration signs. Thus, the images were compatible with more than one vascular territory. There were unremarkable changes on echo/electrocardiogram. Electroencephalogram revealed bilateral occipitotemporal paroxysmal activity with right predominance. Audiometry revealed moderate bilateral sensorineural hearing loss. The genetic testing was positive for m. 3243 A>G heteroplasmy mutation, compatible with Mitochondrial encephalomyopathy, lactic acidosis, and stroke-like episodes (MELAS). This disease results in impaired mitochondrial translation and protein synthesis including the mitochondrial electron transport chain complex subunits leading to impaired energy production. The inability to generate sufficient energy to meet the needs of various organs results in the multi-organ dysfunction observed. At the follow up, the patient is using coenzyme Q10, carnitine, folic acid, riboflavin and lamotrigine (400mg/day), tolerating mild physical exercises without new stroke like events for three years.

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Health profile of elderly people with probable cognitive impairment of primary health care

Tainá Reis Martins[1], Wívian Maria Pires Figueredo[1], Ana Luíza Braga e Silva[1], Luciana Colares Maia[1]

[1]Centro Universitário FIPMoc, Montes Claros MG, Brazil.

Correspondence: Tainá Reis Martins; Email: taina2002@hotmail.com.

Background: Population aging is a reality in Brazil and in the world, generating a need for promoting healthy aging. The change in the epidemiological profile determines that chronic non-communicable diseases (NCDs), such as dementia, are more frequent. Therefore, cognitive decline is a condition ranging from mild cognitive decline to major cognitive impairment or dementia syndrome. **Objective:** To analyze the health profile of the elderly with evidence of cognitive impairment in Primary Health Care (PHC). **Methods:** This is a cross-sectional study conducted in PHC in northern Minas Gerais. Sampling was performed in two stages and by clusters. Initially, the regional health centers of the municipality were drawn and, in each pole, the teams of the Family Health Strategy (FHS) and respective areas of coverage were drawn. 201 elderly people with probable cognitive decline were evaluated, and the Mini Mental State Examination (MMSE) was used for cognitive screening. It is reiterated that the research was approved by the Ethics and Research Committee - Opinion n. 1.628.652. **Results:** We identified 142 women (70.6%) and 59 men, it is noteworthy that the age ranged from 60 to 74 years (41.8%), 75 to 84 years (23.9%) and 85 years and over (34.3%). Through the Clinical Functional Vulnerability Index (CFVI-20), 70.6% (142) were considered fragile, that is with loss of functionality. In addition, 29.9% (60) had urinary incontinence, 53 elderly (31.9%) smoked, and 19.4% (36) were obese. Diseases such as diabetes (21.4%), insomnia (46.3%) and hypertension (67.7%) were also observed in the study population. **Conclusion:** It was found that the profile of the elderly with probable cognitive impairment of the research was characterized

by being the majority of women aged between 60 and 79 years, presenting risk of frailty by the CFVI-20 and also associated diseases such as obesity, diabetes, insomnia, fragility and hypertension. Therefore, cognition can be defined as a predictor of well-being, and its impairment can affect functional capacity in old age, in addition to providing opportunities for loss of autonomy and independence. The importance of NCDs control and the development of promotion strategies, as well as health prevention are essential for healthy and quality aging.

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Healthcare journey for patients with amyotrophic lateral sclerosis (ALS) in a specialized service in Salvador-Bahia

Marie Lidio dos Santos Galvão Ribeiro[1], Ariele Amaral Alves[1], Laís Brandão Mota[1], Luna Souza Silva Vilanova[1], Rafael Amaral Albuquerque[1], Têmis Maria Félix[2], Marcela Câmara Machado-Costa[1], Marilaine de Matos Menezes Ferreira[1], Debora Borges dos Santos Pereira[3], Thiago Godoy de Oliveira[3]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[3]MAPESolutions, São Paulo SP, Brazil.

Correspondence: Marie Lidio dos Santos Galvão Ribeiro; Email: marielidiogr@gmail.com.

Background: The JAV-RARAS study is an integrated initiative within the Brazilian Rare Diseases Network (RARAS), with the objective of measuring the Value of Healthcare Journey (JAV) of patients with rare diseases in Brazil. **Objective:** To evaluate the healthcare journey for patients with Amyotrophic Lateral Sclerosis (ALS) in a specialized service. **Methods:** Patients were followed through the collection of data from medical records and clinical questionnaires. The data obtained was compared to the Clinical and Therapeutic Guidelines (PCDT) defined for ALS, developed by the Brazilian Ministry of Health. **Results:** A total of 30 patients were included in the study, although 2 patients had not undergone any diagnostic tests and 4 patients had no information regarding the types of tests that had been performed. The sample was analyzed for a total of 24 patients. The principal diagnostic tests conducted were as follows: 1. Neurological tests, such as electromyography of four limbs (EMG), magnetic resonance imaging (MRI) of the lumbar and thoracic spine, MRI of the brain and craniocervical junction, and cerebrospinal fluid analysis. 2. Laboratory tests, such as Complete blood count (CBC); measurements of creatinine, alanine aminotransferase (ALT), aspartate aminotransferase (AST), C-reactive protein (CRP), and urea; prothrombin time; and serum protein electrophoresis. **Conclusion:** In accordance with the PCDT for ALS, complementary tests must be conducted to support the diagnosis of ALS and to rule out other systemic disorders that may mimic it. For an accurate diagnosis, every patient suspected of having ALS must undergo complementary tests with results consistent with the disease. These include EMG of four limbs showing denervation in more than one segment and normal motor and sensory nerve conduction (except when there is an associated polyneuropathy); complete blood count, urea, creatinine, ALT, AST, and prothrombin time should be normal. In select instances, at the discretion of the attending physician, the following tests may be requested: An MRI of the brain and craniocervical junction, which should not show any structural lesion explaining the symptoms. Additionally, measurement of the concentration of CRP and serum protein electrophoresis is recommended. These tests should yield normal results. Following a comprehensive analysis, it can be confirmed that the service adheres to the PCDT recommendations for accurate ALS diagnosis. A comparison of this information with the tests performed on the patients in the sample revealed that all recommended diagnostic tests in the PCDT were performed (EMG, MRI, cerebrospinal fluid analysis). Additionally, all recommended laboratory tests were also performed (CBC, renal function, ALT, AST, prothrombin time, CRP, protein electrophoresis). The performance of these tests demonstrates the team's effort to investigate and characterize the clinical manifestations of ALS in this group of patients.

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Healthcare professionals perspectives on palliative care for stroke patients: the passar study

Thales Pardini Fagundes[1], Túlio Maranhão Neto[1], Lucas Gondim Briand Vieira[1], Victor Augusto Zanesi Maciel[1], Juliana Pelegrin[1], Mateus Gustavo Favaro[1], Millene Rodrigues Camilo[1], Octávio Marques Pontes-Neto[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Millene Rodrigues Camilo; Email: millene.camilo@gmail.com.

Background: Palliative care should be addressed to all patients with severe or life-threatening stroke. However, its implementation remains challenging despite guidelines. Understanding healthcare professionals' attitudes and practices regarding palliative care is essential for developing targeted interventions to improve palliative care delivery in stroke management. **Objective:** To explore healthcare professionals' perceptions and practices regarding palliative care for stroke patients, focusing on differences in symptom management and care strategies based on professional experience. **Methods:** A cross-sectional, questionnaire-based study was conducted from August to October 2023. One hundred twenty-four healthcare professionals (47 neurologists, 77 other professionals) with at least one year of stroke care experience were included. Data were collected via online questionnaires and analyzed descriptively. **Results:** (1) Demographics: Among neurologists, 55.32% were male with a median age of 29, while 72.73% of other professionals were female with a median age of 31. 38.3% of neurologists and 15.58% of other professionals had one year of stroke care experience. (2) Palliative Care Experience: 55.32% of neurologists and 53.25% of other professionals reported participating in a few palliative care discussions, while 23.4% of neurologists and 36.36% of other professionals had no contact with the topic. (3) Assessment Tools: On a 5-point scale, neurologists reported using the SPICT tool with a mean score of 1.94, and the PPS scale with a mean score of 2.06. Other professionals scored 2.01 and 2.13 respectively. (4) Patient-centered Care: Neurologists reported asking about patients' illness expectations with a mean score of 3.72 out of 5, compared to 3.08 for other professionals. (5) Symptom Management: On a 5-point knowledge scale, neurologists reported the highest competence in pain management (mean 2.7) and the lowest in hypodermoclysis (mean 1.9). (6) Clinical Decision-making: On a 5-point comfort scale, neurologists felt most comfortable not conducting serial laboratory tests (mean 2.72) and least comfortable performing palliative extubation (mean 2.1). (7) Multidisciplinary Approach: Neurologists reported providing multidisciplinary team support with a mean frequency of 3.8 out of 5 and requesting specialized palliative care team evaluation with a mean of 3.2 out of 5. (8) Spiritual Care: Neurologists offered spiritual support to patients with a mean frequency of 2.4 out of 5, and to families with a mean of 2.2 out of 5. **Conclusion:** The study reveals significant variations in the palliative care approach based on professional background and experience level. These findings highlight the need for targeted education and interdisciplinary collaboration to improve palliative care delivery in stroke management.

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Heidenhain variant of Creutzfeldt-Jakob disease hiding in plain sight: clinical evolution and sequential MRI analysis of brain degeneration

Marco Antônio Delazeri[1], William Alves Martins[1], Lucas Porcello Schilling[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Marco Antônio Delazeri; Email: marco.delazeri@outlook.com.

A 65-year-old woman presented to our service complaining of dizziness and vertigo lasting two weeks. She exhibited horizontal nystagmus and mild unsteadiness of gait. Initial MRI findings were negative for lesions, and she was prescribed anti-vertiginous medications. However, subsequent evaluation due to progressive cognitive decline revealed focal left parieto-occipital hypometabolism on PET scan. During examination, she displayed mild nystagmus, ataxia, and an upper right visual deficit. A week later, she began stumbling on furniture at home, particularly on the right side, attributing it to blurred vision. Re-examination of the initial MRI revealed slight diffusion-weighted imaging (DWI) restriction in the left occipital region, which progressed in subsequent MRI. She was adimitted for further investigation, with cerebrospinal fluid (CSF) studies showing mildly elevated protein levels. Shortly after, she started exhibiting paraphrastic errors (substituting simple words) and behavioral disturbances. This progressed to extreme multifocal myoclonus triggered by light touch, sound, and post-stimulation, accompanied by an intense startle response, aphasia, and bedridden status. A new Protein 14-3-3 returned negative. Initial EEG findings indicated a slowed background evolving into periodic generalized discharges. Intravenous immunoglobulin (IVIG) and methylprednisolone pulse therapy for suspected autoimmune encephalitis proved unsuccessful. Eventually, a diagnosis of probable Creutzfeldt-Jakob disease (Heidenhain variant) was made. Symptomatic medications partially alleviated the myoclonus. Follow-up MRIs demonstrated severe progression of atrophy and leukoencephalopathy, with DWI lesions extending bilaterally to the caudate, putamen, and ipsilateral insula. Over time, the patient became mute, bedridden, and required nasogastric tube feeding. She passed away a year and a half after diagnosis. The Heidenhain variant of CJD, first described in 1954 and clinically characterized by significant visual impairments (hemianopia, cortical blindness, optical hallucinations, palinopsia, or metachromatopsia), occurs in 3.7-20% of CJD cases, with an annual incidence of 1 case per 1 million people. Molecular and histopathological typing in 23 cases has linked this variant to the MM1 sCJD type. In a large series of 370 patients, Heidenhain was diagnosed in only 18 (4.9%), which showed that MM2 may be present and have a more protracted course, such as in our case. Typical brain findings include DWI and FLAIR hyperintensities in the occipital-parietal region, initially overlooked on MRI. PET imaging may aid in early detection by showing hypometabolism. Progressive cortical atrophy, white matter hyperintensities, and loss of gray and white matter characterize disease evolution on MRI. This case underscores the importance of recognizing this rare variant of CJD and provides a clinical-radiological framework for understanding its progression.

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Hemiballism-hemichorea as the initial presentation of type II diabetes refractory to adequate glycemic control: a case report

Luan Yudi Prando Kasuga[1], Melissa Caroline Godoi Prestes[1], Ádria Rodriges da Silva[1], Bruna Beppler[1], Caroline Zapelini[1], Lia Maki Hatisuka Imai[1], Gabriel Rodrigues Espelho Rossi[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Melissa Caroline Godoi Prestes; Email: melissa.caroline@outlook.com.

Case presentation: A 69-year-old male with a history of hypertension, dyslipidemia, and overweight, who had irregular medication use, was admitted to a tertiary hospital due to sudden onset of involuntary movements in his left upper limb that began three days prior, associated with forced blinking and involuntary cervical rotation. On physical examination, hemichorea-hemiballismus was observed in the left upper limb, worsening during distracting maneuvers. There was no nystagmus or signs of meningeal irritation. Muscle strength was globally grade V, and no other abnormalities were noted. Laboratory tests showed a hemoglobin A1c of 12.7% and capillary glucose reaching 534mg/dL, leading to a diagnosis of type 2 diabetes mellitus upon admission. Insulin therapy was initiated, and after four days, with persistence of the movements associated with hemiballism, valproic acid 250mg every 12 hours was started. Due to clinical stability and negative infection screening, the patient was discharged on valproic acid, and outpatient follow-up was initiated. After two months, there was a partial reduction in choreic movements. Cranial magnetic resonance imaging (MRI) showed a hyperintense T1 signal in the right striatum, which may correspond to gemistocytic degeneration related to non-ketotic hyperglycemia-induced hemichorea-hemiballism. After eight months, there was partial improvement of the hemichorea, although movements persisted despite adequate glycemic control, and the patient continued with outpatient follow-up and medication. **Discussion:** Hemichorea secondary to non-ketotic hyperglycemia is one of the manifestations of uncontrolled diabetes mellitus. It has been suggested that hyperglycemia may induce anaerobic metabolism, leading to the utilization of GABA as an alternative energy source in the absence of ketone bodies. The clinical presentation includes hemiballism, which differs from chorea due to the larger amplitude of movements and commonly affects the proximal region of limbs, such as the shoulders. Diagnosis is based on the association of clinical findings with imaging results the presence of a hyperintense T1 signal in the putamen contralateral to the hemiballism on MRI. It is considered a benign phenomenon with a good prognosis, and in most cases, glycemic control is sufficient for the resolution of choreiform movements within days. Persistence is rarer and appears to be related to neuronal death or partial dysfunction resulting from hyperglycemia-induced stress in the striatum. **Final comments:** Undiagnosed diabetes mellitus should always be considered in patients presenting with hemichorea or hemiballism. Restoration of euglycemia leads to symptom resolution in most cases, but persistence may require medication with dopamine receptor antagonists. This case highlights the importance of recognizing neurological symptoms as manifestations of metabolic dysregulation in diabetic patients, emphasizing the complexity of managing this condition.

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Hemiconvulsion-hemiplegia epilepsy syndrome associated with variant in the CACNA1A gene

Lucas Do Rego Barros Correa Martins[1], Caique Alberto Dosualdo[1], Matheus Hisamitsu Facine[1], Humberto Ramos Crispim[1], Nasser Faiçal Abou Zeenni[1], Ricardo Kendy Nishiuchi de Paiva[1], Antonio Lourenço Pires Neto[1], Laura A. M. Montouro[1], Amanda Leme da Silva Campinas[1], Eduardo de Paula Estephan[1,2]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Lucas do Rego Barros Correa Martins; Email: lucasrbcmartins@gmail.com.

Hemiconvulsion-Hemiplegia-Epilepsy syndrome (HHE) is a rare neurological condition characterized by episodes of hemiconvulsions followed by transient or permanent hemiplegia, along with recurrent epileptic seizures. Among few others syndromes the CACNA1A gene have been associated with HHE, but the clinical course, specific features and response to treatment are not totally elucidated. **Case presentation:** 20-year-old Brazilian male has experienced seizures since infancy. Throughout childhood, he experienced two types of seizures: brief episodes of staring and chewing movements occurring weekly, and longer tonic-clonic seizures affecting the right side of the body, accompanied by ocular deviation, head version, and drooling, sometimes lasting up to approximately 15 minutes, necessitating emergency care. Cognitive delay, with difficulties in reading and writing, was noticed by his mother during this period. On first assessment at our center, the patient reported daily focal seizures with impaired awarenes and monthly episodes of hemiconvulsions, which necessitated emergency care frequently. Episodes of agitation and aggression were also common. Physical examination indicated high-arched palate and low-set ears, dysarthric speech, global hyperreflexia, and extensor plantar reflex bilaterally. Throughout his life, the patient has been treated with topiramate, clobazam, levetiracetam, and most recently with valproic acid, lamotrigine, carbamazepine. Diagnostic workup included normal EEGs and MRI showing no structural lesions. New generation sequencing genetic test (panel) identified a heterozygous variant in the CACNA1A gene (p.Tyr62His ENST00000360228). Then patient was put on acetazolamide, with gradually increasing dose, up to 500mg twice a day. The seizures, which were daily before, had a sharp reduction to a frequency of two episodes monthly. **Discussion:** HHE profoundly impacts patients' quality of life. This case illustrates the syndrome's complexity, with frequent seizures resistant to treatment. Managing HHE remains challenging, necessitating multidisciplinary interventions to control seizures and support neuropsychomotor development. Early identification and appropriate intervention are crucial for improving prognosis and quality of life. Seizures response to acetazolamide is not well established on literature, but in our case had a clear effect, suggesting an option to treat this often refractory epilepsy.

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Hereditary coproporphyria: misdiagnosis and iatrogenesis - a case report

Lucas Gonçalves Coelho[1], Victoria Nunes Alvares[1], Clara Ribeiro Rizzo[1], Isabella Navarro Diaz Horta[1], Maria Eduarda Braz Ferreira Leão Caldas dos Santos[1], Bernardo Leal Cupello[1], Luan Henrique Honório Rocha[2], Rayanne da Silva Souza[2], Stephanie de Freitas Canelhas[2], Karina Lebeis Pires[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gafrée e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: Lucas Gonçalves Coelho; Email: lucasgcoelho@edu.unirio.br.

Case presentation: A 45-year-old male patient reports the onset of severe lower back pain in 2013. After numerous visits to the emergency room, he underwent the insertion of a double J catheter due to the diagnosis of nephrolithiasis, with no improvement in the pain. Subsequently, due to a case of constipation, he was admitted for the treatment of paralytic ileus and, with persistent pain, underwent an appendectomy due to suspected appendicitis. With worsening abdominal and lumbar pain, he underwent two exploratory laparotomies, including cholecystectomy, without any diagnostic definition or clinical improvment. The patient developed ascending sensorimotor polyneuropathy and acute respiratory failure, leading to admission to the ICU and dependency on mechanical ventilation. Still during hospitalization, therapy for Guillain-Barré syndrome was conducted without improvement. After two months of symptoms onset, while in the ICU, the hypothesis of hepatic porphyria was raised. Upon confirmation of the diagnosis through porphobilinogen and delta-aminolevulinic acid testing in urine samples, appropriate treatment was initiated, leading to clinical improvement and discharge. During follow-up at the outpatient clinic, molecular genetic testing was performed and was positive for a pathogenic variant of the CPOX gene, confirming the diagnosis of hereditary coproporphyria. **Discussion:** Porphyrias are a group of rare manifestations caused by enzymatic impairment in the heme production pathway, resulting in the accumulation of neurotoxic byproducts. One subtype of this condition is hereditary coproporphyria, caused by the presence of a pathogenic variant of the CPOX gene. This rare disease has a wide spectrum or non-specific manifestations, ranging from cutaneous symptoms to abdominal and neuropsychiatric conditions. In the presented case, it is evident that the lack of pathognomonic symptoms, combined with the rarity of the disease, complicates the diagnosis and initiation of treatment in patients with the condition, often leading to invasive procedures that could have been avoided. **Final comments:** Including porphyrias as a differential diagnosis in acute neurovisceral conditions is crucial to facilitate the early identification of these conditions. Therefore, specific laboratory and molecular genetic testing are essential for the accurate diagnosis. With the proper diagnosis and treatment, it is possible to control the progression of the disease, in addition to significantly improving the quality of life and reducing mortality associated with hereditary coproporphyria.

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Hereditary gelsolin amyloidosis: a case report and brief review of the literature

Caroline Bittar Braune[1], Marina Papais Alvarenga[2], Renato Ambrósio Júnior[3], Luiz Carlos Pinto[4], Marcus Vinicius Rocha Pinto[4], Regina Maria Papais Alvarenga[2,3]

[1]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gafrée e Guinle, Departamento de Neurologia, Rio de Janeiro RJ, Brazil.
[2]Universidade de Vassouras, Vassouras RJ, Brazil.
[3]Universidade Federal do Estado do Rio de Janeiro, Faculdade de Medicina, Rio de Janeiro RJ, Brazil.
[4]Clínica de Neurofisiologia Luiz Carlos Pinto, Rio de Janeiro RJ, Brazil.

Correspondence: Caroline Bittar Braune; Email: caroline.bittar@gmail.com.

Case presentation: A 78-year-old male was evaluated with a history of bilateral facial paralysis for 25 years. He reports that he was asymptomatic until age 53, when he noticed facial asymmetry. He underwent a cranial magnetic resonance imaging, which was normal, and neurophysiological studies showed axonal damage to the facial nerves with signs of reinnervation, but no further investigation was carried out. At the age of 69, he developed new motor difficulties bilaterally in the face, with a diagnosis of new facial paralysis. At the age of 73, he noticed gradual binocular visual loss and was diagnosed with glaucoma. He had a previous diagnosis of systemic arterial hypertension and type 2 diabetes mellitus. Regarding his family history, his mother died at the age of 85 with facial weakness and binocular amaurosis secondary to glaucoma. His neurological examination revealed facial diplegia with bilateral Bell's sign, global apalesthesia, binocular ophthalmoparesis, in addition to "cutis laxa". Neurophysiological studies showed chronic neuropathy of the facial nerves and chronic axonal sensory-motor polyneuropathy. In a new ophthalmological evaluation, corneal biomicroscopy established the diagnosis of corneal lattice dystrophy. Considering the phenotype characterized by slowly progressive cranial neuropathy with

predominant involvement of the facial nerves, associated with polyneuropathy, "cutis laxa" and corneal lattice dystrophy, with positive family history, the diagnostic hypothesis of hereditary gelsolin amyloidosis was established, and genetic test was requested. **Discussion:** Hereditary gelsolin amyloidosis (HGA) is a rare autosomal dominant disease most likely caused by sporadic low-frequency mutations in the gelsolin gene (GSN). It was initially described by the Finnish ophthalmologist Dr. Meretoja, and cases were subsequently noticed in several countries, although the disease may still be underdiagnosed. Gelsolin is a principal actin-modulating protein, implicated in multiple biological processes including the nervous system, e.g. axonal transport, myelination, neurite outgrowth, and neuroprotection. GSN defect causes expression of variant gelsolin, followed by systemic deposition of gelsolin amyloid. HGA symptoms start in the third or fourth decades of life, typically emerging as a classical triad of ophthalmological (corneal lattice dystrophy type 2), cutaneous (cutis laxa) and nervous system involvement, with slow progression. Progressive bilateral facial palsy is typical, with initial involvement of the upper and later lower nerve branches. HGA may manifest with further neurological (i.e., other cranial nerves involvement, peripheral and autonomic neuropathy) or systemic features, including cardiac and renal abnormalities. **Final comments:** HGA is a rare condition, but it presents a characteristic phenotype and should be considered as a differential diagnosis of multiple and progressive cranial neuropathies, especially facial diparesis.

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Hereditary neuropathy with liability to pressure palsies (HNPP): a diagnostic challenge

Geovanna Barros Rocha[1], Lucas Miquilini de Arruda Farias[1], Fernando Tavares Brasil Teixeira[1], Raissa Prata Meireles[1], Douglas Ferreira Pereira[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Geovanna Barros Rocha; Email: geovannaroccha@outlook.com.

Case presentation: F.T.S., 42 years old, male, white. The patient reported a year ago that he noticed loss of strength in his left hand, associated with hypotrophy of the thenar region and also high-frequency tremor, both at rest and in movement. He had a similar episode 10 years ago, which resolved spontaneously with daily activities. Exams: Electroneuromyography of the left upper limb (26/10/23) - Peripheral, sensory-motor neuropathy, with multiple compressive lesions of the ulnar and median nerves. Magnetic resonance imaging of the left elbow (01/23/24) - slight thickening with changes in the ulnar nerve signal. Physical examination of the left upper limb: Strength 3, Mingazzini and Froment positive. Hypoesthesia in the path of the ulnar nerve. Negative Allen, Tinel, Phallen and Adson maneuvers **Discussion:** This is a rare case of Hereditary Neuropathy with Susceptibility to Pressure Palsy (NHPP), with episodic and recurrent characteristics, with an incidence of 2-5/100,000, with autosomal dominant inheritance and involvement of chromosome 17 (80-85%): 17p11.2. The clinical manifestations, despite the variety of phenotypes, are classic, with gradual and progressive weakness, asymmetrical and episodic, with a long evolution, just like the case presented by the patient. The diagnosis is made by performing electroneuromyography, which shows slowness in the conduction fibers, as well as by carrying out genetic tests. There is still no treatment available for peripheral mononeuropathies, only physiotherapy, occupational therapy and some orthopedic procedures to avoid atrophic complications in the future. The presented patient is progressing well with physiotherapy and outpatient follow-up. **Final comments:** Despite the absence of family history and molecular tests, the diagnosis of NHPP can be made with clinical history and examinations. It is necessary to establish the diagnosis as early as possible, with the aim of starting motor rehabilitation treatment and avoiding complications, in addition to allowing genetic counseling to be carried out in the family. Even though it is a rare disease, it represents an important study, due to the complexity of the diagnosis and the possibility of evolution.

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Hereditary spastic paraparesis associated with SPG11 gene: a case report

Victória Nunes Alvares[1], Fernanda Dias Guimarães Almeida[1], Wesley Gabriel Silva de Morais[1], Lucas Gonçalves Coelho[1], Tatyane Andrade de Souza Silva[1], Vanessa Conceição Saar Mautone Lima[2], Luan Henrique Honório Rocha[2], Karina Lebeis Pires[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gafrée e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: Victória Nunes Alvares; Email: victoriaalvares@edu.unirio.br.

Case presentation: A 38 year-old female presented with lower limb weakness, mainly on the right, and daily-long lasting cramps that started after a cesarean delivery at age 30 was referred to our neurology department. On physical evaluation, the patient presented a spastic gait, only being able to walk with support as she was restriced to a wheelchair. She also presented hypertonia and clonus in lower limbs; generalized muscle weakness, specially in the feet; global hyperreflexia with increased reflex area; dysdiadochokinesia and dysmetria in the index-nose test. An in-depth investigation was carried out and, as infectious and rheumatological causes were ruled out, a brain magnetic resonance imaging (MRI) was requested. The exam showed tenuous hypersignal in the white matter adjacent to lateral ventricles in T2 and FLAIR sequences, diffuse thinning of the corpus callosum and a discrete accentuation of the cortical grooves and fissures. Cervical, thoracic and lumbosacral spine MRI didn't show any pathological alterations. The investigation proceeded and an electroneuromyography was performed on the patient, showing results consistent with diffuse preganglionic axonal involvement reaching neural structures that supply the L3-L4-L5-S1 myotomes. Therefore, genetic molecular testing was requested and showed the presence of a mutation of the SPG11 gene (OMIM*610844), consistent with hereditary spastic paraparesis type 11. **Discussion:** Hereditary spastic paraparesis (HSP) represents a heterogenous group of inherited rare diseases in which the main clinical feature is the progressive lower limb weakness and spasticity. Although most studies indicate that the onset of clinical features occur in adolescence, it does not rule out the possibility of a late manifestation of symptoms, as it was seen in the case presented. The diagnosis of HSP is hampered by the difficulty to access the genetic molecular testing and also the wide range of genetic conditions that it may be associated with, as well as the variety of intensity of clinical symptoms that patients with the same mutation may present. Therapeutic options include pharmacological treatments to manage mainly the spasticity and a multidisciplinary approach, including especially physiotherapists and speech-language pathologists, to prevent possible complications, such as paraplegia, dysarthria and dysphagia. In this case report, the late onset of symptoms and late diagnosis compromised the early start of adequate treatment, thus stimulating the progression of the disease. **Final comments:** HSP associated with the SPG11 gene is rare neurological disease that has a challenging diagnosis, even though clinical symptoms and imaging findings may be characteristic. Therefore, we emphasize the significance of early diagnosis as the pharmacological treatments and multidisciplinary approach prove to be beneficial in the improvement of the quality of life of our patients.

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Hereditary spastic paraplegia 76 an extremely rare disease with challenging diagnosis

Juan Sebastián Sánchez León[1,2], Tais Luise Denicol[2], Carolina Matte Dagostini[1,2], Renato Dumba Monteiro de Castro[1,2], Pedro Hall Ruschel[1,2], Karla Sarai Sandoval[1,2], Carlos Roberto de Mello Rieder[1,2], Jonas Alex Morales Saute[3]

[1]Irmandade da Santa Casa de Misericórdia de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[3]Universidade Federal do Rio Grande do Sul, Hospital de Clinicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Juan Sebastián Sánchez León; Email: juansebastian1516@hotmail.com.

Case presentation: 48 years old woman without significant family history and comorbilities presented with a slowly progressive history of weakness in the left lower limb since 30 years old. After few months, she presents involvement of the right lower limb, with gradual worsening of gait, needing to use crutches. Subsequently, present mild dysphagia and dysarthria. The symptoms worsening very slowly and they stabilized the last years. The patient's neurological exam showed crural paraparesis, global hyperreflexia, spasticity in all 4 limbs, ataxia, positive bilateral Babinski and Hoffman signs. In addition to the neurological symptoms she has joint pain, xerostomia and xerophthalmia. We performed MRI of brain and spinal cord without alterations. Blood and serological tests: vitamin B12, serum copper, anti-HTLV-1, anti-CCP, rheumatoid factor, anti-SM, anti-SSa, anti-SSb, anti-histona, anti-RNP, anti-GAD all negative and positive ANA (1:320 BAC-3 pattern). We decided to make a chest tomography, biopsy of salivary glands and ophthalmological evaluation all of them without alterations, ruling out the diagnosis of Sjogren's syndrome. Genetic tests negative for SCA 1,2, 3, 6, 7, 10, 12, 17. Not having diagnosis and high clinical suspicion of hereditary disease, we request complete Genome Sequencing showed: homozygous variant NM_005186.4(CAPN1):c618_619del:p.(Gly208Glnfs*7). The result confirms diagnosis of hereditary spastic paraplegia 76 (HSP76) with involvement by the CAPN1 gene. With the diagnosis we decided to make genetic counseling. **Discussion:** Hereditary spastic paraplegia (HSP) is a heterogeneous group of neurodegenerative diseases with approximately 91 associated loci. HSP76 is an autosomal recessive disease that was described for first time in 2016 and is characterized by presenting spasticity in legs, ataxia, dysarthria and peripheral neuropathy. For the low prevalence and difficulty in accessing to specific tests the diagnosis of HSP is challenging. There's less than 50 cases reported in the world. The disease is caused by CAPN1 gene variants. The phenotype of the disease is characterized for mild or moderated symptoms with early onset and subsequent stabilization of the disease. **Final comments:** Althought there is no disease-modifying treatment, a genetic evaluation is necessary in case suspect of hereditary spastic paraplegia, to start genetic counseling, symptomatic treatment (baclofen, tizanidine, botulinum toxin) and rehabilitative medical therapy.

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Hereditary spastic paraplegia, Kjellin syndrome: clinical findings that may go unnoticed

Caroline Vianna Costa Pinto[1], João Henrique Reis Duque Estrada[1], Luiza Póvoa de Souza Guimarães[1], Victor Waldhelm Cozer[1], Diogo Matheus Terrana de Carvalho[1], Felipe da Rocha Schmidt[1], Sofia Abreu Mermelstein[1]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Caroline Vianna Costa Pinto; Email: carol.vianna.cp@gmail.com.

Case presentation: A 44-year-old man presented to the neurology outpatient clinic complaining of lower back pain and difficulty walking. He reported progressive difficulty over the past 8 years, initially with running, then with dressing, climbing up and down stairs, and eventually with walking. On examination, he exhibited spastic rigidity in the lower limbs with mild asymmetry (worse on the right), symmetrical hyperreflexia with inexhaustible clonus in the lower limbs, and bilateral Babinski sign. Muscle strength was graded 4+ in the lower limbs symmetrically. He denied consanguinity in the family, but mentioned having 6 siblings, including two sisters and four brothers, one of whom had a similar condition. This brother, aged 41, also had spastic rigidity with hyperreflexia, but with exhaustible clonus bilaterally, right-sided Babinski sign, flexor plantar response on the left, and hypoesthesia in a stocking distribution. Both brothers denied visual changes and exhibited intellectual deficit with executive dysfunction, episodic memory deficit, and language impairment. Ophthalmological evaluation revealed retinal pattern dystrophy in both brothers. Cervical and dorsal spine MRI showed no abnormalities, while cranial MRI in both patients revealed discrete foci in the white matter with T2 hyperintensity and T1 hyposignal, suggestive of the ears of the lynx sign. Given the clinical history, and exclusion of diferential diagnosis like infectious and immune-mediated etiologies, the hypothesis of Kjellin syndrome, hereditary spastic paraplegia (HSP), was raised and confirmed by genetic testing showing a mutation in the SPG 11 gene. **Discussion:** HSP is a disease characterized by chronic and progressive spastic paraparesis, with other possible associated symptoms depending on the specific subtype, such as ataxia, dementia, and retinal changes. Currently, there are descriptions of at least 87 genes associated with this disease. The presented case highlights the importance of comprehensive investigation in these patients, including fundoscopy, searching for associated signs and symptoms, and active screening of affected family members. The combination of retinal dystrophy, ears of the lynx sign, and corpus callosum hypoplasia on MRI, should prompt the neurologist to consider hereditary spastic paraplegia due to mutations in the SPG 11 or SPG 15 genes. **Final comments:** The objective of this case report is to emphasize the importance of clinical findings even in a rare disease in which genetic testing plays a prominent role, aiming to reinforce the importance of a comprehensive diagnostic investigation that often brings us very close to the definitive diagnosis.

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Hereditary transthyretin amyloidosis (ATTRv) mimicking motor neuron disease: importance of considering this differential diagnosis

Cleberson Alberceli Vieira de França[1], Mayhara Rosany da Silva Santiago[1], Renata Amaral Andrade[1]

[1]Hospital Pelópidas Silveira, Recife PE, Brazil.

Correspondence: Cleberson Alberceli Vieira de França; Email: cleberson.avf@gmail.com.

Case presentation: Case 1: A 71-year-old male patient presented with symptoms of progressive dysphonia and dysphagia, progressing to fasciculations, weakness, and atrophy in the upper limbs. He reported significant weight loss over the past year and a history of bilateral carpal tunnel syndrome. There was no family history of polyneuropathy. Electroneuromyography (ENMG) revealed chronic pre-ganglionic involvement in bulbar and cervical segments, as well as bilateral median nerve impairment. Exome sequencing: pathogenic mutation in the TTR gene (p.Val142Met). Case 2: A 58-year-old female patient exhibited progressive weakness in the lower limbs, initially distal and evolving over 2 years to involve both proximal and distal muscles of the upper limbs, along with dysphonia and episodes of choking. She had a history of chronic diarrhea. Examination showed tongue atrophy and fasciculations, predominantly distal tetraparesis, global areflexia, and tactile and painful hypoesthesia with a gradient in the lower limbs. ENMG: denervation in bulbar, cervical, thoracic, and lumbar segments, predominantly distal, with absent sensory potentials in the lower limbs and reduced motor potentials dependent on length. Gene sequencing of TTR: pathogenic mutation (p.Ile127Val). Case 3: A 75-year-old male patient presented with mild symptoms of paresthesia in the feet progressing to tetraparesis and dysphagia over 5 years. There was no family history of polyneuropathy. Examination: distal flaccid and areflexic tetraparesis, global muscle hypotrophy, and fasciculations in the tongue, cervical, and lumbar segments. Bilateral hypopallesthesia in the hallux was noted. ENMG: chronic pre-ganglionic involvement in bulbar, cervical, and lumbosacral segments with signs of acute denervation. Associated sensory polyneuropathy was also observed. Gene sequencing: pathogenic mutation in TTR (p.Val50Met). **Discussion:** Hereditary Transthyretin Amyloidosis (ATTRv) is an autosomal dominant disease typically presenting in adulthood with heterogeneous clinical manifestations, classically as a sensorimotor polyneuropathy, with early involvement of small fibers and significant autonomic dysfunction. There is a strong genotype-phenotype correlation with variable penetrance and age of onset with late-onset disease, typically over 50 years old, may present with a predominantly motor phenotype characterized by rapid progression and a worse prognosis. The phenotypic pattern mimicking motor neuron disease (MND), including tongue atrophy and fasciculations, has been documented in the literature. Based on the Gold Coast criteria, all these patients could potentially be misdiagnosed as having amyotrophic lateral sclerosis (ALS). **Final comments:** The predominantly motor clinical presentation in ATTRv patients with polyneuropathy is not commonly recognized by neurologists, leading to frequent misdiagnosis. We currently propose that ATTRv should be considered in the differential diagnosis of patients suspected of having ALS.

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Hereditary transthyretin amyloidosis: a diagnostic challenge

Ana Luíza Xavier Drumond[1], Vivian Proença Lara[1], Gláucia Monteiro Oliveira[1], Igor Pereira Matos de Oliveira[1], Caio Cobucci Esteves[1], Fernando Augusto Bruzzi França[1], Thaís Ribeiro Oliveira Santos de Marcello[1], Gláucia Lara Resende[1], Cíntia Alvarenga Pereira Vieira[1]

[1]Santa Casa de Misericórdia de Belo Horizonte, Belo Horizonte MG, Brazil.

Correspondence: Ana Luíza Xavier Drumond; Email: analuizax95@gmail.com.

Case presentation: Man, 63 years old, previously healthy, retired. In August 2023, he was subjected to a surgical procedure for treating Carpal Tunnel syndrome. As per the standard procedure of the institution where the surgery was conducted, a fragment of the left wrist flexor retinaculum was submitted for histopathological examination, which revealed fibrosis and amyloid deposits. Due to these results, we requested a Transthyretin (TTR) Gene molecular analysis, which detected a pathogenic variant c.424G>Ap. VAl122IIe and a heterozygous mutation in exon 4 of the TTR gene. We postulated the hypothesis of Hereditary Transthyretin Amyloidosis and decided to perform more tests to investigate the possibility of involvement of other organs. According to the electrocardiogram, the patient had already developed first-degree atrioventricular block and nonspecific ventricular repolarization abnormality in the inferior wall. To complement the cardiological propedeutics, we conducted an echocardiogram which revealed atrium enlargement, signs of moderate concentric hypertrophy and grade II left ventricular dysfunction. Furthermore, some laboratory results were also abnormal, such as elevated proBNP levels and reduced glomerular filtration rate. Finally, to investigate the involvement of peripheral nerves, we performed an electroneuromyography of the lower limbs, which demonstrated to be compatible with symmetric axonal sensory polyneuropathy, with mild intensity. After initial propedeutics, we referred the patient to a specialist service for neuromuscular diseases and cardiology, for observing and treating the diagnosed disease. In May 2024, he began the use of the standard medication to treat amyloidosis, Tafamidis Meglumine, 20 mg per day. **Discussion:** Hereditary transthyretin amyloidosis is an autosomal dominant disease caused by a hereditary gene responsible for encoding the mutant form of the TTR protein. Mutation of this protein results in systemic amyloid deposits, and its accumulation occurs mainly in peripheral nerves and in the heart. This is a systemic and consumptive condition that begins in adulthood. Autonomic sensorimotor polyneuropathy is its primary feature. In many cases, Carpal Tunnel syndrome is a common neuropathic manifestation that appears as first symptom of the disease. **Final comments:** To encourage physicians to begin investigating the disease earlier, it is indispensable the conduction of further research to determine symptoms and their prevalence as an initial manifestation of amyloidosis, such as Carpal Tunnel syndrome, since early diagnosis allows for faster treatment and prevention of serious complications.

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Herpes zoster-related hospitalizations and mortality in brazil over 5 years: an epidemiological study

Isadora Carvalho de Sousa[1], Mylena Cordeiro Aranha[1], Luiza Bisognin Marchesan[2], Amanda Fortes Cavalcante de Oliveira[3], Celijane Almeida Silva[4], Mariana Teixeira Mesquita[5], Laiane Rodrigues Macêdo[6], Lara Maia Pereira[1], Luana Dutra Pinheiro da Silva[1], Bárbara Pinheiro Gama de Andrade[3]

[1]Universidade Potiguar, Natal RN, Brazil.
[2]Universidade Franciscana, Santa Maria RS, Brazil.
[3]Universidade Federal do Delta do Parnaíba, Paraíba PI, Brazil.
[4]Centro Universitário Unifacisa, Campina Grande PB, Brazil.
[5]Universidade Estadual de Campinas, Campinas SP, Brazil.
[6]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Mariana Teixeira Mesquita; Email: maritmesquita@gmail.com.

Background: Herpes zoster (HZ) is a chronic disease secondary to the reactivation of the varicella virus. The incidence of HZ increases with age and factors such as immunosuppression, infections, stress, and advanced age. The disease presents various stages and manifestations, some of which show a higher risk of complications such as postherpetic neuralgia, renal and gastrointestinal complications, and vasculitis. Currently, published data on clinical characteristics, severity, and outcomes related to HZ are limited. Understanding these data could potentially increase the impact of HZ management in Brazil. **Objective:** To identify and analyze the epidemiological profile of hospitalizations and deaths due to HZ in regions of Brazil, from 2019 to 2023. **Methods:** This is a descriptive study, with a quantitative approach, of a retrospective nature, regarding hospitalizations and deaths due to HZ in Brazil, between January 2019 and December 2023. The data were collected in April 2024 from the Hospital Information System of the Unified Health System (SIH/SUS), through the DATASUS platform. The selected variables were: processing year,

region/federal unit, age group, race/ethnicity and gender. **Results:** During the period from 2019 to 2023, Brazil registered a total of 20,164 hospitalizations due to herpes zoster. The Southeast region was the most affected, representing 45.5% of the total, with 9,190 cases, followed by the Northeast with 4,177 cases (20.7%), the South with 3,239 cases (16%), the Midwest with 1,876 cases (9.3%), and the North with 1,682 cases (8.3%). The year 2019 recorded the highest number of occurrences, with 5,631 cases, while 2021 had the lowest number, with 2,978 cases. As for age group, the most hospitalized population was those aged between 60 and 69 years, representing 14.5% of the total. The age group with the highest number of deaths was those aged 80 years or older, accounting for 39% of deaths. Regarding gender, females showed a slight predominance over males, with 50.3% of hospitalizations. In terms of ethnic characteristics, self-declared "pardos" (mixed race) were the most affected, representing 41.5% of hospitalizations. As for the total number of deaths, 1,505 cases were recorded, with the highest number also in 2019, with 407 deaths, and the lowest in 2021, with 261 deaths. **Conclusion:** HZ is associated with a significant risk of complications, including zoster-associated pain, impaired quality of life, and increased morbidity and mortality, especially for the elderly and immunocompromised individuals. The findings suggest a greater need for early intervention and prevention programs, such as vaccination, for the population aged 60 and over, women, those of mixed race, and residents in the Southeast region, in order to reduce complications associated with herpes zoster in Brazil.

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Herpetic encephalitis as a differential diagnosis of ischemic stroke

Ashley Cristina da Cruz Costa[1], Gustavo Alves Cangussú[1], Hiago Oliveira Soares[1], Bruna Souza Cardoso[1], Roberto Sérgio Ferreira Nascimento Filho[1], Tamyres Araújo Andrade Donato[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Ashley Cristina da Cruz Costa; Email: cristinadacruzcosta@gmail.com.

Background: Herpetic encephalitis (HSVE) is a severe neurological emergency, representing 15 to 20% of sporadic encephalitis cases in adults, that can also affect children and neonates. The etiological agent is the Herpes Simplex Virus, primarily strains type 1 (HSV-1) and type 2 (HSV-2). Due to viral neurotropism, this condition can lead to cerebral complications, such as strokes, both in primary infection and reactivation after latency. The relationship between ischemic events and HSVE has been reported in several studies across different age groups and encephalic topographies. **Objective:** To analyze the manifestations and ischemic cerebral repercussions in patients with HSVE. **Methods:** This study is a systematic literature review based on articles published at PubMed, Biblioteca Virtual de Saúde (BVS), and SciELO databases. Publications from 2017 to 2024, in English and Portuguese, using the MeSH descriptors: "Encephalitis", "Herpes Simplex", and "Stroke", were considered. Analytical studies related to the theme, providing relevant information on herpetic encephalitis and its potential as a differential diagnosis for stroke, were included. Experience reports and studies not meeting the inclusion criteria were excluded. **Results:** The pathophysiological mechanism of the ischemic presentation of HSVE can be explained by the onset of large vessel vasculitis, leading to multifocal ischemia, which can mimic a stroke. Typical symptoms in this context include fever, chills, memory impairment, and mental confusion due to the involvement of the limbic system, which is common in HSVE. However, even in the absence of these symptoms, a differential diagnosis should be considered in young patients presenting to the emergency department with suspected stroke. Focal neurological deficit may also be present (26%), as well as speech disorders (57%), with receptive aphasia being the most common. Thus, early magnetic resonance imaging (MRI) is important, as atypical presentations are often misdiagnosed, which can delay the initiation of effective treatment or lead to inappropriate therapy. Findings from this exam usually include unilateral edema in the temporal lobe, generally sparing the basal ganglia. Additionally, cerebrospinal fluid (CSF) analysis can be requested, with the gold standard for diagnosis being the detection of viral DNA in the CSF. **Conclusion:** Therefore, the importance of correct diagnosis and management in patients with suspected stroke, especially if young, is evident, considering the differential diagnosis of the ischemic presentation of HSVE. In this context, the MRI and CSF analysis are valuable complementary exams for the exclusion or confirmation of this clinical condition. Thus, this text aims to draw attention to this pathology and expand knowledge in this still underreported area, to reduce its morbidity and mortality, as well as to prevent possible complications.

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Heterozygous GRIN2B mutation as a cause of intellectual developmental disorder: a case report

Rodrigo Ibañez Tiago[1], Guilherme Augusto de Oliveira Soares[2], Isabela de Carvalho Florêncio[2], Rodolfo Antonio Corona[2], Renata Gratão Rezende[2], Carolina Cândida Vaz[2], Liane de Rosso Giuliani[2], Thiago Dias Fernandes[2]

[1]Universidade Federal de Mato Grosso do Sul, Faculdade de Medicina,
[2]Hospital Universitário Maria Aparecida Pedrossian

Correspondence: Rodrigo Ibañez Tiago; Email: rodrigoibaneztiago@gmail.com.

Case presentation: A 30-year-old male patient was referred to the neurogenetics clinic for evaluation of non-syndromic intellectual disability of unclear etiology. The patient was previously diagnosed with global developmental delay during childhood, with no further investigation conducted at that time. Early hypotonia was noted, though he could sit unsupported at 6 months of age. He ambulated with support at 2 years and 7 months, and independently at 3 years and 8 months. He developed cognitive deficits, requiring special education, and was eventually able to read and write with limitations. Physical examination revealed moderate intellectual disability, paratonia, and grade 3 tendon reflexes. The karyotype was 46 XY, with fragile X syndrome ruled out. Electroneuromyography and cranial MRI showed no abnormalities. Whole exome sequencing identified a de novo mutation in the GRIN2B gene at position chr12p13.611.817, with an A>G variation resulting in a missense mutation p.(Phe563Ser) in heterozygosity. The patient was diagnosed with autosomal dominant intellectual developmental disorder-6 with or without epilepsy (MRD6, OMIM# 613970). **Discussion:** A heterozygous mutation in the GRIN2B gene can lead to the phenotypes of both autosomal dominant intellectual developmental disorder-6 with or without seizures, and developmental and epileptic encephalopathy-27 (DEE27, OMIM# 616139). The GRIN2B gene encodes a subunit of the NMDA receptor, which is crucial for synaptic plasticity and neuronal development. Mutations in this gene can alter receptor function. In the present case, the de novo p.(Phe563Ser) mutation in the GRIN2B gene is consistent with the patient's intellectual developmental disorder. **Final comments:** This specific p.(Phe563Ser) variant in the GRIN2B gene has not been previously described in the literature. This case underscores the importance of detailed genetic investigation in patients with nonsyndromic intellectual developmental disorders. Identifying mutations contributes to a more precise diagnosis and informs management strategies, counseling, and prognosis for these patients.

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Hidden focal cortical dysplasia in the frontal lobe: value of repeated mri correlated with electroclinical hypothesis

Fernanda Schuh Martins[1], William Alves Martins[1], Abner Cutrim[1], Vanessa Vargas[1], Fernanda Quadros[1], Eliseu Paglioli[1], Rafael Paglioli[1], Thomas Frigeri[1], André Luis Fernandes Palmini[1]

[1]Santa Casa de Misericórdia de Porto Alegre RS, Brazil.

Correspondence: Fernanda Schuh Martins; Email: fschuhmartins@gmail.com.

Focal cortical dysplasia (FCD) is the leading pathological cause of childhood epilepsy, particularly treatable through surgery in cases of frontal lobe epilepsy (FLE). According to Blumcke's 2011 classification, FCD is divided into three types: Type I, which appears in adults with mild symptoms and temporal lobe changes; Type II, found in children with extensive frontal lobe changes; and Type III, involving FCD with associated lesions like hippocampal sclerosis, tumors, or vascular malformations. FCD-related seizures are often resistant to medication, necessitating surgical intervention such as lesion resection, lobectomies, or hemispherectomies. **Case presentation:** B.P.H., a 6 year old girl with normal developmental presented with drug-resistant epilepsy starting at age five. Her initial seizures involved right ocular deviation, blinking, right cervical lateralization, and oral automatism lasting 10-15 seconds, occurring daily and increasing in frequency despite despite the use of different anti-seizure drug regimens. Physical examination showed no focal signs. The initial brain MRI was reported as normal, however, after a 48-hour vídeo-EEG analysis showing clear left frontal epileptiform activity, image review revealed thickening of the left frontal cortico-subcortical interface and an elongated image in the underlying white matter, suggesting FCD (previously unnoticed). After a clinical discussion, epilepsy surgery was performed on July 3, 2023, involving resection of the left frontal dysplastic area, resulting in complete seizure free. **Discussion:** The patient's history, physical examination, and diagnostic findings indicated FCD manifesting in school age with difficult-to-control seizures. MRI characteristics include cortical thickening, blurred white matter-gray matter junction, abnormal subcortical architecture, altered white matter signals, sulcal or gyral abnormalities, and segmental or lobar hypoplasia. This patient's MRI findings were, upon review of the imaging, consistent with FCD Type II. Literature suggests 60-80% of patients remain seizure-free post-surgery, aligning with this patient's outcome. The most favorable prognostic factor is total resection, defined as the removal of the MRI-visible lesion or the epileptogenic focus identified by intracranial EEG. **Final comments:** Despite advanced neuroimaging methods and effective surgical techniques, FCD remains underdiagnosed due to nonspecific EEG changes and difficulties in detecting FCD on standard MRI. Accurate diagnosis and surgical indication require tools like Vídeo-EEG and high-intensity 3T MRI with standardized epilepsy protocols. Considering FCD in cases of drug-resistant epilepsy with normal MRI findings is crucial. Further research on FCD epileptogenesis could lead to new pharmacological therapies, with advancements in medical technology providing hope for future solutions.

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High values of glycated hemoglobin are associated with the severity of acute cerebral ischemia

Gabriele Babel[1], Thaís Janaína Crukil[1], Teresa Cristina Colvara Mattana[1], João Natel Pollonio Machado[1]

[1]Fundação Universidade Regional de Blumenau. Blumenau SC, Brazil.

Correspondence: Thaís Janaína Crukil; Email: thaiscrukil@gmail.com.

Background: Stroke is the second leading cause of mortality worldwide, affecting more than 100 million people annually. Diabetes mellitus (DM) is one of the main modifiable risk factors, increasing the risk of stroke by 2.5 to 3.6 times. The glycated hemoglobin test (HbA1c) is used as a control and diagnostic marker for DM, as its value reflects the average amount of glucose in hemoglobin over the last 120 days, eliminating the possibility of stress-induced hyperglycemia, which is common in cases of acute stroke. Few studies have explored the relationship between HbA1c levels and ischemic stroke (IS) severity, resulting in a limited understanding of this association. **Objective:** This study aims to evaluate the association between HbA1c values and the severity of acute IS, through a descriptive and retrospective observational study. **Methods:** The variables analyzed included age, sex, race, smoking, dyslipidemia, high blood pressure (HPB), DM and HbA1c values of patients with acute IS admitted to a tertiary hospital in Blumenau, between August 2023 and April 2024. It was approved by the Ethics Committee under opinion 6.633.545/2024. The Modified Rankin Scale and National Institutes of Health Stroke Scale were used to assess disability and severity, respectively. For statistical analysis, P-value <0.05 was considered. The sample consisted of 56 patients, with a mean age of 70.6±12.2 years, predominantly male, white, non-smokers, dyslipidemic, hypertensive, non-diabetic and normoglycemic. The multivariate logistic analysis indentified the following risk factors: age ≤60 years (P=0.02 OR: 8.152), HbA1c ≥7.0% (P=0.03 OR:6.862), hypertension (P=0.02 OR: 4.739) and dyslipidemia (P=0.12 OR: 4.147). DM was present in 39.3% of patients with IS but it was not statistically significant. **Results:** The results suggest that the diagnosis of DM had a smaller impact than having HbA1c ≥7.0%. It was observed that patients with HbA1c ≥7.0% werw more likely to be in groups with greater severity (HbA1c 9.29±1.81%) and disability (HbA1c 10.2±2.49%). Seven patients who were not diagnosed with DM had elevated HbA1c levels, and three had moderate/severe IS. It is important to include HbA1c in the IS care protocol, not only for diagnosing DM when there is hyperglycemia on admission but also for stratifying the risks of cardiovascular events. **Conclusion:** Our study highlighted that high levels of HbA1c were associated with greater severity and disability in IS, regardless of the previous diagnosis of DM. Patients with uncontrolled hyperglycemia had a 6 to 7 fold increased risk of moderate/severe IS. These findings suggest that HbA1c can serve as a prognostic marker to assess the severity of IS and diagnose DM.

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High-resolution ultrasound in the diagnosis of ischemic neuropathy post-arteriovenous fistula surgery: a case report and review

Júlio César Iaia[1], Roberto Pereira Santos[1], Liene Duarte[1], Carolina Rouanet[1], Gustavo Honório[1], Celmir Vilaça[1], Marco Antônio Sales Dantas de Lima[1], Ana Lucia Zuma de Rosso[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Júlio César Iaia; Email: julioiaiaufrj@gmail.com.

Case presentation: A 60-year-old female patient, hypertensive, dyslipidemic, and with chronic kidney disease (CKD), undergoing hemodialysis (HD) three times a week, had an arteriovenous fistula (AVF) created in the left upper limb. After 1 month and 20 days, she complained of pain in the operated limb, distal to the AVF. Physical examination revealed hypoesthesia, paresthesia, paresis, and burning pain in the left hand, in addition to a claw hand with more prominent paresis for flexion of the second and third digits, abduction and adduction of all digits, and wrist flexion. Other neurological aspects were normal. Electroneuromyography (ENMG) demonstrated reduced sensory and motor potentials of the left ulnar, median, and radial nerves. High-resolution ultrasound (HRUS), performed with a 12MHz linear transducer, showed an increased cross sectional area (CSA) of the left median and ulnar nerves. Compressive causes, such as compartment syndrome and nerve section, were excluded through magnetic resonance neurography (MRN). The diagnosis of ischemic mononeuropathy related to the AVF was established. Pain management with amitriptyline was effective. **Discussion:** The creation of AVF in these patients provides greater longevity and reducing the chances of infection in access management. However, ischemic neuropathy (IN) can arise as a complication, involving reduced distal arterial flow due to a steal from increased venous flow in the AVF, resulting in nerve ischemia. This condition is more frequent in women and diabetics. Symptoms of IN include pain, muscle weakness, and loss of sensation, often manifesting acutely. Differential diagnoses for these symptoms include steal syndrome from dialysis access (SSDA) and nerve compression by hematoma. For diagnosis, ENMG and MRN are commonly aplied methods. However, HRUS offers significant advantages such as greater accessibility, lower cost, painless and greater patient comfort. The use of HRUS in the diagnosis of IN is a new and promissing approach that has not been widely explored in the literature. HRUS has demonstrated high sensitivity and specificity in the evaluation of peripheral neuropathies, sugesting its potential as an effective and innovate diagnostic tool in this area. Although ENMG is accurate, combining HRUS with ENMG can enhance diagnostic accuracy. MRN, while useful for evaluating nerve diameter alterations and inflammatory aspects, faces limitations due to cost, availability, and the need for contrast. **Final comments:** To our knowlege, this is the first description of the role of HRUS in IN and compared do ENMG and neurography. HRUS proved to be a valuable tool in diagnosing IN post-AVF surgery for HD. This method is accessible, practical, and non-invasive, providing a viable alternative to traditional methods. This report highlights the need for further studies to better establish the accuracy of HRUS in this context and develop specific protocols for this condition.

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Hirayama disease: a rare case report

Taianara Sampaio Reis[1], Amauri Pereira da Silva Filho[2], Melissa Fernandes Vilela de Freitas[3], Danielle Cortellazzi Colonna Romano[3], Gabriela Yumi Nakamura[3], Sabrina Borges Batista[3], Natália Rebeca Karpejany[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil. [2]Hospital de Emergência e Trauma Dom Luiz Gonzaga Fernandes, Campina Grande PB, Brazil. [3]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Taianara Sampaio Reis; Email: taianarasampaio21@hotmail.com.

Case presentation: A 15-year-old female patient with no significant medical history, currently a student, born in Campina Grande and residing in Carurite, presented with a two-year history of progressive muscle weakness, atrophy, and tremors in her right hand since the age of 13. On physical examination, she was in good general condition with nothing relevant regarding other systems. Neurological examination revealed grade IV muscle strength, fasciculations, and significant atrophy in the right hand, with slight tremors at rest and with effort. The patient reported no improvement or worsening of symptoms over the two-year period. Electroneuromyography indicated an exclusively motor, axonal, multifocal, and asymmetric neuropathic process with signs of acute and chronic neurodegenerative changes in the C7, C8, and T1 myotomes. MRI of the cervical and dorsal spine revealed flattening of the anterior cervical spinal cord from C7 to T1. Based on clinical findings and diagnostic tests, the patient was diagnosed with Hirayama disease (HD). **Discussion:** HD also known as Juvenile Muscular Atrophy of the Distal Upper Extremity, is a rare motor neuron disease that affects the anterior spinal cord region spanning from segments C7 to T1 causing progressive muscle atrophy and weakness primarily affecting the unilateral hand and forearm, typically without sensory deficits. HD was first described in 1959, predominantly affecting asian young males. Several etiological factors have been associated with HD, including viral infections, spinal cord vascular insufficiency, high-impact physical activities, and focal spinal cord atrophy during cervical spine flexion. Electrophysiological and histopathological findings indicate a neurogenic disorder involving anterior horn cells and motor axons, resulting in characteristic signs of lower motor neuron impairment. Clinical diagnostic criteria for HD include weakness and atrophy of distal upper limb muscles, onset typically between ages 10 and 20, unilateral or predominantly asymmetric presentation, insidious onset with gradual progression and stabilization, absence of pyramidal signs or sensory disturbances, and exclusion of other differential diagnoses. **Final comments:** The management of HD typically involves conservative measures such as the use of a soft cervical collar and physiotherapy. Although rare, cervical spondylotic amyotrophy (CSA) should be considered in young patients presenting focal asymmetric atrophy of the upper limbs.

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Histopathological substrate of increased T2 signal intensity of the anterior temporal lobe white matter in temporal lobe epilepsy due to hippocampal sclerosis

Ricardo Chmelnistki Wainberg[1], Bruno Loz[1], Augusto Valadão[1], Ricardo Paganin[2], Ricardo Soder[2], Francine H. de Oliveira[3], Leticia Bassani Devens[2], Eliseu Paglioli Neto[1], William Alves Martins[2], André Palmini[2]

[1]Hospital São Lucas, Serviço de Neurocirurgia, Porto Alegre RS, Brazil. [2]Pontifícia Universidade Católica do Rio Grande do Sul, Brain Institute (Bralns), Porto Alegre RS, Brazil. [3]Universidade Federal do Rio Grande do Sul, Faculdade de Medicina, Departamento de Patologia, Porto Alegre RS, Brazil.

Correspondence: Leticia Bassani Devens; Email: leticiabassanidevens@gmail.com.

Background: Temporal lobe epilepsy due to hipocampal sclerosis (TLE/HS) is the most common type of focal epilepsy (1), and the most common cause of refractory epilepsy in adulthood. Alongside HS, MRI commonly reveals subtle alterations in the anterior temporal lobe white matter (aTLWM) of a significant part of TLE/HS patients, yet histopathological characterization of this abnormality has not been strictly studied. **Objective:** To analyze the histological characteristics of the increased T2 signal intensity (iT2SI) in the anterior temporal lobe white matter (aTLWM) in temporal lobe epilepsy due to hippocampal sclerosis (TLE/HS) patients. And to propose an immunohistochemical Photoshop-assisted method to quantify the proportional area of water content, expressed by vacuoles, in the aTLWM. **Methods:** Quantification of neuronal cells, astrocytes, oligodendrocytes and vacuolization in the white matter of surgical temporal lobe specimens was performed by immunohistochemical reaction (NeuN, GFAP, Olig2 and BMP, respectively). Surgical specimens from TLE/HS patients with iT2SI and normal T2 signal in the aTLWM were included for comparison. The histological samples were divided in three groups according the representativeness of the respective Magnetic Resonance location. G1 = representative

samples of iT2SI; G2 = samples from patients with normal T2 signal; and G3 = representative samples of areas with normal T2 signal in the aTLWM of TLE/HS patients with iT2SI. Clinical and demographic aspect were included in the analysis. **Results:** G1 samples showed larger proportional area of vacuolization than the other (p<0.001), but lower number of neurons (p=0,014). Patients with MRI positive for iT2SI presented younger age at surgery (p=0.018) and epilepsy onset (p=0.005). There was a tendency for right side occurrence of ETL/HS in iT2SI patients (p=0.069). **Conclusion:** The higher proportion of vacuoles in the representative samples of iT2SI could explain, at least in part, the histopathologic substrate of this signal alteration in the white matter of temporal lobe in patients with TLE/HS.

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History of polyneuropathy and musculoskeletal manifestations in patients with transthyretin-mediated amyloidosis with cardiomyopathy in APOLLO-B

Martha Grogan[1], Shaun Bender[2], Varun Kumar[2], Kelley Capocelli[2,3]

[1]Mayo Clinic, Rochester, MN, USA.
[2]Alnylam Pharmaceuticals, Cambridge, MA, USA.
[3]Current Employee of Scholar Rock.

Correspondence: Ana Elisa Rodrigues Marangon; Email: elisarois@yahoo.com.br.

Background: Recent evidence has shown that many patients with ATTR amyloidosis develop a mixed phenotype of both polyneuropathy and cardiomyopathy, with polyneuropathy and musculoskeletal symptoms manifesting first, well before a confirmed diagnosis of cardiomyopathy. The **Objective:** To evaluate the occurrence of polyneuropathy and musculoskeletal manifestations prior to a confirmed diagnosis of ATTR amyloidosis with cardiomyopathy (ATTR-CM) in patients enrolled in APOLLO-B (NCT03997383). **Methods:** Baseline disease characteristics and medical history, coded using Medical Dictionary for Regulatory Activities (MedDRA version 23.0), collected during the screening period from APOLLO-B were analyzed in the overall study population and in patients by ATTR amyloidosis type (hATTR, wtATTR) across both treatment arms. The event prevalence and time from event onset to diagnosis of ATTR amyloidosis with cardiomyopathy were summarized descriptively. Additional analyses of polyneuropathy and musculoskeletal manifestations in a matched cohort of patients, as a comparator group, are ongoing. **Results:** Overall, medical histories of 359 ATTR-CM patients were analyzed, of which there were 72 hATTR and 287 wtATTR patients. Polyneuropathy and musculoskeletal manifestations were found in >20% and >60% of all patients, respectively, with these extra-cardiac events occurring at least 3 years prior to a confirmed diagnosis of ATTR-CM. **Conclusion:** This analysis demonstrates that polyneuropathy and musculoskeletal manifestations may be early signs that are potentially overlooked in patients with ATTR-CM. Multisystem clinical assessment and early identification of non-cardiac manifestations are therefore crucial for management of this debilitating condition.

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HIV-associated neuromuscular weakness syndrome: report of two cases presenting with neuromuscular weakness associated with hyperlactatemia

Luiz Carlos Porcello Marrone[1], Renata Siciliani Scalco[1], William Alves Martins[1], Irênio Gomes da Silva[1], Cássia Elisa Marin Dapper[2], Jefferson Becker[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Universidade de Caxias do Sul, Caxias do Sul RS, Brazil.

Correspondence: Cássia Elisa Marin Dapper; Email: cassiaemarin@gmail.com.

Case presentation: Case 1: A 30-year-old HIV-infected woman with a history of pulmonary tuberculosis presented to Hospital São Lucas with paresthesia and weakness in her lower limbs for 2 weeks. She had been on lamivudine, stavudine, and efavirenz for 10 months. Neurological examination showed grade 3 weakness in lower limbs and absent deep tendon reflexes. Her CD4 count was 229/mm^3, and viral load was undetectable. Within 10 days, symptoms progressed to lower and upper limbs, with lactic acidosis (up to 9.3 mmol/L) and autonomic dysfunction. Normal brain CT and CSF analysis, but nerve conduction studies indicated severe axonal sensorimotor polyneuropathy. HAART was discontinued, and she received ICU care with IVIG therapy. She developed respiratory failure, requiring mechanical ventilation and tracheostomy. Over 3 months, her symptoms improved, and she had only mild weakness and was discharged in with normal lactate levels. Case 2: A 44-year-old HIV-positive woman on stavudine, lamivudine, and efavirenz for 1 year was admitted with nausea, vomiting, stomachache, and diarrhea for 7 days. Her CD4 count was 130/mm^3. Initial evaluations were normal, but abdominal x-ray showed bowel distension. Surgical evaluation ruled out acute pathology, and endoscopy revealed duodenitis. After 10 days, she developed symmetric lower limb paresthesia and increased lactate levels (7.5mmol/L). Stavudine was discontinued, and riboflavin was started. Symptoms progressed to involve the upper limbs and cervical region, with hypotension and tachycardia. Lactate levels rose to 10.1 mmol/L, prompting ICU transfer. She developed respiratory failure with visual symptoms, tachypnea, and diaphoresis. Neurological exam showed generalized areflexia, ascending paresis, diplopia, and bitemporal hemianopsia. Mechanical ventilation was initiated, and IVIG commenced. Nerve conduction showed severe axonal sensorimotor polyneuropathy. She was discharged after 44 days without neurological deficits, continuing medication. Lactate levels normalized to 1.1 mmol/L. **Discussion:** HIV-associated neuromuscular weakness syndrome (HANWS) is a rare neuromuscular syndrome linked to hyperlactatemia and exposure to stavudine and/or didanosine. Severe hyperlactatemia and lactic acidosis are rare but serious complications of antiretroviral therapy, associated with dideoxynucleoside exposure, female gender, advanced immunosuppression, and possibly ethnicity. The pathogenesis of this neuromuscular syndrome in HIV-infected patients is likely multifactorial, involving mitochondrial and immunological mechanisms associated with both HIV infection and HAART. Treatment strategies for HANWS remain uncertain yet critical due to its potentially fatal nature. Most patients exhibit axonal alterations; however, the presence of demyelinating findings may guide therapeutic decisions, suggesting the possible use of intravenous immunoglobulin or plasmapheresis. Neurologists must maintain vigilance for diagnosing this syndrome promptly.

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Home protocol proposal for increased activity in individuals after stroke based on specific and task-oriented motor training

Nicoly Alves Machado[1], Jayne Siqueira Cagliari[1], Marco Túlio Almeida Lenza[1], Luciane Aparecida Pascucci Sande de Souza[1], Gustavo José Luvizutto[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Nicoly Alves Machado; Email: nicolyamachado@gmail.com.

Background: Stroke is the leading cause of chronic disability in adults in Brazil. Therefore, it is necessary to institute increasingly early interventions aimed at preventing the onset of secondary complications and the development of compensatory movements. However, the lack of access to a specific motor training protocol in earlier stages can impair the level of motor recovery in individuals. **Objective:** To develop a guide for task-oriented and specific motor training in a home environment and to create a decision tree based on the levels of the Motor Assessment Scale (MAS) for clinical decision-making in patients in the subacute phase of stroke. **Methods:** This is a feasibility study for the development of a guide of task-oriented and specific exercises for the home environment that encompasses all domains of the core tasks and the creation of a decision tree following the levels of the Motor Assessment Scale (MAS) so that from the evaluation of patients, physiotherapists can choose the best exercises from the motor training guide based on the patient's level and to guide tasks based on mobility and motor impairment. **Results:** The primary outcome of this study was the production of a guide of task-oriented and specific exercises for the home environment, divided into the domains of core tasks with 20 exercises covering rolling (4 exercises), sitting (3 exercises), standing (3 exercises), walking (4 exercises), reaching and grasping (3 exercises), and grasping and manipulation (3 exercises). The decision tree for selecting the best exercise was based on the Motor Assessment Scale (MAS), divided into 8 items: 1- supine to lateral on the intact side; 2- supine to sitting on the side of the bed; 3- sitting balance; 4- sitting to standing; 5- walking; 6- upper limb function; 7- hand movements; and 8- advanced hand activities, with scores from 1 to 5 for classifying the level of mobility and motor impairment. Thus, at the end of the decision tree, we selected the most specific and efficient exercises for each level of mobility and motor impairment. **Conclusion:** We infer that the creation and dissemination of this guide of task-oriented and specific exercises for the home environment and the decision tree following the MAS motor levels to the community and physiotherapists is essential for professionals to have options in their clinical decision-making based on the mobility and motor impairment levels of their patients, and for the community to have an accessible, applicable, and cost-effective guide of task-oriented and specific exercises for the home environment to increase the level of activity post-stroke.

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Hopkins syndrome, a breath of mystery: a case report

Antonio Edvan Camelo-Filho[1], Sabrina Soares Timbó[2], Amanda Camelo Paulino[2], Maria Luiza Miranda Carneiro[2], François Loiola Ponte de Souza[2]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Hospital Infantil Albert Sabin, Fortaleza CE, Brazil.

Correspondence: Antonio Edvan Camelo Filho; Email: edvan.camelo@gmail.com.

Case presentation: A 2-year-old boy presented with progressive left leg weakness following a recent episode of an acute asthma exacerbation. The child had normal prior neuromuscular development and started walking around one year of age. His past medical history was significant for asthma, requiring hospitalization for an acute exacerbation five months before this evaluation. Two weeks after the asthma exacerbation, the patient developed weakness in his left leg, leaving him unable to stand or walk without assistance. He was initially diagnosed with hip synovitis and treated with non-steroidal anti-inflammatory drugs and physiotherapy. Due to a lack of improvement, he was referred to a pediatric reference center for further evaluation. Initial physical examination revealed left lower limb weakness, including hip flexion (MRC 2), ankle dorsiflexion (MRC 2), foot drop, and lateral rotation of the affected limb. This weakness was accompanied by

moderate limb atrophy and absent patellar and ankle reflexes. There were no fasciculations or sensory, cerebellar, or upper limb deficits. Electroneuromyography and muscle ultrasound confirmed a chronic asymmetric anterior horn cell disease with moderate left lumbosacral (L2-S2) compromise without acute ongoing denervation. Further investigations, including serology, spine MRI, and cerebrospinal fluid analysis, ruled out potential infections and compressive and inflammatory causes. The patient was, therefore, diagnosed with Hopkins syndrome. **Discussion:** Hopkins syndrome, characterized by poliomyelitis-like paralysis after an asthma episode, is a rare and challenging condition. This case highlights the differential diagnosis process, distinguishing Hopkins syndrome from other lower motor neuron syndromes. The temporal association between asthma exacerbation and paralysis and the clinical and electrophysiological findings were crucial in reaching the diagnosis. The exact mechanism linking asthma with amyotrophy remains elusive. Several hypotheses have been proposed, including nerve plexus damage from limb immobilization or intravenous cannulation, congestion of the brachial plexus due to severe dyspnea, and neurotoxicity or hypersensitivity reactions to medications. Typically, this condition develops within days to weeks after an acute asthma attack and is characterized by the sudden onset of flaccid paralysis affecting one or both limbs. This paralysis often progresses to severe muscle atrophy in the affected area. Corticosteroid treatment generally proves ineffective, and recurrences are uncommon. **Final comments:** This case emphasizes the importance of considering Hopkins syndrome in the differential diagnosis of any flaccid paralysis in children, particularly those with a history of asthma and asymmetric lower limb weakness. Early recognition and exclusion of differential diagnosis with an extensive investigation are vital for accurate diagnosis and appropriate management.

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Horner's syndrome as a complication of interscalene brachial plexus block: a case report

Maria Fernanda de Abreu Rodrigues[1], Josevânia Fulgêncio de Lima Arruda[1], Armando D'Lucca de Castro e Silva[2] , Maria Eduarda de Abreu Rodrigues[3], Liene Duarte Silva[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Hospital Central do Exército, Rio de Janeiro RJ, Brazil.
[3]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Maria Fernanda de Abreu Rodrigues; Email: maferodriguesufrj@gmail.com.

Case presentation: A 60y female with well-managed hypothyroidism, on a daily regimen of 50 mcg levothyroxine, underwent an elective arthroscopic repair for a traumatic rupture of the right rotator cuff. Anesthesia was administered intravenously, supplemented with an ultrasound-guided interscalene brachial plexus block using 20 ml of 0.3% ropivacaine (60mg). The procedure was initially uneventful. Postoperatively, the patient exhibited ipsilateral eyelid ptosis and miosis, but anhidrosis was not relevant. Notably, these symptoms of Horner's syndrome (HS) resolved spontaneously within 24 hours, without the need for further intervention. The patient was discharged without complications. **Discussion:** HS is a rare condition, characterized by the classic triad of ptosis, miosis/anisocoria, and anhidrosis affecting the ipsilateral face. This syndrome results from disruption of the oculosympathetic pathway and may arise due to iatrogenic causes, systemic diseases or traumatic injury. This case underscores the potential association between HS and interscalene brachial plexus block. The incidence of HS as a complication following an ultrasound-guided interscalene block with ropivacaine is approximately 9%. This adverse event occurs due to inadvertent diffusion of the local anesthetic to the cervical sympathetic chain, leading to a blockade of the ipsilateral stellate ganglion. **Final comments:** Despite its transient nature, HS may cause significant stress and discomfort to patients, particularly if they are not forewarned about this potential complication. It is imperative for clinicians, especially to neurologists, to recognize and appropriately manage this condition. Preoperative counseling regarding the possibility of HS should be considered to alleviate patient anxiety and enhance informed consent.

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Hospital admissions and deaths due to Alzheimer's disease in Brazil: epidemiological analysis 2019-2023

Monyque Pinheiro Charchar[1] , Marcos Lude da Silva Ferreira[2], Alec Peixoto Rabelo[2], Edson Luis de Barros Siqueira[2], Luiz Carlos Figueiredo Filho[2], Luan Nascimento Pereira Amorim[2]

[1]Centro Universitário do Estado do Pará, Belém PA, Brazil.
[2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Monyque Pinheiro Charchar; Email: mcharchar10@gmail.com.

Background: As life expectancy increases, certain diseases have become more prevalent in society. Notably, dementia, particularly Alzheimer's disease (AD), has become a significant concern. By 2020, the number of people living with dementia reached 50 million worldwide, making it a critical public health issue. **Methods:** This study is retrospective, descriptive, and quantitative in nature. Data on hospital admissions and deaths due to AD in Brazil over the last 5 years were analyzed. Variables such as regions, urgent cases, deaths, mortality rate, average annual stay, total cost, age group, gender, and race were examined. Data was obtained from the Hospital Information System of the Brazilian Unified Health System (SIH/SUS) between 2019 and 2023, publicly available via the SUS Information Technology Department (DATASUS) segment TABNET. **Results:** Between 2019 and 2023, Brazil recorded 7,866 hospital admissions due to AD, with 46.6% from the Southeast region and the highest rates in São Paulo (1,815). Rondônia, Pernambuco, Rio Grande do Sul, and the Federal District also had notable numbers, especially in 2023. Urgent admissions predominated in all states except Rio Grande do Norte and the Federal District, totaling 6,288, while elective admissions totaled 1,578. There were 1,905 deaths during this period, with 25.6% in 2023, and 58% in the Southeast region, which also had the highest mortality rate (30.08), peaking in São Paulo. The Midwest region had a significant mortality rate (28.02), especially in the Federal District. The average annual stay was 19.3 days, with higher values in the Southeast (25.4) and Northeast (21.4), particularly in Rio de Janeiro and Rio Grande do Norte. The total cost during the period was 12.3 million Brazilian reais, with the highest expenditure in 2023, representing 64.1% from the Southeast. The average expenditure was 1,565.97 Brazilian reais, peaking in 2020. The most affected age group was 80 years or older, accounting for 59.7% of cases and the highest death rate (1,338). Females were the most affected gender in all states except Piauí, representing 65.5% of cases and also had the highest death rate. The most affected races with the highest death rates were brown in the North, Northeast, and Midwest, and white in the South and Southeast. **Conclusion:** The analysis reveals a significant number of hospital admissions and deaths, especially in the Southeast region and São Paulo. Additionally, the highest expenditure was concentrated in this region, indicating a significant disparity in Brazil. The most affected age group is 80 years or older, which should be considered in health guidelines. Female gender, brown, and white race were identified as relevant risk factors for the disease. This survey is expected to aid in defining more efficient public health policies based on national reality.

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Hospital admissions for migraine and other pain syndromes among children and teenagers in Brazil between 2018 and 2023: an analysis of DATASUS

Arthur Vinícius Feitosa Santos[1], Luana Cristina de Oliveira Lima[1], Vagner Costa da Cunha Júnior[1], Maria Cecília Leal Menezes[1], Sabrina Sthéphanie Ataíde Martins[1], Thiago Pariz da Silva Dumas[1], Arthur Aragão Nunes[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Arthur Vinicius Feitosa Santos; Email: arthur.vinicius04@souunit.com.br.

Background: Migraine is a neurological, chronic and multifactorial disease with genetic predisposition and association with environmental factors (poor quality of sleep, excessive use of caffeine, painkillers, fatty and spicy foods, etc.), hormonal, physical and mental factors. Its main characteristics are unilateral or bilateral throbbing headache, photosensitivity, nausea, vomiting, etc. **Objective:** To analyze hospitalizations for migraines and other headaches in Brazil and its regions during the period from January 2018 to December 2023. **Methods:** Descriptive epidemiological study using secondary data from the SUS Hospital Information System (SIH/SUS) Tabnet, being tabulated in Microsoft Excel, according to data collected from hospitalizations during the period from January 2018 to December 2023. The variables analyzed were federative region, gender, race/ethnicity and age group. **Results:** In the period analyzed, there were a total of 10,313 hospitalizations for migraines and other pains, of which 62.05% (6399 cases) affected females and 37.86% (3904) males. It is noteworthy that, from 0 to 9 years old, this disease had a similar prevalence between both sexes, however, from 10 to 19 years old there was a discrepancy of 1.96 times more for females (2448 hospitalizations), The age group that most contributed to this was 15 to 19 years old. It should be added that the regions with the highest prevalence were the Northeast (2079 hospitalizations) and the Southeast (2060). Regarding ethnicity, it was observed that 50.64% (5223) of the patients analyzed were black, 32.97% (3400) were white and 13.84% (1427) did not self-declare. **Conclusion:** Based on the results, it was found that hospitalization for migraine is more prevalent in the female population, especially those aged 15 to 19, demonstrating an association between this and hormonal factors, as menstrual irregularities, endometriosis, polycystic ovaries and hormone replacement, These may be factors that aggravate this pathology, making it more resistant and more frequent. Furthermore, these pains are more present in the Northeast and Southeast regions, which may correlate with the fact that the first is a place with high temperatures, leading to dehydration and changes in metabolism and, consequently, to crises while the second is marked by intense urbanization and exposure to photo-sound factors and stressors, which can contribute to the high prevalence of the disease. Therefore, the need to create public policies aimed at preventing and monitoring these headaches is evident in order to inform the population about the best control mechanisms.

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Hospital admissions for viral encephalitis: a critical analysis

Felipe Andreani Camago Manduco[1], Bruna Andrello Lameira[1], Gabriel Felipe Lorençato[1], Manuela Silva da Costa[1], Mario Silva Jorge[1], Regina Salles Cauduro[1]

[1]Humanitas, São José dos Campos SP, Brazil.

Correspondence: Felipe Andreani Camargo Manduco; Email: felipemanduco1000@icloud.com.

Background: Encephalitis is an inflammatory process of the brain parenchyma that leads to brain dysfunction. It is usually an acute process, but it can be post-infectious encephalomyelitis, a chronic degenerative disease or a slowly progressing viral infection. Viral infections are the main causes of acute infectious encephalitis and the main viruses are: enterovirus, arbovirus and herpes virus. Although systemic viral infections by the etiological agents most implicated in their development are common, the occurrence of encephalitis is uncommon. Despite being relatively rare, it is a clinical condition that presents high rates of morbidity and mortality, especially when there is a delay in its recognition and management, giving it great epidemiological relevance. It manifests with a typical viral prodrome (fever, chills, myalgias) usually precedes brain involvement, ranging from subtle confusion to obtundation. **Objective:** Updated analysis of hospital admissions from the Basic Health Service (SUS) for viral encephalitis in Brazil between 2008 and 2024. **Methods:** The research was based on information on hospital morbidity from the SUS present in the Ministry of Health database (Tabnet) referring to the numbers of hospitalizations related to viral encephalitis, related to Brazilian regions and age groups. In which, the data collected takes place between the years 2008 and until the first 3 months of 2024, and between the age group of 0 to 14 years old. **Results:** From 2008 to March 2024, viral encephalitis was responsible for 28,507 SUS hospital admissions, with a predominance in the age group between 0 and 14 years old, which corresponds to 44.14% of viral encephalitis admissions. From 2013 onwards, there was an increase in hospitalizations for viral encephalitis of 25%, compared to 2012, and with a concomitant increase in hospitalizations in the Northeast region, with an increase of 115% compared to the previous year. Therefore, it affects more children aged 1 to 9 years (28% of all hospitalizations), mainly in the Northeast and Southeast regions (amounting to approximately 61% in the same age group). Another detail consists of the national stabilization of hospitalizations from 2015 onwards, with a predominance in the Northeast and Southeast regions. However, there was a decrease of 30,6% of the hospitalizations in the year of 2020 and a small variation in 2021, compatible with the pandemic years of COVID, and returning in 2022 to the same stabilization of the last decade. **Conclusion:** We can conclude that there has been an increase in hospitalizations for viral encephalitis in the last decade, suggestive of an increase in case notifications, improvements in the quality of health services and epidemic outbreaks of arboviruses. In the years affected by COVID-19 (2020 and 2021), there was a decrease in numbers, although this was not known to be due to a decrease in registration or lower medical demand due to the isolation resulting from the pandemic.

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Hospital costs associated with malignant brain neoplasms in Brazil: an analysis of the last 5 years

Italo Lafayette Silva[1], Lucas Roque Evangelista[1], Dayanne Garcia de Freitas[1], Renan de Holanda Alves[1], Nicolle Gabrielle Lima Silva[1], Henrique Katsumi Kawano[1], Gilberto de Paula Pinheiro Filho[1], Renato Vieira de Freitas Filho[1]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Italo Lafayette Silva; Email: italolafayette@gmail.com.

Background: Malignant brain cancer is defined by abnormal and uncontrolled cell growth in brain tissue, which can invade the brain locally and potentially spread to other regions of the central nervous system and other organs through metastasis. This condition not only profoundly impacts the lives of affected individuals, but also represents a significant challenge for society. In this context, it is crucial to analyze the hospital costs associated with this disease. **Objective:** The purpose of this study is to comprehensively investigate hospital costs related to malignant neoplasms of the brain in Brazil over the last five years, assessing the progression of these costs, the states with the highest and lowest investments and analyzing regional variations. **Methods:** A descriptive and quantitative study was carried out for this study. The data was obtained from the SUS Hospital Information System (SIH/SUS), made available by the Department of Informatics of the Unified Health System (DATASUS). covering the period from 2019 to 2023. The variables analyzed were: geographic region, federation unit and total hospital expenditure. **Results:** In the last five years, hospital costs associated with malignant neoplasms of the encephalon in Brazil totaled R$195,471,785.44, representing an increase of 23.20% in the period from 2019 to 2023. São Paulo led the way with the highest accumulated expenditure of R$46,635,530.08, followed by Minas Gerais with R$ 22,336,829.92 and Bahia with R$ 11,266,907.39. On the other hand, Amapá had the lowest total expenditure, totaling R$ 128,888.09. Roraima stood out for the largest percentage increase in spending over these five years, growing 1248.24%, from R$ 7,709.19 in 2019 to R$ 103,938.96 in 2023. The highest annual spending averages were seen in São Paulo (R$ 9,327,106.02), Minas Gerais (R$ 4,467,365.98) and Bahia (R$ 2,253,381.48), while Sergipe (R$ 138,387.11), Roraima (R$ 42,421.83) and Amapá (R$ 25,777.62) recorded the lowest averages. The year 2023 saw the highest total disbursement, reaching R$45,022,615.79, while 2020 saw the lowest spending, with R$ 35,529,263.18. São Paulo recorded the highest spending by state and year in 2023, totaling R$ 10,233,749.95, while Roraima had the lowest spending in 2019, with R$ 7,709.19. The regional analysis revealed that the Southeast region led in terms of total accumulated, reaching R$ 108,489,020.68, while the North region had the lowest accumulated, totaling R$ 7,082,581.06. **Conclusion:** The study revealed a significant increase in spending. São Paulo led the way with the highest accumulated spending, while Amapá had the lowest total spending. Roraima stood out with a significant increase in costs, suggesting possible improvements in the early detection of the disease, greater access to specialized health services or even an increase in the number of diagnoses made. This suggests that further research is needed to understand the causes of this significant increase.

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hospitalization due to alzheimer's disease in brazil between 2019 and 2023

Kaike Eduardo da Silva Loboi, Leonardo Bastos Santos[1], Felipe Moraes Costa[1], Ruan Pablo da Silva Gomes[1], Arley Nikson Oliveira Barreto[1], Marcus Paulo Oliveira Lopes[1], João Lucas Lalor Tavares[1], Leonardo Aguiar dos Santos[1], Arthur Moraes dos Santos[1], Flávio Lobo Altieri[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Marcus Paulo Oliveira Lopes; Email: marcus.polopes@aluno.uepa.br.

Background: Alzheimer's disease (AD) is a neurodegenerative disorder associated with aging, characterized by progressive and irreversible cognitive decline. AD manifests clinically with insidious impairment of higher intellectual function, memory, mood and behavior. As the disease progresses, individuals may experience aphasia and disorientation. Epidemiologically, AD predominantly affects individuals over 60 years old, with a higher prevalence among women. Hospitalizations often occur due to the advancing stages of the disease, highlighting its profound impact on patient health and wellbeing. **Objective:** Analyze the epidemiological profile of hospitalizations due to AD in Brazil from 2019 to 2023. **Methods:** This is a descriptive ecological study, using secondary data in the public domain, obtained from the Hospital Information System (SIH-SUS) in the database of the Information Technology Department of the Unified Health System (DATASUS). The variables analyzed were: number of hospitalizations, geographic region, sex, age, race, hospitalization time and financial expenses. Descriptive statistics were performed using absolute (n) and relative (%) frequencies. **Results:** There were 7867 cases of hospitalization due to AD notified during the proposed time period, with an average of 1573 cases per year and an increase from 1600 in 2019 to 2122 cases in 2023. In the distribution by region of the country, it was observed that approximately 46.7% (3673) occurred in the Southeast, 25.5% (2009) in the South, 17.8% (1398) in the Northeast, 6.5% (515) in the Midwest, and 3.4% (272) in the North. The population was majorly female, with 65.84% (5180) of cases, and just 34.16% male (2687). Regarding the age group, the most affected by AD were the ones between 60 and 69 years of age, with 9.05% (712), 70 and 79 years, with 27.9% (2195) and 80 or plus years, with 59.73% (4699). As to race, the most affected were white, with 50.56% (3978), and brown, with 26.42% (2079). The average stay of hospitalization was 19.6 days, with direct healthcare expenses of R$ 2.463.649,8/year. **Conclusion:** Between 2019 and 2023, AD hospitalizations in Brazil grew significantly, with most cases occurring in the Southeast region and predominantly affecting females, whites, and individuals over 80 years old. Given the disabling and debilitating nature of this condition and the substantial public expenditure on its treatment, it is essential to understand the epidemiology of the affected population to develop strategies that enhance their quality of life.

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Hospitalization period in a stroke unit associated with worse functional outcome at 90 days: a cohort study in a reference hospital in Bahia

Gabriel Teles de Oliveira Piñeiro[1], Maria Eduarda Messias Vassoler[1], Gabriel Souza Medrado Nunes[1], Milena Fernandes de Oliveira[1], Luiz Paulo Oliveira Martins[1], Thaís Barbosa de Oliveira[1], João Pedro Fernandes Gonçalves[1], Pedro Antonio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Antonio Lázaro Santos Soares Junior; Email: antoniolssj@ufba.br.

Background: Hospitalization is known to be associated with worse outcomes. However, the correlation between hospitalization and functional outcome at 90 days is not well explored in many populations, including patients with acute neurological injuries. **Objective:** To understand the association between hospitalized duration and functional outcomes in acute stroke patients. **Methods:** This study enrolled patients with acute stroke admitted to a stroke unit at a referral hospital in Bahia, between July/2023 and March/2024. Clinical and epidemiological characteristics were extracted from patients' medical records or obtained through personal interviews. Infection was defined as the administration of antibiotics or documented infection noted in medical records. CAM-ICU was performed by trained researchers and used to assess delirium. The assessment of functional outcome and survival was performed using the modified Rankin Scale (mRS) validated for telephone interviews. Worst functional outcome was defined as an mRS greater than 2. The Mann-Whitney test was used in the univariate regression analyses and a multivariate analysis was made using logistic regression. For predictors in the multivariate analysis, previous studies by the same cohort were used. **Results:** The study comprised 153 patients, of whom 49% were female, with a mean age of 64.9±12.5 years. Within this cohort, 51.6% were mixed-race. Regarding comorbidities, 34.6% had diabetes, 80.4% had hypertension, and 29.4% had dyslipidemia. Among all patients, the median NIHSS at admission was 7 (IQR: 4-11). A total of 142 patients (92.8%) had ischemic strokes, and 90 patients (58.8%) underwent thrombolysis. Infection was diagnosed in 63 patients (41.2%). Hospitalization

had a median length of 7 days (IQR: 5-13). The median mRS at 90 days was 2 (IQR: 0-4), with a total of 8 deaths at this mark. Delirium occurred in 32 patients (20.9%) at least once during hospital stay. The univariate regression showed that hospitalization was associated with higher mRS at 90 days (p<0.001). In the multivariate analysis, adjusted for age, hypertension, delirium, thrombolysis, NIHSS, and infection, the length of hospital stay remained significant (p<0.05; OR 1.07: CI 1.01-1.15). **Conclusion:** In the present study, the length of stay was identified as an independent predictor of worse functional outcomes on the 90th day. However, further research is necessary to gain a deeper understanding of this association.

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Hospitalizations for Alzheimer's disease in the southeast region of Brazil: an epidemiological study (2013-2023)

Italo Lafayette Silva[1], Lucas Roque Evangelista[1], Dayanne Garcia de Freitas[1], Nicolle Gabrielle Lima Silva[1], Gilberto de Paula Pinheiro Filho[1], Renato Vieira de Freitas Filho[1], Henrique Katsumi Kawano[1], Renan de Holanda Alves[1]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Italo Lafayette Silva; Email: italolafayette@gmail.com.

Background: Alzheimer's disease (AD) is a progressive and fatal neurodegenerative disorder that not only has a profound impact on the individuals affected, but also has significant repercussions for their families and society in general. In this context, understanding the pattern of hospital admissions related to AD is extremely important. **Methods:** For this study, a cross-sectional, descriptive and quantitative epidemiological survey was conducted. Data on Alzheimer's-related hospitalizations in the Southeast region of Brazil, from January 2013 to December 2023, were collected using the SUS Hospital Morbidity Information System (SIH/SUS), provided by the Department of Informatics of the Unified Health System (DATASUS). The variables analyzed included aspects such as geographic region, gender, race, type of hospitalization (elective or urgent) and age group. **Objective:** To analyze temporal trends and understand the dynamics of Alzheimer's hospitalizations in the Southeast region of Brazil, providing fundamental scientific input for the development of effective strategies and interventions to tackle this increasingly prevalent neurodegenerative disease. **Results:** Over the last decade, the Southeast region of Brazil has witnessed a total of 8,717 hospitalizations for AD. São Paulo leads the way with 4,633 cases (53.17%), followed by Rio de Janeiro with 2,296 (26.34%), Minas Gerais with 1,458 (16.74%) and Espírito Santo with 330 (3.78%). This period saw a percentage increase of approximately 56% in total hospitalizations. In 2015, there was the highest annual peak, reaching 1,040 episodes, while 2020 marked the lowest number, with 566 hospitalizations, it should be noted that the COVID-19 pandemic may have significantly impacted this number. In terms of patient profile, women accounted for the majority of hospitalizations, with 66% of the total. As for ethnicity, white people were the most affected, accounting for 50.74%, followed by brown people (19.22%), black people (5.84%), yellow people (1.08%) and people with no information on race accounted for 23.12% of hospitalizations. In terms of age group, patients over 80 were the most frequently hospitalized, accounting for 5,225 cases (59.94%) over the decade. In the context of hospitalizations, emergency admissions predominated, accounting for 74.6% of the total (6,501 out of 8,717). Elective admissions accounted for the remaining 25.4% (2,216 out of 8,717). **Conclusion:** This study reveals that, in the Southeast region of Brazil, hospitalizations for Alzheimer's are predominantly for women, from São Paulo, of white ethnicity and aged over 80. The high proportion of emergency admissions underlines the urgent need for effective management strategies for this neurodegenerative disease. There has also been a significant 56% increase in hospitalizations over the last decade, indicating a growing need for resources and policies to address this health challenge.

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Hospitalizations for multiple sclerosis in the Brazilian Northeast: an analysis of DATASUS

Maria Cecília Leal Menezes[1], Luana Cristina de Oliveira Lima[1], Arthur Vinícius Feitosa Santos[1], Vagner Costa da Cunha Júnior[1], Sabrina Sthéphanie Ataíde Martins[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Maria Cecília Leal Menezes; Email: maricecilm00@gmail.com.

Background: Multiple sclerosis (MS) is a demyelinating, autoimmune and chronic neurodegenerative disease, caused by inflammatory and degenerative mechanisms that compromise the neurons of the central nervous system. This health condition can cause difficulties in carrying out activities of daily living and impact the individual's quality of life. Despite MS affecting individuals of both sexes and all ages, there are higher rates of hospitalizations in females and adults. **Objective:** To characterize the profile, according to epidemiological indicators, of hospitalizations for MS in the Northeast in the period between January 2018 and March 2024. **Methods:** This is a study with a quantitative approach, conducted based on an analysis of the database of the Hospital Information System of the SUS (SIH/SUS) and the Brazilian Institute of Geography and Statistics (IBGE) regarding hospitalizations for MS in the Northeast Region of Brazil. Information was collected on the patients' age group, gender, year of hospitalization and federation unit. The quantitative data were organized in a spreadsheet using the Microsoft Excel program. **Results:** The sample studied consisted of 2,941 hospitalizations for MS, with a high prevalence in adults between 30 and 39 years old (29.4%). In relation to Brazilian states, the highest number of cases (847) and numbers per 100,000 residents (38.3) occurred in Sergipe, with emphasis on hospitalizations in the age group between 30 and 39 years old. Furthermore, the Northeast state with the lowest admissions was Alagoas (31), which also had the lowest number of cases per 100,000 inhabitants (0.9). Even more, there was a high number of hospitalizations in women throughout the Northeast (70.5%), especially in Sergipe (655 patients). When analyzing ethnicity, brown color reached a percentile of 59.9% (1,764 hospitalizations) and black color reached 1.6% (49). An 80.08-fold increase in total cases was also observed in 2023 compared to 2017. **Conclusion:** Based on the results, it was found that hospitalizations for MS are more prevalent in women, both in the Northeast region in general, as in Sergipe. Moreover, a majority of admissions were found in adults aged 30 to 39 years, in the mixed-race population and in the state of Sergipe. Therefore, racial, geographic, age factors and the gender of patients were suggested in order to justify the distribution of multiple sclerosis in the Northeast so that effective public policies can be created aimed at preventing the pathology, ensuring better control of it.

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Hospitalizations for neurotic, stress-related, and somatoform disorders in Brazil from 2019 to 2023: an epidemiological study

Ana Clara Lacerda Freitas[1], Aline Teixeira[1], Laysa de Souza Maia[2], Andressa Gonçalves Vicente[3], Ana Alice Lemos Lima[4], Gabriela Hernandez Dumani[5], Ana Cristina Carvalho e Silva[3], Estefany Moreno de Souza[6], Geovana Franklim Gomes e Silva[7], Juliana Braga Rodrigues de Castro[8]

[1]Centro Universitário Funorte, Montes Claros MG, Brazil.
[2]Centro Universitário Aparício Carvalho, Jaru RO, Brazil.
[3]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[5]Universidade Franciscana, Santa Maria RS, Brazil.
[6]Universidad de Aquino Bolivia, Santa Cruz de La Sierra, Bolivia.
[7]Centro Universitário de Patos de Minas, Patos de Minas MG, Brazil.
[8]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Ana Clara Lacerda Freitas; Email: anaclf15@yahoo.com.br.

Background: Neurotic disorders, stress-related conditions, and somatoform disorders represent a broad spectrum of conditions that significantly affect individuals' mental health and quality of life. These disorders, often underdiagnosed and undertreated, are characterized by a variety of symptoms ranging from excessive anxiety and fear to clinically unexplained physiological reactions. In 2020, the prevalence of Post-Traumatic Stress Disorder (PTSD) in Brazil was approximately 8.5% of the population. **Objective:** To analyze the profile of hospitalizations due to neurotic, stress-related, and somatoform disorders across Brazilian states over the past five years. **Methods:** This ecological, cross-sectional time-series study was conducted using data provided by the Hospital Information System of the Brazilian Unified Health System (SIH/SUS) from 2019 to 2023. The variables considered included hospitalizations by demographic region, sex, age group, and race. Data collection and analysis were performed in June 2024. **Results:** There was a notable 37.5% increase in the number of hospitalizations by the public health network from 2019 to 2023, reaching approximately 4,128 hospitalizations in the last year analyzed. The Southeast region had the highest number of cases compared to other regions, while the North region had the lowest number of cases throughout the years analyzed. Regarding sex, there was a higher prevalence of cases among females, with approximately 10,206 (65.8%) cases, compared to 5,297 (34.2%) cases among males. Additionally, individuals aged 20 to 29 years and of white race were the most affected by these disorders. **Conclusion:** There has been a significant increase in the number of hospitalizations over the analyzed years. It was observed that these disorders affect almost twice as many women compared to men. Therefore, the current challenge for the country is the prevention and treatment of these disorders, aiming for early diagnosis and appropriate therapy to ensure the mental and physical well-being of these patients, as well as their greater integration into society.

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Hospitalizations for transient ischemic stroke in women under 60 in the period from 2020 to 2023: an analysis of risk factors and lifestyle habits of women in the pandemic and post-pandemic period

Paola Carolina Santos Foinkinos[1], Maria Eduarda da Silva Santos[1], Matheus Prado dos Santos[1], Suzana Gomes de Oliveira[1], Zulma Catarina dos Santos Silva[1], Matheus de Carvalho Alves[1], Louise Araújo Corrêa[1], Bruna Martyres Gueiros[1], Hellen Jost Evaldt[2], Larissa Ádila Desireé Vieira de Andrade Duarte[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.
[2]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Paola Carolina Santos Foinkinos; Email: paola.csfoinkinos@aluno.uepa.br.

Background: Transient ischemic strokes occur when there is an alteration in brain function caused by a temporary, short-term blockage of the blood supply to the brain. In this sense, there are intrinsic risk factors associated with this pathology, such as gender, age and family history, in which there is a prevalence of risk in women over 55 with a previous family history of stroke. There are also extrinsic factors such as smoking, systemic arterial hypertension, a sedentary lifestyle, type 2 diabetes mellitus, among others, which can be avoided. **Objective:** To identify and analyze data on hospital admissions for strokes between 2020 and 2023 in Brazil, correlating with risk factors and lifestyle habits of women under 60. **Methods:** An ecological study was carried out using secondary data from the SUS Hospital Information System (SIH/SUS), using code G.45 of the ICD10 classification. From the search, information was collected and analyzed on hospital admissions for Transient Ischemic Stroke and Related Syndromes according to the variables regions of Brazil, age group, sex and period: women aged 20 to 59, in the historical series from 2020 to 2023 in the North, Northeast, Midwest, South and Southeast regions. **Results:** Between 2020 and 2023, a total of 10,727 hospital admissions of women aged 20 to 59 due to transient ischemic stroke and related syndromes were recorded in the sum of all regions of Brazil. Among these, the region with the highest prevalence of hospitalizations is the Southeast (4,322), followed by the South, Northeast, North and Midwest, respectively. In addition, there has been an increase over the years, with a total of 2,206 hospitalizations in 2020 and 2,964 in 2023, an increase of approximately 35% when comparing the pandemic (2020/2021/2022) and post-pandemic (2023) periods. **Conclusion:** Therefore, based on the data analyzed, it can be concluded that the Southeast and Midwest regions stand out on the national scene as the regions with the highest and lowest prevalence of hospitalizations, respectively. In addition, the 35% increase recorded between the pandemic and post-pandemic periods is relevant in the analysis carried out. Thus, the data collected reinforces that the pandemic context has had an impact on the lifestyle habits of women under 60 years of age, increasing the risk factors for developing strokes and their clinical complications, thus having implications for hospitalizations for cerebrovascular diseases in Brazil.

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Hospitalizations for viral encephalitis in the northern region of Brazil in the pre-pandemic and pandemic period (2016-2019/2020-2023)

Erminiana Damiani de Mendonça[1], Maria Eduarda Tinti[1], Antônio Marcos Rabelo Barbosa[1], Ruth Léia Souza Alves Gomes[1], Guilherme Magalhães Rezende[1], Guilherme Reis Silva[1]

[1]Universidade Federal do Tocantins, Palmas TO, Brazil.

Correspondence: Erminiana Damiani de Mendonça; Email: erminiana@mail.uft.edu.br.

Background: Viral encephalitis is a condition characterized by inflammation of the brain parenchyma tissue, triggered by a viral infection. In addition to severe neurological impairment, patients often present symptoms such as fever, headache, seizures and altered level of consciousness, and some viruses may be associated with high mortality rates. Geographical distribution plays a crucial role in the spread of viral agents, along with seasonal variations and the individual's immune status. In Brazil, this disease is responsible for high hospitalization rates, emerging as a public health challenge. **Objective:** To identify and quantitatively evaluate the presence of viral encephalitis in the north of the country, based on the number of hospitalizations and the mortality rate of this disease during the pre-pandemic and pandemic periods. **Methods:** This is a quantitative and descriptive study based on data collected from the Hospital Information System of the Unified Health Service (SIH/SUS). Data was collected on the number of hospitalizations and the mortality rate due to viral encephalitis in the federal units of northern Brazil, evaluating age, gender and ethnicity as the main variables. Data collection was limited to the period from 2016 to 2023. **Results:** Between 2016 and 2019, there was a growing increase in the number of cases, with a total of 201 cases in 2019. However, there was a reduction of 11.94% and 16.38% in the number of cases in 2020 and 2021 across the Northern region, respectively, compared to the previous year. From 2022 onwards, the number of cases increased again, with values similar to the pre-pandemic period. The states of Amazonas and Pará led the number of cases throughout the period analyzed. The epidemiology of the affected population remained unchanged in 2020 and 2021, with only a quantitative reduction. Thus, children aged 1 to 4 years were among the most extensively affected age group. With regard to gender and ethnicity, there was a prevalence of male and eastern asian cases. The mortality rate was more prevalent among people aged 70-79. **Conclusion:** It can be concluded that the number of hospitalizations was higher in the pre-pandemic period. Amazonas and Pará led the way in the number of hospitalizations in both periods. In terms of deaths, Pará and Roraima stand out as the states with the highest incidence. In addition, there was a predominance of cases among males and people of asian ethnic and in the 1 to 4 year age group. Mortality rates were higher among the elderly. These findings emphasize the need for continuous monitoring and specific measures for vulnerable groups, as well as strengthening preventive and control strategies to protect susceptible populations.

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Hospitalizations in Brazil due to schizophrenia spectrum disorder in childhood

Helena Rubini Nogueira[1], Helena Rubini Nogueira[1], Giovanna Ferraza Bragante[1], Luísa Montanini Baptista Nunes[1], Mariana de França[1], Milena de Lara Macedo[1], Natália Lima dos Santos[1]

[1]Universidade Municipal de São Caetano do Sul, São Caetano do Sul SP, Brazil.

Correspondence: Helena Rubini Nogueira; Email: helena.rnogueira@live.com.

Background: The pathophysiology of schizophrenia is based on neurochemical and anatomical alterations, as well as genetic and environmental aspects. When it starts early, the symptoms are considered to be non-specific, but the influence of traumatic events on its etiology is emphasized, resulting in the advent of psychic splitting as self-protection, although diagnosis at an earlier age is rare. Childhood schizophrenia is therefore considered to be when symptoms appear before the age of 13, with an estimated incidence of 0.1-1% before the age of 10 (a frequency 50 times lower than in those over 15). The treatments indicated in this scenario consist of typical and atypical antipsychotics, psychotherapy, electroconvulsive therapy (ECT) and transcranial magnetic stimulation (TMS). **Objective:** To characterize hospital admissions in the last 5 years of brazilian children with schizophrenia spectrum disorder by epidemiological variables. **Methods:** Descriptive analysis based on data obtained from DATASUS using the following variables: ICD 10 morbidity list (schizophrenia, schizotypal and delusional disorders); Age group (0-14 years); Brazilian regions; Sex; Color/race; Hospitalizations; Period (Jan/2019 to Dec/2023). **Results:** During the established period, 3.254 hospitalizations in children under 14 years were accounted for the morbidity. The Southeast region accounted for approximately 38% of this total (n=1.231), and the most prevalent age group was 10 to 14 years old (n=1.138). With regard to the sex affected, males were predominant in practically all age groups, except for those aged between 10 and 14, where around 5% more females were hospitalized. Similarly, females under the age of 14 accounted for the majority of hospitalizations in all Brazilian regions, except for the Northeast (where the male population was 2% higher). In addition, children aged 10 to 14 comprise the age group most vulnerable to hospital admissions for this neuropsychiatric condition (91%; n=2.980), with a predominance of individuals of brown color/race (n=1.165), who are also the majority of those affected in the Southeast (n=555). As for the cases that evolved to death, 1 outcome was recorded, corresponding to the Northeast region and to the age group of children under 1 year old. **Conclusion:** The data shows that the largest number of children diagnosed with schizophrenia spectrum disorders and that required hospitalization are between 10 and 14 years old, female, brown and live in the Southeast. The question remains that, in the area of child neuropsychiatry, diagnosis is a notable obstacle, even more so considering schizophrenia spectrum disorders.

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Hospitals costs, average length of stay and numbers of hospitalizations non-specific stroke from 2018 to 2023 in the regions of Brazil

Amanda Almeida Verçosa[1], Luana Almeida Verçosa[1], Lívia Burim Cerreti[1], Cecília Rioja Gomes[1], Tainan Gomes Ferreira[1], Djalma de Campos Gonçalves Júnior[1], Igor Bueno Garrido[1], Carlos Roberto Massella Junior[2], Caio Cesar Nuto Leite França[3], Cezar Kabbach Calaça Prigenzi[1,3]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.
[2]Hospital Militar de Área de São Paulo, São Paulo SP, Brazil.
[3]Conjunto Hospitalar do Mandaqui, São Paulo SP, Brazil.

Correspondence: Tainan Gomes Ferreira; Email: tainanferreira@uni9.edu.br.

Background: Stroke is a cerebrovascular disease characterized by the interruption of blood flow to the brain, caused by ischemia or hemorrhage.It is currently considered one of the leading causes of mortality in the country. Treatment often requires many strategies and a longer stay by care.The average cost per patient for any type of stroke in Brazilian public hospitals is approximately 9.670,00.Over the last 5 years, the average regional costs was divided into, North of 87.393.440,36, Northeast of 409.856.468, Southeast of 749.305.645,05, South of 336.790.531,65 and Central-West of 115.096.499,46. **Objective:** Temporal and geographic analysis of hospital costs for the treatment of non-specific stroke related to the average length of hospital stay. **Methods:** This is an original, descriptive, and cross-sectional study with data from Datasus, comparing the variables hospitalization authorization (HA), average length of stay, and value of hospital services by year of care according to all regions of Brazil between 2018 and 2023. **Results:** The year of 2018 had the lowest number of HA (n=13,613) and the lowest average length of stay (n=11.1), compared to 2019 that had an increase of 9.64%, duo to the higher number of hospitalizations (n=201,131).In 2020, there was a decrease of 0.25% in the total amount spent on lower HA(n=188,797) and in the average length of stay (n=7.3).However, in 2021 and 2022, both HA (n=200,265 and n=22,703) and the average length of stay(n=7.5 and n=7.8) increased, leading to an increase in the total amount spent by 7.69% and 30.7%, respectively.In 2023 there was a reduction in total spent, resulting in a lower HA(n=198,876) and average length of stay(n=7.5).Nonetheless, HA and the average length of stay were evaluated by region, North (n=61,743 and n=7.8), Central-west (n=60,924 and n=7.3), South(n=190,740 and n=6.9),Northeast (n=279,408 and n=7.8) and Southeast(n=432,570 and n=7.8). The average distribution of total expenditures by region, North 48.2%, Centra-West 13.51%, South 39.48%, Northeast 48.2% and Southeast 88.3%.The

Southeast had the highest number of HA and average total expenditures, while North and Northeast also showed higher expenditures compared to the other regions, which presented lower expenses. **Conclusion:** It is concluded that the years 2019, 2021 and 2022 presented higher hospital spending compared to the years 2018, 2020 and 2023 due to their higher number of HA and average length of stay for the treatment of non-specific stroke. When analyzed by region, the South had the lowest number of stays and the Central-West the lowest number of HA compared to the other regions.However, this did not represent the lowest expenditure, which was observed in the North region.The Southeast region had the highest HA and expenditures, with it is average length of stay being equal to or greater than the other regions.

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Hsan type VII in a family from northeastern brazil

Mariana Alcântara Tavares[1], Lívio Leite Barros[1], Nayana Freire de Almeida Fontes[1], Leonardo Elias Araujo dos Santos[1], Francisco de Assis Aquino Gondim[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Mariana Alcântara Tavares; Email: marianaalcantara004@gmail.com.

Case presentation: In 2016, a 65 year-old man sought medical attention due to severe sensory loss in distal lower and upper extremities.According to him, it started back in his childhood, when working in plantations he would often get hurt without noticing, associated with pain and swelling in his wrists and ankles, which got gradually worse.He denied any comorbidities such as diabetes, but reported that his deceased father had a similar neurological condition and had been diagnosed with leprosy. He was admitted for diagnostic work-up that included a normal spinal tap, and a negative panel of autoimmune labs and leprosy screening. During his admission, his neurological exam revealed widespread areflexia with mild distal weakness and multiple finger and toe deformities due to repeated trauma and shortening of distal phalanges in all fingers, mallet finger deformity in both right hallux and right 2° toe, as well as in left hallux, plantar perforating disease in his left foot and ulcerations in his right hallux. An electromyography revealed a sensorimotor polyneuropathy with more severe sensory findings and some demyelinating motor features.The main diagnostic hypothesis became Hereditary Sensory and Autonomic Neuropathy. In 2019, genetic tests were done in order to determine a possible cause for his disease. A genetic panel for neuropathies revealed two uncertain significance mutations: a C>A variation in the gene SCN11A and a C>T variation in the gene SCN9A, both in heterozygosis, 1 copy of each. In the SCN11A gene, the variation happened in the chr3:38.926.843, causing the substitution of glutamine in the position 1000 for histidine, with poor preservation of position and surroundings in the different biological species and "in silico" predictors, demonstrating a potentially deleterious nature. In the SCN9A gene, a variation in the position chr2:167.133.677, promoted the substitution of cysteine in the position 886 for tyrosine, with great preservation of position and surroundings in the different biological species and "in silico" predictors, demonstrating a potentially deleterious nature. **Discussion:** In previous studies, SCN9A mutations have been associated with Congenital Insensitivity to Pain, when in autosomal recessive fashion. However, his heterozygous c.3000G>T - ENST00000302328 SCN11A mutation can potentially be the cause for his neurological findings in autosomal dominant fashion. **Final comments:** In cases with 2 reported variants, it is important to consider the genotype-phenotype correlation in order to make the final diagnosis. In this case, the presence of a single variant rules out the possibility of SCN9A related disease and suggests that HSAN type VII is the most likely etiology of his condition. Unfortunately, we were not able to test his deceased father in order to confirm the familial pattern, although the patient clearly reported a similar clinical course in his father. This study was authorized by the patient.

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HSPB8-associated mutation resulting in myopathy with rigid spine with multiple muscle contractures and multiminicores in biopsy

Mayhara Rosany da Silva Santiago[1], Ana Caroline Paiva Simeão[1], Heloisy Maria Nunes Galvão[1], Raissa Josefa Pereira de Moura[1], Nereu Alves Lacerda[1], Heron Alves Vale[1], Natália Nunes Tenório[1], Maria Luisa Roberto Lima Lisboa[1], Ana Beatriz Roberto Lima Borba[1], Renata Amaral de Andrade Mendonça[1]

[1]Hospital Pelópidas Silveira, Recife PE, Brazil.

Correspondence: Mayhara Rosany da Silva Santiago; Email: santiago.mayhara@gmail.com.

Case presentation: A 15 years old boy presented with a history of proximal lower limb weakness over the last 2 years. He denied dysphagia, sphincter disorders or upper limb weakness. His prenatal, perinatal, and neuropsychomotor development were uneventful. There was no family history that resembled the patient's condition. Physical examination findings included severe proximal bilateral lower limb and axial weakness, with lumbar hyperlordosis, wingeds scapulas, and cervical hyperextension. There were also contractures that limited plantar dorsiflexion, torso and cervical anterior flexion, with a rigid spine predominant phenotype. Clinical examination was unremarkable. Due to the clinical phenotype an investigation of myopathies focusing on genetic causes was conducted. It showed moderate elevation of CK - 778U/L. Heart function evaluation was normal and Electroneuromyography was compatible with diffuse myopathy with proximal predominance. The forced vital capacity assessment revealed a moderate restrictive ventilatory disorder. Muscle biopsy revealed a myopathic pattern with minicores. Lower limb muscle MRI showed fat replacement of adductors, biceps femoris and soleus muscles. Genetic DNA ample panel found a possible pathogenic heterozygous mutation in the gene HSPB8. **Discussion:** Congenital myopathies are rare pathologies defined by genetic analysis, weakness pattern and histopathological findings. Other mutations in the HSPB8 gene were associated with CMT type 2, axial and distal adult onset myopathy, and, proximal limb-girdle rimmed vacuolar myopathy. This case reports a mutation of HSPB8 gene with a new phenotype correlated with a predominant axial myopathy beginning in second decade of life, resulting in a rigid spine and multiples muscles contractures atypical phenotype with minicores at muscle biopsy. **Final comments:** The full function of the HSPB8 gene is still unknown, but it appears to play a very important role to the normal neuromuscular physiology.

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Huntington's disease in a 24-year-old male patient: a case report

Anita Ferreira Das Neves[1], Rafaela Coelho Rachid[1], Lucas Leonardo Alves Cardoso[1], Eduarda Franceschet[1], Gabriela Maria Vicente de Melo[1], Leandro Pereira Janelli da Silva[2]

[1]Universidade Federal do Rio Grande, Rio Grande RS, Brazil.
[2]Universidade Federal do Rio Grande, Hospital Universitário, Rio Grande RS, Brazil.

Correspondence: Anita Ferreira das Neves; Email: atinaneves@gmail.com.

Case presentation: A 29-year-old male was first admitted at age 24 with episodes of syncope, movement disorders, and psychomotor agitation, which had progressively worsened over the past three years. Examination revealed generalized involuntary tremors (rest and intention), bilateral Babinski sign, choreiform movements, plastic rigidity, dysarthria, generalized hyperreflexia, and psychiatric symptoms. His mother had a similar condition since the age of 26, without a confirmed diagnosis. Given the patient's clinical presentation and family history, the direct genetic test for Huntington's disease (HD) was conducted to detect the repeated cytosine-adenine-guanine (CAG) trinucleotide expansion in the HTT gene. The test revealed alleles 18 and 6 with 61 CAG repeats, indicating the patient is heterozygous with full penetrance. Cranial magnetic resonance imaging revealed a slight volumetric reduction in the brain and hippocampus, disproportionate to his age. Cranial computed tomography revealed marked reduction in the volume of the striatal bodies, especially on the left, and of the brain sulci and fissures, unusual for his age, without predominant lobar involvement. After confirming HD, treatment with levodopa and neuroleptics was started for symptomatic relief, although the response was minimal. His most recent hospitalization occurred a year ago for the treatment of acute chorea and the insertion of a gastrostomy tube due to severe oropharyngeal dysphagia, typical of neurodegeneration. Currently, the patient is on levodopa, alprazolam, biperiden, valproate, and haloperidol. **Discussion:** HD is an autosomal dominant neurodegenerative disorder characterized by an abnormal expansion of CAG repeats, which results in a defective huntingtin protein. This defective protein causes progressive neuronal degeneration, leading to a triad of motor, cognitive, and psychiatric manifestations. The age of onset of HD symptoms is inversely related to the number of CAG repeats; more repeats result in earlier symptoms. Most individuals with HD have between 40 and 50 CAG repeats, but our patient has alleles 18 and 6 with 61 repeats, explaining the early onset at age 24. Despite initial treatment with levodopa and neuroleptics, the patient's choreiform movements worsened. Additionally, the need for a gastrostomy a year ago due to severe oropharyngeal dysphagia, a common complication of HD, illustrates the complexity and progression of this disease. Although the patient's mother did not undergo confirmatory genetic testing, her symptoms, similar to those of her son, raise suspicion of HD due to its hereditary nature. **Final comments:** This case is notable for its diagnosis of a rare disease, particularly in a young individual, as HD is more prevalent between the ages of 40 and 50. Therefore, it emphasizes the importance of genetic testing when the triad of symptoms is identified, particularly if there is a suggestive family history.

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Huntington's disease: a two-cases report of atypical presentations

Júlia Pio Fernandes Nery[1], Mariana Beber Chamon[1], Débora Palma Maia[1], Ricardo Oliveira Horta Maciel[1], Francisco Eduardo Costa Cardoso[1]

[1]Universidade Federal de Minas Gerais, Hospital das Clínicas, Belo Horizonte MG, Brazil.

Correspondence: Júlia Pio Fernandes Nery; Email: jupio0107@gmail.com.

Case presentation: The patients are two male brothers, ages 44 and 39 years old. They were referred to our clinic in 2023, being diagnosed as a fatherly-inherited Huntington's disease. The older brother reports a 12-year evolution of hyperkinetic movements, later characterized as chorea, a dystonic posture of the trunk, presence of rest and action myoclonus and cognitive decline. He still reports epilepsy, presenting with focal to bilateral tonic-clonic

seizures one year ago. The diagnosis was confirmed by genetic testing based on the presence of 18 and 45 CAG repetitions in the huntingtin gene (HTT). The younger brother presented 7 years ago with chorea, intentional tremor, generalized myoclonus, dysarthria and difficulty walking. Confirmatory genetic test has shown 18 and 44 CAG repetitions. Both patients have as the main symptom appendicular and axial myoclonus, spontaneous, stimulus sensitive, with postural and action stimulation, leading to great functional impairment. The older brother has laryngeal myoclonus. Their father had a similar clinical history, reported to have myoclonus and chorea, but he was never diagnosed, and couldn't be evaluated since he died at 45 years-old due to complications of the disease. Treatment was started with valproate, taking respectively 1,500 mg/day and 500mg/day, with partial improvement of the myoclonus, and control of the seizures in the epileptic patient. Association of benzodiazepines for both patients promoted greater relief of symptoms. **Discussion:** We presented two case reports of adult onset Huntington's disease with myoclonus as the main clinical feature. Myoclonus is rarely reported as a feature of adult onset Huntington's disease and is not a component of Unified Huntington's Disease Rating Scale (UHDRS), a commonly used scale for following up these patients. Proper diagnosis becomes challenging given this atypical presentation, and in other cases, could raise the possibility of an overlap of another disease. **Final comments:** Valproic acid and clonazepam were partially effective in controlling myoclonus. The knowledge about this atypical presentation of Huntington's disease helps in making the correct diagnosis, establishing an adequate symptomatic treatment and clarifying the prognosis to the patients and their families.

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Huntington's disease mortality in Brazil: an epidemiological overview from 2013 to 2022

Guilherme Carvalho de Souza, Lívia Leite Goes Gitai[1], Alícia Eduarda Rios Soares[1], Antonia Cardoso Silva[1], Cecilia Maria Monteiro Barbosa[1], Eliab Batista Barros[1], Gabriela Barbosa de Sá Rocha[1], João Victor Vasconcelos Tavares Maximiliano[1], Julio César Soares Barros[1], Larissa da Silva Almeida[1]

[1]Universidade Federal de Alagoas, Faculdade de Medicina, Maceió AL, Brazil.

Correspondence: Guilherme Carvalho de Souza; Email: guicadesouza@gmail.com.

Background: Huntington's disease (HD) is a progressive neurodegenerative condition of autosomal dominant hereditary character, with varying phenotypic manifestations grouped into motor, cognitive, and psychiatric categories. The main symptoms are chorea, dystonia, incoordination, cognitive decline, and behavioral changes, typically beginning between 30 and 50 years of age, though they can occur earlier or later. **Objective:** To evaluate HD mortality in Brazil over the ten years from 2013 to 2022, using descriptive variables such as region, age group, and sex. **Methods:** The analysis was conducted based on a cross-sectional, retrospective, and descriptive epidemiological study using data from the Mortality Information System (SIM/DATASUS) for the period 2013-2022. Evaluating HD mortality, an analysis of the variables "Region," "Age Group," and "Sex" was performed. Population data for each region were obtained from the Brazilian Institute of Geography and Statistics (IBGE) and used to calculate mortality per million inhabitants of each region. **Results:** Between 2013 and 2022, a total of 1,310 HD-related deaths were recorded in Brazil. Of these, 47.63% occurred in the Southeast region, 24% in the South, 16.8% in the Northeast, 8.47% in the Central-West, and 3% in the North. However, when calculating mortality per million inhabitants, the highest rates were observed in the South (10.5 deaths per million inhabitants), followed by the Southeast (6.93), Central-West (6.52), Northeast (3.86), and North (2). Regarding age groups, individuals under 19 years old presented the lowest number of deaths during the period. From the age of 20, there is a gradual increase in mortality, reaching 279 deaths between 40 and 49 years old (21.3%) and peaking in the age group of 50 to 59 years old, with 381 deaths (29% of HD mortality). After this peak, a decline in mortality rates by age is observed, with the 60 to 69 age group accounting for 21.35% of deaths, followed by a further decrease in rates with advancing age. It is noteworthy that approximately 63% of deaths occurred in age groups under 60 years old. Regarding sex, the differences were not notable, with females accounting for 55% and males for 45% of the deaths. **Conclusion:** This study provides a deeper understanding of the various epidemiological aspects related to HD mortality. Regarding regional comparisons, the Southern region of Brazil stands out for its higher rates of HD mortality in relation to its population. As for the age at death, the fact that a large portion of deaths occurs before the age of 60 draws attention to the impact of HD on early life and raises questions about possible strategies needed to ensure support for patients with rare neurodegenerative diseases.

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Hyperactivity symptoms are related to functional connectivity alterations in dopaminergic and noradrenergic networks in Alzheimer's disease

Fernanda Burle dos Santos Guimarães[1], Márcio Luiz Figueredo Balthazar[1], Brunno Machado de Campos[1], Rafael Brandes Lourenço[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Fernanda Burle dos Santos Guimarães; Email: fburle.psiq@gmail.com.

Background: Cholinergic, noradrenergic, and dopaminergic dysfunction play an important role in Alzheimer's disease (AD). The basal nucleus of Meynert (BNM), locus coeruleus (LC), and ventral tegmental area (VTA) are its prominent producer nuclei, respectively. Dysfunction of these nuclei seems to contribute significantly to the manifestation of neuropsychiatric symptoms (NPS). **Objective:** First, we aim to identify changes in the functional connectivity (FC) from NBM, LC, and VTA in patients with mild cognitive impairment (MCI) and dementia of AD, compared with healthy subjects (HS). Secondly, to verify potential correlations between the FC maps of these nuclei and NPS in MCI and mildAD patients. **Methods:** We analyzed data from 67 subjects over 50 years old. There were 18 HS and 49 subjects with altered amyloid-beta in the cerebrospinal fluid (25 MCI; 24 DA). Participants underwent rs-fMRI and completed neuropsychological evaluation, including the Neuropsychiatric Inventory (NPI). We compared FC differences among HS, MCI, and AD using ANCOVA test on individual FC maps for each seed region. We then correlated NPI syndromes (apathy, hyperactivity, affective syndrome, psychosis) scores with individual FC maps, adjusting for age and education (SPM12-toolbox). **Results:** In between-group analysis, regarding BNM, compared to HCs, patients with MCI showed significantly decreased FC in the left precentral gyrus and left anterior insula (volume 62.05 and 61.59 voxels, respectively). Similarly, AD patients, when compared to HCs, showed significantly decreased FC in the left and right medial precentral gyrus (volume 104.06 and 82.75 voxels, respectively) and in the right supramarginal gyrus (volume 64.68 voxels). Regarding the VTA network, unexpectedly, we found that, compared with MCIs, DA patients showed increased FC in the left central operculum (volume 92.29 voxels). No other between-group changes reached statistical significance for the LC. Considering the correlation of FC in AD patients and NPI, significant correlations were found exclusively with scores from hyperactivity subsyndrome (agitation, irritability, aberrant motor behavior, euphoria and disinhibition). Regarding right VTA, we noticed an increased FC mainly with the right and left subcallosal area (volumes 22.24 e 20.36 voxels, respectively) and left medial orbital gyrus (volume 26.46 voxels). We found a similar pattern in the left VTA and the right LC, showing increased FC predominantly with right subcallosal area (volumes 18.90 e 11.38 voxels, respectively) and left subcallosal area (volumes 13.84 e 12.19 voxels, respectively). When looking at BNM FC, no correlation reached statistical significance. **Conclusion:** Patients with altered amyloid-beta in CSF presented with decreased FC between precentral gyrus/insula and BNM. Also, the connectivity of noradrenergic and dopaminergic networks to the subcallosal area positively correlates to hyperactivity syndrome in AD.

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Hypereosinophilic syndrome associated with polyneuropathy: a case report

Fabio Victor Vieira Rocha[1], Luísa D'Ávila camargo[1], Lucas Araujo Limongi horta[1], Lívia Valle Dos santos silveira[1], Renann Nunes Pirola[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Fabio Victor Vieira Rocha; Email: fabio.vvrocha@gmail.com.

Case presentation: 36-year-old man complaining of violet-colored edema in bilateral ankles and left hand, associated with restricted movement, pain and paresthesia. After 30 days, he developed bilateral and ascending paresis and paresthesia in the lower limbs, lymphadenopathy, and laboratory tests confirmed eosinophilia and an undetermined septic condition. After 7 days of ceftriaxone, his pain and edema improved. Electroneuromyography showed acute and subacute axonal and demyelinating sensory-motor polyneuropathy. On physical examination, he had dysarthria, right palatal drop, gait imbalance, globally abolished reflexes, decreased muscle strength (grades III and II), and compromised sensitivity. He denied fever and sphincter alteration. After using Pregabalin 300 mg/day and Prednisone 60 mg/day for 6 months, paresthesia and paresis improved, but eosinophilia persisted. **Discussion:** Idiopathic hypereosinophilic syndrome (HES) is a myeloproliferative disorder characterized by unexplained prolonged eosinophilia (>6 months) and specific organ damage. Diagnostic criteria for HES include sustained blood eosinophilia with at least 1,500 eosinophils/μL for more than 6 months, multi-organ involvement, and no other apparent causes of eosinophilia (e.g., parasitic, allergic). In this case, persistent eosinophilia and neurological involvement suggest HES, as other causes were excluded. Neurological manifestations, such as sensory-motor polyneuropathy, paresis, paresthesia, and abolished reflexes, are consistent with HES. Eosinophilia can damage peripheral nerves, contributing to the symptoms. Skin involvement is common in over 50% of HES patients, presenting as pruritic erythematous lesions, papules, nodules, and angioedematous and urticarial lesions, often associated with a better prognosis. Although this patient did not have significant cutaneous symptoms, lymphadenopathy and edema suggest systemic involvement. Thus, hypereosinophilic syndrome may result in a number of neurologic signs and multisystemic affect. Therefore, in cases of neurologic signs with no clinically established cause, especially in younger people, the possible HES should be considered. **Final comments:** This case illustrates the complexity of diagnosing and managing patients with eosinophilia and neurological involvement. HES should be considered in cases of prolonged eosinophilia with multi-organ involvement after excluding other causes. Management includes corticosteroids and other immunomodulatory therapies, with continuous monitoring of clinical and hematological response.

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Hyper-religiousness as a clinical manifestation in Charles Bonnet's syndrome

Sephora Sabrina Candido de Almeida[1], Isabella Marranghello D'Amico[1], Yngrid Dieguez Ferreira[1], Rônney Pinto Lopes[1], Lunielle da Cruz Caldeira[1], Ricardo Salemi Riechelmann[1], Ana Carolina Gomes[1], Isabela Martins de Freitas[1], Alexandre Shoji[1], Diogo Haddad Santos[1]

[1]Irmandade Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Sephora Sabrina Candido de Almeida; Email: sephora_sabrina@yahoo.com.br.

Case presentation: We report the case of a black male 94-year-old patient who was a Catholic in childhood and converted to Spiritism in adulthood. He was a retired farmer and had studied to 1st grade. The patient was only able to see shadows for the past 5 years owing to bilateral cataracts. In the initial assessment, he showed partial orientation for time and space and actively complained of difficulties sleeping with vivid dreams. He also reported hallucinations which he recognized as unreal, but believed he experienced these because he was chosen to have them. These visions always appeared in a messianic context and associated with a religiously based discourse. He reported having medium abilities since the age of 15. His daughter reported good interaction with others and absence of social withdrawal. Family members complained of difficulties in the care process because, when challenged, he became aggressive. They also reported that the religious discourse continued throughout the day and night, hampering feeding and care during the night. The eye test revealed severely limited visual acuity bilaterally, and the patient was unable to count his fingers. He scored 15/30 points on the mini-mental state exame (MMSE). A non-contrast CT scan disclosed global atrophy without acute lesions. These hallucinations were typically worse at night and dark places, improving in brighter spaces. The ophthalmic evaluation indicated corrective bilateral cataract surgery, whereas heart surgery was contraindicated owing to high surgical risk. Atypical neuroleptics were prescribed at a low starting dose and progressive increased as required and according to results on reevaluation. Subsequent improvements in hallucinations (absence), sleep-wake cycle and functioning were reported. Family members cited that care provision was also easier. However, the family noted the patient had no further social contact, particularly at the Spiritist center, na activity which had hitherto had been a source of great pleasure. **Discussion:** Charles Bonnet syndrome (CBS) is a condition in which a visually-impaired individuals with intact cognition experience visual hallucinations in the absence of other sensory stimuli. The condition is generally associated with age-related macular degeneration (ARMD) and ocular diseases that affect central vision, most commonly cataracts, as occurred in the patient reported. These patients are often seen at centers for cognition because the manifestation of visual hallucinations in older persons is often confounded with the onset of early dementia syndromes. The pathogenesis of visual hallucinations remains unclear, although two main theories are accepted. The hypothesis of deafferentation suggests that loss of visual input changes synaptic function adjacent to the affected area, leading to increased excitability, manifesting as visual experiences in the absence of external stimuli.

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Hypertrophic pachymeningitis secondary to granulomatosis with polyangiitis: case report

Stephania Neves Scapim[1], José Víctor Filho de Oliveira[1], Luiz Gustavo Chaves Gomes[1], Angélica Vilela Rodrigues de Alvarenga[1], Luiz Paulo Bastos Vasconcelos[1], Gustavo Guimarães Moreira Balbi[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Juiz de Fora MG, Brazil.

Correspondence: Stephania Neves Scapim; Email: drastephanianeves@gmail.com.

Case presentation: A 53-year-old male, with a history of hypertension, obesity, and bilateral middle ear otitis complicated with chronic mastoiditis, developed a severe headache associated with fever and vomiting. He had a pattern of facial pain in bilateral nasal roots and persistent horizontal binocular diplopia, worsened on right gaze. He presented an episode of complete left-sided hemiparesis and dysarthria, spontaneously resolving within approximately 60 minutes. Initial hypothesis was mastoiditis complicated with meningoencephalitis, and he was treated with Ceftriaxone, Ampicillin, and Doxycycline. After hospital discharge, he experienced recurrence of the headache, associated with visual blurring and diplopia, leading to readmission. He underwent antibiotic therapy with Cefepime, Sulfamethoxazole, Trimethoprim, and Doxycycline, along with corticosteroid therapy with Dexamethasone. Mastoid computed tomography (CT) showed signs of bilateral chronic mastoiditis. Bilateral papilledema was observed on fundoscopy. Brain magnetic resonance angiography (MRA) revealed thickening and enhancement of the right cerebral convexity pachymeninges and interhemispheric falx, associated with subarachnoid hyperintensity on FLAIR sequence and areas of diffusion restriction, as well as signal changes in the right parietotemporal subcortical white matter, consistent with hypertrophic pachymeningitis (HP). Lumbar punctures showed increased opening pressure and ruled out fungal infections and tuberculosis (TB). Due to the hypothesis of an autoimmune disease, an evaluation by the Rheumatology team was performed, and the laboratory workup revealed a positive ANCA-c (1:40) by indirect immunofluorescence, with negative PR3-ANCA, MPO-ANCA, ANA, and RF. A hypothesis of granulomatosis with polyangiitis (GPA) was made. Due to dysphonia, videonasolaryngoscopy was performed to investigate subglottic stenosis, which revealed a vocal cords cyst, with no stenosis. Chest CT revealed a cavitated nodule in the apicoposterior segment of the left upper lobe. Bronchoscopy-guided bronchoalveolar lavage was negative for acid-fast bacilli microscopy and molecular TB testing. Given the diagnosis of severe GPA with meningeal involvement, IV pulse therapy with methylprednisolone and cyclophosphamide was prescribed, according to the CYCLOPS protocol. **Discussion:** HP is a rare condition characterized by inflammation of the dura mater, and GPA is among its etiologies. **Final comments:** The recognition of vasculitis as a cause of meningeal involvement resulting in HP should be part of the diagnostic consideration for clinical syndromes involving secondary headaches of non-infectious etiology, especially those associated with cranial nerve involvement and intracranial hypertension.

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Hypocalcemia and functional outcomes in stroke patients post-thrombolysis

Antonio Lázaro Santos Soares Junior[1], Thaís Barbosa de Oliveira[1], Felipe Asaf Borges Millard[1], Julia Xavier Oliveira[1], Gabriel Souza Medrado Nunes[1], Pedro Antonio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Antonio Lázaro Santos Soares Junior; Email: antoniolssj@ufba.br.

Background: Thrombolysis is a widely recognized treatment for acute ischemic stroke. Despite its effectiveness, some factors have been associated with poorer outcomes following thrombolysis, such as advanced age and higher National Institutes of Health Stroke Scale (NIHSS) scores, however other factors have not been studied yet. Hypocalcemia is a common metabolic disturbance observed in various clinical settings. Its impact on patients undergoing thrombolysis for acute ischemic stroke remains underexplored. Understanding this relationship may enhance patient management strategies and improve prognostic assessments in acute stroke care. **Objective:** To address the association between hypocalcemia and functional outcomes in acute stroke patients post-thrombolysis. **Methods:** This study enrolled patients with acute stroke admitted to a referral hospital's stroke unit in Bahia, between July/2023 and March/2024. Clinical and epidemiological characteristics were extracted from patients' medical records or obtained through personal interviews. Hypocalcemia was defined as total serum calcium inferior to 8.5g/dL. The assessment of functional outcome and survival was performed using the modified Rankin Scale (mRS) validated for telephone interviews. Worst functional outcome was defined as an mRS greater than 2. For statistical purposes, numerical variables were expressed as mean or median, along with their standard deviation or interquartile range (IQR), while categorical variables are expressed as frequencies and percentages, when appropriate. Chi-square test was employed to compare the functional outcome and presence of hypocalcemia in admission. Logistic regression was used to adjust for age and NIHSS at admission. **Results:** Sixty-three participants were included, of whom 39 (61.9%) were male. The predominant color was brown, with 34 (54%) participants, followed by black (30.2%) and white (14.3%). The average age was 64.4 (±12.1) years. Regarding previous comorbidities, 21 (33.3%) individuals had diabetes mellitus, 48 (76.2%) had systemic arterial hypertension, and only 1 (1.6%) had chronic kidney disease. The mean NIHSS on admission was 10.6 (±5.3). At the time of admission, hypocalcemia was present in 13 (20.6%) participants. After 90 days post-stroke, 28 (44.4%) participants had worse functional outcomes. Hypocalcemia correlated with worse functional outcomes in the final model (B=1.35; CI: 1.01-14.71), with a p-value of 0.048. **Conclusion:** Hypocalcemia is correlated with worst functional outcomes 90 days after stroke in patients who underwent thrombolysis.

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Hypoglossal nerve injury resulting from traumatic brain injury

Caroline Dresch Sabadin[1], Lara Auana da Rosa[1], Luisa Bianca Reis Parode[1], Lucas dos Santos Oliveira[1], Gabriel Santos de Almeida[1], Paulo Eduardo Mestrinelli Carrilho[1]

[1]Universidade Estadual do Oeste do Paraná, Cascavel PR, Brazil

Correspondence: Caroline Dresch Sabadin; Email: caroldsabadin@gmail.com.

Case presentation: A 22-year-old previously healthy woman was admitted to a University Hospital on July 2, 2022, due to a mild traumatic brain injury following a car accident. Upon admission, her ectoscopy and vital signs were unremarkable. Neurological physical examination showed isophoto-reactive pupils, preserved extrinsic ocular movement, a Glasgow Coma Scale (GCS) score of 14 (with a point loss in the best verbal response), and muscle strength of grade 5 in all limbs. On the same day, a contrast-enhanced cranial computed tomography (CT) scan revealed a slight obliteration of sulci and gyri, as well as occupied cisterns. The patient was admitted to the ICU for 6 days, and a new CT scan revealed a slight brainstem hemorrhage. She was discharged on July 11, 2022, and continued with outpatient follow-up by neurology. During outpatient follow-up, a cranial magnetic resonance imaging (MRI) showed sequelae changes in the subcortical white matter of the cerebral hemispheres, more evident in the frontal lobes, as well as hemosiderin degradation products in the left infratentorial cisterns. She presented with muscle strength of grade 5 in the upper limbs and left lower limb, and grade 4- in the right lower limb, with reduced thermal and pain sensitivity in the right hemibody. Cranial nerve evaluation revealed a deviation of the

tongue to the left. In a subsequent follow-up visit, there was partial improvement in hemiparetic symptoms on the left side of the body, but the tongue deviation to the left persisted, along with fasciculations in the tongue and hypoesthesia in the right hemiface. Regarding superficial reflexes, the plantar reflex was indifferent on the right and extended on the left. The patient continues to be followed up in the neurology outpatient clinic. **Discussion:** We describe a rare case of hypoglossal nerve palsy in a young patient following traumatic brain injury. The novelty of this case lies in its singularity: while most case reports document hypoglossal nerve injuries following high-impact trauma, typically associated with skull base fractures, this case highlights this complication occurring even in a trauma considered minor, without evidence of bone fracture. This underscores the importance of clinical vigilance in patients with traumatic brain injury. The rapid resolution of the patient's symptoms following steroid therapy and physical therapy suggests a good prognosis for isolated hypoglossal nerve injuries. **Final comments:** According to the literature, post-traumatic hypoglossal nerve palsy is rare. When it does occur, it is associated with high-impact trauma. Therefore, when this condition occurs in low-energy mechanisms, as in the described case, it is noteworthy. The reported case also emphasizes the importance of a complete neurological examination following trauma, as cranial nerve injuries can occur in isolation.

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Hypoglossal paralysis secondary to paraganglioma in the right carotid bulb: a case report

Danielle Cleide da Silva Lima Queiroz[1], Pedro Jatobá Arteiro[2] , João Pedro Lopes da Silva Moraes[1], Gabriela Nogueira Couto[1], Bruno Ramos Levy[1], Amanda Sampaio de Sá[2], Álvaro José Porto Moreira[1]

[1]Real Hospital Português, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Centro de Ciências Médicas, Recife PE, Brazil.

Correspondence: Pedro Jatobá Arteiro; Email: pedro.arteiro@ufpe.br.

Case presentation: We report a case of a 67-year-old woman, who presented with a sudden onset of dysarthria and occipital pressure, seeking neurological emergency services, being evaluated and seen, in addition to dysarthria, tongue deviation to the right with preserved trophy. The remainder of the neurological examination showed no changes, palate, uvula, gag reflex preserved, no changes on strength or sensitivity. A magnetic resonance imaging (MRI) was therefore performed, which revealed a solid formation measuring 3.8 x 3.5 x 2.2 cm, located superior to the right carotid bifurcation and posteriorly to the right internal carotid artery, with low signal on T1 sequence and heterogeneous high signal on T2 and FLAIR, and intense enhancement after contrast injection. Subsequently, the tumor was removed with anatomopathological and immunohistochemical analysis of the sample, which was positive for chromogramin, synaptophysin, CD34 and S100, negative for cytokeratin, Ki67 <1%. The radiological and histopathological findings are compatible with the diagnosis of paraganglioma. **Discussion:** Carotid body tumors (CBTs), also called paragangliomas, are rare neoplasms of the neuroectoderm, responsible for 0.6% of head and neck tumors and 0.03% of all tumors, with a risk of malignancy from 2% to 12.5%. The carotid body is the largest collection of paraganglia in the head and neck and is found on the medial aspect of the carotid bifurcation bilaterally, functioning as a chemoreceptor that helps in the autonomic control of blood pressure, heart rate and respiratory rate. CBTs are more frequent in elderly patients, between 50 and 70 years old, with a slight prevalence in females over males. CBTs are hypervascularized and slow-growing tumors, so that the majority presents as an asymptomatic neck mass, however in some cases there is compression or invasion of adjacent structures. Neurological deficits, such as the case reported, may occur in less than 10% of patients with CBTs, due to tumor compression of the sympathetic chain or adjacent cranial nerves, especially the glossopharyngeal, vagus and hypoglossal nerves. Radiological evaluation is necessary to establish the diagnosis of CBTs, with Doppler Ultrasonography, MRI, computed tomography (CT) and Angiography being the most commonly used. As in the case described, MRI shows high signal lesions on T2 sequence and low signal on T1, with enhancement after contrast injection. Biopsy is contraindicated in CBTs due to the high risk of complications due to the proximity of vascular structures. Treatment consists of surgical excision of the tumor, but radiotherapy can be performed in patients at high surgical risk or with extensive tumors. **Final comments:** CBTs are rare neuroendocrine tumors, often asymptomatic, but, as in the case described, they can cause neurological symptoms, due to compression of cranial nerves. Its diagnosis depends on imaging tests and its treatment is based on surgical resection.

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Hypoglossal paresis in a relapse of multifocal acquired demyelinating sensory and motor neuropathy: a case report

Eduardo Mesquita de Souza[1], Matheus Alves da Silva[1], Rafael Bragança Rodrigues Matias[1], Ana Luiza Vieira de Faria[1], Amanda Miranda de Mendonça[1], Marcelo Freitas Schmidt[1], Fernanda Boldrini Assunção[1], Christiane Monteiro de Siqueira Campos[1], Alex Machado Baeta[1]

[1]Beneficencia Portuguesa de São Paulo, São Paulo SP, Brazil.

Correspondence: Eduardo Mesquita de Souza; Email: eduardoszmesquita@gmail.com.

Case presentation: A 53-year-old woman, with a nine-year history of Multifocal Acquired Demyelinating Sensory and Motor neuropathy (MADSAM), recently presented with paresis of the upper limbs and difficulty in tongue mobility. She had been in remission for two years following monthly immunoglobulin treatment. Neurological examination revealed loss of motor strength in the upper limbs, diffuse hyporeflexia, and left hypoglossal paresis. To investigate secondary causes, magnetic resonance imaging (MRI) and electromyography (EMG) of the tongue were performed, revealing reactivation of the underlying disease. **Discussion:** It's known that patients with MADSAM present with a markedly asymmetric, multifocal condition, sometimes indistinguishable from other forms of multiple mononeuropathy, resulting in diverse sensory and motor symptoms. Although evident manifestations of cranial nerve dysfunctions are rare in this condition, it is estimated that up to 5% of patients may present with cranial neuropathies, particularly in atypical forms. Most reports describe the involvement of the facial, trigeminal, and oculomotor nerves; however, hypoglossal nerve involvement, as in the presented case, is uncommon. Some patients in the spectrum of antibodies against nodal or paranodal proteins may present with combined central and peripheral demyelination, as well as cranial neuropathies. **Final comments:** The presented case highlights a rare and atypical manifestation of MADSAM with hypoglossal nerve involvement, evidencing the complexity and clinical variability of inflammatory neuropathies. Despite a two-year remission with immunoglobulin treatment, disease reactivation underscores the need for continuous vigilance and rigorous follow-up of these patients. This report reinforces the importance of considering the various clinical presentations of demyelinating neuropathies and the possibility of overlap with atypical clinical manifestations. The inclusion of hypoglossal nerve involvement expands the understanding of the extent and impact of cranial neuropathies in multifocal inflammatory conditions, emphasizing the need for comprehensive and personalized diagnostic approaches.

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Hypokalemic periodic paralysis secondary to primary hyperaldosteronism: a case report

Eduardo Abrão Spínola Rezk[1], Natalia de Castro Fim Nakao[1], Yasmim Nadime Jose Frigo[1], Rebeca Aranha Barbosa Sousa[1], Gabriela Rossini Dantas[1], Itamar Meireles[1], José Eduardo Trentim Longo[1], Dominick Esthephanny Monteiro Silva[1], Hélio Aquaroni Farão Gomes[1], Ricardo Silva Carvalho[1]

[1]Universidade Estadual Paulista, Faculdade de Medicina de Botucatu, Botucatu SP, Brazil.

Correspondence: Eduardo Abrão Spínola Rezk; Email: eduardo.s.rezk@gmail.com.

Case presentation: Woman, 34 years old, hypertensive since the age of 30 with depressive disorder, presenting recurrent episodes of diffuse muscle weakness for four years associated with cramps. Initially, the episodes lasted an average of five days with spontaneous resolution, occurring semi-annually. However, over the last 6 months, they have been more frequent, lasting about 10 days each time. Initial evaluation revealed flaccid areflexic tetraparesis and severe hypokalemia (K= 1.8); correction of electrolyte imbalance was performed, and other causes were investigated. Routine electromyography of all 4 limbs showed no abnormalities; channelopathy protocol indicated increased amplitude from the start in short and long exercise tests, but without a pattern consistent with primary channelopathies. During investigation, an alteration in the aldosterone/renin activity ratio of 55 was found, and a nodule in the right adrenal gland on abdominal CT was consistent with aldosterone-producing macroadenoma. Thus, the hypothesis of secondary hypokalemic periodic paralysis due to primary hyperaldosteronism or Conn's syndrome was confirmed. Currently, the patient has suboptimal blood pressure control despite the use of four classes of antihypertensive medications, oral potassium supplementation, and is scheduled for adenoma surgery. There have been no recurrent neurological symptoms. **Discussion:** Episodic muscle weakness can be a clinical presentation of periodic paralysis, and there are known genetic causes for such conditions, such as primary hypokalemic periodic paralysis related to autosomal dominant genetic defects in voltage-gated calcium (type 1) and sodium (type 2) channels of the muscle cell membrane. There are also secondary forms of periodic paralysis described in the context of other conditions like thyrotoxicosis and adrenal diseases. Electromyography channelopathy protocols in these secondary cases may not exhibit a characteristic pattern. **Final comments:** It is important to consider secondary causes in the investigation of periodic paralysis, allowing for specific treatment and prevention of recurrence of neurological symptoms, as well as avoiding further complications from undiagnosed systemic diseases.

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Hypothalamic atrophy and textural changes in polyglutamine ataxias

Marcondes Cavalcante Franca Junior[1], Livia Rodrigues[1], Thiago Junqueira R. Rezende[1], Alberto Rolim Muro Martinez[1], Breno Massurayama[2], José Luiz Pedroso[2], Orlando Graziani Póvoas Barsottini[2], Juan Eugenio Iglesias[3] , Simone Appenzeller[1], Letícia Rittner[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[3]Harvard Medical School, Massachusetts, USA.

Correspondence: Marcondes Cavalcante França Jr.; Email: mcfrancajr@uol.com.br.

Background: Spinocerebellar ataxia (SCA) presents a complex genetic landscape, with over 40 subtypes. These autosomal dominant disorders manifest late onset and severe disability, primarily impacting the cerebellum but also involving other nervous system structures. **Objective:** While a single study has linked hypothalamus atrophy to SCA3, further research is needed to confirm and/or expand this association. **Methods:** The study involved 135 adult patients with confirmed SCA, including subtypes SCA1, SCA2, SCA3, and SCA6 and 117 controls. Image processing utilized H-SynEx for hypothalamus segmentation, delineating 10 distinct subregions. SynthSeg determined total intracranial volume. Analysis focused on hypothalamus subregion volumes and texture features derived from MR images. Non-parametric statistical analysis, included Mann-Whitney U tests with Bonferroni correction ($p<0.05$) and effect size calculation (AUROC). **Results:** The study findings reveal atrophy in various hypothalamic subregions in SCA1 and SCA3 when comparing with controls. Notably, the anterior inferior subregion was most affected, with AUROC values of 0.83 and 0.7 for SCA1 and SCA3, respectively. Texture analysis unveiled significant findings in SCA1 across all attributes and subregions, with AUROC values exceeding 0.8. Correlations were observed between volume and texture attributes and the SARA coefficient for SCA1, SCA3, and SCA6. Furthermore, significant associations were found between volume and texture attributes and the average time from ataxia onset for SCA1 and SCA3. **Conclusion:** This study reveals substantial atrophy in different hypothalamic subregions in SCA1 and SCA3, alongside texture differences in SCA1, SCA2, SCA3 and SCA6, even in areas without visible atrophy. These findings imply complex effects of ataxia on hypothalamic subregions, possibly preceding detectable clinical changes.

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Hypothalamic dysfunctions in patients victims of traumatic brain injury: a systematic review

Laisy Bárbara Souza Silva[1], Danielli Regina da Silva[1], Airton Sena Oliveira Sales[1], Larissa Batista Xavier[1], Bárbara de Mello Galvani[1], Guilherme Victor Rocha Pereira[1], Lorenzo Pereira de Almeida Melo Montes[1], Emerson Oliveira Milhorin[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Laisy Bárbara Souza Silva; Email: d202010950@uftm.edu.br.

Background: Traumatic brain injuries (TBIs) can cause neuroendocrine dysfunctions, which are conditions that have not been extensively studied. Even milder traumas can result in pituitary hormone deficiencies, which may emerge with a certain delay after hypothalamic injury, with 70% of patients showing alterations within the first year of injury. Studies indicate that more than 30% of those affected by TBIs have dysfunction of at least one neuroendocrine hormone, with the combination of deficits being rarer. Hypothalamic injury leads to a diversity of clinical manifestations, given its crucial role in regulating various autonomic and endocrine functions in the body. Furthermore, late diagnoses lead to increased morbidity and mortality in these patients. **Objective:** To conduct a systematic review of studies concerning hypothalamic dysfunctions related to types of traumatic brain injuries. **Methods:** The data for this review were obtained from case studies through the PubMed and SciELO databases. The most prevalent type of injury based on the trauma mechanism and its consequences on the endocrine metabolism of the affected individuals were evaluated. Inclusion criteria were: studies published in the last 10 years, in English, and cases that necessarily presented traumatic injuries with endocrine repercussions after the event. **Results:** Titles and abstracts of 238 articles were analyzed from the search "hypothalamic dysfunction trauma case report," and 15 cases were selected for full analysis and reading. Excluded studies addressed hypothalamic dysfunctions resulting from pre-existing tumors, arteriovenous malformations, snake bites, surgical complications, spontaneous hemorrhages, and other non-traumatic causes. All the articles addressed were in English and published between 2015 and 2024. The sample population consisted of 9 adults (60%), 3 elderly (20%), and 3 children (20%). Regarding the trauma mechanism, 40% of the sample were injuries related to falls, 26.6% to sports injuries, and 20% to traffic accidents. Additionally, 2 studies mentioned injuries from firearm projectiles. Of the cases studied, 10 had a single hormonal deficiency. Furthermore, this evaluation found that 40% of cases had antidiuretic hormone depletion, 33% gonadotropic hormone deficiency, 33% adrenocorticotropic hormone deficiency, and 20% thyrotropic hormone deficiency. **Conclusion:** The scarcity of studies on the topic addressing the Brazilian situation of TBIs and their neuroendocrine repercussions demonstrates a significant gap in the national scientific literature, considering the high prevalence of TBIs in the country, both from traffic accidents and urban violence. The review thus highlights the need for the development of studies that investigate the understanding of TBI casuistry and clinical outcomes when the topic in question is neuroendocrine dysfunction.

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Idiopathic hypertrophic cranial pachymeningitis: a journey from clinical presentation to histological diagnosis

Victor Garcia Ortiz[1], Glauco Cody Nagata[1], Caroline Schiochet Verza[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Victor Alfonso Garcia Ortiz; Email: victorgarcia.or@gmail.com.

Case presentation: A 49-year-old woman was referred to our tertiary hospital for progressive bilateral vision loss, ataxia and cognitive dysfunction. Past medical history revealed a hospitalization in 2004, with diplopia and dizziness, initially diagnosed as CNS toxoplasmosis. Viral serologies – including HIV – were all negative. In 2011, she began with prominent headaches, diplopia and visual loss. She was initially diagnosed with CNS tuberculosis, with slight improvement of symptoms with corticosteroids and tuberculostatics. However, symptoms reemerged and rendered her with bilateral vision loss. Soon after he had an episode of subacute upper right limb weakness, diagnosed as a ischemic stroke. At examination, she presented severe vision loss bilaterally, papilledema, ataxia and infrequent focal seizures. She was admitted for further investigation. Laboratory exams were unremarkable, except for an elevated protein in CSF. Serologies and PCR for multiple infections were all negative (including toxoplasmosis, mycobacterium tuberculosis, borrelia burgdorferi and others). MRI revealed enhancement and thickening of meninges, more preeminent at cerebral falx and tentorium. Also, cerebral edema and enlargement of cerebellum could be noticed. A biopsy of dura-mater showed fibrosis associated to hyalinization and lymphocyte inflammatory infiltrate. She was maintained on corticosteroids (dexamethasone) and initiated on azathioprine, with important improvement of symptoms. **Discussion:** Idiopathic hypertrophic cranial pachymeningitis (IHPM) is an extremely rare inflammatory illness, characterized by recurring cycles of inflammation and fibrosis in the meninges, leading to diffuse hardening and thickening of the dura-mater. First reported in 1869 by Charcot and Joffroy, initially attributed to syphilitic meningitis. Clinical manifestations are wide and include progressive headaches, multiple cranial nerve palsies, cerebellar dysfunction, seizures and myelopathy. Several rounds of inflammation and cicatrization lead to progressive fibrosis and thickening of pachymeninges (dura-mater). This progressive growth leads to compression of increased intracranial pressure, entrapment of cranial nerves and ultimately brain and cerebellar lesions. Soon in the course of this illness, MRI may point to diagnosis, as it shows impressive enlargement and enhancement of meninges, with a 'naïve' or aseptic CSF. Biopsy is the only way to ultimately confirm the diagnosis and must be done early, before irreversible damage has occurred, as in our patient. Recently, it has been shown that IHPM may be the CNS manifestation of IG4-related disease. Further studies may confirm this relationship. Nonetheless, treatment with immunosuppressants should not be delayed and must begin as soon as common infectious etiologies are excluded. IHPM is rare, under-diagnosed and potentially fatal illness. A high suspicion index must be alongside the neurologist, as treatment may avoid great disability

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Idiopathic hypertrophic pachymeningitis: still a diagnostic challenge

Cândida Driemeyer[1], Ana Letícia Amorim de Albuquerque[2] , Vanessa Dantas de Andrade[1], Julia Santos Soares[1], Carlos Roberto de Mello Rieder[1], Arlete Hilbig[1]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[2]Santa Casa de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Cândida Driemeyer; Email: candidaplena@gmail.com.

Case presentation: A 47 year-old female presented to the emergency department with bilateral pulsatile headache and peripheral pattern palsy which began around 45 days ago but with acute worsening in the past 24 hours, when she also started with gaze paralysis. The patient initially visited an ophthalmologist, who ruled out acute ophthalmological complications and referred her to the emergency department. When the first symptoms started, the patient sought emergency care for the first time and was clinically diagnosed with Bell's Palsy secondary to herpes zoster infection and she was prescribed acyclovir 400mg 5 times a day and prednisone 60mg, both for 7 days with no improvement.The patient did not exhibit any skin lesions or vesicles. The patient had a history of systemic arterial hypertension and was taking continuous medication. On neurological examination at the arrival, the patient presented left hemiface infranuclear palsy, left eye ptosis, paralysis of all extrinsic muscles of the left eye and inability to perform convergence movement. The pupils and the right eye showed no abnormalities on physical examination. Magnetic resonance angiography showed pachymeningeal thickening on the left, especially frontoparietal and next to the ipsilateral cavernous sinus, with indeterminate findings. The patient underwent extensive clinical investigation: neoplasia screening and blood serologies were negative (HIV, VDRL, HbsAg, anti-HCV). Autoimmune investigation showed slightly elevated c4 complement (40, reference range 12-36) and no other alteration. Cerebrospinal fluid examination had high opening pressure (354 mmH2O), 1 cells/mm3, proteins 32mg/dL, glucose 71mg/d (blood glucose 106mg/dL). Cultures for bacteria and fungi, VDRL, and alcohol-acid-resistant bacilli were negative in CSF. PCR for herpes (HSV 1 and 2) and for tuberculosis was negative. Additional tests for cryptococcosis, proteinogram, and immunophenotyping were normal. Neoplastic cells were absent. She was then submitted to meningeal biopsy, which showed marked mononuclear infiltrate in areas, sometimes with perivascular arrangement, compatible with the diagnosis of Idiopathic Hypertrophic Pachymeningitis. **Discussion:** Headache improved in the first days of hospitalization with non-steroidal anti-inflammatory drugs. Given the lack of systemic disease findings, it was necessary to perform meningeal biopsy, it was followed by methylprednisolone 1g/day for 5 days, followed by corticoid weaning with progressive improvement of eye movement and facial palsy. Mainly differential diagnosis include nervous system infections, rheumatologic systemic diseases, neurosarcoidosis

and neoplastic lesions. Also, it is not possible to forgo biopsy due to the need to exclude IgG4-related disease. **Final comments:** This case demonstrates the need for attention to cases of headache with cranial nerve palsy. Despite current technology, a biopsy remains the only reliable method for excluding IgG4-related disease.

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Idiopathic intracranial hypertension preceding multiple sclerosis: a case report

Rodrigo Ibañez Tiago[1], Karina Galli Cardoso Mello Roberto[2], Jéssica Feltraco[2], João Ormond de Miranda[2], Isabela de Carvalho Florêncio[3], Thiago Dias Fernandes[3], Glauco Almeida[3]

[1]Universidade Federal de Mato Grosso do Sul, Faculdade de Medicina, Campo Grande MS, Brazil.
[2]Hospital São Julião, Campo Grande MS, Brazil.
[3]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.

Correspondence: Rodrigo Ibañez Tiago; Email: rodrigoibaneztiago@gmail.com.

Case presentation: A 25-year-old female patient was referred due to reduced visual acuity in the left eye associated with ocular discomfort for two days. She reported a weight gain of 30kg over the past two years (33.2kg/m²). Ophthalmological examination revealed: corrected visual acuity in the right eye (RE) of 20/30 and left eye (LE) of 20/80, isochoric pupils, and a relative afferent pupillary defect in the left eye; biomicroscopy showed no abnormalities in both eyes (BE) and intraocular pressure of 12 mmHg (BE). Fundoscopy revealed papilledema in BE. Hematological tests and cranial magnetic resonance imaging (MRI) was within normal limits. Visual field (VF) testing showed reduced superior temporal sensitivity in RE and diffuse sensitivity reduction in LE. Optical coherence tomography (OCT) showed increased thickness of the nerve fiber layers and anterior displacement of the RPE-Bruch's membrane complex, suggestive of intracranial hypertension (IH). Cerebrospinal fluid analysis was unremarkable except for an opening pressure of 23 cmH2O (while on 1.5g of acetazolamide). After 45 days, the patient experienced worsening visual acuity in both eyes (RE 20/60 and LE) and VF. A new cranial MRI showed areas of hyperintensity in the frontoparietal region and spinal cord. In addition to the diagnosis of idiopathic IH, multiple sclerosis (MS) was suspected, and the patient underwent pulse therapy and immunosuppression. Improvement was noted in visual acuity (RE 20/25 and LE 20/60), as well as in VF, OCT, and MRI parameters during follow-up. **Discussion:** Idiopathic intracranial hypertension (IIH) typically presents in obese women of childbearing age and is associated with optic nerve changes, as observed in this case. The coexistence of IIH and MS, although rare, has been documented in the literature. Studies suggest that the chronic inflammation and autoimmunity underlying MS may predispose patients to increased intracranial pressure (ICP), resulting in overlapping conditions. The patient exhibited typical visual symptoms of IIH, such as papilledema and visual acuity loss, but the progression of symptoms and imaging findings raised suspicion of an additional underlying condition. MRI revealed demyelinating lesions characteristic of MS, supporting the diagnosis. **Final comments:** It is essential to be alert to signs of increased ICP in patients with MS due to the overlap of symptoms, which can complicate the diagnosis of MS. The coexistence of IIH and MS requires a meticulous diagnostic approach and careful therapeutic management. Effective management should include stringent control of ICP and appropriate immunomodulation for MS, aiming to preserve visual function and improve quality of life. This rare case highlights the importance of comprehensive differential diagnosis in patients with neuro-ophthalmological symptoms and emphasizes the need for continuous monitoring for associated complications.

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Idiopathic intracranial hypertension presenting with facial palsy: a case report

Kevellyn Cruz Aguilera[1], Paula Camila Alves de Assis Pereira Matos[1], Bruno Nunes Ferraz de Abreu[1], André Borges Ferreira Gomes[1], Amanda Vale Catunda[1], Dina Andressa Martins Monteiro[1], Elisa Gabriela Barros Cunha[1], Marcio Victor de Souza Silva[1], Glauber de Menezes Ferreira[1], Fernanda Martins Maia Carvalho[1]

[1]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Kevellyn Cruz Aguilera; Email: kevellynaguilera@hotmail.com.

Case presentation: We report a case of a female patient, 24 years old, who developed a pressure-type headache in the occipital region in November 2023 that worsened when lying down, associated with binocular diplopia and difficulty blinking to the right. On physical examination, abducent and facial nerve palsy was seen, in addition to bilateral papilledema. Brain magnetic resonance and brain magnetic ressonance angiography was perfomed, demonstrating an empty sella turcica and a lumbar puncture with an opening pressure of 44cmH2O, closing the diagnosis of idiopathic intracranial hypertension. **Discussion:** Idiopathic intracranial hypertension (IIH) is an entity still misunderstanding, in which there is an increase in intracranial pressure in the absence of a secondary condition, such as brain tumors or strokes, for example. It was characterized in criteria for the first time by Walter E. Dandy who listed 22 cases having similar symptoms and signs with increased intracranial pressure seen on lumbar puncture, with no determining factor for this, categorizing the main findings of this condition. These criteria were revised in 2013 by Friedman et al., listing six criteria necessary for the diagnosis, including: the presence of papilledema, without neuroimaging findings that could justify the increase in intracranial pressure, with the remainder of the neurological examination showing no changes, except for involvement of other cranial nerves and lumbar puncture with increased opening pressure (considering 25cmH2O in adults and 28cmH2O in children) with CSF analysis without changes, in addition to incorporating imaging findings in the absence of papilledema or involvement of the sixth pair of cranial nerve. The findings most related to IIH are: headache with characteristics similar to those observed in tension and migraine, bilateral papilledema, paresis of the sixth pair of cranial nerve - a finding that can lead to diplopia - and, sometimes, the presence of pulsatile tinnitus. There are also cases in which unusual symptoms occur, including paresis of other cranial nerves or unilateral papilledema, such as the one presented in the case above, with the presence of paresis of the seventh cranial nerve. When evaluating the concomitance of the facial nerve palsy and IIH, there are some points to be considered, considering that the anatomy of the facial nerve makes the compression process difficult, having a point of probable vulnerability in the intratemporal segment of the facial canal, which may represent a justification for the framework. Facial palsy tends to be unilateral and peripheral, generally associated with abducens nerve palsy. **Final comments:** There are few descriptions in this described clinical context, therefore, with the aim of promoting more about other presentations, the data presented in this case report is relevant.

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Illiteracy and low education levels affect Parkinson's disease screening in an epidemiological study in the administrative region of candangolândia.

Deborah Almeida de Oliveira[1], Letícia Dias dos Santos Silva[1], Flávia Maria Campos de Abreu[2], Bárbara Querem Hapuque da Silva Linhares[2], Clarisse Santos Ferreira[1], Sabrina Simplício de Araújo Romero Ferrari[1], Simoneide Souza Titze de Almeida[1], Pedro Renato de Paula Brandão[3,4]

[1]Universidade de Brasília, Brasília DF, Brazil.
[2]Universidade Católica de Brasília, Brasília DF, Brazil.
[3]Hospital Sírio Libanês, São Paulo SP, Brazil.
[4]Em nome do Probe-PD.

Correspondence: Deborah Almeida de Oliveira; Email: deborah.almeidadeoliveira@gmail.com.

Background: Parkinson's disease (PD) is a chronic, progressive neurodegenerative disorder characterized by motor and non-motor signs and symptoms. Due to the absence of a single definitive test for PD and the overlap of its signs with other health conditions, clinical evaluation is necessary. **Objective:** Identify potential challenges in the screening process due to low education or illiteracy in the studied population of Candangolândia - Federal District, Brazil, and demonstrate that the screening method aids in guiding the diagnosis. **Methods:** The present work is part of the PROBE-PD project (Prodromal and Overt Parkinson's Disease Epidemiological Study in Brazil - approved by the Research Ethics Committee of the Beneficent Society of Ladies Hospital Sírio-Libanês - CEPesq/HSL-5.482.147). The study population consists of individuals aged 60 years or older living in the administrative region of Candangolândia-DF. In the first stage of the study, a questionnaire is administered that includes nine yes-or-no questions and one single-response question from the REM Sleep Behavior Disorder Questionnaire (RBDQ1). Participants who answer 'yes' to two or more items from the Tanner Questionnaire or score for the RBDQ1 question proceed to the second stage of the study, which consists of a clinical evaluation by a neurologist specializing in movement disorders. **Results:** So far, 1253 participants have been screened, with 433 (34.6%) testing positive on the Tanner Questionnaire. In the second stage, a sociodemographic questionnaire was applied. The education levels are as follows: Specialization: 1.32%, Graduates: 3.31%, Incomplete higher education: 1.32%, Complete secondary education: 18.54%, Incomplete secondary education: 12.58%, Complete primary education: 15.23%, Incomplete primary education: 28.47%, Literate: 9.93%, Illiterate: 9.3%. It was observed that due to the varying comprehension skills related to low education levels, participants had difficulty understanding and responding to the stage I screening questionnaire. The questions that had the highest number of positive responses like changes in writing (13.7%), in voice (14.9%), freezing (12%), facial expression (10.1%), and gait (18.4%) required greater interpretative and comprehension abilities. The lack of familiarity with technical terms, despite facilitation by researchers, combined with low education levels, may have contributed to misunderstandings, leading to false positives. These are cases where results indicate the presence of the disease when they are not actually related to it. **Conclusion:** In a screening process, the level of education can affect responses, as some individuals may have difficulty understanding the questions. Therefore, it is important to consider these limitations and find ways to make the questions more accessible and understandable for all participants. This will also facilitate the screening process for the diagnosis of PD.

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Imaging pattern in hypoglycemic encephalopathy: an important differential diagnosis to remember

Carolina Oldoni[1], Rhaná Carolina Santos[2], Victor Hugo Wilhelm Annes[2], Lorenzo Gabriel de Azevedo Viera[2], Enio Bonamigo Junior[3], Ana Letícia Amorim de Albuquerque[2]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade do Vale do Rio dos Sinos, São Leopoldo RS, Brazil.

[3]Digimax Medicina Diagnóstica, Pato Branco PR, Brazil.

Correspondence: Carolina Oldoni; Email: carololdoni15@gmail.com.

Case presentation: A 32-year-old male patient, obese, smoker, alcohol user, and with type 1 Diabetes Mellitus (DM) managed with NPH and regular insulin, presented to an emergency room with generalized tonic-clonic seizures (GTCS) after inadvertently taking extra doses of regular insulin. Upon arrival, he experienced new episodes of GTCS, and was diagnosed with Status Epilepticus (SE) in the context of hypoglycemia (capillary blood glucose=25). Hypoglycemia was corrected, empirical thiamine was started, and the SE medication management was guided by continuous electroencephalogram. Despite the primary cause being evident from the onset, given the refractoriness even with adequate glycemic management, cerebrospinal fluid investigation and cranial CT scan were performed, both normal. The patient was free of seizures after 24 hours and extubated within 3 days. He presented with severe comprehension aphasia. Contrast-enhanced cranial CT was repeated, showing bilateral and symmetrical cortical contrast enhancement in the temporo-parieto-occipital transition, right olfactory sulcus, and insula. Cranial MRI revealed scattered cortical enhancement segments bilaterally, predominantly in the temporal lobes and temporoparietal transition, also involving the right insula and right frontal lobe. T2/FLAIR hyperintensities were observed in the anterior portions of the hippocampi, as well as gadolinium enhancement areas in the hippocampi. On DWI b1000 scans, there were clear bilateral cortical hyperintensity, mainly temporal. A diagnosis of hypoglycemic encephalopathy (HE) was made with MRI and postictal findings. **Discussion:** Imaging changes in the postictal context are widely known and routine in neurology. However, MRI changes in hypoglycemic encephalopathy are not as frequently observed. Classically, there is involvement of the cerebral cortex (mainly parieto-occipital and insula), hippocampus, posterior limb of the internal capsule, and basal ganglia (when involved, associated with a worse prognosis), typically bilateral. The cerebellum, brainstem, and thalami are spared. It is considered that DWI/ADC shows the earliest and most sensitive changes, indicating diffusion restriction, which is often reversible. There is the presence of T2/FLAIR hyperintensity and T1 hypointensity. Corpus callosum (CC) involvement can occur, producing the so-called boomerang sign (cytotoxic edema in the splenium of the corpus callosum, typically seen in cytotoxic lesions of the CC). The gadolinium enhancement changes in this patient can be well explained by the postictal state. **Final comments:** This report highlights the imaging pattern shown on MRI of a patient with hypoglycemic encephalopathy, with the potential for prognostic information depending on the affected areas. Considering the imaging pattern alone, hypoxic-ischemic injury (which would involve the thalami) and Creutzfeldt-Jakob disease (different clinical presentation) are also admitted as differential diagnoses.

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Imbalance in a 44-year-old woman with atrophy cerebelar in MRI: a case report of spinocerebellar ataxia type 42 (SCA42)

Felipe Cenicio Juliatti, Camila Rodrigues Nepomuceno[1], Maria Fernanda Hamamoto Leati[1], Nayara de Paula Guerreiro[1], Julian Leticia de Freitas[1], Maria Sheila Guimaraes Rocha[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Felipe Cenicio Juliatti; Email: felipe_juliatti@hotmail.com.

Case presentation: A 44-year-old female patient without comorbidities was referred from primary care to the tertiary health service, complaining of imbalance for one year. Her husband reported that she walked as if she were drunk. She denied falls or changes in strength. She denied tremors. She did not present dysphagia, sensitivity, or sphincter changes. She has a familiar history; her mother and her sister have the same clinical picture. Neurological examination showed gait with a broad base, normal deep tendon reflexes, and Romberg negative. During the investigation, his laboratory tests were within normal limits. The brain MRI demonstrated the prominence of cerebellar folia, more evident in the upper and middle third of the vermis and cerebellar hemispheres, inferring volumetric reduction/atrophy. The genetic panel for hereditary ataxias revealed the presence of a heterozygous pathogenic variant in the CACNA1G gene (NM_018896.5:C.5144G>A). This variant is associated with spinocerebellar ataxia 42 (SCA42) and early-onset spinocerebellar ataxia 42 with neurodevelopmental deficit (SCA42ND). **Discussion:** SCA42 is a rare, autosomal dominant disease caused by a heterozygous mutation in the CACNA1G gene on chromosome 17q21. CACNA1G is a gene responsible for the formation of calcium channels, and a mutation in this gene can alter the physiological properties of the channel, resulting in more positive (depolarized) action potentials, indicating that it is a channelopathy. Affected individuals present gait instability and cerebellar signs such as dysarthria, nystagmus, and saccadic intrusions. The age of onset and severity of the disease are highly variable, although most symptoms begin in middle adulthood, with slow progression. Imaging examination demonstrates atrophy of the cerebellum and cerebellar vermis. **Final comments:** Hereditary ataxias are rare diseases, and their diagnosis requires evaluation by qualified professionals and confirmatory genetic tests. Delays in clinical suspicion can increase unnecessary tests and mistaken diagnoses. It is essential to correctly diagnose and carry out multidisciplinary monitoring of these patients to avoid greater social stigmatization and futile therapies.

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Immunosuppression in neuromyelitis optica spectrum disorder: a trial sequential analysis and updated meta-analysis

Artur Menegaz de Almeida[1], Maria Eduarda Cavalcanti[2], Francisco Cezar Aquino de Moraes[3], Fernando Westphal Filho[4], ítalo Barros Andrade[5], Paulo Robeto Moss Lopes[4], Clara Rocha Dantas[6], Mariana Rachas Reis[7], Lilianne Fernandes[8], Francinny Alves Kelly[9]

[1]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[2]Universidade de Pernambuco, Recife PE, Brazil.
[3]Universidade Federal do Pará, Belé
m PA, Brazil.
[4]Universidade Federal do Amazonas, Manaus AM, Brazil.
[5]FSA, Vitória da Conquista BA, Brazil.
[6]Universidad de Buenos Aires, Buenos Aires, Argentina.
[7]FSC, São Paulo SP, Brazil.
[8]AFM, Abaetuda PA, Brazil.
[9]DPIC, São Paulo SP, Brazil.

Correspondence: Artur Menegaz de Almeida; Email: arturomenegaz@gmail.com.

Background: Neuromyelitis Optica Spectrum Disorder (NMOSD) is a devastating, disabling, autoimmune disease with clinical features mainly involving the optic nerves and spinal cord. Rituximab (RTX) is a longstanding CD20-positive B-cell depleter agent used for treating NMOSD. Azathioprine (AZA) and mycophenolate mofetil (MMF) are lymphocyte depleters also used as first-line therapy in these patients, both being associated with less safety and efficacy when compared to rituximab. **Objective:** To assess rituximab safety and efficacy and to directly compare it with azathioprine and mycophenolate mofetil. Additionally, we aim to test these findings with a trial sequential analysis (TSA) to investigate their reliability. **Methods:** MEDLINE, SCOPUS, Web of Science, and the Cochrane Library were systematically searched until April 28, 2024. We included observational studies and randomized controlled trials assessing rituximab therapy in NMOSD patients. In addition, we also included studies comparing rituximab to azathioprine and/or mycophenolate mofetil. All confidence intervals were set at 95%. The trial sequential analysis was applied for both continuous and dichotomous outcomes. Rstudio version 4.3.2, Review Manager 5.4.1, and the TSA software were used for statistical analysis. **Results:** 44 studies were included, of whom 27 were retrospective studies, 8 were prospective studies, and 2 were randomized controlled trials. There were 2047 patients, of whom 1336 (65.26%) were treated with RTX, 417 (20.37%) received AZA, and 294 (14.36%) received MMF. Concerning the annualized relapse rate (ARR) and the expanded disability status scale (EDSS), RTX showed a greater effect size in both: (ARR; MD -1.33; 95% CI 1.07 to 1.60; P<0.000001; I^2=93%) and (EDSS; MD -0.89; 95% CI 0.63 to 1.15; P<0.000001; I^2=67%), respectively. The TSA reinforced this result. The comparison between RTX and both AZA and MMF also tended in favor of RTX in the ARR (MD -0.18; 95% CI -0.34 to -0.02; P=0.026922; I^2=90%) but not in the EDSS, which demonstrated no difference between groups (MD -0.10; 95% CI -0.43 to 0.23; P=0.563; I^2=35%). The TSA was not confirmatory for the analysis of RTX vs. AZA but assured the superiority of RTX over MMF. The time to first relapse was statistically significantly lower in patients treated with RTX (HR 1.7465; 95% CI 1.3343 to 2.2862; P<0.001; I^2=26%). Tolerability analysis also tended in favor of RTX administration when compared to AZA and MMF (occurrence of adverse events; 2.83% vs. 10,33%; RR 0.42; 95% CI 0.19 to 0.91; P=0.027; I^2=49%), which was also demonstrated in the TSA. **Conclusion:** This updated systematic review and meta-analysis with trial sequential analysis assessed the safety and efficacy of RTX compared to AZA and MMF for NMOSD. Our study confirms, through TSA, that RTX is associated with a greater effect in improving ARR, EDSS, and time to relapse, and is associated with better tolerability in NMOSD patients.

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Impact of ambient temperature in Teresina/Piauí on multiple sclerosis: an analysis of quality of life, severity of fatigue, sexual performance, and self-medication

Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Francisco Héricles Moreira de Carvalho[1], Samuel de Castro Campos[1], Lara Beatriz Alves Batista[1], Vitor de Deus da Rocha Ribeiro Gonçalves[2], Kayron Rodrigues Lima[1], José Henrique de Melo Feitosa[1], Thalyne Gonçalves Barbosa[1], Hanna Kathrine Vasconcelos Silva Gomes[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Santa Casa de Miserdicórdia de Belo Horizonte, Departamento de Neurocirurgia, Belo Horizonte MG, Brazil.

Correspondence: Ocílio de Deus da Rocha Ribeiro Gonçalves; Email: ocilio41@gmail.com.

Background: Multiple sclerosis (MS) is a neurological disease that impacts functionality and quality of life (QoL). One aspect of the condition is fatigue, which worsens with heat and in warmer climates. Global warming is a worldwide concern, and recent studies suggest that the current climate of Teresina during the hottest months serves as a prototype for the average temperatures estimated for many areas of the planet in ten years, following the current path of climate change. **Objective:** To analyze the degree of severity of fatigue, sexual performance, QoL, and self-medication in patients with MS in Teresina-PI, comparing periods with higher and milder temperatures throughout the year. **Methods:** This is a prospective observational study conducted between january 2022 and november 2022. It includes MS patients according to the modified McDonald criteria from the MS referral center in the city of Teresina-PI. Patients were evaluated during

the hottest and mildest periods using fatigue scales (FSS), EDSS, QoL, and a self-medication questionnaire. Statistical analysis was performed using SPSS software. The paired t-test was used to compare dependent parametric variables (fatigue and EDSS). The Wilcoxon test was used to compare dependent non-parametric variables (sexual performance and QoL). A one-sample chi-square test was used for a single non-parametric categorical variable (self-medication in cold compared to hot periods). A p-value of <0.05 was considered significant. **Results:** The sample consisted of 41 patients with a mean age of 39.6 years, 65.8% of whom were female. The mean EDSS was 3.04 (hot) and 3.11 (mild), and the total (mean) FSS of the patients during the hot period was 88.24 (2.87), while that of the patients during the cold period was 61.24 (2.04) (p<0.001). Regarding self-medication in the mild compared to the hot period, a decrease was observed in 51.6% of the sample, with 45.2% maintaining the same level and only 3.2% increasing usage (p<0.002). For EDSS (p=0.574), impairment in sexual performance (p=0.122), and QoL (p=0.061), comparing patients in the hot and mild periods, no significant difference was observed in the obtained data. **Conclusion:** Comparing periods with higher and milder temperatures throughout the year in Teresina-PI, there is a significant association between worse fatigue severity in MS patients and higher ambient temperature. Patients tend to significantly reduce self-medication during milder periods of the year, despite maintaining the disease's impact as measured by EDSS.

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Impact of benzodiazepine use on quality of life, mental health and cognitive function in the elderly population

Abner Lucas Balduino de Souza[1], João Ricardo Sousa Vasconcellos[1], Déborah Helena Pereira Pinheiro[1], Izabella Gomes de Souza[1], Flávia Gonçalves Vasconcelos[1], Bráulio Brandão Rodrigues[2], Cristiane Teixeira Vilhena Bernardes[1]

[1]Universidade Evangélica de Goiás, Anápolis GO, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Abner Lucas Balduino de Souza; Email: aabnerlucass@gmail.com.

Background: Benzodiazepines (BZD) are among the most prescribed psychotropic drugs in clinical practice, commonly used to treat mood disorders, anxiety, depression, epilepsy, and insomnia. However, their use is increasingly prevalent among the elderly, often without appropriate therapeutic indications, leading to misuse. This misuse is associated with chemical dependence, worsening of delusional conditions, increased risk of falls, and cognitive changes. **Objective:** To compare the aging process of elderly individuals using benzodiazepines with those who do not. **Methods:** This cross-sectional, descriptive study was conducted in Anápolis, Goiás, including individuals over 60 years old. Data were collected after ethical approval using nine questionnaires: sociodemographic, WHOQOL-bref (World Health Organization Quality of Life Instrument), Katz Index, Lawton Scale, GDS-15 (Geriatric Depression Scale), HAM-D (Hamilton Depression Rating Scale), GAD-7 (Generalized Anxiety Disorder 7-item scale), MoCA (Montreal Cognitive Assessment), and Morse Fall Scale. Data were analyzed using IBM SPSS Statistics 29.0.1.0 with Fisher-Freeman-Halton exact test. **Results:** A total of 37 interviews were conducted, with one excluded due to withdrawal, leaving 36 participants (30.6% men). Most were aged 60-69 (50%), married or in a stable union (61.1%), mixed race (69.4%), and had only primary education (63.9%). BZD use was reported by 13 (36.1%) participants, primarily for insomnia (53.8%). Quality of life, measured by WHOQOL-bref, showed that BZD users had lower physical domain scores (mean 56.74%, p=0.058). Depression, assessed by GDS-15, indicated that 75% of those with scores suggesting depression were BZD users (p=0.059). The HAM-D scale also showed a significant association between BZD use and depression (p<0.001). Anxiety levels, measured by GAD-7, showed no significant difference related to BZD use (p=0.610). Cognitive function, assessed by MoCA, revealed that 92.3% of BZD users scored below the cutoff of 26 points, although this was not statistically significant (p=0.114). Fall risk, assessed by the Morse Fall Scale, indicated that 46.1% of BZD users had a low fall risk, and only 15.4% had a high fall risk, compared to 4.3% of non-users (p=0.214). **Conclusion:** The findings suggest that benzodiazepine use in the elderly is associated with poorer physical health, higher rates of depression, and lower cognitive function, although the latter was not statistically significant. These results highlight the need for careful prescription practices to avoid the negative impacts of these drugs on healthy aging.

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Impact of delirium and its timing of onset on stroke outcomes

Gabriel Souza Suzart[1], João Pedro Fernandes Gonçalvez[1], Marivaldo Yslan Silveira Leal[1], Luísa Dias Moreno Aravena[1], Thais Barbosa de Oliveira[1], Alex de Cerqueira Silveira Figueiredo[1], Gabriel Souza Medrado Nunes[1], Pedro Antonio Pereira de Jesus[1], Vinícius Bessa Mendez[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Luísa Dias Moreno Aravena; Email: luisaaravena@ufba.br.

Background: The literature is scarce of studies about the effect of delirium onset on outcome of stroke patients. **Objective:** To determine the frequency of delirium by onset timing, its clinical profile and impact on long-term functional outcome. **Methods:** Prospective cohort of stroke patients admitted to a Brazilian reference stroke unit. Subjects included were older than 18 years with stroke diagnosis within 72 hours of symptoms onset. Delirium was assessed during hospitalization in alternate days during 30 days or discharge with the Confusion Assessment Method for the Intensive Care Unit (CAM-ICU). Delirium was classified according to the day of onset by early (within 3 days from stroke) or late (after 3 days). The main outcome was functional dependence or death in 90 days after stroke, determined by the modified Rankin Scale >2 points. **Results:** A total of 585 patients were included, with a mean age of 62.22 (±13.46) years. Delirium occurred in 114 (19.5%) patients and was associated with infection (p<0.001), with no difference between early and late onset delirium (p>0.05). Patients with Late-onset delirium were more common (71.9%) and had worse outcomes in 90 days compared to those with no delirium [(76.1% vs 34.6%) p<0.001] and no difference compared to early-onset delirium was found [(76.1% vs 70%) p=0.556]. However, there were no significant differences in outcomes in the multivariate analyses, adjusted for age, sex, NIHSS at admission, occurrence of infection during hospitalization and thrombolysis [OR 2.32 (IC95% 0.83-6.49) P=0.108]. **Conclusion:** Delirium presents as a frequent clinical challenge in Stroke Units. Although our study did not find a significant association with poorer outcomes, late-onset delirium may be more prevalent and as morbid as the early-onset. Thus, continuous assessment for delirium during hospital stay remains important, since worse outcomes are present both in early and late delirium. Further research is crucial to fully understand and assess its impact on this patient population.

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Impact of glucagon-like-peptide-1 receptor agonists on non-motor symptoms in Parkinson's disease: a systematic review and meta-analysis

Regina Carneiro Leão[1], Caio Rezende Lima[2], Jorge Ferreira Jasmineiro Pitanga[3], Milena Lima Loures[4], Murilo Marmori Cruccioli[5], Filipe Sarmento[6]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Universidad de Buenos Aires, Buenos Aires, Argentina.
[4]Faculdade Ciências Médicas de Minas Gerais, Belo Horizonte MG, Brazil.
[5]Universidade Evangélica de Goiás, Anápolis GO, Brazil.
[6]University of Florida, Florida, USA.

Correspondence: Maria Regina Vendas Carneiro Leão; Email: mariaregina.leao@ufpe.br.

Background: Glucagon-Like-Peptide-1 Receptor Agonists (GLP-1 RAs), initially designed for diabetes management, show potential for treating motor disability in Parkinson's Disease (PD) due to promising pre-clinical and clinical trial results. Previous studies including PD and non-PD populations also suggest GLP-1 RAs may improve cognition and reduce depression and anxiety. Despite the significant impact of non-motor symptoms (NMS) on PD patients' quality of life, these symptoms are often overlooked or briefly assessed in GLP-1 RA studies. **Objective:** To perform a systematic review and meta-analysis to examine the impact of GLP-1 RA administration on non-motor symptoms in Parkinson's disease (PD). **Methods:** We systematically searched PubMed, Embase, and Cochrane databases for randomized controlled trials (RCTs) comparing GLP-1 RAs to no GLP-1 RAs use in PD. Using a random-effects model, we analyzed studies after critical appraisal. Primary outcomes were the mean difference from baseline in reported non-motor symptoms, assessed using the Movement Disorder Society-Unified Parkinson's Disease Rating Scale (MDS-UPDRS) Part I, Non-Motor Symptoms Rating Scale (NMSS), Beck Depression Inventory (BDI), Montgomery-Asberg Depression Rating Scale (MADRS), Montreal Cognitive Assessment (MoCA), and Mattis Dementia Rating Scale (MDRS). Due to variations in the availability of depression and cognitive scales across studies, we used the standard mean difference (SMD) to compare effect sizes for these outcomes. To account for multiple comparisons and reduce the risk of Type I errors, we applied Bonferroni correction (adjusted p-value of 0.0125). Statistical analysis was conducted using Review Manager 5.4.1, and heterogeneity was assessed with I^2 statistics. **Results:** Analyzing data from five RCTs (569 patients, 59% on GLP-1 RA) with 9-15 month follow-ups, GLP-1 RA significantly improved cognitive functions in PD patients (SMD 1.11; 95% CI: 0.29 to 1.94; p=0.0082; I^2: 94%), maintaining significance after adjusting for multiple comparisons. Depression scores showed a medium effect size change towards improvement (SMD -0.49; 95% CI: -0.95 to -0.02; p=0.04; I^2: 69%), but lost significance after correction. NMSS and MDS-UPDRS Part I changes were non-significant (p=0.99 and 0.29, respectively). High heterogeneity was observed in cognitive (I^2: 94%), depression (I^2: 69%), NMSS (I^2: 68%), and MDS-UPDRS Part I (I^2: 96%) outcomes. **Conclusion:** GLP-1 RAs significantly improved cognitive functions in PD patients, suggesting a potential benefit in treating cognitive impairment. However, the trend towards improvement in depression with GLP-1 RA treatment was not statistically significant after adjusting for multiple comparisons. Substantial heterogeneity among studies may have influenced results, affecting the reliability of findings. Future studies should use comprehensive assessments and longer follow-up to better understand GLP-1 RAs' impact on NMS in PD.

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Impact of neurodegenerative diseases in Brazil: an analysis of mortality from Alzheimer's and Parkinson's from 2012 to 2022

Júlio César Soares Barros[1], Antonia Cardoso Silva[1], Guilherme Carvalho de Souza[1], Larissa da Silva Almeida[1], Mariana Reis Prado[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.

Correspondence: Júlio César Soares Barros; Email: julio.barros@famed.ufal.br.

Background: Alzheimer's disease (AD) and Parkinson's Disease (PD) are neurodegenerative diseases that pose a growing challenge to public health due to the aging global population. AD is characterized by progressive loss of memory and cognitive functions, while PD is marked by tremors, rigidity, and motor difficulties. Both significantly affect the quality of life of patients and their families, besides representing a substantial economic impact on the healthcare system. Analyzing mortality data related to these diseases is essential to understand their prevalence and to develop effective prevention and treatment strategies. **Objective:** To analyze mortality from Alzheimer's and Parkinson's in Brazil between 2012 and 2022, observing growth trends and correlated factors, distribution by age group, and gender. **Methods:** A cross-sectional, retrospective, and descriptive epidemiological study was conducted using data from the Mortality Information System (SIM/DATASUS) for the period between 2012 and 2022. The analyzed variables included "Region", "Age Group", and "Gender". Population data were obtained from the Brazilian Institute of Geography and Statistics (IBGE) to calculate mortality rates per million inhabitants for each region. **Results:** The total number of deaths between 2012 and 2022 was 265,112, with 221,004 from AD and 44,108 from PD. The Southeast and South regions led, respectively, with 138,421 and 54,096 deaths. A nearly linear increase in mortality was observed over the years, with an 89.6% increase in PD and a 123.9% increase in AD. Analyzing mortality per million inhabitants, the South and Southeast regions continued to lead in the number of deaths, with the South ahead of the Southeast in 2022, reaching 248.03 deaths against 197.67. Regarding gender, both men and women had significantly higher mortality from AD compared to PD. However, women had a higher mortality rate compared to men for both diseases, especially for AD (143,239 against 77,743 deaths). In the age group analysis, mortality increased significantly from the age of 50, with a sharper rise from the age of 70. AD had a much greater impact on the elderly, particularly those over 80 years old, with 168,280 deaths compared to 25,752 from PD. **Conclusion:** The increase in mortality during the analyzed period can be attributed to the aging Brazilian population, as these diseases are more prevalent in the elderly. Furthermore, the improvement in diagnostic methods and greater awareness of these diseases may have contributed to the increase in reported deaths. This growth highlights the need for health policies focused on prevention, early diagnosis, and effective treatment of neurodegenerative diseases. Implementing public health strategies that address these factors is crucial to improving the quality of life and reducing mortality associated with these diseases in Brazil.

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Impact of obstructive sleep apnea in post thrombolysis context of stroke patients: a transversal study

Aurea Maria Lago Novais Castello Branco[1], Juliana Cerqueira Gomes[1], Marina Soledad Benitez Bitencourt[1], Thaise Braga de Oliveira[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Aurea Maria Lago Novais; Email: aureanovais20.1@bahiana.edu.br.

Obstructive sleep apnea (OSA) is a disease of upper airways, diagnosed with polisomnography, that is well associated with stroke through diverse mechanisms. Despite that, it still lacks studies about its role in outcomes after thrombolysis in stroke patients. To elucidate the difference of post thrombolysis outcomes in stroke patients with different risks of obstructive sleep apnea (OSA). We performed a transversal study, part of a cohort of stroke patients. Patients were evaluated of risk of OSA with STOP-Bang and SOS scores; also, data of previous comorbidities were collected. Outcomes evaluated were CTI time, internment time, delirium, infection, hemorrhagic transformation and death during internment. We compared groups with T test, Mann-Whitney or chi-square (considering p<0.05). Multivariate analysis with logistic regression was performed for each outcome, considering p<0.05. We obtained 48 patients, 73.5% classified as high risk with STOP-Bang and 49% with SOS score. Patients with high risk of OSA had higher internment (median 11 days; IQR 8-16) and CTI time (median 3 days; IQR 0-5) in comparison with low-risk group (internment median time 9 days IQR 6-14, CTI median time 2 days IQR 0-5). Also, they evolved more to all outcomes, but only delirium was statistically significant (OR=2.2; CI 95% 1.3-3.3; p=0.01). Considering STOP-Bang, age was independently associated to CTI time (OR=1.1;CI 95% 1-1.2;p=0.04) and infection (OR=0.84;CI 95% 0.72-0.98;p=0.02); hypertension with delirium (OR=0.05;CI 95% 0.003-0.95;p=0.04). Although patients with high risk of OSA evolved more to all outcomes, they were not statistically significant, suggesting there is the same safety profile of thrombolysis in both groups. The discrete protector mechanism of hypertension may be due to reduced sample or a maintenance of certain cerebral perfusion that prevents delirium. Further studies are needed to clarify this data.

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Impact of obstructive sleep apnea on immediate outcomes in stroke patients

Marina Soledad Benitez Bitencourt[1], Aurea Maria Lago Novais Castello Branco[1], Juliana Cerqueira Gomes[1], Thaise Braga de Oliveira[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Aurea Maria Lago Novais; Email: aureanovais20.1@bahiana.edu.br.

There is a great association between obstructive sleep apnea (OSA) and cerebrovascular diseases, acting through many pyshiopathological mechanisms. However, the impact of OSA on immediate outcomes in stroke patients remains underdeveloped. This study aims to elucidate which characteristics of patients with OSA significantly affects immediate outcomes. We performed a transversal study, part of cohort of stroke patients. Considering STOP-Bang and SOS scores, we stratified their risk of obstructive sleep apnea. Data of previous comorbidities were collected as well. In the context of immediate outcomes, we evaluated delirium, infection, new stroke occurrence and death during internment. Furthermore, hemorrhagic transformation, CTI time and internment time were also evaluated. We performed an univariate analysis, using T test/Mann-Whitney/chi-square, depending on normality. After, a logistic regression with multivariate analysis. Correlation between scores was done with Spearman or Pearson test. We obtained 430 patients: 69% high risk with STOP-Bang and 50% with SOS. There were significant differences between groups: in the high risk group, 50% were female. In the high risk group of STOP-Bang score, 80% had hypertension, 38.8% had diabetes and 58.4% related previous regular use of medication. Also, median age 65 (IQR 56-72; p=0.003), 19.9% were obese and 22% related alcoholism. About internment time, the results showed median time 10 (IQR 7-16). High risk group evolved more to delirium (p=0.02), infection (p=0.04), death (p=0.04). Age was independently associated to delirium (OR=0.96; CI 95% 0.93-0.98; p=0.003), infection (OR=0.95; CI 95% 0.92-0.97; p<0.001), death (OR=0.95; CI 95% 0.91-0.99; p=0.01); diabetes (OR=2.6; CI 95% 1.11-3.82; p=0.022) to delirium; alcoholism to CTI time (OR=0.32;CI 95% 0.14-0.73; p=0.007). According to SOS score: 50.4% were female. In the high risk group: 76.7% had hypertension, 41% had diabetes, 23% were obese, and BMI median 26.2 (IQR 23.1- 29.6; p=0.015). Besides results, it is noteworthy that patients with high risk of OSA show an absolute number of hemorrhagic transformation, death, and new stroke occurrences during internment. Overall, these observations confirms that STOP-Bang and SOS scoring systems have a similar distribution, reafirming metabolic diseases, hypertension, and diabetes are prevalent in high-risk groups for obstructive sleep apnea (OSA), regardless of the scoring system used. In conclusion, this study elucidates the significant impact of obstructive sleep apnea (OSA) on immediate outcomes in stroke patients due to the observed associations with adverse outcomes such as delirium, infection, death, and new stroke occurrences. Findings reaffirm the need for comprehensive screening and management strategies for OSA in stroke patients to improve outcomes and enhance patient care.

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Impact of oxyhemoglobin desaturation on cerebral compliance in patients with obstructive sleep apnea syndrome: a pilot study

André Caldeira Grobério[1], Gilmar Fernandes do Prado[1], Douglas Sousa Soares[1], Ana Flávia Machado Pitz[1], Davi Dantas Muniz[1], Tainã Paulo Zanata Trombetta[1], Letícia Lambert[1], Lucas Martins Teixeira[1], Karla Carlos[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Davi Dantas Muniz; Email: davimuniz@msn.com.

Background: Obstructive sleep apnea (OSA) is a systemic inflammatory disease considered a risk factor for developing cerebrovascular diseases. Considering its complex pathophysiology, OSA also causes hemodynamic dysfunctions, altering CO2 and O2 concentrations and possibly compromising cerebral autoregulation. In this context, little is known about its impact on cerebral compliance. **Objective:** To assess the impact of desaturation on cerebral compliance in patients with mild obstructive sleep apnea syndrome (OSAS). **Methods:** Cerebral compliance was assessed through P2/P1 ratio of the ICP pulse morphology, using the Brain4care® device during Type 1 polysomnography. Statistical analysis was performed using mean and standard deviation, with Student1s t-test for group comparisons (p<0.05). **Results:** Two patients were included, one diagnosed with mild OSAS and one control. The mean age was 38 years ± 0.7. Both patients were female. The mean recording time of the P2/P1 ratio was 362±33 minutes. The patient with OSAS exhibited 57 hypopneas, with 40 of them accompanied by desaturation. The average duration of desaturations was 18 seconds. The mean P2/P1 ratio during desaturations was 1.12. There was no difference in the P2/P1 ratio when comparing events with and without desaturation (p>0.05). The participant without OSAS presented only 2 hypopneas with desaturation, with an average duration of 45 seconds and a P2/P1 ratio of 0.88. Regarding cerebral compliance behavior, we observed an increase in the P2/P1 ratio in the patient with OSAS compared to the control (1.07 vs. 0.88, p<0.0001). The presence of desaturation did not modify the P2/P1 ratio. However, cerebral compliance appears to be reduced in mild OSAS. According to the morphology of the intracranial pressure pulse, three waves are analyzed: P1, corresponding to arterial pulsation; P2, corresponding to cerebral compliance; and P3,

corresponding to aortic valve closure. Based on morphology, we can classify the P2/P1 ratio as normal (0.65 – 1.00) or suspicious of intracranial hypertension when P2>P1. In our study, we found values suggesting morphofunctional alterations in the patient with mild OSAS, classified as at risk for intracranial hypertension. Idiopathic intracranial hypertension has been studied for a long time, but its pathophysiology has not been well elucidated. It primarily affects obese patients, preferably females, and is believed to be multifactorial in etiology. The patient in this study, despite being female, does not present signs or symptoms of idiopathic intracranial hypertension. It is noteworthy that the intracranial pressure varies, with non-hypertensive indices close to the patient's awakening, which raises questions about a possible manifestation of the disease, although a possible subclinical manifestation cannot be ruled out. **Conclusion:** Cerebral compliance is reduced in patients with mild OSAS, and desaturation did not intensify this alteration.

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Impact of physical activity on the evolution of symptoms in patients with Parkinson's disease

Tatiana Pizzolotto Bruch[1], Camila Pezente Dal Toe[1], Caroline Liberato Pereira[1], Barbara da Silva Becker[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Tatiana Pizzolotto Bruch; Email: tatibruch@yahoo.com.br.

This study investigated the impact of physical activity (PA) on the progression of symptoms in Parkinson's Disease (PD), given the absence of pharmacological treatments that halt its progression. A cross-sectional study was conducted with 41 patients diagnosed with PD in Criciúma, Balneário Rincão, and Içara. The PDQ39 questionnaire was used to assess symptom progression, focusing specifically on the effects of PA. The majority of patients (58.5%) engaged in some form of PA, predominantly aerobic activities. Among participants, 31.7% reported significant improvement in PD symptoms after starting PA. There was a statistically significant association between PA practice and improvements in mobility and communication among patients. Additionally, patients who engaged in more than 150 minutes of PA per week showed better outcomes on the PDQ39 quest. In conclusion, the use of physical exercise as an adjunct to PD treatment can provide significant benefits in emotional, communication, and mobility aspects. Aerobic and resistance activities have shown positive impacts on disease progression, although these effects vary from patient to patient.

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Impact of renal dysfunction on post-stroke rehabilitation in patients at high risk for obstructive sleep apnea

Juliana Cerqueira Gomes[1], Aurea Maria Lago Novais Castello Branco[1], Marina Soledad Benitez Bitencourt[1], Thaise Braga de Oliveira[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Aurea Maria Lago Novais; Email: aureanovais20.1@bahiana.edu.br.

Literature indicates a link between obstructive sleep apnea(OSA)and kidney impairment. Also,stroke patients with OSA often have longer rehabilitation. However, data of renal impact on stroke recovery is limited. Assess the impact of renal dysfunction on rehabilitation outcomes in high-risk post-stroke patients. We performed a cohort of stroke patients stratified by risk of OSA with STOP-Bang and SOS scores. Also, data of previous comorbidities and renal function (creatinine and urea) at admission, peak, and discharge was assessed, alongside functional outcomes (modified Rankin Scale and Barthel Index) at 3.6, and 12 months. We performed a univariate analysis (T test/Mann-Whitney/chi-square), considering p<0.1, and a multivariate analysis with logistic regression (considering p<0.05). We obtained 415 patients (69% high risk with STOP-Bang and 51% with SOS). Functional outcomes data were available for 49 and 40 patients, respectively. In univariate analysis for STOP-BANG, admission creatinine (median 1, IQR 0.8-1; p<0.01), high creatinine (median 1, IQR 0.9-1; p<0.01), peak creatinine (median 1, IQR 28-50.6; p<0.01),discharge urea (median 38, IQR 28-50; p<0.01), and peak urea (median 49, IQR 36-83; p=0.01) showed difference between groups. For the SOS, discharge urea levels (p=0.02). Regarding functional outcomes, STOP-BANG patients had median Rankin scores of 2 (IQR 1-4) at 3 months,1 (IQR 1-4) at 6 and 12 months; with Barthel medians of 85 (IQR 49.5-95), 93 (IQR 69.5-100) and 95 (IQR 68-100) at the same time. For the SOS group, median Rankin scores were 2 (IQR 1-4) at 3 months,1 (IQR 0.2-3) at 6 months, and 1 (IQR 0.2-3) at 12 months with Barthel medians of 85(IQR 55-98), 94 (IQR 71-100) and 96 (IQR 69-100), at the same time. In multivariate analysis, diabetes in STOP-BANG patients was independently associated with worse Rankin (p=0.02, OR=0.2, 95% CI 0.1-0.6) and Barthel (p=0.05, OR=3, 95% CI 1.4-7) at 3 months, and on the Barthel at 6 months (p=0.02, OR=5, 95% CI 1.2-2). For the SOS group, discharge urea levels were associated with worse Rankin (p=0.01, OR=1.0, 95% CI 1.01-10.6) and Barthel (p=0.01, OR=0.9, 95% CI 0.94-0.99) at 3 months, and on the Rankin at 12 months (p=0.04, OR=1.04, 95% CI 1.01-1.08). The study found significant differences in renal function and functional outcomes among high-risk patients. Diabetes predicts poorer outcomes in STOP-BANG patients, while high discharge urea levels predict worse outcomes in SOS patients. Effective management of these factors is crucial for improving long-term outcomes.

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Impact of sars-cov-2 infection on the development of multiple sclerosis

Karoline de Alvarenga Leandro Lobo[1], Saulo Machado Moreira Sousa[1], Sheila Espírito Santo Araújo[1]

[1]Universidade Estadual do Norte Fluminense, Campos dos Goytacazes RJ, Brazil.

Correspondence: Karoline de Alvarenga Leandro Lobo; Email: alvarengalobo@gmail.com.

Background: Multiple sclerosis (MS) is a pathology characterized by inflammatory and autoimmune courses, being the most prevalent and debilitating condition within the spectrum of central demyelinating diseases. As of 2020, its national prevalence was 15 cases per 100,000 inhabitants. Between 2020 and 2022, the world was impacted by a pandemic caused by the SARS-CoV-2 coronavirus, and its infection has been correlated with exacerbation of demyelinating episodes in various autoimmune comorbidities similar to MS. Given this, we question in the scope of this work whether SARS-CoV-2 is capable of triggering MS. **Objective:** Conduct retrospective and prospective analyses of clinical, laboratory, and radiological data from patients infected with SARS-CoV-2, identifying whether this was the trigger for the development of MS. **Methods:** We conducted a survey of the number of hospitalizations for MS and SARS-CoV-2 infections from national data from DATASUS/Tabnet from April 2020 to January 2021. The temporal interval reflects the period from the beginning of the pandemic to the first dose of the vaccine. To determine the existence of a linear relationship between the prevalence of MS and COVID infection during this same period, we performed a regression analysis. **Results:** In the case of the national results obtained from DATASUS, we found a Pearson coefficient of 0.9937 and a p-value less than 0.0001. Similarly, we quantified the number of hospitalizations for MS at Hospital São José do Avaí (HSJA) during the same period and evaluated its correlation with COVID prevalence. With a similar result, the coefficient was 0.9872 and the p-value was less than 0.0001, indicating a strong correlation between the events. **Conclusion:** Therefore, this work contributes to the establishment of the relationship between MS and viral triggers, considering the SARS-CoV-2 virus as one of the responsible triggers for the pathological trigger.

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Impact of sleep deprivation on neurological and cognitive functions

Guilherme Eugênio Gil[1], Danielle Delgado Diaz Medina[1], Heloisa Helou Doca[1]

[1]Universidade de Marília, Marília SP, Brazil.

Correspondence: Guilherme Eugênio Gil; Email:

Background: Sleep is a natural and periodic physiological condition of brain activity. It is a period of reversible loss of consciousness that provides physical well-being and mental rest, restoring energy for physical and mental activities. Sleep deprivation can lead to structural changes in sleep, sleep disturbances, drowsiness, reduced alertness and impaired cognitive functioning, including attention, working memory, reaction time, logical reasoning and decision-making. In addition, sleep deprivation can disrupt functional and structural areas of the brain that are associated with cognition and emotion. **Objective:** To evaluate the evidence on the effects of sleep deprivation on neurological and cognitive functions, its underlying mechanisms and implications for healthy cognitive performance. **Methods:** This systematic review obtained 37 scientific articles with the descriptors "Cognition", "Insufficient sleep" and "Sleep Deprivation", articulated by the Boolean operators AND and OR. The search was carried out in the PubMed and Scientific Electronic Library Online databases. The inclusion criteria were publications between 2020-2024, in Portuguese and English, which directly addressed the topic. Articles with no direct relation to the topic and other systematic reviews were disregarded. Case presentations and randomized clinical trials were evaluated, after which 17 articles were included for final analysis. **Results:** Results indicate that sleep deprivation increases the nocturnal concentration of beta-amyloid peptides in cerebrospinal fluid in approximately 10% of a sample of middle-aged elderly people, without significantly affecting cortisol, lactate or glucose. There is a decrease in gray matter volume in the anterior insula, default mode network and thalamus, with a significant interactive effect of age. In addition, sleep duration may be directly related to the cognitive domain of verbal fluency. In young people with attention deficit hyperactivity disorder, there was a significant increase in errors of omission and cognitive reaction time variability. In athletic performance, partial sleep deprivation caused subtle but important damage. In one of the studies, it is seen that there is better performance in the occupational task of a sample of elderly people after adequate sleep, while sleep deprivation did not reach the expected level of performance. **Conclusion:** Sleep deprivation has a negative impact on neurological functions, reducing the volume of gray matter in the anterior insula, especially in the elderly, which affects emotional and associative processes. Sleep deprivation also impairs memory consolidation by reducing the synthesis of proteins necessary for synaptic plasticity, which increases vulnerability to neurological damage. Maintaining consistent sleep of an average of 7 hours per night improves the general health of the individual, regardless of their age group, in order to mitigate neurobehavioral deficits.

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Impact of the duration of obstructive sleep apnea on cerebral compliance in humans: a pilot study

Davi Dantas Muniz[1], Gilmar Fernandes do Prado[1], André Caldeira Grobério[1], Douglas Sousa Soares[1], Ana Flávia Machado Pitz[1], Tainã Paulo Zanata Trombetta[1], Leticia Lambert[1], Lucas Martins Teixeira[1], Karla Carlos[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Davi Dantas Muniz; Email: davimuniz@msn.com.

Background: Obstructive sleep apnea syndrome (OSAS) is a common disorder marked by recurrent upper airway collapse during sleep, causing hypoventilation and intermittent hypoxia. These episodes lead to systemic complications, including increased oxidative stress, sympathetic activation, and cerebral microvascular remodeling. These changes may be linked to decreased cerebral compliance and elevated intracranial pressure, though further research is needed to clarify these connections. **Objective:** This pilot study investigates the influence of the duration of obstructive apnea events on cerebral compliance using the non-invasive Brain4care® device. **Methods:** The study included two patients: one with severe OSAS and one healthy control. The diagnosis of OSAS was confirmed by type I polysomnography. Cerebral compliance was assessed through the ratio of P2 and P1 peaks (P2/P1) of the ICP pulse morphology, using the Brain4care® device. Statistical analysis was performed using mean and standard deviation, with Student's t-test for group comparisons ($p<0.05$). **Results:** The mean was 45 years ±10, both females. The OSAS patient had an apnea-hypopnea index (AHI) of 41.3, while the control had an AHI of 4.0. The P2/P1 ratio was significantly higher in the OSAS patient (1.24) compared to the control (0.88) ($p<0.0001$). During REM sleep, the P2/P1 ratio in the OSAS patient was higher (1.31) than during NREM sleep (1.23) ($p>0.0001$). The average duration of obstructive events was 23 seconds in the OSAS patient and 18 seconds in the control ($p<0.0001$). The study demonstrates a significant reduction in cerebral compliance in the OSAS patient, potentially due to prolonged hypercapnia from chronic hypoventilation during sleep. The obstructive events lead to sustained elevations in CO2 levels, causing cerebral vasodilation and increased cerebral blood flow, thereby raising intracranial pressure (ICP). The brain, constrained by the rigid skull, has limited capacity to accommodate this increased blood volume without significant rises in ICP, resulting in reduced compliance. During REM sleep, the OSAS patient exhibited higher P2/P1 ratios than during NREM sleep, suggesting that longer and more frequent obstructive events during REM sleep exacerbate compliance reduction. The extended duration of these events prolongs CO2 retention, intensifying vasodilation and ICP elevation. These results align with previous studies indicating that repetitive extended hypoxic events and sympathetic nervous system activation during apneas contribute to cerebrovascular remodeling and compliance loss. **Conclusion:** Obstructive sleep apnea significantly reduces cerebral compliance, with the duration and severity of respiratory events being key factors. Further studies with larger samples are needed to validate these results, better understand the underlying mechanisms and highlight the importance of early diagnosis and treatment of OSAS.

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Impacts of dysarthria on quality of life in Parkinson's disease

Ana Luiza Fabris Moraes[1], Arthur Jun Deguti Hata[1], Vivian Kaori Segawa[1], Emerson Gisoldi[1], Alice Estevo Dias[1], Margarete Jesus Carvalho[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Ana Luiza Fabris Moraes; Email: ana.moraes@aluno.fmabc.net.

Background: Idiopathic Parkinson's disease (IPD) is a chronic, slowly progressive neurodegenerative disorder characterized by plastic rigidity, resting tremor, bradykinesia, and impairment of automatic movements. Dysarthria is one of the most common clinical manifestations in IPD, significantly impacting verbal communication and quality of life. **Objective:** To identify communication difficulties and establish correlations with quality of life in IPD patients. **Methods:** This observational, cross-sectional study was conducted using the "Living with Dysarthria (LwD)" questionnaire in IPD patients, consisting of 10 questions, each with 5 sub-items and 6 response options: 1 (strongly disagree), 2 (disagree quite a bit), 3 (disagree a little), 4 (agree a little), 5 (agree quite a bit), and 6 (strongly agree). All patients were assessed using the Hoehn & Yahr and Unified Parkinson's Disease Rating Scale (UPDRS) parts I, II, III, IV, and V. Pearson's correlation was used to evaluate the relationship between these scales and LwD. **Results:** Responses from twenty-five patients, fourteen women (56%) and eleven men (44%), aged 40 to 80 years, were analyzed. Patients with dysarthria exhibited various communication difficulties: fourteen (56%) had noticeably slower and more slurred speech compared to their pre-disease condition, nine (36%) frequently avoided conversations and had difficulty communicating with family members, eleven (44%) reported a negative impact on self-esteem, sixteen (64%) needed to repeat themselves in conversations, and five (2%) felt treated differently by the people they communicated with. The coefficients found indicated a statistically significant correlation only between the UPDRS scale and the LwD. **Conclusion:** The LwD questionnaire proved effective in analyzing speech difficulties in IPD patients. Dysarthria showed a close correlation with individuals' quality of life, with greater severity of the condition being directly linked to a significant reduction in social well-being. The significant correlation between UPDRS and LwD suggests that worsening symptoms were associated with poorer quality of life. However, this was not the case between the LwD and H&Y, which may indicate that this scale does not capture as well as the UPDRS, regarding the severity of the disease, the quality of life aspects reflected in the questionnaire scores. Finally, the findings highlight the essential need for multidisciplinary intervention, especially speech therapy, in the treatment of this population.

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Impacts of hepatic encephalopathy on transplant patients: a systematic review

Loyslene Maria de Paula Bonifácio[1], Leandro de Souza Cruz[1]

[1]Faculdade Suprema, Juiz de Fora MG, Brazil.

Correspondence: Loyslene Maria de Paula Bonifácio; Email: loyslene.med@gmail.com.

Background: Liver cirrhosis is a chronic disease with high morbidity and mortality rates, and its definitive treatment is liver transplantation. Liver transplantation affects the patient's entire organism and functionality and can lead to severe complications. Hepatic encephalopathy is a brain dysfunction that presents as a neurological complication of liver disease and is associated with high re-hospitalization and morbidity rates in patients. **Objective:** To evaluate the impact of hepatic encephalopathy on transplant patients through a systematic review. **Methods:** Randomized controlled trials published in English over the past decade were analyzed, focusing on human subjects aged 18 and older. Data sources included MedLine and SciELO. The search utilized terms from the Health Sciences Descriptors (DeCS), specifically: hepatic encephalopathy, liver transplantation, and cirrhosis. Studies involving patients who presented with hepatic encephalopathy during the liver transplantation process were included, while those with unclear methodologies, available only in abstract form, or where encephalopathy was unrelated to liver pathology were excluded. The PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) scale was used to enhance the reporting quality of this review. **Results:** Three studies were analyzed, encompassing 477 adult participants with hepatic encephalopathy at some stage of liver transplantation. The effects of hepatic encephalopathy on morbidity, mortality, and neurological functionality during the transplantation process were observed. Statistical tests with a p-value < 0.05 were considered significant. The main symptoms of hepatic encephalopathy are psychomotor deficits, mental confusion, and complications such as intracranial hypertension, edema, and coma. Pre-transplant hepatic encephalopathy directly impacts post-transplant morbidity and mortality and affects the patient's score on the transplant list. Post-transplant hepatic encephalopathy can have multiple causes, such as organ rejection, sepsis, or the neurotoxic effects of immunosuppressants, which increase hospitalization time and impair neurological recovery. **Conclusion:** Hepatic encephalopathy is a significant complication of liver disease, placing a heavy burden on patients, families, and the healthcare system. It is an emergency condition that leads to high re-hospitalization and morbidity and mortality rates, necessitating early diagnosis and management due to its severity. However, further randomized controlled trials are needed to consolidate these observations.

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Impacts of maternal immune activation on neuroinflammation in adult offspring after sepsis: an experimental study

Anita Dal Bó Tiscoski[1], Fernanda Gava[1], Maria Amélia Cechinel da Silva[1], Lucinéia Gainski Danielski[1], Khiany Mathias[2], Larissa Joaquim[2], David dos Santos[1], Richard Simon Machado[2], Solange Stork[2], Fabricia Petronilho[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.

Correspondence: Anita Dal Bó Prudêncio da Silva Tiscoski; Email: anitadalbotiscoski@gmail.com.

Background: Sepsis is considered an infection associated with organic dysfunction resulting from the body's unregulated response. Experts estimate that sepsis globally contributes to approximately 11 million deaths annually. It is known that in sepsis, the exacerbated production of pro-inflammatory mediators amplifies neurological damage, and that an infectious challenge in childhood can program immune responses throughout life. **Objective:** Therefore, the goal of this study was to find out what effect giving lipopolysaccharide (LPS) during pregnancy had on brain dysfunction in the children of males and females who had severe polymicrobial sepsis as adults. **Methods:** Wistar rats were divided into two groups: saline and LPS, with LPS (100 μg/kg) administered in a single dose on gestational day 9.5, intraperitoneally (i.p.). The offspring of males and females (60 days) were subjected to sepsis by cecal ligation and perforation (CLP) and divided into groups: 1) saline+sham, 2) LPS+sham, 3) saline+CLP, and 4) LPS+CLP. After CLP, survival was assessed, and on the 10th day after, behavioral tests of habituation to the open field and forced swimming were carried out. Then, the cortex, prefrontal cortex, and hippocampus were isolated to evaluate levels of interleukins (IL) 6 and 10. **Results:** The Saline+CLP group reduced survival, in addition to inducing damage to habituation memory in both sexes and depressive-like behavior in females, which was prevented by preconditioning to LPS. Furthermore, it

induced an increase in IL-6 levels in the prefrontal cortex and hippocampus in both sexes, and LPS preconditioning prevented this increase only in the hippocampus of males. In addition to decreasing IL-10 levels in the prefrontal cortex and hippocampus in both sexes, preconditioning to LPS prevented this decrease in both structures in males. **Conclusion:** The present study concluded that preconditioning to LPS during pregnancy protected the offspring from a serious infectious challenge in adulthood.

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Impacts of non-prescribed use of methylphenidate by university students: a systematic review

Émile de Carvalho Morais Fraga[1], Giovana Andrade Oliveira[1], Gustavo Alves Cangussú[1], Luídi Neves Nascimento[1], Marcus Vinícius Silva Souza[1], Naila Maria Magalhães Silva[1], Priscila Santos Oliveira[1], Emmanuel Santos Trindade[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Priscila Santos Oliveira; Email: priscilaoliveira35001@gmail.com.

Background: Currently, living in a social context marked by the demand for constant high performance in academic routine, especially in the university environment, associated with the presence in environments that insist on maintaining a parameter of competitiveness, has led students to use psycho-stimulant medications without prescription, such as methylphenidate, used in the treatment of ADHD and narcolepsy. **Objective:** To conduct a systematic review regarding the impacts of non-prescribed use of methylphenidate by university students over the past fourteen years (2011-2024). **Methods:** This is a systematic literature review conducted through articles published in the PubMed and Scielo databases. Publications from 2011 to 2024 were considered, in both English and Portuguese, using the MeSH descriptors "methylphenidate" and "students". Included studies were both observational and analytical, provided they assessed the use of the psycho-stimulant methylphenidate as a performance enhancer in university students rather than for clinical purposes as indicated in the package insert. Exclusion criteria were applied to studies not associated with these criteria. **Results:** 177 articles were found, of which 10 were selected after reading. The analyzed studies revealed that the indiscriminate use of methylphenidate in the academic environment mostly occurs through acquisition from illicit sources, such as peers (56%). However, there is no evidence of significant benefits in the use of this psychostimulant in individuals without a diagnosis justifying therapeutic intervention, with a higher correlation with adverse effects and academic impairment. Individuals who used this drug had a lower mean academic index (7.92) compared to academics who had never used the psycho-stimulant (8.80), suggesting ineffectiveness in enhancing cognitive performance for these students. This low efficacy of methylphenidate may stem from its adverse reactions, such as anxiety (76.7%), tachycardia (40%), rebound effect (30%), fatigue (30%), appetite loss (26.7%), and nausea (13.3%), as well as from incorrect dosing, as in the absence of proper medical prescription, most individuals use sub-doses (12-20mg), insufficient to generate the expected effects of the drug. Among the scarce documented benefits are improved reasoning, memory, and well-being. However, this latter is also related to drug dependence and abuse of other substances. **Conclusion:** The review highlights the dangers of non-prescribed use of methylphenidate by students, emphasizing the academic harm and associated adverse effects. Promoting awareness and implementing control policies are crucial measures to protect student health and face the challenges of this practice in the academic environment, where performance pressure and competitiveness may drive the use of psycho-stimulant medications.

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Impacts of the COVID-19 pandemic on the epidemiological profile of stroke in Brazil between 2016 and 2023

Marcelo Pereira de Souza[1], Arthur Faria Jeveaux[1], Clara de Avelar Portela[1], Érika Yumi Mandú[1], Fábio Eiti Mandú[1], Marçal Pizzico[1], Marinalva Pereira dos Santos[1]

[1]Universidade Nove de Julho, Osasco SP, Brazil.

Correspondence: Marcelo Pereira de Souza; Email: marsouzz@hotmail.com.

Background: Stroke is a medical condition that occurs when there is interruption of blood flow to a part of the brain, causing a lack of oxygen and nutrients in this region. Data were collected during and after the COVID-19 pandemic, from January 2020 to December 2023, and it was observed that this condition was responsible for 696,233 hospitalizations and 105,083 deaths. On the other hand, in the pre-pandemic period, between January 2016 and December 2019, there were 621,900 hospitalizations and 96,127 deaths. From this perspective, there is an 11.97% increase in the number of hospitalizations and, approximately, a 9.31% increase in the number of deaths. Based on this result, we see the importance of a well prepared team to recognize these events and treat patients. **Objective:** To analyze the Brazilian epidemiological profile in relation to the increase in the number of stroke cases after the start of the COVID-19 pandemic. **Methods:** Ecological, quantitative and descriptive study, whose data were obtained through consultations carried out in the Hospital Information System (SIH/SUS), through the DATASUS platform, referring to the period from January 2016 to December 2023. Data on hospitalizations and deaths in Brazil were analyzed and compared in the period before and after the COVID-19 pandemic. **Results:** In relation to the national analysis, this condition is present at all ages, with a greater distribution among women aged 0 to 49 years, but in the age range 50 to 79 years it is more prevalent among men. It was observed that, from 2016 to 2019, the number of hospitalizations of women between 0 and 49 years old was 39,644 and the number of deaths was 3,879, between 2020 and 2023, these numbers increased to 44,010 and 4,139, respectively. Among men, aged 50 to 79, the numbers increased from 233,521 hospitalizations and 32,239 deaths between 2016 and 2019 to 265,231 and 36,125, respectively, between 2020 and 2023. In the general overview, in the period between January 2016 and December 2019, there were 621,900 hospitalizations and 96,127 deaths, but, between January 2020 and December 2023, these numbers increased by 11.97% in the hospitalization rate and 9.31% in the of deaths, resulting in 696,233 hospitalizations and 105,083 deaths. **Conclusion:** Based on the analysis of these data, there was a significant increase in hospitalizations and deaths due to stroke, especially in men aged 50 to 79 years, in Brazil, during the pandemic period.

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Implications of detecting clinical manifestations of dysphagia in Parkinson's disease

Vivian Kaori Segawa[1], Arthur Jun Deguti Hata[1], Ana Luiza Fabris Moraes[1], Margarete Jesus Carvalho[1], Alice Estevo Dias[1], Emerson Gisoldi[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Vivian Kaori Segawa; Email: viviankaorisegawa@gmail.com.

Background: Parkinson's disease (PD) is the second most common neurodegenerative disease in the world and mainly affects the elderly. Dysphagia is one of the most prevalent clinical manifestations (70-100%). **Objective:** To detect the manifestations of dysphagia in patients with PD. **Methods:** An observational, cross-sectional study was carried out using the Dysphagia Detection Questionnaire for Patients with Parkinson's Disease (SDQ-DP), made up of 15 questions about swallowing alterations, 13 of which required the patient to mark the frequency of symptoms from 0-3, 0 being never and 3 being very frequent, and 2 of which required a yes or no answer. The instrument's cut-off point is 11 points; a score equal to or greater than this value indicates a risk of dysphagia. In order to identify the clinical condition of patients with PD, the Unified Parkinson's Disease Rating Scale (UPDRS) parts I, II, III, IV and V and the Hoehn & Yahr (H&Y) scale were used; the statistical test used was Pearson's sample correlation. **Results:** 25 PD patients aged between 40 and 80 years participated, 14 patients (56%) were women and 11 patients (44%) were men. The moderate correlation between UPDRS and SQD-DP (p=0.44) indicates that there is a relationship between the severity of the aspects investigated by the scale and the severity of the statements contained in the evaluation instrument. The weak correlation between H&Y and SDQ-DP (p=0.25) indicates that the H&Y scale does not fully cover the level of disease involvement and its clinical manifestations. It was found that 3 patients (12%) reported food/liquid coming out of their nose after eating/drinking; 5 patients (20%) reported difficulty breathing during meals; 6 patients (24%) reported the presence of leftover food in their mouth after swallowing and that chewed food escaped out of their mouth, and they had difficulty swallowing pasty food; 7 patients (28%) coughed while swallowing solids and felt their voice change after swallowing food or liquids; 8 patients (32%) reported food stuck in their throat while eating; 9 patients (36%) tried to swallow chewed food several times, had difficulty swallowing solids, reported coughing while swallowing liquids and difficulty swallowing saliva outside of meals; 10 patients (40%) noticed an increase in saliva in the mouth and 11 patients (44%) had difficulty chewing solids. There were 19 scores (76%) below 11, representing a low risk of dysphagia, and 6 scores (24%) equal to or greater than 11, with the highest score being 30. **Conclusion:** This study exposes the main complaints and complications related to dysphagia in patients with PD and how they negatively affect quality of life through limiting and uncomfortable aspects. The SDQ-DP proved to be useful in instituting appropriate and early multidisciplinary treatment in order to avoid complications and ensure an improvement in patients' quality of life.

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Importance of the neurological examination: delayed diagnosis of tropical spastic paraparesis with atypical evolution in a young adult

Vinícius Andreata Brandão[1], Vinícius de Pádua Sanders Medeiros[1], Betina Bolina Kersanach[1], Gabriel Andreata Brandão[2], Jovana Gobbi Marchesi Ciríaco[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.
[2]Escola Superior de Ciências da Santa Casa de Misericórdia de Vitória, Vitória ES, Brazil.

Correspondence: Vinícius Andreata Brandão; Email: viniciusandreatab@gmail.com.

Case presentation: Female, 24 years old, from the south of Bahia, seeks orthopedic service complaining of mobility difficulties. At the age of 13, she began to have a progressive mobilizing difficulty of her right lower limb, and after a few years, both limbs were injured, with gradual evolution associated with urinary retention and constipation. She denied pain, paresthesia or anesthesia. The patient was born by vaginal delivery at eight months

and she was exclusively breastfed until seven months of age. Her cognitive, motor and gait development were unchanged. No comorbidities and updated vaccinations. Cardiovascular, respiratory and abdominal examinations were unchanged. The neurological assessment showed grade 2 proximal strength in the right lower limb, grade 3 strength in the left lower limb and grade 5 strength in the upper limbs. Osteotendinous reflexes 4+ globally. Patellar clonus 1+ and mandibular clonus 2+. Plantar cutaneous reflex in bilateral extension. Hofmann and Trömner present bilaterally. Cognitive, cranial nerves and sensitivity assessments were unchanged. Due to the findings of the physical examination, the patient was referred to the neurology department. The patient had an electroneuromyography suggesting paraparesis of the lower limbs, compatible with spinal cord involvement at the lower thoracic level. A magnetic resonance imaging of the neuroaxis revealed atrophy of the thoracic spine, corroborating the diagnosis of spastic paraparesis. The hypotheses of sporadic or tropical spastic paraparesis, caused by HTLV virus infection, were put forward, and then confirmed later by serology for the virus. The treatment was started with botulinum toxin and physiotherapy, with symptom improvement. **Discussion:** Type 1 Human T-cell lymphotropic virus (HTLV-1) associated myelopathy or tropical spastic paraparesis is a severe neurological disease that usually affects adults between the ages of 30 and 50. It leads to chronic and progressive degeneration of upper motoneurons in the Central Nervous System, which can lead to severe motor disability. It is expected that, after 10 years of progression of the disease, 50% of patients will need a wheelchair, while the others will need other devices to help them walk. In this summary, we present a case of an 11-year evolution tropical spastic paraparesis in a 24-years-old patient from an endemic area for HTLV-1. It is important, therefore, to highlight the onset of the condition in a young patient, with 11 years of evolution, who does not depend on a wheelchair for locomotion. In addition, the diagnosis was suspected after a complete neurological physical assessment, highlighting its importance. **Final comments:** In this paper, we reinforce the importance of routine neurological physical examination, especially in patients from an endemic area for HTLV-1, since the findings in the case presented were essential for the patient to be referred to the appropriate service

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Improvement clinic to an unusual drug treatment in short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA): a case report

Leonardo Monteiro Lauria[1], Pedro Augusto Sampaio Rocha Filho[2], Alex de Novais Batista[1], Iago César Lopes Tenório de Cerqueira[1], João Herculano Lins[1], Maria Clara Arcoverde Santana[1], Renato Maciel Freire de Paula e Silva[1], Virgílio Sátyro da Nóbrega Segundo[1], Caio César Alves Lins de Oliveira[1], Lucas de Souza Pessoa[1]

[1]Hospital Universitário Oswaldo Cruz, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Departamento de Neuropsiquiatria, Recife PE, Brazil.

Correspondence: Leonardo Monteiro Lauria; Email: leonardolauria09@gmail.com.

Case presentation: Woman, 60 years-old, reports having right periorbital pain for 1 year, with shock. The pain episodes lasted 3 minutes, were associated with tearing ipsilateral to the pain, and had a frequency of 5 to 10 attacks per day. Furthermore, touch the right face and water in the bath were reported as triggers. MRI and contrast-enhanced cranial MRI angiography showed no significant abnormalities. Also, she used carbamazepine 200 mg/day for 3 months, however, as there was no improvement in pain, this was replaced by valproic acid (250 mg 12/12 h). With this drug, she remained pain free for 2 years and 3 months. After this period, the drug was suspended. Since then, the patient has progressed for over a year without new attacks. **Discussion:** SUNCT (short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing) and SUNA (short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms) are trigemino-autonomic headaches characterized by ultra-rapid attacks that last from 1 to 600 seconds and differ from each other in that SUNA does not present conjunctival hyperemia and tearing at the same time. There are three first-line prophylactic treatment options for these headaches: lamotrigine, topiramate and gabapentin. Our patient evolved with an excellent drug response to valproic acid. Only 6 patients with SUNCT had previously reported improvement with this drug. Our case of SUNA may have been one of the few described with improvement in the patient's pain with the use of valproic acid. **Final comments:** SUNCT (short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing) and SUNA (short-lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms) are headaches traditionally described as difficult to treat. Although valproic acid is not among the first-line medications for the treatment of these headaches, the excellent response in our case to this drug contributes as evidence to consider this drug as a therapy.

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Improvement of neuropathic pain related to spinal cord injury using combined transcranial direct current stimulation and neuromuscular electrical stimulation: a case report

Cezar Sabino Pereira da Silva[1], Ana Clara Dias Pereira[2], Vívian Santos Xavier Silva[1], Jean Marcos de Souza[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Cezar Sabino Pereira da Silva; Email: cezarsabinop@gmail.com.

Case presentation: A 32 Y.O. male with neuropathic pain from a T12/L1 traumatic spinal cord injury (SCI) since 2019 underwent a weekly protocol of combined transcranial Direct Current stimulation (tDCS) at 2mA targeting the motor precentral region and 20 minutes of functional electrical stimulation of the bilateral femoral quadriceps. Initial comprehensive assessments of pain and quality of life were conducted using the Neuropathic Pain Symptoms Inventory (NPSI), McGill Pain Questionnaire (MPQ), Pain Quality Assessment Questionnaire (PQAS), Short-form Health Survey-36 (SF36), and Quality of Life Index Spinal Cord Injury III (SCI III). The American Spinal Cord Injury Association Impairment Scale (AIS) evaluated his spinal cord status. After 12 weeks, these assessments were repeated. The study was approved by the institution's Ethics Committee, and the patient provided written consent. By the end, AIS level and classification remained unchanged, and no side effects were reported. There was a reduction in deep and superficial pain, with evoked and continuous pain remaining stable. Paroxysmal pain showed contradictory results between PQAS and NPSI, precluding definitive conclusions. MPQ showed a 50% reduction in the affective component of pain. Significant improvements in quality of life and functionality metrics were observed in SCI III, along with enhancements in all dimensions of SF-36, some of which were notably enhanced. The questionnaire revealed significant improvements in vitality, sociability, physical condition, and pain, enhancing daily performance and capabilities. Pain characteristics became more benign, with alterations in type, and both recurrence and duration of pain decreased. **Discussion:** Neuropathic pain is a common issue in individuals with SCI, resulting from damage to the somatosensory system leading to neuronal hyperexcitability. Drug-based treatments for neuropathic pain tend not to have satisfactory effects. The intervention reduced some somatosensory pain features while others remained stable. However, the substantial improvement in quality of life suggests an objective decrease in pain through indirect measures, which may be more practical than analog scales. The neuropathic pathophysiology of pain in SCI involves central sensitization, which tDCS may modulate, partially explaining the results. Since chronic pain impacts quality of life through negative emotions, this case report suggests that combined stimulation may modulate the emotional response to pain, not just its sensory characteristics. The decreased recurrence and duration of pain may be related to the patient's improved well-being. **Final comments:** The therapy was safe and feasible, resulting in substantial improvements in quality of life. Therefore, this approach could fill a gap in patient care, given the lack of effective drug-based treatments for neuropathic pain. This case report highlights the importance of future experimental trials with combined neurostimulation.

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Impulse control disorder in patients with Parkinson's disease: a life quality analysis

Ayla Morais Vitório Guerra[1], Gabriel Souza Medrado Nunes[1], Maria Eduarda Messias Vassoler[1], Fernanda Garcia Silva Pereira Nascimento[1], Caio Fontes do Nascimento[1], Gabriel Teles de Oliveira Piñeiro[1], Roberta Borges Gomes Kauark[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Ayla Morais Vitório Guerra; Email: aylaguerra1@hotmail.com.

Background: The therapeutic use of dopaminergic agonists for Parkinson's disease (PD) treatment may cause, as an adverse effect, impulse control disorder (ICD), where the desire to engage in certain activities is independent of social, physical or psychological harm. It can lead to financial difficulties, relationship problems, mental health problems and even legal problems. **Objective:** To describe the frequency of ICD in PD patients and analyze their quality of life (QOL). **Methods:** Cross-sectional study carried out between 2023-2024, patients who responded positively to at least one question on the Questionnaire for Impulsive-Compulsive Disorders in Parkinson's Disease Rating Scale (QUIP-CS), indicating the presence of CDI. Clinical, social and epidemiological data were collected, in addition to the application of the following scales: Parkinson's Disease Questionnaire (PDQ-39), Unified Parkinson's Disease Rating Scale-Part III (UPDRS-III), Mini-Mental State Examination (MMSE), Beck Depression Inventory (BDI), Non-Motor Symptoms Scale (NMSS), and the Hoehn and Yahr scale (HY). **Results:** Sample consisted of 25 PD patients, 72% male, with a mean age of 65.4 years (±8.3) and a mean disease duration of 11.8 years (±3.7). Types of ICD identified included pathological gambling (16%), excessive purchases (16%), hypersexuality (24%), binge eating (20%) excessive medication use (16%) and other behaviors (44%). More than one compulsive behavior were found in 24% and 52% reported that the compulsive disorder had no negative impact on their lives. The median Levodopa Equivalent Daily Dose (LEDD) with 1100mg (450-2422). QOL, measured by the PDQ-39, had a mean total score of 44.2 (±5.5). Severity of motor symptoms, assessed by the UPDRS-III, had a mean of 44.6 (±17.8), we consider a moderate value between 25-50. The MMSE revealed a median of 26 (9-30). Depression assessment, conducted using the BDI, indicated a mean of 15.6 (±10.1). The mean sum of non-motor symptoms, evaluated by the Scales for NMSS, was 92.6 (±52.7) and 56% of participants have disability level 2 on HY scale. Types of ICD didn't demonstrate a difference in impact on QOL.

Bivariate tests showed a positive association between the total PDQ-39 and the sum of NMSS (r=0.67, p=0.00). A moderate correlation was also observed between UPDRS-III and the total PDQ-39 (r=0.54, p=0.05). Additionally, the correlation between the Beck Depression Inventory and the total PDQ-39 was moderate (r=0.49, p=0.01), and the difference in HY is associated with differences in QOL, with post hoc analysis indicating that HY 4 is associated with worse QOL compared to HY 2 (p=0.048). **Conclusion:** ICD has a significant multidimensional impact on the QOL of participants, highlighting the need for comprehensive therapeutic interventions that address the physical and psychosocial aspects of the disease to improve the overall QOL of these patients.

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In addition to Parkinson's disease, a different presentation of the LRRK2 mutation

Sephora Sabrina Candido de Almeida[1], Ana Carolina Gomes[1], Yngrid Dieguez Ferreira[1], Alexandre Shoji[1], Isabela Martins de Freitas[1], Diogo Haddad Santos[1]

[1]Irmandade Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Sephora Sabrina Candido de Almeida; Email: sephora_sabrina@yahoo.com.br.

Case presentation: 58 years old, female, previously healthy , housewife, started experiencing changes in mood and behavior 5 years ago, repeating the same activity several times and becoming disoriented in familiar places. Within 2 years, the patient developed complete loss of self-care capacity. After another year, he presented her first seizure and progressively began to have difficulty with language (naming and repetition). In the mini mental state examination (MMSE) in August 2022, the patient scored 11 (loss in memory, attention, evocation and language) with clinical dementia rating scale (CDR) 1. In April 2024, patient unable to perform MMSE, with CDR 3. Imaging examination (magnetic resonance imaging of the skull) at the beginning of the condition (2021) without changes. After follow-up and exclusion of common causes of dementia syndromes, it was decided to request genetic testing considering a young patient with a progressive condition without other examination findings. LRRK2 gene found – AUTOSOMAL DOMINANT PARKINSON DISEASE. Patient remains under outpatient follow-up. **Discussion:** This case report describes a patient with a mutation in the LRRK2 gene, who presents with a dementia syndrome, but without symptoms of parkinsonism. This clinical presentation is atypical, given that mutations in the LRRK2 gene are most frequently associated with late-onset Parkinson's disease (PD). The absence of parkinsonian symptoms and the isolated presence of dementia in this patient raise important questions about the phenotypic variability associated with LRRK2 mutations and the possible mechanisms underlying the observed cognitive decline. Mutations in LRRK2, especially G2019S, are well documented to cause PD with significant phenotypic variability. Previous studies have demonstrated that, in addition to the typical motor symptoms of PD, some individuals with LRRK2 mutations also experience non-motor symptoms, including cognitive and psychiatric disorders. However, cases of isolated dementia without parkinsonism are rarely reported. The absence of parkinsonian symptoms in a patient with a LRRK2 mutation is unusual but not unprecedented. Isolated case studies indicate that the phenotypic expression of LRRK2 mutations may be influenced by additional genetic factors, epigenetic modifiers, and environmental factors. These cases reinforce the need to consider a broad approach when evaluating the symptoms and prognosis of LRRK2 mutation carriers. **Final comments:** The present case report contributes to the understanding of the phenotypic variability of LRRK2 mutations, highlighting the possibility of an isolated presentation of dementia. This case reinforces the need for further research to elucidate the underlying mechanisms and to develop targeted therapeutic strategies.

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In silico study for optimizing magneto hyperthermia for cancer treatment using the glioblastoma-on-a-chip model, comparing heat dissipation in tumoral versus healthy vascular networks

Giovana Fontanella Pileggi[1], Juan Munoz[1], Mariana Penteado Nucci[2], Arielly da Hora Alves[1], Flavia Maria Morais Pedrini[1], Nicole M. E. Valle[1], Javier Bustamante Mamani[1], Marcelo N. P. Carreño[3], Fernando A. Oliveira[1], Lionel F. Gamarra[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Faculdade Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[3]Universidade de São Paulo, Escola Politécnica, Departamento de Engenharia de Sistema Eletrônicos, São Paulo SP, Brazil.

Correspondence: Giovana Fontanella Pileggi; Email: giovana.pileggi@gmail.com.

Glioblastoma multiforme (GBM) is the most frequent brain cancer in adults and remains incurable despite advancements in cancer therapeutics. The intense angiogenesis in GBM creates a chaotic blood vessel network, contrasting with the organized structure of healthy tissue, contributing to its resistance to conventional therapies. To enhance capacity to treat patients, different therapeutic approaches are required. Among them is magneto hyperthermia (MHT), where magnetic nanoparticles are introduced into the tumor and an alternating magnetic field is applied resulting in localized heating that selectively targets tumor cells at 42°. Preclinical investigation of MHT therapy is crucial, and with the organ-on-a-chip device it is possible to overcome the limitations of in vivo models and replicate the physiological microenvironment of tissues and organs. In silico studies are essential in developing microfluidic devices, addressing the complexity of biological systems by utilizing simulations that predict treatment dynamics, and further investigate the contribution of tumor vascular networks to heat dissipation. This work aimed to analyze the efficiency of heat dissipation in vascular networks under MHT, comparing healthy and tumorous tissue networks. We created two chip designs, based on healthy and tumorous vascularization, grounded on studies of GBM vessels microstructure. The COMSOL Multiphysics software was used to simulate conditions in microchips and analyze the thermal behavior in both geometries.The flow studies analyzed temperature variations over time, mostly focusing on the impact of flow rate variations (0, 1, and 100 μL/min) and their effects on fluid dynamics parameters. Furthermore, we investigated the influence of nanoparticle concentration, from 0.25 to1mg/mL, on temperature distributions over time. Regarding maximum velocity, the healthy design had 4.5 times faster flow at μL/min and 10 times at 100 μL/min than the complex model. In temperature analyses at 0μL/min, no differences were noted between the designs, yet a higher heat concentration was observed compared to the 1 and 100μL/min models. Furthermore, flow analyses revealed a greater heat concentration in the tumorous design, reaching a difference of 4°C, indicating less efficient heat dissipation under irregular pathways. Additionally, higher nanoparticle concentrations led to a linear increase in temperature, suggesting that the tumorous system retains greater heat concentration and reaches higher temperatures within the same timeframe, with a maximum difference of 3.92°C at 0.5mg/mL concentration. This study introduces an innovative microfluidic device for assessing thermal tumor therapies, which allowed the assessment of the considerable influence of vascular architecture, fluid flow, and nanoparticle concentration on the effectiveness of MHT therapy on in vitro studies. Representing a notable advancement in the field of thermal therapy for brain tumors and beyond.

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Inborn errors of immunity: neurological phenotypes, clinical features and genetics of a cohort of 27 patients

Renan Brandão Rambaldi Cavalheiro[1], Kaliny Oliveira Peixoto[1], João Paulo Barile[1], Patrícia Marques Mendes[1], Igor Braga Farias[1] , Paulo de Lima Serrano[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Renan Brandão Rambaldi Cavalheiro; Email: renan.rambaldi@icloud.com.

Background: The inborn errors of immunity (IEI) are a heterogeneous group of monogenic conditions that encompass autoinflammatory syndromes, complement deficiencies, diseases of immune dysregulation and defects of intrinsic and innate immunity and one marrow failure. While individually these conditions can be considered rare, ultra rare (less than 1 case per million people) and some with single-digit case reports, they are common as a group and can present with virtually any constellation of signs and symptoms. A striking feature of some IEIs is the potential of secondary autoimmunity. While IEI are considered multisystem disorders with well described features, such as periodic fevers and arthralgia, there is an increasing body of evidence of isolated neurological presentations, that can be either the sole manifestation of some of these conditions or precede systemic involvement. The recognition of those syndromes is vital, as they are potentially treatable, such as the use of IL-1 inhibitors in TNF receptor-associated periodic syndrome (TRAPS) or the use of TNF blockers in deficiency of adenosine deaminase 2 (DADA2). **Objective:** Describe the demographics, clinical presentation and genetics of a cohort of 27 patients with inborn errors of immunity from an outpatient neuromuscular clinic. **Results:** Of the 27 patients with genetically confirmed IEIs, age at presentation varied from 1 year of age to 62 years of age. 18 patients had a final diagnosis of an autoinflammatory syndrome: 6 patients with Aicardi-Goutières syndrome (AGS), 3 patients with TRAPS, 3 patients with cryopyrin-associated periodic fever syndrome (CAPS), 2 patients with NOD2- related autoinflammatory syndromes. Their phenotypes ranged from pure hypotonia (floppy infant syndrome) to a classic autoinflammatory phenotype with additional neurological findings (dysautonomia, polyneuropathy) and leukodystrophic changes in the AGS patients. 9 patients presented with individually ultra-rare immune dysregulation genes that, while producing immune deficiencies also caused secondary autoimmunity. These patients presented with recurrent sinopulmonary infections and transient arthralgias. The most common secondary autoimmune disorders observed were myasthenia gravis, observed in 4 patients, and stiff-person syndrome (SPS), observed in 3 patients. **Conclusion:** While rare, inborn errors of immunity need to be considered in the differential diagnosis of many neurological conditions, as their presentation can vary substantially. The present data underscore the importance of careful phenotyping of any "outlier" clinical feature, as it may be the protean manifestation of a much rarer disease, and may point the neurologist to a treatable condition.

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Incidence and mortality of congenital malformations of the nervoussysteminbrazil in children under 1 year of age

Emanuel Henrique dos Santos[1], Gabriel Cordova[1], Izabelly Constatinov Lemos[1], Anne Beatriz Ricardo Amaral[1], Kauan Correia Derbli[1], Julia Farto Viana Oliveira[2], Sofia Beatriz Andrade Manfio[1], Gabriel Andre Silverio[1]

[1]Centro Universitario Campo Real, Guarapuava PR, Brazil.
[2]Universidade Estadual do Centro Oeste, Guarapuava PR, Brazil.

Correspondence: Emanuel Henrique dos Santos; Email: hds.emanuel@gmail.com.

Background: Congenital malformations are alterations in the embryonic process. In Brazil, the main ones are congenital heart malformations, Down syndrome, and Neural Tube Defects (NTDs). This study aims to analyze malformations of the nervous system. **Objective:** To expose the prevalence of sex in the incidence of hospitalizations and mortality from these malformations in Brazil. **Methods:** An epidemiological study was conducted using the DATASUS database, focusing on the Hospital Morbidity database of the Unified Health System (SIH/SUS) and Live Births (SINASC). Data collection was carried out from 2019 to 2022, with records of hospitalizations per period and deaths by age group. The variables analyzed were sex and age. The applied ICD was Spina Bifida and Malformations of the Nervous System, with analysis restricted to children under 1 year. Incidence was calculated using the formula: incidence rate = number of cases of the disease in the period / number of people exposed to the risk of acquiring the disease in the period (live births) x 10^5, while mortality followed the standard formula, also with exponent 5. **Results:** The total incidence of hospitalizations was 102.85 per 100,000 inhabitants, with female predominance (104.25) compared to male (101.50). Regarding mortality, females also had higher numbers than males, with 8.25x6.97 deaths per 100,000 inhabitants. Over the analyzed period, the incidence of hospitalizations per one hundred thousand newborns was higher for females in the first two years, becoming higher for males in the following two years: 95.24 for males and 100.39 for females in 2019; 100.09 for males and 100.39 for females in 2020; 108.06 for males and 106.57 for females in 2021; 103.15 for males and 99.61 for females in 2022. However, mortality per one hundred thousand newborns remained higher for females throughout the period: 7.20 for males and 7.83 for females in 2019; 7.30 for males and 8.86 for females in 2020; 6.50 for males and 7.73 for females in 2021; 6.87 for males and 9.03 for females in 2022. **Conclusion:** Females present higher mortality, while the incidence of hospitalizations was higher for females in 2019 and 2020, and for males in 2021 and 2022. It is crucial to highlight the importance of prenatal care to raise awareness among patients about teratogenic substances and preventive measures for neurological malformations, such as gynecological emergencies that can affect blood supply to the fetus, and various infectious diseases. Further studies are needed not only to explore new ways of preventing malformations but also to ensure the physical and psychosocial well-being of patients, as provided by law.

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Incidence and predictors of late seizure after cerebral venous thrombosis: an ambispective cohort

Graziele Carolina Araújo Soares[1], Júlia Barreto de Farias[1], Pedro Fernandes Abbade[1], Lucy Rodrigues Ribeiro[1], Cárita Victória Carvalho de Santana[1], Isabella Reis Vieira[1], Israela Souza Brito Santos[1], Ana Beatriz Cazé Cerón[1], Bruna Guilherme Martendal[1], Jamary Oliveira-Filho[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Graziele Carolina Araújo Soares; Email: grazielemedicina@gmail.com.

Background: Cerebral venous thrombosis (CVT) is a rare cause of stroke with a high incidence of seizures, either acute (AS) or late (LS). The chance of recurrent seizure after an episode of CVT is about 11%, but there is a lack of Brazilian studies on the subject. **Objective:** To evaluate the frequency and predictors of LS after CVT. **Methods:** Ambispective cohort using medical records of consecutive individuals treated at a specialized outpatient center in Salvador-BA between 2008 and 2024. Those with a previous diagnosis of epilepsy and a follow-up time < 6 months were excluded. LS were defined as seizures occurring > 14 days after the diagnosis of CVT. To identify predictors of LS, we used multivariate Cox regression. **Results:** We included 64 participants with CVT, with a median follow-up time of 38 months (IQR: 18-81.5). Seizures were presented by 32 individuals (50%): 30 (47%) had AS and 16 (25%) had LS, and in the LS group, 88% (14/16) of the participants had AS. The baseline characteristics among participants with and without LS were similar, except for a higher frequency in the LS in afrodescendants (53% vs 17%, p=0.035), with hemorrhagic transformation (56% vs 23%, p=0.013), cortical infarction (56% vs 19%, p=0.009) and AS (88% vs 33%, p<0.001). The clinical manifestations of CVT were similar in LS and non-LS participants, except for a lower frequency in the LS group of headache (69% vs 91%, p=0.039) and nausea and vomiting (19% vs 49%, p=0.034). Participants with LS compared to participants without LS had a higher frequency of deep venous thrombosis (27% vs 6.3%, p=0.049) and protein C deficiency (33% vs 0%, p=0.017). In the survival analysis, we identified as multivariate predictors of LS: AS (HR=5.55; 95% CI, 1.05-29.21), use of dabigatran (HR=0.16 compared to warfarin; 95% CI, 0.03-0.81) and visual symptoms (HR=0.19; 95% CI, 0.04-0.89). **Conclusion:** In CVT, 25% of the participants had LS. AS were associated with increased frequency of LS, while dabigatran (vs. warfarin) use and visual symptoms were associated with decreased frequency of LS.

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Incidence and risk factors of viral neuroinfections in children in the Northern region of Brazil

Kayla Dalva Damasceno Bispo dos Santos[1], Fernanda Ferreira Fernandes[2], Norma Rafaella Uchôa Espíndola[3], Evelyn Farias de Oliveira[4], Giovanna Brandão Ferreira[5], Rebeca Leal da Cunha Torres[1], Marianna Leite[6], Amanda Curione de Castro[7]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil.
[2]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[3]Centro Universitário Maurício de Nassau, Recife PE, Brazil.
[4]Uninorte, Rio Branco AC, Brazil.
[5]IDOMED Citta, Rio de Janeiro RJ, Brazil.
[6]Faculdade Santa Marcelina, São Paulo SP, Brazil.
[7]Universidade Católica de Brasília, Brasília DF, Brazil.

Correspondence: Kayla Dalva Damasceno Bispo dos Santos; Email: Kayla.santos@ufpe.br.

Background: Viral neuroinfections present a significant public health challenge, especially in children, due to their potential severity and long-term complications. Factors such as environmental conditions, socioeconomic status, and healthcare access can influence their incidence, particularly in regions with distinct epidemiological profiles, as the Northern region of Brazil. **Objective:** To analyze pediatric hospitalizations in Brazil using data from the Hospital Information System (SIH/SUS) via the DATASUS platform. Employing a quantitative and descriptive approach, it examines variables such as sex, race/ethnicity, epidemiological profile, hospitalizations by state, endemic region, age group, and mortality rate. The goal is to identify patterns and trends to inform effective public health interventions and highlight areas requiring policy attention. **Methods:** This retrospective, quantitative, and descriptive study utilized secondary data from the Hospital Information System (SIH/SUS) via the DATASUS platform. The dataset spans from January 2018 to December 2023, encompassing individuals aged 0 to 14 years, and specifically focuses on hospitalizations within the Brazilian public health system (SUS). Descriptive statistical methods were employed to analyze variables such as the year of hospitalization, sex, race/ethnicity, hospitalizations by state, age group, and mortality rate. **Results:** From 2018 to 2023, in the Northern region, there were 1,031 hospitalizations for viral neuroinfections in children aged less than one to fourteen years old. Pará was the leading state with 391 (37.9%) admissions. From all the cases, 579 (56.2%) were male and 452 (43.8%) were female. The majority (590 or 57.2%) were patients of brown skin color. Acre had the highest mortality rate (5.77), followed by Pará (5.12), whereas Tocantins had the lowest (1.02). Most hospitalizations occurred in children aged 1 to 4 years (33.17%). **Conclusion:** This study provides insights on the incidence and risk factors of viral neuroinfections in Northern Brazil's pediatric population. The data revealed a significant burden of these infections, with Pará being the most affected state. The findings also highlight disparities in mortality rates among different states, underscoring the need for targeted interventions. Overall, this study underscores the importance of continuous surveillance and public health strategies to prevent and manage viral neuroinfections in children, particularly in regions with higher prevalence rates. Further research is warranted to explore the underlying factors contributing to these infections and to develop more effective prevention and treatment measures.

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Incidence of atrial fibrillation in patients with ischemic stroke due to large vessel occlusion undergoing chemical thrombolysis in a stroke reference unit in 2022

Paulo Victor Santos Machado[1], Frederico Otto Flores Moraes[1], Vitor Bonk Rizzo[1], Isadora Candotti Sperotto[1], Sarah Protásio Bonete[1], Julia Duarte Couto de Cristo[1], Felipe Bernardo Costa Ramos[1], Marcos Seefeld[1], Paola R. González[1], Lisseth F. V. Reyes[1]

[1]Hospital Universitário Evangélico Mackenzie, Curitiba PR, Brazil.

Correspondence: Paulo Victor Santos Machado; Email: paulovictorsmachado@gmail.com.

Background: Approximately 30% of ischemic strokes are caused by large vessel occlusion (LVO), characterized as occlusions of the proximal systemic arteries (M1), mid-branch losses (M2), basilar, or intracranial internal carotid arteries (ICA), resulting in greater morbidity and mortality when compared to an ischemic stroke involving smaller vessels. A well-established risk factor is atrial fibrillation (AF), with a doubled prevalence in patients with arrhythmia who have had LVO, when compared to other subtypes of ischemic stroke, with cardioembolism being the cause of almost half of the cases, with more than half of these attributed to AF. Despite this reality, 30% of cases are considered to have cryptogenic etiology, which is defined by criteria for embolic stroke of undetermined origin (ESUS). Initial studies suggest that empirical anticoagulation after ESUS is not superior to antiplatelet therapy, whereas the presence of AF after stroke typically requires anticoagulant treatment by risk classification calculation. Understanding that the identification of AF after stroke typically alters the treatment strategy for secondary stroke prevention, the use of extended cardiac monitoring for AF is therefore critical in the assessment of stroke ESUS. **Objective:** To analyze the incidence of atrial fibrillation and the epidemiological profile of patients who were chemically thrombolysed due to a stroke caused by large

vessel occlusion. **Methods:** Observational, cross-sectional and retrospective study from the year 2022, through analysis of medical records and complementary exams of patients who were treated in a stroke reference unit. **Results:** During the studied period, of the 35 patients with LVO, 12 (34.29%) had AF. The average age of these patients was 75.16 years (ranging from 65 to 92 years). The main occluded branch was the M1 portion of the middle cerebral artery, representing 66.67% of cases, followed by the M2 portion with 25% and the internal carotid artery with 8.33% of cases. **Conclusion:** The results are in line with the literature, with an incidence of approximately 33% of AF known at admission. The epidemiological data are also in agreement, with an average age of 75.16 years found in the present sample, in contrast to the average of 70 years in studies found in the literature. In relation to the affected arterial segment, the M1 portion was the main occluded branch (66.67% of cases in the study and 74% in the literature), with a divergence in relation to the frequency of the other segments, as the base article presents the carotid artery internal as the second most affected, but it occupies third place, after the M2 portion, in the present work.

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Incidence of hemorrhagic transformation in patients with ischemic stroke due to large vessel occlusion, undergoing chemical thrombolysis in a stroke unit in 2022

Paulo Victor Machado[1], Frederico Otto Flores Moraes[1], Vitor Bonk Rizzo[1], Nayara de Oliveira Dias[1], Samuel Marques Starneck Sathler[1], Joao Pedro Gorski Ribas de Araujo[1], Ana Paula G. Barankiewicz[1], Lucas Rodrigues Prim[1], Maria Julia Rodrigues[1], Marcos Seefeld[1]

[1]Hospital Universitário Evangélico Mackenzie, Curitiba PR, Brazil.

Correspondence: Paulo Victor Santos Machado; Email: paulovictorsmachado@gmail.com.

Background: Hemorrhagic transformation (HT) is a serious complication of ischemic stroke,, especially in patients with large vessel occlusion (LVO), being exacerbated by reperfusion through thrombolytic agents and mechanical thrombectomy. This process occurs due to disruption of the blood-brain barrier (BBB), resulting in blood leakage that increases mortality and morbidity from stroke. Thrombolytic therapy, fundamental in the treatment of stroke, faces the significant challenge of the risk of HT, caused both by the recanalization of the obstructed vessel and by non-fibrinolytic inflammatory mechanisms that weaken the BBB. Radiologically, HT is classified according to the ECASS scale into four types: HI-1 (small areas of petechial bleeding without mass effect), HI-2 (more publicized petechial bleeding), PH -1 (clot with moderate mass effect) and PH-2 (extensive clot with significant mass effect). **Objective:** To analyze the radiological pattern and incidence rate of hemorrhagic transformation in patients undergoing chemical thrombolysis, in the context of ischemic stroke due to large vessel occlusion, in a stroke unity. **Methods:** cross-sectional, observational and retrospective study from the year 2022, carried out by collecting data from patients who suffered HT after chemical thrombolysis in the context of LVO in a stroke unit. Tomographic interpretation was performed using the ECASS scale. **Results:** 35 patients had stroke due to LVO during the study period. Of these, 10 (28.57%) presented HT. Considering this population, 70% were women and 30% men, with a mean age of 66.2 years (range between 39 and 89 years). We used the ECASS scale to quantify HT, obtaining 20% of cases classified as HI-2, 30% as PH-1 and 50% as PH-2. The most affected artery was the M1 branch of the middle cerebral artery (MCA) with 70% of cases, followed by the internal carotid artery with 20% and the M2 branch of the MCA with 10% of cases. **Conclusion:** Hemorrhagic transformation was observed in 28.57% of patients undergoing thrombolysis, standing out as a highly relevant incidence factor for an unfavorable prognosis in stroke. Furthermore, it was revealed that HT occurs more frequently in women and older patients, with an average age of 66.2 years. The most affected segments were the M1 portion of the middle cerebral artery, both right and left, covering 90% of cases. Regarding radiological evolution according to the ECASS criteria, half of the patients were classified at the most severe level of the scale (PH-2), followed by 30% with TH PH-1, showing a predominance of more reserved evolutions.

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Incidence of meningitis in bahia from 2013 to 2023: an age group analysis

Jéssica de Andrade Ribeiro Lima[1], Bruna Rafaela Bastos Freire Quixabeira[1], Paulo Vitor Silva Santos[1], Ester Costa Lima[1], Fabrício Rodrigues Barbosa[1], Victor Alex Purificação de Castro Neves[1], Paula Cely da Silva Torres[1], Karollyne de Castro Santos[1], Alexandre David do Nascimento Santos[1], Lucas Granja Kaupert[1]

[1]Universidade do Estado da Bahia, Salvador BA, Brazil.

Correspondence: Ester Costa Lima; Email: ester.enfe@gmail.com.

Background: Meningitis is an inflammation of the meninges, the three membranes that cover and protect the brain and spinal cord. This condition can be caused by various pathogens, including bacteria (such as Neisseria meningitidis and Streptococcus pneumoniae), viruses (such as enteroviruses), and, less frequently, fungi and parasites. Clinically, meningitis manifests with symptoms such as severe headache, high fever, neck stiffness, and photophobia, potentially rapidly progressing to severe cases of sepsis and permanent neurological damage. In Bahia, meningitis represents a significant public health concern, with occasional outbreaks causing alarm among the population. **Objective:** To analyze the incidence of meningitis in Bahia from 2013 to 2023 according to each age group. **Methods:** This is an observational, descriptive, and analytical study, with data collected from confirmed cases reported in Bahia from 2013 to 2023, available in the Notifiable Diseases Information System (SINAN/SUS) as of June 2024. After collecting data according to the ICD-10 chapter on Meningitis in SINAN and population data from the latest demographic census (IBGE), the following analyses were performed according to the demographic variable age group: Incidence rate per 100,000 inhabitants and temporal trend according to the linear regression model using Microsoft Office Excel 2016 software. **Results:** During the analyzed period, 5,740 cases of meningitis were recorded in Bahia, with 24.62% in individuals aged 20 to 39 years, 24.10% in children aged 0 to 4 years, and 15.73% in individuals aged 40 to 59 years. Regarding the meningitis incidence rate in the state, there was a reduction of approximately 54.41% when comparing 2023 with 2013. In turn, the calculation of the incidence rate by age group during the period shows a higher incidence in children aged 0 to 4 years (12.1), followed by children aged 5 to 9 years (5.4), both above the average incidence rate for the period in Bahia (3.5), and a lower incidence in elderly individuals aged 65 to 69 years (0.9). From 2019 to 2020, a decrease in incidence rates was observed, returning to a growth trend in 2022. According to the simple linear regression model, a significant ($p < 0.05$) and decreasing trend in the number of cases over the period was noted, with an average reduction of approximately 1% per year. **Conclusion:** In Bahia, meningitis represents a significant public health concern. The state's health authorities have intensified efforts to control the spread of the disease through vaccination campaigns and case monitoring. Knowing the epidemiological profile and adopting appropriate management are essential to reducing the morbidity and mortality associated with meningitis in the region.

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Incidence of stroke in pediatric population in Joinville (Santa Catarina, Brazil)

Maria Eduarda Angelo de Mendonça Fileti[1], Thábata Emanuelle Martins Nunes[1], Yasmin Ramalho Mandarino[1], Henrique Diegoli[1], Carla Heloísa Cabral Moro[1]

[1]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Maria Eduarda Angelo de Mendonça Fileti; Email: neuro.mef@gmail.com.

Pediatric stroke can lead to significant mortality and morbidity, with 10-25% of children dying, more than 25% with recurrence and up to 66% with sequelae. Epidemiology in the United States in the last century indicated that acute ischemic stroke varies between 0.6 and 7.9 per 100 thousand children. The study by Lynch (2002), from 1979 to 2000, in the United States, indicated an incidence of 11.9 per 100 thousand children aged between 30 days and 18 years of age, with this number referring to the sum of hemorrhagic and ischemic strokes (4.1 and 7.8). Tsze and Valente (2011) estimated 1.2 to 13 cases per 100 thousand children under 18 years of age. A retrospective observational descriptive study was carried out on the incidence of stroke in the pediatric population in a city in the north of Santa Catarina. Data were collected from the JOINVASC database, a program runned out by the Joinville Municipal Health Department (SMS). Age and gender information was entered into Excel spreadsheets for organized observation of the distribution of total cases. Data were grouped every 2 years and applied to the Poisson distribution. Patients under 18 years of age, of both sexes, were included from January 1, 2010 to July 31, 2019 who had sufficient information in the database. Patients over 18 years of age and/or patients who had not suffered a stroke were excluded from the study. Between 2010-2011: Pediatric population: 319,476, 9 cases of stroke (average of 2.82 cases/100,000 inhabitants. CI 1.29 to 5.35). Between 2012-2013: Pediatric population 316,971, 6 cases of stroke (average 1.89 cases/100,000 inhabitants. CI 0.69 to 4.12). Between 2014-2015: Pediatric population: 315,342, 4 cases of stroke (average of 1.27 cases/100,000 inhabitants. CI 0.35 to 3.35). Between 2016-2017: Pediatric population: 313,889, 6 cases of stroke (average of 1.91 cases/100,000 inhabitants. CI 0.70 to 4.16). Between 2018-2019: Pediatric population: 313,007, 1 case of stroke (average 0.32 cases/100,000 inhabitants. CI 0.01 to 1.78). Our results are in line with data found in the literature, which is still scarce on the subject. New studies are needed to improve care for pediatric stroke victims, considering that risk factors, treatment and prognosis are not the same as for the adult population. A special attention should be given to the pediatric population in order to avoid possible sequelae that will be carried out for an entire lifetime.

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Incidental finding of acute disseminated encephalomyelitis in an elderly person: case report

Johnatan Felipe Ferreira da Conceição[1], Gutemberg Augusto Cruz dos Santos[1], Viviane Tavares Carvalho Crelier[1], Nathane Braga da Silva Rezende[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Johnatan Felipe Ferreira da Conceição; Email: johnatanfelipe@id.uff.br.

Case presentation: An elderly patient, 68 years old, reported sudden painless hearing loss in the left ear in December 2022. She was seen by an otolaryngologist with suspicion of herpetic infection, and a cranial MRI

was requested along with specific therapy initiation. Treatment with acyclovir began the same week, resulting in significant improvement, although the MRI was only performed on January 16, 2023. She received vaccinations on January 9, 2023, including COVID-19 (bivalent vaccine - Original+Ômicron BA.4/BA.5), measles, mumps, and rubella. The MRI revealed at least six hyperintense lesions on T2/FLAIR, enhanced with contrast, located in the subcortical white matter in the right parietal lobe, lower portion of the left precentral gyrus, right occipital lobe, and periventricular white matter near the temporal horn of the right lateral ventricle. The patient did not present with neurological complaints or other clinical symptoms, and neurological examination showed no significant abnormalities. Laboratory tests were requested, including inflammatory and autoimmune markers, which were unremarkable. Anti-MOG and anti-AQP4 antibodies (CBA method) were also negative. Cerebrospinal fluid analysis showed no significant changes, with 51mg/dL of proteins and negative oligoclonal bands. The overall clinical impression favored a diagnosis of acute disseminated encephalomyelitis (ADEM). Follow-up imaging showed previous lesions less prominently, with some slight contrast enhancement in the subcortical region of the right parietal high convexity, predominantly perivenular. No new lesions were observed, and analysis of the orbits and spinal cord showed no abnormalities. The patient continued clinical follow-up with an expectant approach and underwent regular monitoring exams, remaining asymptomatic with no new clinical complaints. The latest MRI in May 2024 showed no contrast enhancement in the lesions, which were now even less prominent, with no other abnormalities in the neuroaxis. **Discussion:** ADEM is a rare inflammatory demyelinating disease mainly observed in pediatric cases related to vaccines and post-infections. Its occurrence in adults is less frequent, complicating clinical definition and diagnostic criteria. This case presents incidental MRI findings suggestive of demyelination with a monophasic presentation. However, without typical neurological symptoms, but with a temporal relationship with vaccination, which, together with the exclusion of other pathologies, supports the hypothesis of ADEM, highlighting the diagnostic challenge in adults. **Final comments:** This case emphasizes recognizing atypical presentations of rare demyelinating pathologies in adults. A comprehensive clinical history and neurological examination, along with the exclusion of differential diagnoses, are crucial for accurate diagnosis and treatment.

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Inclusion body myopathy with dysphagia and its expressive improvement with the use of immunoglobulin

Lorena Fernandes Kronbauer[1], Felipe Leonardo Semensati[1], Fabio Gonçalves[1], Letícia de Barros Dias[1], Lucas de Oliveira Pinto Bertoldi[1], Marcelo Simplicio Carvalho[1], Nathalye Fernanda Pedroso Dircksen[1], Bárbara Okabaiasse Luizeti[1], Mônica Meneguette Silveira[1], Damacio Ramon Kaimen Maciel[1]

[1]Irmandade da Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Felipe Leonardo Semensati; Email: felipesemensati@hotmail.com.

Case presentation: Male, 41 years old, with weakness in 4 limbs, without pain, for 3 years, progressively, worse when associated with common clinical decompensations, such as colds and other viral infections. He evolved with moderate dysphagia and restriction to bed. On examination, he demonstrated a generalized loss of strength in the 4 limbs, proximal and distal, with a predominance of the flexor muscles of the fingers, the quadriceps and the extensors of the cervical region, muscle atrophy, without fasciculations, with normal Creatine Phosphokinase, Aldolase and Erythrocyte Sedimentation Rate. Electroneuromyography showing a diffuse myopathic pattern with proximal predominance in the quadriceps. Muscle biopsy demonstrated fiber hypotrophy, liposubstitution, slight endomyosial increase and intracytoplasmic sub-sarcolemic vacuoles. A genetic panel was carried out, without showing any changes. Rheumatological screening showed no changes. There was no improvement in the condition with the use of corticosteroids – so the diagnostic hypothesis was confirmed as Inclusion Body Myositis. Due to associated dysphagia, it was justified to start immunoglobulin at a standard dose, with complete improvement in dysphagia and a significant improvement in limb strength. Patient currently using symptomatic medication and immunoglobulin every 30 days for 4 months until the date of writing this report, maintaining significant improvement. **Discussion:** Inclusion body myositis (IBM) is classified within the idiopathic inflammatory myositis (IIM), a group that also encompasses dermatomyositis, polymyositis and autoimmune necrotizing myositis. Clinically, IBM initially manifests itself with significant weakness in the quadriceps and finger flexor muscles, and is more prevalent in men in a 2:1 ratio. The condition gradually progresses to other muscle groups and, on average, patients lose the ability to walk without assistance within 7 to 10 years after symptoms appear. Furthermore, it can affect the oropharyngeal and esophageal muscles, causing dysphagia. The therapeutic approach to IBM is predominantly empirical due to the limitation of robust clinical studies. Existing literature suggests that IBM tends to be refractory to a range of immunosuppressive agents, including corticosteroids. However, considering the progressive and debilitating nature of the disease, as well as isolated reports of beneficial responses to treatment, with empirical use of rituximab, cyclophosphamide, immunoglobulin, methotrexate, without responses to corticosteroids. **Final comments:** Inclusion body myositis is a disease that is difficult to diagnose, uncommon and has a disabling prognosis. There is generally no good response to treatment, which for now is only empirical with immunosuppressants.

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Inclusion body myositis with exceptional response to immunosuppressive treatment

Mariana Hiromi Manoel Oku[1], André Macedo Serafim da Silva[1], Bruna Moreira de Souza Proença[1], Gabriella Côrrea Dousseau[1], Gabriel Saboia de Araujo Torres[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Mariana Hiromi Manoel Oku; Email: mari.hiromi.mo@gmail.com.

Case presentation: A 60-year-old woman presented with generalized muscle weakness and progressive walking difficulty. She had a previous diagnosis of systemic erythematous lupus due to an episode of nephritis in 2006 with a confirmatory renal biopsy. Neurological examination showed mild proximal weakness with normal reflexes and no sensory or bulbar involvement. She had no other systemic symptoms suggestive of lupus activity. Creatine phosphokinase (CK) levels were around 2800U/L and aldolase was 12U/L. Inflammatory markers were low, complete blood count and renal function displayed no abnormalities. Electromyography was compatible with a myopathic pattern. Magnetic resonance imaging (MRI) showed muscle edema in the biceps, triceps, radial extensors, and pronators bilaterally in the upper limbs and in the anterior and medial compartments of the thighs. Prednisone and methotrexate were initiated considering the possibility of an overlap myositis. The patient presented with complete resolution of the weakness after a few months of immunotherapy. However, muscle biopsy displayed rimmed vacuoles, altered mitochondrial activity, and TDP-43 aggregates, besides inflammatory endomysial reaction (with an increase in CD4, CD8, CD68, and MHC-I expression), a pattern indicative of inclusion body myositis. Repeated electromyography showed no myopathy and MRI had significant edema reduction. Post-treatment CK was 80 U/L and aldolase was 2,9 U/L. **Discussion:** Inclusion body myositis (IBM) is a progressive muscle disease with an uncertain pathogenesis. Some authors defend that an autoimmune T-cell-mediated response has a major role in disease development whereas others believe that neurodegenerative mechanisms are more important. Nevertheless, there is a consensus about treatment refractoriness and how the side effects of immunotherapy outweigh its benefits in most cases of IBM. We present a case with a confirmed diagnosis of IBM which fulfilled diagnostic criteria and showed an excellent response to immunosuppression. We hypothesize that this rare evolution can be due to a more prominent inflammatory component in this particular case as it was observed in the patient's muscle biopsy. Additionally, her rheumatological background might also have played a significant role in the favorable response. **Final comments:** Thus, we believe that it is essential to carefully assess IBM cases, evaluating the importance of the inflammatory component in each case to decide if an immunosuppression trial should be performed.

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Inclusive neuroanatomy education for visually impaired students: development of 3D printed models of the spinal cord

Gabrielle Gomes Garcia[1], Roberto Godofredo Fabri Ferreira[1], Mariana Augusta Penna e Costa de Saldanha da Gama Fischer[1], Luciana Morbach Miranda[1], Vítor Garcia Gomes[1], Raphael Viana Curvelo Sepúlvida[1], Claudio Tinoco Mesquita[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Gabrielle Gomes Garcia; Email: gabriellegarcia3g@gmail.com.

Background: The increasing number of students with disabilities in universities has gained worldwide relevance. When it comes to visually impaired students, teaching morphology becomes a complex process, once visual perception plays an essential role in traditional anatomical knowledge. Neuroanatomy, in particular, is challenging, given the complexity and small size of its structures. Within this context, 3D printing has emerged as an additional tool for the educational inclusion of these students. The 3D printing technology consists of creating a real object from a digital model, which can be adapted in size, shape and texture. **Objective:** Produce and evaluate the applicability of neuroanatomical models created using a 3D printer, with sizes, textures and shapes adapted to the learning of visually impaired students. **Methods:** This is an ongoing experimental study. NOMAD, ZBRUSH and BLENDER software programs were used for three-dimensional modeling of the following anatomical structures: multipolar neurons; spinal cord cross-sections and their tracts; peripheral nerves. The design of the models was based on extensive research into traditional neuroanatomy atlases and textbooks. The 3D models were built using Fused Deposition Modeling (FDM). The materials used were polylactic plastic (PLA) and thermoplastic polyurethane (TPU). Subsequently, the aim of this study is to introduce visually impaired university students to practical neuroanatomy classes, comparing the learning performance of the group that used 3D printed models with the group that did not. **Results:** This project aims to share our experience with creating and using 3D models. We magnified macro and microscopic structures, reaching pieces of about 10 cm. Different three-dimensional planes and varied textures, such as dots and hatches, were used to distinguish white from gray matter and their respective tracts in the spinal

cord segments. Our preliminary experiences with the 3D models reveal that, by bringing the neuroanatomical perception of the visual field into the tactile field, as well as the proportional amplification of the neural structures, spatial understanding by visually impaired students has evolved from an eminently theoretical field to a real and concrete perception. It should also be mentioned that the students' observations and suggestions also guided us in the progressive improvement of the didactic models. Finally, it is worth pointing out that we are still working on a definitive model in order to understand, statistically, the effectiveness of these models in neuroanatomy classes. **Conclusion:** The neuroanatomy models produced by the 3D printer can be a tool for the inclusion of students with and without visual impairments, improving their understanding and learning. However, more studies and evaluation methods are needed to quantify the real impact of these pieces.

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Infantile cerebral venous thrombosis associated with basal ganglia hemorrhage due to protein's deficiency: case report

Clara e Silva Modesto[1], Luana Marcondes Emergente Caproni[1], Renato Ortolani Marcondes de Castro[1], Marcelo Massote de Toledo[2]

[1]Universidade José do Rosário Vellano, Alfenas MG, Brazil.
[2]Universidade do Vale do Sapucaí, Pouso Alegre MG, Brazil.

Correspondence: Clara e Silva Modesto; Email: modestoclara07@gmail.com.

Case presentation: A one-year-old female patient sought medical care due to cephalea associated with projectile vomiting and diarrhea. She had a progress in her condition with changes in level of consciousness, motor deficit and second stage of dehydration after bacterial tonsillitis, she has started treatment with amoxycillin and ceftriaxone 5 days ago. Scans have been conducted, including a cranial computed tomography (CT), which found a cerebral venous thrombosis (CVT) in cavernous sinus, straight sinus, transverse sinus and left great cerebral vein and basal ganglia hemorrhage. Laboratory tests: Hemoglobin (Hb): 10.9; Hematocrit (Ht): 32.7%; Total Iron-Binding Capacity (TIBC): 77.3; RDW: 14.7; Platelets: 700,000; Total Leukocytes: 13,500 (1% Rod cells, 79% Segmented Neutrophils, 15.4% Lymphocytes); C-Reactive Protein (CRP): 32.5; Amylase: 13; Blood Glucose: 137; Potassium (K+): 5.2; Sodium (Na): 135; AST: 51; ALT: 52. Heparinization was performed and patient was hospitalized in Intensive Care Unit, evolving with an improvement of the condition, and then she was discharged after 13 days, when she had no sequela. The patient was guided to continue treatment and was referred to a specialist to investigate the cause of thrombosis, when protein S (PS) deficiency was checked, after laboratory tests performed on February 11, 2010 (PS: 30%). **Discussion:** CVT is a rare subtype of stroke and has an estimated incidence of 1.32 per 100,000 people. About 20% to 30% of the cases are related to genetic factors, and PS deficiency is the main factor and the most difficult to be diagnosed, being found in 1.5 to 7% of the thrombophilic patients. This deficiency is characterized by a defect on PROS1 gene, whose function is crucial when binding to protein C, conducting degradation in the coagulation factors Va and VIIIa. **Final comments:** Mortality rate of CVT can vary from 5 to 30%, but it has some mortality predictors or unfavorable prognosis such as change in level of consciousness and cerebral hemorrhage. The condition in children has a better prognosis, once their brain has a greater neuroplasticity. Thus, we highlight that treatment success comprises a multidisciplinary team and a specialized medical care follow-up.

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Infection sites most associated with the occurrence of delirium during hospitalization in a stroke unit: a cohort study in a reference hospital in Bahia

Maria Eduarda Messias Vassoler[1], Gabriel Souza Medrado Nunes[1], Milena Fernandes de Oliveira[1], Gabriel Teles de Oliveira Piñeiro[1], Luiz Paulo Oliveira Martins[1], Thaís Barbosa de Oliveira[1], João Pedro Fernandes Gonçalves[1], Alex de Cerqueira Silveira Figueiredo[1], Antonio Lázaro Santos Soares Junior[1], Pedro Antonio de Jesus[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Geral Roberto Santos, Salvador BA, Brazil.

Correspondence: Maria Eduarda Messias Vassoler; Email: maria.vassoler@ufba.br.

Background: Delirium is a clinical syndrome linked to various short- and long-term complications in stroke patients. Among many predisposing factors, infections are highly associated with delirium during hospitalization. However, there is still a lack of comprehensive research on the association between the occurrence of infection sites and delirium in hospitalized stroke patients. **Objective:** To investigate the association between infection sites and the occurrence of delirium in stroke patients. **Methods:** This study enrolled patients with acute stroke admitted to a stroke unit at a referral hospital in Bahia, between July/2023 and March/2024. Clinical and epidemiological characteristics were extracted from patients' medical records or obtained through personal interviews. Infection was defined as the administration of antibiotics or documented infection noted in medical records. Delirium was assessed using the CAM-ICU scale, conducted by trained researchers. The chi-square test was employed to ascertain associations, and odds ratios with confidence intervals were derived through logistic regression. **Results:** The study comprised 213 patients, of whom 51.2% were female, with a median age of 66.3 years (IQR: 59-74.3). Within this cohort, 66.7% lacked a completed high school education. Regarding comorbidities, 36.2% had diabetes, 78.95% had hypertension, and 27.7% had dyslipidemia. Delirium occurred in 47 patients (22.1%) at least once during hospitalization. Infection was diagnosed in 95 patients (44.6%), with respiratory infection diagnosed in 34 (35.8%), urinary infection in 53 (55.8%), skin infection in 6 (6.3%), bloodstream infection in 3 (3.2%), other types of infection in 10 (10.5%), non-specified infection in 37 (38.9%), and 38 patients (40%) had more than one type of infection during the hospitalization time. The presence of infection during hospitalization was significantly associated with delirium occurrence ($p<0.001$). Among different infection sites, respiratory tract infection exhibited the strongest association with delirium risk ($p<0.001$; OR: 4.1; 95% CI: 1.8 - 9.4), followed by urinary tract infection ($p<0.05$; OR: 2.6; 95% CI: 1.2-5.4). The presence of multiple infection sites was not more associated with delirium than single sites ($p>0.05$). **Conclusion:** In our study, the respiratory tract was the infection site most associated with delirium, while urinary tract infection was the second. The presence of more than one site of infection was not more associated with delirium. However, more studies are needed to better understand these associations.

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Inflammatory polyneuropathy in the state of São Paulo: a comparative analysis of of the pre-pandemic and post-pandemic period

Aline Marques Barreto Scalioni[1], Maria Luiza Nunes Guimarães[2], Camila Variani[3], Thaís Rigon[4], Fernanda Oliveira Fonseca[5]

[1]Universidade Nove de Julho, Guarulhos SP, Brazil.
[2]Universidade de Franca, Franca SP, Brazil.
[3]Universidade Luterana do Brasil, Canoas RS, Brazil.
[4]Atitus Educação, Passo Fundo RS, Brazil.
[5]Instituto Universitário de Ciências de La Salud Fundacion H. A Barceló, Capital Federal, Argentina.

Correspondence: Aline Marques Barreto Scalioni; Email: alinemscalioni@gmail.com.

Background: Acute inflammatory polyneuropathy is an inflammatory demyelinating neuropathy that affects peripheral nerves. It is characterized by muscle weakness, impairment of reflexes and tactile sensitivity, associated with increased levels of proteins in cerebrospinal fluid. It is a treatable, yet under diagnosed, condition, that affects 1 child in 200,000 and at least 1 to 2 adults in 100,000. **Objective:** To analyze the cases of inflammatory polyneuropathy in the pre-pandemic and post-pandemic periods. **Methods:** This is an ecological, descriptive, and observational study with a quantitative approach. Data were obtained from the Department of Informatics of the Unified Health System (DATASUS), through the TABNET platform, extracting information about the incidence of hospitalizations for inflammatory polyneuropathy in the state of São Paulo from 2018 to 2024, as well as its mortality. The population analyzed includes individuals regardless of age, and the variables analyzed were the month and year of hospitalization. **Results:** From the years of 2018 to 2024, São Paulo had 4,474 hospitalizations for inflammatory polyneuropathy, with 2023 (the last year of the COVID-19 pandemic according to the World Health Organization) having the highest number of hospitalizations (n=922) (about 20.6% of the total). March of 2023 had the highest number of cases (n=97). Analyzing the cases in relation to the pre-pandemic and post-pandemic periods of COVID-19, the years of 2018 and 2019 had 682 and 759 cases, respectively, with a decline in the initial pandemic years, 2020 and 2021, respectively 558 and 629 hospitalizations, which can be due to underreporting of cases. Numbers increased again in the years of 2022 to 2024, with 847 cases in 2022, 922 in 2023, and 77 hospitalizations up to February 2024. After analyzing the results, there was a 28.2% increase in registered cases of polyneuropathy comparing the pre-pandemic and post-pandemic years. Further studies are required to confirm these findings, but a potential relationship has been observed between these pathologies and the immunological side effects associated with COVID-19. **Conclusion:** The findings of this study concluded that the number of notified inflammatory polyneuropathy cases in the years 2022 and 2023, after the pandemic, increased by a difference of 335 cases, compared to the years of 2018 and 2019, before the pandemic. It can be related to the reporting of some cases of Guillain-Barré syndrome after COVID-19 infection (still requiring further studies for certified correlation). One of the limitations of this study was the exclusive use of the DATASUS database

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Influence of gender on IBM: analysis of epidemiological and clinical characteristics

Bruna Moreira de Souza Proença[1], André Macedo Serafim da Silva[1], Ana Lucila Moreira[1], Cristiane de Araújo Martins Moreno[1], Eliene Dutra Campos[1], Mariana Cunha Artilheiro[1], Tatiana Ribeiro Fernandes[1], Rodrigo Holanda Mendonça[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Bruna Moreira de Souza Proença; Email: brunamsproenca@gmail.com.

Background: Inclusion Body Myositis (IBM) is the most common acquired myopathy in individuals older than 50 years. The disorder is slowly progressive, and response to therapies has generally been poor. Clinical heterogeneity may influence treatment responsiveness; however, data

regarding heterogeneity in IBM are limited and often conflicting. Disease progression has been related to the age of onset, with faster progression when the age of onset is more advanced. A recent study revealed that female and black patients have distinct clinical phenotypes. Gender differences may play a role in the variability of clinical severity and progression. **Objective:** To describe epidemiological and clinical characteristics of IBM patients, and to investigate the relationship between disease duration and clinical severity with a focus on gender differences. **Methods:** We evaluated a cohort of IBM patients (6 males, 8 females) from a center of neuromuscular diseases in Brazil using clinical assessments, IBM Functional Rating Scale (IBMFRS), and Motor Function Measure (MFM32). To compare variables between genders and assess the relationships between disease duration and severity, we performed statistical analyses, including Mann-Whitney tests, Pearson correlations, and interaction regressions. **Results:** The median age at onset was 49.5 years for males (45.0 - 66.8) and 51.0 years for females (45.3 - 58.5). The median time to diagnosis was 4.5 years for males (3.3 - 9.0) and 2.5 years for females (1.0 - 4.8). The median disease duration was 7.5 years for males (4.3 - 12.5) and 9.5 years for females (5.8 - 13.8). The median IBM-FRS score was 28.5 for males (20.8 - 34.8) and 31.0 for females (22.0 - 36.5), with no significant difference. Disease duration significantly impacted IBM-FRS scores, reducing by approximately 1.176 points per year (B=-1.176, p<0.001). Gender did not significantly influence the IBM-FRS score (T=-0.128, p=0.901). The median MFM scores for 9 patients showed no significant difference: males had 65.8 (16.1 - 91.7) and females had 67.7 (40.6 - 92.2) (p=0.624). Mobility status varied slightly: 62.5% of females and 50% of males ambulated without support, with no significant association between gender and mobility status (χ^2=0.271, p=0.873). **Conclusion:** Our study indicates no significant differences between male and female IBM patients in terms of epidemiological and clinical data. While disease duration significantly impacts severity, gender does not. As research suggests some differences between genders, future comparisons should investigate other clinical, laboratory, and biopsy characteristics. Further research with larger sample sizes is needed to confirm these findings and explore the underlying mechanisms affecting IBM progression.

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Influence of ischemic stroke on intestinal permeability and systemic immune response: a preclinical study

Fabrício Weinheimer Lippert[1], Maria Amélia Cechinel da Silva[1], Anita Dal Bó Tiscoski[1], Sandra Bonfante[2], Natália Piacentini da Silva[1], David dos Santos[1], Naíla Maciel Andrade[1], Khiany Mathias[1], Richard Simon Machado[2], Fabricia Petronilho[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.

Correspondence: Fabrício Weinheimer Lippert; Email: fabriciowlippert@gmail.com.

Background: Studies reveal that the established peripheral immunosuppression in cases of ischemic stroke may make survivors susceptible to infections. Furthermore, ischemic stroke is increasingly associated with impairment of the intestine-brain axis. **Objective:** To show intestinal permeability as well as indications of peripheral immunosuppression using a preclinical model of ischemic stroke. **Methods:** We divided the animals into two groups: 1) Sham and 2) middle cerebral artery occlusion (MCAO). We evaluated the neurological score, weight, and temperature at 3 h, 24 h, and 48 h after MCAO. After 72 hours of MCAO, we induced painless death in the animals and removed blood through cardiac puncture to assess intestinal permeability, total cell count, body weight, spleen weight, temperature, and neurological score. **Results:** In animals subjected to MCAO, intestinal permeability and blood cell count increased compared to the sham group. There was a significant decrease in weight in all groups within 72 hours after MCAO and sham. A neurological score revealed a cognitive decline in the initial hours after MCAO, but this decline reversed in the following days. There was a decrease in spleen weight, such as in the spleen weight/animal weight ratio in the MCAO group. **Conclusion:** We conclude that ischemic stroke due to MCAO can lead to an increase in intestinal permeability and peripheral immunosuppression in rats.

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In-hospital mortality rate due to meningitis in Brazil, from 2008 to 2022: a joinpoint analysis

Emmanuel Santos Trindade[1], Priscila Santos Oliveira[1], Philip George Glass Andrade[1], Davi Tanajura Costa[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Gustavo Alves Cangussú; Email: canguss.gustavo@gmail.com.

Background: Meningitis, despite public health efforts to reduce its incidence through vaccination against N. meningitidis, S. pneumoniae and H. influenzae, continues to present significant lethality, reaching up to 54% in low-income countries. **Objective:** To analyze the in-hospital mortality rate due to meningitis in Brazil, from 2008 to 2022. **Methods:** This is an ecological time series study of the in-hospital mortality rate due to meningitis in Brazil, stratified by sex, ethnicity and age group. Data on hospitalizations for meningitis from the Hospital Information System (SIH) were used, recorded by ICD-10 codes A17.0, A32.1, A39.0, A87, B00.3, B01.0, B02.1, B05.1, B26.1, B37.5, B38.4, G00, G01, G02 and G03. A temporal analysis was carried out using a joinpoint regression model, through which the Annual Percent Change (APC) and Average Annual Percent Change (AAPC) were obtained with 95% Confidence Interval (95% CI). A p value ≤ 0.05 was considered statistically significant. **Results:** The in-hospital mortality rate from meningitis in Brazil from 2008 to 2022 was 66.12 deaths/1000 hospitalizations. Mortality was higher in females (67.00 deaths/1000 hospitalizations) than in males (65.48 deaths/1000 hospitalizations). The ethnicities in decreasing order of mortality are indigenous (123.60 deaths/1000 hospitalizations), black (98.28 deaths/1000 hospitalizations), brown (70.33 deaths/1000 hospitalizations), yellow (70.10 deaths/1000 hospitalizations) and white (61.86 deaths/1000 hospitalizations). There is a directly proportional relationship between age and mortality rate, ranging from 29.27 deaths/1000 hospitalizations in the 0-9 years age group to 324.88 deaths/1000 hospitalizations in the ≥ 80 years age group. The mortality rate showed an increasing trend (AAPC: 4.26 [2.48; 6.06)]; p:<0.001). This trend was followed by males (AAPC: 4.57 [3.07; 6.10]; p:<0.001) and females (AAPC: 3.84 [1.52; 6.22]; p:0.003). White (AAPC: 4.32 [2.00; 6.70]; p:0.001) and brown (AAPC: 4.60 [2.30; 6.95]; p:<0.001) ethnicities showed an increasing trend. The other ethnicities in turn showed a stationary trend. The age groups of 10-19 years (AAPC: 4.44 [2.87; 6.03]; p:<0.001), 20-29 years (AAPC: 3.61 [0.52; 6.79]; p:0.03) and 50-59 years (AAPC: 2.76 [0.01; 5.58]; p:0.05) showed an increasing trend. The other age groups showed a stationary trend. The 0-9 years age group presented 2 joinpoints, characterizing a decreasing trend from 2008 to 2013 (APC: -10.90 [-13.93; -7.75]; p:<0.001) and a stationary trend both from 2013 to 2016 (APC: 16.31 [-0.40; 35.82]; p:0.055) and from 2016 to 2022 (APC: -1.12 [-3.68; 1.51]; p:0.34). The ≥80 years age group presented a joinpoint in 2010, but characterized a stationary trend in both determined periods. **Conclusion:** Based on the results, it is clear that in-hospital mortality due to meningitis in Brazil showed an increasing trend, mainly among white and brown ethnicities, young adults and middle-aged people.

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Initial neurological manifestations of cryptococcosis in non-HIV adults

Flavia Ana Pacheco[1], Renata Cicci Cunha Castro[1], Bárbara Rodrigues Amaral de Oliveira[2], Ruth Maria Ribeiro Guerra[1]

[1]Universidade de Uberaba, Uberaba MG, Brazil.
[2]Centro Universitário de Brasília, Brasília DF, Brazil.

Correspondence: Flavia Ana Pacheco; Email: flavia.ana.pacheco@gmail.com.

Background: Cryptococcus is an opportunistic yeast with an immunoprotective polysaccharide capsule possessing immunomodulatory properties, which predisposes the organism to neurotropism. The primary infection occurs in the lungs through inhalation of the pathogen, predominantly affecting immunocompromised individuals. It can present asymptomatically or more commonly as meningoencephalitis and pulmonary forms. In immunocompetent individuals, meningoencephalitis is the main presentation. **Objective:** To describe the most frequently presented initial neurological manifestations in non-HIV adults with cryptococcosis. **Methods:** A systematic literature review was conducted with the guiding question: "What are the most frequently presented initial neurological manifestations of non-HIV adults with cryptococcosis?" The Pubmed database was used to search for studies using the descriptors: Cryptococcosis, Neurologic Manifestations, Neurologic Examination and HIV, with the help of Boolean operators "AND", "OR" and "NOT". Articles published between 2019 and 2024 were included and those evaluating cryptococcosis in HIV patients, studies conducted on individuals under 18 and over 65 years and studies not reporting the initial clinical neurological manifestations were excluded. **Results:** The final sample consisted of 19 articles, including 18 case reports and one case-control study with 270 patients, totaling an N of 288. In total, 35 types of neurological alterations were identified, with each article citing between one and nine alterations. Headache was the most frequent manifestation, present in 158 patients (54.7%), followed by meningeal signs in 59 patients (20.4%), decreased auditory acuity in 7 patients (2.5%) and fever in 6 patients (2%). Among the less frequent alterations cited in 2 patients (0.7%) were cognitive alterations, photophobia, nausea, diplopia, ptosis, gait disturbance, altered mental state, seizures, vomiting, visual disturbance, papilledema and ocular movement limitation. Finally, among the alterations reported in 1 patient (0.3%) were tinnitus, dizziness, hemiballismus, blurred vision, ocular deviation, mydriatic pupil, non-reactive pupil, slowed reasoning, altered consciousness level, balance disturbance, retro-orbital pain, lacrimation, hemichorea, confusion, decreased facial sensitivity, facial mimic alteration and dysarthria. **Conclusion:** Cryptococcosis in non-HIV patients is a condition with a challenging early diagnosis. The most common neurological manifestations of the disease include headache, meningeal signs, decreased auditory acuity and fever. Identifying the initial neurological alterations presented in the disease can help guide the diagnostic reasoning towards cryptococcosis in differential diagnosis and direct early treatment for a better prognosis.

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Integrating objective and subjective data in a new outcome scale for epilepsy surgery in people with severe epilepsies (PWSE): the Porto Alegre seizure impact scale (PASIS)

André Palmini[1], Eliseu Paglioli[1], Rafael Paglioli[1], William Martins[1], Fernanda Schuh Martins[1], Thomas Frigeri[1], André Gus[1], Victoria Bizzi Schvartzman[1], Natalie Donida[1], Bibiana Thomé[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Victoria Bizzi Schvartzman; Email: bizzivictoria@gmail.com.

Background: Significant improvement with epilepsy surgery in PWSE -despite seizure persistence- includes shorter and milder seizures, or no longer needing frequent ER visits because of prolonged TCS. These post-op gains beyond complete seizure freedom POSITIVELY IMPACT the lives of PWSE but are not captured by objective outcome scales focused on degree of seizure control, such as the Engel and ILAE outcome scales. **Objective:** Here we propose a new scale -the PASIS- which is more relevant for PWSE in whom seizure freedom is unlikely, yet surgery may bring significant improvement. **Methods:** The PASIS integrates objective and subjective data both pre and post-operatively, considering changes in frequency, severity, duration and circadian distribution of seizures and post-ictal abnormalities, as well as the global impression of the effectiveness of surgery for the life of the patient and the family. PASIS scores are a sum of points from each category and the higher the score the worse the outcome. Subjective items may decrease or increase final scoring. Subjective scores are -1 when there was improvement, +1 when that feature was like before operation and +2 if worse. Thus, when the impact of the operation is minimal, post-op PASIS scores increase. Difference between post- and pre-op scores lead to a 4-tiered outcome classification: 5 or more points less, PASIS 1 (patient much improved); 1 to 5 points less, PASIS 2 (improved); equal scores, PASIS 3 (similar condition); bigger than pre-op score, PASIS 4 (worse). **Results:** Illustrative case: 17-month-old boy with infantile spasms evolved with severe multifocal epilepsy, many daily generalized seizures, protracted post-ictal somnolence and psychomotor regression. There was mild right hemiparesis and bilateral gyral abnormalities yet a gross abnormal area, resembling polymicrogyria, in the left frontal lobe. EEGs had diffuse abnormalities and independent trains of spikes and high amplitude slow waves over one or the other hemisphere, predominating in the left frontal lobe. Pre-op PASIS was 13 (generalized seizures with sudden falls (head drop) = 4 points; daily seizures (≥5/day) = 5 points; seizures also during wakefulness = 2 points; and protracted, disabling post-ictal changes = 2 points). Resection under acute ECoG removed the lesion on the left frontal lobe. 12 months later, the boy is walking and interacting, still with daily seizures, but mild and brief. With no post-ictal. He is Engel IV – but his PASIS went from 13 pre-op to 5 post-op; FINAL PASIS = 1 (much improved), reflecting significant gains despite seizure persistence. **Conclusion:** PWSE suffer from frequent seizures that can lead to recurrent ER visits and protracted post-ictal disability. Many have surgically remediable epilepsies that although responding partially to surgery lead to significant improvements in daily life, not captured by scales focused solely upon degree of seizure control and thus a new scale is proposed to gauge the effect of surgery in this population.

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Intellectual disability and neuropsychomotor delay in a patient with a rare variant 49,XXXXY of Klinefelter syndrome

Viviane Müller[1], Maria Fernanda Peruci Felippe[1], Christian Camatti Menegon[1], Vitor Ritt Xavier[1], Lara Willers Lobato[1], Maria Eduarda Friedrich Pfeifer[1], Maria Eduarda Corrêa Pereira[1], Aline Deborah de Moraes[1], Rafaela Gageiro Luchesi Soares[1], Rafael Fabiano Machado Rosa[1]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Viviane Müller; Email: viviane.muller@ufcspa.edu.br.

Case presentation: The patient was the first child of a young couple. During pregnancy, intrauterine growth restriction was diagnosed in the seventh month. He was born at term, by vaginal delivery, weighing 1950 grams, measuring 43 centimeters, and with an Apgar score of 9 at the fifth minute. At birth, Down syndrome was considered as a diagnosis. Cardiological clinical evaluation revealed a probable pulmonary stenosis. Regarding neuropsychomotor development, he experienced delays, sitting without support at 8 months and walking alone at 3 years and 1 month. At 3 years and 9 months, he was able to pronounce a few words. He later presented learning difficulties. Hand and wrist radiography showed significant bone maturation delay. Physical examination revealed flattening of the occipital skull; low anterior and posterior hairline; upslanting palpebral fissures; posteriorly rotated ears with overfolded helix; high arched palate; retractile testes and small penis hidden in the pubic fat. Karyotyping revealed the presence of a male chromosomal constitution with X chromosome tetrasomy - 49,XXXXY - which was compatible with the diagnosis of Klinefelter syndrome variant. Endocrinological evaluation confirmed the presence of hypergonadotropic hypogonadism. **Discussion:** Reports of variants of Klinefelter syndrome are considered rare. We emphasize the challenge of diagnosing these variants based on clinical findings, given the overlap with those of other chromosomal abnormalities. Therefore, karyotyping is essential for the etiological definition of the syndrome and, consequently, the appropriate management of the patient. **Final comments:** With this report, we highlight the importance of considering variants of Klinefelter syndrome, such as 49,XXXXY, in individuals with neuropsychomotor delay and intellectual deficit, presenting with dysmorphic features and hypogonadism.

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Intensive blood pressure lowering for intracerebral hemorrhage: a systematic review and updated meta-analysis of randomized controlled trials

Pedro Henrique Reginato[1], Gabriel Paulo Mantovani[2], Vinicius Furtado da Silva Castro[3], Giovanna Salema Pascual[4], Letícia Felício Saldanha[5], Leonardo Zumerkorn Pipek[6]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[3]Hospital Universitário Onofre Lopes, Natal RN, Brazil.
[4]University of Bologna, Bologna, Italy.
[5]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[6]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Pedro Henrique Reginato; Email: pedroreginato@ufpr.br.

Background: Intracerebral hemorrhage (ICH) is often complicated by elevated blood pressure (BP), increasing the risk of persistent and recurrent hemorrhage, disability, and death. Numerous research initiatives have yielded divergent results concerning BP management, leading to uncertainty, particularly regarding the influence of treatment timing and the optimal BP target. **Objective:** To perform a systematic review and updated meta-analysis comparing the impact of intensive BP lowering versus standard BP lowering in ICH patients. **Methods:** We searched PubMed, Embase, and Cochrane databases for randomized controlled trials (RCT) comparing intensive BP lowering, commonly defined as BP<140 mmHg, to standard treatment in ICH patients. The protocol was registered on PROSPERO (CRD42024552587). A pooled analysis was conducted to evaluate good functional outcome (defined as a modified Rankin Scale (mRS) score of 0 to 2), major disability or death (mRS score of 3 to 6), all-cause mortality, and substantial hematoma growth. Subgroup analyses examined differences in outcomes concerning studies in which only the intervention group maintained target BP levels below 140mmHg, and according to time-to-treatment (before or after 6 hours from the onset of ICH). Statistical analysis was performed using RevMan 5.4.1. Heterogeneity was assessed with I^2 statistics. **Results:** We included 11 RCTs with a cohort of 13.604 patients, of whom 6.743 (49.56%) underwent intensive BP lowering. Intensive treatment was associated with a reduction in major disability or death (RR=0.93; 95% CI=0.90-0.96; p<0.001; I^2=0%), all-cause mortality (RR=0.86; 95% CI=0.79-0.94; p<0.001; I^2=0%) and substantial hematoma growth (RR=0.83; 95% CI=0.73-0.95; p=0.008; I^2=18%) compared to the standard group, while good functional outcome showed no statistically significant difference (RR=1.08; 95% CI=0.99-1.18; p=0.08; I^2=53%). In the subgroup analyses, there were no statistically significant differences in any outcomes related to studies in which only the intervention group maintained target BP levels below 140mmHg or according to time-to-treatment. **Conclusion:** Our findings suggest intensive blood pressure lowering management in intracerebral hemorrhage patients reduces major disability or death, all-cause mortality, and substantial hematoma growth in comparison to standard treatment.

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Interferon gamma, IL-18 and NFL as potential biomarkers for COVID-19 severity and lethality

Helena França Alcaraz Ferreira[1,2], Isabela Stornelli Teixeira[2], Andreza Salvio Lemos[2], Renan Amphilophio Fernandes[2], Carlos Henrique Ferreira Ramos[1], Carlos Henrique Ferreira Ramos Filho[3], Wagner Luís da Costa Nunes Pimentel Coelho[4], Luciane Almeida Amado Leon[4], Soniza Vieira Alves-Leon[1,2]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Universidade do Estado do Rio de Janeiro, Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.
[4]Fundação Oswaldo Cruz, Rio de Janeiro RJ, Brazil.

Correspondence: Isabela Stornelli Teixeira; Email: isabelastornelli@edu.unirio.br.

Patients infected with SARS-CoV-2 may experience different disease manifestation based on factors like age, sex, comorbidities and the host's immune integrity. Its pathophysiology has been contemplated as a complex web of cell signaling pathways and systemic consequences, presenting the risk of hospitalization and the manifestation of severe symptoms. Aggravated pro-inflammatory cytokine response has been associated with lethality and extensive lesions. Thus, this study aims to investigate the relation between the levels of interleukin-18 and interferon gamma (IL-18, IFN-y), neurofilament light chain (NfL) and disease outcome in patients with acute COVID-19. In order to analyze these biomarkers, peripheral blood was collected, and plasma was examined accordingly. A cohort of 32 acute severe COVID-19 patients, critically ill, was investigated. Statistical analysis was performed using Kruskal Wallis and Mann Whitney U test. This cohort was compared to its homologous healthy control group. Regarding data evaluation, the SIMOA platform was used for NfL, while the cytokines were analyzed using the Luminex 200 system. Our results showed that there were statistically significant differences in NfL (p<0.0001), IFN-y (p=0.0230) and IL-18 (p=0.0014) IFN-y levels (p=0.0056) between severe COVID-19 cohort and healthy control group. When we deeper analyze the severe COVID-1 group, we divide them according to the outcomes: Deceased and Survivor. For those, we also found that NfL (p<0.0001), IFN-y (p=0.0056) and IL-18 (p=0.0008) levels where significantly higher in the deceased sub-group when compared to survivor group.

It has been corroborated that a dysregulated immune response leads to tissular impairment and, in turn, worsening of patient condition. Also, NfL levels may play an important role as a biomarker of cognitive impairment and neurological manifestations associated to disease severity and lethality in COVID-19. For that reason, it may be of interest to explore those biomarkers in larger cohorts as they possibly may be important prognostic tools for COVID-19 positive patients, aiming for a more precise and faster clinical evaluation and, consequently, appropriate treatment.

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Interleukin-6 inhibitors in neuromyelitis optica spectrum disorder: a systematic review and meta-analysis

Giovani Noll[1], Marcos Madeira de Lima[1], Viviana Konzen[1], Alessandro Finkelsztejn[1]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Giovani Noll; Email: nlgiovanimdr@gmail.com.

Background: Interleukin-6 (IL-6) inhibitors recently emerged as a promising therapy in neuromyelitis optica spectrum disorder (NMOSD). A few comparative studies assessing its efficacy and safety are currently available. **Objective:** To perform a systematic review and meta-analysis of efficacy and safety endpoints comparing IL-6 inhibitors to placebo or traditional immunosuppressants in patients with NMOSD. **Methods:** We searched PubMed, Embase and Cochrane Central for studies comparing IL-6 inhibitors to alternative therapies in patients with NMOSD. Efficacy endpoints included hazard ratio for relapse and annualized relapse ratio (ARR). Safety outcomes included any adverse event and infection. Statistical analysis was performed using RevMan Web (Cochrane Collaboration) and R package meta. Heterogeneity was assessed with I^2 statistics. **Results:** 4 studies were included (3 randomized controlled trials and 1 retrospective cohort), comprising 361 patients (228 treated with IL-6 inhibitors) with a mean follow-up time of 20.19 (±7.04) months. Mean age was 45 (±13.54) years, 316 (87.53%) were female and 276 (78.45%) had detectable serum anti-aquaporin-4 antibodies. At baseline, mean duration of disease was 5.12 (±3.68) years, mean EDSS was 4.34 (±1.32) and mean ARR was 1.73 (±1.07). Hazard ratio for relapse (hazard ratio 0.35; 95% CI 0.23 to 0.55); p<0.00001; I^2=0%) and ARR (mean difference -0.79 relapses/year; 95% CI -1.54 to -0.03; p=0.04; I^2=96%) were significantly reduced in patients treated with IL-6 inhibitors when compared to those treated with placebo or traditional immunosuppressants. No significant difference was found in any adverse event (odds ratio 1.10; 95% CI 0.67 to 1.79; p=0.71; I^2=0%) and infection (odds ratio 1.59; 95% CI 0.45 to 5.63; p=0.48; I^2=48%) between the treatment groups. Statistically significant results in efficacy endpoints were restricted to IgG-aquaporin-4 seropositive subgroup of patients. **Conclusion:** This meta-analysis indicates that IL-6 inhibitors are superior to placebo and traditional immunosuppressants in reducing relapses in patients with aquaporin-4 seropositive NMOSD, without increasing adverse events or infection rates.

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Interoperable platform for surveillance of amyotrophic lateral sclerosis cases in Brazil

Ingridy Marina Pierre Barbalho[1], Felipe Ricardo dos Santos Fernandes[1], Aleika Lwiza Alves Fonsêca [1], Danilo Alves Pinto Nagem[1], Karilany Dantas Coutinho[1], João Paulo Queiroz dos Santos[1], Igor Braga Farias[2], Emanuela Coriolano Fidelix[1], Mario Emílio Teixeira Dourado Júnior[1], Ricardo Alexsandro de Medeiros Valentim[1]

[1]Universidade Federal do Rio Grande do Norte, Laboratório de Inovação Tecnológica em Saúde, Natal RN, Brazil.
[2]LAIS. Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Mario Emilio Teixeira Dourado Junior; Email: medourado03@gmail.com.

Amyotrophic lateral sclerosis (ALS) is a rare neurodegenerative disease considered complex due to its heterogeneity. Despite being known for many years, few countries have precise information about its epidemiology and the characteristics of individuals diagnosed with ALS. In Brazil, the lack of information about ALS limits the use of data for advancing research and developing public policies that benefit people affected by this health condition. In this context, the objective to present the development of a platform for the management and monitoring of ALS in Brazil in order to provide intrinsic and timely data to support the formulation of public policies while simultaneously improving the care and monitoring of ALS patients. The development of this platform was supported by the Ministry of Health of Brazil. It was conceived in response to the needs identified in the epidemiological scenario related to the disease, as well as the limitations reported by health professionals specialized in treating ALS patients. The developed platform consists of two systems: the Brazilian National ALS Registry, responsible for collecting structured epidemiological data from ALS patients throughout Brazil; and the Electronic Medical Record for ALS patients, responsible for assisting in the multidisciplinary monitoring of ALS patients safely and appropriately, based on the Clinical Protocols and Therapeutic Guidelines (in Portuguese, Protocolos Clínicos e Diretrizes Terapêuticas - PCDT) for ALS. The systems were developed to be interoperable and capable of securely and effectively exchanging information, considering the security and privacy of patient data in accordance with the General Data Protection Law (in Portuguese, Lei Geral de Proteção de Dados - LGPD). The integration of surveillance and monitoring data presents significant benefits for the development of public policies and the planning of strategies at all levels of health care, in addition to providing a realistic view of the country's situation regarding ALS cases. Therefore, this platform is being used as a tool for research, intervention, monitoring, and strengthening the response to ALS within the Brazilian Unified Health System (in Portuguese, Sistema Único de Saúde - SUS).

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Interregional mapping of stroke in Ceará (IMISC): a proposal of a georeferencing methods of stroke analysis in Northeastern Brazil

David Augusto Batista Sá Araújo[1], Emanuel de Assis Bertulino Martins Gomes[1], Sophia Costa Vasconcelos[1], Cássia Caroline Aguiar da Ponte[1], Bruno Nunes Ferraz de Abreu[2], Hellen Carm de Lacerda Homem[2], Fabrício Oliveira Lima[2]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Emanuel de Assis Bertulino Martins Gomes; Email: emanueldeassis@alu.ufc.br.

Background: Stroke is a serious and disabling disease all over the world. It is a serious medical emergency and reperfusion therapies have been developed with increasing technology in recent years. However, treatment is not offered equally in all regions of the world, including Brazil, where there are marked differences in access to healthcare between different regions. **Objective:** To propose a method for georeferencing and mapping the characteristics of stroke in Ceará state, Brazil, using as an initial analysis the distance and travel time profile between the municipal hospitals in the state and a Stroke Unit located in the capital. **Methods:** Choropleth maps were constructed with R software for statistical analysis, using the "gmapsdistance" package, which estimates the distances between the territories on the geographic map. The geolocation data was obtained from Google Maps. To continue the project, data will be collected from reference hospitals in Ceará, both on the sociodemographic nature/comorbidities of the participants and on stroke characteristics, such as the time of symptom onset and treatment time, scales at hospitalization and discharge (e.g. NIHSS and ASPECTS) and clinical outcomes. A database will be created to assess the profile of stroke care in Ceará, including distances and treatment times. With this data, the extent of stroke in the state and the effectiveness of medical care for those patients will be evaluated. **Results:** Travel time and distance estimates were made between the hospitals in the state's municipalities in relation to the Stroke Unit at the Fortaleza General Hospital (HGF), which is a reference in tertiary care in Ceará for stroke patients. It was observed that the average travel time to the HGF varied between 0.3 and 8 hours, not exceeding the limits for treatment in certain circumstances by indication. The distance profile showed a maximum mean distance of 500 kilometers and a minimum of 5 kilometers between those locations. As the HGF is the only hospital that performs mechanical thrombectomy on an emergency basis in the state, the results described also reflect the disparity in access to this procedure. **Conclusion:** The study presents a method of mapping stroke characteristics in Ceará, Brazil, shedding light on the disparities in access to timely stroke treatment. Using choropleth maps and geolocation data, we identified variations in travel time to the Stroke Unit of the HGF, the sole facility offering thrombectomy in Ceará. Our findings reveal that while some areas have access within acceptable timeframes, others face significant delays, highlighting the need for equitable distribution of stroke care resources. By understanding the geographical profile of stroke care, policymakers and healthcare providers can better allocate resources to ensure prompt treatment and improve outcomes for stroke patients across the state.

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Interventional neuroradiology: epidemiological profile and clinical outcomes in Southern Santa Catarina

Fabrício Weinheimer Lippert[1], Camila Pezente Dal Toe[1], Khiany Mathias[1], Richard Simon Machado[2], David dos Santos[1], Ana Paula Benedet de Carvalho[1], Natalli Studnicka[1], Tatiana Pizzolotto Bruch[1], Fabricia Cardoso Petronilho[1], Luiz Pedro Willimann Rogério[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.

Correspondence: Fabrício Weinheimer Lippert; Email: fabriciowlippert@gmail.com.

Background: Interventional neuroradiology is a subspecialty dedicated to the diagnosis and treatment of cerebrovascular diseases and nervous system tumors. It combines neurosurgical and neuroradiological knowledge, enabling minimally invasive techniques. **Objective:** To investigate the epidemiological profile and clinical outcomes of patients at an interventional neuroradiology service in southern Santa Catarina. **Methods:** Cross-sectional analytical observational study with a quantitative approach and primary and secondary data collection. The study population consisted of all patients undergoing

interventional neuroradiology at a hospital in southern Santa Catarina who were followed up at Clínica Neurosul Neuroespecialistas from January 2017 to July 2022, with a sample of 242 patients. Patient profiles, underlying disease characteristics, and clinical outcomes were evaluated. Secondary data were collected from electronic medical records, and primary data were collected via telephone interviews. **Results:** The mean age of the patients was 55 years, with females predominating (61.6%). According to the Rankin scale, 92.1% remained asymptomatic, 2.9% had symptoms without disability, 1.2% had moderate disability, 1.7% had severe disability, and 2.1% of patients died. Aneurysm was the most commonly treated condition, accounting for 31.1% of all cases. Other commonly addressed conditions included ischemic stroke, carotid stenosis, vertebral stenosis, and arteriovenous malformation. Cerebral angiography was the most performed procedure (75.2%), followed by cerebral aneurysm embolization, intracranial thrombectomy, and carotid angioplasty. Complications occurred in 7% of procedures, considering death the most common complication, followed by pneumonia, aphasia, and brain death. Primary data collection showed that the majority of patients were asymptomatic or had headaches before the procedure, with frequencies of 41.2% and 38.2%, respectively. In the postoperative period, 75.8% of patients reported no symptoms, 9.1% reported headaches, and a few patients experienced contrast allergy, temporary vision loss, dizziness, paresthesia, and seizures. **Conclusion:** Interventional neuroradiology is a growing field with safe procedures and good outcomes. As evidenced in this study, it is primarily involved in the treatment of aneurysms, ischemic stroke, carotid and vertebral stenosis, and arteriovenous malformations. However, there is a potential for further utilization in other conditions, such as tumor preoperative embolization, a procedure that was underrepresented in the current study. The study demonstrates safety through the low complication rate of procedures and the fact that 92% of patients remained asymptomatic according to the Rankin scale post-procedure. These findings reflect the interventional neuroradiology excellence and underscore the need for its expanded use in nervous system pathologies.

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Intestinal microbiome and multiple sclerosis: dysbiosis as a trigger or a reflex of the autoimmune response?

Mariana de Oliveira Trintinalha, André Luiz Fagundes Avila dos Santos[1], Erick Guerra[1], Luciane Filla[2], Débora Marques Herzog[1]

[1]Pontifícia Universidade Católica do Paraná, Hospital Universitário Cajuru, Curitiba PR, Brazil.
[2]Pontifícia Universidade Católica do Paraná, Curitiba PR, Brazil.

Correspondence: Mariana de Oliveira Trintinalha; Email: marianaatrintinalha@hotmail.com.

Background: Multiple sclerosis (MS) stands as a primary demyelinating disease and the second leading cause of disability among young adults in developed nations. This immune-mediated neurodegenerative condition manifests through a myriad of symptoms. Various theories exist concerning the disease's etiology and immune system triggers, with a key focus being the investigation into the dysregulation of the intestinal microbiome. **Objective:** This systematic review aims at unraveling the impact of the intestinal microbiome on the pathogenesis of multiple sclerosis. **Methods:** Employing an integrative literature review approach, this study was conducted between August and December 2023. The search utilized the descriptors "Gastrointestinal Microbiome" and "Multiple Sclerosis." Data were extracted from cohorts, meta-analyses, and cross-sectional studies published in English, Portuguese, or Spanish between 2019 and 2023, sourced from the National Library of Medicine National Institutes of Health (MEDLINE/PUBMED). Exclusion criteria included systematic reviews, content diverging from the microbiome focus, books, protocols, theses, conference publications, and languages not specified. Initial findings yielded 13 articles, with 11 meeting the criteria for analysis. **Results:** The composition of the intestinal microbiome is notably influenced by dietary patterns and disease-modifying therapies, as evidenced by their direct impact on the microbial populations within the gut. Research indicates an association between the development of Multiple Sclerosis and alterations in bacterial populations, such as an increase in Streptococcus and Akkermansia coupled with marked reductions in Faecalibacterium, Bacteroides, Roseburia, and Prevotella. Furthermore, different microbiome patterns were observed based on the different subtypes of Multiple Sclerosis developed by patients. This dysbiosis is linked to individual patient characteristics, including age, antibiotic usage, gender, comorbidities, diet, and probiotic intake. Studies on rodents have demonstrated that fecal transplants can mitigate microglial and astrocytic activation, prevent demyelination, and axonal loss. Additionally, research showcases the immunomodulatory effects of Short Chain Fatty Acids (SCFAs) and tryptophan metabolites originating from the microbiome, facilitating the suppression of autoimmune responses through augmenting T regulatory cell (Tregs) populations. **Conclusion:** Emerging evidence proposes pivotal roles for microbiomes and intestinal immunity in the onset of autoimmune conditions. The comprehension of the functional impact of bacterial populations on the progression of Multiple Sclerosis remains limited. Dietary interventions and disease-modifying therapies might offer benefits in MS management by modulating the microbiome towards an anti-inflammatory composition.

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Intracranial hemorrhage in Brazil

Deborah Araujo Silva[1], Rynele Almeida de Fonseca[1], Larissa Bicanca Borsone[1], Annayza de Oliveira Bessa[1], Amanda Oliva Spaziani[1], Joao Carlos Bizinotto Leal de Lima[1], Gustavo Henrique da Silva[2], Vanessa Maria Gonçalves de Souza[1], Luis Carlos Spaziani[3], Francieli Gomes Lima[4]

[1]Universidade Brasil, Fernandopolis SP, Brazil.
[2]UPA Santa Fé do Sul,Santa Fé do Sul SP, Brazil.
[3]Fundação Educacional de Fernandopolis, Fernandopolis SP, Brazil.
[4]AME Fernandopolis, Fernandopolis SP, Brazil.

Correspondence: Deborah Araujo Silva; Email: araujodeborah@outlook.com.

Background: Intracranial hemorrhage is characterized by initial bleeding within the brain parenchyma, potentially affecting the meningeal space and subsequently the ventricles. It is one of the most significant traumatic conditions due to its severity and potential for neurological impairment, being a leading cause of morbidity and mortality in Brazil. This event is more common in the elderly population, as this age group is more susceptible to falls due to osteomuscular and neural mechanisms involved in maintaining posture. **Objective:** To evaluate the epidemiological profile of intracranial hemorrhage in Brazil between 2020 and 2022. **Methods:** Data collection was performed using publicly available information from the Hospital Information System of SUS (SIH/SUS) in the Tabnet/DATASUS database between June 1 and August 10, 2023. This retrospective longitudinal study employed a quantitative descriptive approach. Data were grouped by age and educational level for the period from 2020 to 2022. Statistical inference was conducted using BioEstat 5.3 software, utilizing the paired T-test for group comparisons, and data presentation included descriptive statistics, measures of central tendency, and dispersion. **Results:** Between 2020 and 2022, there were 91,821 cases of intracranial hemorrhage in Brazil. The highest number of cases was concentrated in the Southeast region (n=13,573), accounting for 44.34% of the total, with annual treatment costs averaging approximately R$ 64,356,563.46. The Southeast region also had the highest number of deaths, accounting for 47.74% (n=10,410) of the total, with a mortality rate of 25.57. The number of hospitalizations varied over the years, with 33,093 cases (36.04%) occurring in 2022 compared to 30.99% (n=28,459) in 2020. Regarding the classification of care, 94.19% (n=86,486) of cases were urgent, while 5.81% (n=5,335) were elective. In terms of sex, 51.81% (n=47,576) of the cases were male and 48.19% (n=44,245) female (test t p=0.0679). Elderly individuals aged 60 to 69 were most affected, accounting for 22.12% (n=20,313) of cases, followed by those aged 50 to 59 at 21.83% (n=20,040). Regarding race/ethnicity, 39.84% of the cases were self-declared as mixed race (n=36,586) and 32.88% as white (test t p=0.1191). **Conclusion:** Intracranial hemorrhage cases are predominantly concentrated in the Southeast region of Brazil, which accounts for the highest proportion of deaths and treatment costs, associated with the highest mortality rate among regions. The majority of cases occur among men aged 50 to 69 years, who are self-declared as white or mixed race, with urgent care being the most prevalent type of hospitalization.

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Intracranial hypotension secondary to dural fistula and complicated in cerebral venous thrombosis (CVT)

Victor Cardoso de Faria[1], Andre Felipe Candêas Amorim[1], Gabriel Pinheiro Martins de Almeida e Souza[1], João Victor Cabral Correia Férrer[1], Márcio Pinheiro Lima[1], Ruan Gambardella Rosalina de Azevedo[1], Adriel Rêgo Barbosa[1], Rafael Tuzino Leite Neves Maffei[1], Lucas de Oliveira Cantaruti Guida[1], Enedina Maria Lobato de Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Victor Cardoso de Faria; Email: victor_cardoso13@hotmail.com.

Case presentation: A 53-year-old woman, previously diabetic and with degenerative spine disease, started a new, frontal, oppressive, mild headache 5 months before admission, which partially improved with analgesics, without nausea, vomiting, photophobia or phonophobia. There was no worsening of pain with decubitus or orthostasis, as with Valsalva. Two days before admission, she developed a headache that progressed to very severe intensity within a few minutes, left sided, radiating to the occiput, associated with nausea and vomiting, which worsened markedly when assuming an upright position and improved in supine. She denied auditory or visual symptoms. On neurological examination, she showed no changes, fundus with well-defined papillae. A head CT was performed with a venous study, showing CVT of the transverse sinuses and confluence of the sinuses, as well as a small frontal subdural hematoma. Investigation progressed for intracranial hypotension, brain MRI showed diffuse pachymeningeal enhancement associated with brain sagging. Lumbar puncture was performed with opening pressure of 6 mmHg, cellularity 3, 10 red blood cells. Myelotomography revealed extrathecal contrast next to the interspinous foramina at multiple cervical and thoracic levels, which may correspond to perineural diverticula/cysts associated with CSF leakage. The diagnosis of spontaneous intracranial hypotension secondary to multiple spinal CSF leaks was confirmed. The patient underwent conservative treatment associated with full anticoagulation, given the difficulty in approaching the multiple lesions with a blood patch and/or surgery. He was discharged with complete clinical improvement. **Discussion:** Spontaneous intracranial hypotension requires for its diagnosis the presence of CSF pressure lower than 6 cm H2O and/or evidence of CSF leak on imaging examination, according to ICHD-3. Patients may present with atypical initial presentations, such as cochleovestibular manifestations, cranial nerve

palsy, personality changes, or even movement disorders. Cases may present with thunderclap headache, patients who developed complications such as subdural hematoma or cerebral venous thrombosis may present a change in the headache pattern, with loss of the typical postural component. Trauma, hypermobility and osteoarthritis of the spine are risk factors. Myelotomography is considered the gold standard in locating spinal CSF leaks. Treatment options are a conservative approach, blood patch (early or if conservative treatment fails) or surgery (refractory cases). **Final comments:** Intracranial hypotension is an important secondary cause of headache, associated with relevant morbidity and mortality and a specific therapeutic approach. This is an underdiagnosed disease and should also be a differential diagnosis for patients with headaches that are daily from onset, refractory to everything and chronic headaches that do not fit into a specific phenotype.

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Intraoperative electrocorticography (ECOG) tailored resection in nonlesional epilepsy and focal cortical dysplasia

Fernanda Schuh Martins[1], William Alves Martins[1], Rafael Paglioli[1], Thomas Frigeri[1], Eliseu Paglioli[1], André Luis Fernandes Palmini[1], Ricardo Paganin[1]

[1]Universidade Católica do Rio Grande do Sul, Hospital São Lucas da Pontifícia, Porto Alegre RS, Brazil.

Correspondence: Fernanda Schuh Martins; Email: fschuhmartins@gmail.com.

Background: Electrocorticography (ECoG) has long been used to guide the resection of epileptic tissue. Acute ECoG offers advantages such as cost-effectiveness, the ability to tailor resections, and the assessment of residual epileptic foci. While ECoG is proven to provide crucial information for tailoring resections in focal cortical dysplasias (FCD), its role in nonlesional patients is less well established. **Objective:** This study investigates the usefulness of ECoG in patients undergoing epilepsy surgery with either nonlesional or FCD-positive MRI. **Methods:** Ninety-four patients from the Porto Alegre Epilepsy Surgery Program (PAESP) were included. All underwent ECoG-tailored surgery from 2012-2019 and had at least 12 months of follow-up. Intraoperative ECoG recordings were performed using long-range, carbon or gold-plated ball-pointed 16-electrode sets, allowing for acute probing of electrical activity from the surface, lateral banks, and depths of gyri. Recordings lasted between 15 and 30 minutes and were performed sequentially at each resection stage. Electrode positioning was customized at each stage under direct visualization of the cortical regions being explored. **Results:** Electrocorticographic epileptiform findings were classified as: 1) continuous/quasi-continuous spikes - rhythmic and frequent bursts of spikes recurring at 0.5-2Hz; 2) recurrent rhythmic polyspikes or burst-suppression - bursts of at least 3 spikes or fast activity followed by attenuation; 3) recruiting ictal rhythm (EEG seizure); 4) frequent non-rhythmic spikes - more than 10 epileptiform spikes/sharp waves per minute; 5) sporadic spikes - fewer than 10 epileptiform spikes/sharp waves per minute. ECoG patterns 1-3 were considered dysplastic and were more commonly found in extratemporal epilepsies (81% vs. 59%, p=0.024). Complete resection of the most active ECoG region significantly improved outcomes, with an OR of 9.5 (CI 95% 2.4-37) for becoming seizure-free (61% vs. 14%, p=0.001). FCD II was found in 21/26 patients (81%) with continuous/quasi-continuous patterns (ECoG 1) and in 7/15 (47%) with recurrent rhythmic polyspikes (ECoG 2), whereas FCD I was seen in 4/11 patients (30%) with frequent non-rhythmic patterns (ECoG 4) and in 5/21 (24%) of those with sporadic spikes (ECoG 5) [p=0.017]. In non-lesional patients, ECoG most often showed sporadic spikes (14 [35%]) and frequent non-rhythmic spikes (11 [27%]), whereas those with lesional MRI had higher rates of continuous/quasi-continuous spikes (23 [40%]) and bursts of polyspikes/burst-suppression (10 [18.2%]; p=0.023).**Conclusion:** ECoG added significant value in tailoring resections in both lesional and nonlesional cases. This surgical strategy provided a reasonable chance of resecting the epileptogenic zone even in patients with nonlesional MRI. However, the pattern of sporadic spikes is common in nonlesional patients, appearing in 35% of intraoperative ECoG recordings, which can complicate the delimitation of the epileptogenic zone.

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Intrathecal pemetrexed for leptomeningeal carcinomatosis in EGFR-mutated NSCLC with severe renal impairment

Thales Pardini Fagundes[1], Amélia Gontijo Velozo de Melo[1], Ingrid Marriel Ramos Novais[1], Victoria Almeida Freire[1], Roberta Martins Queiroz Barbosa[1], Beatriz Olmo Salles[1], Gustavo Dix Junqueira Pinto[1], Carlos Eduardo Baston Silva[1], Flavio Augusto Ferreira da Silva[1]

[1]Fundação Pio XII, Hospital de Câncer de Barretos, Barretos SP, Brazil.

Correspondence: Thales Pardini Fagundes; Email: pardinithales@gmail.com.

Case presentation: A 68-year-old woman with EGFR exon 19-deleted non-small cell lung cancer (NSCLC) developed leptomeningeal disease (LD) after seven years of treatment, including gefitinib and osimertinib. With severe renal dysfunction (creatinine clearance 18 ml/min/m^2), we administered intrathecal (IT) pemetrexed 50 mg on days 1 and 21, with IT dexamethasone, alongside daily osimertinib 80 mg. Treatment was well-tolerated without increased toxicity. Cerebrospinal fluid analysis (CSF) showed an increase in malignant cells from 30% to 36%, and brain MRI revealed communicating hydrocephalus. The regimen continued every 28 days, with close monitoring of safety and efficacy. **Discussion:** This case represents the first reported use of IT pemetrexed in an NSCLC patient with severe renal impairment. LD in EGFR-mutated NSCLC poses significant treatment challenges, even with osimertinib's CNS efficacy. Previous studies have shown IT pemetrexed's efficacy in LD, with doses of 10-50 mg weekly demonstrating response rates of 43.5-80.3%. However, data on renal dysfunction is lacking. Our experience suggests that IT pemetrexed can be safely administered to patients with severe renal impairment without increased toxicity. This finding is crucial, as pemetrexed is predominantly renally cleared, with intravenous administration contraindicated for creatinine clearance under 30 ml/min. The absence of established dose adjustments for IT administration in renal impairment underscores the significance of our case. Despite combining IT pemetrexed with osimertinib, clinical benefit was modest in our heavily pretreated patient. This outcome highlights the need for earlier intervention, novel combinatorial approaches, and individual dosing based on CSF flow dynamics to improve outcomes in LD. Comments: This case expands the limited data on IT pemetrexed in LD with severe renal dysfunction. While our patient tolerated the treatment well, efficacy was limited, possibly due to late-stage disease. Prospective trials are needed to establish the safety, efficacy, and optimal dosing of IT pemetrexed in this challenging population. Our experience opens new avenues for managing LD in EGFR-mutated NSCLC patients with renal impairment, a group often excluded from standard treatments. It underscores the potential of IT chemotherapy in overcoming systemic limitations and targeting the CNS directly. **Final comments:** Future research should focus on earlier intervention, novel drug combinations, and personalized approaches to improve outcomes in this aggressive disease manifestation. This case serves as an important step forward, demonstrating that innovative approaches can expand treatment options for patients with limited choices. It emphasizes the importance of individualized medicine and the need for continuous exploration of new strategies in managing complex cancer presentations.

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Intravenous delivery of human dental pulp stem cells provides neuroprotection and attenuates disease progression in a murine model of amyotrophic lateral sclerosis

Alexandre Hilário Berenguer de Matos[1], Lilian de Oliveira Coser[2], Rodrigo Pinheiro Araldi[3], Luciana Politti Cartarozzi[2], Cristiane Wenceslau Valverde[4], Alexandre Leite Rodrigues de Oliveira[2], Irina Kerkis[5], Marcondes Cavalcante França Jr.[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Departamento de Neurologia, Campinas SP, Brazil.
[2]Universidade Estadual de Campinas, Instituto de Biologia, Departamento de Biologia Estrutural e Funcional, Campinas SP, Brazil.
[3]BioDecision Analytics Ltda., São Paulo SP, Brazil.
[4]Cellavita Pesquisas Científicas Ltda., Valinhos SP, Brazil.
[5]Instituto Butantan, Laboratório de Genética, São Paulo SP, Brazil.

Correspondence: Alexandre Hilário Berenguer de Matos; Email: alexandrehbm@gmail.com.

Background: Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disorder characterized by the progressive degeneration of motor neurons, leading to debilitating muscle weakness, atrophy, and paralysis. Excitotoxicity plays a pivotal role in the neuronal death and dysfunction observed in ALS. Human dental pulp stem cells (hDPSCs) exhibit anti-inflammatory, anti-apoptotic, and neurotrophic properties, suggesting potential therapeutic benefits for ALS. **Objective:** To investigate the therapeutic potential of intravenous delivery of hDPSCs in a murine model of ALS (SOD1G93A mice). **Methods:** This was a case-control study with animals split into 3 experimental groups (hDPSC, n=7; vehicle, n=5; non-transgenic, n=5). To track the distribution of transplanted cells throughout the spinal cord, hDPSCs were labeled with Qtracker 655 (Thermo Fisher) and observed twenty-four hours after transplantation. The treatment was carried out in the asymptomatic phase of the disease (10th week) by a single systemic application of hDPSCs (1x106 cells/kg). The animals were euthanized (n=5) at the 15th week (initial stage of symptoms) of the disease and the lumbar spinal cords were dissected and processed for Nissl staining (neuronal survival) and immunofluorescence (astrocytosis and microglial reactions). Behavioral analyses considering the onset of disease and its progression, neurological score, body weight, and motor control (rotarod test) started on the 10th week and were performed every week until dissection. **Results:** Transplanted hDPSCs were detected throughout the lumbar spinal cord 24 hours after intravenous injection, primarily within the gray matter's Rexed lamina IX, indicating successful target delivery. This treatment resulted in significantly greater neuronal survival (83% vs 27%, p<0.05) compared to the vehicle-treated group, demonstrating a neuroprotective effect. In addition to promoting neuronal survival, the cell therapy also demonstrated a beneficial effect on glial responses. Astrogliosis and microglial reactivity were significantly reduced in the treated group (p<0.05). These findings are aligned with the observed behavioral data, where hDPSC-treated animals had delayed motor deficits and onset the disease, and reduced weight loss compared to the vehicle group. **Conclusion:** Overall, our data demonstrate the therapeutic potential of hDPSCs in a murine model of ALS. Intravenous delivery enhanced neuronal survival, reduced gliosis, and improved structural preservation of the ventral horn. These effects were associated with delayed disease progression, as evidenced by attenuated

motor deficits, reduced weight loss, and a later onset of disease symptoms in treated animals. Thus, our preclinical findings provide evidence that this stem cell lineage may be a promising therapeutic strategy for ALS.

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Intravenous thrombolysis in a patient with embolism in the left middle cerebral artery following recent postoperative osteosynthesis of the distal radius and thoracic spine arthrodesis after severe spinal cord trauma

Maria Júlia Branco Rodrigues[1], Paulo Victor Machado[1], Marcos Seefeld[1], Frederico Otto Flores Moraes[2], João Pedro Gorski Ribas de Araujo[2], João Pedro Greca de Macedo Biasi[2], Guilherme Costa Barghouthi[2], Ana Paula Gonçalves Barankiewicz[1], Lucas Rodrigues Prim[1], Lisseth Fabiola Vallejo Reyes[1]

[1]Hospital Universitário Evangélico Mackzenie, Curitiba PR, Brazil.
[2]Faculdade Evangélica Mackenzie do Paraná, Curitiba PR, Brazil.

Correspondence: Paulo Victor Santos Machado; Email: paulovictorsmachado@gmail.com.

Case presentation: A 32-year-old male patient, without comorbidities, suffered polytrauma from a motorcycle-car collision, resulting in a spinal cord injury (SCI), classified as Frenkel A. He underwent osteosynthesis of the right distal radius and thoracic spine arthrodesis. Despite surgical correction, he maintained paraplegia with a sensory level at T9. On the 10th postoperative day, he experienced an acute focal deficit, witnessed by the medical team, and was transferred to a referral hospital. Upon admission, the patient presented with motor aphasia, in addition to deficits secondary to severe SCI, with an admission NIHSS score of 2 (aphasia). Cranial and cervical computed tomography angiography showed cortical and subcortical hypodensity in the left temporal region, suggestive of an ischemic stroke (IS) in the left middle cerebral artery (MCA), with an ASPECTS score of 10. Despite absolute contraindications to chemical thrombolysis, such as two recent surgeries (<14 days), the patient underwent the procedure. After thrombolysis, the patient showed complete resolution of aphasia, with no bleeding at any point. Two days after the IS, the patient developed deep vein thrombosis (DVT) in the right lower limb, and anticoagulation with enoxaparin was initiated. One day after this diagnosis, he developed fever spikes and tachycardia, and was diagnosed with bilateral pulmonary embolism (PE), confirmed by imaging, despite full anticoagulation. An etiological investigation of the recurrent ischemic events was conducted, including a transesophageal echocardiogram with bubble test, which revealed a patent foramen ovale (PFO) with significant flow. Additionally, thrombophilias were investigated and ruled out. To determine the etiology of the IS, ROPE and PASCAL scores were used, both scoring 9 points, suggesting a high probability that the ischemic events were secondary to the PFO. After 21 days, the patient was discharged with rivaroxaban. **Discussion:** This case involves a severely polytraumatized patient with no known risk factors for thrombotic events, prompting clinical reasoning to find the etiology of the embolic occurrences. Despite being contraindicated, thrombolysis completely reversed the deficit without hemorrhagic complications, opening a discussion about the recurrence rate of embolic events post-thrombolysis in patients with absolute contraindications for such a procedure. **Final comments:** This case highlights the complexity of managing a polytraumatized patient and its developments in a patient with a previously undiagnosed PFO, who experienced multiple embolic events. The identification of the neurological deficit and the decision for immediate thrombolysis, with the consent of the patient and family, successfully reversed the deficit. These facts underscore the necessity of following a clinical reasoning process to reach a diagnosis. The favorable recovery demonstrates the efficacy of an integrated and thoughtful approach.

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Invasive fungal rhinosinusitis: a case report

Érica Paiva Espinosa[1], Isadora Rutkoski Didio[2], Bruno Gernhardt Teneos[3], Isabela Just de Jesus Vanni[2], Yasmin de França[3], Julia Carolina Barbosa Covre[2]

[1]Universidade Católica de Pelotas, Pelotas RS, Brazil.
[2]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.
[3]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Érica Paiva Espinosa; Email: erica.espinosa@sou.ucpel.edu.br.

Case presentation: Aggressive infections such as invasive fungal rhinosinusitis have been most frequently associated with immunocompromised patients, as those with hematologic malignancies, advanced HIV disease, or diabetes mellitus. Upper respiratory tract infections (URTIs) are mostly caused by viruses and bacteria, rarely evoked by funguses. The most prevalent fungal agents responsible for colonizing the upper respiratory tract are Aspergillus, Fusarium, and Mucorales. Clinical manifestations of invasive fungal rhinosinusitis include acute fever, facial pain, nasal congestion, visual alterations, and mental confusion. This report aims to present a rare case report of a young male patient presenting invasive fungal rhinosinusitis caused by Rhizopus oryzae, with important central nervous system (CNS) impairment and surgical outcome. **Discussion:** A 10 year-old-male, from Rio Pardo-RS, developed polyuria, abdominal pain, progressive asthenia, and headache in March 2024, leading to a diagnosis of diabetic ketoacidosis and diabetes mellitus type 1. Additionally, the patient showed signs of nasal congestion, which led us to the sinusitis diagnosis. He developed fungal meningitis, aggressive behavior and agitation, in addition to eyelid edema and visual acuity loss. He was therefore referred to neurosurgical intervention due to suspicion of cerebral edema and optic neuritis. Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) scans indicated signs consistent with fungal infection that had spread to the nasal cavities and CNS. Furthermore, the complication identified included fusiform pseudoaneurysm of the anterior cerebral artery and acute ischemic events. The diagnosis of invasive fungal rhinosinusitis was confirmed after debridement and positive culture for Rhizopus oryzae complex. After craniotomy for brain abscess, the patient still showed signs of complications on imaging tests, such as liquid and gas collections, signs of vasculitis and reduced caliber of the left internal carotid artery. **Final comments:** In this case report, we describe a rare manifestation of invasive fungal rhinosinusitis caused by Rhizopus oryzae involving the central nervous system (CNS) of an immunocompromised patient with type 1 diabetes mellitus. This patient exhibited extensive clinical manifestations of fungal infection, including ocular lesions, cerebral vasculature invasion, and bifrontal granulomatous fungal abscesses. All efforts were made to protect the patient's privacy and confidentiality.

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Investigating proteomic and metabolomic signatures in mesial temporal lobe epilepsy: insights into neurotransmitter dynamics and pharmacoresistance

Amanda Morato do Canto[1], Jaqueline Cruz Geraldis[1], Alexandre Barcia de Godoi[1], Fábio Rogério[1], Marina Koutsodontis Machado Alvim[1], Clarissa Lin Yasuda[1], Enrico Ghizoni[1], Matthew Macdonald[2], Fernando Cendes[1], Iscia Lopes-Cendes[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Campinas SP, Brazil.
[2]University of Pittsburgh, Pittsburgh, USA.

Correspondence: Amanda Morato do Canto; Email: amcanto@unicamp.br.

Background: Mesial temporal lobe epilepsy (MTLE) is the most prevalent form of focal epilepsy in adults, posing significant clinical hurdles due to a high resistance rate to standard antiseizure medications. Delving into the intricacies of synaptic transmission, particularly its processes and potential abnormalities, is pivotal for comprehending the mechanisms underpinning MTLE. Synaptosomes, housing essential machinery for neurotransmitter release, reuptake, and storage, offer a unique window into synaptic function. **Objective:** This study explores the proteomic and metabolomic profiles of synaptosomes isolated from the anterior temporal lobe and hippocampus of patients with pharmacoresistant MTLE and hippocampal sclerosis. **Methods:** In this study, we employed a unique approach to investigate synaptosome proteomic and metabolomic profiles. These synaptosomes were isolated from hippocampal and anterior temporal lobe tissue obtained during epilepsy surgery, including samples from pharmacoresistant MTLE patients (N=20) and post-mortem controls (N=5). Our data acquisition was performed using cutting-edge technologies, including an Orbitrap EclipseTM TribridTM mass spectrometer for proteomic data and an LCMS-9030 quadrupole time-of-flight (Q-TOF) Shimadzu instrument for metabolomic data. The subsequent bioinformatics analysis, conducted using ProteomeDiscoverer and R software, was complemented by SynGO, STRING PPI, and ClueGO analyses to investigate biological alterations. **Results:** Our study revealed significant variations in protein and metabolite compositions among synaptosomes from different brain structures, underscoring the complexity of MTLE pathology. Notably, patient synaptosomes showed a preponderance of presynaptic proteins, with observed alterations in neurotransmitter release cycles and receptor dynamics. These findings, along with evidence of increased sensitivity to toxic substances, suggest potential implications for drug metabolism within the hippocampus of patients with pharmacoresistant MTLE. **Conclusion:** Our study has delineated distinct protein and metabolite profiles within synaptosomes from various brain structures in patients with pharmacoresistant MTLE. The predominance of presynaptic proteins and observed alterations in neurotransmitter release cycles and receptor dynamics highlight the complex molecular underpinnings of MTLE pathology. These findings advance our understanding of MTLE pathogenesis and provide valuable insights into potential therapeutic targets and avenues for further investigation. By shedding light on the molecular landscape associated with pharmacoresistant MTLE, our results may improve management strategies for these patients, potentially leading to the development of novel therapeutic approaches.

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Irreversible bilateral ischemic optic neuropathy in the post-operative myocardial revascularization setting: case report

Pâmela Ritzmann de Lima[1], Laís Cristina Marques Moura[1], Brenda Jahn Henning[1], Patrícia de Morais Della Justina[1], Sabrina Cristof[1], Oscar Nelson Reimann[2], Michael Ricardo Lang[2], Felipe Negrão[2], Chelin Auswaldt Steclan[3], Diogo Pasquali Nones[2]

[1]Universidade do Contestado, Mafra SC, Brazil.
[2]Hospital São Vicente de Paulo e Grupo Neuromax, Mafra SC, Brazil.

[3]Universidade Federal de Santa Catarina, Florianópolis SC, Brazil.

Correspondence: Pâmela Ritzmann de Lima; Email: pamelaritzmann@gmail.com.

Case presentation: Male, 71 years old, white, hypertensive, overweight, with severe coronary insufficiency, heart failure (HF), sleep apnea, dyslipidemia, post-myocardial revascularization surgery (without complications) with extracorporeal circulation (ecc). In the immediate postoperative period (PO), he was sent to the intensive care unit (ICU) due to spikes in arterial hypotension, desaturation (requiring norepinephrine) and anemia. On the 4th PO, in the ward, he developed decreased visual acuity and subsequent loss of complete vision (5th PO), without reaction to light. brain tomography ruled out a stroke, while magnetic resonance imaging (MRI) revealed hypoglycemic encephalopathy, cause of cortical blindness due to amaurosis; by retinography, peridiscal micro-hemorrhages were evident, with a bilateral atrophic area. He was discharged on the 10th PO, with the diagnostic hypothesis of hypoglycemic encephalopathy. However, without showing any improvement in the condition, on the 11th day after vision loss, an eye fundus examination and fluorescein angiography revealed: edema, pallor of the papilla, signs of hypo fluorescence and candle flame hemorrhage, considering hypothesis of anterior ischemic optic neuropathy (AION). On the 17th day after amaurosis, skull MRI showed nonspecific changes in optic nerves, corroborating NAION. Inflammatory tests were conducted, with results within normal limits. Pulses with methylprednisolone (3x/day) were then performed, however, without improvement. On the 50th day after visual loss, a new fundus examination revealed: pallor and reduction in papillary edema; optic nerve atrophy; confirming bilateral non-arteritic AION (NAION). After one year of PO, the patient developed a diagnosis of total blindness, remaining in bilateral amaurosis and without perception of light. **Discussion:** Perioperative visual loss is a rare and feared complication in the postoperative period of non-ophthalmic surgeries, which can result in serious and irreversible damage. The patient, here in the PO of cardiac surgery, had preoperative risk factors (comorbidities), which in addition to postoperative risk factors (hemodynamic decompensation and use of vasoactive drugs) contributed to NAION. This condition is related to optic nerve infarction, resulting in hypoperfusion and ischemia. It has been shown that NAION is a diagnosis of exclusion, which makes its diagnosis difficult. Treatments do not have good evidence of effectiveness so far, so the prognosis tends to be darker, considering the prevention of risk factors essential. **Final comments:** Recognizing the previous clinical history, immediate anatomical and etiological diagnosis is essential to guide the diagnosis, management, and prognosis of NAION. Case reports like this tend to contribute to the recognition of rare complications, decentralizing health information, as well as its applicability.

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Irreversible paraplegia following benzathine penicillin injection: a case report

Monique Melo Fortaleza[1], Felipe Melo Leal[2], Victor José Campelo Vilanova[1], Janine Midori Figueiredo Watanabe[1], Fabbricio Santos Castelo Branco[3], Tibério Silva Borges dos Santos[3], Cristina Reis de Sousa Brito Rocha[2], Jonh Heberthon de Souza Araújo[1]

[1]Universidade Estadual do Piauí, Teresina PI, Brazil.
[2]Universidade Federal do Piauí, Teresina PI, Brazil.
[3]Hospital Getúlio Vargas, Teresina PI, Brazil.

Correspondence: Monique Melo Fortaleza; Email:

Case presentation: A 30-year-old male with no comorbidities or prior allergies presented to a local hospital with fever and odynophagia, where he received a dose of benzathine penicillin in the left gluteal region. Eight minutes later, he developed paralysis and cyanosis of the lower limbs, abdominal pain, melena, and hematuria. He was promptly admitted to intensive care, exhibiting signs of tissue injury at the injection site, paresthesia, non-fixed cyanosis, and livedo on the lower limb extremities, with amputation under consideration. His condition deteriorated presenting anuria and decline in overall condition. Two weeks later, he was transferred to a referral hospital where hemodialysis was initiated, leading to the restoration of diuresis, and had no further episodes of melena. In the ward, ten days later, he continued to experience paraplegia and developed ecchymotic patches on the trunk, with improvement in paresthesia and cyanosis. In light of the suspicion of transverse myelitis, he underwent thoracolumbar spine MRI, confirming hypersignal in T11-L1, electroneuromyography showing severe plexopathy, in addition to laboratory evaluation, serological tests, venous Doppler ultrasound, and lumbar puncture, ruling out other etiologies. Given the symptoms and their temporal association with a medication injection, the diagnosis of Nicolau syndrome was established. The patient was discharged with a catheter, experiencing dual sphincter incontinence and no motor improvement, for physiotherapy follow-up. **Discussion:** Transverse myelitis is characterized by acute or subacute spinal cord injury causing motor, sensory, and autonomic alterations. It is estimated to affect approximately 1 to 4 new cases per million individuals annually, with no clear genetic or gender predisposition. Etiologically, it can be autoimmune, infectious, drug-induced, and rarely associated with medication injection, particularly non-steroidal anti-inflammatory drugs or benzathine penicillin, precipitating the complication known as Nicolau syndrome. This clinical entity involves skin, subcutaneous, and muscular tissue reactions, attributed to medication embolism or arterial vasospasm due to intra-arterial injection. Diagnosis is clinical and exclusionary, and although it is more common in women and obese individuals, the symptoms align with the patient's clinical picture, including: livedoid vasculopathy, a pathognomonic sign; renal failure from rhabdomyolysis; gastrointestinal bleeding; hypoesthesia and paraplegia caused by plexopathy and myelopathy. There is no established standard treatment for the syndrome, largely due to its low incidence. **Final comments:** Although Nicolau syndrome is rare, it is extremely harmful, emphasizing the need for healthcare professionals to exercise vigilance in its prevention. This includes selecting injection sites in the gluteal region that have smaller-caliber vessels, given the frequent administration of medications in this area.

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Is artificial intelligence ready to teach epilepsy through images?

Felipe Guimarães Kanda[1], Luma Hussein Salem[1], Raquel Guimarães Kanda[2], Rafael Guimarães Kanda[3], Paulo Afonso Medeiros Kanda[4]

[1]Universidade de Taubaté, Taubaté SP, Brazil.
[2]Universidade Federal do Cariri, Barbalha CE, Brazil.
[3]Universidade Estadual de Campinas, Campinas SP, Brazil.
[4]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Felipe Guimarães Kanda; Email: felipekanda99@gmail.com.

Background: Artificial intelligence (AI) has shown significant potential in various areas of medicine, including neurology. With the increasing availability of data and advances in machine learning algorithms, the question arises: is AI ready to contribute to the teaching and understanding of epilepsy? Epilepsy is a complex neurological condition characterized by recurrent episodes of seizures, the understanding of which is essential for healthcare professionals, patients, and their families. This study investigates the feasibility of using AI to generate illustrative images of epileptic seizures from descriptions provided in command prompts. **Objective:** To explore the ability of artificial intelligence, specifically language models like GPT-3.5, in conjunction with AI-assisted design tools to generate illustrative images of epileptic seizures from textual descriptions and base images. We aim to assess the quality and usefulness of these generated images in facilitating the understanding of epilepsy by healthcare professionals, patients, and families. **Methods:** We used the GPT-3.5 language model to generate command prompts based on descriptions of different types of epileptic seizures, including physical, behavioral, and temporal characteristics. Image Generation: We tested AI image generators (Copilot, Getimg.ai, bing.com, shakker, aigenerator) apps to translate the command prompts into illustrative images of epileptic seizures. **Results:** The generated images were evaluated by a neurophysiologist, PhD MD titular member of the Sociedade Brasileira de Neurofisiologia Clínica (SBNC) to determine images accuracy, clarity, and utility in an educational context. Results Preliminary results indicate that the combination of GPT-3.5 and AI image generators were able to generate illustrative images of epileptic seizures visually informative and similar to reality. Nevertheless, some images were bizarre, inaccurate and deceptive. Besides, in all cases, the research team modified and adjusted GPT-3.5 prompt, while testing image generation, until images were minimally useful. **Conclusion:** This study demonstrates the potential of artificial intelligence to assist in the teaching and understanding of epilepsy. The generation of illustrative images from textual descriptions offers an innovative and accessible approach to the study of epilepsy. However, further research is needed to validate the accuracy and effectiveness of these images in clinical and educational contexts. The ongoing advancement of AI promises to further improve our ability to teach and manage complex neurological conditions such as epilepsy.

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Is Lhermitte's sign in the first acute episode associated with earlier diagnosis of multiple sclerosis?

Felipe Asaf Borges Millard[1], Thaís Barbosa de Oliveira[1], Antonio Lázaro Santos Soares Junior[1], Julia Xavier Oliveira[1], Vinícius Bessa Mendez[1], Gabriel Souza Suzart[1], Pedro Antonio Pereira de Jesus[1,2], Bruno Bacellar Pedreira[2], Thiago Gonçalves Fukuda[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Felipe Asaf Borges Millard; Email: felipemillard@ufba.br.

Background: Multiple sclerosis (MS) is among the most common causes of neurological disability in young adults worldwide. Among its variety of symptoms, Lhermitte sign (LS) is a sudden electric shock sensation in the cervical region, usually when flexing the neck. Despite being described in other myelopathies, LS is more commonly described in MS, a fact that raises questions about its clinical and diagnostic significance for MS. The influence of LS in time-to-diagnosis of MS is unknown. **Objective:** To evaluate the association between the manifestation of LS in the first acute episode of the disease and the time between the first visit to the doctor and the definitive diagnosis of MS. **Methods:** Cross-sectional study with patients diagnosed with MS (according to the McDonald 2017 criteria), who were followed up at a neuroimmunology reference center in Bahia, from January 2019 to June 2024. The presence of LS in the first acute episode and the period between the search for the first doctor until the definitive diagnosis of MS were reported by clinical interview. The inclusion criteria were patients with definitive diagnosis of MS and without report of flare-up in the last three months until admission. The Mann-Whitney test was used to compare the period from the first medical consultation to the definitive diagnosis between individuals with LS+

and LS-. **Results:** A total of 203 patients were included in the study, of whom 141 (69.5%) were female, the mean age of the sample was 41.6 years (SD±14) and the most frequent ethnicity was brown (47.3%), followed by white (26.1%) and black (25.1%). The most frequent level of education was complete higher education (33.3%), followed by complete secondary education (30.5%) and incomplete higher education (14.3%). The most commonly described clinical form of MS was relapsing-remitting, with 176 individuals (86.5%). LS was present in 60 (29.6%) patients in their first acute illness. The median time between seeking the first doctor and the definitive diagnosis of MS was not different between LS+ and LS- groups [7 (2-25) vs 4 (0-20) months; p=0.078]. **Conclusion:** The diagnosis of MS and its timing is an important challenge for this population. Although a considerable number of patients presented with LS in the first acute episode, it was not associated with an earlier diagnosis or a shorter time to diagnose MS.

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Is there a relationship between atopic dermatitis and stroke? A systematic review and meta-analysis is there a relationship between atopic dermatitis and stroke?

Luiz Carlos Figueiredo Filho[1], Walter Oliveira Verissimo[2], Maria Fernanda de Almeida Cavalcante Aranha[1], Ricardo Ormanes Massoud[1], Isadora Rocha Rosa[1], Lucas Silva Cabeça[2], Rafael Augusto Silva Cabeça[1], Ingrid Jade Muniz Wanderley[1], Alexandre Marques da Rocha[1], Andrew Moraes Monteiro[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil. [2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Walter Oliveira Verissimo; Email: walterverissimo17@gmail.com.

Background: Atopic dermatitis (AD) has a chronic inflammatory character and intense pruritus that affects the skin tissue, afflicting 13% of children and 7% of adults. Recently, atopic dermatitis has been associated with other immunological and cardiovascular morbidities, with stroke being one of the most prevalent globally, with a 25% risk of occurrence after the age of 25, making it the second leading cause of mortality. **Objective:** To analyze the association between atopic dermatitis and stroke. **Methods:** This is a systematic review and meta-analysis of observational studies focused on the prevalence of stroke and atopic dermatitis in the same population. Articles were selected from the PubMed, VHL, Embase, Cochrane Database, Web of Science and Clinical Trial.gov databases. The PRISMA flowchart was used for data collection, with Covidence software for screening and selection. The risk of bias analysis was conducted using the ROBINS-I tool. Data meta-analysis was carried out using Review Manager 5.4 software. For continuous variables, the inverse variance method was used to group mean differences (MDs) with 95% confidence intervals (CIs). Heterogeneity was assessed using the chi-squared test. **Results:** Of the 365 articles found, 12 were included for qualitative and quantitative analysis. The primary endpoint of stroke was performed globally followed by a subgroup analysis according to the type of stroke. This outcome had a higher risk for ischemic stroke (HR = 1.19, 95%CI 1.13 - 1.26) compared to the risk of hemorrhagic stroke (HR = 1.13, 95%CI 1 - 1.29), with no statistical significance in the increased risk in patients with AD with hemorrhagic stroke. Furthermore, for the second analysis, patients with moderate AD had a lower risk of stroke (OR = 1.06, 95%CI 0.88 - 1.27) compared to severe AD (OR = 1.06, 95%CI 0.88 - 1.27). For the Risk Ratio analysis, only one study had data that were used for grouping, showing a correlation in the overall risk of stroke (RR = 1.04, 95%CI 1 - 1.07). The funnel plot showed significant heterogeneity in the HR results, which was lower in the OR results. **Conclusion:** In summary, there is a relationship between Atopic Dermatitis and Ischemic Stroke. However, there is a need for studies in different populations, to better describe the disease and other risk factors for stroke associated with the participants. Limitations of the study include the limited number of studies for analysis, risk of bias and heterogeneity. Thus, more studies on the subject are needed, including the relationship between AD treatment and stroke risk reduction, as well as the prevalence of other associated factors.

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Ischemic myelopathy, a rare vascular syndrome: case report and literature review

Monique Evelyn Mendonça do Nascimento[1], Marcelo Ataíde de Lima[1], Mariana Holanda Gameleira[1], Ricardo Martins Silva[1], Maria Eduarda Moreira Cardoso[1], Gregory Ramon Bandeira da Silva[1], Rosevaldo Alves Junior[1], Stefan Welkovic Junior[1], Ana Dolores Firmino Santos do Nascimento[1], Noelle Ventura Jordão[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Monique Evelyn Mendonça do Nascimento; Email: moniqueemnascimento@gmail.com.

Case presentation: A 64-year-old female patient with cardiovascular comorbidities (hypertension and dyslipidemia) presented with sudden low back pain followed by weakness and reduced sensation in her lower limbs. A neurosurgeon ruled out any compressive injury or surgical pathology. A CSF analysis was normal. Treatment with methylprednisolone pulse therapy for 5 days did not improve her symptoms. Tests for schistosomiasis were negative. MRI of the thoracic and lumbar spine showed hyperintensity in the dorsal medulla and the medullary cone, indicating anterior involvement of the medulla, leading to a diagnosis of ischemic myelopathy. There was a slight tumefactive effect in the medullary cone, but no significant enhancement after venous contrast. The MRI also revealed extensive atheromatosis in the aorta. Overall, the findings suggest that the patient has ischemic myelopathy, potentially caused by atherosclerosis in the aorta. **Discussion:** The spinal cord, especially the thoracic portion, is particularly dependent on radicular vascular contributions and may be the most vulnerable to infarction. The most prominent thoracic radicular artery is the artery of Adamkiewicz. Spinal cord ischaemia is a rare occurrence caused by the occlusion of part of the complex vascularization network of the spinal cord. Ischaemic myelopathy accounts for less than 1.0% of all strokes. It has a higher prevalence in elderly patients and those with comorbidities: atherosclerosis, hypertension and diabetes mellitus. The ischaemic myelopathy is associated with diffuse atheromatous disease of the aorta. Also, most cases are caused by aortic disease and repair (after an aortic surgery). MRI plays a crucial role in the diagnostic investigation and can help differentiate it from other myopathies. T2-weighted and diffusion-weighted MRI have proved to be the most useful diagnostic tools for establishing a definitive diagnosis. The clinical course after spinal cord infarction is variable. **Final comments:** Ischaemic myelopathy is a rare neurological emergency that lacks specific management guidelines. We highlight the importance of rehabilitation, reducing vascular risk factors and maintaining a healthy lifestyle. Despite severe deficits, approximately 50% of patients end up walking unaided. The patient returned to the neurology clinic after 60 days, still with significant gait impairment (Rankin 4).

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Ischemic or hemorrhagic unspecified stroke: an analysis of death profile and mortality rate in the Northern region of Brazil, 2019 - 2023

Juliana Alencar Isacksson Vieira[1], Vinícius Gabriel Nunes de Freitas[1], Arthur Andrade da Silva[1], Vitor Bidu de Souza[1], Caroline Lima de Freitas[1], Agnus Gabriel Bitencourt Magalhães[1], Thiago da Costa Brito[2]

[1]Universidade Federal do Amapá, Macapá AP, Brazil. [2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Juliana Alencar Isacksson Vieira; Email: julianaisacksson17@gmail.com.

Background: Unspecified Cerebrovascular Accident (UCA) is a condition where it is not possible to determine the classification between the types of strokes, which may have been caused by an anoxic-ischemic condition - encephalic vasogenic failure leading to lack of supplies to the brain - or hemorrhagic. Furthermore, this condition is included in the list of "garbage "codes (GC), which refer to causes of death not considered primary or those that are intermediate, terminal, or undefined. Epidemiological evidence indicates that the northern region of Brazil has a high incidence of UCA cases. Therefore, it is important to analyze the epidemiological profile of these cases to contribute to the prevention of prevalent risk factors. **Objective:** To analyze the profile of deaths and the mortality rate in the last five years of the population in the northern region affected by unspecified ischemic or hemorrhagic cerebrovascular accidents. **Methods:** This is a descriptive and quantitative study, in which secondary data were selected from the Hospital Information System of the Ministry of Health (SIH/SUS) through the Department of Informatics of the Unified Health System (DATASUS). The following resources were analyzed: mortality rate and number of deaths, according to age group, color/race, sex, and unit of the Federation. **Results:** During the period from 2019 to 2023, 7,536 deaths from UCA were reported in the northern region. Among these, the most affected age group was 80 years or older (29.15%), as expected, with a prevalence of males (52.87%) and brown people (70.60%). Notifications increased during this period, with a slight decrease in 2020, probably due to underreporting because of the Covid-19 pandemic, and peaked in 2023 (22.87%). The state with the highest occurrence was Pará (PA) with 4,076 deaths (56.96%), followed by Amazonas (AM) with 1,020 (13.53%). Another relevant data point is the mortality rate, with 2019 having the highest coefficient of 17.64, with a prevalence in females (16.56), the age group of 80 years or older (22.48), and race/color with close data between whites and blacks (19.54 and 19.53 respectively), in the 2019-2023 interval. Additionally, the states with the highest mortality rates during this period were Amapá (AP) (21.62) and Acre (AC) (19.76). **Conclusion:** Therefore, between 2019 and 2023, unspecified ischemic or hemorrhagic cerebrovascular accidents had the highest number of deaths among elderly people over 80 years old, males, and brown people,with the highest notification in the state of Pará and a peak in 2023. Furthermore, concerning the mortality rate, the most prevalent profile was also among the elderly over 80 years old, however, females and with a slight discrepancy between whites and blacks, having the highest coefficient in the state of Amapá. Thus, it is extremely relevant for public health to recognize this epidemiological profile to contribute to effective prophylactic measures for this disease.

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Ischemic stroke as a diagnostic key for takayasu's arteritis: case report

João Pedro Matos de Santana[1], Allefy Beltrão Albano[1], Bruno Henrique Carneiro Costa Filho[1], Evelliny Gomes da Silva[1], Fabrício Martins Veras[1], Letícia Klabinske Marques Monteiro[1], Luísa Couceiro de Albuquerque Macêdo[1], Víctor de Moraes Chagas[1], José Luiz de Miranda Coelho Inojosa[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.

Correspondence: Allefy Beltrão Albano; Email: allefymed@gmail.com.

Case presentation: A 27-year-old woman with recurrent episodes of fever, holocranial headache and high blood pressure since childhood, which presents with a sudden event characterized by speech changes associated with reduced strength and sensitivity in the right side that occurred in 2023. Admitted to a health care service emergency after 4 days from the onset of symptoms. She denied a similar condition and relevant family history. There are reports of four cardiorespiratory arrests during surgical procedures. On physical examination, presents asymmetry of brachial pulses, dysarthric speech, right central facial paralysis, proportional hemiparesis on the right associated with tactile hemihypoaesthesia, hemihypopalesthesia and ipsilateral heightened reflexes. Magnetic Ressonance imaging of the brain showed ischemic nucleocapsular area on the left and pons on the right, besides to angiography showing dissection of the left carotid bulb and occlusion of the right subclavian artery. In addition, there is a CT angiographic study with fusiform ectasia of the abdominal aorta and stenosis of the renal arteries with sequelae retraction of the bilateral renal parenchyma. Laboratory tests showed changes in renal function (GFR 45ml/min), negative rheumatological tests, without other findings. In this context, she was diagnosed with Takayasu's Arteritis (TA) and started treatment with Methotrexate weekly. In the onset of 2024, there were transient episodes of worsening strength in the right side of the body and rotational vertigo, with improvements occurring after a few minutes. She was admitted again for investigation and the occurrence of a transient ischemic attack represented the main occurrence suggested, considering the general clinical context and stability of the changes in neuroimaging. **Discussion:** TA is an inflammatory vasculopathy of large vessels that usually has an insidious course, with a higher prevalence in young women and asiatic population. The distribution of vascular involvement differs between sexes, with greater involvement of the abdominal aorta in men and the aortic arch in women. It is estimated that neurological manifestations such as headache and dizziness affect up to half of patients, with stroke/TIA occurring in a minority of cases (10-20%). In the present report, the patient already had long-standing symptoms, however the onset of the ischemic event corresponded to the starting point for investigation and adequate treatment of the underlying disease. **Final comments:** Takayasu's arteritis is a challenging condition considering that the presence of initial non-specific symptoms causes a delay in diagnosis and a worse clinical outcome. Therefore, it is necessary to raise such a differential diagnosis, especially in the case of stroke in young patients without established cardiovascular risk factors.

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Ischemic stroke due to free-floating internal carotid thrombus in a patient with COVID-19: case report

Daniel Lacerda da Costa[1], Gisela Tinone

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Daniel Lacerda da Costa; Email: dlacerdadacosta@gmail.com.

Case presentation: A.C.S, 53 years old, female, has a history of type 2 diabetes mellitus and was taking metformin. On June 21, 2022, the patient developed a sudden onset of weakness in the right side associated with dysarthria. Upon arriving at our institution, the patient was already outside of the window for reperfusion therapies (intravenous and mechanical thrombectomy), and investigation and clinical management were chosen. On physical examination, the patient had a drop in the deficit maneuvers of the right side. The patient underwent a head CT upon admission, and the angiotomography showed a free-floating thrombus of the left internal carotid artery. Per institutional protocol, a PCR for COVID-19 was requested during hospitalization, with a positive test result. Cardiac investigation, serology, rheumatological examinations and investigations for thrombophilia were all negative. Anticoagulation was performed with enoxaparin 1mg/kg every 12 hours for 15 days, and control Doppler ultrasound showed no changes. She was discharged asymptomatic with 100mg aspirin and 40mg atorvastatin. **Discussion:** Free-floating thrombi (FFT) are defined as clots attached to the arterial wall with blood surrounding the thrombus. They are considered infrequent causes of stroke (1.6% of ischemic stroke and TIA cases combined), and the majority of cases are symptomatic (92%). Regarding the natural history of FFT, potential outcomes include progression to occlusion, distal embolization, stabilization, or disappearance. Underlying atherosclerosis is the most common cause. During the COVID-19 pandemic, reports of an increase in cases emerged, especially in mild cases, in patients with relatively healthy vessels. Thrombophilia related to COVID-19 could be one of the reasons to justify the events. Various antithrombotic approaches (anticoagulants, antiplatelet agents and fibrinolytics) and early or late carotid interventions (endarterectomy, bypass, thrombectomy and stent) have been performed. However, neither medical nor surgical treatment was clearly superior to the other. In the patient's imaging control on the 15th day, the Doppler ultrasound revealed no changes, in line with what has been reported in the literature of cases of patients who had a stroke with relatively healthy vessels. **Final comments:** Stroke related to FFT is an uncommon etiology. Before the pandemic, the reported cases had atherosclerosis as the main underlying cause, and during the pandemic there was an emergence of cases with relatively healthy vessels. Thrombophilia related to COVID-19 is one of the justifications. The treatment of this condition is very diverse in different services, with many preferring anticoagulation. In the case described here, the patient had mild symptoms and underwent anticoagulation with enoxaparin for 15 days, with the thrombus disappearing on imaging control. The patient was discharged asymptomatic, as is the case in most cases of FFT-related strokes.

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Ischemic stroke in young adult and left ventricular non-compaction cardiomyopathy: a case report

Bárbara Borges de Oliveira[1], Brenda Letícia Lopes Batista[1], Victor Souza Torres de Vasconcelos[1], Yanne Almeida Aguiar[1], Ana Carolina Paiva Farias[1], Mariana Holanda Gameleira[1], Rosevaldo Alves Júnior[1], Monique Evelyn Mendonça do Nascimento[1], Renato de Amorim Guedes[2], Ana Rosa Melo Correa Lima[1]

[1]Hospital da Restauração, Recife PE, Brazil. [2]Procape, Recife PE, Brazil.

Correspondence: Bárbara Borges de Oliveira; Email: barbaraborgesneurologia@gmail.com.

Case presentation: A 23-year-old male patient presented with right-sided weakness and inability to speak for 2 days. He had no known previous comorbidities (cardiovascular, thrombophilia or immunocompromised). A brain CT scan revealed a decrease in density in the left middle cerebral artery area, as well as the nucleocapsular and periventricular regions. Doppler ultrasound of the carotid and vertebrate arteries showed no significant abnormalities, and viral sorologies were negative. However, a transesophageal echocardiogram revealed left ventricle dilatation and a region of non-compacted myocardium in the apical lateral and apical inferior segments, meeting the criteria for non-compacted myocardium. As a result, the patient was started on warfarin for anticoagulation. **Discussion:** Left ventricular non-compaction is a rare cardiomyopathy affecting the left ventricle, in which the two-layered myocardium has a non-compacted, abnormally thick, sponge-like trabecular layer and a thinner, compacted myocardial layer. This disease may not be considered an independent risk factor for stroke, but it should be remembered as a condition that may be associated with the risk of cardiac embolism and other additional cardiovascular abnormalities. Therefore, the risk versus benefit of starting oral anticoagulation in the absence of atrial fibrillation should be weighed up, bearing in mind that the incidence of stroke in non-compacted cardiomyopathy is around 10-15%. **Final comments:** His deficits gradually improved after he was monitored by motor physiotherapy and speech therapy. He was discharged from hospital with advice to maintain regular INR control, as well as outpatient follow-up with neurology and cardiology.

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Ischemic stroke secondary to dissection of the right common carotid artery resulting from clamping during pre-hospital care for a stab wound in cervical zone II

Caroline Dresch Sabadin[1], Marcos Henrique Lima Galles[1], Jordana Martins Américo de Souza[1], Leonardo Kenzo Cury Nogata[1], Isabella Maria de Almeida Goulart[1], Laura Bergenthal[1], Gabriella Colodel Bazzi[1] , Júlia Bortolozzo Cazari[1], Marcius Benigno Marques dos Santos[1]

[1]Universidade Estadual do Oeste do Paraná, Cascavel PR, Brazil

Correspondence: Caroline Dresch Sabadin; Email:

Carotid artery dissection (CAD), a rare complication of penetrating cervical vascular trauma, results from the laceration of the tunica intima or the rupture of the vasa vasorum, leading to bleeding within the tunica media and the creation of a false lumen, with one of its main consequences being ischemic stroke (IS). We describe a case of IS secondary to CAD resulting from clamping of the right common carotid artery (CCA) in a young, healthy patient with no history of cerebrovascular disease. **Case presentation:** The patient is a 26-year-old female, a victim of a 10-centimeter transverse stab wound in the right anterior cervical region (zone 2), managed via clamping of the right CCA during pre-hospital care. On initial examination, she presented with a Glasgow Coma Scale score of 15, no deficits, patent airways, and no other abnormalities. She underwent surgical treatment consisting of ligation of the internal jugular vein and suture of the CCA's adventitia layer. The postoperative period was managed in a hospital setting with antiplatelet therapy using acetylsalicylic acid and prophylactic anticoagulation with full heparinization. Follow-up cranial CT images showed a hypodense area in parts of the right middle cerebral artery territory, consistent with IS. Eco Doppler highlighted a 1.0-centimeter discontinuation in the intima layer of the CCA. After sixteen days of hospitalization, the patient developed left hemiparesis and grade III muscle strength in the left upper limb, with other neurological examinations remaining unchanged. **Discussion:** Penetrating cervical injuries account for 5 to 10% of traumas, with the most common mechanism being stab wounds. Arterial involvement occurs in 25% of cases, primarily affecting the carotid artery (80%). **Final comments:** Given the above, appropriate pre-hospital care is crucial for a favorable clinical outcome, as the mortality rate for penetrating cervical injuries can reach 10%. Bibliographic data suggest simple external compression or tamponade with a Foley catheter as an initial approach for cervical hemorrhages. Clamping of the CCA should be avoided as it may increase the risk of dissection, which accounts for 2% of all IS cases.

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Ischemic stroke with bilateral internal carotid artery dissection in a 30-year-old woman: a case report

Giovanna Nina Ueda[1], Maria Eduarda Cosme Bongiovani[1], Juliana Alcântara Brugnoli[1], Laura Bianchini Fogulin[1], Alice Jardim Zaccariotti[1], Matheus Spilari[1], Raíssa Cariello[1], Mayko Nascimento Merscher[1], Wagner Mauad Avelar[1], Danilo dos Santos Silva[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Giovanna Nina Ueda; Email:

Case presentation: A 30-year-old woman, with no prior medical history, undergoing uterine preparation therapy for her second attempt at in vitro fertilization, and previous episodes of paresthesia in the right upper limb, presented with acute onset headache, that progressed to reduced verbal fluency and decreased grasp force in her right upper limb. She had partial recovery and a week later had another event, this time associated with aphasia, right facial palsy and right upper limb paresis. Computed tomography and Magnetic resonance imaging indicated acute ischemic injury in the territory of the left middle cerebral artery. There was also reduced flow-void of the intracranial left internal carotid artery on MRI. Angiotomography suggests a bilateral internal carotid artery dissection (ICAD), which was then confirmed with arteriography. Double antiplatelet therapy was started and, after two months, mild right facial palsy and expression aphasia were noted. **Discussion:** Carotid artery dissection, a tear in the intimal layer of the artery with intramural thrombus, is the leading cause of ischemic stroke in individuals under the age of 50. Most ischemic strokes from dissections actually occur in the extracranial carotid and vertebral arteries, potentially blocking the artery or causing distal embolism due to cervical traumas or even relatively minor actions such as coughing or sneezing. When no major traumatic event is evident, ischemic stroke is typically attributed to spontaneous carotid artery dissection (SCAD). Most SCAD cases are unilateral, while bilateral dissections are rare, accounting for about 10% of all patients and the exact causes remain unknown. Nevertheless, certain factors, such as migraines, hypertension, smoking, and the use of oral contraceptives, are associated with an increased risk of dissection. Additionally, connective tissue and collagen diseases like fibromuscular dysplasia, Ehlers-Danlos syndrome and Marfan's syndrome can predispose to SCAD, even when minor trauma is present, indicating that these conditions may render the blood vessels more susceptible to injury. However, these disorders are rarely identifiable with current tests, as screening for underlying connective tissue disorders is not routinely conducted. **Final comments:** We encountered a bilateral ICAD case in a 30-year-old woman. Simultaneous spontaneous dissections of both carotid arteries are rarely reported but represent a noteworthy cause of ischemic stroke, especially in young patients, even with minor trauma.

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Isolated conus medullaris infarction: a case report

Gabriel Abrahão Stoliar[1], César Minoru Toita Koga[1], Valmir Vicente Filho[1], Francisco Manoel Branco Germiniani[1], Valéria Cristina Scavasine[1], Caio César Diniz Disserol[1,2,3]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[2]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[3]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Gabriel Abrahão Stoliar; Email: gabrielstoliar@gmail.com.

Case presentation: Female patient, 62 years old, hospitalized for treatment of acute chronic kidney disease, awakens in the morning with weakness of the lower limbs associated with reduced sensitivity, with an acute evolution. She had an episode of hypotension prior to the event. He presented with symmetrical paraparesis (strength grade 1/5 bilaterally), tactile, painful and vibratory hypoesthesia with a sensory level at L1-L2. She progressed with urinary and fecal retention in the following days. She had undergone cardiac catheterization via the femoral artery 3 months before the current situation. On magnetic resonance imaging of the thoracic and lumbosacral spine, hypersignal was observed on T2/FLAIR at the level of the conus medullaris, with restricted diffusion on DWI. Angiotomography of the chest and abdomen demonstrated the presence of significant atherosclerotic disease throughout its extension, with the presence of ulcerated plaques at the level of the affected topography. **Discussion:** Spinal cord infarction represents approximately 1% of strokes and 6 to 8% of acute myelopathies. Conus medullaris infarction is particularly rare, but potentially disabling, and may cause impairment in ambulation and sphincter control. Isolated infarcts of the conus medullaris are rare events, being described in the literature only in case reports and small case series. Due to the adequate blood supply to the conus medullaris through this artery and anastomosis of the anterior and posterior spinal arteries, injuries of this nature to the conus medullaris are uncommon. Clinically, it presents with acute or sudden back pain associated with bilateral sensory deficits, motor and sphincter deficits, the latter of which can develop within 12 hours. Its main etiology is atherosclerosis, as well as fibrocartilaginous embolism, vasculitis and surgical procedures in the aorta. Arterial hypertension was demonstrated as the main risk factor. Magnetic resonance imaging is useful for diagnosing spinal cord infarction. It can reveal abnormalities within approximately 3 hours of the onset of symptoms, and is recommended as it assists in appropriate treatment, reducing neuronal death and subsequent neurological deficits. **Final comments:** Isolated conus medullaris infarction is a rare entity that can be extremely disabling. Its etiology is mainly related to atherosclerosis and aorta procedures. Early clinical identification and diagnostic confirmation through neuroimaging examination are essential to prevent impairment to the patient.

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Isolated oculomotor nerve palsy, stroke and syphilis: a reported case

Renan Macionil Cotrim[1], João Márcio Borgue de Oliveira[1], Itamar Meireles Andrade Santos[1], Rebeca Aranha Barbosa Sousa[1], Ana Beatriz Marangoni Baston[1], José Eduardo Trentim Longo[1], Pedro Machry Pozzobon[1], Natália Cristina Ferreira[1], Rodrigo Bazan[1], Laura Cardia Gomes Lopes[1]

[1]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Renan Macionil Cotrim; Email: renan.cotrim@unesp.br.

Case presentation: A 68-year-old man experienced sudden onset of bilateral ptosis, more pronounced on the left, along with horizontal and painless binocular diplopia. Neurological examination revealed left-sided paresis of the medial rectus, superior rectus, and inferior rectus muscles, as well as bilateral ptosis, worse on the left. There were no pupillary abnormalities, cranial nerve deficits, or involuntary movements noted. Arterial CT angiography showed redundancy in the left posterior cerebral artery and stenosis in the right, with a soft plaque detected in the middle third of the basilar artery. Brain MRI identified an ischemic lesion with restricted diffusion in the left paramedian mesencephalic region. During the investigation into the stroke's cause, serological testing confirmed syphilis, accompanied by hyperproteinorrachia in the cerebrospinal fluid analysis. The patient was treated for neurosyphilis with crystalline penicillin 4 million IU every 4 hours for 14 days. Consequently, the patient's condition was classified as TOAST undetermined due to two potential causes: intracranial stenosis from the basilar artery plaque and meningovascular syphilis. Upon discharge, the patient received dual antiplatelet therapy and a high-potency statin. **Discussion:** Isolated oculomotor nerve palsy is rare, with limited national studies available. A population-based study conducted in Olmsted County, United States, from 1978 to 2014 reported an overall incidence of acquired third nerve palsy of 4.0 per 100,000 (95% CI, 3.3-4.7), with higher rates in those over 60 years old. Ischemic neuropathy is the primary cause in adults, but investigations should consider other etiologies causing acute clinical manifestations such as stroke. Although involvement in this neuroanatomical region typically results in additional neurological deficits, such as choreoathetotic movements and contralateral upper limb tremors with red nucleus involvement, the patient presented an atypical course without additional deficits on admission. **Final comments:** Isolated third cranial nerve palsy, though uncommon, warrants consideration of various etiologies. A comprehensive clinical history and neurological examination are crucial for accurate diagnosis. Despite the prevalence and comorbidities suggesting ischemic neuropathies in adult patients, thorough investigation into less common causes is essential for precise diagnosis and treatment planning.

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Isolated oculomotor nerve paralysis after mild traumatic brain injury

Emanuelly Victória Andrade Fonseca[1], Gustav Barbosa Falcão[1], Luma Santos de Oliveira[1], Kézia de Souza Pinheiro[1], Melissa dos Santos Matos[1], Igor Flavio Gonçalves do Vale[1], Daniel Mariani Favalessa[1], Bruno Vasconcelos Sobreira Guedes[2], Marina de Angeli[2], Bruno Batitucci Castrillo[2]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.
[2]Hospital Universitário Cassiano Antonio Moraes, Vitória ES, Brazil.

Correspondence: Emanuelly Victoria Andrade Fonseca; Email: mn.andradeduarte@hotmail.com.

Case presentation: A 37-year-old woman was involved in a motorcycle accident with head trauma while wearing a helmet in July 2023. She reported post-traumatic amnesia for 3 hours without loss of consciousness. She related difficulty to open her left eye and diplopia. She denied ethylism, smoking and other comorbidities. Physical examination revealed complete left ptosis, divergent strabismus, absence of ocular elevation, depression and adduction in the left eye, anisocoria with fixed mydriasis in the left eye. Cranial resonance imaging showed pathological enhancement of the left 3rd cranial nerve (CN III), from the interpeduncular fossa to the lateral wall of the left cavernous sinus, along the entire cisternal pathway, configuring isolated neuropathy of the left NC III after trauma. The serology was negative for infectious causes. The diagnosis of left oculomotor nerve traumatic neuritis was confirmed with clinical data, physical examination and neuroimaging. Prednisone was prescribed for 1 month with partial improvement and slight opening of the palpebral rima. **Discussion:** The CN III is divided into 4 segments: fascicular, cisternal, cavernous and orbital. Its general somatic efferent fibers arise from the oculomotor nerve nucleus and general visceral efferent fibers from the Edinger-Westphal nucleus, located at the level of the superior colliculus in the brainstem. It has an apparent origin in the interpeduncular fossa, penetrates the cavernous sinuses and enters the orbit through the superior orbital fissure. Isolated palsy of the CN III can be complete, when all voluntary movements are absent, resulting in exotropia, diplopia, ptosis, or incomplete, when it affects part of the muscles innervated by the CN III. The pupil can be involved with an enlarged and nonreactive pupil (mydriasis). The main causes in adults are ischemia,

aneurysm, neoplasm and trauma, with only 10% being traumatic etiology. Isolated involvement of the CN III by traumatic brain injury (TBI) that is not caused by mass effect lesions, such as subdural or extradural hematomas, is rare, especially in mild TBIs. Pathogenic mechanisms leading to this outcome can be direct (abnormal nerve stretching from high kinetic energy trauma, compression by adjacent structures altered by injury, such as a pseudoaneurysm) or indirect (mainly ischemia from immediate vascular changes or metabolic alterations post-trauma). Our case shows a woman with isolated CN III palsy after mild TBI. The etiological hypothesis is distension and contusion of the left CN III at the posterior petroclinoid ligament, due to movement of the brainstem and consequent axonal stretching during the impact. **Final comments:** In conclusion, the case highlights a rare but significant neurological complication following mild traumatic brain injury, emphasizing the importance of detailed clinical evaluation, imaging, understanding of anatomical and pathophysiological aspects in managing such cases effectively.

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Isolated ophthalmoplegia in a patient with Miller-Fisher syndrome

Luísa Stradioto Sartor[1], Maysa Rebelo Crestani[2], Pietro Turella[3], Tanisi Rebelo Crestani[1], Fabrício Diniz Kleber[1]

[1]Universidade de Caxias do Sul, Caxias do Sul RS, Brazil.
[2]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.
[3]Universidade do Oeste de Santa Catarina, Joaçaba SC, Brazil.

Correspondence: Luísa Stradioto Sartor; Email: lssartor@ucs.br.

Case presentation: A 34-year-old male patient was admitted to the hospital due to sudden onset diplopia. The initial neurological examination shown bilateral ophthalmoplegia suggestive of III nerve palsy, associated with mydriasis, dysarthria, and paresthesia on the left side of the face. The first impression of the attending neurologist was a brain stem syndrome with internuclear ophthalmoplegia due to the sudden onset of symptoms. Gradually, during the first 10 days of hospitalization the patient developed complete paralysis of the extrinsic ocular muscles and started with bilateral facial palsy and worsening of the dysartria, the reflexes were also widespread decreased. There was no significant ataxia on examination. The investigation progressed with a lumbar puncture and electroneuromyography that shown elevated protein with normal cellular counts on the cerebrospinal fluid and alterations suggestive of Miller Fisher syndrome on the electroneuromyography. Treatment with Intravenous Immunoglobulin was administered with some improvement of the facial palsy at discharge from hospital. Physical examination at discharge showed mild left facial paresis, mild dysarthria, and he continued with the bilateral ophthalmoplegia, worst on his left side. At follow-up revision by phone the patient stated that he has improved considerably, but remaining with the ophthalmoparesis. **Discussion:** The Miller-Fisher syndrome (MFS) is usually a rare syndrome characterized by the classical triad: ophthalmoplegia, ataxia and areflexia. It is considered a variant of the Guillain-Barré syndrome, with incidence worldwide around 1-2 cases each 100.000 people. Typically it presents with the acute or subacute onset of the triad, starting with the ophthalmoplegia, diplopia, following with ataxia and generalized areflexia. The diagnoses is usually made by the presence of elevated proteins in the cerebrospinal fluid and electroneuromyography typical findings, and we can also identify the presence of anti-GQ1b IgG antibodies on the cerebrospinal fluid. At the present case we do not have this test available what the public system. The GQ1b antibody is thought to have a direct effect on the neuromuscular junctions of the ocular muscles. The GQ1b ganglioside is also an important component of oculomotor nerve myelin. In the afore mentioned case it is remarkable the unusual presentation of hyper acute symptoms with the association with marked mydriasis at the onset. There was no significant ataxia or gait abnormalities at the onset. Furthermore, it is uncommon for the patient with MFS to present with an hyperacute onset associated with mydriasis and paresthesias. **Final comments:** The case report related here, shows a rare presentation of the MFS. It elucidates a rare presentation of a variant of GBS, that is an important cause of diplopia and ophthalmoplegia caused by a demyelinating peripheral nerve autoimmune desease that could affect the young population.

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Isolated optic neuritis: an atypical manifestation of bartonellosis

Amanda Carvalho de Oliveira[1,2], Lucas Leroux de Ricchezza[2], Marcela de Moraes Serpa[2], Marco Antonio Sales Dantas de Lima[1], Carolina Rouanet[1,2], Nayane Gentil[2], Rosa Batistella[2], Vinicius Soares[2], Luciane S. Oliveira[2]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Hospital Copa D'or, Rio de Janeiro RJ, Brazil.

Correspondence: Amanda Carvalho de Oliveira; Email: amanda-c16@hotmail.com.

Case presentation: A 49-year-old female experienced an acute onset reduction of visual acuity in her right eye, with a predominantly central scotoma, which progressively worsened over seven days, accompanied by pain on eye movement. She also reported daily chills and headaches over the past month. She had a relevant history of being scratched by a rescue kitten two months before her first evaluation. The kitten passed away a few days later without a known cause. Neurologic examination was notable for perception of movement only in her right eye. Fundoscopic examination showed right optic disc (OD) edema without retinal changes. During her hospital stay, she consistently had elevated serum inflammatory markers (ESR and CRP) and daily low-grade fever of unknown origin. MRI showed mild optic nerve (ON) sheath distension, right ON tortuosity, bulginig of the right nerve optic disk, contrast enhancement of the ON-globe junction, and restricted diffusion of the right OD. CSF workup showed 42 white blood cells with lymphomononuclear predominance. She was started on doxycycline, rifampin, and prednisone for suspected bartonellosis with CNS involvement, with gradual improvement of symptoms, ESR, and CRP. On day 15 of her hospital stay, serum IgG and IgM for bartonellosis came back positive, and the patient was discharged for outpatient follow-up. A repeat MRI two weeks after discharge showed significant improvement. **Discussion:** Cat scratch disease is a systemic disease caused by Bartonella henselae, acquired through scratches, bites, or exposure to fleas from infected cats, and mostly causes mild systemic symptoms in immunocompetent individuals. Neuroretinitis is a rare manifestation of the disease (1-2%) and causes acute unilateral visual loss due to OD edema and macular exudates, which are often star shaped, but it can take weeks to appear. Other possible neurologic manifestations include encephalopathy, transverse myelitis, radiculitis, and cerebellar ataxia. We report a case of isolated optic neuritis initially seen in an ophthalmologic emergency unit, which identified OD edema and transferred the patient for suspected autoimmune disease. However, a history of fevers and chills and persistently elevated serum ESR and CRP led us to withhold immunosuppressive treatment and focus on a potential infectious etiology. The patient's history of being scratched by a kitten two month before and characteristic optic disk bulging seen at MRI led us to start empiric treatment for neurobartonellosis while awaiting serologic confirmation. **Final comments:** This case highlights the importance of considering infectious etiologies of isolated optic neuritis, especially in the presence of warning signs such as fever and elevated inflammatory markers, as high-dose corticosteroids may worsen such cases. Additionally, it underscores the need to consider cat scratch disease even without the characteristic stellate macular exudate and retinal changes.

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Isquemic stroke concomitant with pulmonary thromboembolism: when to anticoagulate ?

Lucas Rodrigues Prim[1], Ana Paula Gonçalves Barankiewicz[1], Maria Julia Branco Rodrigues[1], Marcos Seefled[1], Lisseth Fabiola Vallejo Reyes[1], Frederico Otto Flores Moraes[1], Eduardo Takahashi Sunaga[1], Joao Pedro Greca de Macedo Biasi[1], Gustavo Schneck de Jesus[1], Vitor Bonk Rizzo[1]

[1]Hospital Universitário Evangélico Mackenzie, Curitiba PR, Brazil.

Correspondence: Ana Paula Gonçalves Barankiewicz; Email: anapaulaapgs@gmail.com.

Case presentation: A 69-year-old woman with no comorbidities presented to the Emergency Department (ED) on April 29th for a suspected stroke. Family members who witnessed the episode reported that she had a same-level fall at home in the morning, followed by right-sided hemiparesis, leftward gaze deviation, and global aphasia. On admission, her NIH Stroke Scale (NIHSS) score was 18, with a time of evolution of 1 hour and 19 minutes. She received thrombolysis due to the absence of contraindications. Her general condition was poor, with a blood pressure of 140/81 mmHg, heart rate of 78 bpm, respiratory rate of 18 breaths per minute, and oxygen saturation of 87% on room air, requiring supplemental oxygen via a nasal cannula. Neurological examination revealed global aphasia, right hemineglect, right lower limb plegia, and grade 1 weakness in the right upper limb with global hyporeflexia. An admission CT angiogram showed a left M1 occlusion. The cranial computed tomography (CT) scan had an ASPECTS score of 10 and also revealed bilateral pulmonary embolism (PE) in the peripheral pulmonary arteries. A follow-up CT scan 24 hours after thrombolysis showed evidence of bleeding without NIHSS change, consistent with ECASS-PH 1. Anticoagulation with enoxaparin was initiated on May 1st due to the risk of cardiogenic shock from PE. A subsequent cranial CT revealed intraparenchymal hemorrhage in the left basal ganglia with surrounding hypodense edema, compression of the left lateral ventricle, and hyperattenuating material suggestive of hematoma. There was a midline shift to the right (5 mm). On May 10th, the cranial CT showed reduced areas of hemorrhagic transformation and decreased intraventricular hematoma. The patient's consciousness level improved (Glasgow Coma Scale score of 10), but she continued to have right hemiplegia and global aphasia. She was not using supplemental oxygen and had no desaturation, and she was referred for rehabilitation and outpatient follow-up. **Discussion:** Stroke is highly prevalent and a leading cause of mortality in Brazil, ranking second in women and third in men. Similarly, PE is common, with a prevalence of up to 16.6% and mortality rates of up to 30%. The occurrence of concurrent PE and ischemic stroke (IS) is rare, lacks clear therapeutic guidelines, and poses treatment challenges. Anticoagulation for PE is typically contraindicated in IS patients receiving thrombolysis due to an increased risk of bleeding. Clinically, the simultaneous presence of neurological deficits and respiratory symptoms should raise concern. **Final comments:** Simultaneous PE and IS is extremely rare but has a significant impact on patient health. Although guidelines do not recommend early resumption of anticoagulation after thrombolysis in IS patients, the risks and benefits should be evaluated in cases of concurrent PE. Interdisciplinary assessment and treatment decisions are essential in this clinical context.

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James Parkinson's legacy on child abuse two centuries after his passing.

Emanuelle Campos Amaral[1], Fernanda Almeida Carvalho[1], David Franca Ferreira Cruz[1], Maria Esther Lima Rios Evangelista[2], Danilo Lemes Reis[1], Manuela Brandao Ferreira Teles[1], Victor Hugo Soares dos Santos[1], Samuel Natal Rodrigues[1], Diogo Rabelo de Lima[1], Marcus Vinicius Della Coletta[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Emanuelle Campos Amaral; Email: eca.med22@uea.edu.br.

Background: Well-known English doctor James Parkinson is credited with providing the initial explanation of Parkinson's disease. He was influential in the development of the present specialty of pediatrics because, despite his extensive study in the field of neurology, he was interested in the healthcare, safety, and protection of children. He was a significant figure in many fields, studying fossils, geology, medicine, politics, and raising awareness of child abuse and its harmful effects on children's health. **Objective:** The primary goal is to draw attention to James Parkinson's importance in the battle against child abuse, because he was among the first medical professionals to speak out about the problem. **Methods:** The methodology was a systematic review based on 3 articles found on Scopus, Scielo and PubMed, to highlight his significance outside of the medical field. The inclusion criteria were articles that included the terms "James Parkinson", "child" and "abuse" in the title or in the abstract. The exclusion criteria were inappropriate methods and articles that emphasized his accomplishments in different fields. **Results:** James Parkinson noticed the abusive behavior when he began to connect head injuries, primarily from falls, with the development of hydrocephalus or "watery head". After seeing a case of child abuse, that he detailed in his 1804 pamphlet "The Villager's Friend and Physician", he noticed that another major contributing factor to hydrocephaly was improper child discipline, in which parents would strike their children severely in the head. Consequently, following the aggressive conduct, the youngster would complain of typical hydrocephaly symptoms, like cephalalgia, restless sleep, nausea and fever. He wrote the book "Medical Admonitions to Families" in 1807 to warn the public, in which he candidly said that the parents' inappropriate educational methods were the root of many health problems. He labeled those actions as child abuse and emphasized that, to properly correct someone, there must be mercy and open communication, not suffering and terror. He openly spoke about different and improved ways to educate children, defying the habits of the nineteenth century. Furthermore, he explained that the fear experienced during these violent acts would result in mental illnesses. **Conclusion:** In summary, it is evident that Parkinson acted outside of the Neurology field, with accomplishments beyond the description of Parkinson's disease, which had positive impacts, even 200 years after his death. He was ahead of his time in recognizing the negative effects of this kind of maltreatment on a child's development and health. It was a tremendous move for him to openly address the matter and share his opinions, starting a conversation that would result in future rights enforcement and protection for this vulnerable group as well as avoid any psychological and physical harm.

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Journey of value of raras (JAV RARAS): cost analysis of the treatment of amyotrophic lateral sclerosis (ALS) using the time-driven activity-based costing (TDABC) methodology in a specialized service in Salvador-Bahia

Laís Brandão Mota[1], Luna Souza Silva Vilanova[1], Maria Rodrigues Lima de Souza Pessoa[1], Stefania Zingone Andrade Carvalho[1], Temis Maria Felix[2], Marcela Câmara Machado Costa[1], Marilaine de Matos Menezes Ferreira[1], Camila Azevedo[3], Marcelo Nita[3], Thiago Belina[3]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[3]MAPES Solutions, São Paulo SP, Brazil.

Correspondence: Laís Brandão Mota; Email: bmota.lais@gmail.com.

Background: Amyotrophic lateral sclerosis (ALS) has many ongoing research efforts and multidisciplinary care initiatives aimed at improving the management of this disease. In this context, the Time-Driven Activity-Based Costing (TDABC) methodology has proven to be an effective approach for healthcare organizations to obtain an accurate view of the care process and the costs associated with the services provided. The use of TDABC allows for the identification of costly activities, enabling the optimization of processes and the reduction of costs without compromising the quality of patient care. This approach is crucial for supporting decisions and efficiently allocating resources in the treatment of rare diseases such as ALS. **Objective:** To quantify the costs related to the treatment of ALS in a specialized service in Salvador, Bahia, using the TDABC method and to contribute to a more detailed understanding of the annual cost of care for patients with ALS. **Methods:** This study describes the implementation of the TDABC methodology in the rare disease care journey (JAV RARAS). Cost data were collected through interviews and surveys with administrative, nursing, and healthcare professionals from the institution. Direct costs were analyzed by identifying activities in disease management, allocating consumed resources, estimating the cost per unit, and measuring activity time. Additionally, the study allowed for the validation of protocols that cover the care journey of patients with rare diseases, contributing to a more accurate understanding of the annual cost of care. **Results:** The average annual cost per patient with ALS is $7.506.20. Most of this value, $7.238.15, is dedicated to the treatment itself. The main cost components are human resources ($1.227.46), materials ($16.43), medications ($2.252,51), tests ($138.20), procedures ($348.63), and equipment ($3.251.97). Regarding the source of financial resources, approximately 98.5% comes from the Unified Health System (SUS), while about 1.5% is funded by the institution itself. **Conclusion:** This study investigated the annual costs of ALS treatment in a specialized service. The results indicate that the average annual cost per patient is $7.506.20, with most of this value dedicated to the treatment itself. These findings highlight the importance of understanding and optimizing the use of these public resources to ensure the sustainability of the health system and equitable access to ALS treatment.

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Juvenile Huntington's disease: the influence of CAG repeat number on clinical manifestations - 2 case reports

Víctor de Oliveira Costa[1], Iasmim Portela Maifrede[1], Márcio Alves da Cruz Júnior[1], Daniel Sabino de Oliveira[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, EBSERH, Juiz de Fora MG, Brazil.

Correspondence: Victor de Oliveira Costa; Email: victordeoliveiracosta82@gmail.com.

Case presentation: Case 1: A 21-year-old woman began exhibiting heteroaggressiveness, inattention, difficulty with social interaction, and information processing at the age of 10. By 19, she developed mild appendicular bradykinesia and rigidity. At 21, she presented with upper limb bradykinesia, cogwheel rigidity in all four limbs, gait with dystonic posture of the right upper limb and lower limbs, choreiform movements of the upper limbs, and myoclonus, along with abnormal cognitive test results for her age and educational level. Behavioral evaluation revealed severe depression with previous suicidal ideation. Concurrently, her father developed aggressiveness and choreic movements. A brain MRI showed bilateral atrophy of the caudate nuclei. Both the patient and her father underwent genetic testing for Huntington's disease (HD), revealing 61 and 47 CAG repeats, respectively. Case 2: A 10-year-old male experienced difficulty sitting without support, with backward falls and right-sided imbalance while running since 7 years of age. He progressed to frequent falls associated with rhythmic involuntary movements of the right upper limb. He exhibited mild dystonic posture, bradykinesia, and backward falls in tandem. His school performance was delayed, particularly in reading. A brain MRI showed hyperintensity of the striatum and putamen, along with atrophy of the caudate nuclei and putamen. His father was diagnosed with HD at the age of 38. Genetic testing for HD revealed 79 CAG repeats. **Discussion:** Huntington's disease is characterized by motor and cognitive dysfunction, along with psychiatric and behavioral changes. Individuals with age of onset ≤20 years are considered to have juvenile Huntington's disease (JHD), accounting for 4-10% of HD patients. Juvenile forms are associated with alleles containing more than 50 CAG repeats. There is an inverse relationship between CAG repeat length and age of onset for JHD, with higher repeats indicating earlier onset and faster disease progression. Moreover, JHD is divided into those with highly expanded repeats (≥80 CAG repeats) and those with fewer repeats. Those with higher repeat counts exhibit earlier and faster symptom progression, as seen in these cases. The most common pathological change is diffuse and marked atrophy of the neostriatum, often worse in the caudate than in the putamen. There is no cure or disease-modifying treatment for HD, focusing instead on symptom management and supportive care. **Final comments:** JHD is a rare disease requiring further studies to better define its parameters and compare it with adult-onset HD for improved diagnostic approaches and therapeutic interventions.

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Juvenile myasthenia gravis positive acetylcholine anti-receptor in a male child: a case report

Maria Luiza Vasconcelos Montenegro[1], Mariana Lucena Loureiro[1], Déborah Emmily de Carvalho[1], Maria Clara Arcoverde Santana[2], João Eudes Magalhães[2], Fabíola Lys de Medeiros[2]

[1]Universidade de Pernambuco, Recife PE, Brazil.
[2]Hospital Universitário Oswaldo Cruz, Recife PE, Brazil

Correspondence: Maria Luiza Vasconcelos Montenegro; Email: marialuizavasc534@gmail.com.

Case presentation: Male, 11 years old, started experiencing muscle weakness, frequent falls, and fatigue in the lower limbs (LL), associated with difficulty walking and eating, at the age of 6. He denied other complaints and had no similar family history. On neurological examination: bilateral semiptosis, ophthalmoplegia, reduced facial expression, nasal speech, normal static and dynamic balance. Grade 4 strength in the upper and lower limbs, normoactive reflexes, trophism, and muscle tone without changes. Computed tomography of the chest, to evaluate the thymus, was normal. Total creatine phosphokinase (CK) dosage was 81u/L. Electroneuromyography showed a maximum decrease of 30.1%, compatible with myasthenic syndrome. Measurement of antibodies against

acetylcholine receptors (anti-AChR), in 2019, was negative. But in the face of post-synaptic myasthenic syndrome, Mestinon (120mg/day) was prescribed. Due to the persistence of symptoms, Prednisone was added at 5mg/day, resulting in an improvement in lower limb weakness and nasal speech. In 2020, a new measurement of antibodies against the muscle-specific tyrosine kinase receptor was carried out, which was normal. In 2021, due to the persistence of bilateral eyelid semiptosis, Mestinon was increased to 180 mg/day. In 2024, a new measurement of anti-AChr antibodies was positive, with Azathioprine 50 mg/day being associated with therapy. Currently, the patient denies muscle weakness or new complaints, however, due to the persistence of bilateral eyelid ptosis and ophthalmoplegia, the doses of Mestinon and Azathioprine were increased to 210 mg/day and 75 mg/day, respectively. **Discussion:** Myasthenia gravis (MG) is a chronic autoimmune disease that affects neuromuscular transmission, leading to progressive muscle weakness and excessive fatigue. The most common form of MG is that associated with anti-AChR antibodies, which attack acetylcholine receptors at the neuromuscular junction, resulting in the characteristic muscle weakness. This symptom usually occurs mainly in the eye muscles, leading to ptosis and diplopia. The incidence of MG with positive anti-AChR antibodies is higher in adults, especially young women and older men. However, it can also occur in children and adolescents, although it is less common. The usual treatment involves the administration of anticholinesterase medications to increase the concentration of acetylcholine at the neuromuscular junction, thus improving the transmission of nerve stimuli to the muscles. Immunosuppressants may be necessary to control the underlying autoimmune response, as seen in the reported case. **Final comments:** Juvenile MG, although uncommon, is a complex condition that requires multidisciplinary and personalized management. With early diagnosis, appropriate treatment, and ongoing support, many children can achieve a good quality of life, allowing them to fully participate in everyday activities.

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Kennedy disease: a case report

Antonio Marco Duarte de Albuquerque[1], Sarah Câmara Ferreira[1], Sophia Alves de França[1], Jonatas Pinto Pinheiro de Sousa[1]

[1]Faculdade de Medicina de Olinda, Olinda PE, Brazil.

Correspondence: Sarah Câmara Ferreira; Email: sarahcamaraf99@hotmail.com.

Kennedy's disease (KD), or bulbospinal muscular atrophy, is a hereditary recessive condition linked to the X chromosome. It is characterized by the degeneration of lower motor neurons, resulting in atrophy, muscle weakness, and tremors. A mutation in the androgen receptor gene is associated with the disease, leading to the expansion of CAG trinucleotide repeats. Additionally, endocrinological manifestations such as gynecomastia, testicular atrophy, and oligospermia are common. **Case presentation:** A 68-year-old male patient with difficulty walking, initially in his right foot and later affecting both feet. He presents with decreased sexual capacity and testicular atrophy. Physical examinations reveal a paraparesis gait and gynecomastia. Laboratory tests show an increase in CPK. Breast ultrasound and electromyography of the lower limbs confirm the gynecomastia and strongly suggest KD, respectively. **Discussion:** Discussion highlights that, even without access to genetic testing, it is possible to diagnose rare pathologies through clinical exams and low-cost tests. The increase in CPK may indicate myopathic changes secondary to denervation. **Final comments:** The conclusion emphasizes the importance of genetic counseling and monitoring the disease's progression in the absence of molecular diagnostic evidence.

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Kennedy's disease (X-linked spinal and bulbar muscular atrophy): follow-up of 18 patients in a neuromuscular outpatient clinic

João Paulo Barile[1], Kaliny Oliveira Peixoto[1], Patrícia Marques Mendes[1], Renan Brandão Rambaldi Cavalheiro[1], Daniel Delgado Seneor[1], Igor Braga Farias[1], Paulo de Lima Serrano[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, Setor de Doenças Neuromusculares, São Paulo SP, Brazil.

Correspondence: João Paulo Barile; Email: barile.jpaulo@gmail.com.

Background: Kennedy's disease (KD), or X-linked spinal and bulbar muscular atrophy, is a rare lower motor neuron disease associated with abnormal CAG trinucleotide repeat expansion (≥38 CAG repeats) in the AR gene. The onset of symptoms is earlier in cases with a greater number of CAG repeats. Symptoms generally begin between 30-50 years of age with slowly progressive lower motor neuron syndrome, weakness of the proximal and distal muscle groups of the upper and lower limbs, bulbar compromise with dysarthria and dysphagia, fasciculations, myokymia and postural tremor. Sensory symptoms, such as paresthesia, may be also observed. Androgen insensitivity frequently occurs leading to gynecomastia, infertility, erectile dysfunction, and metabolic disturbances (glucose intolerance, dyslipidemia). **Objective:** Present the clinical, genetic and neurophysiological aspects of an original case series of 18 patients with KD. **Results:** The average age at onset of symptoms was 44 years (25-60) and the average age at diagnosis was 50 years (38-62). The average duration of symptoms until diagnosis was 4.8 years. The main clinical complaints included: limb weakness (n=14); dysphagia (n=7); muscle fatigue (n=6); and neck weakness, hand tremors, fasciculations, and cramps (n=1, each one). Clinical course was progressive in 72.22% of cases. A positive family history was identified in 77.7% of cases. Regarding clinical involvement, the following profile was seen: proximal muscle weakness of the lower limbs with reduced tendon reflexes (n=18; 100%); tongue atrophy (n=17; 94.4%); proximal quadriparesis (n=15; 83.3%); fasciculations in the tongue n=14 (77.7%); dysphagia (n=12; 66.6%); distal tremor (n=11; 61.1%); facial diparesis (n=9; 50%); dysphonia (n=8; 44.4%); neck fasciculations (n=4; 22.2%). Ankle clonus, extensor plantar responses, spasticity and dementia were not observed in any patients. Non-invasive ventilation dependency was seen in only 11.1% of cases. Other findings observed included: gynecomastia (n=15; 83.3%), sexual dysfunction (n=4; 22.2%), hyperkyphosis or scoliosis (n=4; 22.2%), and pes cavus (n=2; 11.1%). Diabetes or pre-diabetes and dyslipidemia were seen in 44.4% of cases. Needle electromyography disclosed chronic denervation involving the bulbar, cervical and lumbosacral myotomes (n=18) as well as reduced amplitude of sensory nerve action potential (n=17). Mean serum creatine kinase levels were 772 (450-1300). Genetic testing disclosed an average value of 48 CAG repeats (44-55) in the AR gene. No significant brain and spine abnormalities were seen on Magnetic Resonance Imaging (MRI) studies. All patients used leuprorelin acetate (n=18) in their treatment. **Conclusion:** KD is a rare X-linked inherited disease with complex neurological and systemic features. Future studies are needed to develop modified disease therapies.

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Ketogenic diet applied to myoclonic status epilepticus in progressive myoclonic epilepsy

Atilio Gomes Romani[1], Miriam Eva Koch[1], Ruda Alessi[1], Eduardo Macedo de Souza Tieppo[1], Rayanne Pereira Mendes[1], Renato Morais Bueno[1], Thainá Louise Rodrigues[1], Vitor Luiz Selva Pinto[1], Bruno Graziosi[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Atilio Gomes Romani; Email: atilio_romani@hotmail.com.

Case presentation: Man, 26 years old, son of consanguineous parents with no family history of genetic or epileptic diseases. Asymptomatic until the age of 14, when his school performance declined. At the age of 18, he presented startle with myoclonic jerks, falls from his own height, difficulty walking and his first generalized seizure. Subsequently, despite the use of anticonvulsants, he progressed with generalized myoclonic tremors, dysarthria and decreased strength in the upper and lower limbs. Within a few years he was bedridden due to severe ataxia. The neurological examination showed: paratonia, sialorrhea, sardonic smile, presence of dystonic tremor of the four limbs, facial dystonia, moderate cognitive dysfunction (difficulty understanding), grade II general muscle strength. Computed tomography and magnetic resonance imaging were normal. The EEG: revealed bitemporal irritative activity independent of Sherriton. The genetic test was not available to define hypotheses, but neurological signs and clinical history corroborate the diagnosis of progressive myoclonic epilepsy. During hospitalization for pneumonia, the patient developed myoclonic status epilepticus and maintained the condition even after optimization of topiramate, valproic acid, levetiracetam, clobazam, cannabidiol and an infusion pump with ketamine and midazolam. Therefore, it was decided to introduce a ketogenic diet via a nasoenteral tube, showing an express response in the following days with the possibility of gradual withdrawal of sedatives and continuation of anticonvulsant drugs. **Discussion:** Progressive myoclonic epilepsy (PME) is a rare and heterogeneous group of epileptic syndromes characterized by myoclonus, instability, muscular instability, intellectual disability and epileptic seizures. PMEs are often caused by genetic changes, including genes such as PME1 and PME2. Clinically, PME is characterized by onset in childhood or adolescence with myoclonus and seizures, followed by progressive cognitive and motor decline. The diagnosis of PME requires a multidisciplinary approach, integrating clinical history, genetic and histological tests. Its treatment is based on symptoms control, but does not change the progression of the disease. The ketogenic diet (KD) is a diet rich in fat and low in carbohydrates, which induces a state of metabolic ketosis. Studies suggest that ketosis can stabilize neuronal excitability, reduce seizure frequency and, specifically for PME, decrease the severity of myoclonus. The relevance of KD in PME is particularly notable given the limitations of traditional anticonvulsants in controlling seizures in this population. Furthermore, KD may offer neuroprotective benefits by slowing the progression of neurological extension associated with PME. **Final comments:** In this report, we describe a case that was sucessful in controlling seizures using a ketogenic diet, which can be used as a basis for other studies and as a therapeutic tool for similar cases

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Kleine-Levin syndrome after COVID-19: a clinical report of the use of clarithromycin during the crisis of hypersomnia

Aline Vieira Scarlatelli Lima Bardini[1], Silvia Rosa Neu[1], Maria Luiza Ataide Cardoso[1], Julia Pinter Del Castanhel[1], Victoria Pietsch[1], Gabriele Tartari de Souza[1]

[1]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.

Correspondence: Aline Vieira Scarlatelli Lima Bardini; Email: alinevieiralima@hotmail.com.

Case presentation: A 15-year-old male, with no personal or family history of neuropsychiatric disease or perinatal/developmental complications, has developed symptoms of hypersomnia, with episodes of severe daytime sleepiness, associated with hyperphagia and altered social behavior different from his typical pattern of sleep or humor. During the same period, he was diagnosed with hemolytic anemia, requiring hospitalization and frequent transfusions with human immunoglobulin. All these symptoms have been developed few days after a mild COVID-19 infection. Two months later, there were changes in the pattern of hypersomnia, characterized by long hours of sleep over a 7-day period, with episodes of partial awakening characterized by automatic behavior, along with hyperphagia and an altered mood. These episodes have occurred at least once a month, and a total of 3 episodes occurred before the neurological evaluation. Neurological examinations, blood exams and polysomnography were normal. The diagnose of Kleine Levin was done and treatment with lithium 300mg was prescribed. During the dose adjustment, the patient has showed a new episode of hypersomnia, and during the crisis the lithium was increased to 450mg and associated Clarithromycin 500mg, twice a daily, reducing the duration of episodes to 3 days with mild symptoms. After the symptoms improved, only lithium 450mg was maintained. There have been no new events since then. **Discussion:** Kleine-Levin syndrome is a rare condition of unknown origin. The clinical scenario is defined by intermittent and repetitive cycles of sleepiness associated with behavioral disturbances such as aggressiveness, mental confusion, and hypersexuality. Regarding its management, there are divergences in the literature, as its pathophysiological and etiological mechanisms are not yet clarified. Previous research has mentioned the use of lithium and carbamazepine due to the similarity of Kleine-Levin Syndrome with affective disorders related to Gabaergic receptors and monoaminergic neurotransmitters. Clarithromycin could be used during the crisis to reduce the episodes. Besides being an antimicrobial, there are reports of its action as a GABAa receptor antagonist, decreasing its inhibitory action on GABA. However, it crosses the blood-brain barrier only in situations of meningeal inflammation, which explains why the action in remitting the condition is temporary. **Final comments:** The pathophysiology, etiology, and treatment of Kleine-Levin Syndrome remain unknown. In the present case, the syndrome developed after the COVID-19 infection. The use of clarithromycin associated with lithium reduced the severity of symptoms and reduced hypersomnia episodes. This suggests that, in addition to Gabaergic and antimicrobial action, there may be a third mechanism that has not yet been clarified in the literature.

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Knowledge of patients about the risk factors of ischemic stroke

Natalli Studnicka[1], Tatiana Pizzolotto Bruch[1], Amanda Honorato Matos[1], Nathalia Copetti Guzmán[1], Camila Pezente Dal Toe[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Natalli Studnicka; Email: natalli.stu@gmail.com.

The study investigated the knowledge of attendees at a university outpatient clinic regarding the risk factors for ischemic stroke (IS). A total of 385 participants were interviewed, predominantly female (77.1%), aged between 18 and 84 years. Most had a monthly income between 2 and 4 minimum wages (49.4%) and had completed high school (20.8%). Approximately 69.4% reported previous contact with ischemic stroke victims. The risk factors most recognized by participants were systemic arterial hypertension, smoking, sedentary lifestyle, obesity, and chronic alcohol consumption, all identified by over 90% of respondents. Overall knowledge level was deemed moderate, especially among those with prior contact with IS victims. It was concluded that there is a significant gap in the understanding of ischemic stroke and there is a need to enhance public education on prevention and management of these risk factors., suggesting the need for measures to promote chronic disease control and adoption of healthy habits.

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Laminopathy diagnosis: the importance of clinical correlation when considering a VUS mutation

Jéssica Cristina Silveira Damasceno[1], Laura Altomare Fonseca Campos[1], Júlia Lima Vilela[1], Marcelo Sobrinho Mendonça[1], Thiago Cardoso Vale[1]

[1]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.

Correspondence: Jéssica Cristina Silveira Damasceno; Email: drajessicadamasceno@gmail.com.

Case presentation: A 13-year-old boy reported proximal weakness in the lower limbs with a pelvic tilt gait that began at 4 years old, followed by proximal weakness in the upper limbs at 6 years old. His father died at the age of 45 due to complications of a suspected muscular dystrophy, but without a molecular test. Physical exam showed hypotrophy of the biceps and triceps brachii bilaterally and muscle tone preservation. Electroneuromyography of the 4 limbs showed normal motor and sensory nerve conductions. Lower limb myography was also normal and the patient did not tolerated the exam in the upper limbs due to pain. Duchenne muscular dystrophy (DMD) testing ruled out the presence of deletions/duplications in the dystrophin gene, DMD locus XP, associated with Duchenne/Becker muscular dystrophy. A broad DNA panel of neuromuscular diseases was then carried out, which revealed a mutation with 1 heterozygous copy of the lamin A/C (LMNA) gene, variant of uncertain significance (VUS), raising the diagnostic hypothesis of laminopathy. At the last consultation, he presented preserved global strength, bilateral humeral atrophy, shortening of the bilateral Achilles tendon with walking on tiptoe, and noted initial peroneal hypotrophy. **Discussion:** The patient had a negative genetic test for DMD, the main diagnostic hypothesis in boys with proximal weakness. Electrophysiological study came out within normal limits, excluding the diagnosis of motor neuron disease. It was necessary to expand the diagnostic work-up with a broad genetic panel that detected a VUS mutation in the LMNA gene, establishing the diagnostic hypothesis of laminopathy, due to the clinical presentation shown by the patient combined with the exclusion of other diagnoses. Laminopathies can present with an extensive phenotypic spectrum, the main ones being: Emery-Dreifuss muscular dystrophy (EDMD), limb-girdle muscular dystrophy (LGMD) with cardiac conduction disorders, dilated cardiomyopathy and lipodystrophies. In EDMD, weakness occurs with a humeroperoneal pattern with early tendon contractures, which may occur even before the weakness, in addition to cardiac conduction disorders in adulthood. The patient, despite not presenting any classic phenotype of laminopathies, presents significant and unequivocal humeral atrophy and incipient peroneal hypotrophy, characteristic of EDMD. **Final comments:** This case illustrates the importance of the correlation between clinical picture and molecular test. After extensive investigation of myopathies, including dystrophies, it became important to value the VUS mutation, since a clinical phenotypic correlation could be established with the evidenced genetic alteration.

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Langerhans cell histiocytosis involving the corpus callosum and spinal cord in an elderly patient

Mateus Pinto Marchetti[1], Isabela Louise Weber[1], Lucas Shigueo Hiromoto[1], Pedro Arthur Possan[1], Leandra Schneider[1], Karina Carvalho Higa[1], Karen Luiza Ramos Socher[1], José Eduardo Thomaz[1], Johny Bard de Carvalho[1]

[1]Hospital Nossa Senhora das Graças, Curitiba PR, Brazil.

Correspondence: Mateus Pinto Marchetti; Email: mateus.marchetti@live.com.

Case presentation: APF, 74-year-old female, admitted for an investigation of a 3 month onset ataxic gait, progressing to abasia in 1 week, associated with mood changes. Initial assessment showed left upper limb ataxia, grade 3 lower limbs paraparesis, abasia, upper limbs hyperreflexia, lower limbs hyporreflexia, hypopalesthesia, tactile, termal and pain hypoesthesia up to L1 level in lower limbs, dysarthric, dysprosodic, with liquid dysphagia and neurogenic bladder. Neuroaxis magnetic resonance imaging (MRI) revealed altered signal in corpus callosum's splenium (15x11mm) and left posterior fornix branch (10x6mm), characterized by T1 hypointensity, FLAIR/T2 hypersignal, DWI and ADC hypersignal, and gadolinium contrast enhancement; nodular foci with contrast enhancement were seen in cervical spinal cord, dorsal spinal cord, and conus medullaris, associated with diffuse spinal cord enhancement and thickening of cauda equina roots. Cerebrospinal fluid analysis showed 144mg/dL proteins, 29/mm^3 leukocytes (90% lymphocytes, 10% macrophages), 7mg/dL glucose, 10.4mmol/L lactate, and normal immunophenotypic analysis. Solid and hematologic neoplasm investigations (bone marrow biopsy and immunophenotyping) were negative. During hospitalization, the patient showed increased lesions on magnetic resonance Imaging, progressing to lower limb plegia. Central nervous system pathological diagnosis was consistent with Langerhans cell histiocytosis (positive for Vimentin, S100, CD68, and CD99 markers). There was no response to pulse therapy, starting then, treatment with Trametinib and Dabrafenib. **Discussion:** Langerhans Cell Histiocytosis is an inflammatory myeloid neoplasm. It commonly affects children aged 1 to 3 years, more frequently males. Its incidence in adults is not well defined. The disease predominantly involves bones (80%), and central nervous system involvement (LCH-CNS) is rare, with the hypothalamic-pituitary-adrenal axis being the most commonly infiltrated site. Clinical presentation of tumorous LCH-CNS depends on the location and type of central nervous system involvement. MRI is the imaging modality of choice for central nervous system evaluation, typically showing expansive lesions that are hypointense on T1, hyperintense on T2, and enhance post-contrast. Biopsy is necessary for diagnosis confirmation, revealing clonal neoplastic proliferation with CD1a, CD207, and S100 expression. Another important diagnostic marker is the BRAFV600E mutation, seen in 25 to 60% of patients. Prognosis depends on the extent of involvement and is worse if located in the central nervous system. **Final comments:** Faced with an extremely rare case of Langerhans Cell Histiocytosis, both due to the location of lesions and the patient's age and sex, this prompts us to consider this disease in the differential diagnosis of expansive central nervous system lesions, aiming for better prognosis through early diagnosis and treatment.

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Langerhans cell histiocytosis presenting with pachymeningitis and epilepsy: a case report

Luiz José Malta Gaia Ferreira Júnior[1], Fernanda Ferreira de Abreu[1], Gabriela Rodrigues Tomaz[1], Daniel Delgado Seneor[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Leonardo Villaverde Buback Ferreira[1], Fernanda Lustosa Cabral Gomez[1], Larissa Braga Mendes[1], Orlando Graziani Povoas Barsottini[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Luiz José Malta Gaia Ferreira Júnior; Email: jr_malta@hotmail.com.

Case presentation: A 57-year-old patient with a history of recurrent pruritic vulvar lesions spanning 14 years presented initially with physical examination findings of friable erythematous-papular lesions in the vulvar and perineal regions. Infectious screening was negative. A biopsy of the lesion revealed Langerhans cell histiocytosis, confirmed by immunohistochemistry with the following markers: positive for S100 and CD1A, and negative for CD68. Treatment commenced with topical therapy, coupled with regular monitoring for systemic involvement. Neurological evaluation was prompted by the patient's recent onset of episodes involving brief behavioral arrests over the past year. Despite a comprehensive neurological examination yielding no abnormalities, an EEG demonstrated discharges in the left temporal region. Subsequent MRI of the brain identified expansive nodular formations with multilobular enhancement along the tentorium. Additionally, a nodular subcortical expansive lesion with paramagnetic contrast enhancement was noted in the right parietal lobe, indicating potential central nervous system involvement from the underlying disease. Furthermore, radiographic findings revealed ground-glass changes in the bony walls of the paranasal sinuses, suggesting concurrent bone involvement. Given the extensive systemic manifestations, the hematology team opted to initiate chemotherapy with cytarabine and dexamethasone. Initial treatment for focal epilepsy with carbamazepine proved ineffective, leading to a switch to levetiracetam. Upon reaching a daily dosage of 1500 mg, the patient experienced no further episodes of focal dyscognitive seizures. **Discussion:** Histiocytic neoplasms are uncommon, affecting approximately 5 individuals per million population.Central nervous system (CNS) involvement in Langerhans Cell Histiocytosis poses a significant challenge in clinical management. Pituitary lesions are prevalent, affecting up to half of LCH patients, often resulting in endocrinopathies, whereas dural lesions are less frequent (5% of cases). The presence of parenchymal lesions, as observed in our case with a subcortical lesion in the right parietal lobe, is even rarer but may indicate a more severe prognosis. The diagnosis of CNS involvement, indicated by pachymeningitis and parenchymal lesion, along with the visualization of skull bone lesions, prompted chemotherapy initiation, reflecting disease progression to a multisystemic form. This case highlights the critical importance of early recognition and treatment of neurological manifestations in LCH to enhance clinical outcomes and mitigate associated mortality. **Final comments:** Central nervous system involvement in LCH is rare; however, it can present with various clinical manifestations, many of which impact patients' quality of life and lead to a worse prognosis for the disease. Therefore, early recognition is crucial so that treatment can be effective in improving patient outcomes.

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Late onset ataxia: Gerstmann-Straussler-Scheinker disease a diagnosis to consider

Verena Miranda de Souza[1], Alberto Rolim Muro Martinez[1], Tania Aparecida Marchiori de Oliveira Cardoso[1], Marcondes Cavalcante França Junior[1], Mayko Nascimento Merscher[1], Rafael Saliba Helmer[1], Maria Paula Oiticica de Jesus[1], Kaio Cezar Ferreira[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Verena Miranda de Souza; Email: vemirandasouza@gmail.com.

Case presentation: A 46-year-old previously healthy woman presented with a 4-year history of a subacute onset, but progressive, loss of balance and gait instability with frequent falls. Two years after the symptoms onset, she experienced, also in a progressive manner, dysphagia and dysarthria, along with a hearing loss. Neurological examination disclose scanning speech, dysarthria, square-wave-jerks, bidirectional nystagmus, in addition to vestibular hyporeflexia. There was also limb uncordination, more pronounced in the left segments and unstable gait requiring unilateral support. Motor strength and sensory perception were preserved. The Scale for the Assessment and Rating of Ataxia (SARA) score was 21.5. Complementary investigation was unremarkable and included cerebrospinal fluid annalisis, infectious, paraneoplastic, autoimmune and metabolic screening. Brain and spinal cord MRI revealed signs of mild cerebellar atrophy. A genetic test revealed a heterozygous mutation in the PRNP gene (p.Pro102Leu), associated with Gerstmann-Straussler-Scheinker disease (GSS), consistent with the late-onset progressive cerebellar ataxic syndrome. A follow-up cranial MRI conducted four years after disease onset showed evidence of decreased T2 signal in the dentate nucleus, striatum and midbrain. **Discussion:** GSS is a slowly progressive hereditary autosomal dominant neurodegenerative disease, caused by several different point mutations on chromosome 20. The most common mutation is at condon 102 of the prion protein gene (PRNP) leading to a substitution of proline by leucine. The prevalence between 1-10/100.000.000 and the clinical onset typically occurs in the fifth and sixth decades of life. Neurological signs and symptoms resulting from this mutation can be highly heterogeneous even between different family members. The clinical hallmark is a progressive cerebellar ataxia and/or parkinsonism accompanied by cognitive impairment. Dysarthria, abnormal eye movement, movement disorders (myoclonus and athetosis) pyramidal signs and symptoms can also be found in the disease course. The average illness duration is 40-50 months, which is longer than other prion diseases but relatively shorter than other genetic ataxias. Definite diagnosis requires genetic testing, however, brain MRI, RT-Quick and EEG findings can be helpful. Low signal intensity in the basal ganglia on T2-weighted MRI sequencies, similar to our case, is reported as a characteristic finding in GSS and is attributed to abnormally increased amounts of iron deposition. The loss in signal intensity on T2-w images due to iron deposition may mask the increase in signal intensity from spongiform degeneration, common in Creutzfeldt-Jakob disease, a different prion disease. **Final comments:** Late-onset ataxia is a challenging phenotype regarding its specific diagnosis. GSS, despite its rarity is a etiology to be considered specialy in those cases with familiar history and characteristic MRI findings

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Late onset multiple acyl-coenzyme a dehydrogenase deficiency mimicking inflammatory myopathy: a case report

Catarina Dantas Corrêa[1], Delcio Caran Bertucci Filho[2], Claudia Kamoi Kay[1], Paulo Jose Lorenzoni[1], Renata Dal Prá Ducci[1], Paula Raquel Do Vale Pascoal Rodrigues[1], Otto Jesus Hernadez Fustes[1], Lineu Cesar Werneck[1], Rosana Herminia Scola[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil. [2]Universidade Estadual de Ponta Grossa, Ponta Grossa, PR, Brazil.

Correspondence: Catarina Dantas Correa; Email: catarinadcorrea@gmail.com.

Case presentation: Male, 23 years old. Six-month onset of spontaneous myalgia, progressive strength loss and difficulty climbing stairs. Non-intentional weight loss of 17kg/5 months. Denied using anti-obesity medications (topiramate, metformin or triiodothyronine). No consanguinity. Used to practice football and had a history of obesity in childhood'. Maternal grandfather and four great uncles with Wilson's disease. Exam showed bilateral symmetric thigh atrophy, cervical muscles, upper and lower limbs hypotrophy associated with motor impairment: cervical 3+/5, upper limbs 4-/5 proximal and lower limbs 3+/5 proximal (MRC). Global arreflexia, except from bilateral bicipital hyporeflexia. No sensory abnormalities. Bilateral resting tremor. Myopathic gait. Blood test revealed elevated creatine phosphokinase (CK), aspartate amino transferase (SGOT), glutamic-pyruvic transaminase (SGPT) and serum B12 vitamin. Hemogram, erythrocyte sedimentation rate, C-reactive protein, renal and thyroid function, lactic acid, aldolase, ceruloplasmin and serum copper were normal. Abdominal ultrasound showed moderate hepatic steatosis. Normal thig MRI with gadolinium. Electroneuromyography with positive fibrillation and waves including paraspinal muscles, suggesting inflammatory myopathy. Multiple acyl-coenzyme A dehydrogenase deficiency (MADD) diagnosis was made after acylcarnitine dosage showed increased levels of C12, C14, C14-1 and Ca4-1/C4 and genetic b-fatty acid oxidation panel revealed a pathogenic mutation for ETFDH gene. The patient was treated with riboflavin, Q10 coenzyme, B12 vitamin, increased carbohydrate intake and reduced fat and protein intake with complete improvement of symptoms. **Discussion:** MADD is a rare, autosomal-recessive disorder caused by the lack or deficiency of the electron transport flavoprotein (ETF) or its dehydrogenase (ETFDH) in the mitochondrial flavoprotein chain. A mitochondrial flavoprotein deficiency leads to the accumulation of acylcarnitine of all chain lengths. The most common involved genes are ETFDH, ETFA or ETFB. There are three categories of MADD phenotypes: a neonatal onset form with congenital anomalies (type I) or without congenital anomalies (type II) and a mild and/or late-onset form (type III). Late-onset cases are common, with age presentation raging from adolescence to old age. Mainly manifestation is myalgia, symmetrical limb weakness with or without neck weakness and respiratory muscle weakness associated with increased CK. Our patient presented type II phenotype also affecting the liver. Due to its broad phenotypes, MADD can mimic inflammatory myopathy, Guillain Barré syndrome or myasthenia gravis. Electromyography typically reveals a myogenic pattern whereas a muscle biopsy usually shows lipidosis within type I muscle fibers. **Final comments:** MADD must be considered whenever there is acute or chronic muscle involvement associated with metabolic disorders.

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Late onset Pompe's disease as a differential diagnosis in muscular dystrophy in young adults: a case report

Tatyane Andrade de Souza Silva[1], Leandro Araújo Gomes Filho[2], Ligia de Almeida Fernandes[2], Natanael Arnor da Silva Barros Goveia[2], Patricia Gomes Pinheiro[3], Carolina Garcia Nuñez Carrijo[3], Vanessa C S Mautone Lima[2], Karina Lebeis[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gaffrée e Guinle, Rio de Janeiro RJ, Brazil.
[3]Casa Hunter, São Paulo SP, Brazil.

Correspondence: Tatyane Andrade de Souza Silva; Email: tatyane16andrade@gmail.com.

Pompe's disease (PD) is a type of glycogen storage disease (GSD) that culminates in the accumulation of glycogen in the lysosome of various tissues, especially muscles.

Its transmission is autosomal recessive and located on chromosome 17. PD is classified into two forms: Infantile-Onset Pompe Disease (IOPD), considered the classic form, and a late-onset or non-classic form (Late-Onset Pompe Disease). , LOPD), which can occur in young or adult age with manifestations of skeletal myopathy and progressive respiratory and peripheral muscle weakness. This case reports a 32-year-old male patient who arrived at the neurology outpatient clinic in 2021 complaining of difficulty performing any activity that required physical strength. The condition began at the age of 26, with paresis in the upper limbs, which progressed over a period of 1 year with difficulty climbing stairs, getting out of bed and chairs and combing one's hair. He did not present dysphagia, choking or respiratory symptoms. The patient also reported the occurrence of an episode of low back pain diagnosed with a herniated disc in 2016, having undergone physiotherapy without improvement, which led to his initial referral to neurology outpatient clinics. On this occasion, ENMG and laboratory tests were requested, which diagnosed him as having muscular dystrophy of etiology to be clarified. It showed laboratory tests from September 2020, showing dosages of 2.8nmol/h/mL of acid alpha-glucosidase (GAA), alpha-galactosidase (GLA) of 4.66nmol/h/mL and CPK of 1,000U/L . At that time, corticosteroids were prescribed, in a decision motivated by the CPK dosage, and research for Pompe disease was indicated using a molecular panel examination to investigate muscular dystrophy. The panel was carried out in August 2021 and indicated a heterozygous pathogenic variant in the GAA gene. The following month, their enzymes were measured again, resulting in GAA of 1.40nmol/h/mL and GLA of 2.76nmol/h/mL, showing a reduction in GAA levels. With the diagnosis, the patient's clinical follow-up was based on the prescription of Enzyme Replacement Therapy (ERT) with the use of GAA (Myozyme). After starting to use the medication, a new dosage of GAA and GLA activities was carried out, which presented results, respectively, of 2.58mcmol/L/h and 4.07mcmol/L/h, showing an increase in their levels with appropriate replacement. enzymatic and, consequently, therapeutic efficacy. In many cases there is a delay between the first symptoms and diagnosis, which can last up to 10 years in the adult form. Unlike what occurs with muscular dystrophies of genetic origin, PD has a well-targeted treatment, which is ERT, which degrades accumulated glycogen, stopping the progression of the disease. Therefore, early diagnosis and therapy are crucial for a significant improvement in the quality of life of patients with PD.

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Late onset Tay-Sachs disease in bBahia's countryside: a case report

Lukas Santos Freire[1], Emmanuel Santos Trindade[1], Érica Otoni Pereira Miranda[1], Giovana Andrade de Oliveira[1], Guilherme Rocha Cardoso[1], Priscila Santos Oliveira[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Érica Otoni Pereira Miranda; Email: ericaotoni15@gmail.com.

Case presentation: A male patient, 34 years old, son of consanguineous parents, reports that 5 years ago he began to suffer from progressive stiffness in the left lower limb and left upper limb, presenting difficulties with mobility and apprehension. The patient currently reports improvement in the symptoms of stiffness and weakness in the left hemibody with antiparkinsonian drugs (levodopa + benserazide 400/100mg/day and amantadine 300mg/day), but began to present similar symptoms on the right hemibody. He reports a sister and paternal uncle with similar symptoms, who are now deceased. On physical examination, there is evidence of Parkinsonian gait, resting tremor in the upper limbs, bradykinesia and significant asymmetric rigidity, more evident on the left, with preserved strength, sensitivity and coordination. Radiological examinations of the brain and spine did not provide any diagnostic information. In December 2022, mutation research was carried out on a gene panel for parkinsonism syndromes. This panel identified the missense variant c.533G>A (p.Arg178His) in heterozygosity in exon 5 of the HEXA gene, classified as pathogenic. A second pathogenic variant was not identified. Multiplex Ligation-dependent Probe Amplification (MLPA) and complete EXOMA sequencing also identified only the pathogenic variant described above. **Discussion:** Tay-Sachs disease is an autosomal recessive disorder caused by a deficiency of hexosaminidase A (HEXA), a lysosomal enzyme that normally degrades GM2 ganglioside. As a result, GM2 ganglioside accumulates in the lysosomes of nerve cells. Around 78 mutations in the HEXA gene have been described, which impair the activity of beta-hexosaminidase to varying degrees. Although the infantile form of the disease is the classic one, there are juvenile and adult presentations, due to residual HEXA activity, grouped together as Late Onset Tay Sachs Disease (LOTS). With regard to LOTS, its clinical presentation can be composed of cerebellar and neuromuscular symptoms, such as tremors, rigidity and spasticity, and psychiatric symptoms. Currently, LOTS management aims at relieving symptoms, which is why the case has shown a good response to antiparkinsonian drugs. **Final comments:** Family history, clinical presentation and identification of a pathogenic variant in the HEXA gene allow raising high suspicion of LOTS. However, as Tay-Sachs disease is autosomal recessive, requiring homozygosity or compound heterozygosity of pathogenic variants, we cannot be certain about this diagnosis at the moment, although it is very suggestive. It should be emphasized that LOTS should be considered in unusual cases of parkinsonism, although this adult form of the disease is even rarer than the others. Even without specific therapy, early diagnosis favors correct management, with antiparkinsonian drugs and psychiatric and genetic monitoring, with measurement of HEXA activity, which had not been performed, until this report, due to cost.

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Late-life education's impact on adult executive functions

Ana Luisa Carvalho Santiago[1], Clarisse Vasconcelos Friedlaender[2], Vivian Proença Lara[3], João Victor de Faria Rocha[2], Augusto Braga Fernandes Antunes[4], Carlos Leandro Silva dos Prazeres[5], Lea T. Grinberg[6], Francisca Izabel Pereira Maciel[2], Paulo Caramelli[2], Elisa de Paula França Resende[2]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[2]Universidade Federal de Minas Gerais, Faculdade de Medicina, Belo Horizonte MG, Brazil.
[3]Faculdade de Medicina Ciências Médicas de Minas Gerais, Belo Horizonte MG, Brazil.
[4]Alliar Medicina Diagnóstica, São Paulo SP, Brazil.
[5]Universidade Federal de Sergipe, São Cristovão SE, Brazil.
[6]Global Brain Health Institute

Correspondence: Ana Luisa Carvalho Santiago; Email: analuisabrasil@hotmail.com.

Background: Dementia is a progressive neurodegenerative disorder characterized by cognitive decline and its incidence in Brazil is 5 times higher in illiterates. Early life education is a well-established modifiable risk factor for dementia prevention. However, whether learning to read and write in adulthood contributes to brain health is still unknown. **Objective:** To elucidate the effects of a late-life literacy intervention on cognitive reserve and brain connectivity in illiterate adults. We conducted a longitudinal clinical trial within an adult-literacy program targeting low-educated adults which is called PROAME ("Programa de Alfabetização e Memória") **Methods:** First, 130 illiterate adults were screened at the basic education program for adults called EJA ("Educação de Jovens e Adultos"). After applying the exclusion criteria, 108 were recruited. Participants were divided into two groups: the control group (50 participants) attended the regular classes (literature, math, geography, history and sciences) and the intervention group (58 participants) attended part of the regular classes plus a specific training focused on learning how to read and write. Both groups attended their activities 4 days a week for 6 months. At the 6 month follow up visit, 77 participants successfully completed all required clinical assessments and interviews. Interviews were conducted to gather basic socio-demographic information, past and current medical history, use of tobacco, alcohol and illicit substances, physical activity levels and the presence of anxiety and depression. Participants also underwent a comprehensive cognitive assessment, which included executive functions assessment (digit span backwards, rapid number naming and phonemic verbal fluency), a functional brain MRI was performed to assess brain connectivity, especially the frontoparietal network, and a blood sample collection to measure plasma neurofilament levels. **Results:** We observed a significant increase in the episodic memory and executive functions in all participants, without a significant difference between the groups (control x intervention). When we adjusted the model for age, sex, reason for illiteracy and executive functions at baseline, we saw a significant improvement of executive functions in the intervention group. Factors that influenced in this result were attendance to classes combined with intensive literacy training, previous illiteracy related to rural areas residency, low executive performance at the baseline and lower levels of serum neurofilaments at baseline. In terms of brain connectivity, we saw that the intervention group had an increase in the frontal parietal and in the left hippocampal-temporal connectivity. **Conclusion:** Late life literacy training seems to improve executive functions, with and underlying rearrangement of the hippocampal and frontal connectivity.

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Late-onset biotinidase deficiency mimicking adult leigh syndrome: case report

Kaliny Oliveira Peixoto[1], João Paulo Barile[1], Renan Brandão Rambaldi Cavalheiro[1], Patrícia Marques Mendes[1], Igor Braga Farias[1], Paulo de Lima Serrano[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Acary Souza Bulle Oliveira[1], Marco Antônio Trocolli Chieia[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Kaliny Oliveira Peixoto; Email: kalinyop355@gmail.com.

Case presentation: 20-year-old woman, she was born cesarean section, with prolonged expulsive period and circular of cord, birth weight 2500 grams, apgar 1/3/3, she presented with neonatal hypoglycemia, transient neonatal jaundice, probable perinatal sepsis, and infant hypotonia. She had delayed neuropsychomotor development. Since childhood, she had difficulty walking, dystonia in the lower limbs and later in the upper limbs, dysphagia and dysarthria. On examination, she presented with mild spastic hypertonia in the lower limbs superimposed on a dystonic component when walking; global hyperreflexia; cerebellar tests with dysmetria and dysdiadochokinesia in the upper limbs, with the presence of a dystonic component, predominantly on the left; and ataxic gait. The brain resonance showed global cortical atrophy, hypersignal in the pyramidal tracts bilaterally, focal signal changes in the bilateral thalamus, symmetrical, and in the posterior putamen bilaterally, spectroscopy without lactate peak, but with a small inversion of the creatinine/choline peaks in the basal ganglia. Muscle biopsy demonstrated selective atrophy of type II fibers and

subsarcolemmal mitochondrial proliferation. Organic acid profiles, electroencephalogram and optical fundoscopy disclosed no abnormalities. Laboratory screening showed pre-prandial lactate at the upper limit of normality; elevated post-prandial lactate; mild hyperammonemia; mild metabolic acidosis with a slight increase in anion gap; normal uric acid, creatine phosphokinase and aldolase. Gene panel disclosed the presence of the pathogenic variant c.1330G>C (p.Asp444His) in homozygosity in the BTD gene, confirming the diagnosis of biotinidase deficiency. The use of biotin 10 milligrams/day was started, with important improvement in fine motor coordination, in gait and reduction in the frequency of falls. **Discussion:** Late-onset biotinidase deficiency is an underdiagnosed inherited neurometabolic disorder, which can present with several neurological phenotypes, such as optic neuropathy, isolated optic atrophy, recurrent myelopathy, axonal polyneuropathy, cerebellar ataxia, sensorineural hearing loss and rarely mimicking neuroimaging features seen in Leigh syndrome. As biotinidase deficiency represents a metabolic disorder currently included in newborn screening programs, late-onset phenotypes may become probably rarely described in the following decades. Leigh syndrome is a complex neurogenetic group of metabolic disorders which can be caused by several distinct genetic variants, among which BTD pathogenic variants stand out. **Final comments:** The present report raises the diagnostic suspicion index for biotinidase deficiency, a potentially treatable condition, faced with Leigh Syndrome in adults. The timely treatment institution, consequently, provides an improvement in the patient's functionality.

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Late-onset GM2 gangliosidosis due to mutations in GM2A gene mimicking spinal muscular atrophy type 3

Rodrigo de Holanda Mendonça[1], Juliana Gurgel Giannetti[2], Ronnyson Gativvol[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil. [2]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Rodrigo de Holanda Mendonça; Email: rodrigo.holanda@hc.fm.usp.br.

Non-5q spinal muscular atrophy (SMA) presents a diversity of genes described in the literature. Previously, GM2 gangliosidosis mimicking spinal muscular atrophy (related to mutations in the HEXA gene) was described. However, motor neuron disease related to mutations in the GM2A gene, which encodes a ganglioside activator protein, has not yet been described. Here, we describe a late-onset motor neuron disease phenotype mimicking SMA type 3 or 4, whose genetic investigation resulted in the finding of GM2A gene mutations. Case 1 is a male, 36 years old, history of parental consanguinity. He started progressive proximal muscular weakness in lower limbs at ten years old – weakness presented with slow progression over the years to the scapular girdle. Clinical evaluation at that time with 15 years old showed a motor neuron disease, being diagnosed as Kugelberg Welander disease (SMA type 3). At that time, neurological evaluation was remarkable for the presence of preserved tendon reflexes in 4 limbs. Weakness progressed slowly, and the patient lost his gait around the third decade of life. At the age of 35, he presented proximal and distal muscle atrophy, and tendon reflexes are now abolished, with exuberant polyminimyoclonus on examination. Cognitive is normal. Molecular investigation for deletions in the SMN1 gene ruled out that it was 5q SMA - it had 02 SMN1 copies and 00 SMN2 copies. Exome sequencing revealed a homozygous missense mutation at position c.428G> A (p.Gly143Glu) in the GM2A gene. Case 2 is a female, 43 years old, started muscle weakness of proximal predominance, exclusively affecting the lower limbs, around ten years ago and with electroneuromyography indicating a neurogenic process restricted to the quadriceps femoris and gastrocnemius muscles bilaterally. MRI of the thighs and legs showed exclusive involvement of the quadriceps femoris and gastrocnemius muscles, with a neurogenic pattern. Exome NGS showed two variants in compound heterozygosity (p.Gly143Glu and p.Gly166Arg) in the GM2A gene. Case 3 is a male, 40 years old, adult-onset muscle weakness of the lower limbs, mimicking SMA type 3, and investigation with NGS also showed two variants in the GM2A gene (p.Gly166Arg and p.Asp116His). The three cases presented here have in common a late-onset motor neuron disease mimicking SMA 3, with preferential involvement of the quadriceps and recessive mutations in the GM2A gene.

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Late-onset Pompe's disease in children: case report and genetic particularities

Maria Luiza Vasconcelos Montenegro[1], Patrícia Trindade de Lucena[2], João Eudes Magalhães[2], Fabíola Lys de Medeiros[2]

[1]Universidade de Pernambuco, Recife PE, Brazil. [2]Hospital Universitário Oswaldo Cruz, Recife PE, Brazil.

Correspondence: Maria Luiza Vasconcelos Montenegro; Email: marialuizavasc534@gmail.com.

Case presentation: Male, 9 years old, started walking after 2 years of age, at the age of six, he progressively presented global muscle weakness and difficulty walking. Similar family history was denied. Neurological examination: mild facial biparesis, hypophonia, weak cough, global hypotonia, grade 4- proximal strength in the upper limbs and grade 3 in the lower limbs, grade 4+ distal strength in all limbs, strength deficit when standing up in the supine position, reflexes hypoactive and anserine gait. He also presented thoracic kyphosis, hypotrophy of the abdominal muscles, winged scapula, and slight atrophy of the shoulder and pelvic girdles. Electroneuromyography revealed myopathy. CPK dosage was 692 U/L. Complete sequencing of the GAA gene found the presence, in heterozygosity, of the variants described as chr17:80.118.209 CCA>C (2501_2502del];p.[?(;)Thr834Argfs*49]) and Chr17:80.104.542 T>G (NM_000152.5(GAA):c.[-32-13T>G]), both classified as pathogenic, in the GAA gene, associated with alpha-glucosidase deficiency (OMIM: 232300), autosomal recessive inheritance. The acid alpha-glucosidase dosage was 0.52 mcmol/L/h, with normal being greater than or equal to 2.10 mcmol/L/h. Transthoracic echocardiogram and spirometry were normal. Pompe Disease was diagnosed and he has been receiving an infusion of 12 vials containing 50mg of alpha-glucosidase, every 15 days for a year, undergoing motor physiotherapy. He has improved trunk strength, can stand up without supine support, strength grade of 4+ overall. Currently, he rides a bicycle. **Discussion:** The infantile form affects children in the first months of life, with cardiomyopathy, hypotonia, and rapidly progressive muscle weakness, and progresses to death within the first year of life. Late-onset disease can manifest between the first year and the sixth decade of life. Typically the late form does not present severe cardiomyopathy and disease progression is slower. Muscle weakness predominates in the proximal muscles with significant impairment of the lower limbs, resulting in impairment in carrying out daily activities, as seen in the reported case. The c.-32-13T>G variant is the most common GAA mutation associated with the childhood/adult phenotype. Almost 90% of patients affected by this phenotype carry this mutation on at least one allele. The chr17:80,118,209 CCA>C variant is quite rare, being present in heterozygosity in 2 of approximately 125 thousand individuals in the world population. Both pathogenic. **Final comments:** Pompe disease described is possibly caused by these two heterozygous variants in the GAA gene. The relevance of this case demonstrates that the disease started early, at least after 18 months, with a very slow progression and affecting the trunk muscles and that one year of enzyme replacement brought a lot of independence to this child. We also emphasize the Compound Heterozygous Mutations and rarity of the chr17:80.118.209 CCA>C variant, found in this patient.

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Lateralized periodic discharges (LPDS) with identical periods

Taissa Ferrari Marinho[1], Luciana Rodrigues de Marchi[1], Luis Otavio Caboclo[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Taissa Ferrari Marinho; Email: taissaferrari@yahoo.com.br.

Case presentation: A 77-year old woman with high grade glioma on splenium of the corpus callosum was admitted in the hospital with cough, dyspnea, mental confusion and fever. After investigation, she started treatment for pneumocystosis. Five days after antibiotic theraphy initiation, she developed apathy, inattention, and presented an epileptic focal seizure characterized by behavioral arrest and right hand automatisms. She received 5mg IV diazepam and was taken to the intensive care unit. After two more clinical seizures, she received phenytoin. Patient evolved to consciousness impairment and started continuous electroencephalogram monitoring that showed diffuse disorganization of background activity and asymmetry, slower in the left hemisphere, infrequent triphasic waves, frequent epileptiform abnormality in the left temporal region, with a periodic pattern lateralized periodic discharges (LPDs) with identical periods of 0.5-0.6Hz, and eletrographic seizures on the left temporal region. Cerebral fluid showed herpes simples virus type 2. Patient had status epilepticus, received two more antiseizure medications and was sedated with propofol with seizure control. Although patient had seizure control, she persisted with severe sequelae of consciousness impairment. **Discussion:** LPDs are a frequent EEG pattern that have been described as an interictal, ictal and post ictal pattern, therefore the real significance of this pattern may be challenging in clinical practice. Recent studies showed that LPDs occur with a variety of etiologies and have the highest association with seizure risk, regardless of frequency, mainly when the Plus modifier is present. Our patient showed LPDs with identical periods; to the best of our knowledge, this pattern was not found in the literature before. LPDs with identical periods may have specific pathophysiology. Our patient had a high grade glioma that compressed the thalamus, a structure called the brain "pacemaker" by some authors, due to its relation to sleep spindles and periodic discharges. In addition, she had herpes encephalitis, which is a severe brain infection frequently associated with periodic pattern on EEG. Both diseases are of poor prognosis. Comatose patients after cardiac arrest can show malignant EEG patterns, that predict a poor outcome; burst-suppression with identical bursts, is one of these patterns. Would the identical periods seen in this case of LPDs be an indicative of poor prognosis, as seen with burst suppression with identical bursts in post anoxic patients? Would the identical repetition of morphology and time be of predictive value in patients with severe diffuse brain lesions? **Final comments:** LPDs is an EEG pattern frequently seen in critically ill patients, and show a high risk of seizure occurrence. LPDs with identical periods is a new specific pattern that could be associated with poor prognosis. Further studies will be needed to prove this hypothesis.

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Left arm palsy and cerebellar ataxia revealing mycobacterium avium infection in a patient with HIV and systemic lupus erythematosus: a case report

Alex Araújo Fernandes[1], Matheus Alves da Silva[1], Matheus Luís Braga Munareto[1], Eduardo Mesquita de Souza[1], Rafael Bragança Rodrigues Matias[1], Ana Luísa Faria Donadon[1], Amanda Miranda de Mendonça[1], Christiane Monteiro de Siqueira Campos[1], João Antônio Gonçalves Garreta Prats[1], Alex Machado Baeta[1]

[1]Beneficência Portuguesa, São Paulo SP, Brazil.

Correspondence: Matheus Luís Braga Munareto; Email: matheusbragamunareto@gmail.com.

Case presentation: A 38-year-old woman presents with headache, myalgia, diarrhea and non-specific malaise for 3 days. She is diagnosed with systemic lupus erythematosus, using hydroxychloroquine, and 3 months ago she was diagnosed with HIV infection, undergoing treatment with dolutegravir, lamivudine and darunavir/ritonavir and currently with a viral load of 593 copies and CD4 of 248. Initially, the hypothesis of dengue was made, but the NS1 antigen test was negative 2 times. Stool culture and investigation of opportunistic agents in feces were requested, with negative results. Due persistence of headache and appearance of paresis of the left upper limb associated with appendicular and axial cerebellar ataxia, a head CT was made with a finding of focal hypoattenuation in the subcortical white matter of the right precentral gyrus. A MRI performed showed a cortico-subcortical nodular lesion in the right precentral gyrus with peripheral gadolinium enhancement and a central area with diffusion restriction associated with a double halo sign on T2. Also, there was nodular thickening of the pachymeningeal plane in the left temporo-occipital convexity, associated with infiltration of the adjacent bone plate with gadolinium impregnation and signs of diffusion restriction and leptomeningeal enhancement of the pial surface of the cerebellar vermis and in grooves between cortical gyri in the superior aspect of the cerebellum, associated with T2/FLAIR hyperintensity in the adjacent parenchyma. The cerebrospinal fluid analysis showed 6 cells, 69 proteins and 35 glucose, tests for microorganisms, including GeneXpert for tuberculosis, and antibodies were negative. Due to the characteristics of the precentral lesion on the MRI being suggestive of neurotoxoplasmosis, treatment with Sulfamethoxazole and Trimethoprim was chosen empirically, however, in radiological control after 15 days, a worsening of the lesions was observed with an increase in size and she had no clinical improvement. It was decided to perform a brain biopsy that showed a chronic granulomatous inflammatory process with central necrosis, and a positive culture for Mycobacterium avium. **Discussion:** Non tuberculous mycobacteria are rare human pathogens and usually infects immunocompromised hosts. These bacteria are divided in two main groups: Low-growing mycobacteria (Mycobacterium avium complex (MAC) and rapidly growing mycobacteria (Mycobacterium abscessus). The neurological manifestation of these diseases includes meningitis and/or parenchymal lesions. The diagnosis is usually made by tissue sampling. **Final comments:** With this case it was possible to observe a clinical and radiographic presentation of Mycobacteria avium infection, a rare central nervous system infection. This disease is usually difficult to diagnose and early diagnosis is essential for successful treatment.

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Left oculomotor nerve paralysis after dengue infection: a rare neuroinfectious journey

Melissa dos Santos Matos[1], Daniel Mariani Favalessa[1], Guilherme Valiate Picoli[1], Emanuelly Victoria Andrade Fonseca[1], Ivo Gabriel Ribeiro Carvalho[1], Jovana Gobbi Marchesi Ciriaco[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Melissa dos Santos Matos; Email: melissa.matos@edu.ufes.br.

Case presentation: An 82-year-old female patient presented with vertical diplopia in the left eye and frontal headache at night. She came to the emergency room in February 2024. Initial examinations, including magnetic resonance imaging (MRI), ruled out stroke or aneurysm, and she was released. However, four days later, she was readmitted due to continuing symptoms. A new MRI revealed asymmetry of the left cavernous sinus with homogeneous contrast uptake. After further investigation, Tolosa-Hunt Syndrome was ruled out, based on the 2004 International Headache Society diagnostic criteria, and corticosteroid therapy was prescribed with methylprednisolone, 1 g/day, for three days. However, her condition progressed to complete left eyelid ptosis. Previous pathological history revealed an episode of dengue fever in January of the same year, confirmed by IgM. After careful analysis, she was diagnosed with left cranial III nerve palsy as a result of the dengue infection. **Discussion:** The dengue epidemic in Brazil is one of the main public health challenges today. Although the range of clinical signs of the disease is widely known, rare complications can occur, especially related to the DENV2 and DENV3 serotypes. In the case reported, the left oculomotor nerve palsy was manifested by vertical monocular diplopia, eyelid ptosis and frontal headache. The MRI findings, together with the exclusion of compressive lesions of the nerve and the absence of other infectious or inflammatory causes, establish the link between III nerve palsy and dengue infection. The most common causes of third cranial nerve palsy are ischemic, but they can also be secondary to viral infections. The pathophysiology of dengue-related neuritis is not yet clearly defined, although there are indications of immune-mediated mechanisms. The diagnosis should be made by excluding other etiologies, taking into account the history of viral infection. Treatment is controversial, generally opting for support, with the use of corticosteroids in refractory cases. Complete remission of symptoms can occur in up to 6 months. **Final comments:** Due to its long intracranial course, the oculomotor nerve is susceptible to injury at various anatomical points. Although it shows obvious clinical signs, III nerve palsy can have multiple causes, including neoplasia, demyelination, trauma and infections such as dengue fever, which often makes defining the etiological diagnosis a challenge. The growing incidence of neurological manifestations related to the immune-mediated reaction following dengue infection highlights the need to consider this etiology in cases of cranial nerve palsy - although they are still poorly described in the literature. Therefore, this case report highlights the importance of investigating infectious causes, especially in endemic areas such as Brazil, where dengue is prevalent, alerting us to the relevance of a broad and timely approach in similar cases.

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Left temporal arteritis associated with ipsilateral amaurosis

Fábio Gonçalves[1], Felipe Leonardo Semensati[1], Letícia de Barros Dias[1], Marcelo Simplicio Carvalho[1], Nathalye Fernanda Pedroso Dircksen[1], Lorena Fernandes Kronbauer[1], Mônica Meneguette Silveira[1], Bárbara Okabaiasse Luizetti[1], Lucas de Oliveira Pinto Bertoldi[1], Damacio Ramon Kaimen Maciel[1]

[1]Irmandade da Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Felipe Leonardo Semensati; Email: felipesemensati@hotmail.com.

Case presentation: T.A.B.S., 57, female, started with sudden pain in the left eye, ipsilateral amaurosis and hemicranial headache. Presented, as a previous comorbidity was hypertension, diabetes, dyslipidemia and glaucoma, using Losartan, Anlodipine, Metformin, Simvastatin and insulin. Attended at the Eye Hospital the following day with evaluation: Normal eye movement; Bilateral visual acuity 20/20; Bilateral intraocular pressure without changes. Fundus of eye: right eye without alterations and left with alteration of the foveal coloration. Referred for neurological evaluation, with hospitalization. At the entrance, at the neurological examination, an alteration was found in the ocular examination: visual turbidity in the left eye with predominance in the temporal field. Without changes on head CT and with changes in the cerebrospinal fluid in: Glucose 155; Proteínas 56; Lactate 3.3. 20 CmH20 manometry. In laboratory tests, increased VHS (56) and normal PCR (0.1). Following etiological investigation, skull resonance - which showed microangiopathy, and orbits resonance - was performed with hypersignal in the intra-orbital portion of the optic nerve on the left. Proposed diagnostic hypothesis of optic neuritis, instituted treatment with methylprednisolone 1 gram/day for 5 days. He maintained left amaurosis and ipsilateral hemicranial headache, being discharged with prednisone and outpatient follow-up. Following in etiological investigation, oligoclonal bands and anti-aquaporin 4 were performed, both without alterations. Left temporal artery biopsy was performed confirming temporal arteritis of giant cells. Maintained outpatient follow-up, evolved with improvement of headache. **Discussion:** Giant Cell Arteritis (ACG) is a granulomatous vasculitis that affects medium- and large-caliber arteries, particularly temporal arteries. Pathogenesis involves infiltration of the arterial wall by inflammatory cells, including T lymphocytes and macrophages, leading to inflammation and vascular damage. It manifests itself with systemic and specific symptoms. Systemic: fever, fatigue and weight loss and specific: headache, sensitivity in the scalp and lameness of the jaw. The most serious complication is sudden monocular amaurosis due to ischemia of the optic nerve. The diagnosis is clinical, supported by laboratory tests (VHS and PCR) that will often be elevated. Diagnostic confirmation can be obtained by biopsy of the temporal artery, which shows granulomatous inflammation. The recommended treatment is corticosteroid therapy to reduce inflammation and prevent ischemic complications. **Final comments:** Sudden monocular amaurosis, resulting from GCA, is an emergency that requires immediate diagnosis and treatment to preserve vision. Therefore, early recognition and aggressive treatment is essential to avoid complications and improve the prognosis of patients.

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Leptomeningeal disease in genitourinary tract cancer manifesting as intracranial hypertension: case reports

Pedro Henrique de Oliveira Silveira[1], Verônica Guerra Matuella[1], Thales Pardini Fagundes[1]

[1]Fundação Pio XII, Hospital de Câncer de Barretos, Barretos SP, Brazil.

Correspondence: Thales Pardini Fagundes; Email: pardinithales@gmail.com.

Case presentation: We described two patients with genitourinary tract cancer complicated by leptomeningeal disease (LD). The first case involves a 61-year-old male with metastatic, castration-resistant prostate adenocarcinoma. After completing chemotherapy with docetaxel in December 2023, he presented in February 2024 with acute bifrontal headache, cognitive-behavioral fluctuation, and nausea. Examination revealed sixth cranial nerve paresis, psychomotor slowing, and disorientation. Brain MRI demonstrated post-gadolinium enhancement in the sulci of the right frontal and parietal regions, loss of signal suppression in post-contrast FLAIR sequence, and leptomeningeal enhancement in post-contrast T1 sequence, confirming LD. The second case is a thirty-four-year-old female with mixed squamous and adenocarcinoma cell cervical cancer. Post platinum-based chemotherapy in September and November 2023, she presented with right-sided hearing and visual loss, vertigo, and headache. Examination revealed a lateralized

Weber's test, papilledema, and optic nerve pallor. Brain MRI was inconclusive for LD, but cytology of three CSF samples showed neoplastic cells and elevated opening pressure of 60 cmH2O. Her lumbar puncture also revealed an elevated white cell count, suggestive of an inflammatory process secondary to the neoplastic involvement. The patient's management included corticosteroids to reduce intracranial pressure and alleviate symptoms, in addition to whole-brain radiotherapy. **Discussion:** LD diagnosis significantly impacts overall survival, which ranges from four to eight weeks. In genitourinary tract malignancies, LD is extremely rare. A retrospective review at MD Anderson Oncology Center revealed that among 41,830 prostate cancer patients from 1979 to 2011, only seven (0.016%) had LD. Similarly, in cervical cancer, only 0.03% of metastatic cases are associated with LD. Early recognition of neurological symptoms is vital for prompt diagnosis. Cytological CSF analysis and MRI with post-contrast enhancement are the most reliable diagnostic tools, with post-contrast FLAIR sequence being more sensitive than T1-weighted sequence. Effective supportive measures are essential due to the poor penetration of systemic therapy in the central nervous system and the rapid decline in performance status in these patients. **Final comments:** The diagnosis of LD is critical for prognostic and therapeutic decision-making, given the rapid deterioration in these patients. Current systemic antineoplastic therapies have limited central nervous system penetration. Clinical, diagnostic, and therapeutic discussions of LD in genitourinary cancers can provide valuable references for similar future cases, highlighting the need for more robust scientific evidence and effective treatments.

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Levamisole-induced leukoencephalopathy

João Victor Cabral Correia Férrer[1], Lara Santos Machado[1], Luiz José Malta Gaia Ferreira Júnior[1], Luccas Almeida Andrade[1], Marianna Pinheiro Moraes de Moraes[1], Pedro Henrique Almeida Fraiman[1], Gabriel Pinheiro Martins de Almeida e Souza[1], Marcio Pinheiro Lima[1], José Luiz Pedroso[1], Orlando Graziani Povoas Barsottini[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: João Victor Cabral Correia Férrer; Email: jvccferrer@gmail.com.

Case presentation: A 62-year-old woman presented a 15-year-old history of progressive gait abnormality, with imbalance and frequent falls. After 5 years of disease progression, she required bilateral support to walk and, after 10 years, the patient was restricted to a wheelchair and exhibited cognitive dysfunction. Cocaine consumption was associated with de first clinical symptoms, and clinical stability was obtained since drug abstinence, which was approximately 2 years ago. It was noted that the patient's husband exhibited similar symptoms and also had a history of cocaine addiction. Physical examination evidenced axial and appendicular ataxia, spastic paraplegia and cognitive impairment primarily affecting executive functions. Brain magnetic resonance imaging (MRI) showed symmetrical leukoencephalopathy, including U-shaped fibers, with white matter cavitations, especially in periventricular regions, and thinning of the corpus callosum (FIGURE 1). Cerebrospinal fluid (CSF) evaluation was normal. Serum infectious, metabolic and autoimmune investigation was negative. LILE was diagnosed considering the clinical findings e its stability linked to drug abstinence, the presence of related person with similar exposure and symptoms, and the MRI typical abnormalities. **Discussion:** LILE predominantly affects females, between the 4th and 5th decades of life. Its progression is generally subacute and there are reports of recurrence of associated syndromes with re-exposure to levamisole, possibly due to a genetic predisposition. The three main clinical manifestations are muscle weakness, with or without hypertonia or hyperreflexia, language impairment and cognitive dysfunction, but cerebellar ataxia, altered level of consciousness and sphincter symptoms are also described. Brain MRI most commonly shows T1-hypointensity and T2/FLAIR-hyperintensity abnormalities in cerebral white matter characterized by asymmetric involvement in periventricular area, predominantly in the frontal region, with or without contrast enhancement related to blood-brain barrier disruption. CSF evaluation shows inflammatory pattern in 41% of cases, and elevated IgG index is the most frequent finding, followed by lymphomononuclear pleocytosis and hyperproteinorrachia, but oligoclonal bands were present in a minority of cases. Treatment consists in drug abstinence and pulse therapy with methylprednisolone and/or intravenous immunoglobulin and/or plasmapheresis. The prognosis is favorable in 96.7% of patients. **Final comments:** Thus, it is evident that both cases present clinical history and neuroimaging findings described in cases of LILE. However, the chronic course due to frequent re-exposure to this substance poses a diagnostic challenge. This atypical form of progression should be recognized and considered as a differential diagnosis in cases of chronic leukoencephalopathies when pertinent.

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Light chain neurofilament in biofluids and its application as a potential biomarker in amyloidosis: preliminary findings

Larissa Bittencourt de Carvalho Chaffin[1], Ligia Rocha Andrade[1], Renan Amphilophio Fernandes[2], Soniza Vieira Alves-leon[2], Andreza Salvio Lemos[2], Izabela Jardim Rodrigues Pitta[3], Clarissa Neves Spitz[1], Robson Teixeira Vital[1], Salim Lázaro Balassiano[1], Marcia Maria Jardim Rodrigues[1]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Fundação Oswaldo Cruz, Rio de Janeiro RJ, Brazil.

Correspondence: Larissa Bittencourt de Carvalho Chaffin; Email: laribit@hotmail.com.

Background: Peripheral neuropathies are a very common clinical condition and are associated with numerous cases of functional disability. There is a wide range of differential diagnoses associated with polyneuropathies, which can make it difficult to identify their etiology. The increasing emergence of treatments for these conditions demands the need to monitor the ideal therapeutic response. This monitoring often does not end with clinical and neurophysiological assessments and requires the use of biomarkers for neuronal damage, such as the measurement of neurofilament levels. **Objective:** To evaluate neurofilament light chain levels in biofluids as a biomarker of disease activity in patients with familial amyloid polyneuropathy associated with transthyretin. Investigate whether there is an increase in the dosage of neurofilaments in carriers of the transthyretin gene mutation, even if asymptomatic and after the first symptoms of the disease. **Methods:** Patients with a mutation in the transthyretin gene, whether symptomatic or asymptomatic, were selected. Patients who had comorbidities potentially causing peripheral neuropathies were excluded. All of them underwent electroneuromyography. Physical examination and clinical history data were used to classify patients on functional scales. Blood was collected in 4 milliliter samples and deposited in 2 to 3 tubes with ethylenediaminetetraacetic acid anticoagulant. The material is then centrifuged to separate plasma and serum. After centrifugation, the plasma was separated from the buffy coat and added to suitable tubes. Plasma, serum and buffy coat samples must be stored in a -80 °C freezer for later use. Plasma neurofilament concentration was measured by ultra-sensitive single molecule array technique. Preliminary **Results:** Of the 19 patients evaluated, only 2 had neurofilament levels comparable to healthy controls. Both were asymptomatic. All Coutinho stage 2 patients had plasma neurofilament levels about 4-fold higher at baseline relative to healthy controls. One of the patients showed a reduction of approximately 70% in plasma neurofilament levels 6 months after starting tafamidis, although he experienced clinical worsening. The use of biomarkers has been suggested as a means of stratifying patients and monitoring treatment. This is important because neurofilament is an integral component of the axonal structure of neurons, and the neuropathy observed with transthyretin amyloidosis is often characteristic of axonal injury. A 2021 publication showed that serum neurofilament dosages were four times lower in healthy patients than in patients diagnosed with transthyretin amyloidosis. **Conclusion:** The recent emergence of disease-modifying therapies demands the development of biomarkers so that treatment can be initiated earlier. This might prevent the development of disability by these individuals and greater success of therapy.

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Limb girdle muscular dystrophy: autosomal recessive calpainopathy, case report

Laura Altomare Fonseca Campos[1], Jéssica Cristina Silveira Damasceno[1], Marcelo Sobrinho Mendonça[1], Thiago Cardoso Vale[1]

[1]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.

Correspondence: Laura Altomare Fonseca Campos; Email: lauraaltomare8@gmail.com.

Case presentation: A 45-year-old previously healthy male patient, was evaluated with a history of mainly proximal weakness in the lower limbs with onset around the age of twelve with progressive worsening until the age of twenty-five, when he became dependent on the use of a wheelchair. He later developed proximal weakness in his upper limbs. There was no report of a similar family history or consanguinity. Physical examination showed proximal flaccid tetraparesis, predominant in the lower limbs, accompanied by global hypotonia with proximal atrophy in all four limbs. Electroneuromyography (ENMG) was compatible with myopathy in axial muscles, proximal upper and lower limbs, corroborating the diagnostic hypothesis of limb-girdle muscular dystrophy. A broad DNA panel was carried out and revealed a pathogenic mutation in homozygosity in the gene calpain 3 (CAPN3). This gene is responsible for the autosomal recessive limb girdle muscular dystrophy type 1 (LGMDR1). Magnetic resonance imaging (MRI) of the left hip, showed marked fatty degeneration of the muscle bellies of the pelvic girdle, proximal thigh and lower abdominal wall. Laboratory tests showed an increase in muscle enzymes. **Discussion:** Autosomal recessive calpainopathies are caused by pathogenic mutations in the CAPN3 gene, a protease involved in the cytoskeleton of muscles, and are the most prevalent subtype of limb girdle muscular dystrophy. It usually manifests in childhood, with variable phenotypes, from asymptomatic to those with proximal muscle weakness in the pelvic and scapular girdles, contractures and gait disorders. Patients are cognitively normal in most cases and progress from childhood with weakness, muscle atrophy, slow and progressive changes in gait, to loss of ambulation. Although the facial, cardiac and pulmonary muscles are generally spared in autosomal recessive calpainopathy, cardiac and pulmonary involvement can occur in girdle dystrophies, so it is necessary to carry out cardiological tests to screen for cardiomyopathies, abnormalities in the cardiac conduction system and ventilatory disorders. **Final comments:** It is important to make a differential diagnosis with other causes of proximal muscle weakness, in order to carry out the correct approach, screening tests for possible complications in the natural history of the disease, laboratory alterations, as well as appropriate rehabilitation and genetic counseling.

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Limb levitation and alien limb as a rare initial manifestation of high grade glioma (glioblastoma multiforme)

Julyana Leoni do Prado[1], Mariana de Oliveira Trintinalha[1], Felipe Cardoso Garcia[1], Carlos Alberto Mattozo[1], Marina Farah[1]

[1]Pontifícia Universidade Católica do Paraná, Hospital Universitário Cajuru, Curitiba PR, Brazil.

Correspondence: Julyana Leoni Prado; Email: julyana.leoni@gmail.com.

Case presentation: Female patient, 61 years old, right-handed, treated on an outpatient basis complaining of weakness in the left hand and episodes of involuntary movements and a sensation of lack of control over the left upper limb. The patient describes that those movements interfere with daily activities, such as eating and in her job as a cook, especially performing unintentional activities, such as handling food while eating. There is still a feeling of weakness dropping objects. The patient had history of breast cancer treated with left breast quadrantopsy, radiotherapy and tamoxifen and had family history of neoplasia, 2 cousins of maternal origin diagnosed with neoplasia of the central nervous system. Neurological examination: grade IV left upper limb strength with distal predominance, ideomotor apraxia and slow elevation of the left upper limb triggered by tendon percussion and tactile stimulation, interrupted with interference from the examiner. Investigation with Skull Magnetic Resonance Imaging showed a lesion with mass effect on the right parietal lobe and signs of intracranial hypertension. She underwent resection and biopsy, confirming grade IV Glioblastoma multiforme (high-grade). After surgery, she obtained complete resolution of involuntary movements and, at that moment, grade IV left hemiparesis. Radiotherapy and adjuvant chemotherapy was performed. Patient remains under follow-up. **Discussion:** High-grade gliomas correspond, on average, to 2% of all gliomas. Peak incidence: 65 to 74 years. There is a slight predilection for males, with a ratio of 1.5 men to 1 woman. They may have different clinical presentations depending on topography and volume. Larger ones can develop intracranial hypertension. Seizures and focal neurological deficits are common presentations at diagnosis. One reported patient presented with atypical limb levitation and hand syndrome, which are more frequent in neurodegenerative diseases such as Corticobasal Degeneration (CBD). Alien hand syndrome can be divided into anterior, present in lesions of the frontal lobe, manifested by difficulty in holding objects, or in callous lesions, in which the affected limb makes conflicting movements in relation to the unaffected one. And into posterior syndrome, generally located in the parietal lobe, that presents with less complex movements of the affected limb (levitation, for example); this last variant was manifested by the patient described. Standard treatment for glioblastoma includes resection of the largest tumor margin, followed by radiotherapy and chemotherapy with Temozolomide (TMZ), a 5-year survival rate of 7.2%. In the case described, the patient relapsed after 4 months. **Final comments:** We describe a case of Glioblastoma Multiforme with an atypical manifestation of limb levitation, an unusual finding as a manifestation of significant neoplasms, highlighting the importance of investigating other non-degenerative causes in such presentations.

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Limb-girdle muscular dystrophy TRAPPC11 related (LGMDR18) and central nervous system involvement

Diego Silva Figueiredo[1], Nadson Bruno Serra Santos[1], Alexandre Motta Mece[1], Aldrin Pedroza Martins[1], Izadora Vieira Bolzani Lopes Lima[1], Marcondes Cavalcante França Junior[1], Alberto Rolim Muro Martinez[1], Anamarli Nucci[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Diego Silva Figueiredo; Email: ddiegofigueiredo@gmail.com.

Case presentation: A 22-year-old man was attend to our outpatient clinic at age of 14 due to myopathy without an etiological diagnosis. His parents are consanguineous (first cousins). The patient presented normal development, except for speech delay, which he improved with the help of speech therapy. Since age 3 when he began experiencing motor incoordination, with difficulty walking in a straight line and postural instability. Furthermore, he had some difficulty learning and memorizing at school. At 14-years -old, the patient denied weakness, but reported myalgia, especially in the lower limbs after long walks and sports. On physical examination, he presented MRC grade IV+ in proximal upper and lower limbs, mild global hypotonia, normoactive deep tendon reflexes, dysmetria on the index-nasal-index maneuver without worsening with eyes closed, bidirectional horizontal nystagmus, ataxic gait and choreoathetoid movements in the distal region of the upper limbs. Complementary exams: all basic biochemical exams were within normal limits, except for elevated CK, up to 1096, LDH 532, aldolase 24.2. Negative acanthocyte test. ENMG showed a myopathic pattern in proximal and distal muscles in the four limbs, without detection of fibrillations or positive waves. Muscle biopsy and brain MRI were normal. He remained with stable symptoms over the years, with a diagnosis of myopathy plus central nervous system involvement without a specific diagnosis. In 2021, na exome detected a biallelic pathogenic variant in the TRAPPC11 gene, c.1287+5G>A, colocar a proteína também, compatible with the diagnosis of LGMDR18 (p.Ala372_Ser429del) **Discussion:** This case report presents a patient with myopathy, cerebellar ataxia, hyperkinetic disorder and speech delay. Myopathic pattern is compatible with LGMD. An exome revealed a homozygous pathogenic TRAPPC11 gene mutation (c.1287+5G>A) similar the description by Bogershausen N et al, 2013, in Hutterites families. These authors emphasized the disease spectrum of LGMD (latter named LGMDR18) and myopathy with movement disorder and intellectual disability. Muscle biopsy and brain MRI may not show changes. There is no specific treatment for this condition. **Final comments:** To date, we are unaware of any Brazilian case report of LGMDR18. Increasing knowledge of the pathogenic variants in LGMD and myopathy "plus" enable the diagnosis of many patients previously under follow-up and without a defined etiology, as occurred in our patient.

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Linfoproliferative malignancy recidive as a stroke mimics

Maria Luiza Levindo Coelho Martinis[1], Bárbara Luisa Coraspe Gonçalves[1], Luiz Guilherme Gerhardt Fernandes[1], Marina Brasileiro Cesar Leitão[1], Bárbara França Amorim[1], Osvaldo Pantoja de Oliveira[1], Ana Carolina Andorinho de Freitas Ferreira[1], Cristiane Borges Patroclo[1], Marcelo Cagy[1], Jamison Menezes de Souza[1]

[1]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.

Correspondence: Maria Luiza Levindo Coelho Martinis; Email: mlevindocoelho@gmail.com.

Case presentation: Male patient, 19 years old, woke up with paresthesia in the right upper limb. He went to the emergency room and a brain CT disclosed an extensive hypodense area in the left hemisphere. He received the diagnosis of ischemic stroke and was discharged. Then, he sought hospital for better comprehension about his condition. On neurological examination, the patient presented subjective paresthesia without objective sensory impairment, and there was only a slight pronator deviation on his right. His anamnesis included a prior history of T-type lymphoblastic leukemia supposedly cured when he was 15 years old. Therefore, the clinical radiological dissociation observed elicited the suspicion of leukemic recurrence, this time in the central nervous system (CNS). Lumbar puncture was performed, and the cerebrospinal fluid was analyzed by the hematologist who identified 12 cells (atypical lymphocytes and blasts), suggestive of hematological disease recurrence, in the CNS this time. Intratechal chemotherapy was started. **Discussion:** Stroke mimics (SM) are defined as acute onset of focal neurological symptoms, which later is diagnosed with a non-vascular origin. Based on review articles, up to one-third of stroke admissions can be labelled as mimics. They are likely a misdiagnosis for: peripheral vestibular dysfunction, toxic/metabolic disturbance, seizures, functional disorder and migraine. SM is associated with lower NIHSS and affect prevailingly younger female individuals. In a large database review, 15,595 patients were analyzed, and space-occupying lesion represented <5%. In parallel, only up to 5% of patients with intracranial tumors seem to develop sudden onset of neurological features. Gliomas, meningiomas, and metastases are the main tumors masquerading as acute stroke. The mechanisms underlying acute "tumor attacks" include acute intracranial pressure changes leading to reduction in cerebral blood flow, vascular steal phenomena and vascular compression with resultant infarction. Thrombolysis may be prescribed in more than 1/4 of the cases. Although this procedure has very few complications in most of the SM, in case of brain tumor, it an absolute contraindication. Moreover, ignoring the actual etiology delays the proper treatment of a severe condition. **Final comments:** The case is an example of stroke mimics caused by an infiltrative tumor lesion. In the present case, the low NIHSS and the striking clinical radiological dissociation was another red flag to interrogate another etiology for the acute deficit. The reported case highlights the importance of emergency doctors being aware of stroke mimics and their possible critical etiology.

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Livedo reticularis as a dysautonomic manifestation in bulbar ischemia

Aline Paredes Cardoso Fukuda[1], Bernardo Leite Ponde da Luz[1], Jacy Bezerra Parmera[1], Luiz Roberto Comerlattu[1], Fernando Freua[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Aline Paredes Cardoso Fukuda; Email: alineparedesc@gmail.com.

Case presentation: A 46-year-old female patient presented with acute and non-limiting onset of headache, vertigo, and tetraparesthesia. On the following day, she awakened with altered speech and right side weakness. She was admitted to the emergency unit and soon after progressed into hypoxia leading to cardiac arrest that was assisted until return to sinus rhythm. Upon evaluation, the patient presented with high blood pressure (206x114mmHg). She was alert, responsive to orders and able to reply non-verbally to simple questions. She had right-side hemiplegia and hypoesthesia, bilateral upper limb ataxia with prominent dysmetria, left-side Horner syndrome, nystagmus with a rapid phase to the left and worsening in right side gaze, and, skew deviation with hypotropic left eye. Facial sensitivity unimpaired and evaluation of bulbar nerves was hindered due to intubation. Notably, the patient presented with left-side livedo reticularis, strictly respecting the midline and sparring the right side. Initially, extubation was possible, however, hours later, she progressed with rapid respiratory failure and returned to ventilatory support. She persisted with

fluctuating vital controls and dysautonomia throughout the following days culminating in refractory shock and death. In investigation, cervical angiography revealed a left vertebral artery occlusion, and, MRI was compatible with ischemia in the left medulla, concordant with a Dejerine- Roussey syndrome. These findings were later confirmed in necropsy results. **Discussion:** Bulbar infarcts are associated with autonomic dysfunction due to lesions in the vagus nucleus and solitary tract. These structures are pivotal in the relay between hypothalamic outputs affecting cardiovascular, respiratory and gastrointestinal reflexes. The ventrolateral and ventromedial medulla contain sympathoexcitatory neurons that intermediate blood pressure reflex control in response to internal stressors. Acute vascular lesions in such topography can culminate in increased sympathetic activity promoting neurogenic hypertension and loss of baroreceptor regulation. Livedo reticularis is a skin manifestation of impaired perfusion, and in this case, its distribution was congruent with the topography of the ischemic lesion. Dysautonomic manifestations in midbrain infarcts are typically perceived as altered vital signs, however, this case shows how such manifestations can also affect other systems. **Final comments:** Autonomic dysfunction is an important manifestation of midbrain ischemic lesions and may lead to catastrophic outcomes. Patients with such pathology benefit from intensive monitoring and immediate interventions to stabilize vital controls. It is important to recognize that autonomic dysfunction may have an atypical clinical presentation in other organ systems, including the skin.

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Lively contributions: 150 years after Johannes (Jan) Evangelist Purkinje passing

Isabelle Pastor Bandeira[1], Catarina Dantas Corrêa[1], Camila Emi Fujiwara Murakami[1], Helio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Complexo do Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Isabelle Pastor Bandeira; Email: isabellepbandeira@gmail.com.

Background: Johannes Evangelist Purkinje was a remarkable Czech scientist who made discoveries concerning the structure and function of the eye, brain and heart which gave him countless eponyms. Beside biological science, Purkinje translated poetry from German, Russian and Polish to Czech and was an active Czech patriot. **Objective:** Brief review of Purkinje life and accomplishments in honor of his 150 th death anniversary. **Methods:** Based on literature research performed at PubMed. **Results:** On December 17, 1787, Purkinje was born in Libochovice Castle, then Bohemia – Empire Austro-Hungarian (now Czech Republic). Since a young child, Purkinje highlighted himself by his wit and intelligence. Attended the Piarist order at the Stará Voda monastery (Moravia), not to became a priest but to acquire advancer education. He then moved to Prague where attended physical science courses at the Philosophical Institute of Universitas Carolo-Ferdinandea and after enrolled as a student at Medical Faculty of the University of Prague which culminated with his dissertation, Contributions to the Knowledge of Vision from the Subjective Point of View in 1818 Acted as an anatomy instructor at Prague Medical Faculty between 1819 and 1822 and, in 1823, was accepted as Professor of Physiology and Pathology at the Medical Faculty of the Royal Prussian University of Breslau. In November 1839, Purkinje founded the Wroclaw's Institute of Physiology, which was not solely dedicated to research but also to Czech and Polish culture. In 1850, back in Prague, accepted the nomination of Professor of Physiology at the Medical Faculty of Prague, where he graduated. Honored with the Imperial Austrian Order of Leopold in 1868, conferred for merit and moral integrity, Purkinje died a year later, on July 28, 1869, in Prague. Despite his primary focus not being on cardiology, one of Purkinje's discoveries was the identification of a specialized conduction pathway in the heart known as Purkinje fibers. These fibers are now recognized as cardiomyocytes specialized in conducting electrical signals from the His bundle to the contractile ventricular myocytes. Among the many eponyms attributed to the Czech physiologist, perhaps one of the most celebrated are the Purkinje cells: the largest neurons of the cerebellum. Purkinje and his group examined slices of sheep cerebellum fixed with alcohol in a series of experiments that would accurately describe each of the three layers of the cerebellar cortex. Through these experiments, attributed to the cerebellum the role of precision of motor movements. Beyond histological descriptions, Purkinje also dedicated himself to investigating the mechanisms of balance and vertigo, giving rise to Purkinje's law of vertigo. **Conclusion:** Throughout his brief 52 years of life, Purkinje left a legacy which goes beyond the medical field. The year of 2024 marks 155 years of his death, however his contributions remain relevant and alive.

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Logopenic variant of primary progressive aphasia: case report

Francisco de Assis Pinto Cabral Júnior Rabello[1], Giovana Sena Veloso Stefani Moreira[2], Adriana Ruphael de Freitas[2]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[2]Faculdade de Medicina de Barbacena, Barbacena MG, Brazil.

Correspondence: Francisco de Assis Pinto Cabral Júnior Rabello; Email: francisco.cabral.neuro@gmail.com.

Case presentation: A.C.F.S, male, 58 years old, already diagnosed with unspecified dementia and using memantine 10mg/day for the last 3 years. His wife reports that 7 years ago he began to have progressive difficulties for communication, with hesitancy in the middle of sentences, which caused social isolation. He started to writ wrong, which was an issue since he works as a hospital receptionist and filled out forms. At time, he also had difficulty recalling recent events. Falls, signs of parkinsonism, behavioral problems, hallucinations and delusions weren´t present in the symptoms. In the first evaluation patient was completely dependent for daily activities, global cognition was moderately affected (CDR 2.0), despite the somatic neurological exam was normal. Neuropsychological evaluation exhibited marked difficulties in repeating verbal sentences, comprehension of long phrases and errors in writing and reading irregular words. Cerebrospinal analysis was normal, except for mild protein elevation (65mg/dl). Search for presenilin mutation (PSEN 1 and 2) was negative and the brain PET FDG-18F showed glycolytic hypometabolism in the posterior aspect of the cingulate gyrus as well as in the cortical topography of the bilateral temporal region, extending to the parahippocampal gyrus, inferior, middle and superior temporal gyri. Glycolytic hypometabolism was also present in the frontoparietal region, extending to the precuneus, more prominent on the left. **Discussion:** Logopenic variant of primary progressive aphasia (LV-PPA) is generally associated with atrophy of the posterosuperior temporal (perisylvian) and inferior parietal regions, which are areas involved in the auditory processing of speech and in the activation of phonological representations for language expression (phonological loop). The patient in this case presented hypometabolism in this region in the dominant hemisphere, which reinforces the diagnosis. Difficulty in lexical access during spontaneous speech results in hesitations and pauses in patients with LV-PPA. A more comprehensive assessment of language with specific tasks (comprehension of words and phrases; naming by visual confrontation; repetition of words and phrases with variable length; reading of regular and irregular words) may be necessary, as performance differs between the variants. Additionally, research into structural and molecular neuroimaging biomarkers may help, especially when demonstrating focal patterns of atrophy or hypometabolism. **Final comments:** The most common underlying neurodegenerative pathology in LV-PPA is Alzheimer's disease, with the language alteration attributed primarily to a deficit in the phonological component of working memory, which explains the difficulty in repeating sentences, the preservation of understanding of isolated words (which helps to differentiate from the semantic variant of PPA) and the relative preservation of fluency when compared to the non-fluent variant.

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Longitudinal analysis of 173 long-COVID subjects reveals worsening of work capacity and neuropsychiatric symptoms 20 months after diagnosis

Gabriel Monteiro Salvador[1], Ana Paula Beppler Lazaro Lino[1], Fernando Cendes[1], Guilherme Vieira Nunes Ludwig[1], Hildete Prisco Pinheiro[1], Marcia Bandini[1], Clarissa Lin Yasuda[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Gabriel Monteiro Salvador; Email: gabrielsalvador1906@gmail.com.

Background: Although neuropsychiatric symptoms are commonly reported in LONG-COVID syndrome, fewer studies evaluated their impact on work capacity. Here, we analyzed the persistence of symptoms of anxiety, depression, fatigue and sleepiness and the reduction in work capacity in a homogeneous group of workers with validated questionnaires. **Objective:** To investigate the longitudinal course of persistent neuropsychiatric symptoms and occupational vulnerability of post-survivors of COVID-19. **Methods:** We analyzed 173 individuals with confirmed COVID-19 (hospitalized (17.9%) and non-hospitalized (82.1%)) at two different times after acute infection (interval of 227 vs 614 days). The information was extracted from the database of a longitudinal study. Patients answered the "Chalder Fatigue Scale (CFQ)", "Epworth Sleepiness Scale (ESS)", "Hospital Anxiety and Depression Scale (HADS)" Patient Health Questionnaire-9 (PHQ9), and "Work Ability Index (WAI)". We used SPSS22 for statistical analysis with Chi-Square tests for analyses of proportions. **Results:** Our patients presented average age of 45 years (25 to 66 years), sex (59% female), education (43.9% complete undergraduate, 43.4% post-graduation, and 10.4% complete high school), function at work (68.2% contact with the public and 31.8% administrative work), reinfection (13.3%), vaccination (86.1%). With an average interval of 614 days between diagnosis and the second online interview, subjects SELF-reported several symptoms, including related fatigue (38.7% VS 45.1%; P=0.128), anxiety (41% VS 38.7%; P=0.594), depression (20.8% VS 18.5%; P=0.467), cognitive dysfunction (37% VS 42.2%; P=0.001); some reported to be asymptomatic, without post-COVID symptoms (3.5% VS 17.3%; P=0.614). However, the quantification of symptoms with validated questionnaires revealed unexpected changes. The comparison of symptoms between the two applications revealed an increase of excessive daytime somnolence (ESS) (58.4% to 75.1%; P=0.001), fatigue (CFQ) (55.5% to 79.9%; P<0.001), symptoms of anxiety (HADS) (6.4% to 35.5%; P<0.00) and symptoms of depression (PHQ9) (42.2% to 77.5%; P<0.00). The results reveal a reduction of WAI (23.4% to 63%; P<0.00). **Conclusion:** The results of this longitudinal study suggest that some subjects persist with neuropsychiatric symptoms almost two years after the acute infection, even those with mild disease. Surprisingly, this group of individuals presented worsening symptoms and working ability, which should raise the concern of potential long-term dysfunction in

post-COVID-19 individuals. As the responders volunteered for the study, we speculate that responders for the second interview were motivated by their persistent symptoms. However, despite this limitation, these results are valuable as we do not understand entirely the natural course of SARS-COV-2 infection. The study reveals a reduction in work capacity several months post-infection, suggesting economic impacts

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Longitudinal analysis reveals the persistence of reduced work capacity and neuropsychiatric symptoms 25 months after infection in post-survivors

Gabriel Monteiro Salvador[1], Ana Paula Beppler Lazaro Lino[1], Fernando Cendes[1], Márcia Bandini[1], Clarissa Lin Yasuda[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Gabriel Monteiro Salvador; Email: gabrielsalvador1906@gmail.com.

Background: While numerous studies confirm that neuropsychiatric symptoms are prevalent in LONG-COVID syndrome, fewer have analyzed them longitudinally or investigated changes in work capacity after COVID-19. Here, we interviewed a group of survivors longitudinally to quantify neuropsychiatric symptoms and the reduction in work capacity in post-covid individuals with validated questionnaires. **Objective:** Our primary aim was to shed light on the course of persistent neuropsychiatric symptoms and the longitudinal variation in work capacity after COVID-19 disease. This research is crucial in understanding the long-term effects of the virus and its impact on individuals' ability to work. **Methods:** We analyzed longitudinal data from one study's database of 602 individuals with a confirmed diagnosis of SARS-COV-2 infection ((hospitalized (16.8%) and non-hospitalized (83.2%)) with an average of 216 days after acute infection (T1), followed by 173 individuals (hospitalized (17.9%) and non-hospitalized (82.1%)) with 614 days after the acute infection (T2). Finally, 46 subjects (hospitalized (21.7 %) and non-hospitalized (78.3%)) underwent the third application 774 days after the acute infection (T3). Participants completed the "Chalder Fatigue Scale (CFQ)", "Epworth Sleepiness Scale (ESS)", "Hospital Anxiety and Depression Scale (HADS)", Patient Health Questionnaire-9 (PHQ9), and "Work Ability Index (WAI)". Statistical analysis was conducted using SPSS22, employing Chi-Square tests to analyze proportions. **Results:** Our patients presented average age of 44 years (25 to 66 years), sex (53.5% female T1 vs 59% in T2 vs 58.7% in T3), education (43.9% complete undergraduate, 43.4% graduate, and 10.4% complete high school), reinfection (9.3% in T1 vs 13.3% in T2 vs 13% in T3), vaccination (83.4 % in T1 vs 86.1% in T2 vs 87% in T3). The comparison of symptoms between the three applications revealed persistent levels of excessive sleepiness (ESS) (70.4 % VS 75.1% VS 58.7%), fatigue (CFQ) (79.8% VS 79.9% VS 80.4%), symptoms of anxiety (HADS) (39.4% VS 35.5% VS 41.3%) and symptoms of depression (PHQ9) (30.9% VS 42.2% VS 28.3%). In addition, the results reveal the persistence of reduced WAI (64.8% VS 63% VS 63%). **Conclusion:** As expected in longitudinal studies, the number of responders reduced over time. Interestingly, the smaller groups of participants in T2 and T3 revealed long-term persistence of neuropsychiatric symptoms, which may impair daily life functioning. The elevated proportion of individuals with reduced work capacity raises the concern of a significant economic impact at both individual and collective levels. Given the uncertainties related to the natural course of SARS-COV-2 disease and LONG-COVID syndrome, these results raise the concern of potential long-term sequela associated with the new virus. More extensive studies are necessary to understand this course and provide data for prevention and specific treatments.

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Longitudinal clinical and spinal cord imaging in adult patients with SMA type 3

Felipe Franco da Graça[1], Cristina Iwabe[1], Rodrigo de Holanda Mendonça[2], Edmar Zanoteli[2], Marcondes Cavalcante França-Jr[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Felipe Franco da Graça; Email: felipefgraca@hotmail.com.

Spinal muscular atrophy (SMA) is an autosomal recessive disorder of lower motor neurons caused by biallelic loss-of-function SMN1 variants. SMA type 3 is a milder phenotype of the disease (patients are able to walk) and divided into types 3a (disease onset before 3 years old) and 3b (disease onset after 3 years old). The natural history of the more severe phenotypes (ie, SMA types 1 and 2) is well known, but much remains to be investigated regarding the clinical course of late-onset patients. Such knowledge gap represents a major limitation in the clinical management of these patients. This is particularly troublesome in the novel therapeutic landscape of SMA, where effective disease-modifying treatments are available. In this study, 17 healthy controls and 17 adult patients with SMA type 3 confirmed by genetic testing and clinical evaluation underwent spinal cord (SC) MRI. All subjects were scanned on a 3T Phillips Achieva Scanner using a 16-channel neurovascular coil for all SC images in two visits separated by one year. These were acquired in the axial-oblique plane with the center of the imaging volume positioned at the level of C2-3 intervertebral disc. To measure the cross-sectional (CSA) and the gray matter (GM) areas, we automatically segmented the cervical SC using a deep-learning algorithm. All MRI data were processed using the open-source software Spinal Cord Toolbox v4.0.1. The MFM scale was used to quantify disease severity. In the patient group, there were 12 men and 5 women with an average age of 32.1 years. A comparative analysis was conducted across-groups on the cross-sectional area of the spinal cord at the C3 level in patients during visits 1 and 2. No significant differences were observed neither in the comparison between patient groups across visits (p=0.89) nor in the comparison between patients and controls (p=0.20). Similarly, an evaluation of the GM area of the SC was performed in patients during visits 1 and 2, as well as in the controls. Once again, there was no significant change between visits both in motor scales and area, but there was a significant difference in the comparison between patients and controls (p<0.0001) In the combined evaluation with other variables, a strong correlation was found between the GM area and the values of the MFM scale (r=0.79, p=0.0002) Quantitative SC MRI emerges as promising neuroimaging biomarker in late-onset SMA. SC GM atrophy is the most promising candidate as it was found not only to distinguish patients and controls, but also to correlate with motor function in this disease.

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Longitudinal evaluation of fatigue in adult patients with spinal muscular atrophy and the impact of treatment with disease-modifying drugs

Felipe Franco da Graça[1], Cristina Iwabe[1], Marcondes Cavalcante França Jr[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Felipe Franco da Graça; Email: felipefgraca@hotmail.com.

Fatigue is a prevalent symptom in Spinal Muscular Atrophy (SMA) and significantly affects the quality of life of individuals, particularly in performing repetitive daily activities. In adult patients, fatigue is a frequent complaint and a key factor that diminishes quality of life. This study aimed to evaluate fatigue in adult patients with SMA and correlate it with the use of disease-modifying drugs. We conducted a longitudinal study involving 25 adult patients (17 men and 8 women) diagnosed with 5q Spinal Muscular Atrophy confirmed by genetic testing. Each patient underwent an initial assessment using the Fatigue Severity Scale (FSS) to quantify fatigue levels, considering a score above 4 as indicative of fatigue. Additionally, the Hammersmith Functional Motor Scale Expanded (HFMSE) was used to assess motor function. Patients were reevaluated using the same scales after one year, with their treatment status documented at both visits. At the first visit, the mean FSS was 4.6, and at the second visit it was 4.9. As for HFMSE, the means were 20.8 and 20.3 at the first and second visits, respectively. We did not observe significant differences between the groups when all patients were evaluated together. Regarding treatment status, at the first visit, only 4 patients were using disease-modifying medications (all with nusinersen), while at the second visit, 9 patients were being treated (8 with nusinersen and 1 with risdiplam). Of the 5 patients who started medication between visits, 1 did not have significant fatigue previously. After treatment, 4 out of these 5 patients did not experience significant fatigue. The higher number of patients using disease-modifying medications at the second visit allowed comparisons between treated and untreated groups, showing that the proportion of patients with fatigue was significantly higher in the untreated group (81.2%) compared to the group that received specific therapy (33.3%) (p=0.03). No significant differences were observed in HFMSE among the same individuals. Our study population has a lower frequency of fatigue among treated patients compared to the untreated, although we did not observe significant differences in motor scales between the groups. Despite limitations due to the small number of patients, these findings support the clinical impression that treated adult patients experience a reduction in fatigue compared to untreated patients, providing an improvement in quality of life, regardless of changes in functional scales. Given that motor progression may be slow in adult patients, evaluating other factors such as fatigue may be relevant for clinical follow-up and assessment of therapeutic response.

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Longitudinal T2-weighted MRI relaxometry in patients with temporal lobe epilepsy

Fernando Bruno de Aquino Cassol[1], Marina Koutsodontis Machado Alvim[1], Brunno Machado de Campos[1], Clarissa Lin Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Fernando Bruno de Aquino Cassol; Email: f242155@dac.unicamp.br.

Background: Temporal lobe epilepsy with hippocampal sclerosis (TLE-HS) is associated to higher rates of resistance to anti seizure medication (ASM). It is already known that these patients exhibit widespread white and gray matter structural abnormalities, such as volume reduction, extending beyond the temporal lobe and hippocampus. MRI T2-relaxometry can add information in the analysis of structural abnormalities in these patients, mainly in the hippocampus. **Objective:** Evaluate whether T2 relaxometry changes progress over time in patients with unilateral (L-HS and R-HS) and bilateral (Bil-HS) TLE-HS. **Methods:** Were included 131 TLE-HS patients from a tertiary center epilepsy outpatient clinics, divided into 3 groups, according to clinical data and HS visual analysis: 71 L-HS (42 women), 49 R-HS (31 women) and 11 Bil-HS (9 women). All patients had 2 MRIs with at least 1 year interval, including a high resolution 3D T1-weighted image

and a T2 multi-echo image (voxel= 0.45x0.45-0.45x0.9 mm³, gap= 3 mm, TE= 30, 60, 90, 120,150,180 ms, TR= 3300 ms and 36 coronal slices aligned to the hippocampi). The T2 maps were quantified using single exponential fit. The quantified maps were coregistered to the T1 WI, normalized to the MNI space and labeled using Desikan atlas. We chose the thalami, amygdalae and hippocampi and tested for longitudinal differences using paired t-test. **Results:** The groups had similar age at first MRI, the mean age of the patients was 46.21 (17.30 - 72.75). The mean interval between MRIs was 6.4 years (1.17 - 12.18) in L-HS group, 6.17 years (1.09 - 11.84) in R-HS and 7.88 years (6.34 -11.67) in Bil-HS group. In R-HS patients, we identified significant increase in their contralateral and ipsilateral thalamus (p=0.0001; p<0.0001) and hippocampus (p=0.0004; p<0.0001), with no significant changes in the ipsilateral and contralateral amygdala. In L-HS group, there was a significant increase signal in the contra and ipsilateral thalamus (p<0.0001; p<0.0001), hippocampus (p<0.0001; p<0.0001), ipsi (p=0.0002) and contralateral (p=0.0158) amygdala. Finally, in patients with bilateral hippocampal sclerosis, there were increases in the left (p=0.0180) and right hippocampus (p=0.0445), with no significant differences in their thalamus and amygdala, on both sides. **Conclusion:** There are changes over time in T2-signal in patients with TLE-HS in both hippocampi, regardless of whether it is unilateral or bilateral HS. In R-HS and L-HS patients, there were also changes in both thalami and in the L-HS group in both amygdalae. Whether these changes reflect gliosis or axonal or neuronal damage, remains to be confirmed.

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Longitudinally extensive myelitis secondary to neuroschistosomiasis: case report

Stephania Neves Scapim[1], José Víctor Filho de Oliveira[1], Iasmim Portela Maifrede[1], Leopoldo Antônio Pires[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Juiz de Fora MG, Brazil.

Correspondence: Stephania Neves Scapim; Email: drastephanianeves@gmail.com.

Case presentation: A 65-year-old man with benign prostatic hyperplasia and essential tremor presents with a history of fatigue and unintentional weight loss in the past year. Subsequently, he developed hypoesthesia and progressive lower limb weakness, associated with urinary retention and fecal incontinence. After 6 months, he progressed to tetraparesis. He was diagnosed with transverse myelitis, confirmed by spinal cord magnetic resonance imaging (MRI) and cerebrospinal fluid (CSF). He underwent pulse therapy with methylprednisolone for 5 days with improvement in strength in his upper limbs. He was then referred to our service for further investigation. Physical examination revealed spastic crural paraparesis and tetrahyperreflexia with bilateral Babinski sign, signs of pyramidal release in upper limbs, bilateral hyperesthesia in T7-T12 dermatomes and hypoesthesia below T12 for all modalities, except for pain. Brain MRI revealed chronic intraparenchymal hematoma in the subcortical white matter of the right superior temporal gyrus. Cervical and thoracic MRI showed diffuse signs of spondyloarthrosis, with mild spinal canal stenosis highlighted at the levels C4-C5, C5-C6. In addition, it was noted an hyperintensity on T2/STIR compromising the central portion of the dorsal spinal cord from the level of D1 to the conus medullaris. Foci with faint enhancement by intravenous contrast were noted sporadically in the right lateral funiculus and central portion of the spinal cord at the level of D4, D6 to D7. CSF showed glucose 50 mg/dL, 27 cells (97% mononuclear), protein 133 mg/dL, with presence of oligoclonal bands, type 2 pattern. Anti-AQP4, ANCA-c and ANCA-p were negative. Screening tests for HIV, HTLV, syphilis, toxoplasmosis and hepatitis were also negative. Immunological test for schistosomiasis was positive in serum (1:128) and CSF (1:2), suggesting the hypothesis of longitudinally extensive transverse myelitis (LETM) due to neuroschistosomiasis (NE). Treatment with praziquantel and pulse therapy with methylprednisolone was performed, followed by prednisone. The patient showed complete improvement of symptoms in the upper limbs and partial improvement in the lower limbs. He continued to experience urinary retention and constipation. **Discussion:** We present a case of NE as the cause of spastic paraparesis with hypoesthesia below T12, and sphincter dysautonomic syndrome. NE is uncommon and can cause serious neurologic complications, involving the spinal cord and/or the brain. Patients with myelopathy may present with lower limb pain, lower motor dysfunction, bladder paralysis, and bowel dysfunction. **Final comments:** Differential etiologic diagnosis for LETM should include schistosomiasis, especially in endemic areas.

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Longitudinally extensive transverse myelitis after hand-foot-and-mouth disease in an adult, immunocompetent patient: a case report

Víctor de Moraes Chagas[1], Bruno Henrique Carneiro Costa Filho[1], Luísa Couceiro de Albuquerque Macêdo[1], João Pedro Matos de Santana[1], Lucas de Andrade Alexandre[1], Mariana de Almeida Cruz Silva[2], Luziany Carvalho Araújo[3], Clélia Maria Ribeiro Franco[1], Álvaro José Porto Moreira[1], Eduardo Sousa de Melo[2]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Serviço de Neurologia, Recife PE, Brazil. [2]Universidade Federal de Pernambuco, Centro de Ciências Médicas, Recife PE, Brazil. [3]Universidade Federal de Pernambuco, Hospital das Clínicas, Serviço de Radiologia, Recife PE, Brazil.

Correspondence: Mariana de Almeida Cruz Silva; Email: mariana.almeida20021@gmail.com.

Case presentation: A 46-year-old male patient was admitted to the Neurology infirmary with a history of acute urinary and fecal retention, symmetrical weakness in the legs and torso, and hypoesthesia below his chest, initiated 30 days prior. On admission, he had grade II symmetrical spastic paraparesis of the legs, brisk patellar and Achilles reflexes, bilateral Babinski sign; brisk arm reflexes, with no weakness; and urinary retention requiring indwelling urinary catheter. By the time of admission, axial weakness, hypoesthesia, and fecal retention had resolved. No prior similar symptoms, chronic illnesses, or substance use were reported. However, two weeks before these symptoms, he had malaise, muscle pains, low fever, hyporexia, and erythematous, pruritic rash on both hands. His 8-year-old son had presented similar lesions on his hands, feet, elbows, and around the lips, days before. Spine MRI showed bilateral T2-hyperintense lesions on grey matter from C3-T12. CSF analysis showed a high protein level (70 mg/dL) with normal cell count. PCR for myelitis-associated viruses, collected one month after onset, was negative. He was treated with methylprednisolone 1 g/day for 5 days, with improvement of symptoms. Six months after treatment, he had no sequelae, and a new spinal MRI was normal. **Discussion:** Hand-foot-and-mouth disease is usually benign, caused by enteroviruses, most commonly by Coxsackie A16, a well-established cause of aseptic meningitis, encephalitis, Guillain-Barré syndrome, and acute flaccid paraparesis; transverse myelitis is reported, but rare. Viral myelitis commonly causes only mild alterations on CSF analysis and spinal cord imaging, and clinical history is paramount to diagnosis. History must include recent travel to endemic areas, immune suppression, and associated symptoms. Our patient had not traveled recently, nor had a history of immune alterations. Skin rash, accompanying or preceding neurological symptoms, favors the possibility of enterovirus or herpesvirus infection. Negative viral DNA investigation on CSF can be explained either by the delayed time from symptom onset to sample collection; or by an immune-mediated lesion with no direct viral invasion to the nervous system. The absence of previous similar symptoms and normal follow-up MRI suggest monophasic acute myelitis, compatible with our hypothesis. Our patient had an excellent response to corticosteroid therapy, which was not seen in previously reported cases. **Final comments:** We present the case of an adult, immunocompetent patient who developed monophasic longitudinally extensive transverse myelitis after symptoms of hand-foot-and-mouth disease. Despite negative viral analysis of CSF, enterovirus is a highly likely etiology, considering associated symptoms and clinical history. Because of the very few cases reported, it is impossible to determine the optimal time and treatment course for similar cases.

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Longitudinally extensive transverse myelitis as a manifestation of systemic lupus erythematosus

Paulo Victor Machado[1], Ana Paula G. Barankiewicz[1], Lucas R. Prim[1], Paola R. González[1], Lisseth F. V. Reyes[1], Maria Julia B. Rodrigues[1], Isabela Wandarti Ribas[1], Lucas Voigt Machado[1], Bruna Conti[1], Marcos Seefeld[1]

[1]Hospital Universitário Evangélico Mackenzie, Curitiba PR, Brazil.

Correspondence: Paulo Victor Santos Machado; Email: paulovictorsmachado@gmail.com.

Case presentation: Female patient, 29 years old, previously diagnosed with systemic lupus erythematosus (SLE), using hydroxychloroquine and mycophenolate mofetil, with a previous episode of transverse myelitis in 2022, at the time treated with methylprednisolone pulse therapy and 6 pulses of cyclophosphamide. Admitted to the service in 2023, with tetraparesis and urinary incontinence, in addition to sensory level at T1. Subjected to neuraxial MRI, longitudinally extensive myelitis was demonstrated, with the lesion extending from the medulla oblongata to the level of C6. Examination of the cerebrospinal fluid showed a slight increase in proteins and negative oligoclonal bands. Negative anti-aquaporin 4 antibody in serum. Pulse therapy with methylprednisolone 1g/day was started. The patient developed hypercapnic respiratory failure, requiring orotracheal intubation and transfer to the intensive care unit (ICU) on the 3rd day of hospitalization. Finished 5 days of treatment with corticosteroid pulse therapy, without improvement in muscle strength. The patient underwent plasmapheresis and developed septic shock with a pulmonary focus. Culture-guided antibiotics were started, plasmapheresis was suspended due to the infectious context, and pulse therapy with cyclophosphamide was chosen, with partial improvement in muscle strength. The patient underwent tracheostomy and developed a new septic shock with a pulmonary focus, refractory to clinical measures. **Discussion:** Systemic lupus erythematosus (SLE) can affect the central and peripheral nervous system. Occasionally, patients present spinal cord involvement through inflammatory activity. Transverse myelitis is a relatively rare manifestation of the disease, with an incidence of around 1-2% of patients with SLE. Magnetic resonance imaging (MRI) is the imaging modality of choice for investigating myelopathies. Lesions that involve three or more contiguous vertebral bodies are classified as longitudinally extensive. In a review of 63 cases of myelitis associated with SLE, 71.4% of patients manifested longitudinally extensive myelitis. The measurement of oligoclonal bands in CSF can help distinguish transverse myelitis related to multiple sclerosis. Likewise, it is necessary to measure anti-aquaporin 4 antibodies in the patient's serum, to exclude myelitis associated with neuromyelitis optica. The treatment of lupus myelitis derives directly from the treatment of SLE and idiopathic myelitis. Methylprednisolone and cyclophosphamide are commonly used as first line when there is serious and threatening target organ involvement. Plasmapheresis is an option in refractory cases. Other

options are represented by mycophenolate, rituximab, azathioprine and human immunoglobulin. **Final comments:** Cases of myelitis associated with lupus can occur regardless of the degree of activity of the underlying disease. Rapid recognition of the condition and early initiation of treatment are associated with better outcomes.

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Long-term cardiac magnetic resonance imaging study in Friedreich's ataxia

Thiago Junqueira Ribeiro de Rezende[1], Alberto Rm Martinez[1], Mariana R de Brito[1], Renan Ff Nunes[1], Luis M Silva[1], Karen Ag Takazaki[1], Melina P Martins[1], Fernando Cendes[1], Michael Jerosch-Herold[2], Otávio R. Coelho-Filho[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Harvard University, Boston MA, USA.

Correspondence: Thiago Junqueira Ribeiro de Rezende; Email: thiago.jrezende@gmail.com.

Background: Friedreich's ataxia (FRDA) demands the identification of biomarkers crucial for clinical management and clinical trial planning. Despite heart failure being the primary cause of mortality in FRDA, there remains a lack of dependable cardiac biomarkers. While cardiac magnetic resonance imaging (cMRI) has emerged as a promising option, longitudinal cMRI data for the disease are limited. **Methods:** Initial cMRI scans were conducted on 18 healthy individuals and 42 FRDA patients, with 17 of the latter group receiving repeat cMRI scans after a 5-year interval. The study involved the measurement of left ventricle ejection fraction (LVEF), mass, and volumes, as well as the evaluation of late gadolinium enhancement (LGE), extracellular volume fraction (ECV), and intracellular water-lifetime (τic). Neurological deterioration was quantified using the FRDA rating scale (FARS). **Results:** Analysis of the data revealed that FRDA patients displayed preserved LVEF but increased left ventricle volumes, thickness, ECV, and τic in comparison to the control group. Nine out of 41 patients exhibited epicardial LGE. Furthermore, the left ventricle volumes showed correlation with FARS scores. Longitudinal assessment demonstrated no significant decline in LVEF; however, an increase in left ventricle diastolic volume and a decrease in the left ventricle volume/mass ratio were observed. Five additional patients displayed epicardial LGE after the 5-year follow-up. **Conclusion:** The cross-sectional analysis highlighted that FRDA patients maintain preserved LVEF but exhibit increased left ventricle volumes and thickness due to myocardial interstitium expansion and cardiomyocyte size enlargement. Noteworthy findings from the longitudinal follow-up suggest evolving heart damage and a transition from a hypertrophic cardiomyopathy phenotype to a dilated cardiomyopathy phenotype. In conclusion, cMRI has unveiled valuable insights into cardiac involvement in FRDA and could serve as a valuable biomarker for monitoring disease progression.

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Long-term effect of subthalamic nucleus deep brain stimulation on freezing of gait in Parkinson's disease: a single-arm meta-analysis of prospective studies

Marianna Leite[1], Jordana Borges C. Diniz[2], Lizen Clare André Moreira[3], Helena do Amaral Barreto[3], Ana Maria Porto Laxe[3], Maria Eduarda Silva[4], Lucca Lopes[5], Carolina B. Moura[3]

[1]Faculdade Santa Marcelina, São Paulo SP, Brazil.
[2]Instituto Neurológico de Goiânia, Goiânia GO, Brazil.
[3]Universidade Federal Fluminense, Niterói RJ, Brazil.
[4]Escola Estadual Antônio Silva, Timóteo MG, Brazil.
[5]Johns Hopkins Bloomberg School of Public Health, Baltimore, United States of América.

Correspondence: Marianna Gerardo Hidalgo Santos Jorge Leite; Email: msmariannaleite@gmail.com.

Background: Numerous observational trials have shown the use of deep brain stimulation on freezing of gait in patients with Parkinson's disease (PD). However, it remains unclear what is the long-term effect of this intervention. **Objective:** Therefore, we conducted a meta-analysis to assess the long-term effect of subthalamic nucleus deep brain stimulation (DBS) on freezing of gait in PD. **Methods:** We systematically searched PubMed, Embase and Web of Science for studies using subthalamic nucleus deep brain stimulation on freezing of gait in PD. Primary outcomes were Gait item Axial Symptoms items in the Unified Parkinson's Disease Rating Score (UPDRS), measured by mean and SD scores. For this single-arm meta-analysis a random effects model was used to calculate the pooled proportions with 95% confidence intervals. Statistical analysis was performed using the R program (version 4.3.2). Heterogeneity was assessed with I2 statistics. **Results:** Fifteen studies were included in this meta-analysis, comprising 407 patients with PD treated with subthalamic nucleus deep brain stimulation. Mean score on Gait item 29 on medication off DBS was 7.39 points (95%CI = -4.27-23.68; I2 = 97%), while mean score on Gait item 29 on medication with DBS was 7.05 (95%CI = -0.78-14.87; I2 = 96%). Combined Axial symptoms score, defined as the sum of the motor scores: item 18, 22 (rigidity of the neck) and items 27-30, revealed a mean score of 9.71 points (95%CI = -4.27-23.68; I2 = 97%) on medication off DBS, and 9.01 score on medication on DBS (95%CI = -1.02-19.04; I2 = 99%). **Conclusion:** In PD patients with freezing of gait symptoms, long-term subthalamic nucleus deep brain stimulation was associated with better scores of Gait and Axial Symptoms items in the Unified Parkinson's Disease Rating Score (UPDRS).

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Long-term memory trajectories in seizure-free patients operated for hippocampal sclerosis

Roberta Gomes[1], Eduardo Leal-Conceição[1], Caroline Schiochet Verza[1], Wyllians Vendramini Borelli[1], Mirna Portuguez[1], William Alves Martins[1], Eliseu Paglioli[1], Andre Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Caroline Schiochet Verza; Email: carolinesverza@gmail.com.

Background: Antero-mesial TLE surgery is usually safe, although the possibility of significant decline in memory function should be considered. Several predictors have been advanced, the most relevant being the pre-resection structural and functional status of these structures as verified by MRI and neuropsychological evaluation. In general, the more preserved the structure and /or the function, the higher the risks for post-operative memory deficits. However, post-operative memory is usually assessed only in the first few years and function in thevery long-term is seldom addressed. Furthermore, there is a tendency to group patients with TLE due to distinct pathologies and achieving variable post-operative seizure outcome **Objective:** To study memory trajectories over the years in patients with temporal lobe epilepsy and unilateral hippocampal sclerosis (TLE/HS) seizure free since surgery. **Methods:** Memory tests were applied before (T1), 1 to 4 years (T2) and 10 to 15 years after operation (T3) in 54 patients with TLE and pathology-verified HS (34 left), seizure free since surgery. Logistic regression evaluated variables correlated with memory function at each point in time. A Reliable Change Index (RCI) was performed to identifty changes at individual measures. **Results:** Forty-four to 52% of patients with left HS had normal either preoperative verbal learning (VL) or memory (VM) and this associated with worsening at T2 and T3 (p=0.029 and 0.045 for VL; p<0.015 and p<0.001 for VM, respectively). Between 50 and 70% of those with left HS and normal preoperative VM or VL sustained function at T2, although the majority worsened in the long term (T3). Around 30% of patients with left HS improved function in VM or VL at T2, but most worsened at T3. In contrast, 15% of patients reversed from dysfunctional to normal performance in VL between T2 and T3. Visual memory (VisM) following non-dominant surgery also worsened at T3 for patients who improved or sustained normal performance between T1 and T2. Worsening in VM or VL correlated with normal preoperative function, younger age, low schooling, surgery in the dominant side and nonselective surgical technique. Around 91% of the operated patients showed no reliable change in memory tests across T1-T2, T2-T3 and T1-T3. **Conclusion:** Memory preservation or improvement in the first few post-operative years is usually not sustained in the long term, suggesting that disease progression surpasses plasticity over the years.

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Long-term neurological impact of boxing: sensitivity and specificity of diagnostic tools for chronic traumatic encephalopathy

Sephora Sabrina Candido de Almeida[1], Renato Anghinah[2], Carla Cristina Guariglia[2], Tarcila Marinho Cippiciani[2], Ana Carolina Gomes[1], Isabela Martins de Freitas[1], Diogo Haddad Santos[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
Universidade de São Paulo, Faculdade de Medicina, Centro Ambulatorial de Reabilitação Cognitiva para Traumatismo Craniano, São Paulo SP, Brazil.

Correspondence: Ana Carolina Gomes; Email: gomesac90@gmail.com.

Background: The long-term neurological consequences of cumulative head trauma were initially recognized in professional boxers. Over many years, these athletes are on the receiving end of thousands of blows to the head of varying intensity, in sparring and matches. The initial description of what is now known as chronic traumatic encephalopathy (CTE) was reported in boxers in 1928. The clinical features of CTE include various symptoms affecting the pyramidal and extrapyramidal systems, which manifest most often as disturbed gait and coordination, slurred speech and tremors, as well as cerebral dysfunction causing cognitive impairments and neurobehavioural disturbances. **Objective:** To evaluate the sensitivity and specificity of neuroclinical, neuropsychological and electroencephalogram (EEG) findings in the diagnosis of chronic traumatic encephalopathy in a cohort of retired boxers. **Methods:** A prospective observational study was conducted. Eigth retired boxers and 14 controls were evaluated and submitted to neurological evaluation, neuropsychology evaluation and EEG. **Results:** In the retired boxers group the average age was 64.3 years (55.7 in the CG), average of 9.8 schooling years (13.7 in the CG). The average results in MMSE was 22.8 (29 in the CG), the average verbal fluency for letter F was 11.3 (13,5 in the CG), the average digit span in backwards was 2.5 (3.8 in the CG), the average verbal fluency for animals was 13.8 (24 in the CG), the average clock-drawing test was 8.6 (9 in the CG) and the average FAB (Frontal Assessment Battery) was 13.3 (17.5 in the CG). Visual EEG analysis showed a predominance of posterior alpha activity with 9.5 Hz average in the boxer participants and 11 Hz in the control group (CG). The analysis with low resolution electromagnetic tomography (LORETA) showed that 5 boxers had hippocampal asymmetry (3 had less activity

in the right hippocampus and 2 had less activity in the left hippocampus). Of the 9 boxers, 3 had previous TBI history. Our findings showed that the boxers scored in the MMSE and verbal fluency below the expected average for schooling, which would be 26 and 13 points, respectively. **Conclusion:** In general, the boxers performed within the normal ranges in the tests, except for MMSE and VF. Of the 9 boxers, 5 presented hippocampal asymmetry, consisting with the memory complaints. Three of 9 boxers presented neurological alterations - had parkinsonian syndrome. This can be explained by the mechanism of head trauma they suffer in boxing.

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Long-term outcome of Guillain-Barré syndrome associated with zika virus infection

Mario Emílio Teixeira Dourado Junior[1], Heric Weverton dos Santos[1], José Victor de Carvalho Dantas[1], Fátima de Menezes Dantas[1], Miguel Inácio Silva Gomes[1], Cássio Ricardo de Medeiros Souza[1], Selma Maria Bezerra Jerônimo[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Mario Emilio Teixeira Dourado Junior; Email: medourado03@gmail.com.

Background: The incidence of Guillain-Barré syndrome (GBS) has increased in Brazil since 2015, which initially was speculated to be secondary to the ZIKV and other newly introduced pathogens. The prognosis and overall outcome of Zika virus-associated GBS remain unclear. **Objective:** To investigate long-term neurological residua after GBS after ZIKV infection in the state of Rio Grande do Norte, Brazil **Methods:** The study included all GBS subjects diagnosed in 2015. Cases were considered having ZIKV disease if they had a rash with at least two other ZIKV-like symptoms in addition to positive anti ZIKV IgM or IgG. Information on signs or symptoms during the acute phase of the disease was retrieved from medical records. Patients were personally interviewed and re-examined or were contacted by phone 3 to 9 years after disease onset about residual symptoms and changes in daily living. Disability and handicap were assessed using the Hughes scale. **Results:** 34 patients were diagnosed with GBS during this period, 22 (64.7%) were included and examined at mean 52.81 months (SD 28.84) after disease onset. In the acute phase: mean age, years, was 44.41 mean (SD 15.79); 11 (50%) gender male; AIDP was the most frequent subtype, 18 patients (81.1%). The nadir varied from one day to 3 to 25 days (mean 8.5; SD 5). At that time 15 (68.18%) were severely disabled (grades 4 or 5 of the Hughes' scale). Four (18.8%) participants required ventilator support. All received IVIG treatment. At follow-up, the facial paralysis found in 3 (13.6%) participant and 3 (13.6) participants experienced paresthesia. 18 (81.8%) participants had completely recovered (Hughes 0); 3 (13.6%) patients able to run with minor signs or symptoms (Hughes 1) and 1 (4.5%) participant were incapable of running (Hughes 2). **Conclusion:** Over 90 % of GBS patients diagnosed when ZIKV was first introduced in Brazil had a more or less complete functional recovery. These data suggest that long-term outcomes and prognosis of Zika virus-associated GBS do not differ in severity from GBS due to other triggers., but of also importance was the prompt IVIG treatment provided timewise.

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Long-term survival of an 11-year-old boy with brainstem ganglioglioma associated with severe obstructive and central sleep apnea

Sayuri Aparecida Hirayama[1], Mariana Graça Nasr[2], Maria Clara Foloni[1], Rodrigo de Carvalho Pazeto[1], Lucas Martins Teixeira[1], Letícia Lambert[1], Gilmar Fernandes do Prado[1], Karla Carlos[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.
[2]Hospital Central da Aeronáutica, Rio de Janeiro RJ, Brazil.

Correspondence: Sayuri Aparecida Hirayama; Email: sah.hirayama@gmail.com.

Case presentation: An 11-year-old boy experienced intense snoring since the age of 5, along with waking up at night with difficulty breathing and restless sleep. By the age of 6, he also had excessive drooling, swallowing difficulties, and vertigo with an unsteady gait trending downward to the left. At 7, he started complaining of severe headaches. An MRI discovered a wide lesion spreading from the medulla to the fourth ventricle and upper cervical spinal cord. He was referred to Neurosurgery with the final diagnosis of brainstem ganglioglioma (WHO grade 1). After starting chemotherapy, the patient was referred to our service for desaturation and dyspnea during sleep. Polysomnography revealed an obstructive sleep apnea (OSA) index of 90.8 and a central sleep apnea (CSA) index of 22.1, with a severe desaturation of 61%. The CPAP (continuous positive airway pressure) titration was recommended and a pressure of 10 cmH2O abolished the obstructive apneas without changing the central apneas. Tracheal dead space increasing (respiratory center CO2 modulation) was also inefficient. He started CPAP treatment, what improved some of his symptoms, but his mother observed peripheral desaturations of up to 69% with tachycardia. Upon waking up, he presented symptoms of carbonarchosis, requiring frequent in-hospital noninvasive ventilation. Hospitalizations were also needed due to infections. Once the CPAP machine monitoring system persistently showed high central apnea index, supplemental nocturnal oxygen was recommended and a servo-ventilation titration was performed, aiming to treat central and obstructive sleep apnea, that was succesful with a EPAP = 6, PS = 6 to 13 cmH2O. **Discussion:** Patients with brainstem gangliogliomas typically had only a few months of survival after surgery. Our patient has completed one year of follow-up in our service, showing significant improvement in his oncological condition, school activities, and marked body development. CSA is a rare disorder in children, but it can be associated with underlying conditions such as brain tumors. Gangliogliomas are rare tumors accounting for 0.33-1.3% of all primary brain tumors, mostly affect children and young adults. Only 3% of gangliogliomas occur in the brainstem. Symptoms of children with brainstem gliomas vary depending on the location of the lesion. In our case, the tumor was located in the medula oblongata, which contains the respiratory center. The treatment of CSA depends on the severity of symptoms but includes optimization of underlying etiology. The best option for our patient was surgery, followed by servo-ventilation, and we believe that his long-term survival relies on this approach. **Final comments:** CSA is a rare disease in the pediatric population. Detailed history taking and complementary exams, such as polysomnography, should be performed, and a thorough etiological investigation should be conducted. Individualized management can improve CSA and the clinical condition in most patients.

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Loss of bilateral voluntary eyelid opening in a patient with bilateral thalamic infarction: apraxia or dystonia? A case report

Thamires Marx da Silva Santos[1], Rodrigo Santiago Gomez[1], Ricardo Oliveira Horta Maciel[1], Leonardo Dornas de Oliveira[1], André Luis de Oliveira Silveira[1], Italo Pena Guimarães[1], Nathália Luísa Carlos Ferreira[1], Gustavo Carvalho Oliveira Gonçalves Machado[1], Maria Luiza Nóbrega Lima de Albuquerque[1], Sabrina Gesteira Souza[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[2]Universidade Federal de Minas Gerais, Hospital das Clínicas, Belo Horizonte MG, Brazil.

Correspondence: Thamires Marx da Silva Santos; Email: marxthamires@gmail.com.

Case presentation: We report the case of a 72-year-old male patient admitted with sudden onset of mental confusion and inability to open both eyelids. He had a history of alcohol and tobacco use, hypertension, and coronary artery disease, with no prior cognitive impairments reported. Upon admission, he exhibited disorientation and confused speech without signs of aphasia, slight left hypotropia without diplopia, and an inability to voluntarily open his eyelids, with evident contraction of the frontal muscles during attempts and no involuntary eyelid contractions, raising the possibility of eyelid opening apraxia. Magnetic resonance imaging showed posterior circulation infarction with bilateral thalamic involvement. Electromyography (EMG) of the levator palpebrae superior muscles, which were functional, and the orbicularis oculi muscles, which showed sustained contraction at rest, was performed. One month after the ictus, the patient showed partial improvement, spontaneously opening about one-third of his eyelids, but continued to have cognitive deficits. **Discussion:** Eyelid opening apraxia is defined as a non-paralytic motor disorder characterized by difficulty in voluntarily opening the eyelids in the absence of visible contraction of the orbicularis oculi muscle, but its classification as true apraxia is debatable. EMG studies suggest that the pathophysiology of this phenomenon involves involuntary inhibition of the levator palpebrae superior muscle and/or sustained contraction of the orbicularis oculi muscle. Under normal conditions, the contraction of the levator palpebrae superior muscle leads to the reciprocal relaxation of the orbicularis oculi muscle, allowing eyelid opening. In the reported case, sustained contraction of the orbicularis oculi muscle prevented its relaxation, despite the functioning of the levator palpebrae superior muscle, thereby hindering eyelid opening. According to the literature, this observed pattern may be more appropriately classified as blepharospasm-related eyelid dystonia. The anatomical bases are not well defined, but there is evidence of an association with alterations in the basal ganglia, the rostral midbrain and the frontal lobes. Case reports in patients with bilateral thalamic infarction suggest that the paramedian thalamic nuclei may play a role in the neural network responsible for voluntary eyelid movements. **Final comments:** The loss of voluntary eyelid opening is a rare condition that can significantly impair patient functionality. Its pathophysiological and anatomical bases are not fully understood and may present different patterns, making the term eyelid opening apraxia inappropriate. More EMG and functional imaging studies are needed to better characterize this condition.

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Low frequency repetitive stimulation as a tool to differentiate Eaton-Lambert myasthenic syndrome from myasthenia gravis

Lucas Marenga de Arruda Buarque[1], Nicolas Ruan dos Santos Cavalcante[1], Jose Pedro Soares Baima[1], Ian Felipe Barbosa Souza[1], Stefano Machado[1], Fernando Cavalcanti de Sá e Benevides Falcão[1], Andre Macedo Serafim da Silva[1], Rodrigo de Holanda Mendonça[1], Carlos Otto Heise[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Lucas Marenga de Arruda Buarque; Email: l.marengabuarque@gmail.com.

Background: It's has long been stated that high frequency repetitive stimulation (HFRS) is a valuable tool to differentiate myasthenia gravis (MG) from Lambert-Eaton Myasthenic Syndrome (LEMS), nevertheless it is not performed routinely in neuromuscular junction (NMJ) studies. **Objective:** We describe the use of low frequency repetitive stimulation (LFRS) with a train of six stimuli in order to differentiate between LEMS and MG with antibodies to acetylcholine receptor (AChR). **Methods:** The ratio of the 4th and the 6th amplitude potential were compared between 04 patients diagnosed with LEMS (group 1) and 11 MG AChR patients (group 2). **Results:** The amplitude ratio was 1.21 in group 1 and 0,92 in group 2. Of 11 patients in group 2, only one had a decremental index above 1; and, in group 1, only one patient had a decremental index under 1, with whom was diagnosed myasthenia gravis lambert-eaton overlap syndrome. **Conclusion:** We suggest that an amplitude ratio above 1 in LFRS (train of 6 stimuli) in patients with a neuromuscular junction syndrome for screening LEMS. A relation above 1.0 of the 4th and the 6th amplitude potential is suggestive of LEMS.

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Lower pain and reduced strength in lower limbs as a complaint in stiff-person syndrome, a rare autoimmune condition: a case report

Victor Xavier da Silva[1], Camila Maria Barreto Ribeiro[1], Lucas Vergne Cheade Lins[1], Julia Cordeiro Braga[1], Adriana Alves Rapp[1], Marcel Leal Ribeiro[1], Brenda Francisca Cardoso Brito[1], Paulo Ricardo Sampaio de Melo[1], Karla Oliveira Couto[1], Eduardo de Oliveira Costa[1]

[1]Hospital Santa Izabel, Salvador BA, Brazil.

Correspondence: Victor Xavier da Silva; Email: victorxavier.vxs@gmail.com.

Case presentation: Patient 22 years old, male, with moderate low back pain, without other associated symptoms for 6 months, evolving for approximately 1 week with weakness in the lower limbs which he reports to be progressive in onset, with worsening for the last about 24 hours. Denies trauma, fever, weight loss. He had a previous diagnosis of Wolff-Parkinson-White syndrome. First-degree family member with vitiligo. In this context, the patient was admitted to a hospital unit for diagnostic elucidation. On admission physical examination, muscle strength was grade 5 in the upper limbs (UL) and grade 4+/5 in the lower limbs (LL), predominantly proximal. Grade 3 osteotendinous reflexes in the LL, with an increase in the reflex area and a discrete bilateral exhaustible Achilles clonus. Sensibilities preserved globally, without sensitive level. Preserved coordination. Bilateral plantar flexor cutaneous. Signs of pyramidal release present. MRI of the skull and spine showed no changes, CSF study showed protein-cytological dissociation (0.3 cells and 113 proteins) suspecting inflammatory polyradiculopathy, starting human immunoglobulin for 5 days, with clinical improvement. There was a search for neoplastic foci, rheumatological tests, myopathy markers (CPK, aldolase) in addition to serology, all of which were negative. After discharge from hospital within 90 days, he developed contractures and spasms in the lower back and right lower limb (RLL). Electroneuromyography demonstrated coactivation of agonist-antagonist muscles in RLL, without other changes, and a Stiff-Person syndrome (SPS) test was then carried out, with a posteriori reagent test for anti-GAD. Diazepam was started with symptom improvement and subsequent sessions of human immunoglobulin were scheduled, in addition to defining a rehabilitation strategy, screening for Type 1 Diabetes Mellitus (DM1) was negative. **Discussion:** Stiff person syndrome or SPS is an uncommon disorder characterized by progressive muscle stiffness, rigidity, and spasm involving the axial muscles, resulting in severely impaired ambulation. SSP is often associated with DM1, which may be a reflection of autoimmunity. Rarely occurring as a paraneoplastic syndrome. It has an estimated prevalence of 1-2 cases/1,000,000, and an incidence of 1 case/1,000,000/year. The majority of patients present between the ages of 20 and 50 years, being more common in women (ratio of 3:1). Diagnosis includes clinical characteristics and complementary exams. Some patients with SPS spectrum disorders require multiple immunological therapies. For example, intravenous immunoglobulin. **Final comments:** The case in question shows how presentations such as low back pain and complaints of weakness on the part of patients can, in fact, be attributable to other conditions, which in this case was attributed to the SPS condition, opportunely anticipating the diagnosis and so the treatment.

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Lupus encephalitis: a case report

Rodrigo Batista Irikura[1], Anouar Sacca Colognesi[1], Bruna Santos Silva Guzzardi[1], Sarah de Souza Chinelato[1], Gustavo Carvalho Viveiros[1], Isabella Brasileiro Quintella Migueis[1], Guilherme Leonel Gomes da Cunha[1], Talles Camin Rajab[1], Fábio de Araujo Pereira[1], Luiz Henrique Soares Santos Stefano[1]

[1]Faculdade de Medicina de Marília, Marília SP, Brazil.

Correspondence: Rodrigo Batista Irikura; Email: rodrigoirikura@gmail.com.

Case presentation: A 65-year-old female patient was referred to the Emergency Department due to progressive functional decline that began 4 months prior. Previously healthy and independent, she started experiencing febrile episodes interspersed with fluctuations in consciousness and disorientation. After one month, she developed motor impairment with gait difficulties and reduced verbal interaction with family members, progressing to mutism and global spasticity over two months, becoming bedridden. Initial evaluation found no significant findings, prompting a referral to the Neurology team for further investigation. Neoplastic screening revealed no abnormalities. An electroencephalogram showed global slowing of waves with periodic complexes. Cerebrospinal fluid analysis showed normal cell count and a protein level of 70mg/dL. Laboratory tests indicated moderate anemia, thrombocytopenia, elevated erythrocyte sedimentation rate (ESR), anti-DNA at 1:160, high-titer anti-Ro positivity, and consumed complement levels, consistent with active Lupus. The patient was prescribed pulse therapy with methylprednisolone 1g for 5 days, resulting in transient improvement. Intravenous human immunoglobulin was then administered, leading to progressive improvement after the infusion began. The patient was discharged alert, oriented, communicative, and with grade IV strength globally. **Discussion:** Lupus is a chronic autoimmune disease characterized by periods of activity and remission, more prevalent in women under 40. Neuropsychiatric symptoms occur in about 40% of cases, with a minority involving mental status changes, constituting a medical emergency. Anti-DNA antibody positivity is associated with encephalitis targeting the NMDA receptor. Treatment focuses on disease control with continuous immunosuppression, often requiring multiple immunosuppressant classes. Prognosis varies depending on the extent of central nervous system involvement. **Final comments:** The patient had no prior diagnosis of any autoimmune disease, presenting with subacute changes in mental status and consciousness. She was admitted bedridden and mute, but showed significant clinical improvement and return of functionality with the instituted therapy. This case presented a diagnostic challenge and a robust clinical response, highlighting its relevance.

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Machine learning analysis of gray matter and disease severity in post-COVID patients

Thais Maria Santos Bezerra[1], Ítalo Karmann Aventurato[1], Lucas Scárdua Silva[1], Brunno Machado Campos[1], Ana Paula Beppler Lázaro Lino[1], Marina Koutsodontis Machado Alvim[1], Mateus Nogueira[1], Fernando Cendes[1], Benjamin James Sinclair[2], Clarissa Yasuda[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Departamento de Neurologia, Campinas SP, Brazil.
[2]Monash University, Melbourne, Australia.

Correspondence: Thais Maria Santos Bezerra; Email: t243567@dac.unicamp.br.

Background: Although cognitive dysfunction and structural brain changes have been described after Sars-Cov-2 infection, little is known about the impact of the severity of acute infection. Whether gray matter changes are severe enough to automatically differentiate between post-COVID and non-infected individuals is unknown, and neither is the power to classify individuals according to the acute severity of COVID infection. **Objective:** Here, we applied machine learning to investigate if grey matter alterations can distinguish between individuals after mild or moderate infections and non-infected controls. **Methods:** We acquired high-resolution T1-weighted images (3T) from 327 post-COVID subjects (285 mildly infected non-hospitalized and 42 moderately infected) and 151 healthy controls (non-infected) at the University of Campinas. The T1 images were segmented using Freesurfer 7.1.1, which yielded 68 cortical thickness measures and 14 subcortical volumes. Age and sex were normalized using a reference cohort of 37,407 individuals. Machine learning models were built using Python3 and scikit-learn. The final models were selected based on the area under the receiver operating characteristic curve (AUC), the f1 score, and accuracy. The reported results are from cross-validation scoring, and hyperparameter tuning was performed. **Results:** The SVM achieved an accuracy of 69.1%±0.6% in differentiating patients from controls based on thickness and subcortical volumes. This accuracy was observed to be 69.3% when both parameters were considered. In differentiating between mild cases and controls, SVM yielded accuracies of 66.1%±2.6% with thickness and 65%±1.8% with subcortical volumes, while 65.8%±2.5% was observed with both. The separation between moderate versus controls or mildly infected patients was not identified, as the f1-score was zero, despite the high accuracies observed on both (78.2% and 87.2%). **Conclusion:** The application of machine learning techniques enabled the differentiation between patients diagnosed with COVID-19 and those in the control group, as well as between patients exhibiting mild symptoms and those in the control group. These preliminary results indicate the potential for structural brain changes even after a mild infection. Additionally, the cortical and subcortical information appears to exhibit comparable patterns, as evidenced by the similar accuracies and the lack of significant improvement in results when both are included. As the disease progressed, the models were unable to differentiate moderate infected patients from controls or from mildly infected patients, despite achieving high accuracy rates, even after tuning the hyperparameters of the models. This is due to the fact that the dataset is severely imbalanced. Consequently, in the event of datasets exhibiting potentially intricate interactions between data points, all instances are ultimately classified as the majority class, resulting in a f1-score of zero.

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Machine-learning models applied to independent cortical folding variables distinguish tle subgroups

Thais Maria Santos Bezerra[1], Lucas Scárdua Silva[1], Ítalo Karmann Aventurato[1], Ricardo Brioschi[1], Marina Koutsodontis Machado Alvim[1], Brunno Machado Campos[1], Fernando Cendes[1], Guilherme Ludwig[2], Benjamin Sinclair[3], Clarissa Yasuda[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Departamento de Neurologia, Campinas SP, Brazil.
[2]Universidade Estadual de Campinas, Instituto de Matemática, Estatística e Computação Científica, Campinas SP, Brazil.
[3]Monash University, Melbourne, Australia.

Correspondence: Thais Maria Santos Bezerra; Email: t243567@dac.unicamp.br.

Background: Although gray matter atrophy in temporal lobe epilepsy (TLE) has been repeatedly demonstrated, advanced in-vivo MRI analyses of cortical gyrification may aggregate information about complex pathological processes. A recent study validated a cortical folding universal model based on metrics of cortical thickness (T), total surface area (At) and exposed surface area (Ae), describing a universal scaling law: At*sqrt(T)=k*Ae^(5/4). This model was advanced to a vector base space describing the human cortex with 3 perpendicular and dimensionless variables: axonal tension (K), cortical shape complexity (S) and brain isometric size (I). So far, little is known about the pathological changes of these cortical variables in TLE and their capacity to discriminate subgroups. **Objective:** To compare pathological changes in newly derived measures between groups of mesial temporal lobe epilepsy (TLE) and healthy controls. **Methods:** Cortical measures were extracted from T1-weighted images of 337 controls and 165 patients (95 LTLE and 69 RTLE) using Freesurfer 7.4.1. Novel measures were derived with the CorticalFoldingAnalysisTool/MATLAB2019. Metrics I, K, and S were computed as scalar metrics of vectors that represented each lobe in a log-normalized space (At,Ae,T^2) as (1,1,1), (1,-5/4,1/4) and (3/2,3/4,-9/4), respectively. Value changes of I represent size, K represents tension/pressure and S represents shape. Metrics were corrected for age and sex and standardized relatively to the controls. Group differences were evaluated with repeated measures ANOVA and Bonferroni's post-hoc pairwise correction using SPSS26. Machine learning classification models were built to distinguish between groups using Python3/scikit-learn. Models were scored with 5-fold cross-validation using accuracy, f1-score, and area under the curve (AUC). **Results:** Compared to controls, bilateral hemispheres were affected in LTLE and RTLE. For LTLE, ipsilateral I for parietal and temporal lobes were reduced (p-value=0.03, p-value<0.001), and K for the temporal lobe was augmented (p-value<0.001). LTLE contralateral hemisphere had a bigger K for the temporal lobe (p-value<0.001). For RTLE, ipsilateral I for temporal (p-value=0.015) and occipital (p-value=0.033) lobes were reduced (p-value=0.015, p-value=0.033), while K for temporal and S for parietal lobe were augmented (p-value<0.001, p-value=0.006). RTLE contralateral hemisphere had a bigger S for the parietal lobe (p-value=0.005). Machine learning models consistently separated TLE patients and controls (AUC=0.71±0.02) and RTLE from LTLE (AUC=0.79±0.08) using logistic regression. **Conclusion:** Although both temporal ipsilateral regions were affected in TLE, side of hippocampal atrophy differed in disease progression, as different measures were abnormal for LTLE or RTLE. These metrics, although sparse, can distinguish TLE patients from controls.

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Magnetic resonance imaging abnormalities in idiopathic intracranial hypertension: a study of 40 female patients

Annelise Akemi Higa Lee[1], Ana Carolina Montouro Storarri[1], Lorrane Campidelli Arthuzo[1], Luis Armando Sawada[1], Beatriz Marques dos Santos[1], Thassia Cavalcante de Oliveira Moraes Marques[1], Bruno Pissolati Mattos Gonzaga[1], Renan Barros Domingues[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Ana Carolina Storarri; Email: anastorarri@gmail.com.

Background: Idiopathic intracranial hypertension (IIH) is a condition characterized by increased intracranial pressure with no detectable cause. MRI is an importat tool in diagnosing and understanding IIH, providing insights into the anatomical changes associated with this condition. This study aims to describe the MRI findings in a cohort of 40 female patients diagnosed with IIH. **Objective:** To evaluate the frequency and type of abnormalities detected on MRI in female patients with IIH. **Methods:** We conducted a retrospective analysis of MRI findings in 40 female patients diagnosed with IIH. The average age of the patients was 33.9 years (standard deviation of 11.5 years). The MRI abnormalities evaluated included sella turcica deformities, optic nerve abnormalities (including optic nerve tortuosity, distended optic nerve sheath, and optic nerve head protrusion), globe flattening, and transverse sinus stenosis. The data were analyzed to determine the prevalence of each abnormality. All the patients were evaluated at the Headache Outpatient Clinic of Santa Casa de Misericórdia de São Paulo. **Results:** The analysis revealed the following abnormalities:

- Sella turcica deformities in 14 patients (35%)

- Optic nerve abnormalities in 35 patients (87.5%)

- Globe flattening in 8 patients (20%)

- Transverse sinus stenosis in 14 patients (35%).

Conclusion: Our study demonstrates that optic nerve abnormalities are the most frequent MRI finding in female patients with IIH, observed in 87.5% of cases. This is a higher frequency compared to sella turcica deformities, which were present in 35% of the patients. These findings differ from other series in the literature, where sella turcica abnormalities are often reported as more common. This highlights the variability in anatomical manifestations of IIH and underscores the importance of comprehensive MRI evaluation in these patients.

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Main factors related to the development of post-avc headache - a systematic review

Sávio Roberto Silva Costa[1], Fernando Gabriel dos Santos Santiago[1], Mariangela Moreno Domingues[1], Danusa Neves Somensi[1], Emanuel de Jesus Soares de Sousa[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Sávio Roberto Silva Costa; Email: saviorsc20@gmail.com.

Background: Stroke is one of the main causes of morbidity and mortality in the world. In Brazil, there is an estimated prevalence of 2,231,000 cases, and the prevalence of post-stroke disability is 29.5% for men and 21.5% for women. Among the clinical manifestations after stroke, headache is a common complaint, with observational studies showing frequencies of 11 to 39%. However, despite the high prevalence of post-stroke headache, few studies characterize it and relate it to predisposing factors. **Objective:** To identify which factors may predispose to the development of post-stroke headache. **Methods:** This is a systematic literature review based on the following guiding question: what are the main factors that can predispose to the development of post-stroke headache? The databases used were PubMed and Scielo. The descriptors were divided into two groups and permuted as long as one of each group was included in the search; thus, the groups were "disease": Stroke, Ischemic Stroke and Hemorrhagic stroke; "Sequela": headache disorder and headache; and their respective descriptors in Portuguese; the groups were joined by the Boolean operator AND. Full articles published between 2018 and 2022 were included. The PRISMA method was used to construct the review; in addition, the Joanna Briggs Institute tool was used to analyze the quality of the selected articles. **Results:** The studies showed a prevalence of headache after stroke of between 10.4% and 50% and converged on most of the characteristics, describing pain of moderate intensity and unilateral or bilateral localization of a pattern similar to tension headache. To a lesser extent, they identified nausea and photophobia associated with the headache, a typical symptom of migraine. However, these patients proved to be unresponsive to triptans, a common drug in the abortive treatment of migraine. In addition, patients who already had a headache before the stroke episode are more likely to develop post-stroke headache, which occurs with a change in the characteristics of the initial symptom. With regard to the location of the stroke and headache, only one study found a statistically significant relationship, with infarcts in the cerebellar region. It is also worth mentioning that good condition at the start of the stroke, lack of sleep, the presence of pathological changes in the intracranial arteries (mainly dissections) and strokes that affected large cerebral arteries are statistically significant factors in the development of post-stroke headache. **Conclusion:** Post-stroke headache is a chronic, recurrent condition that tends to be tension-type, moderate in intensity and unilateral or bilateral. It should also be noted that vascular pathologies may be closely related to this condition. More studies are needed to assess the relationship between topography and post-stroke headache.

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Malignant brain neoplasms in the Brazilian midwest from 2014 to 2023: an epidemiological study

Gustavo Hayasaki Vieira[1], Giovana de Oliveira Araújo[1], Gustavo Cartaxo de Sousa Melo[1], Isabela Maciel Mesquita[1], Milena Almeida Barbosa[1], Paulo André Silva Miranda[1], Paulo Sérgio Machado Diniz[1], Rafaella Rodrigues de Freitas Vieira[1], Samira Dionísio Maia[1], Valentina Melo Rosa Centofanti[1]

[1]Pontifícia Universidade Católica de Goiás , Goiânia GO, Brazil.

Correspondence: Gustavo Hayasaki Vieira; Email: gushayasaki@gmail.com.

Background: Malignant Brain Neoplasms represent 1.4 to 1.8% of malignant tumors worldwide. It involves the proliferation of cells within the brain, presenting symptoms that vary depending on the location and extent of tissue involvement. Although rarely metastatic to other organs, this tumor can destroy brain tissue as it grows. Due to the scarcity of studies on malignant brain tumors in the Brazilian Midwest, it is important to conduct an epidemiological analysis of the patients of this region, focusing on identifying the frequency of this condition and enhancing diagnostic and treatment measures. **Objective:** To comparatively analyze the epidemiological profile of patients with Malignant Brain Neoplasms in the regions of the Brazilian Midwest. **Methods:** This is a descriptive, retrospective, and quantitative study based on data from the Hospital Information System (SIH/SUS), provided by the Department of Information Technology of the Unified Health System (DATASUS), regarding patients over the past ten years in Goiás, Mato Grosso, Mato Grosso do Sul, and Distrito Federal. Filters applied included: ICD-10 morbidity list (malignant brain neoplasms), midwest region, race/ethnicity, gender, age group, number of deaths, mortality rate, available period (January 2014 to December 2023), and year of data processing. Data were tabulated in Excel and subjected to descriptive statistical analysis using R 4.40 with RStudio 2024.04.1 + 748. Since this study utilizes publicly available anonymous data, it is exempt from the Ethics Committee's approval. **Results:** During the study period, 1,420 deaths were confirmed,

with an annual average of 142 deaths (±22.07). The year 2022 exhibited the highest mortality rate (12.88%), with indigenous populations being the most affected (25%). In this context, the state of Goiás had the highest number of deaths (40.99%), while Mato Grosso do Sul had the lowest (13.66%). However, Mato Grosso do Sul had the highest overall mortality rate (15.85%) and a 100% mortality rate among the Indigenous population. The age group with most deaths was 60-69 years (339 cases). Regarding hospitalizations, Goiás led with 41.4% of cases, while Mato Grosso do Sul had the lowest proportion, 9.94%. Furthermore, the disease was more predominant in males (761 cases) compared to females (659 cases). **Conclusion:** Indigenous people show the highest mortality rate and are a vulnerable community to the disease, reflecting the effects of social inequality, and territorial segregation. Moreover, the state of Goiás had the highest number of hospitalizations alongside the second lowest mortality rate. Despite having fewer deaths compared to Goiás, Mato Grosso do Sul has a smaller population, thus proportionally it is the most affected state by the disease as it exhibits the highest mortality rate. Further research is required to explore the causes of regional differences in hospitalizations and mortality rates throughout the Brazilian Midwest

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Malignant neoplasm presenting as subacute longitudinally extensive myelopathy

Andre Felipe Candêas Amorim[1], Gabriela Rodrigues Tomaz[1], João Vitor Gerdulli Tamanini[1], Enedina Maria Lobato de Oliveira[1], Isabella Vargas Baldon[2], Henrique Ballalai Ferraz[1], Carolina Candeias da Silva[1], Heitor Raia Bottura[1], Adrialdo José Santos[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Andre Felipe Candêas Amorim; Email: andfcamorim@gmail.com.

Case presentation: A 22-year-old man reports the beginning of pain and weakness in the lower right limb, affecting the contralateral limb a few days later. After 10 to 15 days, the patient started to need help to walk. In addition, progressively, he began to present numbness up to the nipple level bilaterally, associated with urinary retention and constipation. After 3 months, the patient noted neck pain, which intensified with head flexion radiating to the lumbar region. On examination, he presented hypotonia in the lower limbs. The strength was 0/5 bilaterally in the lower limbs and 4/5 in the upper limbs. About the reflexes Biceps, brachioradialis, triceps were 2/4 bilaterally and patelar and Achilles were 0/4 bilaterally. No clonus were presente and Cutaneous plantar reflex was absent bilaterally. Tactile, painful, proprioceptive and vibratory sensation were absent bilaterally up to the level of T6, remaining hypoesthesia in the left side up to C2. Lhermitte's sign was present. **Discussion:** Initially, a complete spinal cord syndrome was raised as a syndromic diagnosis. Due to the subacute presentation, mainly inflammatory and infectious conditions were identified as etiologies. The general lab revealed an VHS of 25mm/h. Rheumatological tests came back normal (Antinuclear antibody with dense fine speckled pattern 1:160; Negative Anti-dsDNA; Negative Rheumatoid Factor; Complement C2 fraction 122 (Normal), Total complement 103 (Normal). Serology showed a VDRL of 1:1 with a positive treponemal test that were seen as a serological scar, since the patient had previous treatment. In the sequence of investigation, a MRI was performed and revealed an extensive heterogeneous intramedullary lesion from the medulla oblongata to the conus medullaris, centrally located and tumefactive in appearance. It showed gadolinium enhancement and areas of hydrosyringomyelia. In the sequence, a cerebrospinal fluid was collected with the presence of 11cells/mm^3 of lymphomonocyte predominance, glucose 44mg/dL, protein 159mg/dL, lactate 22.71mg/dL, negative GeneXpert, negative microorganism cultures and research for neoplastic cells positive. At the end of the investigation, a spinal cord biopsy was performed and the anatomopathological analysis, together with immunohistochemistry, revealed a Malignant Neoplasm with a Small Cell morphological pattern, with an Embryonic Tumor as the main diagnosis. **Final comments:** The case above points to a rare differential diagnosis of cases of longitudinally extensive myelopathy. Thus, despite a rapid evolution over 3 months with involvement of the entire length of the spinal cord, which initially suggested an inflammatory or infectious condition, it was ultimately revealed to be an aggressive malignant neoplasm.

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Management of myopathic symptoms with oral dantrolene in a pregnant patient susceptible to malignant hyperthermia

Gabriel Saboia de Araújo Torres[1], Mariana Hiromi Manoel Oku[1], Gabriella Correa Dousseau[1], Filipe Tupinambá Di Pace[1], Cristiane Araújo Martins Moreno[1], Clara Gontijo Camelo[1], Rodrigo de Holanda Mendonça[1], Edmar Zanoteli[1], André Macedo Serafim Silva[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.

Correspondence: Gabriel Saboia de Araújo Torres; Email: gabriel.saboiat@gmail.com.

Case presentation: A 30-year-old female presented to the Neuromuscular Clinic with a history of muscle cramps, exercise intolerance, and fatigue since childhood, accompanied by persistently elevated creatine kinase (CK) levels reaching up to 2257U/L. She had presented a few events of myoglobinuria after physical efforts. Her physical examination did not reveal weakness or other associated findings. Whole exome sequencing revealed two variants in the RYR1 gene c.8197G>T; p.(Gly2733Cys) and c.7393G>A; p.(Asp2465Asn). Due to significant symptoms, treatment with oral dantrolene 25mg twice daily was initiated, resulting in substantial improvement in muscle cramps and post-exercise pain. Over subsequent months, the dose was adjusted to 25mg three times daily, leading to a reduction in CK levels to 237U/L. After two years of follow-up, the patient became pregnant with twins. Concerned about the safety profile of dantrolene, treatment was initially suspended, resulting in an increase in CK levels. Dantrolene was resumed at 29 weeks of pregnancy, leading to significant clinical and laboratory improvement. Despite a complex delivery at 36 weeks due to placental abruption and subsequent puerperal hemorrhage necessitating intensive care support for four days, the patient eventually recovered. Uterine hypotonia possibly related to dantrolene was considered. One month post-delivery, dantrolene was reintroduced with continued good response. Both children were born healthy and demonstrated normal neurodevelopmental milestones. **Discussion:** Malignant hyperthermia is characterized by a hypermetabolic state triggered by genetic variants affecting intracellular muscular calcium metabolism, notably in the RYR1 gene encoding the ryanodine-1 receptor responsible for calcium release from the sarcoplasmic reticulum during excitation-contraction coupling. Intravenous dantrolene is the cornerstone of acute malignant hyperthermia treatment, acting by blocking calcium release via the ryanodine receptor. Oral dantrolene has been explored in patients with RYR1 variants for managing muscle symptoms and reducing CK levels, showing promising results in case series. Limited literature exists on the use of oral dantrolene during pregnancy. The obstetric complications observed in our patient, including placental abruption and hemorrhage, may have been influenced by dantrolene use or the challenges inherent in a twin pregnancy, which carries a higher risk of peripartum complications. **Final comments:** Oral dantrolene appears beneficial in controlling symptoms of RYR1-related myopathy and malignant hyperthermia susceptibility, including during pregnancy. However, its safety in pregnant women warrants further investigation.

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Management of neurogenic bladder and bowel as a strategy to reduce epileptic seizures in a child with congenital zika syndrome: case report

Larissa Coutinho de Lucena[1], Amanda da Silva Santos[1], Vitor dos Santos Rodrigues[2], Jhulia Thaysa Macena da Costa[1], Marília da Costa Paiva[1], Amanda Georgia Diniz de Campos[3], Milena Thaisa Silva de Lima[2], Fernanda Gomes Mourão[1], Gabriel Alves Mendes Vasiljevic[1], Lilian Lira Lisboa[1]

[1]Instituto Santos Dumont, Macaíba RN, Brazil.
[2]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[3]Centro Universitário Maurício Nassau, Aracaju SE, Brazil.

Correspondence: Larissa Coutinho de Lucena; Email: larissa.lucena@isd.org.br.

Case presentation: 8-year-old girl with congenital zika syndrome (CZS), born by cesarean section at 39 weeks, 2,116g, 38cm, cephalic perimeter of 29cm and Apgar of 5/8. She had alterations in the Central Nervous System (CNS) as evidenced by tomography, supratentorial hydrocephalus and calcification of the basal nuclei. At 14 months, she was diagnosed with pharmacoresistant epilepsy, with an average of 5 seizures a day despite taking Sodium valproate 50mg, Phenobarbital 40mg and Nitrazepam 5mg. Motor development has functional limitation level V by Gross Motor Function Classification System.The patient has severe diffuse spasticity, bilateral hip luxation, laryngeal stenosis, gastrostomy diet dependency and lack of intentionality in her movements. Regarding bladder and bowel function, ultrasound (USG) of the upper urinary tract showed no alterations, but urodynamics revealed a neurogenic bladder with low compliance, a cystometric capacity of 37% of that expected and hyperactivity from 28ml on.The urinary diary showed low water intake (300ml/day) and constant use of diapers (average of 14 per day). The bowel movement diary showed a bowel movement frequency of less than once a week with the use of laxatives, type 1 and 2 stools on the Bristol Scale and previous formation of fecaloma. There were no signs of urinary infection or renal failure in the laboratory tests. Treatment included behavioral approaches and daily parasacral neuromodulation (10 Hz, 300 ms, 30 minutes). The study was approved by the Ethics and Research Committee (CAAE 51865521.5.0000.0129) and funded by the CNPq/MCTI/FNDCT-18/202, UNIVERSAL (403498/2021-0). **Discussion:** after two months of treatment, water intake increased to 1 liter/day, bowel function improved with an average bowel movement frequency of 4 times/week without laxatives, and stools type 5 to 6 on the Bristol Scale. The use of diapers was reduced to 8 a day. From the tenth day of treatment, there was a reduction in the frequency of epileptic seizures, which ceased completely from the twenty-sixth day of neuromodulation. Studies indicate that there is a two-way communication between the CNS, the gut and the intestinal microbiota, influencing the production of neurotransmitters and inflammatory responses that affect the brain. Findings in the literature indicate that changes in diet, such as ketogenic diets, can reduce indicators of refractory epilepsy. These findings suggest that the reduction in epileptic seizures may be related to changes in water and food intake during behavioral therapy. **Final comments:** this case suggests the possibility of a relationship between epilepsy and urinary and bowel dysfunction in children with CZS. These findings indicate the importance of proper management of urinary and bowel dysfunction in this population in order to reduce or control the epileptic seizures that are so frequent.

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Management of NMOSD in Brazilian public scenario

Rafaela Pessoa Alves Luso[1], Izabela Cristina de Faria Martins[1], Rodrigo Gonçalves Kleinpaul Vieira[1], Jessica Vaz de Melo[1], Gustavo Carvalho Oliveira Gonçalves Machado[1], Maria Luiza Nobrega Lima Albuquerque[1], Alexandra de Oliveira Lopes[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Rafaela Pessoa Alves Luso; Email: lusorafaela@gmail.com.

Background: Neuromyelitis optica spectrum disorder (NMOSD) is an uncommon immune-mediated disease that affects the central nervous system and presents with relapses frequently leading to disability. Cases clinically manifested as NMOSD may include aquaporin-4 antibody (AQP4-Ab) seropositive disease, myelin oligodendrocyte glycoprotein antibody disease (MOGAD), and double-seronegative disease. Diagnosis is based mainly on core clinical features, but serologic testing and magnetic ressonance imaging should be evaluated. These exames, as well as most therapies available are costly, therefore managing NMOSD in the public scenario of emerging countries represents a challenge. **Objective:** Analyze epidemiological data from a Brazilian cohort in a neuroimmunology referral public service in Belo Horizonte (Minas Gerais) and establish correlations about challenges regarding diagnosis and treatment. **Methods:** This study was retrospective and observational, analyzed data from electronic medical records from patients at a neuroimmunology referral service from January 2022 to June 2024, including demographic information, laboratory findings and treatment. Patients aged between 18 and 70 years were included, who had a neurological event suggestive of relapse and met the MOGAD and the NMOSD criteria. Patients with multiple sclerosis were excluded. Descriptive analyses of numerical and categorical data were conducted. **Results:** The sample included a total 32 patients, comprising 28 women and 4 men, with mean age of 43 years, 25% AQP4-Ab seronegative disease, 46% AQP4-Ab seropositive disease, 13% MOGAD, 13% double-seronegative disease and 3% did not undergo any sorological testing. Among current treatments prescribed to prevent relapses in NMOSD, azathioprine was the most used immunotherapy (31%). It was discontinued due to irregular use associated with financial matters, myelotoxicity and hepatotoxicity. Cyclophosphamide was the next choice (16%), discontinued mostly due to accumulated dose, but also due to hepatotoxicity. Rituximab, steroids alone and methotrexate were also prescribed. **Conclusion:** This study shows that access to targeted antibody testing remains limited in Brazil, even at referral services, postponing definitive diagnosis and treatment in many cases. Cyclophosphamide and steroids are available in the public health system at a tertiary center, but these do represent long term solutions. Azathioprine and Rituximab have shown to reduce relapses and disability, but are hard to maintain due to its high costs. There is urgent need for better options to treat NMOSD and MOGAD patients in the public scenario in Brazil, including hope for Humanized Monoclonal Antibody for aggressive and refractory AQP4-Ab disease. This strategy might prove to be cost-effective since it is expected to reduce expenses regarding young adults disabilities, hospital admissions and relapses treatments.

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Management of severe intracranial hypertension in a patient with cerebral venous thrombosis

Ana Carolina Montouro Storarri[1], Annelise Akemi Higa Lee[1], Renan Barros Domingues[1]

[1]Hospital Samaritano Paulista, São Paulo SP, Brazil.

Correspondence: Ana Carolina Storarri; Email: anastorarri@gmail.com.

Case presentation: We report the case of acute management of intracranial hypertension with lumboperitoneal shunt in a young, obese, smoker female patient, who presented with an acute case of intracranial hypertension, with headache, nausea, vomiting, diplopia, papilledema and severe bilateral visual loss (counting fingers less than 1 meter) secondary to extensive venous thrombosis involving the right sigmoid sinus and internal jugular vein. Upon admission, the patient underwent neuroimaging with study of intracranial vessels (magnetic resonance imaging and magnetic resonance venography) in addition to a diagnostic lumbar puncture - the latter showing an opening pressure greater than 100 cm H2O. Initial management included anticoagulation with heparin. Despite this, the patient worsened the signs of elevated intracranial pressure (ICP) and progressive neurological deterioration. To manage the intracranial hypertension, high-dose acetazolamide was administered, providing partial relief of symptoms. Mannitol was also used intermittently to reduce acute spikes in ICP. However, due to persistent elevated ICP and the risk of permanent visual loss and herniation, a lumboperitoneal shunt was placed. This intervention resulted in significant clinical improvement and stabilization of the patient's condition. **Discussion:** Intracranial hypertension (ICH) is a common complication of cerebral venous thrombosis (CVT). The management of ICH in CVT is complex and requires a tailored approach. Recommendations regarding the management of intracranial hypertension in cerebral venous thrombosis are scarce due to the lack of controlled studies. High-dose acetazolamide can be effective in reducing ICP by decreasing cerebrospinal fluid (CSF) production. Mannitol, an osmotic diuretic, is useful for acute reduction of ICP but must be used cautiously due to potential side effects. In some cases of ICH, relief lumbar punctures are an option, however, in this case this strategy would be incompatible with the need for full anticoagulation. In cases where medical management is insufficient, surgical options such as lumboperitoneal shunting offer an alternative to alleviate pressure and prevent neurological deterioration. **Final comments:** This case underscores the importance of a comprehensive management strategy for intracranial hypertension in cerebral venous thrombosis, integrating pharmacological and surgical interventions. Each case requires individual assessment to determine the most effective combination of treatments, aiming to mitigate the risks and enhance patient outcomes.

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Marchiafava-Bignami disease: a diagnostic challenge

Roberta Monteiro de Souza[1], Carolina Rocha da Silveira Brugger Cardoso[1]

[1]Universidade Federal do Rio de Janeiro, Instituto de Neurologia, Rio de Janeiro RJ, Brazil.

Correspondence: Carolina Rocha da Silveira Brügger Cardoso; Email: carolinarbrugger@gmail.com.

Case presentation: A 43-year-old female presented behavioral changes after irregular follow-up of bariatric surgery, in 2018. The patient started drinking excessively and taking prescription medications without supervision. In 2019, she was found unconscious at home, being admitted to the ICU for further investigation, but no neurological or psychiatric diagnosis was found at the time. Since then, family members have observed a progressive worsening of her cognition, behavior and functionality. Her neurological exam was difficult due to lack of cooperation, but it was seen an ataxic gait, Romberg sign falling backwards, slowed saccades, dysarthria, global hyperreflexia. In cognitive screening, she scored: Mini-Mental State Examination 13/30; Clock drawing test 0/5; Verbal Fluency Test by semantic category (2 animals) and by phonemic (0 "F" words). Brain MRI (2020) showed hypersignal in T2 and FLAIR in the splenium of the corpus callosum. Accentuation of cerebellar grooves and cisterns, disproportionate to the age group. The newest brain MRI (2024) showed diffuse thinning of the corpus callosum, generating ectasia of the lateral ventricles, associated with areas of low and high signal affecting its extension on FLAIR. **Discussion:** Marchiafava-Bignami disease (MBD) is a rare disorder of demyelination or necrosis of the corpus callosum and adjacent subcortical white matter. It is associated with alcoholism, malnutrition or both together, generating vitamin deficiencies, particularly vitamin B1. The course may be acute, subacute or chronic. Patients frequently show an altered mental state, which include confusion, delirium, unconsciousness, impaired memory and/or disorientation. Impaired walking, dysarthria, mutism, pyramidal signs, primitive reflexes, rigidity, incontinence, sensory symptoms, gaze palsy or diplopia, psychotic disorders and seizures were also described. Progressive dementia is the major clinical manifestation of the chronic form. The diagnosis is essentially clinical and radiological. In general, MBD is characterized by hyper-intense lesions without mass effect within the corpus callosum accompanied by swelling of the genu and splenium. In some cases, the anterior and posterior commissures, centrum semiovale and other white matter tracts may also be affected. Treatment involves replacement of B complex vitamins, in order to avoid disease progression. Some authors recommend treatment with corticosteroids, proposing that it prevents the demyelination process. **Final comments:** The current lack of knowledge about the mechanisms underlying demyelinating lesions of the corpus callosum is a barrier to establishing an effective therapy. Also, it is important to pay greater attention to alcoholic patients, in order to avoid serious nutritional deficiencies which can impact their functionality. Furthermore, new approaches for the prevention, early diagnosis and treatment of MBD are essential.

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Marchiafava-Bignami disease: a case report with favorable outcome occurred in a quaternary hospital in Southern Minas Gerais in the year 2022

Joselene de Sales Costa[1], Augusto Cesar Marrafon[1], Bruna Mendanha Reis[2], Carla Hawthorne Reis[2], Tamiris Araujo Silva[1], Franciane Couto Soares[1], Ana Clara Rodrigues Santos[1], João Batista Macedo Vianna[1], Lucas Porto Ferreira[1], Luiza Serafini Balestrassi[1]

[1]Hospital de Clínicas de Itajubá, Itajubá MG, Brazil. [2]Faculdade de Medicina de Itajubá, Itajubá MG, Brazil.

Correspondence: Joselene de Sales Costa; Email: joselenesales.c@gmail.com.

Case presentation: LAR, 50 years old, male, right-handed, diabetic, hypertensive, chronic alcoholic for 28 years, smoker, epileptic and under investigation for polyneuropathy. Admitted on 09/04/2022, appearing disoriented, with periods of psychomotor agitation interspersed with drowsiness and torpor and unable to walk. On neurological examination, he appeared drowsy, disoriented in time and space. Presence of optic ataxia and right esotropia, dysarthric. Active movement of all four limbs, overall reduced strength, MRC grade 3, heightened deep tendon reflexes, with bilateral Babinski sign. Presence of appendicular and trunk ataxia. Laboratory tests: Elevated mean corpuscular volume and mean corpuscular hemoglobin, thrombocytopenia. Head CT: Hypoattenuation throughout the corpus callosum. Magnetic resonance: Diffusion restriction throughout the corpus callosum, hyperintense lesion in the corpus callosum and bilateral subcortical

frontal region, symmetric periventricular edema, typical of Marchiafava-Bignami disease, corroborating the diagnosis. The patient received intravenous B complex and oral thiamine supplementation, presenting clinical and neurological improvement during hospitalization. On the day of hospital discharge, he was alert, with improvement in muscle strength, MRC grade 4, deep tendon reflexes alive in the upper limbs and normoactive in the lower limbs, ataxic, indifferent plantar cutaneous reflex. Still disoriented in time and space and unable to walk. He was discharged from hospital with vitamin supplementation and outpatient follow-up. After three months, at a return visit, he presented normal laboratory tests, superior cortical function and cranial nerves without changes; muscle strength grade 4+; normoactive deep tendon reflexes in the upper limbs and decreased in the lower limbs; preserved appendicular coordination; bilaterally indifferent cutaneous-plantar reflex and ataxic gait. **Discussion:** Marchiafava-Bignami syndrome results from the neurotoxicity of alcohol and deficiency of B vitamins, mainly B12. The clinical presentation varies, with reports of interhemispheric disconnection syndromes and possible synergy with Wernicke's encephalopathy, as in this case. The imaging findings corroborate the diagnosis, showing characteristic changes in the corpus callosum. Treatment typically involves B complex, thiamine and folic acid supplementation, as performed in this case. In summary, favorable results are attributed to rapid initiation of treatment, cessation of alcohol consumption, adequate nutrition. **Final comments:** Although rare, Marchiafava-Bignami syndrome is a serious and potentially fatal condition, highlighting the importance of immediate diagnostic investigation and appropriate treatment. In this case, rapid diagnosis and treatment led to a favorable outcome. Case reports like this contribute to raising awareness about this disease, the importance of early treatment, differential diagnoses and management strategies.

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Mechanical thrombectomy in an extended window for more than 24 hours: a case report

Camilla Vanessa Araújo Soares[1], Ana Luísa Castelo Branco Gomes[2], Tiago César Costa da Silva[1], Luciano Gonçalves do Nascimento Júnior[2], Felipe Nunes de Araújo[2], Gilmar Leite Pessoa Filho[2], Matheus Gurgel Saraiva[2], Juliana Magalhães Leite[2], Rafael Gonçalves[2], Alex Tiburtino Meira[3]

[1]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[2]Hospital Metropolitano Dom José Maria Pires, João Pessoa, PB, Brazil.
[3]Universidade Federal da Paraíba, Departamento de Medicina Interna, João Pessoa PB, Brazil.

Correspondence: Camilla Vanessa Araújo Soares; Email: camilla.vanessa46@gmail.com.

Case presentation: A 53-year-old male patient with a history of heart failure with decompensated reduced ejection fraction presents with right facio-brachio-crural hemiplegia, global aphasia and combined gaze deviation on the left. On admission, he had an NIHSS of 24. Head computed tomography showed a hypoattenuating area in the subcortical region of the left precentral gyrus, ASPECTS 9; and computed tomography angiography revealed occlusion of probable thrombotic nature in the proximal third of the cervical segment of the left internal carotid artery, with absence of flow in the distal cervical and intracranial segments and in the horizontal segment (M1) of the ipsilateral middle cerebral artery, in addition to the carotid web. Given the diagnosis of ischemic stroke, mechanical thrombectomy (MT) was performed 24 hours and 40 minutes after the ictus. MT was performed with total opening of the left internal carotid, left middle and anterior cerebral arteries, with total reperfusion (TICI III) of the aforementioned arteries. The patient's neurological status improved significantly after the interventions and he was discharged with an NIHSS of 0. **Discussion:** Ischemic stroke is an acute focal insult, which is characterized by impaired arterial supply to the cerebral territory. It is the second cause of morbidity and mortality in Brazil. In the absence of contraindications, treatment should be carried out with intravenous thrombolytics and MT, the latter being the standard treatment in of large vessel obstruction in the anterior circulation within 6 hours of ictus. Studies in developed countries have already shown that there is benefit in performing the procedure within an extended window of up to 24 hours, but there is still no solid evidence of benefit beyond this interval. In this case, the patient presented ASPECTS 9 even 24 hours after the ictus, which favored to MT despite exceeding the extended window time. Extending the therapeutic window is important, especially in developing countries, as it allows more patients to have access to the procedure. Considering that patients in the sistema único de saúde have less access to rehabilitation due to the long queues and that many users have difficulty accessing services within the ideal time window, it is interesting to consider the extended window in selected cases. However, it is a procedure that is prone to complications, which requires qualified professional to make decisions like this. **Final comments:** Early diagnosis is essential to determine the therapy to be initiated, whether thrombolytic or MT, with time being a crucial factor to avoid serious and irreversible neurological sequelae. However, in selected cases, MT may be beneficial if performed in an extended window. This case is relevant because of the therapeutic success of performing MT after the extended 24-hour window.

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Median nerve impairment in leprosy: how does it differ from the classic carpal tunnel syndrome?

Lucas Figueira Vieira[1], Caio Luiz Nazar Cunha[1], Ana Clara Nogueira Cezar[1], Ariane Aparecida Correa de Miranda[1], Laura Caroline Felipe de Freitas[1], Eduardo William Farinha Carvalho[1], João Paulo Moreira Fernandes[1], Pedro Henrique Sirotheau Corrêa Alves[1], Isabela Maria Bernardes Goulart[1], Diogo Fernandes dos Santos[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Lucas Figueira Vieira; Email: lucasfigueira@ufu.br.

Background: Carpal tunnel syndrome (CTS) has already been described as a possible form of neural leprosy presentation. However, the median nerve can be involved in this neuropathy in proximal segments and, sometimes, with an asymmetric impairment of the digital branches. **Objective:** To detail the pattern of median nerve impairment through a careful electroneuromyographic and ultrasound evaluation. **Methods:** This cross-sectional study comprises two groups, encompassing 15 primary neural leprosy (PNL) patients and 14 patients with CTS who underwent peripheral nerve ultrasonography and electroneuromyography evaluation. The median nerve was evaluated at the tunnel (Mt) and 4cm proximal to the carpal tunnel (Mpt). **Results:** 92.8% of patients with CTS and 80.0% of patients with PNL had bilateral impairment of the median nerve, totaling 27 nerves affected in each group. Considering the cross-sectional area (CSA) of the Mt segment, 63.0% of the nerves with CST and 74.1% of the nerves in the PNL group were found to be thickened, with an average CSA of 13.4 (± 4.4) and 12.4 (±4.6) respectively (p=0.18). The CSA of the Mpt segment showed the presence of thickening in only 3.7% in the CST group and 96.3% in the PNL group (p<0.0001), with an average of 6.6 (±1.3) and 17.0 (±6.7) respectively (p<0.001). 88.9% of the nerves in the PNL group and only 7.4% (2/27) showed a reduction in conduction velocity in the distal forearm, with an average of 41.0 (±6.3) and 53.2 (±5.2), respectively (p<0.0001). **Conclusion:** The presence of neural thickening and demyelinating impairment in the segments proximal to the carpal tunnel favors the diagnosis of leprosy.

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Memory impairment post expermental pneumococic meningitis mediated by the actvation of NLRP3 inflammasome

Gabriel de Marco[1], Amanda Christine da Silva Kursancew[2], Beatriz Brandão Lima[1], Cristiano Julio Faller[2], Diogo Dominguini[2], Fabricia Petronilho[2], Tatiana Barichello[3], Vijayasree V. Giridharan[3], Jaqueline S. Generoso[2]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]Universidade do Extremo Sul Catarinense, Laboratório de Neurologia Experimental, Programa de Graduação em Ciências da Saúde, Criciúma SC, Brazil.
[3]The University of Texas Health Science Center at Houston, McGovern Medical School, Translational Psychiatry Program, Faillace Department of Psychiatry and Behavioral Sciences, Houston, TX, USA.

Correspondence: Gabriel de Marco; Email: gabrieldemarx@gmail.com.

Bacterial meningitis is considered a life-threatening condition with high mortality rates. In response to the infection, signaling cascades, producing pro-inflammatory mediators trigger an exacerbated host immune response. Another inflammatory pathway occurs through the activation of inflammasomes. Studies highlight the role of the NLR family pyrin domain containing 3 (NLRP3) in central nervous system disorders commonly involved in neuroinfammation. We aimed to investigate the role of NLRP3 and its inhibitor MCC950 on neurochemical, immunological, and behavioral parameters in the early and late stages of experimental pneumococcal meningitis. For this, adult male Wistar rats received an intracisternal injection of Streptococcus pneumoniae or artifcial cerebrospinal fuid as a placebo. The animals were divided into control/saline, control/MCC950, meningitis/saline, and meningitis/MCC950. Immediately after the meningitis induction, the animals received 140ng/kg MCC950 via intracisternal injection. For the acute protocol, 24 h after induction, brain structures were collected to evaluate cytokines, NLRP3, and microglia. In the long-term group, the animals were submitted to open feld and recognition of new objects tests at ten days after the meningitis induction. After the behavioral tests, the same markers were evaluated. The animals in the meningitis group at 24 showed increased levels of cytokines, NLRP3, and IBA-1 expression, and the use of the MCC950 signifcantly reduced those levels. Although free from infection, ten days after meningitis induction, the animals in the meningitis group had elevated cytokine levels and demonstrated behavioral defcits; however, the single dose of NLRP3 inhibitor rescued the behavior defcits and decreased the brain infammatory profle.

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Meningeal carcinomatosis in gastric cancer: a case series and narrative review from a single center in Brazil

Gabriela de Oliveira Tavares[1], Letícia Dias Costa[1], Thales Pardini Fagundes[1]

[1]Fundação Pio XII, Hospital de Câncer de Barretos, Barretos SP, Brazil.

Correspondence: Thales Pardini Fagundes; Email: pardinithales@gmail.com.

Background: Leptomeningeal carcinomatosis (LC) is a rare and devastating complication of cancer, affecting approximately five to ten percent of patients with solid tumors, with particularly low incidence in gastric cancer (0.14 to 0.24 %). This study presents the first case series of LC in gastric cancer in Brazil, combined with a comprehensive literature review to contribute to the understanding of this condition. **Objective:** To describe the clinical course of patients with gastric cancer who developed LC through the first Brazilian case series and a literature review. To conduct a literature review on LC in gastric cancer, describe possible clinical, laboratory, and imaging findings that aid in diagnosis, and explore the best therapeutic approaches. **Methods:** This study combines the first Brazilian case series of patients diagnosed with LC from gastric cancer in a single center with a comprehensive literature review. A literature search was conducted using the Virtual Health Library (BVS) database, with no period restrictions, limited to articles in English and Spanish. Data extracted included age, sex, histological type, MRI findings, CSF analysis results, treatment regimens, survival time, and presenting symptoms. These data were combined with additional cases managed by the authors. Descriptive statistics were used to summarize the data. **Results:** Nine patients with LC from gastric cancer were analyzed, including six cases from the literature and three novel cases managed by the authors, representing the first Brazilian case series. The median age was 56 years (range: 32 to 84 years), with a predominance of females (7 out of 9). The most common histological type was adenocarcinoma (7 out of 9), followed by signet ring cell carcinoma (2 out of 9) and early-stage gastric cancer (1 out of 9). MRI findings varied, with several cases showing leptomeningeal enhancement, while others presented unremarkable or negative findings. CSF analysis frequently revealed the presence of malignant cells or adenocarcinoma, confirming the diagnosis of LC. Treatment regimens were diverse and included intrathecal chemotherapy with agents such as methotrexate, cytarabine, and prednisolone, systemic chemotherapy, radiotherapy, and targeted therapy. Three patients received no treatment. The median survival time was two months (range: 0.5 to 29 months). **Conclusion:** This study provides valuable information on the clinical characteristics, diagnostic findings, therapeutic approaches, and outcomes of patients with LC from gastric cancer, presenting the first Brazilian case series. Further research is needed to improve the management of this rare and challenging complication.

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Meningococcal disease and vaccination in the child and youth population in Brazil (2010-2022): an epidemiological study

Maria Eduarda Garcia Marvulle[1], Gabriella de Oliveira Barboza[1], Thairon Guilherme de Santana[1], Victória Maria Rabelo Lopes[1]

[1]Universidade de Marília, Marília SP, Brazil.

Correspondence: Maria Eduarda Garcia Marvulle; Email: mariaeduardamarvulle@outlook.com.

Background: Meningococcal disease, with meningitis as a common presentation, is caused by Neisseria meningitidis, with serogroup C being the most prevalent in Brazil. This disease carries significant rates of lethality and morbidity, leaving sequelae such as deafness, seizures, paralysis, and mental disability. Considering these factors and given that the population under 5 years old is the most affected, in 2010 the conjugated meningococcal C vaccine was included in the National Immunization Program, being applied at 3 and 5 months, with a booster between 12 and 15 months of age. In 2017, vaccination of adolescents aged 12 to 13 years was introduced and, in 2018, it was extended to individuals aged 11 to 14 years. Therefore, it is important to clarify the relationship between vaccination coverage and the number of deaths by meningococcal infection in recent years. **Objective:** To associate meningococcal C vaccination with the number of deaths by meningococcal infection in the Brazilian child and adolescent population, considering the period from 2010 to 2022. **Methods:** This is a cross-sectional, descriptive, and retrospective epidemiological study conducted through the analysis of data made available by the DATASUS health information database, from 2010 to 2022, selecting individuals up to 19 years of age. **Results:** The total number of deaths from meningococcal infection among individuals up to 19 years old in this period was 1,801, with approximately 52.42% of these deaths occurring in children under 4 years old. The year 2010 had the highest number of deaths from meningococcal infection, corresponding to about 18.6% of the total, and between 2010 and 2022 there was an 87.46% reduction in the number of deaths. Between 2019 and 2021, the number of deaths was reduced by 69.9%, however in 2022 this number started to increase, totaling 11 more deaths than in 2021. The number of meningitis cases, considering the same population and the same period, followed the increases and decreases in the number of deaths by meningococcal infection. **Conclusion:** According to the data analyzed, it can be inferred that, after the introduction of the conjugated meningococcal C vaccine, there was a reduction in the number of deaths from meningococcal infection and confirmed cases of meningitis. However, although the infant population is the main target of vaccination, it still represents the highest death rate. In addition, in the years most affected by the COVID-19 pandemic, there was a decrease in the number of deaths from meningococcal infection in all age groups, which may be associated with social isolation and the probable underreporting that occurred during this period. Finally, as meningococcal disease is still a relevant public health problem, especially in children and adolescents, new preventive and immunization actions aimed at this population and the strengthening of epidemiological surveillance are necessary, considering the post-pandemic scenario.

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Meningoencephalitis caused by Varicella-zoster virus complicated with cerebral venous thrombosis: presesentation in an imunocompetent patient without previous rash

Gabriel Augusto Wandekoken[1], Clara Moreira Gonzaga[1], Julia de Oliveira Xavier Ramos[1], Carolina Candeias da Silva[1], Enedina Maria Lobato de Oliveira[1], Henrique Ballalai Ferraz[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Gabriel Augusto Wandekoken; Email: gabrielwandekoken@hotmail.com.

Case presentation: A 58-year-old woman with hypertension, type 2 diabetes, dyslipidemia, and childhood epilepsy, was brought to the hospital due to a 12-day history of confusion, inattention, spatial disorientation, and incoherent speech, with an insidious onset and progressive course. Two days before admission, she developed aggressive and hypersexualized behavior, nausea and emesis. Upon admission, she had stable vital signs, no skin lesions, was agitated, disoriented and aphasic. An initial cranial CT scan showed no abnormalities. Lumbar puncture revealed an opening pressure of 43 cmH2O, cerebrospinal fluid (CSF) analysis showed 355 cells/μL (83% lymphocytes), protein of 219 mg/dL and positive PCR for Varicella-zoster virus (VZV), with other microbiological tests negative. An EEG at admission showed lateralized rhythmic delta activity and bilateral frontotemporal epileptiform discharges. Cranial MR angiography revealed cerebral venous thrombosis in the left sigmoid sinus and internal jugular vein. She was treated with intravenous acyclovir for 14 days and enoxaparin 1mg/kg bid, showing clinical and electroencephalographic improvement, and was discharged without sequelae for outpatient follow-up on anticoagulation with direct oral anticoagulant (DOAC). **Discussion:** VZV is a neurotropic herpesvirus. It causes varicella (chicken pox) as a primary infection, after which the virus remains latent in different ganglionic neurons, capable of reactivating during immunosuppression or advanced age. CNS involvement by VZV is uncommon, particularly in immunocompetent population, occurring in both primary infection and reactivation, and can manifest as diffuse encephalitis, acute cerebellar ataxia, myelitis, stroke, polyneuropathy, and aseptic meningitis. Acute diffuse encephalitis more frequently affects adults, presenting with delirium, seizures, and focal neurological signs. The most accurate diagnostic method for viral encephalitis is VZV PCR in CSF. Current data suggest VZV as an emerging cause of encephalitis in young, immunocompetent patients. Although skin lesions are a hallmark of the disease, up to 50% of reported cases of VZV neurological complications occur without cutaneous manifestations. This case features two unusual characteristics of VZV CNS involvement: varicella encephalitis not preceded by a rash, in an immunocompetent patient younger than 60 years. **Final comments:** We describe an atypical case of an immunocompetent patient with acute varicella-zoster meningoencephalitis without preceding rash, complicated by a cerebral thrombotic event. This condition is classically described more frequently in immunosuppressed individuals. However, This case highlights the importance of considering VZV in the differential diagnosis of encephalitis presentations in immunocompetents.

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Meningoencephalitis due to free-living amoebae in an immunocompetent patient: case report

Danielli Regina da Silva[1], Laisy Bárbara Souza Silva[1], Airton Sena Oliveira Sales[1], Hágda Pires Moreira Damasceno[1], Giovani Zago Borges[1], Gabriela Adami Ribeiro[1], Giovana Bertin Mira[1], Larissa Batista Xavier[1], Emerson Oliveira Milhorin[1], Mario Leon Silva Vergara[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Danielli Regina da Silva; Email: d202011051@uftm.edu.br.

Case presentation: Woman, 67 years old, mixed race and previously healthy. Admitted to the Clinical Hospital with a progressive history of visual blurring, memory changes, unestimated weight loss and paresis in the lower limbs for 2 months. A cranial magnetic resonance imaging was performed, which revealed two expansive corticosubcortical lesions in the left hemisphere, with intense and heterogeneous post-contrast enhancement, partially defined limits, with surrounding vasogenic edema, foci of calcifications and hemosiderin deposits. The initial diagnostic hypothesis proposed was oligodendroglioma. The patient underwent a surgical procedure, during which it was observed that the lesion had an inflammatory/infectious appearance. Pathological examination: no evidence of neoplasia and extensive necrotizing encephalitis with inflammatory infiltrate, xantomized macrophages and rounded, small, eosinophilic structures, with discretely basophilic nuclei suggestive of cysts and free-living amoebae trophozoites. Empirical treatment with metronidazole, trimethoprimsulfamethoxazole and fluconazole was initiated. Miltefosine and albendazole were later added. The patient's neurological condition worsened and 13 days after starting treatment, she died. **Discussion:** Free-living amoebae considered pathogenic to humans are: Naegleria fowleri, Acanthamoeba spp. and Balamuthia mandrillaris. The last two species can affect both immunocompetent

and immunocompromised individuals and are associated with subacute or chronic granulomatous meningoencephalitis with an unfavorable outcome in most cases. Meningoencephalitis caused by free-living amoebas is a rare condition, late diagnosed and lethal in more than 90% of cases, given the unavailability of effective treatment. The regimen prescribed for this patient was based on the suggestion of several authors, but with a very limited level of evidence. The clinical history and chronic evolution of this case, combined with the anatomopathological findings, support the diagnosis of granulomatous amoebic meningoencephalitis caused by Balamuthia mandrillaris. The diagnosis of this condition represents a challenge and is generally performed postmortem and is based on the histopathology of the brain lesion. Defining the amoebae species requires specific molecular, immunohistochemical and serological methods, which are only available in a few places. **Final comments:** Protozoal meningoencephalitis is rare and presents a challenging diagnosis, especially in immunocompetent patients, given its nonspecific clinical presentation. Thus, this report clearly illustrates the difficulties in early diagnosis of meningoencephalitis caused by free-living amoebae and the importance of brain biopsy for diagnostic elucidation.

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Metabolic syndrome in patients with Duchenne muscular dystrophy

Marcela Misao Miyata[1], Marcondes Cavalcante França Júnior[1], Mariana Rabelo de Brito[1], Anamarli Nucci[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Marcela Misao Miyata; Email: marcelamisao@hotmail.com.

Background: Duchenne muscular dystrophy (DMD) is the most frequent muscle dystrophy in children. It is an X-linked condition caused by loss-of-function variants in the DMD gene. During disease course, affected patients lose lean body mass, become severely motor disabled and are often treated with long term steroids. All these factors pose them at increased risk for developing metabolic syndrome (MS), even as children. Surprisingly, there are few studies addressing this aspect in DMD cohorts, particularly in Brazil. **Objective:** To estimate the frequency and risk factors for DMD-related MS in a Brazilian cohort. **Methods:** This is a cross-sectional study involving 46 patients with DMD regularly followed at Universidade Estadual de Campinas. All subjects underwent detailed clinical (Blood pressure, BMI, Motor function) and laboratorial (Serum glucose, HbA1C, cholesterol and triglyceride levels) evaluation. We also recorded demographic, genetic and steroid use (cumulative dose) data from each subject. MS was defined according to the international federation of diabetes criteria for children/teenagers. In addition to these data, we also measured the volume of each patient hypothalamus and compared it with healthy controls, using the HypAST pipeline developed at Universidade Estadual de Campinas. **Results:** mean age of patients was 11.1 years and there were 60.8% wheelchair bound. Most of them had either DMD deletions (n=26) or duplications (n=5). Four of the patients fulfilled the entire set of criteria for MS. However, particular clinical features of the syndrome were frequently found: obesity in 41.8% and 62.7% of body mass index above percentile 85, central obesity in 65.9% and increased blood pressure in 18.2%. Hb1AC was normal in all individuals and glucose was elevated in one participant. However, age-adjusted increased cholesterol and/or triglyceride levels were noticed in 65.2% of the patients. No significant differences were found when comparing the volumes of the hypothalamus between patients and controls. **Conclusion:** metabolic abnormalities are frequent and should be actively investigated in DMD. The existing MS diagnostic criteria may not be adequate for these patients and contribute to underdiagnosis.

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Methodological quality of experimental studies with cannabinoids in Alzheimer's disease: a systematic review.

Maria Eduarda Kobayashi Teixeira[1], Maria Antônia Vianna Santos[1], João Lucas Lima Diogo[1], Ana Clara Costa de Siqueira Campos[1], Ruben Costa Santos Neto[1], Lara Millena Miranda Frota[1], Thiago de Faria Junqueira[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Maria Eduarda Kobayashi Teixeira; Email: mariateixeira20.1@bahiana.edu.br.

Background: Alzheimer's disease (AD) is a progressive neurodegenerative disease that results in a significant decline in cognitive, mnemonic and behavioral functions, leading to chronic impairment of patients' quality of life. However, currently available pharmacological treatments, such as acetylcholinesterase inhibitors (e.g. Rivastigmine) and glutamatergic drugs (e.g. Memantine) - which have been previously validated in experimental models - are still not capable of stopping the functional deficits of the disease. Experimental models, in turn, are crucial for the development and discovery of new therapeutic approaches. Therefore, exploring the differences between experimental and clinical results in the treatment of AD with cannabinoids can clarify and generate new hypotheses, expanding investigations into the medicinal use of this class of substances. **Objective:** To evaluate the methodological quality of experimental studies on Alzheimer's Disease with rodents, which used cannabinoids as a therapeutic intervention. **Methods:** The systematic review followed the guidelines of the PRISMA Protocol. The searches were carried out on the MEDLINE/Pubmed, Scielo and LILACS platforms, using the descriptors ("NAME OF THE EXPERIMENTAL MODEL WITH RODENTS") AND ("CANNABINOIDS"). Information was extracted from the articles about the type of experimental model, the therapeutic intervention with cannabinoids and the assessment of cognitive deficits in rodents. The ARRIVE Protocol was applied to assess methodological quality and the SYRCLE's RoB Tool to check the risk of bias. **Results:** After applying the inclusion and exclusion criteria, five articles were included in the present review: two used 5xFAD transgenic mice and the others used APP/PS1 mice. The article by Wang et. al stood out from the others for being the only one with moderate methodological quality (with the highest score assigned) and low risk of bias, however the main clinical outcome tested was spatial memory, without other cognitive implications. Researchers have applied cognitive tests in water mazes or open spaces and some studies included immunohistochemical analysis of mouse tissues. It was also possible to identify important differences in cannabinoid dosages, which ranged from 0.002 to 10mg/kg. **Conclusion:** The systematic review revealed that the methodological quality of experimental studies with cannabinoids for the treatment of Alzheimer's disease is unsatisfactory. None of the studies analyzed achieved a good assessment in the ARRIVE Protocol and only one was classified as low risk of bias by SYRCLE's ROB. Furthermore, the studies presented significant methodological heterogeneity. Therefore, given the results of this review, the use of cannabinoids to treat Alzheimer's in experimental studies with rodents is still a topic that needs to be explored in the scientific literature.

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Metronidazole intoxication. A important differential diagnosis of dizziness and encephalopathy in the elderly: a case report

Rafael Bragança Rodrigues Matias[1], Eduardo Mesquita[1], Ana Luisa Vieira de Faria Donadon[1], Amanda Miranda de Mendonça[1], Alex Araujo Fernandes[1], Matheus Luís Braga Munareto[1], Alex Machado Baeta[1], Hennan Salzedas Teixeira[1], Matheus Alves da Silva[1]

[1]Beneficiência Portuguesa, São Paulo SP, Brazil.

Correspondence: Rafael Braganca Rodrigues Matias; Email: rafaelwaa@gmail.com.

Case presentation: An 87 year old man presented to the hospital for evaluation of a traumatic brain injury due to a fall secondary to the worsening of a previous chronic dizziness and consciousness impairment. He had advanced Alzheimer disease, high grade atrioventricular block, systemic arterial hypertension and chronic pulmonary obstructive disease. He had a diagnosis of pseudomembranous colitis in the last month, and was using oral metronidazole and vancomycin for two weeks before the hospital evaluation. His neurological examination showed bilateral dysmetria and dysdiadochokinesia. Due to the cerebellar features, a magnetic resonance imaging was performed and showed bilateral hypersignal in dentate nucleus, inferior colliculus, subthalamic nucleus and in the splenium of the callosum body, compatible with metronidazole intoxication. He had a neurological improvement after metronidazole suspension. **Discussion:** Metronidazole-induced neurotoxicity is a rare adverse effect that can lead to cerebellar ataxia, dysarthria, and peripheral neuropathy. These symptoms may arise from either short- or long-term metronidazole use. The exact mechanism of neurotoxicity remains unclear, but neuroimaging plays a crucial role in diagnosis. To reach an accurate diagnosis, clinicians should maintain a high clinical suspicion, and sometimes brain help in the differential diagnosis and to identify specific abnormalities. Withdrawing metronidazole promptly is necessary to avoid additional morbidity. Discontinuation of the offending agent is currently the only effective treatment. Early referral to rehabilitation services is essential for optimizing functional outcomes in affected patients. **Conclusion:** This case highlights the importance of a right history when evaluating the elderly, because sometimes, unlike our case, the magnetic resonance imaging may not show the classic abnormalities and the diagnosis may be missed. A good clinical history, with neuroimaging as a complementary exam, is fundamental in theses cases.

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Microdeletion syndrome 8q22.2q23.3: a rare case in Brazil.

Beatriz Medeiros Correa[1], Paulo Afonso Ortega[1], Marcio Melegaro[1], Elisangela Melegaro[1], Walter Arayjo[1], Jemerson Silva[1], Maria Fernanda Pontelli[1], Khadige Arabi[1]

[1]Faculdade de Medicina Unoeste, Guarujá SP, Brazil.

Correspondence: Beatriz Medeiros Corrêa; Email: beamedneuro@gmail.com.

Case presentation: This case report describes a pediatric patient referred for evaluation due to Neuropsychomotor Developmental Delay (NPMD) and pondero-statural deficit, resulting from a rare genetic condition due to microdeletion 8q22.2q23.3. **Discussion:** Female patient, was referred to the pediatrics department for evaluation of NPMD and pondero-statural deficit. The gestational history did not reveal significant complications, with normal growth until the last trimester of gestation, when ultrasound identified the presence of diaphragmatic hernia. Born via cesarean section at term (39 weeks), weighing 2570g, with a length of 45cm, head circumference of 33cm, and chest circumference of 31cm. At birth, the patient had respiratory difficulty, requiring immediate ventilatory support and was transferred to the neonatal ICU without further complications. Neuropsychomotor development revealed early signs of hypotonia and delay in motor milestones, standing with support only at 22 months, without

ambulating or babbling words, and delayed dentition with anomalous dental eruption. The patient had an evaluated height of 79 cm (below the 3rd percentile), weight of 8255 g (below the 3rd percentile), head circumference of 45 cm (between the 10th and 25th percentiles), with a broad forehead, hypertelorism, alternating divergent strabismus, short palpebral fissures with epicantus, global hypotonia, preserved muscular strength, and severe intellectual deficit. Complementary exams revealed, Blood karyotype (20 cells): 46, XX, t(7;8) (p15;q22) - presence of apparently balanced translocation involving chromosomes 7 and 8; SNP-Array: presence of chromosomal deletion 8q22.2q23.3 and duplication 15q26.3; Urinary screening for inborn errors of metabolism: within normal limits; TSH: within normal limits. **Final comments:** Clinical examination associated with complementary exams indicated a diagnosis of chromosomopathy due to chromosomal deletion 8q22.2q23.3. A review of the phenotype-genotype correlation of 8q22.2-q23.3 was seen in seven other cases in the literature showing presence of cleft palate or facial, abnormal intrauterine growth, epilepsy, trichorhinophalangeal syndrome, cryptorchidism or micropenis which differ from the presented clinical picture. Parental exams did not reveal the presence of 8q microdeletion, indicating a low risk of recurrence for future pregnancies. There is no specific treatment for this condition, but rehabilitation is a valuable tool for the development of affected children.

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Migraine and other headache syndromes in Brazil: an epidemiological analysis over the last 5 years

Caroline Patrícia Amaral Costa[1], Matheus Prado dos Santos[2], Fernando Silva Galvão[1], Daniel Lucas Araujo Ramos[1], Rafael Hendrick Freitas de Paula[2], Luan Nascimento Pereira de Amorim[2]

[1]Universidade Federal do Pará, Belém PA, Brazil.
[2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Caroline Patrícia Amaral Costa; Email: caroline.costa@ics.ufpa.br.

Background: Migraine is one of the most common neurological conditions in the population, significantly impacting the quality of life of those affected. In Brazil, the types of headache syndromes vary epidemiologically, affecting health policies and resource allocation. Understanding the epidemiological differences between regions to identify distribution profiles is essential for developing strategies to control the current scenario of headache syndromes in the country. **Objective:** To analyze the epidemiological profile of patients affected by migraine and other headache syndromes in Brazil between 2019 and 2023. **Methods:** This is an ecological, cross-sectional, and descriptive study with a quantitative approach, based on secondary data from the Department of Informatics of the Unified Health System (DATASUS), recorded from 2019 to 2023 in Brazil. The following variables were analyzed: region, deaths per year, mortality rate, gender, ethnicity, and age group. **Results:** In 2019, there were 11,996 hospitalizations, decreasing to 8,778 in 2020 and 8,505 in 2021. They then increased to 10,293 in 2022 and 11,697 in 2023, totaling 51,269 over the period. The Northeast led with 15,864 hospitalizations, followed by the Southeast (14,946). The most affected age group was 30-39 years old (9,295). In the Northeast, hospitalizations were concentrated in the 30-39 age group (2,898). In the Southeast, the 30-39 age group was prominent (2,866). In the South, there was a peak in the 30-39 age group (2,249). Hospitalizations were predominantly among women, with a total of 33,584 cases, compared to 17,685 cases in men. Black individuals led with 22,600 hospitalizations, followed by whites with 19,398. Regarding deaths, there were 79 in 2019, rising to 71 in 2020 and 113 in 2021, stabilizing at 112 in both 2022 and 2023, totaling 487 deaths. The Northeast region recorded the highest number of deaths during the period (257). The average mortality rate was 0.95. The majority of deaths occurred among individuals aged 60-69. In the Northeast, deaths predominantly occurred among black individuals, with 204 cases. In the Southeast, the majority were white, with 24 cases, and in the South, whites predominated, with 33 deaths. In the North, there were 113 deaths among black individuals, and in the Midwest, there were 6 deaths. **Conclusion:** The study concludes that there was significant variation in the number of hospitalizations during the observed period, while the number of deaths increased steadily over the last two years. Additionally, the Northeast region accounted for the majority of hospitalizations and deaths resulting from the condition. In terms of race, black individuals led in the number of hospitalizations and deaths, and in terms of gender, females were the most affected. These data highlight the need for policies aimed at the treatment and prophylaxis of migraines and other headaches, especially in the Northeast.

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Migraine with aura and paradoxical embolism: case report

Assis Porfirio Furtado Nogueira[1], Tulio Marlus Castro Lucena[1], Vitor Gustavo Souto Leão[2], Mariana Cota Bastos[2], Renato Maciel Freire de Paula e Silva[3], Letícia Januzi de Almeida Rocha[2], Maria Júlia Tabosa de Carvalho Galvão[2], Geovane Brito Affonso[2], Érika Santos Machado[1], Alden Rodrigues de Albuquerque[1]

[1]Hospital Geral de Alagoas, Maceió AL, Brazil.
[2]Universidade Federal de Alagoas, Maceió AL, Brazil.
[3]Hospital Universitário Oswaldo Cruz Recife PE, Brazil.

Correspondence: Túlio Marlus Castro Lucena; Email: tuliomarlus.neuro@gmail.com.

Case presentation: A 29-year-old female patient, a smoker (1 pack/day) with a history of migraine with visual aura since the age of 15, was admitted to an emergency service with a sudden onset of dysarthria and facial asymmetry associated with intense headache, followed within a few hours by complete right-sided hemiparesis. She was admitted with 11 points on the NIHSS (National Institute of Health Stroke Scale) and underwent a cranial Computed Tomography (CT) scan, which revealed hypodense lesions in the right frontal, insular, and nucleocapsular regions, suggesting an acute ischemic event. Cranial CT angiography identified areas of narrowing and luminal irregularities in the M2 branch of the left middle cerebral artery. Further etiological investigation for ischemic stroke (IS) in a young patient included: Carotid and Vertebral Doppler (CVD) without evidence of stenosis; Transthoracic Echocardiogram showing regular rhythm, normal left atrial volume, 61% ejection fraction, and the presence of an aneurysmal interatrial septum; and Transcranial Doppler with microbubble test revealing the presence of a right-to-left shunt. The patient is awaiting scheduling for a transesophageal echocardiogram and 24-hour Holter monitoring. She remains on prophylaxis with Aspirin 100 mg and Simvastatin 40 mg. **Discussion:** Stroke is one of the leading causes of death and acquired disability worldwide. Brazil has the fourth highest mortality rate from this condition among Latin American countries, with an incidence of 150 cases per 100,000 inhabitants. Migraine with aura is a well-established risk factor for ischemic stroke and constitutes an important differential diagnosis in emergency settings. The exact pathophysiological mechanism for this association remains uncertain, but the main hypotheses include oligemia, vasospasm, endothelial dysfunction, genetic predisposition, paradoxical embolism, among others. Some meta-analyses have found that people with migraine with aura have approximately double the risk of having a stroke compared to those without migraine or those with migraine without aura. Some studies reaffirm this association and highlight that the risk is particularly high in young women, especially those who smoke or use oral contraceptives, as well as its association with right-to-left cardiac shunts, a finding commonly related to patent foramen ovale (PFO). **Final comments:** The association between migraine with aura and ischemic stroke should be commonly addressed among healthcare professionals and the general public. The described case supports these findings and reinforces the importance of this type of headache as an established risk factor for cerebral ischemia, as well as highlighting the relevance of PFO research in this group.

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Mild cognitive impairment and multiple sclerosis: a shift in cognitive disturbances

Caio Hussid de Góes[1], Gabriella Chinaglia Zanin[1], Diego Olympio Peixoto Lopes[2], Maria Tereza Martinez[3], Fernanda Herculano[4], Diogo Haddad Santos[4]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Universidade Santo Amaro, São Paulo SP, Brazil.
[3]CP Quali.
[4]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Caio Hussid de Góes; Email: caio.goes@aluno.fcmsantacasasp.edu.br.

Multiple sclerosis (MS) is the most common demyelinating disorder, and typically manifests at a highly productive period of life. Common findings among patients is progressive fatigue and cognitive function impairment, severely impacting quality of life and economic burden of disease. MS cognitive impact on several domains, mainly information processing speed and learning disability, differ from the patterns present in dementias such as Alzheimer, though clinical presentation of both syndromes may overlap at later stages of MS. While studies have been carried out on older patients (above 50 years), no study was found focusing on younger populations and the transitional stage between normal cognition and Alzheimer, denominated mild cognitive impairment (MCI). The study's objective is to evaluate the risk of a patient below 50 years receiving an MCI diagnosis within 10 years of a MS diagnosis in a Brazilian population. An electronic medical record aggregator called TriNetX was used to anonymously obtain the patient data used in the study, using exclusively hospital related records. The Brazilian cohort was built using a Brazilian-only database via selecting patients who received the MS diagnosis in the last 10 years time frame, had never received an MCI or MS diagnosis before, and have less than 50 years. The MS diagnosis was considered the index event, while the outcome was a MCI diagnosis within 10 years of the index event. Risk was obtained by calculating the fraction of patients with the selected outcome from the cohort's total. Test results were a cohort of N=2,220, of whom 10 patients had the selected outcome, with a risk of 0.45% in 10 years. The mean age of the cohort was 37 years, with standard deviation of 8 years and median follow-up was 455 days. While some studies in older age groups suggest a higher cognitive impairment and risk of dementia in later stages of MS, the data gathered in the study shows a low risk of MCI in young patients with MS, specially when compared to the expected 30 to 70% of cognitive impairment in early stages of the disease. Rather, this dual pattern of cognitive disturbances may corroborate a shift over time in the underlying pathology of the cognitive impairment.

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Miller Fischer syndrome post dengue fever: a case report

Isabela da Costa Rodrigues[1], Matheus Zago[1], Ivo Marcos Darella Lorenzin Fernandes Neto[2], Eduarda Basso Badalotti[2], Angelo Marcelo Wosniacki Filho[2], Gabriel Angelo Garute Zenatti[2], Renato Endler Iachinski[2], Antonio Carlos de Andrade Soares[2]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Fundação Hospitalar São Lucas, Cascavel PR, Brazil.

Correspondence: Isabela da Costa Rodrigues; Email: isacostarod.2001@gmail.com.

Case presentation: Female, 29 years old, manifesting ascending and progressive paraparesis on lower limbs, starting on feet, evolving with paresthesia, paresis and paresthesia on upper limbs, peripheral facial paralysis on the right and transfer dysphagia. The patient was diagnosed with dengue fever 2 weeks before the onset of symptoms. Physical examination: peripheral facial paralysis on the right, tetraparesis (4-/5) and symmetrical global hyporeflexia. Skull CT with no alterations and lumbar puncture revealed hyperproteinorrachia with protein-cytological dissociation on CFS, suggesting Guillain-Barré syndrome. Terefore, 25 vials of immunoglobulin, 5g/vial, were started for 5 days. During hospitalization, the patient developed bilateral peripheral facial paralysis. After finishing the immunoglobulin infusion, there was partial improvement in paraparesis (4+/5) and facial diplegia. **Discussion:** Miller Fisher syndrome (MFS), considered a multifocal neuropathy, presents with the triad: ataxia, ophthalmoplegia and areflexia. However, there are many clinical variations for this pathology, with involvement of the facial nerve being the most common. This syndrome is a benign variant of Guillain-Barré syndrome (GBS), generally associated with previous gastrointestinal or respiratory tract infections and presenting proteino-cytological dissociation in the cerebrospinal fluid as the diagnostic criterion. There are some reports in the literature about the association of GBS and MFS with arboviruses such as Dengue and Chikungunya, especially in epidemic regions. This disease is treated in the same way as GBS, with intravenous immunoglobulin (IVIg) or, alternatively, plasmapheresis. As it is a benign condition, patients generally progress well after treatment, with complications or sequelae of the disease being rare. **Final comments:** Being a rare variant of GBS, MFS usually appears after gastrointestinal or respiratory tract infections. The case discussed demonstrates the emergence of MFS after infection with an arbovirus very common in tropical regions, Dengue. As this is a rare association, there are few similar reports in the literature with bilateral peripheral facial paralysis associated. Therefore, it is essential to record them when they arise.

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Miller-Fisher syndrome: a case report

Maria Carolina Marçon Barroso[1], Laura Alonso Matheus Montouro[1], Cibele Alexandra Ferro[1], Guilherme Guimarães Coelho[1], Thaís Delamagna Bordonal[1], Marcus Vinicius Rodrigues de Mello Junior[1], Juliana Arantes Calil[1], Marina de Carvalho Bofo[1]

[1]Faculdade de Medicina de Catanduva, Catanduva SP, Brazil.

Correspondence: Maria Carolina Marçon Barroso; Email: barrosomariacarolina@gmail.com.

Case presentation: A 35-year-old male patient admitted to the emergency department due to symmetrical flaccid paraparesis, hypo/arreflexia, symmetrical and ascending after a history of airway infection, as well as ophthalmoplegia, ataxia and generalized arreflexia. On physical examination, he presented with symmetrical flaccid tetraparesis, worse in the lower limbs, global areflexia and dysphonia, associated with cerebelar symptoms. CSF was collected and showed slight protein-cytological dissociation. During hospitalization, immunoglobulin was indicated and then was a progressive clinical improvement. Even without an antibody test, the diagnosis was Miller-Fisher Syndrome, due to the clinical presentation and the improvement after immunoglobulin. **Discussion:** Miller-Fisher syndrome (MFS) is a variant of Guillain-Barré syndrome (GBS), an acute autoimmune inflammatory demyelinating polyradiculopathy. This variant is characterized by at least 2 of this classic triad: ataxia, ophthalmoplegia and areflexia. The pathophysiology involves autoimmune mechanisms that can be triggered by external factors, such as respiratory or gastrointestinal infections, by attack to gangliosides. The presence of IgG anti-GQ1b antiganglioside antibodies in the acute phase of the disease is a marker of MFS, but its value decreases with the duration of the disease. Just as the distribution of GQ1b is not homogeneous. Both factors can explain the seronegativity of 10 to 30% of cases. The Brighton criteria uses clinical history, physical examination, laboratory and neuroimaging for diagnosis and does not include this antibody. In the case described, the characteristic clinical picture of the disease, albuminocytological dissociation in the CSF, normal brain and spine MRI allowed the diagnosis. Anti-GQ1b antibody test is unavailable at the service. It is important to highlight that investigation through anti-GQ1b antibody research may be useful in suspected cases of Miller-Fisher syndrome with atypical evolution or unusual clinical manifestations, given that it is a disease that can be treated with the use of plasmapheresis and/or human immunoglobulin. Therefore, the possibility of including the anti-GQ1b antibody test in the tests offered by the Brazilian public health system for selected cases should be considered and evaluated, improving the prognosis and lowering the long-term cost for the system. **Final comments:** The diagnosis of typical MFS is made based on clinical findings associated with CSF analysis and exclusion of other probable causes. The dosage of Anti-GQ1b should be reserved for atypical cases and the incorporation of this diagnostic tool into the Brazilian public health system for these cases should be discussed.

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Minimally invasive treatment of intraventricular hemorrhage

Marco Antônio Delazeri[1], Eduarda Rosa Fernandes[1], Giuseppe Dick Bonato[1], Carollina Danezi Felin[1], Carolina Heck Haubrich[1], William Alves Martins[1], Eliseu Paglioli[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Marco Antônio Delazeri; Email: marco.delazeri@outlook.com.

Case presentation: A 77 year-old female is brought to the emergency unit due to left hemiparesis and hemispacial neglect. Previous medical history include hypothyroidism, cardiac arrhythmia and arterial hypertension, and she was regularly taking edoxaban, metoprolol, amlodipine and methimazole. Her symptoms started 3 hours before admission and she had no history of traumatic injury. A brain computed tomography promptly showed extensive right thalamic hematoma associated with vasogenic edema and loss of telencephalic sulci-gyri differentiation, as well as extensive right lateral, third and fourth ventricle hemorrhage and subdural bleeding. There was mass effect leading to lateral ventriculus compression and 0.7 midline deviation, besides hydrocephalus and intracranial hypertension signs. Angiography showed no aneurysm, arterial occlusion or venous thrombosis. At first it was opted for conservative intensive care, and during the following hours she began to show profuse vomits and severe consciousness impairment (Glasgow Coma Scale of 3), undergoing intubation. New computered tomography showed a total bleeding volume increase. The three following days, imaging showed no signs of new bleeding and she had no awakening despite sedatives discontinuation. Continuous electroencephalographic monitoring showed six epileptic episodes beginning at left frontocentral region, during which it was evident right hand automatisms and tonic posturing. Seizures were controlled with levetiracetam, clobazam and phenytoin. Due to no bleeding absorption evidence and lack of consciousness improvement despite anti-epileptic treatment, she underwent external ventriculostomy, with hematoma drainage associated with intraventricular alteplase infusion (4mg three times a day for 3 days). She began to recover consciousness as hematoma and intraventricular hemorrhage resolved. Nowadays, after 4 years of the procedure, she still presents thalamic pain, left hemiparesis and major depression. **Discussion:** A role of minimally invasive surgery has demonstrated significant benefits, sometimes even superior depending on the type of intracerebral hemorrhage: hematoma volume, lobar or ganglionic location, time from hospital arrival to procedure, midline shift, vasogenic edema, and hemoventricle, among others. The standard procedure involves placing an external ventricular catheter guided by CT scan, clot aspiration, and, if necessary, infusion of thrombolytics for clot dissolution. Thrombolytics used mostly include alteplase (rtPA) and urokinase (uPA). The major complication is aspiration with immediate risk of rebleeding. **Final comments:** We present a case in which consciousness impairment had potentially multiple hemorrhagic causes and only resolved after invasive treatment. Minimally invasive neurosurgical procedures in similar cases are not standard practice, although they show potential to help bleeding resolution and quicken recovery, while diminishing long-term sequelae.

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Misdiagnosis in multiple sclerosis (MS): the influence of the first doctor's specialty at the disease onset

Nara Samara Silva dos Santos[1], Alex de Cerqueira Silveira Figueiredo[1], Gabriel Souza Medrado Nunes[1], Maria Eduarda Messias Vassoler[1], Milena Fernandes de Oliveira[1], Júlia Xavier Oliveira[1], Luana Araujo Passos[1], Caio Fontes do Nascimento[1], Bruno Bacellar Pedreira[2], Pedro Antonio Pereira de Jesus[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Universidade Federal da Bahia, Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Nara Samara Silva dos Santos; Email: nara.samara@ufba.br.

Background: Multiple sclerosis (MS) is a chronic autoimmune condition that affects the central nervous system with variable clinical presentation both in terms of neurological functions affected and the intensity of disability generated. These characteristics create a barrier that leads to difficulty in recognizing, suspecting, and confirming the diagnosis of MS, resulting in misdiagnosis, a common and potentially dangerous condition, as it delays the start of disease-modifying therapies and increases the possibility of worse outcomes. **Objective:** To investigate the relation between misdiagnosis and the first doctor's specialty at disease onset in MS patients. **Methods:** This cross-sectional study was conducted with patients diagnosed with MS according to the McDonald criteria, from July/2021 to May/2024. Clinical and demographic data were collected through interviews in a reference center in Bahia, Brazil. Binary logistic regression was used to identify the association between misdiagnosis and the first doctor's specialty. We adjusted the model for other variables such as ethnicity, comorbidities, region of residence and cranial nerve alterations, and motor and sensitive symptoms at onset. **Results:** A total of 313 individuals were selected, of whom 216 (69.0%) were female. The median age at disease onset was 31 (22-39) and at admission to the study was 40 (30-48). Of the patients analyzed, 113 (36.1%)

received an alternative diagnosis before MS; of these, 86 (76.1%) had a non-neurologist as their first doctor while of those without misdiagnosis (200), 122 (61.0%) went to a non-neurologist (p=0.009). Regarding past misdiagnosis, the most prevalent were stroke (21.2%) and labyrinthitis (19,5%); other atypical and less prevalent alternative diagnoses were depression, stress, and flu. In the adjusted model analysis, the specialty of the first doctor as a non-neurologist (OR 1.98 [95% CI 1.14-3.42]) was the factor related to the presence of erroneous previous diagnoses (p=0.015), while ethnicity of African descent (OR 0.49 [95% CI 0.27-0.86]) was associated to less misdiagnosis (p=0.014). **Conclusion:** Misdiagnosis remains an important barrier in the patient's journey to the definitive MS diagnosis in our population. Most of the patients who received previous alternative diagnoses looked for a non-neurologist as their first doctor and this factor could be associated with misdiagnosis in MS. These results highlight a possible worrying gap in knowledge about the presentations of this disease and the importance of addressing it during medical education, especially given the increasing global prevalence of MS.

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Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) - like syndrome associated with polg gene mutations: a case report

Rhanice Soares Huguenin Câmara[1], Roberto Pereira Santos[1], Filipi Brito[1], Ana Carolina Leite[1], Barbara Pina Aiello[1], Matheus Jorand Cecatto[1], Amanda de Morais Affonso[1], Caio Tardin[1], Victor Evangelista Rodrigues Pereira[1], Marcos Raimundo Gomes de Freitas[1]

[1]Universidade Federal do Rio de Janeirotal, Hospital Universitário Clementino Fraga Filho, Serviço de Neurologia, Rio de Janeiro RJ, Brazil.

Correspondence: Roberto Pereira Santos; Email: r.santos.ufrj@gmail.com.

Case presentation: Mitochondrial neurogastrointestinal encephalomyopathy (MNGIE) arises from mutations in the gene encoding thymidine phosphorylase (TP), a mitochondrial enzyme. This case involves a 55-year-old woman with lifelong gastrointestinal issues, including abdominal pain, intestinal obstruction, nausea, dysphagia, early satiety, and vomiting. Concurrently, she developed progressive muscular weakness, eventually leading to inability to walk independently. Despite extensive medical consultations and investigations spanning 55 years, no definitive diagnosis was reached. Physical examination revealed intolerance to walking, proximal weakness in all limbs with predominant proximal involvement, generalized areflexia, and distal hypoesthesia across all sensory modalities. Her parents were not consanguineous, and her sibling reported a similar but asymptomatic condition. The clinical presentation aligned with MNGIE syndrome criteria (Hirano, 2021). Exome sequencing identified a pathogenic variant in the POLG gene, c.752C>T (p.Thr251Ile), and a variant of uncertain significance, c.752C>T (p.Pro587Leu), confirming MNGIE syndrome with a novel association with POLG mutation. **Discussion:** MNGIE results from mutations in the TYMP gene, impairing TP function and causing thymidine (dThd) and deoxyuridine (dUrd) accumulation, leading to mitochondrial DNA (mtDNA) abnormalities. Onset typically occurs around 17.9 years, with gastrointestinal symptoms manifesting early and progressing to severe complications like diverticulosis, diverticulitis, and hepatopathy. Neurological symptoms include ocular signs, polyneuropathy, hearing loss, and leucoencephalopathy, with cognitive impairment in some cases. The disease is progressive, with a median survival of 35-37 years, primarily due to GI and liver complications exacerbated by cachexia. **Final comments:** This case suggests a novel MNGIE-like phenotype associated with a POLG gene mutation, highlighting the complexity and diversity of mitochondrial disorders. Further research is crucial to elucidate the underlying mechanisms and refine diagnostic and therapeutic strategies for such rare genetic conditions.

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Modeling dendritic integration with non-linear dynamics and eigenmodes

Jose Reynaldo Walther de Almeida[1], Thalles Olivi de Almeida[2]

[1]Instituto de Neurociencias de Araraquara, Araraquara SP, Brazil.
[2]Hospital Sirio Libanes, São Paulo SP, Brazil.

Correspondence: Thalles Olivi de Almeida; Email: almeida.thalles@outlook.com.

Background: Dendritic integration is crucial in neuronal function, where excitatory and inhibitory signals are non-linearly summed to determine the generation of an action potential. Besides synaptic interactions, external electric fields also modulate neuronal activity. This study presents an extended differential equation that includes eigenmodes, providing detailed modeling of dendritic integration. **Objectives:** Develop and test a differential equation that predicts the generation or inhibition of action potentials in the axon, considering non-linearity and the effects of electric fields and eigenmodes under different stimulus conditions. **Methods:** The differential equation used is: CmdVmdt=−[gNa(Vm−ENa)+gK(Vm−EK)+gCa(Vm−ECa)]+∑igsyn,i(Vm−Esyn,i)+ extVm+α(Iion·Isyn)+βIext1+e−VmC_m frac{dV_m}{dt} = -left[g_{Na}(V_m - E_{Na}) + g_{K}(V_m - E_{K}) + g_{Ca}(V_m - E_{Ca}) ight] + sum_{i} g_{syn,i}(V_m - E_{syn,i}) + epsilon_{ext} V_m + alpha (I_{ion} cdot I_{syn}) + eta frac{I_{ext}}{1 + e^{-V_m}} CmdtdVm=−[gNa(Vm−ENa)+gK(Vm−EK)+gCa(Vm−ECa)]+∑igsyn,i(Vm−Esyn,i)+ extVm+α(Iion·Isyn)+β1+e−VmIext The equation was numerically solved using the Euler method. Three conditions were simulated: increased excitatory synaptic conductance, increased inhibitory synaptic conductance, and increased influence of external electric fields. **Results:** Simulations under three distinct conditions showed the following findings: 1. Excitatory Synaptic Conductance: A moderate increase resulted in a rapid and pronounced depolarization, bringing the membrane potential closer to the firing threshold but still sub-threshold. 2. Inhibitory Synaptic Conductance: Increased inhibition caused continuous hyperpolarization, moving the membrane potential away from the firing threshold and stabilizing the cell. 3. External Electric Fields: Enhanced influence of electric fields provided moderate depolarization, indicating that while electric fields have significant influence, it is still controlled and insufficient to trigger an action potential. **Conclusion:** The results demonstrate that membrane potential depolarization is strongly influenced by changes in stimulus parameters. The inclusion of eigenmodes in the differential equation provides detailed and precise modeling of dendritic integration. Simulations showed how different stimulus conditions affect membrane potential depolarization. These results help better understand neuronal excitability modulation and open avenues for practical applications in theoretical neuroscience, including memory studies, synaptopathies, and other areas.

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Modifiable risk factors in dementia: literature review

Kathleen Torres Tenorio Monteiro[1], Maria Lucia Veluttini Pimentel[1], Ricardo Novis[1], Leticia Fezer[1], Filipe Freitas[1], Raquel Molina Costa[1], Barbara Sampaio de Souza Dias[1], Rodrigo Moreira Pereira[1], Gabriel Rivadavia[1], Cicera Simplicio Machado Leandro[1]

[1]Santa Casa de Misericórdia do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Kathleen Torres Tenório Monteiro; Email: kathleen_tenorio@hotmail.com.

Background: Dementia is a general term used to describe a group of conditions that affect brain function through neuronal loss, leading to a progressive decline in memory, thinking, behavior and the ability to perform daily activities. In the world, around 57 million people live with dementia, and this number is expected to increase to 153 million by 2050 due to global aging, increasing especially in low- and middle-income countries, where the predicted increase in the number of cases is greater. due to the low sociodemographic index. The risk of developing cognitive decline is influenced by a combination of genetic and environmental factors. Although some risk factors that negatively influence cognitive reserve cannot be modified, such as age and genetic predisposition; identifying and addressing modifiable risk factors can help reduce the likelihood of developing dementia or slow its progressain. **Methods:** Systematic review based on applied research in the Pubmed, LILACS, Scielo and Uptodate databases. We selected 203 articles, of which 34 articles were included in this study. The inclusion criteria were articles published and indexed in the aforementioned databases from 2013 to nowaday that addressed the relationship between dementia and modifiable risk factors, in patients who started with cognitive impairment. **Results:** Modifiable risk factors are responsible for around 40% of dementias worldwide, which could theoretically be avoided or delayed. Adopting a healthy lifestyle, with a balanced diet, regular physical activity and control of chronic diseases, is essential in preventing dementia. **Conclusion:** Prevention has to do with public politics through individually adapted health programs and population-based strategies. People in good physical health can sustain a greater burden of neuropathology without experiencing cognitive impairment. It is important to investigate factors that may be protective and delay its development.

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Möebius-Polland syndrome associated to epilepsy

Caroline Schiochet Verza[1], David Guillermo Carriazo Galindo[1], Lucas Porcello Schilling[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Caroline Schiochet Verza; Email: carolinesverza@gmail.com.

Case presentation: A 24-year-old male presented to our epilepsy clinic with recurrent spells of loss of conscience. Past medical history revealed a diagnosis of Möebius syndrome, developmental delay, mild cognitive impairment and recurrent suicidal thoughts. Seizures had begun at 8 eight years-old and never remised. Semiology was diverse and multiple seizure types were suspected, including dyscognitive spells, tonic upper limps seizures and tonic-clonic seizures. Neurological examination revealed bilateral facial and VI nerve palsies, along with right great pectoralis atrophy and tonge fasciculations. At this moment, we initiated valproic acid up to 1500mg a day. This was followed by a significant improvement in seizure rate. Brain MRI demonstrated a right hippocampal atrophy associated to straightening of floor of the 4th ventricule and loss of facial colliculus bilaterally. Electroencephalogram sowed only diffuse disorganization. A further increase in valproic acid rendered seizure-free. **Discussion:** Möebius syndrome is a congenital

disorder characterized by total or partial absence of VI and VII cranial nuclei, leading to an 'expressionless' face and horizontal gaze paralysis. It is estimated to occur 1 in every 100.000 births. This disorder may be caused by several different etiologies, including some medications and congenital infections. About to 90% of patients present normal neurodevelopment, and most of disease symptoms are present at birth. However, rarely it may be associated to other features. Here, we present a case of Möebius-Polland syndrome associated to focal epilepsy. Rarely, other nerves may be affected, such as our patients, including hypoglossal, accessory and even absent of pectoralis major muscle, syndactyly, barchydactyly and hypoplasia of the hand – which added to facial palsy comprise the Möebius-Polland syndrome, seen in our patient. There are only two cases of Möebius syndrome associated to epilepsy, one of which was diagnosed as a psychogenic non-epileptic seizures (PNES) due to the stress of coping with such a devastating congenital illness. MRI findings are a reflection of a hypoplastic brainstem, with loss of VII nuclei and consequently the facial colliculus, leading to a straight 4th ventricule floor. Other findings may include calcification of the pons and absence or cerebellar hypoplasia. This fascinating disorder has many physiopatological theories, but the most accepted is an early ischemic brainstem lesion, causing underdevelopment of cranial nuclei. **Final comments:** Epilepsy and Mobius syndrome is extremely rare, but this association should be kept in mind when dealing with this rare syndrome.

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MOGAD optic neuritis possibly triggered by bartonellosis: a case report

Weverton Carlos da Silva Teixeira[1], Aline Paredes Cardoso Fukuda[1], Julia Haddad Labello[1], Mário Luiz Ribeiro Monteiro[1], Leandro Tavares Lucato[1], Guilherme Diogo Silva[1], Tarso Adoni[1], Samira Luisa Apóstolos Pereira[1], Luiz Henrique Martins Castro[1], Dagoberto Callegaro[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Weverton Carlos da Silva Teixeira; Email: weverton.teixeira@hc.fm.usp.br.

Case presentation: A 31-year-old woman presented with fever, malaise, and headache three days after hiking at night in Minas Gerais, Brazil. Five days later, she developed acute visual impairment and pain with eye movements in the left eye (OS). Neuro-ophthalmological evaluation showed 20/70 visual acuity, altered color vision, central scotoma, relative afferent pupillary defect, and mild optic disc hyperemic edema OS, but no other ocular findings. Initial laboratory screening for infections, including syphilis, and CSF analysis were normal. MRI showed signs of optic neuritis (ON) in the intraorbital segment of the left optic nerve, while brain and spine MRI were normal. Treatment with methylprednisolone 1g for three days led to complete visual improvement, followed by rapid corticosteroid tapering. Three weeks later, she presented with a right-sided headache, vision loss, and painful eye movements in the right eye (OD). Visual acuity was 20/800 OD and 20/25 OS. OCT showed bilateral loss of ganglion cells in the macula without other retinal abnormalities. New CSF analysis showed only mild lymphocytic pleocytosis (8 cells). MRI indicated residual alterations in the left optic nerve and acute signs of ON in the entire orbital segment of the right optic nerve. Laboratory evaluation revealed high titers of serum anti-myelin oligodendrocyte glycoprotein (MOG) IgG (1:320) detected by cell-based assay and anti-Bartonella IgG (1:256). Serum anti-AQP4 IgG and CSF PCR for Bartonella henselae were negative. Therefore, treatment included doxycycline 100 mg twice daily for five weeks, methylprednisolone 1g for five days followed by prednisone 60 mg daily, and five sessions of plasmapheresis, resulting in complete visual recovery within two weeks. Anti-Bartonella IgG titers decreased to 1:128 and 1:64 at two and four months, respectively. Gradual corticosteroid tapering has been ongoing for six months. **Discussion:** Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) has been linked to various infectious triggers, including EBV, CMV, Borrelia burgdorferi, and SARS-CoV-2. B. henselae infection, although more commonly associated with neuroretinitis, emerged as a potential trigger in this case. After a presumed flea bite, initial febrile disease and high serological titers strongly indicated active or recent infection. In contrast, the absence of other ocular abnormalities and a negative CSF PCR for B. henselae reduce the likelihood of direct optic nerve infection. The presence of a core clinical demyelinating event and a clear positive anti-MOG IgG test meet the 2023 MOGAD diagnostic criteria. Longitudinal extensive bilateral ON and the initial response to high-dose glucocorticoids further support the diagnosis of MOGAD. **Final comments:** This case raises the hypothesis that Bartonella infection can lead to optic neuritis not only through direct infection (neuroretinitis) but also as a trigger for MOGAD, an immune-mediated disease.

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MOGAD rhombencephalitis and myelitis: a rare report of a lethal case

Larissa Silva Teixeira[1], Ana Flavia Souza Freire da Silva[1], Artur Pedreira de Andrade Souza[1]

[1]Hospital da Bahia, Salvador BA, Brazil.

Correspondence: Larissa Silva Teixeira; Email: dralarissa.neurologia@gmail.com.

We present a case of a 32-year-old brown man, cocaine user, who in November 2023 presented with paraparesis and urinary retention that lasted 7 days. Neuraxial MRI showed an extensive area of hypersignal in the pons tegmentum and middle cerebellar peduncles, in addition to longitudinally extensive signal changes affecting the cervicothoracic spinal cord. CSF showed lymphomonocytic pleocytosis (15 cells), and extensive infectious screening was unremarkable. The patient underwent methylprednisolone (MP) 1g for 3 days, with almost full recovery, including regaining the ability to walk. After 5 days, the patient worsened again, this time with tetraparesis, facial diparesis, dysphagia and dysautonomia. A new MRI showed an increase in the extent of hypersignal areas on T2/FLAIR that compromise the brainstem, in addition to a longitudinally extensive myelitis from the bulbomedullary transition to the conus medullaris. At this time, CSF showed pleocytosis of 60 cells. Molecular viral panel, PCR for tuberculosis, HIV and syphilis serology, schistosomiasis and listeria tests, rheumatological markers and anti-aquaporin antibody were negative. Anti MOG antibody come up positive 1:40. A 7-day course of pulse therapy with methylprednislone and associated therapy with human immunoglobulin was immediately completed, but the patient's dysautonomia worsened, requiring intubation and worsening to cardiorespiratory arrest. After cardiopulmonary resuscitation and numerous complications during hospitalization, such as septic shock and convulsive status after cardiac arrest, the patient progressed with progressive improvement, return of level of consciousness, improvement of tetraparesis, regaining strength up to grade 3. We chose to complete a 2nd cycle of immunoglobulin after 60 days, still during hospital stay, and the patient maintained significant improvement. On the day of hospital discharge, however, he developed septic shock which resulted in death after 10 days. The fact that MOGAD mainly involves the optic nerves and spinal cord involvement in MOGAD often affects the thoracic region and the conus medullaris may contribute to its lower mortality rate. We report a serious case of medullary bulb involvement that resulted in death despite initially have showed a clear improvement after agressive immunotherapy.

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MOGAD syndrome: a differential diagnosis between pediatric demyelinating encephalopathies - a case report

Vanderson Carvalho Neri[1], Thaynná Alvim Gomes[2], Ana Carolina Monteiro Malafaia Carvalho[2]

[1]Hospital Federal da Lagoa, Departamento de Neurologia, Rio de Janeiro RJ, Brazil.
[2]Faculdade de Medicina de Campos, Departamento de Neurologia, Campos dos Goytacazes RJ, Brazil.

Correspondence: Vanderson Carvalho Neri; Email: vandersoncn@yahoo.com.br.

Case presentation: Female, mixed race, 12 years old. In February 2020, he developed a sudden onset of aphasia, sialorrhea, and distal paresis of the right upper limb. Skull MRI: large hypersignal lesion on Flair and T2 with bilateral digitiform edema pattern; discrete diffusion hypersignal and areas of contrast enhancement with a pattern of perivascular/perivenular uptake in the subcortical white matter of the frontal regions; optic neuritis on the left and, after 15 days, spontaneous remission of aphasia. After two months, a new skull MRI showed significant improvement in the reported lesions, which no longer captured contrast, but there were new lesions on the right parietal surface and in the right temporo-occipital subcortical white matter. In November 2021, focal convulsive crisis. In 2022, generalized tonic-clonic seizure. She remained asymptomatic until December 2022, when she developed distal paresis of the right upper limb, changes in coordination, and dysarthria. Laboratory Investigation: MOG+, AQP4-, CSF unchanged. **Discussion:** Recently, the international diagnostic criteria for MOGAD (Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease) were published, and the diagnosis of MOGAD is now made based on the presence of at least one of the core clinical demyelinating events (optic neuritis, myelitis, ADEM, cerebral monofocal or polyfocal deficits, brainstem or cerebellar deficits, and cerebral cortical encephalitis often with seizures), a positive MOG-IgG test, and the exclusion of alternative diagnoses including MS. Approximately 30% of MOGAD cases are in the pediatric age group, and MOGAD comprises approximately 35-40% of cases of acquired CNS demyelinating syndrome in the pediatric population. When a MOG antibody is low-positive in a patient with a core clinical demyelinating event, at least one supporting clinical or MRI feature should be met to make a diagnosis of MOGAD since alternative diagnoses including MS can be low-positive. Approximately 35-50% of MOGAD cases have a relapsing course, with the relapse risk being slightly higher (~60%) in the young adult-onset group. Optic neuritis is the most common onset phenotype (~40%). Age-related onset phenotype is a feature of MOGAD, where ADEM is more common in pediatric patients <10 years old, while myelitis and brainstem encephalitis are more common in adult patients. **Final comments:** We describe a case of a pediatric patient with encephalopathy and optic neuropathy associated with anti-MOG antibodies, with extensive brain lesions, without spinal cord injury, but with a good response to pharmacological treatment. Recurrent condition, with the presence of the antibody persisting, even during treatment with corticosteroid therapy.

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Molecular analyses of cell cycle-related genes in Rasmussen encephalitis

João Ismael Budelon Gonçalves[1], Marciéli Gerhardt[2], Vinicius Rosa de Castro[2], William Alves Martins[2], Fernando Xavier[1], Jaderson Costa da Costa[1], Eliseu Paglioli Neto[2], André Palmini[2], Daniel Marinowic[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Instituto do Cérebro, Porto Alegre RS, Brazil.

[2]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Marciéli Gerhardt; Email: marcieligerhardt@yahoo.com.br.

Case presentation: A 13-year-old boy presented a bilateral tonic-clonic seizure a few weeks after a self-limited viral illness. Neurological symptoms began 2 years earlier, with spasms in the left limbs, eventually associated with generalized tonic-clonic seizures. His medical history and development had been otherwise uneventful. After failing several antiseizure drugs, he was referred to our tertiary epilepsy center. At arrival, neurological examination showed spastic left hemiparesis and moderate to severe dysarthria, with continuous left hemiclonic motor seizures (epilepsia partialis continua [EPC]). MRI showed progressive right hemisphere and caudate atrophy, as well as hyperintensities in the right temporal and frontal lobes, extending throughout the right insula. EEG showed progressive slowing in the right hemisphere with periodic sharp waves over the right frontotemporal and parietal regions and frequent electrographic seizures. His symptoms led us to the diagnosis of probable Rasmussen's encephalitis (RE). Therefore, he underwent a right functional hemispherectomy. On the first postoperative day, he was awake and responsive and had no further seizures, although developed a transitory right third nerve palsy. He remained seizure-free six months after surgery (Engel IA) and has shown progressive motor improvement of the left leg and arm. Cortical tissue sample was obtained through functional hemispherectomy, and histopathology confirmed RE. Excised tissue was subjected to molecular analysis to evaluate the expression of cell cycle-related genes and protein levels of inflammatory cytokines, as well as neurotrophic factors. **Discussion:** RE stands as a rare neurological disorder marked by progressive cerebral hemiatrophy and epilepsy resistant to medical treatment. In this case report, we present a patient who underwent hemispherectomy and has remained seizure-free for over six months, experiencing gradual motor improvement. Despite extensive study, the primary cause of RE remains elusive, while its histopathological features comprehend cortical inflammation, neuronal degeneration, and gliosis. In our analyses, we found a general decreased expression of cell cycle-related genes in the brain tissue from our patient. The BRCA1, BIRC5, CASP3, E2F4 and CCDN genes were more than 5-fold downregulated. Alongside, levels of BDNF and TNF-α proteins were elevated. These findings suggest a potential involvement of cell cycle regulators in the progression of RE. **Final comments:** Molecular analyses may play an important role for understanding RE, as cell cycle-related genes might be involved in its physiopathology.

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Molecular and clinical aspects of a Brazilian cohort of patients with variegate porphyria

Michelle Abdo Paiva[1], Beatriz Carneiro Gondim Silva[1], Rodrigo de Holanda Mendonça[1], Cristiane de Araujo Martins Moreno[1], Andre Macedo Serafim Silva[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Michelle Abdo Paiva; Email: michelle.abdop@gmail.com.

Background: Acute hepatic porphyrias (AHP) are a group of metabolic disorders of the heme biosynthesis Clinical aspects are very similar among acute intermittent porphyria (AIP), variegate porphyria (VP) and hereditary coproporphyria (HCP), regarding acute attacks and hereditary autosomal dominant pattern However, VP and HCP present with cutaneous symptoms. In most countries, AIP is believed to be the most common type of AHP followed by VP. Data from Brazil are still scarce due to lack of available specific biochemical exams and genetic studies. **Objective:** To describe epidemiological, clinical, and genetic profiles of patients with VP in a single tertiary center in Brazil. **Methods:** We included patients from our center with AHP as defined by the consensus of 2023, meaning typical heme precursors accumulation and/or a pathogenic variant in genetic exam. We reviewed medical history, medical records and complementary exams of all patients. A severe attack was defined with one or more of the following - significant hyponatremia, peripheral neuropathy, urinary retention or incontinence, central nervous system involvement and arrhythmias. **Results:** A total of 26 genetically confirmed AHP patients were identified in our hospital. VP was diagnosed in 13 patients from six different families. Ten patients were female, and 3 patients were male, with a mean age of 40 (22-55). The age at first symptoms ranged from 17 to 46 years. Four patients had two severe attacks, five patients had one severe attack, one patient had recurrent symptoms, not being possible to define the exact number, and three patients were asymptomatic. During the attack, seven patients experienced acute flaccid paralysis. Of these, three patients had seizures. Six patients presented psychiatric symptoms (anxiety, depression and/or psychosis). Acute phase electroneuromyography available for six patients with findings of axonal sensorimotor polyneuropathy for four cases, one patient with intermediate polyneuropathy, meeting CIDP criteria and one patient with only autonomic alterations. Acute confirmatory biochemical exams were available for five patients. Remaining cases had biochemical non conclusive biochemical exams or results not available. Skin lesions and/or photosensitivity were reported in five patients. Four families shared the same missense variant in exon 6, c503G> A of the PPOX gene, one patient with a deletion in exon 5-6 and one patient with a missense variant in exon 5 c.454C>T. **Conclusion:** Data from our center may suggest that VP is more common in our population than previously reported, as more than half of our patients with AHP had a pathogenic variant in PPOX. However, many from the same family, which may indicate a difference in penetration between AHPs. Clinically, patients may present similarly to other forms of AHP. Skin injuries may not be present in all patients. The lack of biochemical studies in our country may influence diagnostic accuracy.

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Molecular therapy with gapmer antisense oligonucleotides (ASOS) in the treatment of Angelman syndrome

Giovana Pereira[1], Mariana Marques Rodrigues de Almeida[1], Luísa Ribeiro Dias Godoy[2], Vitória Aparecida Caetano[1], Thairine Alves Nascimento[1], Maria Fernanda Pascholati Penteado[1], Fernando Soares de Morais[1], Júlia Ribeiro Dias Godoy[2], Livia Maria Rodrigues de Almeida[3], Eduardo Freire Oliveira Figueiredo[1]

[1]Universidade Nove de Julho, Bauru SP, Brazil.
[2]Universidade do Vale do Sapucaí, Pouso Alegre MG, Brazil.
[3]Universidade Metropolitana de Santos, Santos SP, Brazil.

Correspondence: Giovana Pereira; Email: giovanapereira61@outlook.com.

Background: Angelman syndrome (AS) is a neurogenetic disorder caused by mutations that lead to the deactivation of the maternal allele of the UBE3A gene, present on chromosome 15. In neurons of the central nervous system (CNS), the UBE3A gene is subject to genomic imprinting, in which the maternal allele is expressed while the paternal allele is silenced by a long non-coding RNA called UBE3A-AS. Therefore, all people depend on the allele inherited from the mother to express the final product of the UBE3A gene, the E3A ubiquitin-protein ligase, responsible for directing certain proteins for degradation in brain tissue. The combination of paternal silencing of UBE3A and loss of maternal UBE3A expression results in the complete loss of expression of the neuronal protein UBE3A, compromising neuronal function and plasticity, justifying the severe neurological symptoms present in people with AS, including developmental delay, motor and cognitive deficits and seizures. Hyperactivity and hyperexcitability happen in 100% of cases, and they react to the slightest physical and mental stimuli with uncontrolled explosions of laughter, vocalizations (screams of excitement) and shaking hands (waving hands). **Objective:** Report the genetic pathogenesis and analyze the current perspectives for the management of AS based on molecular therapy with Gapmer antisense oligonucleotides (ASOs). **Methods:** Systematic literature review based on the PRISMA methodology. It was searched for articles indexed on the PubMed platform, in English language, in the last 10 years. 153 results were gathered, which were refined by the Ryyan platform and detailed by three independent reviewers. At the end of two analyses, 15 documents were fully reviewed. **Results:** Currently, there is no specific approved treatment for Angelman Syndrome. However, evidence suggests that molecular therapy with Gapmer antisense oligonucleotides (Gapmer ASOs) may be an effective therapeutic strategy. These modified oligonucleotides can modulate gene expression, increasing levels of functional proteins or improving the structure of altered proteins. Studies have shown that Gapmer ASOs can efficiently suppress the expression of the UBE3A-AS transcript and reactivate the expression of the paternal UBE3A allele in cultured human neurons, improving neurological deficits in mouse models of AS. These studies also demonstrated the feasibility and potential clinical benefit of a disease-modifying strategy for individuals with AS. **Conclusion:** Reactivating expression of the paternal UBE3A allele could be a powerful modifier treatment for AS, although further studies are needed to confirm efficacy in the human brain.

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Monoclonal gammopathy: an uncommom, but not rare cause of peripheral neuropathy

Márcio Alves da Cruz Júnior[1], Carla Cristina Teixeira[1], Felipe Henriques Monteiro[1], Iago Collins Tavella Paulino[1], Liz Rocha Paes Hellbrügge[1], Lorenzo Batista Oliveira[1], Pietro Soares Motta[1], Suzana Rosa Silva[1], Daniel Sabino de Oliveira[1], Marcelo Maroco Cruzeiro[1]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Márcio Alves da Cruz Júnior; Email: marcio.jr14@yahoo.com.br.

Case presentation: Case 1: white woman, 75 years old, hypertensive, depressive disorder and hypothyroidism with bilateral paresthesia in the lower limbs (LL) up to the level of the knees and progressive worsening within 8 months, paraparesis with distal predominance and burning pain in the feet. There were weight loss (8kg in 3 months), pain in the right hypochondrium, pleural effusion on the left side and was hospitalized. Tetraparesis with a predominance in the lower limbs, hyporeflexia, thermal and painful hypoesthesia up to the knees, reduced vibratory sensitivity up to the pelvic girdle, and ataxic-talonant gait were observed. Electromyography (EMG) revealed demyelinating sensory-motor neuropathy. Electrophoresis showed monoclonal peak, and bone marrow biopsy had hypercellularity, interstitial plasmacytosis, presence of CD138 and preferential expression of IgA/lambda light chain, referring to multiple myeloma. The diagnosis of POEMS Syndrome (polyneuropathy, organomegaly, endocrinopathy, monoclonal plasma cell disorder, and skin changes) has been established. Treatment with melphalan and dexamethasone was instituted (bone marrow transplant refused), evolving with partial motor clinical improvement. Case 2: A 67 year-old wthite Woman, with

hypertensive, diabetic, chronic venous insufficiency and fibromyalgia developed global weakness of all four limbs, numbness in boots and gloves, imbalance and frequent falls. The gait was ataxic, there was global areflexia, painful hypoesthesia and distal abscence of vibratory sensitivity of all four limbs, and EMG showed sensory-motor polyneuropathy of a demyelinating nature. Cerebrospinal fluid (CSF) had protein ncreased, and protein electrophoresis identified IgM/Lambda monoclonal peak. Multiple myeloma and lymphoproliferative diseases have been discarded. The measurement of antibodies against myelin-associated glycoprotein (anti-MAG) identified reactive IgM and non-reactive IgG. Chronic Demyelinating Inflammatory Polyneuropathy (CIDP) with distal variant (anti-MAG DADS) was diagnosed. Methylprednisolone pulse therapy monthly with partial improvement of the condition. **Discussion:** Monoclonal gammopathies (GM) are a heterogeneous group of diseases of hematopoietic system, which result from the production of immunoglobulins (Ig) by differentiated lymphocytes or monoclonal plasma cells and affect the peripheral nervous system by mechanisms not yet fully understood, such as deposition of Ig or amyloid substance, and its prevalence increases with age, reaching 10% around 80 years of age, especially in men. Both cases are due to the deposition of antibodies. Treatment is with corticosteroid therapy, intravenous human immunoglobulin, or immunosuppressants. **Final comments:** Monoclonal gammopathies, although uncommom, are not rare and should be part of the investigation in people from the sixth decade with neuropathy.

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Mononeuritis multiplex following crotalid envenomation: a case report

Francisco Luciano Honório Barreto Cavalcante[1], Antônio Edvan Camelo Filho[1], Pedro Lucas Grangeiro de Sá Barreto Lima[1], Pedro Vitor Ferreira Rodrigues[1], Oliver Reiks Miyajima[2], Nahiana Bezerra de Menezes Cruz[3], Pedro Braga Neto[4], Paulo Ribeiro Nóbrega[4]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.
[3]Centro Universitário Christus, Fortaleza CE, Brazil.
[4]Universidade Federal do Ceará, Departamento de Medicina Clínica, Fortaleza CE, Brazil.

Correspondence: Francisco Luciano Honório Barreto Cavalcante; Email: francski2304@gmail.com.

Case presentation: A 34-year-old man was bitten on his left hand by a Crotalus durissus snake (rattlesnake) and suddenly developed pain and edema. The patient evolved in a few hours to a confusional state with lethargy, nausea, blurred vision, and dark urine. He was admitted to a reference center with laboratory tests revealing elevated liver enzymes and creatinine with myoglobinuria. He was treated with 20 anticrotalic serum (ACS) vials, each containing 10mL in a 1,5 mg/mL concentration. The patient had an allergic reaction to the serum, manifesting as periorbital and tongue edema. He was discharged after stabilization with normalization of liver and renal function. However, after three weeks, he presented with paresthesia and "needle-like" pain in his left hand and arm, which later progressed to his right hand. A week later, those symptoms progressed to his legs, which the patient described as a sensation of fatigue. An electroneuromyography (ENMG) was performed on the patient, showing an axonal, subacute, sensorimotor mononeuritis multiplex (MNM) in his four limbs, with ongoing denervation signs. An extensive investigation of potential causes of MNM was conducted, and the results were negative. He was treated with prednisone (1mg/kg) and pregabalin. Six months later, a new ENMG was performed with an expressive improvement of sensorimotor conduction without denervating activity in that exam. **Discussion:** Neurotoxicity due to crotalid venom usually occurs by a neuromuscular blockade, causing a myasthenic syndrome that presents with ptosis, mydriasis, ophthalmoplegia, and acute flaccid paralysis. Peripheral neuropathy (PN) is an atypical feature of snake envenomation. The literature points out that snake envenomation could cause local sensitive axonal mononeuropathy adjacent to the bite site, either due to the bite itself, compartment syndrome, or post-traumatic neuroma. Acute axonal sensorimotor polyneuropathy, both as an early or as a delayed presentation of envenomation, has been previously reported, with partial or complete recovery in months. As far as we know, this is the first case of MNM associated with crotalid snake envenomation. Another hypothesis is that a hypersensitivity reaction to the ACS could cause the MNM. Usual side effects of ACS include IgE-mediated reactions and serum sickness, which could cause PN. However, normal complement values and the lack of fever and skin changes weaken this hypothesis in the present case. **Final comments:** Snake envenomation is a common health issue in Brazil. Elusive manifestations of ophidic accidents or even unknown side effects of serum therapy, such as MNM, are still a challenge for physicians. Therefore, further studies are needed to better understand the relationship between those factors and improve snakebite victims' quality of life.

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Morbidity and mortality from traumatic brain injury in infants under 1 year old in the state of São Paulo between 2018 and 2023

Yasmim Melo Costa de Azevedo[1], Victhor Luiz Senna de Moraes[2], Paula Ester de Oliveira Almeida[3], Victor Hugo da Silva Dias[4], Danielle de Souza Mometto[5], Sergio Gonçalves de Oliveira[6]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.
[2]Universidade Federal Dos Vales do Jequitinhonha e Mucuri, Diamantina MG, Brazil.
[3]Faculdade de Minas, Belo Horizonte MG, Brazil.
[4]Universidade Cidade de São Paulo, São Paulo SP, Brazil.
[5]Universidade Potiguar, Natal RN, Brazil.
[6]Hospital Socor, Departamento de Neurocirurgia, Belo Horizonte MG, Brasil.

Correspondence: Yasmim Melo Costa de Azevedo; Email: yaxmelo@outlook.com.

Background: Traumatic brain injury (TBI) is one of the main causes of death and disability in childhood. It occurs due to an external trauma that results in anatomical changes and functional impairment. This incident is more serious in children, especially younger ones, than in adults, since the cervical ligaments and muscles are weaker, the skull is thin and flexible and the sutures are not yet fused. Common scenarios of occurrence in newborns are associated with the birth process, while in children they are associated with falls, aggression, or car accidents. However, despite its relevance, there is a gap in studies that investigate this topic and epidemiological data in Brazil. **Objective:** To analyze the epidemiological information on morbidity and mortality from traumatic brain injuries in infants under 1 year old in the state of São Paulo. **Methods:** Consists of a transversal and descriptive study with a quantitative approach. Based on information from the Hospital Admissions System (SIH/SUS) linked to DATASUS. The analysis involves raw data on hospitalizations, the number of cases that resulted in death, and the corresponding mortality rate due to Traumatic Brain Injury in infants under 1 year old in São Paulo from January 2018 to December 2023. This data was entered into Excel to establish the comparative evolution using percentages, aiming to organize temporal trends. **Results:** It was found that in the period analyzed, there were a total of 4273 hospitalizations. Regarding this result, we have that: 702 cases occurred in 2018, 739 in 2019, 749 in 2020, 718 in 2021, 639 in 2022, and 726 in 2023. Between 2021-2022 is the period with the greatest reduction, around 11%. During the entire period, simple variations were observed with the maintenance of trends proven by the 3% increase between 2018-2023. Regarding data on cases that evolved into deaths, there was a total of 42 in which the municipality that presented the highest number of deaths was São Paulo, with 9 cases, which represented around 21% of the total. Regarding the mortality rate, the municipality of São Paulo had the lowest (0.70) and Catanduva the highest (50.0). **Conclusion:** Thus, the effectiveness of therapeutic management in the good prognosis of patients is significant when comparing the number of total cases with those with the worst health outcome. Therefore, it is clear that the highest number of registered cases occurred in 2020, with 794 (17.5%). The drop in registration occurred in 2022, with 639 (14.9%). Furthermore, the polarity of results stands out when related to the variables that analyze the number of deaths and the mortality rate, with the capital of São Paulo being the highlighted representative. In this sense, a more compact data analysis, not yet widely explored, aims to promote impact through information and generate discussions to optimize the scenario of the injury presented in the state.

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Mortality due to intracranial trauma in the elderly in Brazil: an ecological study (2008-2024)

Anna Gabriella Venâncio Neves de Moraes Tosta[1], David Cohen[2], Larissa Farah de Oliveira[3], Hadassa Lucena Sales Santos[4], Paulo Fernando Katsuo Ogatha Ito[5]

[1]São Leopoldo Mandic, Campinas SP, Brazil.
[2]Universidade Luterana do Brasil, Canoas RS, Brazil.
[3]Universidade Municipal de São Caetano do Sul, São Caetano do Sul SP, Brazil.
[4]Centro Universitário do Maranhão, Imperatriz MA, Brazil.
[5]Universidade para o Desenvolvimento do Estado e da Região do Pantanal, Campo Grande MS, Brazil.

Correspondence: Anna Gabriella Venâncio Neves de Moraes Tosta; Email: annagabriellatosta@gmail.com.

Background: Intracranial trauma (ICT) is one of the main emergencies and causes of morbidity and mortality in the elderly. Factors that favor a worse prognosis in the elderly include bone fragility, use of anticoagulants and antiplatelets, comorbidities, and previous level of consciousness. **Objective:** To analyze the morbidity and mortality due to intracranial trauma in the elderly in the Southeast Region between January 2008 and March 2024. **Methods:** This is an ecological, cross-sectional, descriptive study with a quantitative approach, conducted in May 2024, using data collected from the Hospital Information System of SUS (SIH/SUS - Sistema de Informações Hospitalares do Sistema Único de Saúde), available from the Department of Informatics of the Unified Health System (DATASUS - Departamento de Informática do Sistema Único de Saúde). The variables used were hospitalizations, total cost, deaths, and mortality rate. The collected data were organized in Microsoft Excel spreadsheets and analyzed using descriptive statistics. **Results:** The analysis revealed mortality rates, deaths, hospitalizations, and total cost, respectively, for each state in the Southeast Region. In Minas Gerais, the mortality rate was 13.10% (6,765 deaths and 51,626 hospitalizations) with a total cost of R$116,313,880.46; in Espírito Santo, the mortality rate was 13.51% (1,135 deaths and 8,399 hospitalizations) with a total cost of R$18,792,052.87; in Rio de Janeiro, the mortality rate was 21.79% (5,723 deaths and 26,269 hospitalizations) with a total cost of R$48,345,157.04; in São Paulo, the mortality rate was 17.82% (18,116 deaths and 101,633 hospitalizations) with a total cost of R$206,647,673.22. Thus, it was possible to analyze that, although Rio de

Janeiro and Minas Gerais have a similar total population, Rio de Janeiro has the highest mortality rate. This suggests differences in socioeconomic conditions, access to healthcare, or quality of medical services. Factors such as income distribution, education, employment, distribution of hospitals and healthcare professionals, as well as the efficiency and effectiveness of treatments, can influence these variations in mortality rates. **Conclusion:** The data examined from January 2008 to March 2024 highlight the discrepancy in morbidity and mortality between Minas Gerais and Rio de Janeiro. Therefore, greater attention and search for prevention strategies, such as investment in elderly care policies, as well as medications capable of preventing conditions that may lead to possible brain trauma and reduce morbidity and mortality, are necessary.

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Mortality due to malignant neoplasm of the CNS in children under 20 years old in Brazil

Gabriel Cordova[1], Emanuel Henrique dos Santos[1], Izabelly Constantinov Lemos[1], Kauan Correia Derbli[1], Anne Beatriz Ricardo Amaral[1], Sofia Beatriz Andrade Manfio[1], Julia Farto Viana Oliveira[2], Gabriel André Silvério[1]

[1]Centro Universitário Campo Real, Guarapuava PR, Brazil.
[2]Universidade Estadual do Centro-Oeste, Guarapuava PR, Brazil.

Correspondence: Gabriel Cordova; Email: gabrielcordovah15@gmail.com.

Background: Malignant Neoplasm of the Central Nervous System (CNS) comprises a spectrum of malignant brain tumors that detrimentally impact neurological functions. It is typified by the invasive and unbridled proliferation of aberrant cells within the encephalic tissue, exhibiting aggressive infiltration into the adjacent tissues, with the capacity to originate in both the encephalon and the spinal cord. **Objective:** To compare the mortality rate of children under 20 years old due to CNS cancers across the different Brazilian regions. Additionally, the study aims to analyze which gender is most affected by this condition, as well as to determine the most common anatomical location of this neoplasm within the CNS. **Methods:** An epidemiological study using the DATASUS database, covering 2017-2021. Constants included deaths by region and age group. Brazil was divided into North, Northeast, Southeast, South, and Central-West. Variables included sex, age (0 to 19 years), and neoplasm location. ICD codes used were C70, C71, and C72. Additionally, Tabnet data of the resident population in each region for the same period was used to determine the exposed population (n= 100,000). **Results:** From 2017 to 2021, the number of total deaths was respectively: 642, 593, 609, 552 and 590. Of these, 5.09% occurred in children under 1 year. The Southeast had the highest rate of deaths, responsible for 37.61%, followed by the Northeast (29.71%), South (14%), North (10.25%) and Central-West (8.44%). Among the ones under 20 years old and over 1 year, the average mortality rates during the period of the study, in descending order was, 1.06 in the Central-West, 1.05 in the South, 0.99 in the Northeast, 0.96 in the Southeast and 0.91 in the North. Thus, the Central-West and the South regions had the highest mortality rates. In those, under 1 year, the Northeast (1.23) and North (1.16) led the ranking of the average mortality rate, followed by the South (1.15), Southeast (1), and Central-West (0.75). Of all deaths due to malignant neoplasm of the CNS, 53.48% occurred in males (n= 1597) and 46.52% in females (n= 1389). Regarding the anatomical location, 81.42% of neoplasms were located in the encephalon, followed by 18.04% located in the spinal cord, cranial nerves, and other parts of the CNS, and 0.54% in the meninges. **Conclusion:** The Central-West and South regions had the highest mortality rates, in those under 20 years and over 1 year. On the other hand, the North and Northeast regions had the highest mortality rates among those under 1 year. The sex with the highest number of deaths was male. In addition, more than 80% of neoplasms were located in the encephalon. Thus, further research is required to understand the mortality rates of children in each region, ensuring an accurate grasp of epidemiology and promoting the physical, mental, and social well-being of the Brazilian population.

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Mortality from Alzheimer's disease in Bahia, 2017-2022

Mariela Sousa dos Santos[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Mariela Sousa dos Santos; Email: mariellasanttos5@gmail.com.

Background: Alzheimer's disease (AD) is the most usual type of dementia among the elderly and it's cognitive and neuropsychiatric manifestations result in progressive disability and incapacitation. It is characterized, histopathologically, by massive synaptic loss and neuronal death observed in brain areas responsible for cognitive functions, including the cerebral cortex, hippocampus, entorhinal cortex and ventral striatum. It is the main cause of functional dependence and mortality among the elderly population. Socioeconomic aspects and genetic modifications may be linked to AD. **Objective:** Measuring the prevalence of Alzheimer's disease mortality rate and potential associated socioeconomic determinants among the population of Bahia, 2017-2022. **Methods:** A descriptive, retrospective, time series study was carried out to evaluate Alzheimer's mortality rate evolution in Bahia, from 2017 to 2022. Death due to Alzheimer's disease happens when the disease is listed as the cause of death in the Death Declaration (DO). The G30 code of the International Classification of Diseases, 10th revision (ICD-10) was considered. To calculate mortality rates, we used the number of deaths available in the International Mortality System (SIM), population data from 2000 to 2021 censuses and the 2022-panorama census, from the Brazilian Institute of Geography and Statistics (IBGE). Standardized mortality rates per 100,000 inhabitants with Alzheimer's disease as the underlying cause used the census population from 2017 to 2022. Data were analyzed using SPSS software, version 17.0 **Results:** In the period between 2017 and 2022, 600,760 deaths were registered in the SIM in Bahia, of which 1% had Alzheimer's disease as the cause mentioned in the DO. Considering all central nervous'"s system deaths between 2017 and 2022, the general prevalence of mortality from Alzheimer's was 45.5% among all deaths, being more frequent in women, mixed race/color, and age group ≥80 years old. **Conclusion:** High prevalence of mortality from Alzheimer's disease. Socioeconomic factors associated with the outcome were age group, race/color and sex.

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Mortality from cerebral palsy in Brazil between 2013 and 2022

Ana Carolina Ribeiro de Araujo e Araujo[1], Aeriberto Carlos Lindoso de Souza[1], Consuelo Penha Castro Marques[1], José Carlos Gomes Patriota Neto[1]

[1]Universidade Federal do Maranhão, Pinheiro MA, Brazil.

Correspondence: Ana Carolina Ribeiro de Araujo e Araujo; Email: anacarolinaraa@hotmail.com.

Background: Cerebral palsy is the main cause of disability in children, caused by permanent and non-progressive damage to the central nervous system during the fetal period, at birth or in early childhood, generating serious complications in the development of skills, tone, reflexes and the posture. In Brazil, for every 1,000 children born, 7 are carriers of this paralysis, in addition, it is estimated that there are around 30,000 to 40,000 new cases per year. Diagnosis involves a complete neurological examination, finding delay in motor development, persistence of primitive reflexes and presence of abnormal reflexes. **Objective:** To investigate mortality from cerebral palsy in Brazil between the years 2013 and 2022. **Methods:** Quantitative, descriptive, time series and ecological study of deaths from cerebral palsy in Brazil, during the period from 2013 to 2022, with data from the System of Information on Mortality - SIM, available from the Information Technology Department of the Unified Health System (TABNET/DATASUS). The variables used were: region, age group, sex, education, marital status and place of occurrence. **Results:** n=14,371 deaths due to cerebral palsy were reported in Brazil during the study period, with the region with the highest incidence being the Southeast, with n=5,803 deaths, followed by the Northeast n=3,846 and the South n=2,656, affecting mainly age ranges from children under 1 year of age to 29 years of age, representing 70.6% of cases. Furthermore, cerebral palsy was mostly found in males (56.7%), especially in individuals with little or no education (69.3%), in singles (78.9%), being the main place of occurrence of deaths in hospital n=9,010, followed by home n=4,144 and other health establishments n=951. **Conclusion:** Given the above, it can be concluded that cerebral palsy has a higher prevalence of deaths in individuals younger than 29 years of age, showing a lower life expectancy of these patients. That said, supportive treatments and delaying the progression of the pathology become necessary for the well-being and life expectancy of these individuals. Given the evolution of treatments and technologies focused on the neurological area, it is necessary to further evolve the neurological treatment of specific paralysis.

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Mortality from CNS tumors in the state of Minas Gerais over the past 25 years

José Victor Ribeiro Silva Gomes[1], Kananda Oliveira Garcia Ruiz[1], Cínthia Emanuelle da Costa Sousa[1], Rodrigo Chávez-Penha[1]

[1]Universidade Federal de São João del-Rei, São João del Rei MG, Brazil.

Correspondence: José Victor Ribeiro Silva Gomes; Email: josevrsgomes@gmail.com.

Background: Central Nervous System (CNS) tumors are neoplasms originating from tissues in the brain and in the spinal cord. They are estimated to be the 11th most common type of cancer in Brazil between 2023 and 2025. Its rising number of cases and deaths worldwide claims for better understanding of its trends. **Objective:** Describe mortality due to central nervous system (CNS) neoplasms in adults in the Brazilian state of Minas Gerais (MG). **Methods:** this is an ecological time-series analysis with data from DATASUS regarding deaths from all kinds of CNS neoplasms in adults of 20 or more years old residing in MG between the years 1998 and 2022, by age group and reported cause of death as in the ICD10. Joinpoint analysis was used to verify trends in age adjusted mortality rate per 100.000 people (MR) by annual percentage change (APC). **Results:** 19,537 deaths were recorded in MG in the period, most of them in the 60-69 age group (24.5%) and in men (52.2%). 77.7% of deaths (15,185) occurred due to malignant neoplasms of the brain (ICD C-71). Overall MR due to CNS tumors showed 2 identifiable trends: an increase (APC 2.44) from 1998 (MR 5.86) to 2006 (MR 6.89) and a decrease (APC -0.73) from 2006 to 2022 (MR 5.54). MR trends in men followed that pattern, but with an increase (APC 1.32) from

1998 (MR 6.28) to 2012 (MR 7.29), followed by decrease (APC -1.32) until 2022 (MR 6.23). Women MR trend was of reduction through the whole period (APC -0.13; 1998 MR 5.64 to 2022 MR 4.90). MR by age group showed very different patterns, with an increase for the 50-59 (APC 0.31) and 80 or more years old (APC 0.83) groups, and decrease in the other groups (APCs: 20-29: -0.15; 40-49: -0.81; 60-69: -0.02; 70-79: -0.28). As for identified deaths related to CNS tumors, malignant tumors were responsible for the highest MR and all showed an increase in different patterns followed by reductions. Trends of MR for malignant neoplasm of meninges (C70) increased (APC 3.72) from 1998 (MR 0.25) until 2010 and then decreased (APC -5.50) up to 2022. As for trends of MR for malignant neoplasms of the brain (C71) raised (APC 1.83) from 1998 (MR 10.3) till 2006 (MR 11.25) to then reduce (APC -1.70) until 2022 (MR 7.54). Finally, trends of MR for malignant neoplasms of spinal cord, cranial nerves and other parts of central nervous system (C72) increased (APC 8.14) from 1998 (MR 0.35) until 2017 (MR 2.25) and then decreased (APC 8.73) until 2022 (MR 1.50). **Conclusion:** Deaths from CNS neoplasms, although less common compared to other tumors, have become more frequent in Minas Gerais. The identified trends may raise concerns on the attention to CNS tumors in men, elderly people and malignant neoplasms of the brain through the studied period. Further investigation is needed to assess if the MR and trends identified relate to changes in the population age distribution, quality and accessibility to medical care.

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Mortality from viral meningitis in Brazil from 2011 to 2020

Larissa Bianca Borsone[1], Deborah Araujo Silva[1], Rynele Almeida de Fonseca[1], Annayza de Oliveira Bessa[1], Amanda Oliva Spaziani[1], João Carlos Bizinotto Leal de Lima[1], Francieli Gomes Lima[2], Vanessa Maria Gonçalves de Souza[1], Gustavo Henrique da Silva[3], Luis Carlos Spaziani[4]

[1]Universidade Brasil, Fernandópolis SP, Brazil.
[2]AME fernandopolis, Fernandópolis SP, Brazil.
[3]UPA Santa Fé do Sul, Santa Fé do Sul SP, Brazil.
[4]Fundação Educacional de Fernandópolis, Fernandópolis SP, Brazil.

Correspondence: Larissa Bianca Borsone; Email: larissa.borsone@hotmail.com.

Background: Viral meningitis results from inflammatory processes affecting the membranes that cover and protect the central nervous system, including the brain and spinal cord. It is a significant public health problem due to its high morbidity and mortality rates, complicated neurological sequelae, and transmission potential, which can lead to epidemic outbreaks. **Objective:** To evaluate the mortality profile from viral meningitis in Brazil between 2011 and 2020. **Methods:** Data collection was performed using publicly available information from the Mortality Information System (SIM) of the Ministry of Health in the Tabnet/DATASUS database between May 1 and July 22, 2023. This retrospective longitudinal study employed a quantitative descriptive approach. Data were grouped by age and educational level for the period from 2011 to 2020. Statistical inference was conducted using BioEstat 5.3 software, utilizing the Friedman test for group comparisons, and data presentation included descriptive statistics, measures of central tendency, and dispersion. **Results:** Between 2011 and 2020, there were 608 deaths from viral meningitis in Brazil. The highest number of deaths occurred in the Southeast region (261), corresponding to 42.93% of the total, and 132 (21.71%) cases in the South (p<0.0001). The Central-West (n=50, 8.22%) and North (n=52, 8.55%) regions had the lowest number of deaths. In terms of annual distribution, 75 (12.34%) deaths occurred in 2017 compared to 8.06% (49) in 2011, with no significant increase in deaths between 2011 and 2020 (p=0.1921). Regarding sex, 57.07% (n=347) of the deaths occurred among males and 42.93% (n=261) among females (p=0.0269). In terms of age, 293 cases (48.19%) occurred among individuals aged 20 to 59, and 29.64% (162) were aged 60 and over. The least affected age groups were those up to 9 years (15.63%, n=95) and 10 to 19 years (9.54%, n=58) (p<0.0001). Regarding education, 134 cases had 8 to 11 years of schooling (22.04%), and 129 (21.22%) had 4 to 7 years (p<0.0001). In terms of race/ethnicity, the majority were white (295, 48.52%), followed by mixed race (237, 38.98%), black (51, 8.39%), yellow (4, 0.66%), indigenous (4, 0.66%), and 17 (2.80%) were not reported (p<0.0001). **Conclusion:** In conclusion, patients who died from viral meningitis in Brazil tended to be from the Southeast and South regions, aged between 20 and 59 years, predominantly male, and of white or mixed race, with an average of 4 to 11 years of schooling. Keywords: Mortality; Viral Meningitis; Brazil.

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Mortality of viral encephalitis in children and adolescents in the Brazilian Midwest: an epidemiological study over the past 15 years

Gustavo Hayasaki Vieira[1], Giovana de Oliveira Araújo[1], Gustavo Cartaxo de Sousa Melo[1], Isabela Maciel Mesquita[1], Milena Almeida Barbosa[1], Paulo André Silva Miranda[1], Paulo Sérgio Machado Diniz[1], Rafaella Rodrigues de Freitas Vieira[1], Samira Dionísio Maia[1], Valentina Melo Rosa Centofanti[1]

[1]Pontifícia Universidade Católica de Goiás, Goiânia GO, Brazil.

Correspondence: Gustavo Hayasaki Vieira; Email: gushayasaki@gmail.com.

Background: Viral encephalitis is a rare condition affecting approximately 1.4 individuals per 100.000 annually worldwide. It is characterized by inflammation of the brain parenchyma due to common viral infections in Brazil, such as dengue, Zika, and chikungunya. Given the high mortality associated with viral encephalitis and the lack of studies focusing on the Brazilian Midwest and, more specifically, the pediatric population, conducting an epidemiological analysis of mortality cases in this region is crucial. **Objective:** To analyze the epidemiological profile of mortality cases due to viral encephalitis in the pediatric population of the Brazilian Midwest from 2008 to 2023. **Methods:** This is a descriptive, retrospective, and quantitative epidemiological study based on data from the Hospital Information System (SIH/SUS) provided by the Department of Information Technology of the Unified Health System (DATASUS), regarding the mortality from viral encephalitis in pediatric patients from 2008 to 2023 in Goiás, Mato Grosso, Mato Grosso do Sul, and Distrito Federal. Filters applied included: ICD-10 morbidity list (viral encephalitis), midwest region, race/ethnicity, gender, age group, available period (January 2008 to December 2023), and year of processing. Data were tabulated in Excel and subjected to descriptive statistical analysis using R 4.40 with RStudio 2024.04.1 + 748. Since this study utilizes publicly available anonymous data, it is exempt from the Ethics Committee's approval. **Results:** During the study period, the average mortality rate was 2.85 %, with 2012 having the highest mortality rate (6.9%) followed by 2009 (6.45%). On the other hand, records from 2023 show the lowest mortality rate, 1.02%. Among the analyzed regions, Mato Grosso exhibited the highest mortality rate (4.12%), whereas Distrito Federal had the lowest (1.93%). Notably, individuals of indigenous ethnicity were most affected, with a mortality rate of 12.5%. Additionally, males showed a higher mortality rate compared to females, with rates of 3.49% and 1.95%, respectively. The most affected age group was 10-14 years, with a mortality rate of 4.62%. **Conclusion:** This study identified the indigenous population as vulnerable to viral encephalitis, likely due to geographical isolation and insufficient healthcare services available. Therefore, there is a clear need to invest in public health policies targeted at this social minority. It is also worth mentioning that Mato Grosso had the highest mortality rate, while the Federal District had the lowest. Furthermore, the highest mortality rates were concentrated about 10 years ago, notably in 2009 and 2012, while there is a decreasing trend in mortality rates, as demonstrated by the 2023 rate, the lowest recorded in the study. Given the data analysis, further studies are needed to investigate the factors influencing the correlation between mortality and viral encephalitis in the Brazilian Midwest regions.

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Mortality rate and average length of stay of hospitalizations for the treatment of polyneuropathies in Brazil by region in the last 6 years

André Luiz Borges Silva[1], Arthur Cauã Silva Santos[2], Igor José Balbino Santos[1], Isaias Felipe dos Santos[1], João Gabriel Dantas Cunha[2], Crisdan Cainã Costa Chagas[2], Helton Benevides Santana de Oliveira[1]

[1]Universidade Federal de Sergipe, São Cristóvão SE, Brazil.
[2]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Andre Luiz Borges Silva; Email: andre.borges297@gmail.com.

Background: Polyneuropathies refer to a group of conditions characterized by diffuse damage to peripheral nerves, typically associated with symmetric sensory loss and a length-dependent pattern. With a heterogeneous neurological presentation and potentially severe outcomes, they often require hospital admission for appropriate etiological investigation, treatment of underlying causes, and management of possible complications arising from the neurological condition. **Objective:** To compare the mortality rate and the average length of stay of hospitalizations for the treatment of polyneuropathies in Brazil by region over the past 6 years, in order to contribute to future strategies for addressing these diseases. **Methods:** This is a descriptive study with secondary data, retrospective and quantitative in nature. The data were obtained from the Department of Informatics of the Unified Health System (DATASUS). Mortality rates and average lengths of stay for hospitalizations due to polyneuropathies in Brazilian regions between 2018 and 2023 were analyzed. The data are public domain, hence there was no need for submission to the Ethics and Research Committee. **Results:** In the Northern region, the average annual mortality rate for hospitalizations for the treatment of polyneuropathies was 2.22. In the Northeast, this value was 2.22; in the Southeast, it was 2.26; in the South, 1.54; and in the Midwest, 2.80. Regarding the average length of stay per hospitalization, in days, the Northern region had an average of 8.93 days; in the Northeast, this average was 8.75; in the Southeast, 9.61 days; in the South, 6.90; and in the Midwest, 9.76 days. Considering that these values involve averages and percentage data, the total sample size (resident population) does not significantly influence the reliability of the data. **Conclusion:** The study showed significant regional variations in the mortality rate and the average length of stay for hospitalizations for the treatment of polyneuropathies in Brazil between 2018 and 2023. The Midwest region had the highest mortality rates and average lengths of stay, while the Southern region had the lowest numbers in both variables, indicating the need for interventions to reduce significant differences in the quality of care between different regions of the country. The limitations of the type of research conducted include the impersonal nature of the study, making it difficult to seek causality for the data found. Therefore, a recommendation for future studies is direct research to identify more specifically the needs of each region.

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Mortality rate in patients with neuromyelitis optica at a university hospital

Mackeyle Siqueira de Souza[1], Sávio Cesar Rodrigues Pestana[1], Vinícius Andreata Brandão[1], Igor Flavio Gonçalves do Vale[1], Victória Pagani Samora Sousa[1], Paula Zago Melo Dias[1], Bruno Batitucci Castrillo[1], Carlos Alberto Magirius Peixoto[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Igor Flavio Gonçalves do Vale; Email: igor.f.vale@edu.ufes.br.

Background: Neuromyelitis optica spectrum disorder (NMOSD) is an autoimmune condition characterized by acute attacks of optic neuritis and transverse myelitis, often associated with autoantibodies against aquaporin 4 (AQP4-IgG). Commonly used preventive therapeutics, including Azathioprine, Mycophenolate, and Rituximab, have helped control the disease by reducing immune system activity. Although these treatments have demonstrated clinical significance in the stabilization of NMOSD, their effectiveness is not absolute. **Objective:** Evaluate the mortality rate in patients with positive anti-AQP4 NMOSD at the university hospital **Methods:** This retrospective study was conducted at a public neurology outpatient clinic in Espírito Santo, Brazil, over a two-year period. Electronic medical records of patients diagnosed with NMOSD and receiving treatment were reviewed to assess the mortality rate within the local neurology service. **Results:** Of the 37 patients studied (6 men and 31 women), there were 2 deaths representing 5.4% of the cohort. One male and one female died after complications from a severe spinal cord attack. Both were using azathioprine and had therapeutic failure, and were awaiting release of rituximab. **Conclusion:** At the beginning of the neuromyelitis optica descriptions in the 1920s, mortality in 5 years was 32%. With preventive treatments, there was a significant reduction in mortality, which in our sample was 5.4%. However, it still represents a high mortality rate, similar to that of Guillain-Barré, demonstrating the need for public policy to treat this condition.

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Motor alterations in children with autism spectrum disorder level 3, according to the operationalized portage inventory: a cross-sectional study

Gabriela Batista Santos[1], Laércio da Silva Paiva[1], Débora Bachin Carvalho[2,3], Noelia Barbosa de Oliveira[3], Lívia Joela Almeida de Lima e Silva[3], Clara Criez de Lima Santos[3], Lucas Tomita Sodré[3], Marina Ortega Golin[1]

[1]Centro Universitário Faculdade de Medicina do ABC, Santo André SP, Brazil.
[2]Universidade Nove de Julho, São Paulo SP, Brazil.
[3]FisioPETI, São Paulo SP, Brazil.

Correspondence: Débora Bachin Carvalho; Email:

Background: Autism Spectrum Disorder (ASD) is a neurodevelopmental disorder classified into three levels of severity. Level 3 is the one that requires the most support, with the greatest restrictions on independence. In order to identify motor alterations in these children, it is necessary to use assessment scales. The most widely used in this population is the Operationalized Portage Inventory (OPI), which assesses six areas of development (gross and fine motor skills, cognition, socialization, language and self-care). **Objective:** To analyse overall motor performance of children with ASD level 3, according to the OPI, and to check the correlation with performance in language, cognition, self-care and socialization. **Methods:** This is a retrospective observational cross-sectional study analyzing 40 medical records of children aged between three and six years old diagnosed with ASD level 3, seen at FisioPETI Rehabilitation Clinic in São Bernardo do Campo / SP. The project was approved by the Research Ethics Committee of the Faculdade de Medicina do ABC University Center, protocol no. 70212423.6.0000.0082. The following data was collected: age; gender, Childhood Autism Rating Scale (CARS) score, classification of the development (in years) and quantification of developmental delay (in months) in each dimension of the OPI. Pearson's correlation test was used to relate the delay in overall motor performance to the other dimensions. The sample power calculation to quantify the delay in global motor performance was 100%. **Results:** Among the 40 patients, 62.50% were male, 97.50% showed delay in global motor development and self-care and 100% in language, cognition and socialization. The median age was 4.0 years old, global motor development, 2.1 (years); self-care, 2.2; socialization, 1.4; cognition, 1.2; and language, 0.4. The extent of delay in motor development (average) was 21.67 months; in self-care, 23.12; in socialization, 33.1; in cognition, 34.6 and language, 43.62. Delayed motor development showed a significant correlation with all other areas of development assessed by the OPI ($p<0.001$). **Conclusion:** Delayed global motor development was identified by the OPI in almost all children with ASD level 3. However, it was less severe, especially when compared to cognitive and language delays. The extent of developmental delay in all dimensions was correlated.

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Motor and respiratory function in young children with LAMA2 congenital muscular dystrophy

Mariana Cunha Artilheiro[1], Alulin Tácio Quadros Santos Monteiro Fonseca[1], Tatiana Ribeiro Fernades[1], Cristiane Araújo Martins Moreno[1], Umbertina Conti Reed[1], Edmar Zanoteli[1], Clara Gontijo Camelo[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Alulin Tácio Quadros Santos Monteiro Fonseca; Email: alulin.fonseca@gmail.com.

Background: The manifestation of weakness in the skeletal and respiratory muscles in patients with LAMA2 Congenital Muscular Dystrophy (LAMA2-CMD) typically occurs after birth or during early childhood. Data related to the young children population are less frequently published compared to older patients. However, assessments at young ages are crucial for prevention, early detection and treatment of complications. **Objective:** To identify motor and respiratory changes over the period of 2 years in young children with LAMA2-CMD. **Methods:** Eight LAMA2 children (6 female, 2 male) were assessed annually for a period of 2 years to monitor the evolution of their range of movement (ROM), motor function (Motor Function Measure - MFM D1, D2, D3 and total score -, and Revised Upper Lim Module - RULM) and respiratory function (measured by spirometry over 4 years old). The mean age for the first assessment was 3 - 1.75 years old and 4.62 - 1.19 years old for the second assessment. The analysis was conducted using descriptive statistics. **Results:** ROM: for hip extension, 4 children showed a worsening of 5-250, while 4 maintained their ROM; for knee extension, 4 children showed a worsening of 10-200 and 4 also maintained their ROM. Motor function: for upper limb function, 3 children showed a decrease of 3-9 points in the RULM score, while 3 children increased their RULM score by 3-6 points; two children maintained their score; for MFM D1 (standing position and transfers), 4 children showed worsening of 1-5%, 2 have increased the score between 4-8% and other 2 maintained their score; for MFM D2 (axial and proximal function), 7 children showed a decrease between 3-33%, while only one maintained their score; for MFM D3 (distal function), 3 children showed a decrease of 6-17%, while 3 children increased between 5-10% and 2 children maintained their percentages; for MFM total score, 4 children showed a decrease of 1-7%, while 2 children increased between 1-3% and 2 children maintained their percentages. Respiratory function: assessments were obtained only in 2 children; one child showed maintenance of the forced vital capacity (FVC) and another one increased the FCV in 5%. **Conclusion:** In two years, LAMA2 children between 2 and 6 years old showed the most evident deterioration in hip and knee ROM, as well as in the MFM D2 score. Upper limb function assessed by RULM and MFM D3 showed variable changes, which could either remain stable or change significantly, above 3 points up or down. On the other hand, MFM D1, MFM total score and respiratory function showed few significant changes over a 2-year follow-up.

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Motor neuron disease and Sjogren syndrome overlap: a case series

Mariana Hiromi Manoel Oku[1], Adriana Alves Rapp[2], Filipe Tupinamba Di Pace[1], Julia Cordeiro Braga[2], Marcel Leal Ribeiro[2], Marcela Camara Machado Costa[3], Diego Sant'ana Sodré[4], Acary Souza Bulle Oliveira[5], Edmar Zanoteli[1], André Macedo Serafim da Silva[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[2]Hospital Santa Izabel, Salvador BA, Brazil.
[3]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[4]Universidade Federal da Bahia, Salvador BA, Brazil.
[5]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Mariana Hiromi Manoel Oku; Email: mari.hiromi.mo@gmail.com.

Background: Sjogren syndrome (SS) is an autoimmune disease with a predilection for the peripheral nervous system that can precede in years systemic involvement. Motor neuron disease (MND) has been described in association with SS in a few case reports but it represents a rare presentation. **Objective:** To describe a case series of patients with an overlap between MND and SS. We discuss diagnostic clues and challenges, our treatment experience, and neurological response at follow-up. **Methods:** Data was collected from medical records between February 2021 and May 2024. We describe five patients, including epidemiologic and clinical characteristics, rheumatologic investigation, electromyography, magnetic resonance imaging (MRI), and treatment. **Results:** We identified five caucasian female patients with signs of MND and a concomitant diagnosis of SS after systemic review. Median age was 40 years old. Three developed both upper and lower motor neuron symptoms (LMN), the other two presented LMN symptoms only. Asymmetrical weakness was the initial symptom in three patients while the others had symmetric weakness. Two also displayed prominent bulbar and respiratory involvement. Electromyography of all patients showed signs of active denervation and reinnervation in at least one segment. All patients presented with Sicca Syndrome in addition to a positive Anti-Ro/SSA antibody and four met the European League Against Rheumatism (EULAR) diagnostic criteria by either having a confirmatory salivary gland biopsy and/or a positive Schirmer test. Three cases had a positive antinuclear antibody. Plexus MRI was performed in two patients, with plexus enhancement in one case and plexus thickening with no enhancement in the other. Cerebrospinal fluid (CSF) of three patients exhibited albuminocytologic dissociation (average protein level of 65 mg/dL). All patients received corticotherapy and immunoglobulin, three received rituximab, and one received cyclophosphamide. Disease progression was observed in three patients

despite any treatment whereas the other two had a partial response. The best outcome was observed in one patient who received immunoglobulin with no initial response followed by corticosteroids and rituximab, which led to neurological stabilization. She displayed only LMN symptoms, had plexus enhancement in MRI, and had the highest CSF protein level (99 mg/dL). The other patient who showed weakness improvement also had LMN symptoms exclusively and received corticotherapy and immunoglobulin. **Conclusion:** Sjogren syndrome seldom presents with MND. In our case series, most patients displayed both signs of upper and lower motor neuron symptoms. Response to immunotherapy was minor if compared to other neurological manifestations of SS, inferring a possible worse outcome related to this presentation. Further investigation is paramount for a better understanding of the association between the two diseases.

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Motor performance in children with autism spectrum disorder level 1, according to the portage inventory: a cross-sectional study

Vitória Hikari Ucima[1], Laércio da Silva Paiva[1], Débora Bachin Carvalho[2,3], Noelia Barbosa de Oliveira[3], Lívia Joelma Almeida de Lima e Silva[3], Clara Criez de Lima Santos[3], Lucas Tomita Sodré[3], Marina Ortega Golin[1]

[1]Centro Universitário Faculdade de Medicina do ABC, Santo André SP, Brazil.
[2]Universidade Nove de Julho, São Paulo SP, Brazil.
[3]FisioPETI, São Paulo SP, Brazil.

Correspondence: Débora Bachin Carvalho; Email: dbachinc@gmail.com.

Background: Autism Spectrum Disorder (ASD) includes individuals who have global neurodevelopmental disorder. The severity of the commitment in ASD is classified in three levels On level 1, the individual shows problems while interacting, but with considered ability to be reajusted to the social environment. The Operationalized Portage Inventory (OPI) assesses the functions performed by these children. Most studies report only cognitive and social alterations in this population. **Objective:** To analyse the global motor performance of children with ASD level 1, according to the OPI, and to check the correlation with language, cognition, self-care and socialization performances. **Methods:** This is a retrospective observational cross-sectional study, analyzing medical records from FisioPETI Rehabilitation Clinic, São Bernardo do Campo / SP. The project was approved by the Research Ethics Committee of the Faculdade de Medicina do ABC University Center, protocol no. 70199623.0.0000.0082. A total of 49 medical records was selected and the following data was collected: age, gender, Childhood Autism Rating Scale (CARS) score, developmental classification (in years) and quantification of developmental delay (in months) in each dimension of the OPI. Pearson's correlation test was used to relate the delay in overall motor performance to the other dimensions. The power of the sample to quantify delay in overall motor performance was calculated at 100%. **Results:** Among the 49 children, 77.55% were male, 97.96% had delay in overall motor development, 100% in language development, 95.92% in cognition, 100% in socialization and 95.92% in self-care. The average age was 4.04 years old; CARS score, 25.32; global motor development, 3.22 (years); self-care, 2.93; cognition, 2.64; socialization, 2.36 and language, 1.80. The average delay severity was: 11.53 months in global motor development, 14.91 in self-care, 18.48 in cognition, 21.83 in socialization and 28.67 in language. A significant correlation was found between the severity of the delay in global motor development and the other areas of development ($p<0.001$). **Conclusion:** The majority of children with ASD level 1 showed delay in motor development, but with less severity than in other areas of development. The areas with the highest proportion of impairment and extent were language and socialization. The severity of motor impairment was correlated with that of language, socialization, cognition and self-care.

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Motor unit number index (MUNIX) as a biomarker for disease progression in late-onset 5q-spinal muscular atrophy treated with nusinersen

Rodrigo de Holanda Mendonca[1], Jose Pedro Soares Baima[1], Antonio Edvan Camelo Filho[1], Graziela Jorge Polido[1], Carlos Otto Heise[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.

Correspondence: Rodrigo de Holanda Mendonça; Email: rodrigo.holanda@hc.fm.usp.br.

Background: The Motor Unit Number Index (MUNIX) has emerged as a more sensitive biomarker for detecting motor unit loss in 5q-spinal muscular atrophy (SMA) compared to compound muscle action potential (CMAP). Recent studies have shown correlations between MUNIX values of various muscles and motor scales used to assess disease progression and treatment response. **Objective:** This study is an investigation of the feasibility of MUNIX as a biomarker for disease progression in SMA types 3 and 4 treated with nusinersen. **Methods:** A four-year study involved 20 patients diagnosed with SMA types 3 and 4, ten treated with nusinersen and ten non-treated. Patients underwent evaluations using the Hammersmith Functional Motor Scale Expanded (HFMSE), the 6-minute walk test (6MWT), and assessments of CMAP and MUNIX in the abductor policis brevis (APB), adductor digiti minimi (ADM), and tibialis anterior (TA) muscles. **Results:** A significant change was observed between the first and fourth years in CMAP values of the ADM muscle ($p=0.012$) in treated patients. In contrast, untreated patients experienced a significant reduction in mean CMAP values of the TA ($p=0.015$). Significant decreases in mean MUNIX values were found only in untreated patients, in the ADM ($p=0.022$) and TA ($p<0.001$). Treated patients experienced an average increase of 24.9 points in the MUNIX of the ADM ($p=0.024$) compared to non-treated patients. CMAP and MUNIX values for the TA muscle correlated with HFMSE scale results. **Conclusion:** The MUNIX values of ADM and TA muscles are biomarkers for disease progression in 5q-SMA types 3 and 4 over a four-year follow-up period. Despite a significant increase in ADM CMAP values among treated patients, only ADM MUNIX could identify motor unit loss in untreated patients. Furthermore, MUNIX values remained stable among treated patients over the four-year follow-up period.

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Movement disorders in patients with systemic lupus erythematosus: cross-sectional study

Camilla Vanessa Araújo Soares[1], Tiago César Costa da Silva[1], Alessandra de Sousa Braz[2], Danielle Christinne Soares Egypto de Brito[2], Ana Karla Guedes de Melo[1], Adriana Meira Tiburtino Nepomuceno[1], Alex Tiburtino Meira[2]

[1]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[2]Universidade Federal da Paraíba, Departamento de Medicina Interna, João Pessoa PB, Brazil.

Correspondence: Camilla Vanessa Araújo Soares; Email: camilla.vanessa46@gmail.com.

Background: Systemic lupus erythematosus (SLE) is an autoimmune, multisystemic disease characterized by autoantibodies production and immune complexes deposition, triggering an multiorgan inflammatory process. Approximately 50% of patients with SLE develop neuropsychiatric symptoms, including movement disorders (MD). **Objective:** To investigate the prevalence of MD in patients with SLE, in a SLE outpatient clinic in a quaternary hospital in the state of Paraíba. **Methods:** This is a cross-sectional observational study, carried out in a quaternary hospital in the state of Paraíba between September 2023 and May 2024. Patients were selected at random from the SLE outpatient clinic. Seventy individuals fulfilled the inclusion criteria, which encompassed a confirmed diagnosis of SLE for a minimum duration of one year as per the American College of Rheumatology's standards. Additionally, participants were required to be 18 years of age or older and under regular monitoring by the Rheumatology team within the service. One patient was excluded due to a previous history of subarachnoid hemorrhage and two patients refused to be examined by a neurologist. **Results:** 67 patients were included in the final sample, the majority of whom were female (90%). The prevalence of neurological changes related to movement disorders detected on physical examination was 35.3% (n 24). No problems of coordination and balance were found. Action and postural tremor were the most prevalent (n 16; 23.5% of cases), probably secondary to hydroxychloroquine in 15 of them (mostly action worse than postural) and 1 case secondary to levothyroxine. Seven patients presented a discrete rigidity, although only 2 with bradykinesia. Only one patient presented spasticity. Despite being the well-known movement disorder associated with SLE, chorea was only found in 2 patients (2.9%), one presented generalized, including the hung-up knee jerk, and the other presented hemichorea. One patient presented myoclonic jerk movements on the foot, discrete and only with distraction (1.49%). Except for one patient within the tremor subgroup, the majority experienced mild tremors. No drug intervention was necessary in most cases, given the mild nature of the symptoms and their lack of impact on daily activities, except in two cases of parkinsonism and in one patient with hemichorea. One patient presented considerable exacerbation of essential tremor after the use of levothyroxine. **Conclusion:** MD has a prevalence higher than expected in patients with SLE (35.3%), with drug-induced tremor being the most prevalent, followed by chorea and parkinsonism. In light of this, it is imperative for healthcare professionals assisting these patients to recognize the presence of MD, enabling tailored management strategies to enhance their quality of life.

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Moyamoya disease in a patient with previous bilateral carotid atherosclerosis and who underwent radiotherapy after breast cancer: a case with an uncommon evolution

Elvis Lucas Rabelo Shintay Chan[1]

[1]Universidade Federal de Ouro Preto, Ouro Preto MG, Brazil.

Correspondence: Elvis Lucas Rabelo Shintay Chan; Email: elvislucaschan2011@hotmail.com.

Case presentation: A 41-year-old pregnant woman receiving usual risk prenatal care is admitted to labor, which occurs uneventfully, on 01/26/2018. On the fifteenth day of the postpartum period, a sudden onset of paresthesia and hemiparesis in the right side began, but she did not seek medical attention because she thought the cause had been a "scare" and the symptoms resolved spontaneously. On 06/23/2019, he presented new hemiparesis in the right side, in addition to dysarthria with deviation of the lip commissure. The magnetic resonance imaging showed an ischemic stroke in the perirolandic regions

and left superior frontal gyrus, as well as marked thinning of the petrous and cavernous parts of the left internal carotid artery and prominence of the pericarotid leptomeningeal circulation ("ivy sign"), a pattern suggestive of Moyamoya disease. She was discharged, persisting only with grade IV hemiparesis and braquicrural on the right. On 12/17/2023 she presented a third ischemic event with worsening of hemiparesis and dysarthria, with computed tomography showing hypodensity in the high brain convexity bilaterally and in the left semi-oval center. She maintained sequelae of dysarthria, urinary incontinence and right crural monoparesis, having been approached by Neurosurgery only in 2024. **Discussion:** Moyamoya disease has a well-documented association with genetic factors, especially in Asian populations. Although the patient was not of Asian origin, she had a family history of stroke, but without further investigation. Otherwise, it is worth highlighting that there are a series of conditions associated with this pathology, such as trauma, meningitis and neoplasms, as well as two others that were present: atherosclerosis (which is bicarotid) and exposure to radiotherapy (the patient had a malignant neoplasm of breast in 2015). Regarding imaging exams, there are typical findings: stenosis in the final portion of the internal carotid artery; a collateral and dilated arterial network branching from the circle of Willis; and evidence of previous small cerebral infarcts. The age distribution of the incidence is bimodal, with a peak at 10 years old and the other at 40 years old. The diagnosis is often made after a stroke. However, the disease can progress with convulsions or remain asymptomatic. Surgical revascularization is indicated, but it depends on weighing risks and benefits. **Final comments:** Moyamoya disease, although low in incidence, is a pathology of great repercussions as it can lead to recurrent strokes, causing great harm to the individual. The importance of speed in investigation and treatment is essential to prevent new deficits. The genetic association makes it necessary to pay attention to neurological events in family members, which are often little investigated. Furthermore, this case demonstrates a possible late side effect of radiotherapy, which illustrates the importance of being aware of the association between neoplasms and strokes.

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Moyamoya disease in early puerperium: a case report

Gabriel Abrahão Stoliar[1], César Minoru Toita Koga[1], Samia Talise El Horr de Moraes[1], Alexandre Henrique Scheibe[1], Alana Bacelar Limeira Sales[1], Talita Aparecida Conte[1], Gabriella Favero[1], Valéria Cristina Scavasine[1], Caio César Diniz Disserol[1,2,3]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[2]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[3]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Gabriel Abrahão Stoliar; Email: gabrielstoliar@gmail.com.

Case presentation: A 23-year-old female patient, born in Venezuela, on the 2nd day of postpartum post-vaginal delivery, presented with a sudden onset of right-sided hemiparesis and confusion, followed by a short-lasting focal epileptic seizure, progressing to refractory status epilepticus. Cranial magnetic resonance imaging demonstrated acute ischemia in both cerebral hemispheres. Cranial and cervical tomography angiography with occlusion of the intracranial portion of the internal carotid artery from the paraophthalmic segment with extensive collateral circulation and prominence of pial vessels, suggesting an appearance of Moyamoya syndrome. Cerebral angiography confirmed the findings. **Discussion:** Moyamoya syndrome is a cerebrovascular disorder associated with progressive stenosis of the internal carotid arteries and their proximal branches. As a result, an extensive network of collateral is developed. The main clinical manifestations are neurovascular injuries. Moyamoya is an angiopathy represented by Moyamoya disease and Moyamoya syndrome. It mainly affects people of Asian origin. It has incidence in 2 age groups (children aged 5 years and adults around 40 years old), affecting almost twice as many female patients as male patients. Approximately half of adult patients present with intracranial hemorrhage and half with ischemic complications. Pregnancy, including the prenatal, intrapartum and postpartum periods, is probably associated with the risk of hemorrhagic and ischemic stroke in women with Moyamoya disease, with the onset of the condition more frequently in the postpartum period, especially among 3 and 7 days after birth. On cerebral angiography, a distinct appearance characterized by stenosis of the distal intracranial internal carotid artery, extending to the proximal anterior and middle arteries. This stimulates a development of collateral vessels at the base of the skull, which have a "smoke breath" appearance (translating as moyamoya in Japanese) and are therefore called moyamoya vessels on cerebral angiography. **Final comments:** Moyamoya disease is a rare condition, but represents an important differential diagnosis of stroke in young people. Its neuroradiological characteristics are remarkable and its early identification can help in the management and prognosis of patients.

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MS treatment in Brazil in 2022: medication dispensation by the public health system using big data

Alexandre Coelho Marques[1], Marina Martins Siqueira[1], Gabriely Rangel Pereira[1], Lucas Hernandes Corrêa[1], Victoria Lin[1], Luiz Eduardo Betti Paiva[1], Lívia Almeida Dutra[1]

[1]Faculdade Israelita de Ciências da Saúde Albert Einstein, São Paulo SP, Brazil.

Correspondence: Alexandre Coelho Marques; Email: acmarques.neuro@gmail.com.

Background: Multiple sclerosis (MS) is a chronic inflammatory disease. Within a decade of onset, about 50% of those who do not receive any treatment develop severe disabilities. Disease-modifying drugs (DMDs) prevent disabilities. Treatment modalities include escalation (starting with low-efficacy DMDs and escalating to high-efficacy DMDs) or initiating with high-efficacy drugs, which carry risks such as opportunistic infections and the need for infusion units. High-efficacy drugs like natalizumab and alemtuzumab are linked with better outcomes and are available through Brazil's Unified Health System (SUS), though they require administration at specialized centers. **Objective:** To describe the dispensation of MS medications by SUS in Brazil in 2022 and analyze regional differences. **Methods:** A retrospective, observational study using public databases from Data SUS was conducted to analyze patient demographics and MS medications dispensed. Data were extracted from the Authorization of Ambulatory Procedures (APAC) for MS drugs in 2022. All APACs dispensed in Brazil during this period were analyzed using a Big Data system for data extraction, processing, and modeling. **Results:** In 2022, 28,012 MS patients received treatment by SUS, with 72.4% being female. The regional distribution was as follows: Southeast (16,006 patients, 57%), South (5,840, 21%), Northeast (3,456, 12%), Midwest (2,230, 8%), and North (480, 2%). The highest patient prevalence was in the South (19.8 per 100,000 inhabitants), while the lowest was in the North (2.8 per 100,000 inhabitants). A total of 268,814 DMDs were dispensed, corresponding to the patient distribution across regions. There were 194 dispensation centers, with half of the states having only one center. Fingolimod, dimethyl fumarate, and interferons were the most dispensed drugs, at 22.3%, 20.2%, and 19.4% respectively. There were regional differences; interferons were predominantly dispensed in the North and Northeast, whereas natalizumab were most dispensed in three specific states (DF, CE, and SE). High-efficacy drug dispensations totaled 44,650 (16.6%), with alemtuzumab accounting for only 9 treatments. The median distances traveled by patients from their municipality of residence to the municipality of DMD dispensation were 84.2 km (±149.8) in the Midwest, 77.6 km (±253.2) in the North, 63.4 km (±113.7) in the South, 81.2 km (±232.0) in the Northeast, and 29.5 km (±70.4) in the Southeast. **Conclusion:** Patients with MS treated by SUS receive less high efficacy medications when compared to other countries. Most patients travel more than 100 km to receive medications, which could be a barrier to high efficacy medications. Regional differences highlight the need for greater attention to ensure equitable access to MS medication.

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Mucormycosis as a cause of extrinsic eye musculation impact: rhino-cerebral form

Lorena Fernandes Kronbauer[1], Letícia de Barros Dias[1], Marcelo Simplicio Carvalho[1], Nathalye Fernanda Pedroso Dircksen[1], Mônica Meneguette Silveira[1], Bárbara Okabaiasse Luizeti[1], Damacio Ramon Kaimen Maciel

[1]Irmandade da Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Lorena Fernandes Kronbauer; Email: lorenafkronbauer@gmail.com.

Case presentation: Male, 51 years old, 10 days after admission, right eye movement began with pain, of severe intensity, with a progressive decrease in visual acuity until reaching amaurosis, ptosis and ophthalmoplegia. There was weight loss of 12kg in 60 days. Previous pathologies: only diabetes mellitus. Ophthalmological examination: RE with papillary edema, macular hypopigmentation, fixed mydriasis, acuity without light perception or counting fingers. Magnetic Resonance Imaging of the orbits showed an infiltrative mass compromising the intraconal fat of the right orbit with slight contrast enhancement. Orbital tomography demonstrated a bulging nodular lesion in the left maxillary sinus, areas of bone discontinuity in the ethmoidal fovea and cribriform plate on the right, ethmoidal sinusopathy and mucous thickening of the right maxillary sinus. The patient underwent pulse therapy with corticosteroids, without clinical or radiological improvement. A biopsy of the right extraocular muscle was performed, with a positive result for mucormycosis. The patient was treated with Voriconazole and Amphotericin and referred to the responsible center for treatment of his pathology. **Discussion:** Mucormycosis is an infection caused by fungi from the Mucoraceae family - saprophytes often found in soil. Tissue invasion and rapid dissemination of the pathogen occurs in deep tissues, giving rise to an intense inflammatory process and tissue necrosis, especially in immunosuppressed patients. Epidemiology: affected age group is under 60 years old, but with more mortality in the elderly - 33.3%. Predominance of males, but with no significant difference in mortality between the sexes. Ocular symptoms, with edema and pain, are the most common symptoms, associated with paranasal sinuses involvement. Around 94% patients are diabetic. Patients who receive high doses of corticosteroids and when preceded by COVID-19 infection, have relatively higher mortality rates. The clinical forms: rhinocerebral, pulmonary, gastrointestinal, central nervous system (CNS), subcutaneous and disseminated form. The form of rhino-cerebral mucormycosis (RCM) is the most common of these forms and is subdivided into three types: rhino-maxillary, rhino-orbital and rhino-orbito-cerebral. Treatment must be aggressive, given its lethal nature, and involves treatment of the underlying cause (diabetes mellitus with or without diabetic ketoacidosis or neutropenia), appropriate antifungal

therapy, mainly with liposomal amphotericin B, and surgical debridement of the tissues involved. **Final comments:** Mucormycosis is a serious fungal infection, with classic involvement involving the sinuses, orbits and brain. Generally associated with immunosuppression, diabetes, and appears to have increased severity with recent COVID-19 infection. The most common form - rhinocerebral - was the one described in this case, with bilateral sinus involvement, but with involvement only of the ocular muscles on the right.

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Multidimensional functional decline throughout Parkinson's disease development

Nathalia de Brito Pereira[1], Katia Cirilo Costa Nobrega[1], Isaíra Almeida Pereira da Silva Nascimento[1], Rafaela Pires Bocicovar[1], Carolina Gonçalves Santana[1], Letícia Rocha Matos[1], Fernando Fachine da Silva Fidelis[1], Luiza Mattos Aranha[1], Graziele Cardoso dos Santos[1], Maria Elisa Pimentel Piemonte[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Nathalia de Brito Pereira; Email: nathaliabpereira@usp.br.

Background: To manage diseases such as Parkinson's Disease (PD), guide new intervention frameworks and inform patients, it is important to identify the decline in different dimensions of functionality. The functionality analyzes the complex interaction among biological, psychological, and social factors, that is why it is one of the main health indicators. The World Health Organization based on International Classification of Functioning, Disability and Health (ICF), developed the World Health Organization Disability Assessment Schedule 2.0 (WHODAS 2.0), a generic assessment instrument to provide a method for measuring the impact of different health conditions in terms of functionality and disability across cultures. **Objective:** A cross-sectional study was carried out to measure how functional decline is associated with the progression of PD using WHODAS 2.0. Furthermore, a characterization of the participant functionality profile was made. **Methods:** There were 352 participants (55% male, 45% female), assessed with early to advanced PD (H&Y I-IV), aged between 35-85 years age, using dopaminergic medication and without other neurological disorders than PD. The instruments used were: the Telephone - Montreal Cognitive Assessment (T-MoCA), Movement Disorder Society – Unified Parkinson's Disease Rating Scale (MDS-UPDRS) Part I - II and WHODAS 2.0. **Results:** The descriptive analysis indicated that the most affected WHODAS 2.0 domains were domestic activities (ADL) representing 46% of the total score and subsequently mobility and participation representing 43% of the total score. The motor activities where participants found the most difficulties were: carrying out household tasks as quickly as possible (87%), getting dressed (74%), and getting up from a sitting position (71%). Furthermore, the time spent on healthcare and the emotional impact caused by the health condition also created difficulties for the participants, with 77% and 72% complaining respectively. In the Mobility, ADL, Self-Care and Participation domains, there is a significant difference between all H&Y stages confirmed by the post-hoc test. However, for the total scores of the Interpersonal Relationships, Cognition and ADLw (activities of daily living related to work and school) domains, there was no significant difference between H&Y I and II according to it. Besides that, a high positive correlation was found between WHODAS ADL with MDS-UPDRS I (R=0.53; p-value<0.00001) and MDS-UPDRS II (R=0.65; p-value<0.00001), WHODAS Self-Care and MDS-UPDRS II (R=0.82; p-value<0.00001), and between WHODAS 2.0 total scores and MDS-UPDRS I (R=0.67; p-value<0.00001) and MDS-UPDRS II (R=0.68; p-value<0.00001). A negative correlation was found between WHODAS 2.0 Cognition and T-MoCA (R=0.33; p-value<0.0001). **Conclusion:** Pharmacological and non pharmacological interventions should consider the clinical characterization of the different affected dimensions of functionality in people with PD.

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Multifocal and longitudinal extensive transverse myelitis after Varicella zoster virus infection: a case report

Jessica Vaz de Melo[1], Lucas Alexandre Santos Marzano[1], Luiza Fontes e Souza[1], Giovanna Moraes Cardoso Oliveira[1], Luiz Felipe Lopes Campos[1], Vinícius Rezende Avelar[1], Mariana Figueiredo Miranda[1], Thiago Michael Ferreira Ibiapino[1], Pedro Henrique Dias Silva[1], Rodrigo Gonçalves Kleinpaul Vieira[1]

[1]Hospital Governador Israel Pinheiro, Belo Horizonte MG, Brazil.

Correspondence: Lucas Alexandre Santos Marzano; Email: lasmarzano@gmail.com.

Case presentation: We report an unusual case of transverse myelitis (TM) in a 65-year-old immunocompetent woman occurring two weeks after a varicella zoster virus (VZV) infection. The patient presented with painful vesicular eruption on her back involving the right T8-T10 dermatomes, consistent with VZV infection and started treatment with topical acyclovir. Two weeks later, she developed right inguinal paresthesia progressing to bilateral leg involvement, paraparesis and urinary retention. Upon admission, neurological exam revealed bilateral lower limb weakness graded 4 on the Medical Research Council. Sensory system examination indicated decreased pinprick, touch, temperature, joint position and vibration sensation below the T10 level bilaterally. Deep tendon reflexes were exaggerated at the knee and ankle joints. No further neurological abnormalities were observed. Cerebrospinal fluid analysis demonstrated elevated leukocyte counts with normal glucose and mildly elevated protein. Magnetic resonance imaging of the spine revealed hyperintense T2 lesions at C5-6 and T6-10 levels, the first one with gadolinium enhancement on T1 sequence. No lesions were identified in the brain or optic nerves. The patient was treated with intravenous acyclovir at 10 mg/kg every 8 hours for 14 days and methylprednisolone 1g every 8 hours for 5 days. Upon discharge, there was no immediate improvement in the neurological deficits, however, at one-month follow-up, significant improvement was noted in both motor strength and sensory function. **Discussion:** VZV is known to be responsible for a broad spectrum of neurological diseases, but TM is a rare and severe complication. It occurs in less than 1% of VZV reactivation situations and usually affects immunocompromised patients. It typically manifests days after the onset of the rash, although delayed presentations may occur even in the absence of active skin lesions. While there is usually a correlation between the dermatomal rash and the motor weakness, this is not always the case, and there may be a topographic dissociation in around 10% of cases. In our case, two peculiarities were observed: multifocal and longitudinally extensive spinal cord involvement. The pathogenesis of VZV-related TM is postulated to be related to direct viral invasion. Vascular and autoimmune mechanisms have also been proposed in its pathophysiology. Treatment strategies often involve intravenous acyclovir, corticosteroids and symptomatics. **Final comments:** This case underscores the importance of early recognition and comprehensive management strategies in improving outcomes for TM secondary to VZV infection. Early therapeutic intervention with acyclovir prevents multiplication and spread of the virus in the spinal cord contributing to good prognosis.For this reason, correct identification and early treatment are very important.

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Multifocal motor neuropathy: a two-decade case report

Gabriel Belo Cavalcante Linhares[1], Samila Marissa Pinheiro Gomes[1], Aglagilson Fernandes das Chagas[1], Yasmin Luíse Lucena e Silva[1], Moisés Vasco de Freitas[1], Emanuela Maria de Aguiar Azevedo[1], Jonnathan Camara da Silva Vianna[1], Eduardo Martiniano de Lima[1]

[1]Universidade Federal Rural do Semi-Árido, Mossoró RN, Brazil.

Correspondence: Gabriel Belo Cavalcante Linhares; Email: gabriel.belo@alunos.ufersa.edu.br.

Case presentation: Multifocal motor neuropathy (MMN) is a rare autoimmune disease characterized by progressive and asymmetric muscle weakness due to conduction blocks in motor nerves. This report details a case initially diagnosed as Guillain-Barré Syndrome (GBS), but later identified as MMN following further testing. Patient A.J.G.X., a 51-year-old female, presented rapidly progressing tetraparesis with predominant right-side weakness in 2004. After treatment with human immunoglobulin and physiotherapy, she partially regained mobility, though significant improvement was not observed. In 2017, her symptoms worsened, peaking in 2021, after which she required assistance for walking and daily activities. Her family history includes an uncle with GBS. The patient, a former alcoholic, quit drinking five years ago. A neurological examination revealed grade IV strength in the upper limbs and proximal left lower limb, grade III in the proximal right lower limb, and grade 0 in bilateral plantar dorsiflexion. Patellar and Achilles reflexes were absent bilaterally. There was bilateral rigidity (1+/4+) and slight postural tremor in the lower limbs, with a resting tremor in the right hand. Imaging and neurophysiological tests indicated cerebellar tonsillar herniation (Chiari type I). In 2023, electromyography confirmed multifocal motor neuropathy with proximal motor conduction block in the bilateral median and ulnar nerves, and abolished motor potentials in the bilateral fibular nerves. After undergoing immunoglobulin therapy, consisting of three monthly sessions at a dose of 2g/kg intravenously each, the patient reported clinical stability but no significant improvement in weakness. She could walk with assistance only for short distances. She experienced occasional cramps and resting and kinetic tremors, especially in the right hand, with slight improvement. She currently attends motor physiotherapy twice a week. **Discussion:** MMN is a rare and diagnostically challenging condition. In this case, GBS was initially the main diagnosis. However, progressive motor deficits and lack of stabilization increased suspicion of MMN, confirmed by electromyography in 2023. This report underscores the need for heightened suspicion for pathologies that, despite initially resembling GBS, show continuous symptom progression. It is crucial for neurologists to consider MMN in the differential diagnosis of motor neuropathies with persistent progression to avoid treatment delays and improve patient outcomes. **Final comments:** This case highlights the complexity of differential diagnosis among progressive motor neuropathies, where MMN should be considered in patients with asymmetric and progressive motor weakness that does not stabilize. Understanding and differentiating MMN from other neuropathies is essential to avoid delays in diagnosis and improve patient prognosis, emphasizing the importance of detailed clinical evaluation and the judicious use of complementary tests.

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Multifocal motor neuropathy: different clinical presentation of three cases

Danilo Marinho Pereira[1], Ana Marina Dutra Ferreira da Silva[1], Alessandra Lima Nogueira Tolentino[1], Igor Dias Brockhausen[1], Alzira Maria Siqueira de Carvalho[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Danilo Marinho Pereira; Email: danilomarinho14@gmail.com.

Case presentation: We present three male patients with different manifestations of multifocal motor neuropathy. Patient 1: 56-year-old male with history of right upper limb weakness starting at age 31, progressing to weakness in both upper limbs over two years, followed by weakness in the lower limbs with a distal predominance after three years. Motor nerve conduction studies showed conduction block and uneven reduction in conduction velocity in all four limbs. Myography revealed positive sharp waves, fibrillations, and rarefaction of motor units' recruitment. Tests for GM1 antibodies were negative. Patient 2: 58-year-old male experiencing since the age of 45 weakness in the right lower limb, progressing within a year to bilateral weakness with drop feet. Nerve conduction studies indicated reduced motor conduction velocity in all four limbs with conduction block and no changes in the sensory conduction study. Myography showed fasciculations and rarefaction of motor units' recruitment. Patient 3: 58-year-old male with a seven-year history of distal weakness in the upper limbs, accompanied by cramps and pain. The first motor conduction study was unremarkable, and myography indicated chronic and acute denervation in four limbs compatible with motor neuron disease. After 01-year, the nerve conduction study was repeated in our service, showing motor conduction blocks, while sensory conduction remained normal. Myography during voluntary contraction indicated signs of chronic reinnervation in the proximal and distal muscles, with rarefaction of motor units in all four limbs. Complete exome sequencing revealed an alteration in the TTN gene variant chr2: 178,560,016 G>A, unrelated to the clinical findings. All three patients underwent treatment with intravenous human immunoglobulin and showed satisfactory symptom response. The treatment began at 25, 13, and 7 years after the onset of symptoms, respectively. **Discussion:** Multifocal motor neuropathy (MMN) is an uncommon, purely motor neuropathy characterized by asymmetric deficits with a preference for upper limb involvement. However, it is crucial to emphasize the importance of electrodiagnostic examination in the presence of atypical clinical presentations of the disease. **Final comments:** Multifocal motor neuropathy is a treatable condition. However, it is challenging to diagnose and can be confused with motor neuron disease (ALS and its variants). Hence, early diagnosis and treatment are crucial for improving the prognosis. It is important to note that symptom improvement may occur after many years of illness.

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Multiple ischemic infarcts mimicking Opalski syndrome

Heron Alves Vale[1], Raissa Josefa Pereira de Moura[1], Mayhara Rosany da Silva Santiago[1], Nereu Alves Lacerda[1], Ana Carolina Martins Fragoso Carneiro[1], Maria Luisa Roberto Lima Lisboa[1], Natália Nunes Tenório[1], Ana Beatriz Roberto Lima Borba[1], Humberto Cabral de Oliveira Filho[2], Igor Figueiredo Gonçalves[1]

[1]Hospital Pelópidas Silveira, Recife PE, Brazil. [2]Universidade Federal do Rio Grande do Norte, Escola Multicampi de Ciências Médicas, Caicó RN, Brazil.

Correspondence: Heron Alves Vale; Email: heron_vale@hotmail.com.

Case presentation: A 50-year-old male patient was admitted to the neurology emergency department due to a sudden onset of motor and sensory deficit syndrome, associated with ataxia and dysphagia. His symptoms included: right ptosis, vertigo, vomiting, dysarthria, dysphonia, hypoesthesia in the right hemiface and left half of the body, peripheral pattern facial paralysis on the right, right hemiparesis, and hemiataxia. His medical history revealed hypertension, alcohol abuse, severe obesity, and smoking. He denied regular medication use. There was a positive family history of cardiovascular diseases, with his mother passing away at 52 years from acute myocardial infarction. Laboratory findings showed relative polycythemia (Hemoglobin 19.5g/dl). Cranial CT revealed hypodensity in the right cerebellar and lower bulbar regions. Angiotomography showed occlusion of the distal segment of the right vertebral artery and ipsilateral posterior inferior cerebellar artery. Further diagnostic assessment with MRI demonstrated multiple recent ischemic insults in the right caudate nucleus, right corona radiata, left centrum semiovale, and a subacute event in the right cerebellar hemisphere. **Discussion:** The array of clinical findings upon admission raised suspicion of Opalski syndrome, a rare manifestation of Wallenberg syndrome, characterized by ipsilateral hemiplegia/paresis with lateral bulbar syndrome. Wallenberg syndrome typically results from ischemic involvement of the arterial branches supplying the dorsolateral portion of the medulla, marked by ipsilateral limb ataxia, vertigo, vomiting, nystagmus, dysarthria, dysphonia, dysphagia, and possibly ipsilateral facial hypoalgesia, thermoanesthesia, and Horner syndrome. **Final comments:** The presentation of ipsilateral motor deficit associated with dorsolateral medullary syndrome suggests involvement of medullary penetrating arteries, branches of the vertebral artery supplying the pyramidal fibers post-decussation, potentially characterizing Opalski syndrome. However, the involvement of the facial nerve would not typically be explained by this syndrome. In this context, MRI is crucial for definitive diagnosis. In this case, confirmation of vascular territories and distinct chronologies suggests a possible thromboembolic etiology.

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Multiple mononeuropathy in eosinophilic granulomatosis with polyangiitis: a case report

Manoela Moreira Guerreiro[1], Maria Fernanda Batalha Marques[1], Maria Eduarda Vieira Ribeiro Garcia[1], Elisa Fernandes Melo[1], Karolina Gouveia César Freitas[1]

[1]Universidade de Taubaté, Taubaté SP, Brazil.

Correspondence: Manoela Moreira Guerreiro; Email: manoelamguerreiro@gmail.com.

Case presentation: Male patient, 44 years old, asthmatic for 4 years, without other comorbidities, sought care reporting paresthesia in the left lower limb that began in the tibial region, extending to the foot. After two days, the symptom spread to the right lower limb and upper limbs, reporting loss of strength and allodynia in all four limbs. The condition was preceded by a fever of 38°C lasting two days, two bacterial pneumonias and two dengue fever events in the last 60 days. He was admitted to the nursery with an initial suspected diagnosis of Guillain-Barré Syndrome, but after being examined the syndromic condition became mononeuritis multiplex. Examinations showed hypereosinophilia (2,352), negative serology, unremarkable cerebrospinal fluid, non-reactive anti-cytoplasmic antibodies (ANCA), increased rheumatoid factor (16), negative ANA and increased ESR in the first hour (62). After three days, he continued to complain of decreased strength, paresthesia and allodynia. On physical examination, he presented diffuse subcrepitant rales on lung auscultation, multiple diffuse erythematous non-pruritic papules on his back, he was alert, oriented, without language and speech changes, with a mild paretic gait on the left, proximal grade IV strength in the lower limbs III/IV distal on the left and IV on the right, patellar, achilleus and styloradial reflexes were absent, vibratory hypoesthesia in the left lower limb, without other changes. The hypothesis of Eosinophilic Granulomatosis with Polyangiitis (EGPA) was considered based on signs, symptoms and late-onset asthma; thereforethe treatment was initiated with prednisone and cyclophosphamide. He was discharged after fourteen days of hospitalization, with improvement in lung auscultation and complaints of pain, reporting only maintenance of paresthesia. Continued on outpatient follow-up and electroneuromyography was performed, which showed diffuse and incipient impairment of peripheral sensory-motor nerve fibers, primarily axonal, asymmetrical and subacute, corroborating the clinical suspicion of vasculitis. DISCUSSION:It is very important to investigate rheumatological conditions such as EGPA in the presence of mononeuritis multiplex. EGPA belongs to the group of ANCA-associated vasculitis,although considered rare, it is the most common cause of neuropathy, characterized by the clinical triad asthma, hyper eosinophilia and systemic necrotizing vasculitis affecting small vessels. Peripheral neuropathy may be present in up to 72% of cases, expressed as multiple mononeuropathy or peripheral polyneuropathy. **Final comments:** This clinical case portrays the importance of early diagnosis of EGPA and the relevance of identifying peripheral nervous involvement in this process. Therefore, recognizing the clinical pattern of this type of peripheral neuropathy is essential for preventing comorbidities, reducing mortality, reducing neurological sequelae and consequently improving the patient's quality of life.

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Multiple sclerosis (MS) and MOGAD familial cases share genetic variant rs118004742 in EFCAB13 previously associated as potential MS risk factor

Karla Pinto Coelho Fadel[1,2], Renan Amphilophio Fernandes[2,3], Elielson Veloso da Silva[2], Andreza Lemos Salvio[2], Jessica Raposo Vedovi[2], Larissa Araujo Duarte[2,3], Helena França Alcaraz Ferreira[2,3], Elisa Gouvea Gutman[2,3], Valéria Coelho Santa Rita Pereira[3], Soniza Vieira Alves-leon[2,3]

[1]Universidade Federal do Estado do Rio de Janeiro, Centro de Ciências Biológicas e da Saúde, Programa de Pós-Graduação em Neurologia, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Instituto Biomédico, Laboratório de Neurociências Translacional, Rio de Janeiro RJ, Brazil.
[3]Universidade Federal do Rio de Janeiro, Hospital Universitário Clementino Fraga Filho, Centro de Referência em Doenças Inflamatórias Desmielinizantes, Rio de Janeiro RJ, Brazil.

Correspondence: Karla Pinto Coelho Fadel; Email: karlafadel@gmail.com.

Genetic studies of CNS autoimmune diseases, such as multiple sclerosis (MS), and Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD) aim to identify genetic variants that influence disease susceptibility and progress. In this study, we performed whole-exome sequencing on a family with a history of MS and MOGAD. The proband, a 24-year-old male, presented with paresthesia on the left side of his upper and lower limbs. MRI revealed two hyperintense white matter lesions, one periventricular and one at the medullary bulb junction, both gadolinium-enhancing. Cerebrospinal fluid (CSF) examination showed oligoclonal bands with normal cell and protein counts and no serological or molecular evidence of infection, including SARS-CoV-2. Serum testing was positive for anti-MOG antibodies and negative for anti-AQP4 antibodies. Tests for other metabolic and autoimmune diseases were negative. Despite being asymptomatic, the patient tested positive for COVID-19 in serum and was treated with intravenous glucocorticoids, leading to complete recovery. The proband, diagnosed with MOGAD, and his mother, diagnosed with MS, were both found to be heterozygous for the rs118004742 (T>G) variant in the EFCAB13 gene. In contrast, his healthy father

and sister were homozygous for the reference allele. This variant, previously identified as potentially associated with MS susceptibility, introduces a premature stop codon at position 546 (p.Tyr546Ter), disrupting protein function and reducing EFCAB13 expression in various brain regions according to FIVEx and GTEx datasets. EFCAB13 encodes a protein with a calcium-binding domain essential for neuronal function and plasticity, suggesting that its reduced expression may contribute to MS pathogenesis, although the full function of this gene remains unknown. This familial pattern highlights the potential significance of the EFCAB13 variant in autoimmune conditions. Further investigation is warranted to better understand the role of EFCAB13 in MS and other autoimmune diseases, such as MOGAD, for which genetic risk factors are currently not widely explored. This finding reinforces the overlapping genetic aspects of CNS autoimmune diseases, offering insights into shared mechanisms of disease and potential cross-disease therapeutic approaches.

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Multiple sclerosis and Behçet's disease: case report and literature review

Karina Carvalho Higa[1], Juliana Mathias Netto Khouri[1], Jussara Mathias Netto Khouri[1], Carlos Cesar Conrado Caggiano[1], Leandra Schneider[1], Isabela Louise Weber[1], Lucas Shigueo Hiromoto[1], Pedro Arhur Possan[1], Mateus Pinto Marchetti[1]

[1]Hospital Nossa Senhora das Graças, Curitiba PR, Brazil.

Correspondence: Karina Carvalho Higa; Email: karinacarvalho.fdc@gmail.com.

Case presentation: T.R.G, a 43-year-old female, has been diagnosed with antiphospholipid antibody syndrome (APS) and Behçet's disease (BD) since 2002. She was diagnosed with relapsing-remitting multiple sclerosis (MS) (EDSS 3) in 2019. The patient presents with left hemiparesis, optic neuritis, and hyperreflexic bladder. The MS diagnosis was confirmed by magnetic resonance imaging (MRI) showing multiple typical lesions and cerebrospinal fluid (CSF) analysis revealing oligoclonal bands. Previous treatments included beta-interferon 1a, glatiramer acetate, cyclophosphamide, dimethyl fumarate, and azathioprine, without favorable results. The patient is awaiting initiation of treatment with ofatumumab, an anti-CD20 therapy that may benefit both MS and BD. **Discussion:** The coexistence of MS and BD is rare and presents diagnostic and clinical management challenges. MS is an autoimmune disease of the central nervous system characterized by inflammation, demyelination, and neurodegeneration, resulting in varied neurological symptoms. BD is a systemic autoinflammatory disease with recurrent oral and genital ulcers, skin lesions, and rarely, central nervous system involvement. Differentiating MS from Neuro-BD (NBD) or primarily neurological BD (pNBD) is crucial, as each condition has specific diagnostic and therapeutic criteria. The McDonald criteria for MS require demyelinating lesions disseminated in space and time in the central nervous system, with findings on MRI and CSF. Patients with BD+MS tend to be women and exhibit a clinical course similar to MS, with relapses and remissions, unlike the progressive course of NBD or pNBD. This suggests that a personalized therapeutic approach, considering treatments for both conditions, may be beneficial. **Final comments:** A minority of patients with BD exhibit lesions similar to those in MS and meet the 2017 McDonald criteria. These BD+MS patients show clinical manifestations more aligned with MS than with NBD or pNBD, responding favorably to typical MS treatments. Findings suggest that patients with BD who present with MRI lesions, clinical characteristics, and CSF findings meeting the McDonald criteria should be treated considering both MS and BD. A multidisciplinary approach involving neurologists and rheumatologists is crucial for accurate diagnosis and appropriate management, aiming to provide the best treatment and quality of life for patients.

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Multiple sclerosis and CIDP: navigating the challenges of coexistence

Filipe Brito Porto[1], Isabela Caldeira de Oliveira[1], Jéssica Semler de Sá Lima[1], Gabriella Corrêa Dousseau[1], Daniel Thomas Pereira Lopes[1], Arthur de Medeiros Dias[1], Maria Sheila Guimarães Rocha[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Gabriella Corrêa Dousseau; Email: gabrielladousseau@gmail.com.

Case presentation: A 65-year-old man with a prior diagnosis of primarily progressive multiple sclerosis at age 51 began experiencing tingling and numbness in his feet at age 58. Over several months, this progressed to significant difficulty in moving his feet, such as an inability to use the pedal when driving. Additionally, he reports noticing muscle flaccidity in previously spastic lower limbs. His neurological examination showed generalized hypotonia and muscular atrophy. Deep tendon reflexes were hypoactive in the upper limbs, patellar and achilleus reflexes were abolished, although there was a bilateral Babinski sign. There was a decrease in superficial sensitivity in the feet and a loss of vibratory sensation in the upper and lower limbs. An electroneuromyography examination was performed in which motor nerve conduction studies revealed a decrease in motor conduction velocities (>30% of the lower limit of normal) and prolonged distal motor latencies (>50%) in more than two nerves. Following targeted investigation, a diagnosis of overlapping CIDP was made. Treatment with intravenous methylprednisolone for 5 days a month over 6 months showed no measurable improvement. Subsequently, Rituximab was initiated, and the disease remained stable without further progression. **Discussion:** Multiple sclerosis is an inflammatory autoimmune demyelinating disease that affects the central nervous system (CNS) and may have a relapsing or progressive course. Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a peripheral autoimmune disease with also a relapsing or progressive course and in its classical form may cause symmetrical, proximal and distal, sensory-motor impairment, mainly in the lower limbs. Although the accidental occurrence of multiple sclerosis and CIPD or other polyneuropathies is possible, there is evidence that common antigens (e.g., Neurofascin 155, myelin basic protein, and myelin-associated glycoprotein) can trigger an autoimmune response in central and peripheral myelin proteins. Furthermore, considering combined central and peripheral demyelination as a distinct condition increases the need for specific treatment. Some previous studies have identified rituximab and alemtuzumab as possible treatments. **Final comments:** Therefore, given the importance of early recognition of clinical manifestations, a clinical case of CIDP in a patient with multiple sclerosis is presented below to improve the awareness of practicing neurologists for better treatment outcomes.

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Multiple sclerosis and pregnancy: a relevant approach

Guilherme Vedovato Vilela de Salis[1], Gabriel Pina Paiva[1], Andressa Regina de Mello Galego[1], Renato Arantes Lima Simões[1], Waldir Antonio Maluf Tognola[1]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Guilherme Vedovato Vilela de Salis; Email: guilhermesalis@hotmail.com.

Case presentation: A 30 years old woman was diagnosed with RRMS in July 2001. For 3 years she had fluctuating multifocal neurological symptoms and the initial MRI showed multiple lesions suggesting demyelinating substrate with some of them showing contrast enhancement. During initial hospitalization she underwent corticosteroid therapy and plasmapheresis. After hospital discharge, Natalizumab (NTZ) was started with the patient reporting persistence of significant ataxia and asthenia. She underwent a total of 17 NTZ infusions, later abandoning treatment in January 2023. Therefore, she returned in April 2023 reporting worsening of the symptoms when MRI demonstrated areas suggesting inflammatory activity. She was subjected to a new corticosteroid therapy and presented remission of them. In November 2023, she discovered pregnancy and suspended the use of NTZ on her own, returning to emergency care in May 2024, 30 weeks pregnant, with a new outbreak, when MRI without contrast was compatible with previous exams but as a new outbreak was considered and pulse therapy was performed. There was symptomtic remission and she was discharged with pregnancy resolution 9 weeks later. The newborn was healthy and there were no new clinical outbreaks until the outpatient return in May 2024, when breastfeeding was suspended to resumption of NTZ. **Discussion:** The use of DMT, particularly NTZ, during pregnancy, lactation and its planning is a relevant issue in neurological practice and requires cleared neurological monitoring and discussion due to the lack of consistent data. The magnitude of risk of pregnancy-related MS relapses following NTZ cessation is unclear as whether this risk is reduced by pregnancy or other modifiable factors. Fetal morbidity and mortality associated with it and the joint decision regarding therapy should be an important factor in planning and approaching the patient. Our case illustrates a common and relevant clinical situation to the current state of the art of treatment of MS, considering the average age of prevalence of the disease. The inadvertent discontinuation of NTZ by the patient, motivated by her insecurity about the medicine and the lack of information regarding its use, resulted in 2 possible outbreaks within 3 years of her initial diagnosis, one of which was during pregnancy. Furthermore, the suspension of breastfeeding to resume use of DMTs is still a point of discussion in the literature. **Final comments:** The risk of new outbreaks of MS following the interruption of NTZ in the context of pregnancy planning, its implementation and breastfeeding must be discussed with the patient and considered in a broad context of possibilities so that the decision joint may benefit the patient in their treatment and reduce any fetal risks involved. The lack of information about this and the resulting patient's insecurity can increase the risks involved and be detrimental to the patient's clinical approach, depriving them of the best treatment

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Multiple sclerosis and treatment with natalizumab in pregnancy

Rafael Bastos Delgado[1], Sandra Regina Alonso Gonçalves[1], Regina Maria Papais Alvarenga[1], Marina Papais Alvarenga[1], Lilian Bezerra do Nascimento[2]

[1]Universidade de Vassouras, Vassouras RJ, Brazil. [2]Hospital Federal da Lagoa, Rio de Janeiro RJ, Brazil.

Correspondence: Rafael Bastos Delgado; Email: 16rafaelbastos@gmail.com.

Case presentation: A 38-year-old patient, brown, lawyer with Relapsing-remitting Multiple Sclerosis diagnosed in 2009, using natalizumab (NTZ) since 08/2013, returns to the Reference Center for the treatment of Multiple Sclerosis, reporting that she has been pregnant with a gestational age of eight weeks. Although the routine care for pregnant MS patients was to suspend the use of disease-modifying drugs (DMD), in this patient, with a high lesion burden and already in the third month of pregnancy, it was decided by common agreement that the medication would be maintained. The last NTZ infusion occurred on 01/2018 (gestational age of 28 weeks, beginning of the 3rd trimester of gestation). On 24/03/2018, she

went into labor, giving birth to a baby girl weighing 2,300g 48cm long, by normal delivery after 38 weeks and four days of gestation. There were no relapses during pregnancy, only nausea and gestational diabetes. The daughter was born healthy and did not show any changes in the hematological tests and exams. The patient did not breastfeed her daughter and returned with the NTZ 15 days postpartum. NTZ was indicated due to the failure of glatiramer acetate and recombinant interferon-1a beta-to-control relapses. In 2013, with the worsening of disability after a new outbreak (EDSS = 6.0), the first infusion of NTZ was started, and since then, it has been stable (last follow-up on 03/2024). **Discussion:** Among the DMD, NTZ is the 4th line drug made available by the Unified Health System for the treatment of MS patients with high lesional burden after proven lack of efficacy in controlling relapses by immunomodulatory and oral drugs. It is a monoclonal antibody and has been approved since 2012 for RRMS. Its action is to prevent the migration of leukocytes to the brain, reducing the inflammation on the disease. Recent studies have shown that its use before conception or during pregnancy does not contribute to the increase in the rate of miscarriage, risk of malformations, and premature birth. Concerning newborns, low birth weight and hematological alterations have been identified when exposed to NTZ. However, it is challenging to abruptly discontinue the use of NTZ in pregnant MS patients as there is a high risk of rebound. **Final comments:** We report the case of a patient with Multiple Sclerosis and high lesion burden who, after becoming pregnant without planning, used NTZ during pregnancy without complications. Only this patient used NTZ during pregnancy in our referral center, where we have already followed more than a hundred cases of pregnant women in whom we suspended the use of these drugs until the puerperium. The British Consensus highlights the importance of discussions between patient and physician, presenting risks and benefits evidenced in studies, recommending the maintenance of natalizumab before conception, pregnancy, postpartum, and breastfeeding, and respecting the specificities found in each phase of the pregnancy-puerperal period.

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Multiple sclerosis deaths in Brazil: a comparison between age groups and regions in the last 5 years

Izabelly Constatinov Lemos[1] , Gabriel Cordova[1], Emanuel Henrique dos Santos[1], Anne Beatriz Ricardo Amaral[1], Sofia Beatriz Andrade Manfio[1], Kauan Correia Derbli[1], Julia Farto Viana Oliveira[2], Gabriel André Silvério[1]

[1]Centro Universitário Campo Real, Guarapuava PR, Brazil.
[2]Universidade Estadual do Centro Oeste, Guarapuava PR Brazil.

Correspondence: Izabelly Constantinov Lemos; Email: med-izabellylemos@camporeal.edu.br.

Background: Multiple sclerosis (MS) is a neurological condition that affects the Central Nervous System, causing damage to the myelin and impairing the transmission of electrical impulses to the brain. It is believed that genetic susceptibility and environmental factors play a triggering role in the disease. Symptoms, including asthenia, paresthesia, weakness, dizziness, bladder dysfunction, and cognitive impairment, are variable and can manifest differently in each individual. **Objective:** To analyze the number of deaths related to MS in Brazil from 2019 to 2023, comparing age groups and regions. **Methods:** This observational epidemiological study used data from the Department of Epidemiological Analysis and Surveillance of Non-Communicable Diseases (DAENT) on deaths from MS (G35 multiple sclerosis) in Brazil between 2019 and 2023. Variables included age groups (from 0 to over 80 years) and regions (North, Northeast, Midwest, South, Southeast), excluding the unknown category. Deaths were categorized by residence, without distinction of sex and race. Results were compared to determine the most affected age groups and regions. **Results:** A total of 1329 deaths were recorded from 2019 to 2023. The highest number of deaths occurred in 2022 (n=325), and the lowest in 2023 (n=110). The most affected age groups were 60-69 years (highlighted in 2020, 2021, 2022) and 50-59 years (highlighted in 2019 and 2023). The Southeast region had the highest concentration of deaths, representing 48.9% of national cases. **Conclusion:** The Southeast and South regions were the most affected by MS deaths, possibly due to the genetic inheritance of European descent. The most affected age group was 60-69 years between 2019 and 2022, with a significant reduction in deaths in 2023 (64%). Further studies are needed to clarify etiological factors and improve treatment in different Brazilian regions.

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Multiple sclerosis diagnostic delay in Brazil: related factors and the impact on functional outcome

Alex de Cerqueira Silveira Figueiredo[1], Maria Eduarda Messias Vassoler[1], Gabriel Souza Medrado Nunes[1], João Pedro Fernandes Gonçalves[1], Luana Araujo Passos[1], Luísa Dias Moreno Aravena[1], Júlia Xavier Oliveira[1], Bruno Bacellar Pedreira[2], Pedro Antonio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Alex de Cerqueira Silveira Figueiredo; Email: alexcsf@ufba.br.

Background: Multiple sclerosis (MS) is a demyelinating disease marked by neurological deficits that are often disabling. Even with well-defined criteria, early diagnosis is a challenge worldwide. Knowing the factors related to late diagnosis enables timely recognition and treatment, allowing a better functional outcome. However, no studies yet investigated the factors associated with delayed diagnosis and its consequences in Brazil. **Objective:** To identify the factors associated with late diagnosis of MS in Brazil and its impact on functionality. **Methods:** A cross-sectional study was conducted with patients diagnosed with MS according to the McDonald criteria, followed up from July/2021 to April/2024 in a reference center in Bahia, Brazil. Clinical-demographic information was collected by interview and EDSS was defined after neurological examination performed by neurologists or neurology residents and was used to categorize functionality from symptom-free (0) to severe disability (≥5). Diagnosis after 3 months from symptom onset was considered late, following the NICE recommendation. Logistic regression was used to identify factors related to late diagnosis and ordinal logistic regression was performed to determine the impact of this delay on functionality. **Results:** 265 RRMS patients were recruited, of whom 178 (67.2%) were female. The median (IQR) age at symptom onset was 30 (22-38) and at diagnosis, 32 (24-40). One hundred and fifty-nine (60%) individuals were diagnosed more than 3 months after symptoms onset. The mean (SD) time from symptom onset to diagnosis was 24.1 (±43.1) months and the median (IQR) was 7 (2-29). Living outside the capital and metropolitan region (OR 2.477 [95%IC 1.382-4.439]), symptom onset before the 2017 McDonald criteria actualization (OR 2.090 [95%IC 1.134-3.607]), previous misdiagnosis (OR 2.057 [95%IC 1.131-3.743]) and presence of comorbidities (OR 2.925 [95%IC 1.639-5.222]) were the factors related to late diagnosis while first presentation with visual symptoms was related to early diagnosis (OR 0.522 [95%IC 0.292-0.933]). The delayed diagnosis was independently associated with worse functional outcomes (OR 1.724 [95%IC 1.086-2.737]) in a model adjusted for total relapse rate, disease duration, and age. **Conclusion:** There is still a lack of information on the delay in diagnosing MS in Brazil, and the time to diagnosis found in the present study is one of the longest ever reported in international literature. The majority of individuals with MS were diagnosed late and this was associated with difficult access to the health system, and pitfalls in the recognition and application of diagnostic criteria by both specialists and non-specialists. Finally, a longer time to diagnosis was related to worse functional outcomes, highlighting the importance of timely diagnosis and treatment for a favorable prognosis in MS patients.

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Multiple sclerosis functional composite (MSFC) as a possible more sensitive tool for changes in functional evaluation than expanded disability status scale (EDSS) in multiple sclerosis (MS) patients: a Brazilian cohort study in Bahia

Maria Eduarda Messias Vassoler[1], Gabriel Souza Medrado Nunes[1], Alex de Cerqueira Silveira Figueiredo[1], Thaís Barbosa de Oliveira[1], Antônio Lázaro Santos Soares Junior[1], João Pedro Fernandes Gonçalves[1], Júlia Xavier Oliveira[1], Pedro Antonio Pereira de Jesus[2], Bruno Pedreira Bacellar[2], Thiago Gonçalves Fukuda[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Maria Eduarda Messias Vassoler; Email: maria.vassoler@ufba.br.

Background: The multiple sclerosis functional composite (MSFC) is a tool used to assess functionality based on limb mobility and cognition in individuals with multiple sclerosis (MS). Periodic evaluation of functional capacity allows for monitoring disease progression. Although previous national studies have evaluated this tool, its sensibility has yet to be addressed, particularly in our heterogeneous population. **Objective:** To investigate the MSFC in evaluating variation in functional outcomes in patients with MS. **Methods:** This cohort study enrolled patients diagnosed with MS, according to the 2017 McDonald criteria, conducted between April 2023 and March 2024, that had no variation in the Expanded Disability Status Scale (EDSS) and no disease relapses between evaluations. Clinical and demographic information was collected through interviews at a reference center in Bahia. The EDSS was determined following a neurological examination performed by a neurologist, and the MSFC was administered by researchers at the same consultation. The MSFC consists of the Timed 25-Foot Walk (T25W), 9-Hole Peg Test (9HPT), and Paced Auditory Serial Addition Test (PASAT-3). We normalized the tests into Z-scores using the population mean established by the NMSS Task Force database and calculated the MSFC score. The Wilcoxon signed-rank test for dependent samples was performed. **Results:** The study comprised 85 patients, of whom 70.6% were female, with a median age of 40 years (IQR: 31.5-49). Within this cohort, 44.7% had completed high school education and 35.3% had higher education. The median duration of the disease was 6 years (IQR: 3-10), and the median follow-up time was 5 months (IQR: 4-6). The median EDSS evaluation in our sample was 1 (IQR: 0-4.75), while the median total Z-score of MSFC was -0.41 (IQR: -1.32-0.19) in the first evaluation and -0.20 (IQR: -1.34-0.28) in the second. When comparing the values of the different assessments with MSFC, a statistically significant difference was found ($p<0.05$). However, analyzing the components, T25W and PASAT-3 did not show a significant difference between evaluations ($p>0.05$), which was found in the different evaluations of the 9HPT ($p<0.001$). **Conclusion:** The MSFC is a tool that can possibly be more sensitive to small variations in the assessment of functional capacity in patients with MS when the EDSS evaluations do not vary. The 9HPT was identified as the test within the MSFC that showed the most significant differences between evaluations. More studies are needed to better understand the differences observed.

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Multiple sclerosis in Brazil: an analysis of death and hospitalization trends using joinpoint regression

Alisson Rafael de Oliveira Pereira[1], Ana Júlia Alves de Oliveira Andrade[1], José Sidinei Rodrigues Dias[1], Rodrigo Ferreira de Moura[1]

[1]Universidade Federal de Lavras, Lavras MG, Brazil.

Correspondence: Alisson Rafael de Oliveira Pereira; Email: alissonropereira@gmail.com.

Background: Multiple sclerosis (MS) is one of the neurological diseases with guidelines described in SUS's Clinical Protocol and Therapeutic Guidelines (PCDTs). Although external factors may interfere, as it is a chronic noncommunicable disease without seasonality, trend variations in this type of time series can be more likely attributed to public policy interventions. **Objective:** To analyze joinpoints in the time series of deaths and hospitalizations due to MS in Brazil. **Methods:** A study was conducted using data recorded at the SIH using DATASUS. Time-series analysis was performed using a joinpoint model, and all PCDTs published on MS were reviewed. **Results:** Hospitalizations for MS varied annually with rates ranging from 0.85 (2010) to 3.39 (2021). The selected regression model suggested the presence of two joinpoints (WBIC= -3.29) and three bands of behavior in relation to the annual rates of change. In the first stretch, from 2008 to 2014, there was a non-significant increase (APC: 0.89; 95%CI: -9.69 to 5.84; p=0.91); in the second section, from 2014 to 2017, there was a considerable increase (APC: 27.5; 95%CI: 13 to 36.9; p<0.02); and in the third section, from 2017 to 2023, there was another increase, although less intense than the previous one (APC: 9.18; 95%CI: -5.37 to 12.76; p=0.12). During the study period, there was a significant increase in hospitalization for MS (AAPC: 8.72; 95%CI: -5.37 6.64; p<0.001). Regarding the death rate, joinpoints were not identified (APC: -1.24; 95%CI: -4.65, 2.28; p=0.44). As for Brazilian policies, the first PCDT for MS was instituted in 2010, guiding the use of β-interferon and glatiramer as 1st line of disease-modifying drugs (DMDs) and natalizumab in case of refractoriness. Updates to this plan were made in the following years: inclusion of fingolimod as DMD in 2014; mandatory monitoring of the 1st dose of fingolimod in a specialized center in 2015; inclusion of teriflunomide and dimethyl fumarate as DMD in 2018; Update of the McDonald criteria in 2019; possibility of plasmapheresis during outbreaks in 2021; and inclusion of alentezumab in high-activity RR-MS. The results of the ARIMA models (0,1,0) showed that the modification of the McDonald criteria in 2019 had a significant effect on deaths due to MS (B=0.014, SE=0.005; p<0.02), although the same analysis was not statistically significant for hospitalizations. **Conclusion:** Maintenance of the downward trend in MS deaths illustrates the effectiveness of therapies in preventing disease progression. An increase in MS hospitalizations was identified with joinpoints in 2014 and 2017. In both cases, the modification in the PCDT occurred in the year following the change in the trend. The need to monitor the administration of fingolimod may explain the increasing trend in hospitalizations that began in 2014, and the inclusion of teriflunomide and dimethyl fumarate in the PCDT may explain the decrease in the slope of this regression.

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Multiple sclerosis resulting from the use of adalimumab for psoriatic arthritis

Fabricia dos Santos Almeida[1], Amanda Lima Sampaio[1], Lucas Medeiros Dunga[1], Artur Rebouças de Souza[1], Bianca Etelvina Santos de Oliveira[2], Iury Hélder Santos Dantas[1], Tiago Lameque de Sousa e Silva[1], Tereza Cristina Batista Dias[1], Matheus Procópio Guimarães[1], Helmut Kennedy Azevedo do Patrocínio[1]

[1]Universidade Federal do Rio Grande do Norte, Caicó RN, Brazil.
[2]Centro de Referência em Esclerose Múltipla na Paraíba, João Pessoa PB, Brazil.

Correspondence: Lucas Medeiros Dunga; Email: lucasdunga2000@gmail.com.

Case presentation: Female patient, 40 years old, diagnosed with psoriatic arthritis being treated with adalimumab (ADM). She developed paresthesia in her lower limbs, which was initially attributed to an anxiety disorder. After six months, there was a decrease in visual acuity in her left eye, and she was diagnosed with optic neuritis. She then underwent pulse therapy with methylprednisolone and began investigation for multiple sclerosis. She underwent a magnetic resonance imaging of the brain revealed several high signal density lesions in T2/FLAIR in both periventricular regions. In addition, imaging of the thoracic spine showed high signal intensity in the posterolateral cords at levels T5 to T9, these findings met McDonald's criteria. The neurological examination revealed mild paresis of the left lower limb, hyperreflexia in the lower limbs,mild hypopalesthesia in the right leg and bilateral plantar-cutaneous reflexes. Treatment with ADM was then suspended, and there were no more signs of psoriatic arthritis, except for a worsening of the skin condition. **Discussion:** The patient had an underlying autoimmune disease, psoriatic arthritis, one of whose treatments is the use of tumor necrosis factor (TNF) antagonists. However, one of the serious adverse effects of this type of medication is the risk of developing demyelinating diseases, such as multiple sclerosis. **Final comments:** Neurological disorders resulting from adverse effects of TNF, such as ADM, are rare, but should also be analyzed. And if these disorders occur during treatment, the drugs should be discontinued immediately.

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Multiple sinus cerebral venous thrombosis secondary to Neuro-Behçet: a case report

Patrícia Marques Mendes[1], Eduardo Mendonça Werneck da Silva[2], Caio Gonçalves Gomes[2], Maycon Melo Lopes[2], Laís Aparecida Machado de Siqueira[2], Johnny Teixeira Neves[2], Yan da Silva Raposo[2], Igor Pereira Matos de Oliveira[3], Vitor Hugo Machado Silva[4], Enzo Barreto Viana[4]

[1]Universidade Federal de São Paulo, SaoPaulo SP, Brazil.
[2]Hospital das Clinicas Samuel Libâneo, Pouso Alegre MG, Brazil.
[3]Santa Casa da Misericórdia de Belo Horizonte, Belo Horizonte MG, Brazil.
[4]Universidade do Vale do Sapucaí, Pouso Alegre MG, Brazil.

Correspondence: Eduardo Mendonça Werneck da Silva; Email: eduardomwerneck@gmail.com.

Case presentation: In the present work we presente a 29-year-old male patient who presented with a pulsatile hemicranial headache with limitation to physical exertion, along with nausea, phonophobia, photophobia and analgesic refractoriness, due to such complaints he initiated investigation with a neurologist. The patient presented in his pathological history a previous event of cerebral venous thrombosis and recurrent genital and oral ulcers. In his Venous MRI, signs of extensive venous thrombosis of recent/subacute appearance were evident, affecting a large part of the superior sagittal dural sinuses, torcula of herophilus, straight sinus and transverse and sigmoid sinuses bilaterally, with extension to the bulbs of the jugular veins; In his CSF examination, no changes were found. **Discussion:** Neuro-Behçet is defined as the manifestation of neurological signs and/or symptoms in patients with a defined diagnosis of Behçet's disease, which can manifest itself in multiple ways, including multiple sclerosis syndrome, cerebral venous sinus thrombosis, meningoencephalitic syndrome or others, being subsequently divided into parenchymal or non-parenchymal forms depending on the brain involvement, the case in question being the non-parenchymal type. Presenting in its list of possible clinical changes symptoms, similar to idiopactic venous thrombosis, the patiet may experience headache, nausea, convulsions , focal deficits or even intracranial hypertension. **Final comments:** In cases with atypical epidemiology of cerebral venous thrombosis, investigation of possible secondary etiologies should be carried out with the aim of early diagnosis and thus avoiding new events and possible morbidity and mortality secondary to the underlying pathology.

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Multisystemic fabry disease in female patient with irregular treatment: a case report

Maria Eduarda Braz Ferreira Leão Caldas dos Santos[1], Bernardo Leal Cupello[1], Clara Ribeiro Rizzo[1], Isabella Navarro Diaz Horta[1], Lucas Gonçalves Coelho[1], Victoria Nunes Alvares[1], Vanessa Conceição Saar Mautone Lima[2], Karina Lebeis Pires[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Hospital Universitário Gaffrée e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: Maria Eduarda Braz Ferreira Leão Caldas dos Santos; Email: meduardaleao@edu.unirio.br.

Case presentation: A 40-year-old female presented in 2010 a crisis of pain and burning sensation in the upper and lower limbs that worsened with temperature extremes, in addition to asthenia, abdominal pain, bilateral amaurosis for 1 week and a diarrhea episode that lasted 6 months. Two years later, she presented a case of hemiparesis. The complementary tests were performed for further investigation and the results were consistent with Fabry disease: genetic molecular testing showed the heterozygous mutation c.823delC in the sixth exon of the GLA gene and ophthalmological evaluation that identified cornea verticillata. Furthermore, laboratory tests showed microalbuminuria (55.54mcg/min) and proteinuria (0.19g/24h) in 24-hour urine collection and a brain MRI showed deposits of hemosiderin/ferritin in the right cerebellum hemisphere. Currently, our patient is on irregular betagalsidase enzyme replacement therapy, referring daily episodes of migraine with aura, frequent nausea, dysgeusia, worsening pain in her limbs and anemia. On current neurological evaluation, she presents monocular diplopia, painful hypoesthesia in hands and feet and touch hypoesthesia in feet consistent with superficial sensitivity syndrome compatible with polyneuropathy. **Discussion:** Fabry disease is a rare lysosomal deposition disorder linked to chromosome X, that affects the GLA gene. It's caused by a partial deficiency of the alpha galactosidase A enzyme, resulting in the excessive storage of globotriaosylceramide in the vascular endothelium and visceral tissue, which the skin, heart, kidney and the central nervous system are the most affected. Due to being a genetic inheritance linked to X chromosomal, every male patient with the genetic alteration manifests the disease symptoms. Otherwise, heterozygous female subjects may not present symptoms for a long while and, when manifested, these usually are less frequent and less intense. Therefore, this case warns of the occurrence of systemic manifestations in women, such as anemia, renal insufficiency, mood disorders, neuropathic pain and migraines, especially without the rightful treatment. **Final comments:** Fabry disease patients must join enzyme replacement therapy, since the treatment can halt disease progression, decreasing systemic manifestations. Thus, with early diagnosis and long-term adequate enzyme therapy, it is possible to guarantee the patient's well being.

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Muscle ultrasound changes in pre-symptomatic and type 1 spinal muscular atrophy in the first months of life

Ana Lucila Moreira[1], Rodrigo Holanda Mendonca[2], Edmar Zanoteli[2], Raquel Diógenes Alencar Sindeaux[2]

[1]CENEC Neurofisiologia Clínica, Campinas SP, Brazil. [2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Ana Lucila Moreira; Email: ana.lucila.moreira@gmail.com.

Background: Spinal muscular atrophy (SMA) is a life-threatening motor neuron disease and better outcomes are possible with both early diagnosis and treatment. **Objective:** We investigated the ultrasound pattern of muscle involvement in treated pre-symptomatic and type 1 SMA patients along the first months of life, to evaluate the evolution of muscle compromise secondary to the disease **Methods:** We examined longitudinally the biceps brachii, rectus femoris and intercostal muscles of three pre-symptomatic (2 copies of SMN2) and one type 1 SMA (2 copies of SMN2) patients diagnosed in the newborn screening, since the diagnosis (in the first month of life) along the first months of life (3-8 months), using B-mode ultrasound for mean grey scale analysis (GSA). All patients were initially treated with Risdiplam. **Results:** All patients had higher GSA in the rectus femoris compared to biceps brachii around the age of 3 months, even though in three of them the rectus femoris GSA was lower than the biceps brachii at the moment of diagnosis. In all but one patient the biceps brachii GSA increased in the first months of life. Three out of four patients had been followed of at least six months, showing stabilization of GSA in all studied muscles in two and continuous increase in one. **Conclusion:** Preliminary results of quantitative muscle ultrasound analysis in pre symptomatic and type 1 SMA patients demonstrated abnormalities in even clinically non symptomatic patients, and stabilization during the treatment. However, inclusion of more patients in the study and a longer follow-up period are necessary to obtain definitive conclusions.

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Muscle ultrasound evaluation on congenital myasthenic syndrome related to CHRNE

Eduardo de Paula Estephan[1], Ana Lucila Moreira[1], Beatriz Carneiro Gondim Silva[1], André Macedo Serafim Silva[1], Antonio Alberto Zambon[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Eduardo de Paula Estephan; Email: eduardo.estephan@gmail.com.

The assessment of muscle disorders and myopathies is greatly enhanced by the addition of imaging to identify structural abnormalities and provide an assessment of muscle quality. In congenital myasthenic syndromes (CMS), muscle changes and their histopathological basis are not well understood. In fact, this issue has rarely been addressed. In one study with different types of CMS, no muscle alteration was found with magnetic resonance. In recent publication, myopathic changes on biopsy and magnetic resonance image have been reported, mainly in patients with CHRNE CMS. Muscle imaging showed at least mild alterations in the majority of confirmed cases, with preferential involvement of soleus, especially in CHRNE cases. However, muscle echogenicity in this disease group has not been evaluated. We qualitatively measured muscle echogenicity in patients with congenital myasthenic syndrome with receptor deficiency due to CHRNE mutations. Sixteen CHRNE patients were included. The patients' muscle echo intensity (IE) was classified as normal, mildly myopathic, moderately myopathic, or severely myopathic. Cranial, respiratory and unilateral appendicular muscles were evaluated. Fourteen patients had at least one altered muscle (87%). The most frequently affected muscles were: genioglossus (75%), lateral gastrocnemius (68%), tibialis (68%) and medial gastrocnemius (62%). Some distal muscle of the lower limb was altered in 81% of the patients. Among respiratory muscles, the external oblique was the most affected (56%). Surprisingly, the diaphragm was normal in all 16 cases, while intercostals were affected in 50%. On orbicular oculli, only five patients (35%) showed mild alteration. Other rarely affected muscles were: neck extensors (18%), geniohyoid (25%), internal oblique (31%) and digastric (37%). In all muscles, alterations were mild, except for 8 lateral gastrocnemius (50% of all cases), 6 medial gastrocnemius (37%), 4 tibial (25%) and 1 soleus (5%), on which moderate or even severe alterations were seen. In conclusion, complete normal muscle echogenicity is not common in CHRNE congenital myasthenic syndrome, being tongue and distal lower limb muscles the most affected. In the other hand, diaphragm is consistently spared among this population but intercostals can be affected. Then, muscle ultrasound may be useful to characterize this group of disease, and can be helpful to deeply identify the phenotype.

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Music therapy as a complementary strategy for behavioral and cognitive enhances in dementia: a systematic review

Gabriel Thomaz Candido[1], Brener Raad Pereira[1], Caio Araujo de Souza[1], Carla Diovanna Spadine Pinhataro dos Santos[1], Gabrielle Gomes Garcia[1], Henrique Sarlo Pezzin[1], Marcus Vinicius Teixeira Calejon Stumpf[1], Matheus Guilherme Marques Vidal Barboza[1], Sofia Gameiro de Souza[1], Izabela Jardim Rodrigues Pitta[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Gabriel Thomaz Candido; Email: gabriel_candido@id.uff.br.

Background: The increasing prevalence of dementia, due to the rise in the population life expectancy, represents a public health and socioeconomic challenge, given the demands for individualized and multidisciplinary care. Music therapy, a practice that aims to promote the maintenance and improvement of physical, mental and emotional health through music stimuli, emerges as a non-invasive and complementary treatment alternative. The purpose of this systematic review is to evaluate musical practice as a tool, based on an analysis of the main findings in the literature. **Objective:** To provide evidence concerning the potential therapeutic role of music therapy in improving, behavioral and cognitive symptoms in elderly people with dementia diseases. **Methods:** This study is a literature review based on the methodological recommendations of the Preferred Reporting Items for Systematic Review (PRISMA) guidelines. The databases used were PubMed, SciELO, Cochrane and Lilacs. The search strategy was: music therapy OR song AND dementia AND elderly. The inclusion criteria were: randomized clinical trials, from 2014 to 2024, in English, covering the elderly population and meeting the objective of the study. The exclusion criteria were studies that used another non-pharmacological intervention simultaneously and studies not available in full version. **Results:** A total of 119 articles were found in the databases, with 14 duplicates. After double-blind screening, with a third evaluator to cases of disagreement, 25 were selected for analysis. Among these, 10 were excluded for not presenting results or for not being available in full version. The analysis of the 15 remaining articles showed a population aged 65 years or older, of both sexes and diagnoses with mild to very severe dementia. The type of therapeutic intervention varied between music listening, singing and choral singing. Nine articles, out of the 13 that analyzed behavioral issues, showed an improvement in patients` mood or a decrease in anxiety/agitation/depression. Six articles, out of eight that evaluated cognitive issues, showed an improvement in memory, executive functions or general cognitive functions. Among these, one article aimed to analyze the beta-amyloid levels and telomerase length, showing a slight increase in protein concentration and enhancement in cognitions and mood. In one article, lower keys increased patients` feelings of sadness, and faster rhythms increased their excitement. **Conclusion:** Music therapy has shown itself to be a possibility for the non-invasive treatment of symptoms related to cognitive and behavioral function, capable of improving patient's quality of life. Possible limitations of the studies analyzed relate to different etiologies of dementia diseases, which may respond differently to the therapy, the degree of dementia, that may influence the response to the proposed therapy and the variation in the music interventions, which can provide different results.

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Mutation in the FHL1 gene and X-linked myopathy with postural muscle atrophy

Camilla Rocha Medeiros Pinho[1], Rafael Amaral de Matos Albuquerque[1], Guíniver Couto Almeida[1], Gabriel Negreiros de Camargo Martins Moreira[1], Marcela Câmara Machado Costa[1], Marilaine Matos de Menezes Ferreira[1], Adriele Ribeiro França Viriato[1], Thainá Gonçalves Tolentino de Figueiredo[1], Laís Brandão Mota[1], Luna Souza Silva Vilanova[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Rafael Amaral de Matos Albuquerque; Email: rafaelalbqq@outlook.com.

Case presentation: Consanguineous parents, first-degree cousins, had 8 children (2 females and 6 males) - 3 of the males died, two at 29 and 21 years (with similar histories). A.S.J., 21 years old, and L.S.J., 24 years old, children of the couple and patients at the outpatient clinic, began to manifest progressive muscle weakness at age 12, in the lower limbs and scapular-waist region. Both report episodes of falls and pain with physical exertion, as well as exertional dyspnea, requiring intermittent rests and non-invasive ventilatory support (biPAP). Gait in waddle, winged scapulae, and limited movements due to joint contractures are additional shared signs and symptoms. At 13 years old, A.S.J. presented severe right thoracolumbar scoliosis. Genetic testing identified a mutation in the FHL1 gene, with a suspected diagnosis of X-linked myopathy with postural muscle atrophy. **Discussion:** Both siblings present a complex clinical picture characterized by progressive muscle weakness, respiratory involvement, and cardiovascular changes, including structural cardiomyopathy with myocardial fibrosis. The mutation in the FHL1 gene that suggests a diagnosis of X-linked myopathy with postural muscle atrophy is a progressive condition with no known disease-modifying treatment, requiring multidisciplinary management involving neurologists, physiotherapists, and speech therapists to improve quality of life and delay disease progression. **Final comments:** This case report highlights the challenges in diagnosing and managing progressive myopathies, especially in families with complex genetic histories. Therapeutic approaches focused on respiratory support, muscle rehabilitation, and management of cardiac complications are crucial to minimize symptoms and improve the quality of life for these patients. Continuity in medical follow-up and adaptation of therapies as the disease progresses are essential to address the challenges faced by these patients throughout their lives.

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Myasthenia gravis after hormonal blockade in a prostate cancer patient: case report

Pedro Vinicius Brito Alves[1], Riccardo Casarotti[1], Luiz Gustavo Brenneisen Santos[1], Aida Maria Negrão Esteves[1]

[1]Prevent Senior Brasil, São Paulo SP, Brazil.

Correspondence: Pedro Vinicius Brito Alves; Email: pvbalves@gmail.com.

Case presentation: A 66-year-old male was admitted with a history of bilateral asymmetric ptosis, complex ophtalmoparesis and head drop, with fluctuating symptoms, which began 3 weeks prior to admission and progressed to a myasthenic crisis. The patient had a previous medical history of multiple cardiovascular comorbidities and prostate adenocarcinoma with local lymph node invasion. Four weeks before the onset of symptoms, he received androgen deprivation therapy (ADT) with leuprorelin. Treatment with intravenous immunoglobulin therapy was started with substantial clinical improvement. Further investigation revealed a positive serum anti-acetylcholine receptor blocking antibodies and negative binding antibodies andnormal muscle enzyme levels. Chest imaging studies was unremarkable. He was diagnosed with myasthenia gravis based on clinical syndrome and antibody positivity. While his ocular and bulbar symptoms resolved, he still experienced fatigue and impaired axial support. He was referred to neuromuscular rehabilitation department and continues his oncological treatment without any other complications. **Discussion:** Myasthenia gravis (MG) is the most common neuromuscular junction disorder, with a bimodal prevalence, being more prevalent in young women and older men. Primary clinical manifestations include muscle weakness with fatigability, which can be explained by the saturation of a reduced number of acetylcholine receptors available on the postsynaptic membrane. MG produces autoantibody- and cell-mediated destruction of acetylcholine receptors (AChRs) at the neuromuscular junction of the skeletal muscles. Symptoms arise once the number of AChRs is reduced to approximately 30% of normal. Often known as a primary autoimmune disease, it may also occur in the context of active neoplasia, such as thymoma, which is frequently associated with anti-AchR receptor antibodies. There are rare case reports of myasthenia gravis associated with prostate cancer. Although androgen deprivation therapy is associated with a reduction in chronic inflammation and cancer progression, literature reports show patients developing myasthenia gravis during ADT. The detection of an atypical antibody and its response to immunotherapy suggest an immunological mechanism for the condition. However, the persistence of fatigue and generalized muscle weakness with absent muscle injury markers could be the caused by residual hypoandrogenism. **Final comments:** Oncological disease represent a major impact and has a increasing prevalence in the general population. This case emphasizes the importance of keeping high awareness of neurological complications related to cancer, as well as the adverse effects of oncological therapies. The association of disease activity and androgen deprivation medication may be a link between the state of functional hypogonadism in elderly men and development of MG. We highlight the role of the steroid hormonal system in modulating the immune system.

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Myasthenia gravis as a hidden diagnosis in a patient with chronic pain syndrome and fatigue: a case report

Vanessa Conceição Saar Mautone Lima[1], Ricardo Cezare Araujo[1], Victória Nunes Alvares[2], Lucas Gonçalves Coelho[2], Clara Ribeiro Rizzo[2], Carlos Alberto de Souza Moreira[1], Mariana Gonçalves Musauer[1], Raphael Breder[1], Caroline Bittar Braune[1]

[1]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gaffrée e Guinle, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Victória Nunes Alvares; Email: victoriaalvares@edu.unirio.br.

Case presentation: A 20 year-old female presented with generalized myalgia and migratory arthritis following an acute episode of gastroenteritis. She reported progressive worsening of the pain and fatigue, associated with predominantly proximal muscle weakness, all of which worsend through the day and limited her daily activities. She was evaluated by the pain clinic, with a diagnosis of chronic pain syndrome, and was referred to our neurology department for further investigation. Severe pain greatly limited neurological assessment and interpretation of examination findings. The neurological examination demonstrated a slow gait, inability to walk on toes and heels, proximal appendicular weakness in upper and lower limbs, and complaint of fatigue during ocular motility evaluation, without ptosis and/or ophthalmoparesis. Complementary investigation included normal levels of creatine phosphokinase, aldolase and lactate dehydrogense, normal neurophysiological studies with repetitive nerve stimulation (acessory, ulnar and deep fibular nerves) and normal electromyography. Considering the possibility of generalized Myasthenia Gravis, the anti-acetylcholine receptor antibody level was requested, which was positive (3.41 nmol/L; negative <0.40nmol/L), thus confirming the diagnosis. **Discussion:** Myasthenia gravis (MG) is an autoimmune diasease of the neuromuscular junction mainly caused by anti-acetylcholine receptor antibodies (AChR). AChR-MG has typical bimodal age pattern, with a peak in young adults, as in the present case report, and then a steady increase in incidence with age older than 50 years. Muscle weakness is a major feature, and fatigability is a hallmark. Approximately 60% of patientes with MG have ptosis and/or diplopia at onset, but the absence of ocular findings should ot rule out the diagnosis, as ocurred in our case. Furthermore, symptoms related to chronic pain syndrome and proeminent fatigue obscured the clinical finding of muscle weakness in our case, reiterating the importance of having a high clinical suspicion for the diagnosis of MG in patientes complaining of predominantly proximal muscle weakness without defined etiology. Low-frequency repetitive nerve stimulation has sensitivity of 53-100% for the diagnosis of generalized MG, and accuracy is higher when weak muscles are evaluated. In forms of MG with proximal muscle weakness, the study of distal muscles of the limbs may be negative, which may have occured in our case. Single fiber eletromyography was not available on our institution by the time of her evaluation. **Final comments:** MG is a highly heterogeneous disease with different possible phenotypes. The absence of ocular findings should not exclude the diagnosis, and nonspecific findings such as fatigue are common, and can often obscure the identification of classic muscle weakness, thus delaying the diagnosis of a treatable disease.

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Myasthenia gravis induced by immune checkpoint inhibitor

Alberto Pereira Firmino Filho[1], Maria Weryca de Souza Belo Silva[1], Mario Emilio Teixeira Dourado Junior[1], Miguel Inacio Silva Gomes[1], Felipe Toscano Lins de Menezes[1], Mario Teruo Yanagiura[1], Nathalia Lindsay Pereira de Queiroz[1], Francisco Gil Sarmento Neto[1], Felipe Olobardi Freire[1], Gabriel Soares Dutra de Souza[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Alberto Pereira Firmino Filho; Email: firminofilho.ap@outlook.com.

Case presentation: 70 year-old male with unresectable multinodular hepatocellular carcinoma secondary to the hepatitis C virus, had been taking atezolizumab and bevacizumab every 3 weeks, starting in November 2023. In March 2024, he noticed diplopia and eyelid ptosis on the left, progressive in intensity, affecting the contralateral eye over the course of days. Symptoms fluctuated, worsening throughout the day and the side most affected varied as the day progressed. On physical examination, he presented a mild divergent strabismus, with slight limitation of eye movement in both the vertical and horizontal planes, unable to achieve convergence, associated with bilateral semiptosis. His pupils were small, but with correct photomotor response. Chronic bilateral hearing loss in use of hearing aid device. No change in appendicular assessment of strength, sensitivity or coordination. Fatigability tests did not increase strabismus or ptosis. Ice testing showed no improvement. Repetitive effort did not intensify strength or myotatic reflexes. Electroneuromyography was performed without evidence of decrement after repetitive low-frequency stimulation. Evaluated once again with concentric needle single-fiber electroneuromyography, the jitter analysis was suggestive of neuromuscular junction disease. Magnetic resonance imaging of the skull and CSF study showed no pathological findings. The patient was diagnosed with ocular Myasthenia Gravis, had atezolizumab suspended and started on prednisone associated with pyridostigmine. **Discussion:** Immune checkpoint inhibitors (ICIs), such as atezolizumab, have been increasingly used in clinical practice, with good effectiveness and tolerability, significantly improving the treatment prognosis, increasing survival and quality of life for users. However, in the follow-up of patients who use it, increased rates of side events related to autoimmunity were noted, including myasthenia gravis. Its occurrence has been reported in 1.2% of individuals who were exposed to treatment, with the majority of cases starting within the first 6 weeks of therapy. Two-thirds of cases are considered serious. Specific antibodies are negative in the vast majority of patients. Treatment involves the interruption of ICIs associated with immunotherapy, with corticosteroid therapy being the first choice. In refractory cases, human immunoglobulin or plasmapheresis are indicated. After improvement, selected patients can be treated with ICIs depending on the therapeutic need for the underlying disease, evaluating risk and benefits. **Final comments:** The use of ICIs is associated with autoimmune events, such as myasthenia gravis. Appropriate management includes temporary or permanent suspension of medication, which is only possible through early and accurate recognition of these possible associated complications.

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Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) mimicking a stroke: case report

Andreia Lira de Oliveira[1], Nise Alessandra de Carvalho Sousa[1], João Victor de Chagas Rossin[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Andreia Lira de Oliveira; Email: andreialira.med@gmail.com.

Case presentation: Woman, 53 years, presenting with an acute intense burning pain in her left arm, developing left hemiparesis over 15 minutes, resulting in a fall from her own height. After being taken to an Emergency Department, upon arrival she also noticed hypoesthesia on her left hemibody. The condition was treated as a stroke, and after ruling out intracranial hemorrhage, the patient underwent thrombolysis. After the procedure, she presented intense holocranial headache, which peaked in less than 1 minute. She was transferred to the intensive care unit, while maintaining neurological deficits and after 1 day, developed weakness in her right lower limb. Despite that, she was discharged from the hospital, returning

after 2 days due to persistent headache. A new cranial CT scan revealed subarachnoid hemorrhage and she was readmitted. Fifteen days after the onset of symptoms, she was transferred to a tertiary service, where neurological examination revealed flaccid paraplegia, associated with paresis in her left upper limb, along with areflexia in the lower limbs and left upper limb, with normal reflexes in her right upper limb. Regarding sensitivity, she presented anesthesia, hypoesthesia and anarthresthesia on her left hemibody up to C2 level, with preserved sensitivity on the right side. MRI of the skull, cervical and thoracic spine showed hyperintensity on T2/STIR sequences in the central region of the spinal cord, affecting from C2 to D3 level, with mild local mass effect and no contrast enhancement. Cerebrospinal fluid analysis revealed a cell count of 10cells/mm^3, 100% mononuclear cells, protein of 40g/dL and Glucose of 53mg/dL. Other tests did not show significant abnormalities. The patient received pulse therapy with methylprednisolone 1g/day for 2 days, which was suspended due to urinary tract infection. Two days after suspension of pulse therapy, she developed visual blurring in the right eye, prompting reintroduction of pulse therapy, completing a 7-day course. Subsequently, she underwent 7 sessions of plasmapheresis, resulting in partial improvement of strength in her left upper limb and sensitivity on her left hemibody. During treatment, her Anti-Aquaporin 4 antibody test was negative, while anti-mog was positive. She started maintenance therapy with Rituximab 1g every six months. **Discussion:** MOGAD is an autoimmune inflammatory disease associated with antibodies directed against a glycoprotein exclusive to the myelin sheath within the central nervous system (CNS). It presents with diverse clinical manifestations and typically shows significant response to corticosteroid therapy. The case described represents an atypical presentation of the disease, mimicking the presentation of a stroke in an emergency context. **Final comments:** It is necessary to consider the hypothesis of inflammatory CNS diseases in acute neurological contexts that present with atypical and recurrent symptoms.

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Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) with infectious trigger of neurosyphilis: case report

Camila Martins Soares[1], João Marcus de Lima Brito Alves[1], João Pedro Izidoro Gomes[1], Vanessa Lauanna Lima Silva[1], Charles Maroly Lessa Mantovani[1], Camila Aquino Cruz[1], Katharina Vieira Messias[1], Vanessa Daccach[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.

Correspondence: Camila Martins Soares; Email: camila.m.soares25@gmail.com.

Case presentation: Male, 57 years old, smoker, with amblyopia in the right eye (OD) and late latent syphilis treated with 3 doses of benzathine penicillin. He presented with left eye pain, temporal headache ipsilateral and progressive low visual acuity, initially 0.8 progressing in 10 days to counting-fingers at 4m. Proptosis was observed in the left eye (OS). On fundoscopy, pale optic nerve (ON), swelling of the left optic disc, with hyperemia and blurring of the disk margins, in addition to candle flame hemorrhages. Additional investigation showed black visual fields bilaterally, OCT with OD atrophy and edema of the retinal nerve fiber layer (OS). Laboratory results showed positive FTA-ABS and CSF with hyperproteinorrhaquia (52.4) and negative VDRL. Brain and orbits MRI with signs of optic neuritis on the left, highlighting perineuritis, as well as signs of right ON atrophy. Spine MRI showed no lesions. Therefore, treatment was initiated for neurosyphilis with ocular involvement with intravenous ceftriaxone 2g/d for 14 days and pulse therapy with methylpredisolone 1g/d for 5 days. After 8 days of starting treatment, the patient showed improvement in pain and visual acuity of OS of 0.7. However, 2 days later, he presented a new sudden visual loss and left eye pain, with finger-counting visual acuity in front of his face. A new pulse therapy was chosen, concomitant with plasmapheresis, with an improvement in visual acuity (0.5 in OS).The anti-MOG antibody (cell-based assay) test came back positive in the blood (1:10), confirming the diagnosis of myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD), with an infectious trigger of neurosyphilis being hypothesized. **Discussion:** Ocular syphilis is a clinical manifestation that can occur at any stage of the disease. It is known that some autoimmune diseases have an overlying infectious condition as trigger for the onset of the disease, as the MOGAD. There is another report of anti-MOG optic neuritis associated with neurosyphilis, in addition to other etiologies, such as HIV infection, SARS-COV-2 and CMV. It is known that MOGAD can present with atypical optic neuritis, transverse myelitis, acute disseminated encephalomyelitis and focal cortical lesions. Optic neuritis can present as perineuritis, making a differential diagnosis with other inflammatory optic neuropathies. However, incomplete response or recurrence after treatment of the infectious condition is an alert for an additional pathophysiological mechanism. **Final comments:** The association of neurosyphilis with MOGAD suggests it as a possible trigger, being the second case reported in the literature. The association of autoimmune diseases with infectious conditions must be remembered when faced with atypical progression, highlighting the importance of investigating differential diagnoses. In patients with neuritis and neurosyphilis, anti-MOG testing may be considered.

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Myelitis as a paradoxical manifestation of tuberculosis in an immunocompetent patient

Jesângeli de Sousa Dias[1,2], Larissa Blanc Morais[2], Betânia Mara de Freitas Nogueira[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Secretaria de Saúde do Estado da Bahia, Instituto Couto Maia, Salvador BA, Brazil.

Correspondence: Jesângeli de Sousa Dias; Email: jesangelidias@gmail.com,

Case presentation: A 22-year-old immunocompetent woman was readmitted to the hospital complaining of acute bilateral leg weakness which began seven days earlier and progressed to complete paralysis within 48 hours associated with fecal and urinary retention, however without pain, fever, or trauma. Fifty days ago, she was diagnosed with disseminated tuberculosis including meningeal involvement which has improved after taking anti-tuberculosis drugs associated with prednisone (40mg/d). Of note is that the prednisone dose was gradually decreasing when symptoms began. The neurological examination showed a bilateral lower limb flaccid paralysis with normal patellar-deep tendon reflexes, but absent ankle-deep tendon reflexes. The sensitivity to pain and touch below a T8 level and a position sense in her feet were reduced. A thoracolumbar spine CT and CSF examination with the molecular test were normal. Previously, there was no abnormality on neurological examination, while a CSF study showed 30 cells/mm3 (86% neutrophils, 10% lymphocytes, and 4% monocytes), glucose 38mg/dL, protein 50mg/dL, and lactate 3,20 mmol/L. GeneXpert MTB/RIF detected M. tuberculosis in CSF and urine, also the bacteria grew in CSF culture. The tuberculosis treatment was continued, and prednisone was replaced by intravenous dexamethasone (16mg/day) without improvement. A thoracolumbar spine MRI spine performed 7 days after her admission showed edema and nodular lesions suggestive of granuloma inside the spinal cord at T8 and T12-L1 transition levels. After it, myelitis as a paradoxical reaction was confirmed and she was treated with an intravenous methylprednisolone (1 g/d) for three days. A few weeks later, the patient began to improve. She could walk without assistance and had better fecal and urinary control 8 months later. **Discussion:** The paradoxical reaction occurs during tuberculosis treatment. It results from an exaggerated host immune response to the mycobacterial antigen manifesting as worsening or appearance of new signs, symptoms, or radiographic manifestation of tuberculosis following a good response to the treatment. It appears between 2 weeks to 8 months after starting the medications, mainly within 12 weeks. Patients with tuberculous meningitis (TBM) are prone to this reaction, as well as those with high bacillary load. Corticosteroids used within the first 2 months to reduce TBM mortality, may prevent these complications. In this case, an early reduction of prednisone dose could trigger the reaction. Although reactions are treated with high corticosteroid doses, there is no definition of which treatment is best. A methylprednisolone pulse therapy was prescribed, and she improved. **Final comments:** Tuberculosis affects the nervous system in several ways, including paradoxical reactions. The diagnosis and treatment are challenging. Physicians should be aware of this manifestation, and that early diagnosis as well as appropriate treatment can prevent neurologic damage.

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Myeloradiculitis from wasps swarm sting

Luis Fernando Guimarães[1], Osvaldo Pantoja de Oliveira[1], Adriane Paz Rocha[2], Jeferson Henrique Pereira[2], Marina Brasileiro Cesar Leitão[2], Bárbara Luisa Coraspe Gonçalves[1], Maria Luiza Levindo Coelho Martinis[1], Yasmin de Freitas Santos[1], Luiz Guilherme Gerhardt Fernandes[1]

[1]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.
[2]Universidade Estadual de Montes Claros, Montes Claros MG, Brazil.

Correspondence: Luis Fernando Guimarães; Email: luisguimaraesneuro@gmail.com.

Case presentation: A 53-year-old man, resident of a rural area, arrived at the emergency room with weakness in his lower limbs that had started two years ago. He reports that he was affected by numerous wasp bites on his lower limbs. He developed paraparesis and urinary retention hours later. It was necessary to use an indwelling urinary catheter. On physical examination: preserved tactile sensitivity, epicritic touch and impaired proprioception distally, in the lower limbs, worse on the right; paraparesis more evident in dorsiflexion (3/5 on the right and 2/5 on the left) and dorsoextension (4/5 on the right and 4/5 on the left); Achilles areflexia on the right. Complementary exams: magnetic resonance imaging (MRI) of the dorsal spine and skull; cerebrospinal fluid; tomography of the lumbar spine, abdomen and chest; electroneuromyography (ENMG); serology for syphilis, HIV, HTLV, hepatitis B and C, herpes simplex; research for ANA, Anti-Ro, anti-La and protein electrophoresis, Anti-MOG, Anti-Aquaporin-4; fecal parasitology (Kato-Katz) and rectal biopsy for research into schistosomiasis. Only MRI and ENMG showed changes. MRI of the dorsal column showed enlargement of the spinal cord in conus medullaris from D10 to L1 (hyposignal on T1, hypersignal on T2 and STIR), heterogeneous myeloradicular contrast enhancement, suggesting inflammatory-infectious disease. Brain MRI showed gliosis. ENMG showed acute axonopathy bilaterally, more evident on the left with L5 and S1 root involvement. The diagnosis of myeloradiculopathy resulting from wasp bites was confirmed. He received methylprednisolone, 1gram/day, for five days. The patient progressed with partial improvement of the paraparesis, but continued to require intermittent bladder catheterization. He is undergoing neurological monitoring. **Discussion:** Millions of cases of hornet bites occur every year around

the world. They are accompanied by local inflammatory reactions and, less frequently, by severe systemic reactions as a result of an IgE-mediated allergic reaction, which can lead to death. Anaphylaxis is a known consequence, but there are other unusual systemic reactions that can occur hours to weeks after stings. Among these uncommon reactions, we find neurological ones such as: myelitis, encephalitis, optic neuropathy, encephaloradiculoneuritis, cerebral infarction, acute inflammatory polyradiculopathy and axonal demyelination. We present the case of a patient diagnosed with myeloradiculitis secondary to wasp stings. **Final comments:** We emphasize the possibility of nervous system involvement in patients who have suffered wasp stings and, therefore, the main findings of the physical examination and complementary tests that, related to the clinical history, confirm the diagnostic hypothesis of wasp sting myeloradiculitis. Neurological changes in these cases are rare, little described in the literature and vary between affected individuals.

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Myokines as biomarkers of severity and progression in spinal muscular atrophy

Ana Letícia Amorim de Albuquerque[1], Júlia Kersting Chadanowicz[1], Arthur Carpeggiani Weber[1], Giovanna Câmara Giudicelli[2], Marina Siebert[2], Thayne Woycinck Kowalski[2], Michele Michelin Becker[2], Jonas Alex Morales Saute[2]

[1]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Ana Letícia Amorim de Albuquerque; Email: analeticia.neurologia@gmail.com

Background: Spinal muscular atrophy (SMA) is a neurodegenerative disease characterized by progressive muscle atrophy and weakness due to the death of spinal cord motor neurons. The identification of biomarkers is essential for prognostic prediction and response to advanced specific therapies. **Objective:** Since muscle damage secondary to muscle denervation is one of the main clinical manifestations of SMA, we aimed to characterize the potential of myokines, myostatin, follistatin, and irisin as biomarkers. **Methods:** Differential gene expression of myokines was evaluated in a denervation lesion model of the gastrocnemius muscle in mice, as well as in a meta-analysis of RNA-seq datasets from three studies in Smn/;SMN2 SMA mice and two studies in SMA patients. Following this, a case-control study was conducted with 27 SMA patients and 27 controls, followed by a 12-month cohort study with the cases (n=25). Serum levels of myostatin, follistatin, and irisin were measured using enzyme-linked immunosorbent assay (ELISA). **Results:** Gene expression of myostatin and irisin was reduced, while follistatin was increased after muscle denervation in mice. The results were similar in the meta-analysis of the transgenic SMA mouse model and in the iliopsoas of 5 type 1 SMA patients. Consistent with these results, serum myostatin levels were markedly reduced in SMA patients, with a median of 98 (5-157) pg/mL compared to 412 (299-730) pg/mL in controls ($p<0.001$). Lower myostatin levels correlated with greater disease severity according to different clinician-rated outcomes (Rho=0.493-0.812; $p<0.05$). After 12 months, there was a further reduction in myostatin levels among cases ($p=0.021$). There was no statistically significant difference between cases and controls or correlation with severity variables in levels of follistatin or irisin. **Conclusion:** Serum myostatin level is a promising new biomarker for assessing the severity and progression of SMA. The reduction in myostatin levels and the consequent cellular environment favorable to muscle growth can be attributed to denervation-induced motor neuron dysfunction.

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Mysterious case of subacute corticosteroid-dependent rhombencephalitis

Thire Baggio Machado Marazzi[1], Bruna Pereira Correia[1], Ellen Silva de Carvalho[1], Renato Ramon da Cruz[1], Soraia Fábio Ramos Cabette[1], Francisco Antonio Coletto[1], Raissa Gontijo Cabral de Souza[1], Gabriel Rosa Vilela[1]

[1]Hospital Unimed, Ribeirão Preto SP, Brazil.

Correspondence: Bruna Pereira Correia; Email: brunapereirac@hotmail.com.

Case presentation: A 67-year-old male from a rural area with no significant previous medical history was admitted to a tertiary care hospital with a 30-day history of progressive headache, vertigo, cerebellar ataxia, and diplopia. His brain MRI demonstrated diffuse leptomeningeal enhancement, an expanding lesion with hypersignal on T1/FLAIR affecting both cerebellar hemispheres, cerebral peduncles, mammillary bodies, and bilateral basal ganglia. CSF analysis showed 80 cells (98% lymphocytes), 113 protein glucose 67, cultures, and direct testing for bacteria and fungi were harmful. A hypothesis of rhombencephalitis associated with chronic meningitis was suggested, and the patient underwent treatment with ampicillin and ceftriaxone for 21 days, with slight clinical improvement and expansion of the 4th ventricle. Approximately 45 days after the onset of symptoms, the patient developed uncontrollable vomiting with signs of decompensated intracranial hypertension. Corticotherapy was started with excellent clinical response. Investigations were expanded to include multiple fungal serologies, CSF FilmArray® Meningitis/Encephalitis (ME) Panel and metagenomic next-generation sequencing (mNGS), all negative. A search for a hidden neoplasm was carried out using serial images and markers, and a PET CT was completed without significant findings. Brain biopsy was considered high-risk and contraindicated. Approximately four months after the onset of symptoms, the patient developed a progressively worsening headache and bilateral papillary edema. New CSF demonstrated increased opening pressure (40 mmHg, all previously normal) and positive latex agglutination for fungi with an antigen titer of 1:1280. Definitive treatment was performed with liposomal amphotericin, highlighting the immense challenges in diagnosing and treating this rare case. **Discussion:** This report presents a unique and challenging case of neurocryptococcosis in an immunocompetent patient. Despite high-accuracy tests, the infectious agent was only detected in the CSF after immunosuppression induced by corticosteroid therapy. This highlights the complexity and challenges in diagnosing such cases and the need for further research in this area. Previous literature reports of false negatives with film array and metagenomics have already been described but are scarce. **Final comments:** Although this report was developed with an immunocompetent patient, an infectious agent was only detected in CSF after immunosuppression induced by corticosteroid therapy. Despite being highly accurate, we must underline the potential for false negatives in molecular tests, a crucial point for further research and caution in similar cases.

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Narcolepsy, an atypical presentation of neuromyelitis optica: a case report

Fabrício Martins Veras[1], Gabriel Farina Giovani[2], Laura Luiza Barbosa Menezes da Mota[3], Evelliny Gomes da Silva[1], Allefy Beltrão Albano[1], Letícia Klabinske Marques Monteiro[1], José Luiz de Miranda Coelho Inojosa[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Recife PE, Brazil.
[3]Universidade de Pernambuco, Recife PE, Brazil.

Correspondence: Evelliny Gomes da Silva; Email: evellinygomes1@hotmail.com.

Case presentation: A 20-year-old woman reported excessive sleepiness, being confined to bed most of the time and dependent on others to carry out daily activities. She was initially diagnosed with a mood disorder and treated with lithium carbonate, with no significant improvement in her condition. After about 6 months, she began to lose her visual acuity and was admitted to a neurology ward for further investigation. On admission, she had bilateral visual acuity of 20/200, pallor of the optic papillae on fundoscopy, isophotoreagent pupils and no relative afferent pupillary defect. During the anamnesis and physical examination, it was noted that the patient suddenly had episodes in which she appeared to fall into deep sleep. No other alterations were found on physical examination. Magnetic resonance imaging revealed bilateral sequelae of gliotic lesions in the paramedian region of the thalamus, with apparent extension to the posterior hypothalamus and an impression of optic nerve blurring on the left. In view of the suspicion of neuromyelitis optica spectrum disease, a positive anti-AQ4 test was requested. The patient was started on modafinil 100 mg/day to control sleepiness and rituximab, with guidance on laboratory follow-up. At the outpatient clinic, the patient reported an improvement in sudden sleep episodes and no new outbreaks of the disease. **Discussion:** Narcolepsy is a clinical syndrome characterized by excessive daytime sleepiness with cataplexy. Its cause may be related to the loss of orexin signaling, produced in the lateral hypothalamus, whose excitatory effects are important for maintaining wakefulness. Genetic factors and rare brain lesions can also cause the disease. As it has a non-specific clinical course, it is commonly diagnosed as a mood disorder or functional disorder, which is detrimental to the individual living with the disease. Neuromyelitis optica (NMO) and narcolepsy are two distinct diseases, but both can manifest with unusual clinical phenotypes, which creates several challenges in diagnosis and treatment. The discovery of the anti-aquaporin-4 (AQP4) antibody in patients with NMO has transformed the understanding of the disease, allowing the identification of cases beyond the traditional phenotype. These patients can exhibit a wide range of neurological symptoms, including narcolepsy associated with hypothalamic lesions. Early detection of patients with atypical presentations of NMO and narcolepsy is essential in order to start treatment quickly, with the aim of reducing disability and preventing further attacks. This underscores the importance of a thorough investigation of neurological symptoms and complementary tests for an accurate diagnosis and effective clinical management of these conditions. **Final comments:** It is important to consider differential diagnoses in patients with atypical neurological symptoms, such as excessive daytime sleepiness and loss of visual acuity.

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NCSE in the elderly: clinical-EEG variables and prognosis at 30 days according to acute and remote etiology

Gloria M. A. S. Tedrus[1], Rodrigo Barros de Pádua Andrade[1], Bruno Ferreira Faria[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Gloria M. A. S. Tedrus; Email: gmtedrus@uol.com.br.

Background: In the elderly, nonconvulsive status epilepticus (NCSE) is frequent, however there are still gaps in knowledge about the types of presentation, ictal EEG patterns and prognosis. It is questioned whether the prognosis is related to the etiological factor. **Objective:**

To evaluate the prognosis and clinical-EEG variables in older adults with NCSE according to remote or primary acute etiology. **Methods:** Clinical-EEG data and the occurrence of death at 30 days in 90 elderly patients diagnosed with NCSE of structural etiology (primary acute or remote) treated at the clinical neurology service of the PUC-Campinas Hospital were evaluated and compared. **Results:** The mean age was 72.6±8.3 years and 57(63.3%) cases were female, 53(58.9%) cases had acute etiology and 37(41.1%) had remote etiology. The NCSE type was - NCSE-coma in 41(45.6%) cases and NCSE-without coma in 49(54.4%) cases. No significant difference was observed in gender and age according to the type of etiology. There was no difference in the ictal EEG pattern (Salzburg Criterion - A1, A2, A3, A4) according to acute or remote etiology. Death within 30 days occurred in 32(35.6%) cases, with no difference according to etiology. Previous history of seizures/epilepsy occurred more and significantly in older adults with NCSE of remote etiology (Fisher's exact test, 6(11.3%) vs 17(45.9%), p< 0.000). There was a difference in the type of NCSE according to the etiology, with a higher occurrence of NCSE-coma in cases with acute etiology (chi-square test; 30 vs 11, p=0.012). **Conclusion:** In the elderly with NCSE, there were differences in the clinical variables according to the acute/remote etiology. Previous history of seizures/epilepsy occurred more frequently in the NCSE of remote etiology, and the clinical presentation of coma was more frequent in the elderly with NCSE of acute etiology. Mortality in this sample was high, with no difference according to acute/remote etiology in elderly patients with NCSE. FAPESP 2023/01226-4.

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Needle electromyography in patients with amyotrophic lateral sclerosis: a cohort with 81 patients

João Vítor Ribeiro dos Santos[1], Rômulo Cerqueira Heckert dos Santos[1], Salim Lázaro Balassiano[1], Eduardo Rodrigues Davidovich[1], Robson Teixeira Vital[1], Clarissa Neves Spitz[1], Larissa Bittencourt de Carvalho Chaffin[1], Livia de Almeida Afonso Abihaila[1], Isadora Versiani de Lemos[1], Marcia Rodrigues Jardim[1]

[1]Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.

Correspondence: João Vítor Ribeiro dos Santos; Email: jvriber@gmail.com.

Background: ALS represents the main entity within the group of Motor Neuron Diseases (MND). It is characterized by the progressive, painless degeneration of the first and second motor neurons. It is a disease that imposes a significant burden of morbidity and mortality on patients. Therefore, it is important to have tools that assist not only in the diagnosis but also in the prognosis of these patients. The diagnosis of ALS is based on clinical findings, however the neurophysiological study can support the diagnosis when it is not defined by the clinic. Furthermore, the neurophysiological study is an important tool for the differential diagnosis of ALS. **Objective:** To demonstrate the neurophysiological findings of patients diagnosed with ALS (Amyotrophic Lateral Sclerosis) at the Pedro Ernesto University Hospital. **Methods:** Eigthy one patients were diagnosed with ALS (Revised El Escorial Criteria). Patients with comorbidities that lead to Peripheral Nervous System impairment or Central Nervous System diseases were excluded. The criteria analyzed were: 1) fibrillations and positive sharp waves; 2) fasciculations; 3) signs of chronic axonal loss. The findings were topographically mapped in the following segments: 1) cervical; 2) thoracic; 3) lumbar; 4) bulbar. The patients were classified according to the El Escorial criteria: 1) Suspected; 2) Possible; 3) Probable; and 4) Definite. **Results:** 81 patients, findings were present in 1 segment in 5% (4) of patients; in 2 segments in 25% (20) of patients; in 3 segments in 32% (26) of patients; and in 4 segments in 36% (29) of patients. Among the patients with definite ALS, 3% showed alterations in 1 segment, 20% in 2 segments, 28% in 3 segments, and 50% in 4 segments. For those with probable ALS, 18% had neurophysiological findings in 2 segments, 73% in 3 segments, and 9% in 4 segments. Patients classified as having possible ALS showed a wider range of results 11% had normal exams, 22% had alterations in 2 segments, 33% in 3 segments, and another 33% in 4 segments. Finally, among the 21 patients with suspected ALS, 5% had normal exams, 14% showed alterations in only one segment, 38% had two segments affected, and 24% had findings in all four segments. In cases with suspected ALS, 43% of patients had findings in 3 or 4 segments. In patients with possible ALS, 66% of patients (2/3) have findings in 3 or 4 segments. Chronic denervation (long polyphasic motor unit potentials with or without large amplitude and neurogenic recruitment) were found in 91% of the total 81 patients studied. Membrane instability findings as acute fibrillations / positive sharp waves, and fasciculations were observed in 68% and 63%, respectively. **Conclusion:** The findings from the neurophysiological study of patients with ALS can be a tool to increase the safety of diagnosing a disease that has a poor prognosis. Furthermore, these findings allow us to propose future studies to evaluate the loss of motor units in patients with ALS.

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Nephrogenic diabetes insipidus following carbamazepine withdrawal in the treatment of epilepsy: case report

Isabela de Carvalho Florêncio[1], Rodrigo Ibañez Tiago[2], Carolina Cândida Vaz[1], Rodolfo Antonio Corona[1], Mateus Magalhães Barbosa[1], Renata Gratão Rezende[1], Guilherme Augusto de Oliveira Soares[1], Gabriel Pereira Braga[1], Elder Yamazane Oda[1]

[1]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.
[2]Universidade Federal de Mato Grosso do Sul, Faculdade de Medicina, Campo Grande MS, Brazil.

Correspondence: Rodrigo Ibañez Tiago; Email: rodrigoibaneztiago@gmail.com.

Case presentation: A 60-year-old woman, on carbamazepine for 49 years due to focal epilepsy, was admitted to the emergency department because of refractory status epilepticus, requiring sedation, analgesia, and orotracheal intubation. The accompanying person reported that the medication had been discontinued a week prior and valproic acid initiated without apparent reasons. Following medication cessation, she began experiencing daily episodes, escalating to a bihemispheric tonic-clonic seizure lasting over 30 minutes on the day of admission. Brain tomography showed no abnormalities. EEG post-sedation did not reveal electrographic status epilepticus. She developed right-sided bronchoaspirative pneumonia, treated with piperacillin and tazobactam. During weaning off mechanical ventilation and sedation, she developed polyuria (maximum urine output 10,200 mL/24h) associated with refractory hypernatremia and hypokalemia. Nephrogenic Diabetes Insipidus (NDI) was diagnosed, resolving upon carbamazepine reintroduction. She was discharged after 11 days of hospitalization. **Discussion:** NDI is a rare condition characterized by an inadequate response to vasopressin, resulting in the kidneys' inability to concentrate urine and subsequent electrolyte disturbances. Carbamazepine-induced hyponatremia is common due to upregulation of vasopressin V2 receptors (V2R) in the collecting duct and consequent upregulation of aquaporin 2 (AQP2), which is why it can be used in the treatment of Diabetes Insipidus. Abrupt withdrawal of the medication may have disrupted homeostasis, reducing V2R and AQP2 expression. To the best of our knowledge, there are no reports of NDI caused by this medication discontinuation. **Final comments:** The presented case illustrates a rare and potentially serious complication associated with prolonged use and inadequate management of carbamazepine. Early diagnosis and medication reintroduction were essential for resolving the patient's clinical condition. Attention to the adverse effects of antiepileptic medications is crucial for the effective and safe management of patients with long-standing epilepsy.

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Nervous system tuberculosis in Bahia and Brazil: an ecological study

Fabrício Rodrigues Barbosa[1], Ester Costa Lima[1], Victor Alex Purificação de Castro Neves[1], Lucas Granja Kaupert[1], Paula Cely da Silva Torres[1], Paulo Vitor Silva Santos[1], Jéssica de Andrade Ribeiro Lima[1], Karollyne de Castro Santos[1], Bruna Rafaela Bastos Freire Quixabeira[1], Alexandre David do Nascimento Santos[1]

[1]Universidade do Estado da Bahia, Salvador BA, Brazil.

Correspondence: Ester Costa Lima; Email: ester.enfe@gmail.com.

Background: Tuberculosis (TB) is a serious public health problem, especially in developing countries. Although it mainly affects the lungs, TB can spread to the Central Nervous System (CNS), resulting in neurotuberculosis, a severe form of extrapulmonary TB. This condition causes sensory and motor deficits, seizures, and cognitive changes. Understanding the epidemiological profile of hospitalizations and deaths due to neurotuberculosis is crucial for early diagnosis and prevention of permanent neurological damage. **Objective:** To compare the epidemiological profile of hospitalizations and deaths due to tuberculosis in the nervous system in Brazil and Bahia from 2013 to 2023. **Methods:** This ecological, retrospective, cross-sectional, descriptive study used data from the Hospital Admission System (SIH/SUS-DATASUS) from 2013 to 2023. ICD-10 codes were used to select data on total costs, hospitalizations, and deaths by sex, color/race, and age group in Brazil and Bahia. Data analysis was performed using Microsoft Office Excel® 2016. **Results:** From 2013 to 2023, Brazil spent R$7,323,229.72 on CNS TB hospitalizations, with Bahia accounting for 4.8% of this. The average length of hospital stay in Brazil was 15.9 days, higher for women (16.2) than men (15.7). Mixed-race individuals had the longest stays (16.4 days), and indigenous people the shortest (8 days). In Bahia, the average stay was 17.5 days, with women averaging 18.2 days and black individuals 19.3 days, while white individuals had the shortest stay (5 days). The mortality rate in Brazil for CNS TB was 10.92 deaths per 100,000 inhabitants. Evaluating by age group, the rate increased with each decade above 40 years: 40-49, 11.34; 50-59, 12.15; 60-69, 13.24; 70-79, 24.55; and over 80, 28.95. In Bahia, the mortality rate was lower than the national average (10.46), but the increasing trend with age was maintained: 30-39 years, 13.04; 50-59 years, 21.05; and 70-79 years, 33.33. No data were available for those under 15. Nationally, female mortality was higher (11.10/100,000) compared to males, while in Bahia, male rates (10.53) were slightly higher than female rates (10.39). Indigenous people had the highest mortality rate nationally (13.33), while white people had the lowest (9.20). In Bahia, black individuals had the highest rate (25), followed by mixed-race individuals (8.62). Data for other races were not available for Bahia. **Conclusion:** Bahia had a slightly lower mortality rate than the national average but longer hospital stays. Women and black individuals are at higher risk, with higher hospitalization and mortality rates. These disparities highlight the need for public policies to reduce inequalities and improve access to early diagnosis and treatment of neurotuberculosis. More studies are needed to understand these factors and guide effective interventions.

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Neuro-Behçet disease mimicking viral rhomboencephalitis: a case report

Renato Morais Bueno[1], Rudá Alessi[1], Miriam Eva Koch[1], Eduardo de Souza Tieppo[1], Victor Alexandre Dantas de Miranda[1], Ana Claúdia Piccolo[1], Victor Ezaki Alves Silva[1], Guilherme Bitencourt Barbosa[1], Rayanne Pereira Barbosa[1], Atílio Gomes Romani[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Renato Morais Bueno; Email: renatombueno@hotmail.com.

Case presentation: A 35-year-old female from São Bernardo do Campo (SP, Brazil) was admitted to hospital due to diplopia associated with headache and drowsiness 15 days previously. On physical examination, she presented bilateral oculomotor involvement, more evident on the left, as well as trochlear and abducens involvement. Initially treated as viral meningoencephalitis, with no clinical response. A cerebrospinal fluid puncture showed an increase in cellularity of 120 (82% neutrophils and 18% lymphocytes), with proteins of 38 and other parameters within the reference value. Rapid molecular test for turberculosis, VDRL and viral serologies in the cerebrospinal fluid were negative. After extensive investigation, with no evidence of an infectious process, methylprednisolone was administered for 5 days, with a significant clinical response. The patient was admitted to hospital 2 years later with recurrence of symptoms and reported oral and genital ulcers. Magnetic resonance imaging of the skull showed hyperintensity in the right thalamus, midbrain and pons on T2 and FLAIR scans, in a "Waterfall sign" - or Cascade Sign - with no diffusion restriction and no contrast uptake, suggestive of an inflammatory process in the brainstem. Other imaging studies and vessels were unchanged. Once again, the patient showed a good response to pulse therapy and, due to the data presented, the hypothesis of Neuro Behcet's was made. **Discussion:** Involvement of the hindbrain is a challenge for neurologists due to the broad spectrum of diseases that can trigger signs and symptoms typical of this area. Following the flowchart of the most prevalent diseases, investigation of the cerebrospinal fluid is essential in these cases, but is often not very elucidating, especially when the etiology cannot be determined, either due to lack of resources or lack of confirmatory tests. In the case above, what contributed to the diagnosis was the set of clinical, anamnesis and imaging findings. It is worth remembering that the "Waterfall Sign" - an involvement of the midbrain on MRI, extending to the thalamus or internal capsule - is a suggestive sign, but not pathognomonic of Neuro-Behçet. Furthermore, the presence of other systemic manifestations, such as oral and genital ulcers, corroborates the diagnosis - having, of course, ruled out other pathologies such as syphilis. Finally, the good response to pulse therapy is another factor that points to the pathology in question. **Final comments:** Investigating autoimmune diseases and central nervous system vasculitis is a challenge for any neurologist. This becomes more evident when there is no specific laboratory test for the pathology. The clinical history and the progression of the disease are essential in these cases for investigation. In this case, the patient scored 5 points on the International Criteria for Behcet's disease (ICBD-2006), requiring at least 4 to make the diagnosis.

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Neuro-Behçet with subacute onset accompanied by classic neuroradiological sign: a case report

Ana Luisa Suqueno[1], Pedro Simões dos Santos Pilau[1], Antônia dos Santos Bobrowski[1], Barbara Suman Bahlis[1], Bernardo Sommer Cimirro[1], Guilherme Vanik Pinto[1], Marco Antônio Delazeri[1], Maria Caterina Bortolazo Pepato[1], Maria Paula Hanel[1], Vitor Augusto Cioquetta[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Ana Luisa Suqueno; Email: analuisasuqueno@gmail.com.

Case presentation: A 57-year-old female patient presented to the emergency department due to weakness and loss of tactile sensation in her right hemibody, with a subacute onset and progressive worsening over the past 3 weeks. Physical examination revealed grade IV hemiparesis and tactile hemihypoesthesia on the right side, associated with hyperreflexia of the ipsilateral bicipital, patellar, and achilles reflexes. Basic laboratory tests and non-contrast head CT were normal. To further investigate, a contrast-enhanced MRI of the head and lumbar puncture with basic cerebrospinal fluid studies were performed. The MRI showed a poorly defined hyperintense lesion on T2/FLAIR and hypointense on T1 with subtle diffusion restriction, extending to the internal capsule, left cerebral peduncle, and ipsilateral superior cerebellar peduncle, crossing the midline at the thalamomesencephalic junction: the waterfall or cascade sign. Cerebrospinal fluid studies were normal. Due to the characteristics of the lesion, additional investigation included another MRI with spectroscopy and perfusion, blast cell search, cerebrospinal fluid cytology, and rheumatological profiling. The new MRI showed no capillary permeability alteration, but demonstrated reduced N-acetylaspartate peak and increased lactate/lipid and choline peaks. Blast cell search and cerebrospinal fluid cytology were negative for malignancy. Rheumatological profiling was normal. Neurosurgical biopsy of the lesion was difficult due to its location. Considering the clinical picture and complementary tests, three main hypotheses were considered: vasculitis (specifically Neuro-Behçet's disease), inflammatory/infectious processes, and low-grade glial neoplasm. Vasculitis, particularly Neuro-Behçet's disease, was deemed the most probable diagnosis. Thus, intravenous pulse therapy with methylprednisolone 1g/day for 5 days was initiated, resulting in significant improvement of the aforementioned complaints. Additionally, maintenance treatment with cyclophosphamide every six months was decided. **Discussion:** Neuro-Behçet's disease can be divided into parenchymal and non-parenchymal types, with the former being more common, presenting with brainstem syndromes characterized by acute or subacute onset, slow progression, and neuroradiological manifestations as described above. Elevated proteins and absent oligoclonal bands in cerebrospinal fluid are characteristic. The disease has a broad variety of differential diagnosis, requiring extensive investigation to exclude similar pathologies. Acute-phase treatment usually involves intravenous methylprednisolone, while maintenance therapy with immunosuppressants aims to prevent relapses. **Final comments:** Neuro-Behçet's disease is a rare condition with extensive differential diagnosis and varied clinical presentations, posing a high risk of morbidity and mortality. Accurate diagnosis and treatment are crucial to prevent major deficits and loss of patient functionality.

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Neuro-Behçet's syndrome presenting as hemorrhagic lesions: a case report

Bruna Silva Lima[1], Beatriz Pires Paes[1], Poliana Lima Campos[1], Beatriz Ballarin Costa[1], Anthony Magalhães Morais Santiago[1], Ernane Pires Maciel[1], Camilo Carvalho[1]

[1]Hospital de Base do Distrito Federal, Brasília DF, Brazil.

Correspondence: Bruna da Silva Lima; Email: brunasilval@yahoo.com.br.

Case presentation: A 37-year-old male, smoker, presents episodes of fever, progressive headache, vertigo, weight loss of 6kg, blurred vision in the right eye, and sudden hemiparesis on the right side. At the time, a CT of the brain disclosed a hemorrhagic lesion in the left thalamus, internal capsule, and mesencephalon, and contrast-enhanced cranial MRI showed low-intensity lesions on T1 with areas of hyperintensity, as well as heterogeneous T2 images with areas of hyperintensity/hypointensity on the left cerebral peduncle extending to basal pons, thalamus, and internal capsule. He was then admitted to the hospital 6 months after the initial symptoms and progressed with decreased consciousness, requiring endotracheal intubation. After extensive investigation of the previous pathological history, it was discovered that the patient presented with recurrent painful and ulcerated oral and genital lesions in the past 5 years that were treated as sexually transmitted infections. During his stay at the hospital, he presented new aphthous ulcers and acneiform papulopustular lesions associated with altered inflammatory tests. In this case, the hypothesis of Neuro-Behcet's disease (NBD) was raised, and the patient underwent treatment with Cyclophosphamide and high-dose corticosteroid. He showed neurological improvement and resolution of oral lesions. **Discussion:** Behcet's disease (BD) is a rare autoimmune vasculitis disorder. More prevalent in males aged 30 to 40 years. The most commonly used diagnostic criteria require recurrent oral ulcerations plus two of the following: recurrent genital ulcerations, skin or eye lesions, or a positive pathergy test. NBD is defined as a combination of neurologic symptoms and signs in patients with definite BD. Neurological lesions can be classified into parenchymal, non-parenchymal, and peripheral nervous system involvement. Non-parenchymal lesions involve thrombosis and other stroke syndromes, intracranial hypertension, and meningeal syndromes, being venous sinus thrombosis the most common presentation. Typically, the neurological manifestations result from vasculitis-induced thrombosis, and the most commonly affected region is the mesodiencephalic junction, followed by the pontobulbar region. At the mesencephalic, diencephalic, and pontine levels, thrombosis, necrosis, and rupture of small vessels can cause hemorrhagic lesions. However, intracranial hemorrhages are rare, with most occurring within ischemic lesions. Diagnosis of NBD incorporates clinical symptoms, MRI findings, CSF analysis, and exclusion of other causes since no criteria have been validated. **Final comments:** In cases with suspected NBD in a patient without a previous diagnosis of BD, an extensive guided history and detailed physical exam have to be made. Diagnosis and management can be challenging, especially in cases of atypical presentations such as hemorrhagic lesions, which can delay treatment and worsen the patient's prognosis.

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Neuro-chikungunya: a diagnostic challenge

Victor Hugo de Souza Silva Gomes[1], Letícia Leão Alvarenga[1], Danyelle Rute Francisco e Silva[1], Lucas Wilson Matos Gomes[1], Gabriel Santos de Castro[1], Taysa Alexandrino Gonsalves Jubé Ribeiro[1], Denise Sisterolli Diniz[1], Aracelle Victor do Carmo Faria[1], Delson José da Silva[1]

[1]Universidade Federal de Goiás, Goiânia GO, Brazil.

Correspondence: Victor Hugo de Souza Silva Gomes; Email: victorhhugo2005@gmail.com.

Case presentation: DLAB, 48 years old, female, mixed race, evaluated by the Neurology Service in May 2024 with a history of asthenia, hyporexia, fever (38-38.5°C), myalgia and polyarthritis more than 1 month ago. As symptoms had persisted for around 10 days, she sought medical attention when she developed a sudden onset of generalized paresthesia followed by descending bilateral paresis and reduced visual acuity in the left eye. She denies altered level of consciousness, behavioral disturbance or epileptic seizures. Upon admission, she

had disproportionate tetraparesis with crural predominance, in the right lower limb, tactile hypoesthesia on the left in the topography of V1 and V2 of the trigeminal and in the right hemibody. She also noted bilateral patellar and achilles hyporeflexia. At the ophthalmological evaluation: signs suggestive of retinitis, with visual acuity 20/25 on the right and 20/200 on the left. The CSF examination showed no other changes, except for detectable Chikungunya PCR. On magnetic resonance imaging: signal changes in the left hippocampus, optic neuritis on the left with signs of inflammatory activity and polyneuroradiculopathy. During hospitalization, the patient progresses with worsening paresis and urinary retention. Pulse therapy was administered with methylprednisolone in addition to immunoglobulin infusion for 5 days. It evolves with progressive improvement in the medullary syndrome and, more slowly, in visual changes. **Discussion:** Chikungunya virus (CHIKV) is an arboviral infection caused by Chikungunya virus, an RNA virus from the Togaviridae family. It typically causes fevers and artharlgias and neurological complications are rare. Encephalopathy is the most common CHIKV-associated neurological complication, however, cases of Guillain-Barré syndrome (GBS), myelitis, neuro-ocular diseases (such as optic neuritis), acute disseminated encephalomyelitis and myeloradiculitis have also been reported. Age has been consistently reported as a significant risk factor for severe manifestation of CHIKV, both in the elderly and in infantis. Recognition of CHIKV in patients with neurologic disease requires an understanding of classic presentation of the virus (fever and arthalgias) in people from endemic regions. The CHIKV IgM can be tested in the serum and CHIKV RNA can be assessed via PCR, which is highly specific. Optimal treatment remains not well defined, but case series of patients with GBS or myeloradiculopathy have shown improvement with steroids or intravenous immunoglobulin, as well as with plasma exchange. There are no specific antiviral agents or vaccines at the moment. **Final comments:** Our case report highlights the fact that CHIKV fever may be associated with diverse neurological dysfunctions and should be investigated in patients who live in endemic areas.

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Neurodegenerative profile and MMSE score in patients at a geriatric outpatient clinic

Camila Pezente Dal Toé[1], Debora Dagostin Casagrande[1], Tatiana Pizzolotto Bruch[1], Natalli Studnicka[1], Gabriela Serafim Keller[1], Maria Laura Zanette Naspolini[1], Natália Effting Crema[1], Fabrício Weinheimer Lippert[1], Ana Paula Benedet de Carvalho[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Camila Pezente Dal Toé; Email: camilapezente@hotmail.com.

Evaluate the neurodegenerative profile of first-time patients seen at a geriatric outpatient clinic. Data were extracted from medical records concerning scores obtained on the Mini-Mental State Examination (MMSE), which assesses cognition, in both healthy patients and those diagnosed with neurodegenerative diseases. The study population consisted of 192 individuals, with a mean age of 76.67 years; 34% had a diagnosis of neurodegenerative disease, with 48.4% of these cases being Alzheimer's disease, which showed cognitive impairment in orientation, registration, recall, and language. In Parkinson's disease, significant findings were noted in orientation, registration, attention and calculation, and language. The use of validated scales during consultations aids in recognizing the needs presented by the analyzed patients. This allows for the identification of which cognitive and functional areas require more attention from healthcare professionals in the recovery and systematization of multiprofessional treatment and in promoting healthy aging.

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Neurodemyelinating diseases as a differential diagnosis of ischemic myelopathy with atypical manifestations: a case report

Gabriela da Silva Cremonese[1], Rafael Marques Mendes[1], Cristine Koehler[1], Joana Laurindo da Silva[1], Andre Luiz Pereira Martins[1], Olga Cassol Silva[1], Mariana Costa Borges[1], Maria Eduarda Andreatta[1], Carolina Aparecida Donofrio Santos Costa[1], Gladys Lentz Martins[1]

[1]Hospital Governador Celso Ramos, Florianópolis SC, Brazil.

Correspondence: Gabriela da Silva Cremonese; Email: gabrielacremonese@gmail.com.

Case presentation: A 54-year-old male patient, construction worker, used to experience episodic shock-like sensations through the body for over one year before developing paresis and paresthesia in the upper limbs, progressing to the lower limbs. Due to the worsening of the paresthesia in a "boot and glove pattern", associated with myoclonus in the upper limbs and torso, a neuro demyelinating disease was considered. Electroneuromyography showed an increase in the area of insertion, fibrillation, and fasciculation potentials, with a cluster of fasciculations over the vastus medialis on the right, associated with long polyphasic potentials and reduced muscle recruitment in most motor potentials, and suggested changes in the bulbar and segments paraspinal muscles with signs of active and chronic degeneration of the cervical to lumbosacral segments, more evident in the right hemibody and mainly in the cervical segment and simulating signs of lower motor neuron disease. In addition, non-rhythmic discharges during muscle contraction in the biceps, triceps and dorsal interosseous indicated myoclonus. The myoclonus finding broadened a range of differential diagnoses and represents an uncommon complication of spinal cord infarctions. The MRI indicates radiological signs compatible with ischemia in the internal part of the cervical spinal cord between C4 and C7, predominating in the posterior horns. **Discussion:** Myelopathy refers to any pathological process that affects the spinal cord, often mimicking multiple diseases with rare manifestations. Beyond trauma, the most common etiologies of myelopathy include vascular, autoimmune, infectious, neoplastic, hereditary, neurodegenerative, and idiopathic diseases. As an uncommon symptom of this condition, myoclonias are rapid and involuntary muscle contractions that predominantly represent central nervous system injuries. Spinal cord injuries leading to myoclonus are rare, but they must be taken into consideration for diagnosis and consequently better therapy. **Final comments:** The diagnosis of ischemic myelopathy can be challenging and requires additional tests such as neuroaxis MRI and Electroneuromyography. It is essential to identify the cause of ischemic myelopathy to define the best therapeutic proposal. Therefore, further studies on the different manifestations of this syndrome are essential to bring attention to this differential diagnosis for an effective and early treatment.

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Neurodevelopmental disorders in patients with Duchenne and Becker muscular dystrophy: new evidences for genotype-phenotype correlation

Fabrício Maciel Soares[1], Bruna Faria Rosa[1], Gabriela Marchisio Giordani[1], Daniele Lima Rocha[1], Ana Carolina Brusius-Facchin[2], Michele Michelin Becker[2], Jonas Alex Morales Saute[1]

[1]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.

[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Jonas Alex Morales Saute; Email: jsaute@hcpa.edu.br.

Background: Neurodevelopmental disorders are frequently and heterogeneously diagnosed among patients with dystrophinopathies. The clinical manifestation of intellectual development disorder had already been associated with the underlying DMD genotype, however correlation between others frequently diagnosed comorbidities and genotype is uncertain. **Objective:** We aimed to evaluate how the diagnostic of Attention-Deficit/Hyperactivity Disorder (ADHD), Obsessive-Compulsive Disorder (OCD), or Autism Spectrum Disorder (ASD) and patients genotype are related, as well as whether the prevalence of these comorbidities differ among the genotypic subgroups established. **Methods:** Childhood Autism Rating Scale (CARS), Yale-Brown Obsessive Compulsive Scale for Children (CY-BOCS), and Swanson, Nolan and Pelham IV (SNAP-IV) scale were applied to 50 participants and their caregivers, mainly from the Hospital de Clínicas de Porto Alegre (n=38) or other Brazilian centers (n=12). Participants were grouped according to their genotype and affected dystrophin isoforms. **Results:** The overall diagnostic rate for ASD was 34%, similar to OCD (35.5%). Half of the patients (51.4%) were diagnosed with ADHD. Cerebral isoforms were affected in more than half of the participants (52%). The diagnosis of ASD, CARS scores, and diagnosis of OCD were associated with genotype and impairment of cerebral isoforms. ADHD diagnostic rates were elevated across all the genotypic groups and showed no association with muscular or cerebral isoforms. **Conclusion:** Our results show that patients with DMD and impairment of cerebral dystrophin isoforms have an increased risk for ASD and OCD, and the identification of a patient with a genotype that affects these isoforms should prompt neuropsychological assessment and multidisciplinary follow-up.

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Neurogenetic intersection: a fascinating CMT-CIDP overlap in a Brazilian cohoort

Carolina Braga Moura[1], Ana Luiza Morgado Costa[1], Maria Clara Lemes[1], Gabriel Thomaz Candido[1], Gabrielle Sales[1], Camila Castelo Branco Pupe[1], Osvaldo Jose Moreira do Nascimento[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Ana Luiza Morgado Costa; Email: anamorgado@id.uff.br.

Discerning inherited neuropathies from CIDP, particularly in instances with a subacute onset, poses a diagnostic challenge. In the presence of cases with non-classical clinical findings of CMT, it is important to clinically suspect other entities such as CIDP. It is necessary to search for mutations, such as in the PMP22 gene. **Case presentation:** We described four patients exhibiting the CMT phenotype, including three siblings. The eldest, a 51-year-old female, experienced a subacute onset of proximal weakness at 40 years old, a nerve conduction study demonstrated conduction block, alongside a cerebrospinal fluid analysis indicating albumino-cytological dissociation. These findings led to the diagnosis of CIDP, for which she received IVIG treatment resulting in partial symptom improvement. The second sibling, a 48-year-old female, received a CIDP diagnosis at 30 years old, confirmed by CSF analysis and nerve conduction study, also experiencing improvement after IVIG treatment. The youngest brother presented inflammatory CSF and proximal weakness at 39 years old, with improvement observed following glucocorticosteroid therapy. A fourth patient from our outpatient clinic not belonging to this family nucleus, aged 46, began presenting symptoms in

childhood with changes in gait, accompanied by frequent falls; at 30 years old, an investigation was initiated due to worsening weakness patterns, resulting in a diagnosis of CIDP via NCS and CSF. **Discussion:** All four patients underwent genetic testing for hereditary neuropathies. On physical examination, all four exhibited bilateral pes cavus, legs with an inverted champagne bottle appearance, balance issues, while the three siblings had distal atrophy, and the fourth patient had scoliosis . All tests confirmed CMT 1A with pathogenic mutation in PMP22. The eldest sister demonstrated significant balance impairment and carried a heterozygous PMP22 mutation at chr17:14.236.572 - 15.574.201, while the other sister primarily struggled with neuropathic pain and possessed a PMP22 Gain (Entire coding sequence) copy number of 3. The youngest brother faced progressive balance difficulties and weakening over subsequent years, with a PMP22 mutation at chr17: 14.095.305 - 15.492.541. The fourth patient, similar to the youngest, presents a pattern of imbalance and loss of strength and carries a duplication of the PMP22 gene. **Final comments:** This scenario unveils a rare overlap of CMT and CIDP within a group, underscored by PMP22 mutations. Despite distinct clinical diagnoses, genetic testing provided unequivocal confirmation of this intriguing overlap, shedding light on the complex interplay between genetic neuropathies and CIDP.

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Neurogenic stuttering as a clinical manifestation of acute ischemic brain stroke: a case report

Moacir Pereira Leite Neto[1], Ricardo Queiroz Gurgel[2], Kelly Rejane Leite Figueiredo de Oliveira[3], Cláudio Gleidiston Lima da Silva[1], Isabel Felipe Vásquez[1]

[1]Universidade Federal do Cariri, Barbalha CE, Brazil.
[2]Universidade Federal de Sergipe, São Cristovão SE, Brazil.
[3]Afya Educação Médica, São Paulo SP, Brazil.

Correspondence: Moacir Pereira Leite Neto; Email: moacir.leite@ufca.edu.br.

Cerebrovascular accident (CVA) can cause various neurological deficits affecting speech. We report the clinical case of a woman, who presented neurogenic stuttering as a sequel after an acute ischemic stroke. **Case presentation:** C.O.A, 63 years old, female, married, farmer, from Iguatu-Ce. On 2013 december, she suddenly presented "sensations of numbness in the left side of the face and heaviness in the tongue", mild dysarthria and stuttering that was unusual for the patient until then. In 4 days, only the stuttering persisted. Previously, she was a smoker, had high blood pressure and was diabetic. On general physical examination there were no changes, and the neurological examination of the superior cortical functions was normal, but speech was developed with difficulty. Assessment of the motor system, sensitivity, balance, normal gait and coordination. Complementary exams revealed normal electrocardiogram and echocardiogram, Doppler of the carotid and vertebral arteries showed calcified atheromatous plaques in the right internal carotid artery, causing stenosis of less than 50%, and in the left internal carotid artery, causing 55% stenosis. Laboratory tests performed showed normal blood count, AST, ALT, sodium, potassium, calcium, urea, creatinine, triglycerides and uric acid, LDL cholesterol 145, fasting blood glucose 122. Skull MRI revealed T2 and FLAIR hyperintensity in the left superior and middle frontal gyri, without restriction to diffusion, due to ischemia in the territory of the left middle cerebral artery, and ischemic microangiopathy. **Discussion:** Stuttering is a disorder characterized by disturbances in the normal flow of speech fluency, causing blockages and prolongations in the sounds of words. The neural circuits underlying stuttering as a symptom associated with stroke injury remain unknown, however available evidence suggests that stuttering is not localized to a specific focal region, but rather is the result of a disruption at any level of the cortico-striato-integrative pathway. cortical layer that mediates the execution of speech, and has been described as a symptom of subcortical and cortical infarctions in all lobes, which may be due to disarticulation of speech or impairment of phonology processing. In this case, the presence of sequelae of the neurogenic stuttering type was concluded after an acute ischemic stroke in the left superior and middle frontal gyri, and ischemic leukomicroangiopathy. It was recommended to use acetylsalicylic acid 100 mg per day, atorvastatin 40 mg per day, adjust antihypertensive and hypoglycemic medications, and speech therapy. **Final comments:** A stroke can cause focal neurological deficits that can cause various manifestations, including communication problems, and stuttering is a not very common symptom, whose pathophysiology requires further study, and which requires medical attention for referral to rehabilitation treatment speech therapy.

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Neurolinfomatosis secondary to Burkitt lymphoma

Leonardo Villaverde Buback Ferreira[1], Larissa Braga Mendes[1], Daniel Delgado Seneor[1], Gabriela Rodrigues Tomaz[1], Lara Santos Machado[1], Lucas Toshio Uenishi[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Leonardo Villaverde Buback Ferreira; Email: leonardo.buback@gmail.com.

A 33 year-old man was admitted referring diplopia initiated 1 month prior and insidiously worsening in the following weeks, associated with weakness in both upper and lower limbs, face and with intense lumbar pain. On examination he had lost weight, had inguinal palpable lymphonodes, left ptosis, ophtalmoplegia and midriasis; and right ophtamoparesis with only partial abduction of the eye spared. He had facial diplegia and right hemiface hypostesia, right jaw deviation upon opening and right palatal paresis. An asymmetric tetraparesis was evident, worse in the right upper limb and left lower limb, not being able to offer resistance in any of the four limbs. Deep tendon reflexes were absent and he exhibited intense lumbar pain upon lower limbs movements. His MRI of the brain, cervical, thoracic and lumbosacral spine demonstrated thickening and contrast enhancement at multiple cranial nerves and spinal roots, in addition to leptomeningeal enhancement. A lumbar puncture with cerebrospinal fluid analysis showed 880 cells with atypical characteristics, identified through immunophenotyping as mature B lymphocytes. A bone marrow biopsy was obtained, confirming the presence of a malignant mature B cell neoplasm expressing CD10 and CD38 antigens, compatible with the diagnosis of Burkitt Lymphoma, and characterizing the neurological manifestations as a neurolinfomatosis secondary to the infiltration of lymphoma cells. Central nervous system lymphomas can be further categorized as primary or secondary to systemic disease, as in the present case. In the latter situation, 90% are due to diffuse large B cell lymphoma, and 10% to other types, like T cell lymphomas and Burkitt lymphomas. Their most common neurological manifestations are focal deficits, mental state changes, intracranial hypertension, epilepsy and visual alterations. Up to 40% of central nervous system lymphomas present on imaging as unique or multiple parenchymatous lesions, with restricted diffusion, contrast enhancement and minimal perilesional edema. The leptomeningeal involvement is seen in up to 30% of cases, being more common in secondary lymphomas. Neurolinfomatosis, the cranial nerve, spinal roots and nerves infiltration is rarely seen. The central nervous system secondary infiltration is a severe complication of systemic hematological neoplasms, with multiple possible presentations, among those the rare neurolinfomatosis.

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Neurological challenges in Burkitt lymphoma: a case of methotrexate-induced myelopathy or central nervous system invasion?

César Minoru Toita Koga[1], Anne Caroline de Brito Edir[1], Elenaide Coutinho Nunes[1], Francisco Manoel Branco Germiniani[1], Hélio Afonso Ghizoni Teive[1], Sâmia Talise El Horr de Moraes[1], Gabriel Abrahão Stoliar[1], Camila Emi Fujiwara Murakami[1], Luis Daniel Silva Pilatti[1], Caio César Diniz Disserol[1,2,3]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[2] Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[3]Hospital Israelita Albert Einstein, Curitiba PR, Brazil.

Correspondence: César Minoru Toita Koga; Email: cesarmtkoga@gmail.com.

Case presentation: A 57-year-old male presented with progressive subacute gait impairment, bilateral lower limb numbness, and urinary incontinence. Physical examination revealed impaired pain, vibration, and position sense with a T10 sensory level, brisk deep tendon reflexes in the lower limbs, mild paraparesis, and bilateral extensor plantar response. The patient had been recently diagnosed with Burkitt lymphoma with bone marrow involvement and multiple bone lesions and was undergoing treatment with hyperfractionated cyclophosphamide, vincristine, doxorubicin, dexamethasone, methotrexate and cytarabine chemotherapy (hyper-CVAD regimen), along with alternating intrathecal injections of methotrexate (ITM) and cytarabine for CNS prophylaxis. Symptoms were mild and nonspecific at first, but worsened after the first ITM doses and became severe after the sixth cycle. Laboratory results and cerebrospinal fluid (CSF) with immunophenotypic analyses were unremarkable, including normal levels of vitamin B12 and homocysteine. Magnetic resonance imaging (MRI) of the thoracic spine revealed new longitudinally extensive T2-weighted hyperintensities in the posterior and lateral columns of the thoracic spinal cord, consistent with methotrexate-induced myelopathy. Empiric replacement of vitamin B12 and folate were administered, but no improvement was observed. At the end of the initially proposed therapy, the patient developed complete oculomotor nerve palsy in the right eye. Brain and orbit MRI along with new CSF with immunophenotypic analyses disclosed no new findings. Brain vascular neuroimaging was unremarkable. Neuroaxial MRI revealed improvement of bone lesions, but no new CNS findings. Due to the neurological symptoms, empirical treatment with high-dose intravenous methotrexate was initiated. A whole-body PET scan one month later showed new diffuse bone lesions, and the patient succumbed three months later due to systemic complications from the hematological disease. **Discussion:** This case highlights the complex neurological manifestations in Burkitt lymphoma patients undergoing chemotherapy. It remains unclear whether the patient experienced ITM-induced myelopathy, CNS lymphoma infiltration, or both, given the limitations of CSF analysis and neuroimaging accuracy. ITM is commonly used in hematological malignancy treatment, yet its neurotoxicity can lead to rare neurological complications, such as subacute myelopathy, in 0.8% to 3% of cases. In our patient, initial symptoms worsened post-ITM, suggesting potential neurotoxicity. However, subsequent oculomotor nerve palsy and poor prognosis raised suspicion of lymphoma spreading along the CNS. The lack of highly accurate and specific biomarkers hinders differential diagnosis of CNS lymphoma, delaying early targeted interventions. **Final comments:** Future advancements in biomarker profiling and imaging technologies are crucial to improve outcomes in these complex cases involving aggressive blood marrow neoplasms.

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Neurological complications of cerebrospinal fluid hypotension: a case of cerebral venous thrombosis

Eduardo Rosa da Silva[1], João Victor Meneses de Aguiar[1], Laís Bissoli Perini[2], Bruna Garcia da Silveira[3], Adaucto Wanderley da Nóbrega Junior[1], Bruna Macena Cutti[1], Edson Pilotto Duarte[1]

[1]Hospital Homero de Miranda Gomes, São José SC, Brazil.
[2]Instituto de Assistência Médica ao Servidor Público Estadual de São Paulo, São Paulo SP, Brazil.
[3]Universidade do Sul de Santa Catarina, Palhoça SC, Brazil.

Correspondence: Laís Bissoli Perini; Email: laisbperini@hotmail.com.

Cerebral venous thrombosis is a rare complication of spontaneous intracranial hypotension (SIH). The primary symptom of SIH is severe headache worsened by orthostatic positioning and improved by lying down, often accompanied by cervical pain, nausea, and auditory changes. Diagnosis is confirmed by cerebrospinal fluid opening pressure <6cm H2O and/or evidence of cerebrospinal fluid leakage on imaging studies. Etiology can be spontaneous or secondary to trauma or medical procedures, with cerebral venous thrombosis being a rare complication of this secondary headache type. **Case presentation:** A 38-year-old female presented with fronto-parietal headache associated with neck pain, unresponsive to common analgesics, worsened by orthostasis and relieved by recumbency. There was no alteration in consciousness, fever, or ear pain. She had a history of migraines for 8 years, managed with amitriptyline with good response to prophylactic therapy. There was a change in the characteristics and intensity of the headache compared to her usual pattern. A history of post-spinal anesthesia headache 10 years ago resolved after a blood patch. She denied hormonal contraceptive or therapy use. Neurological exam was unremarkable. Laboratory tests, including negative thrombophilia screening (AntiBeta2glycoprotein IgM and IgG, antiCardiolipin IgG, Lupus anticoagulant, and Antithrombin III 116%), hemogram, and coagulation studies were normal. During hospitalization, she was refractory to optimized combined analgesia. Cranial MRI showed signs of cerebrospinal fluid hypotension, small old infarcts in the periphery of cerebellar hemispheres, with cortical vein filling defect in the left paracentral high convexity, indicative of recent venous thrombosis. **Discussion:** Based on clinical history and findings, the diagnosis was cerebral venous thrombosis secondary to cerebrospinal fluid hypotension. Following anesthesiology consultation, she underwent a blood patch with significant headache improvement. She was also started on full in-hospital anticoagulation and discharged for outpatient follow-up, receiving Xa factor antagonist. **Final comments:** This case report highlights the rare occurrence of cerebral venous thrombosis secondary to cerebrospinal fluid hypotension. Early detection, thorough investigation, and identification of potential triggers are crucial for accurate diagnosis and initiation of appropriate treatment.

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Neurological diseases associated with work absences in a Brazilian public university

David Greco Varela[1,2], Antonio de Souza Andrade Filho[1,2], Frederico Luiz da Silva Figueroa[1,3], Vitoria de Araujo Carneiro Santos[1,4], Adonay Ferreira Lisboa[1,3], Vitor Raphael Heliodoro Barreto[1,3], Natalia Lino Dorea da Silva[1,3], Ana Carolina Campos Carvalho[1,3]

[1]Fundação Baiana de Neurologia e Neurocirurgia, Salvador BA, Brazil.
[2]Universidade Federal da Bahia, Salvador BA, Brazil.
[3]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[4]União Metropolitana de Educação e Cultura, Lauro de Freitas BA, Brazil.

Correspondence: David Greco Varela; Email: dgvarelabr@yahoo.com.br.

Background: Neurological diseases are an important cause of morbidity and serious sequelae in Brazilian workers. Many of them need to remain away from their activities for long periods. Public servants do not yet have a fully structured Occupational Health network. It is necessary to have knowledge about the illness profile of this population to choose the best approach to such diseases, whether they are general or specific and related to professional routine. **Objective:** To verify the frequency of neurological diseases associated with prolonged work absence in Brazilian public university employees between 2019 and 2023. **Methods:** It was a cross-sectional study in which a retrospective survey was carried out, including work absences lasting fifteen days or more associated with neurological diseases in the period from 01/01/2019 to 12/31/2023. The following variables were analyzed: gender, occupation and kind of neurological disease. The population was 5,564 public university employees, with a predominance of women (53.8%). The majority of staff were between 31 and 59 years old (75,8%). **Results:** Neurological diseases accounted for 3.5% of the total of 5,197 absences due to illness over the five years of the study. The frequency in women was 3.5% (132/3844) and 3.7% (51/1353) in men. The year with the highest incidence was 2022, corresponding to 25.1% (46/183) of the total cases and the lowest was 2020, with 13.6% (25/183). Cerebrovascular diseases in women corresponded to 26.5% (35/132) of absences and in men they corresponded to 47.05% (24/51). There was also emphasis on weakness caused by migraines, which affected 20.4% (27/132) of women, and mononeuropathies, which affected 13.7% (7/51) of men. Illness among healthcare professionals was more frequent among women, ranging from 42% in 2019 to 86% in 2020, with nursing assistants being the most affected position. In men, there was a similarity between absences among health, teaching and administration professionals. **Conclusion:** There was a high frequency of absences due to cerebrovascular diseases in men and women. Such diseases have a great potential for morbidity and risk to life. A high frequency of involvement of health professionals was noted, especially among women. Preventive measures and health promotion programs must be adopted to protect and rehabilitate the population of workers identified as more vulnerable to neurological problems.

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Neurological health of refugees in Brazil: results and perspectives

Diogo Haddad Santos[1], Flávia Esper Dahy[1], Maria Aldilene Gonçalves[1], João Eduardo Nunes Salles[1], Fares George Abdelmassih[1]

[1]Associação Médica Líbano-Brasileira, São Paulo SP, Brazil.

Correspondence: Diogo Haddad Santos; Email: dhaddadsantos@gmail.com.

Forced migration is a growing reality worldwide, and Brazil has become an important destination for many refugees. According to the latest official report from the Brazilian government, the country received 58.628 requests for refugee status recognition in 2023, an increase of 16.4% compared to the previous year. This increase reflects the overcoming of the most severe period of the Covid-19 pandemic and the resumption of migratory dynamics. Most of these requests came from Venezuelans, followed by Cubans and Angolans, highlighting the diversity of nationalities among the applicants. Importance of Neurological Evaluation in Refugees The health of migrants is a global concern affecting millions of people. It is known that migrants face a series of health vulnerabilities due to factors such as poverty, malnutrition, poor living conditions, dangerous work, and psychological problems related to the migration process. These vulnerabilities increase the incidence of communicable and non-communicable diseases, including neurological and psychiatric disorders. Studies indicate that refugees have a higher prevalence of neurological diseases compared to the local population. Chronic neurological diseases, such as epilepsy and cerebrovascular diseases, are common in refugee camps, surpassing infectious neurological diseases. American data show that resettled refugees in the United States of America often present psychiatric comorbidities associated with neurological diseases, such as headaches, sleep disorders, and head trauma. The Saha (Health in Arabic) Brazil project is a philanthropic initiative conceived by Prof. Dr. Riad Younes and developed by the Lebanese Brazilian Medical Association (AMLB) board to provide specialized medical care to refugees in our country. Results Over a period of four years, 43 patients with neurological conditions were evaluated. Most are adults, 33/43 (76.75%), the majority are women, 25/43 (58.14%), and the mean and median age at the time of first care are, respectively, 34.21 (SD 16.25) and 38 years. These patients come from thirteen different countries, the majority from Venezuela, 14/43 (32.56%). The main neurological condition was headache, presented in 18/43 patients (41.86%), followed by epilepsy, 6/43 (13.95%), which was most diagnosed in children and adolescents 4/6 (66.67%). Conclusion Neurological evaluation of refugees is crucial to identify and treat conditions that may be neglected due to migration and resettlement circumstances. The prevalence of neurological diseases and their psychiatric comorbidities among refugees highlights the need for specific and coordinated interventions. The preliminary results of this project underline the importance of comprehensive neurological evaluations in refugees, contributing to the improvement of health and quality of life for this population.

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Neurological immune-related adverse events of checkpoint inhibition therapy: insights from a large-scale electronic medical database

Diogo Haddad Santos[1], Fernanda Grôppo Nogueira Herculano[1], Natalia Silva Fernandes[1], Cheng Tzu Yen[1], Ariel Galapo Kann[1], Ana Carolina Gomes[1]

[1]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Diogo Haddad Santos; Email: dhaddadsantos@gmail.com.

Background: Immune checkpoint inhibitors (ICIs) have revolutionized cancer therapy by enhancing the immune system's ability to recognize and attack tumor cells. Despite their clinical benefits, ICIs are associated with a range of immune-related adverse events (irAEs), including neurological toxicities. Our study, with a cohort of 114,317 patients, significantly surpasses all the previous studies, providing a more comprehensive understanding of the neurological adverse effects associated with ICIs. TriNetX is a global federated health research network providing access to electronic medical records, across large healthcare organizations (HCOs). This study leverages data from the Global Collaborative Network, which includes 119 HCOs. **Objective:** To evaluate the incidence, types, and severity of neurological adverse effects in this population. **Methods:** Identify patients treated with ICIs and setting up and running the analysis by defining the index event, outcomes criteria, and the time frame for the analysis. The analysis included four types: Risk, Survival, Number of Instances, and Lab Result Distribution. Inclusion Criteria: Patients must have received any of the following medications: pembrolizumab, atezolizumab, avelumab, cemiplimab, durvalumab, ipilimumab, nivolumab,

dostarlimab, relatlimab, tremelimumab, or retifanlimab. Exclusion Criteria: Patients must not have had diagnoses of the neurological diseases before the use of the ICIs. Outcomes were analyzed from 1 day to 60 days after the index event. **Results:** The analysis revealed the incidence rates of neurological immune-related adverse events (n-irAEs) among the cohort of 114,317 patients treated with immune checkpoint inhibitors (ICIs). The composite outcome of n-irAEs was observed in 3,643 patients, representing a risk of 0.032. Within this group, several specific events were identified. Encephalitis was observed in 114 patients, corresponding to a risk of 0.001. Demyelinating diseases were identified in 91 patients, also corresponding to a risk of 0.001. Meningitis occurred in 161 patients, with a risk of 0.001. Neuromuscular junction disorders were recorded in 327 patients, with a risk of 0.003. Although the specific number of patients and the corresponding risk for polyneuropathies were not detailed here, they are included in the composite outcome of n-irAEs. Similarly, the specific number of patients and the corresponding risk for skin-limited vasculitis were not detailed here, but they are part of the composite outcome of n-irAEs. **Conclusion:** This study, utilizing the largest cohort available in the literature provides a comprehensive analysis of the neurological adverse effects associated with ICIs in oncology patients. The findings underscore the importance of vigilant monitoring and proactive management of these adverse effects to optimize patient outcomes.

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Neurological manifestations in chikungunya fever: a systematic review

Pedro Lucas Grangeiro de Sá Barreto Lima[1], Pedro Vitor Ferreira Rodrigues[1], Leonardo José Rodrigues de Araújo Melo[1], Lucas Soares Radtke[1], Emanuel de Assis Bertulino Martins Gomes[1], Sophia Costa Vasconcelos[1], Pedro Henrique Azevedo Damascena[1], Júlia Viana de Castro[1], Francisco Luciano Honório Barreto Cavalcante[1], Paulo Ribeiro Nóbrega[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Pedro Lucas Grangeiro de Sá Barreto Lima; Email: pedro.lucas@alu.ufc.br.

Background: Chikungunya is an arboviral infection caused by the Chikungunya Virus (CHIKV) and is typically transmitted to humans by infected Aedes spp. mosquitoes. The disease is distributed worldwide, occurring more often in outbreaks in Asia and Americas. Neurologic involvement is highly heterogeneous, with manifestations involving the central and peripheral nervous systems. Information regarding the effective burden of this impairment is still lacking. **Objective:** To summarize the existing data on neurological manifestations of this arbovirus, providing new insights on prevalence and characterization. **Methods:** A systematic review and meta-analysis were conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) reporting guideline. We included studies with patients diagnosed with Chikungunya by acute serology or PCR who developed neurological manifestations during or after the disease course. Articles reporting patients with headache as only neurological manifestation were not included. **Results:** A total of 126 unique articles out of 1941 were included for review. These articles enrolled 2970 patients presenting neurological impairment caused by CHIKV. The overall incidence of neurological manifestations in CHIKV varied from 16.3 to 75% of the patients. A study reported a cumulative incidence rate of 8.6 per 100,000 persons of neurological CHIKV in the general population during an outbreak. Most patients present these manifestations in the first week of the disease. However, a minority of patients may exhibit impairment weeks or even months after the initial infection. The main reported neurological manifestations of CHIKV are encephalitis, myelopathy, peripheral neuropathy, myeloneuropathy, and myopathy. Other conditions, such as isolated cranial neuropathy, chronic neuropathic pain, horner syndrome and hemorrhagic stroke may occur much less frequently. The infection may also act as a trigger for other neurological dysfunctions, being associated with manic episodes in bipolar disorder patients, generalized anxiety disorder and autoimmune encephalitis. Few articles have reported specific neuroimaging features in neurological CHIKV. The "glass eel sign" is a radiological pattern of spinal cord lesion presenting with peripheral hypersignal on T2 images and central sparring. The "boomerang sign" is characterized by hyperintensity of the splenium of corpus callosum also in T2 imaging. Both of these signs are consistently reported in cases of CHIKV infection. **Conclusion:** We systematically reviewed the neurological disorders caused by CHIKV infection, describing the prevalence and characteristics of the main associated manifestations. Larger samples of the most uncommon manifestations are still necessary to provide a proper description of the whole CHIKV spectrum. This knowledge helps to identify future Neurological CHIKV cases and to improve the outcomes of the patients.

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Neurological manifestations in the context of dengue

Ana Vitória Alves de Souza Gripp Laizo[1], Mariana Vaz de Melo Diniz[1], Eustaquio Claret dos Santos[1], Anderson Noboru Mitsui[1]

[1]Hospital Socor, Belo Horizonte MG, Brazil.

Correspondence: Ana Vitória Alves de Souza Gripp Laiz; Email: anavitoriagripp@hotmail.com.

Background: During the second quarter of 2024, there was an increase in the number of dengue cases and associated complications, including involvement of the central nervous system. Diseases reported in the literature include encephalopathy, ischemic and hemorrhagic stroke, polyneuropathies, among others. **Objective:** To analyze neurological manifestations in patients with dengue fever, treated at a private hospital in Belo Horizonte - MG, between February and May 2024, and compare them with the literature. **Methods:** This was a descriptive, retrospective study conducted through the review of medical records from February to May 2024. **Results:** During the study period, 15 cases of neurological manifestations due to dengue were observed. These included 5 cases of encephalitis (33.33%), two cases of refractory migrainous headache (13.33%), one decompensation of a patient with multiple sclerosis (6.67%), two decompensations of patients with epilepsy (13.33%), one hemorrhagic manifestation of dengue with subarachnoid hemorrhage (6.67%), one patient with transient intracranial hypertension with VI nerve palsy (6.67%), one case of ischemic stroke (6.67%), and two cases of transient ataxia (13.33%). The age of patients ranged from 19 to 87 years, with a mean age of 62 years. Male patients predominated (66.67%) over female patients (33.33%). Common comorbidities included systemic arterial hypertension, diabetes, epilepsy, and history of previous ischemic stroke. Most patients fully recovered from their symptoms (86.67%), while a small portion experienced partial resolution (13.33%). There were no fatalities. Regarding the temporal relationship between neurological symptoms and dengue fever, both were concurrent in 11 cases (73.33%), while in 4 cases (26.67%), neurological symptoms followed dengue fever immediately. **Discussion:** A review of the literature reveals that the incidence of neurological manifestations of dengue varies widely, with encephalitis predominating, consistent with findings in this study. An important consideration is the lack of specific treatment directed at the arbovirus itself, particularly in cases of encephalitis, necessitating supportive care focused on complications such as epilepsy, headache, subarachnoid hemorrhage, and ischemic stroke. Ischemic strokes pose a challenge in terms of the risks associated with antiplatelet therapy, particularly in the context of thrombocytopenia. **Conclusion:** Based on the present analysis, there is a clear need for thorough diagnostic investigation, including dengue testing, in patients presenting with acute neurological conditions in the current epidemiological context. Additionally, the importance of neurological monitoring and appropriate ancillary testing for patients already diagnosed with dengue is underscored. Lastly, further studies are needed to explore the treatment of such conditions.

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Neurological manifestations of leprosy: a complex clinical case

Isabelle Pastor Bandeira[1], Catarina Dantas Corrêa[1], Camila Emi Fujiwara Murakami[1], Mariana Marques Wolski[1], Allinson Lidemberg Ribeiro[2], Caio César Diniz Disserol[1], Paulo José Lorenzoni[1], Marcos Christiano Lange[1], Rosana Hermínia Scola[1], Carlos Eduardo Soares Silvado[1]

[1]Universidade Federal do Paraná, Complexo do Hospital de Clínicas, Curitiba PR, Brazil.
[2]Universidade Estadual de Ponta Grossa, Ponta Grossa PR, Brazil.

Correspondence: Isabelle Pastor Bandeira; Email: isabellepbandeira@gmail.com.

Case presentation: O.L., a 63-year-old man, presents a complex medical history marked by progressively worsening neurological symptoms since 2017. His previous medical conditions included hypertension, dyslipidemia, and an ischemic stroke at age 50. In May 2019, the patient began experiencing burning sensations in his hands, noted only after the appearance of blisters, without accompanying pain. This hypoesthetic symptom progressed from distal to proximal areas, now affecting both upper limbs up to the middle third of the arms, accompanied by a sensation of swollen fingers. Subsequently, he reported burning on the sole of his right foot in January 2020, coinciding with initial signs of foot anesthesia. By November 2021, burning sensations became frequent, with worsening anesthesia in both lower limbs extending up to the knees, associated with reports of imbalance but no falls. Upon admission to our service, we encountered an elderly patient with asymmetric reduction of superficial and deep sensitivity, progressively worsening over 6 years. Further investigation revealed length-dependent peripheral neuropathy, predominantly involving small fiber impairment. The final diagnosis was neuropathy due to Multibacillary Leprosy (10% intact bacilli) with Type 1 leprosy reaction. Multidrug therapy treatment commenced promptly on September 5, 2023. **Discussion:** Leprosy, or Hansen's disease, manifests in diverse clinical forms, ranging from mild skin lesions to severe nerve damage and deformities, as illustrated by this clinical case. This chronic infectious disease, caused by *Mycobacterium leprae*, primarily affects the skin and peripheral nerves, leading to progressive hypoesthesia and loss of sensation. Our patient presentation of burns on hands and feet without pain perception highlights the characteristic insensitivity to injury, a hallmark of leprosy neuropathy. Diagnosing leprosy relies on clinical suspicion complemented by dermatological findings and histopathological confirmation. In this case, the diagnosis of multibacillary leprosy with a type 1 reversal reaction was confirmed through clinical evaluation, EMG findings consistent with sensory-motor axonal polyneuropathy and lymph bacilloscopy. Treatment of leprosy revolves around multidrug therapy (MDT), comprising rifampicin, dapsone, and clofazimine, aimed at eliminating *Mycobacterium leprae* and preventing disability. **Final comments:** This clinical case exemplifies the intricate clinical spectrum and diagnostic complexities of leprosy. By understanding the pathophysiology, clinical presentation, and management challenges highlighted in this case, healthcare providers and policymakers can enhance their approach to leprosy control efforts. Continued research, advocacy, and community-based interventions are essential to achieving sustained progress towards eliminating leprosy as a public health threat globally.

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Neurological repercussions in dengue infection: a systematic review

Alice Cristina Souza Cunha[1], Fernanda de Abreu Silva[1], Cristina da Costa Oliveira[1]

[1]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.

Correspondence: Alice Cristina Souza Cunha; Email: alicescunha262@gmail.com.

Background: Dengue is the aborvirus with the greatest epidemiological importance in the Brazilian context today, resulting from infection by the arbovirus of the Flaviviridae family, which is transmitted by female vectors of the genus Aedes spp. Associated symptoms may include severe neurological manifestations. The prevalence and pathophysiology of these manifestations still need to be further clarified. **Objective:** To analyze the profile of neurological repercussions in dengue infection. **Methods:** This is a systematic review of the literature, qualitative and descriptive. The PubMed, SciELO and Lilacs databases were used, using the search terms: ("Dengue" OR "Dengue Virus") AND ("Neurologic Manifestations"), selected according to the Health Sciences Descriptors (DeCS) and Medical Subject Headings (MeSH/PubMed). The research included articles with publication dates between 2014 and 2024, available in Portuguese, English or Spanish. Articles published before 2014 and/or in languages other than those mentioned were excluded. **Results:** After a careful analysis of the thematic adequacy of the articles found, in addition to the comparison between the impact factor and Qualis Capes of the scientific journals in which they were published, we selected 12 articles. We were able to observe that the neurological consequences in dengue infection are more prevalent in serotypes 2 and 3, although they can also occur in serotypes 1 and 4. The pathophysiological mechanism involves disruption of the blood-brain barrier and hematogenous invasion by the pathogen, followed by activation of factors autoimmune diseases that trigger metabolic and bleeding disorders. The main complications detected were: (i) encephalitis; (ii) generalized cerebral edema; (iii) or focal cerebral edema. In addition to these, other less common neurological problems associated with dengue were also mentioned, namely: (i) Guillain-Barré Syndrome; (ii) Miller-Fisher syndrome; (iii) Acute Disseminated Encephalomyelitis; (iv) Neuromyelitis Optica; (v) and Cerebellitis. We also identified conditions considered rare, such as: (i) hypokalemic paralysis, a rarely described record, where there is potassium equal to or less than 3mmol/L, with acute onset of flaccid quadriplegia without cranial nerve palsy; (ii) and parkinsonism, an extremely rare manifestation of dengue, with only one case recorded in the literature, accompanied by brachial plexopathy, multiple cranial nerve palsies and ataxia. **Conclusion:** Dengue is a viral infection that can present a complicated evolution, including neural manifestations, with encephalitis and generalized or focal cerebral edema considered the main neurological repercussions. Determining the epidemiological order of these complications is extremely important for early diagnosis and consequent appropriate therapeutic management. Improving pathophysiological knowledge of these possible complications is still necessary.

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Neurological repercussions of acute intermittent porphyria and its importance as a differential diagnosis of Guillain Barré syndrome: a case report

Júlio César Soares Barros[1], Antonia Cardoso Silva[1], Guilherme Carvalho de Souza[1], Larissa da Silva Almeida[1], Maria Adélia de Albuquerque Barros[1], Marcelo Augusto Vieira Jatoba[1], Maria Erigleide Bezerra da Silva[1], Isabelle Galvão de Oliveira[1], José Maria Gonçalves Fernandes[1], Fernando Tenorio Gameleira[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.

Correspondence: Júlio César Soares Barros; Email: julio.barros@famed.ufal.br.

Case presentation: A 28-year-old woman presented with fever, nausea, vomiting, and right iliac fossa pain for one month. She was referred to the surgical emergency service, where she underwent an appendectomy due to suspected acute appendicitis. One week later, she developed nausea, abdominal pain, occipital headache, and progressive ascending paresis in the lower limbs (LL), with significant difficulty in walking. She progressed to respiratory failure and was admitted to the ICU with suspected Guillain-Barré Syndrome (GBS) and Acute Intermittent Porphyria (AIP). Neurological examination revealed isocoric and photoreactive pupils, paraparesis (muscle strength grade I in LL), muscle strength grade III in upper limbs, moderate hypopallesthesia in LL, and preserved superficial sensitivities. Plantar and patellar reflexes were present, while other reflexes were absent. Cerebrospinal fluid analysis ruled out GBS hypothesis (no albumin-cytological dissociation), and darkening of urine after sun exposure strengthened AIP possibility. A 24-hour urine test confirmed the diagnosis with positive delta-aminolevulinic acid and porphobilinogen. Initial treatment included a carbohydrate-rich diet, but hematin was not used due to supply issues. The drug was later introduced into therapy, but the patient was already in a critical state, evolving with tetraplegia, dysautonomia (inappropriate sinus tachycardia), worsening respiratory failure, persistent seizures, and death. **Discussion:** AIP is a rare autosomal dominant disease characterized by an enzyme deficiency in heme biosynthesis. This condition leads to the accumulation of harmful precursors causing severe neurovisceral and neurological symptoms. The porphyric crises can mimic acute abdomen and neurological disorders such as GBS. In this case, cerebrospinal fluid analysis helped rule out GBS. Urine darkening after sun exposure supported the AIP hypothesis, as exposure to sunlight and acidic urine pH can produce porphyrins and other pigments, making the urine appear redder. Confirmation came with the presence of porphyrin precursors in the 24-hour urine test. The initial treatment with a carbohydrate-rich diet and intensive care measures did not yield a satisfactory clinical response, necessitating the introduction of hematin in therapeutic management. This medication is essential as it suppresses the production of porphyrin precursors. The difficulty in obtaining hematin and its late introduction contributed to the unfavorable outcome in this case. **Final comments:** The central and peripheral nervous systems can be affected in AIP, leading to peripheral neuropathies, seizures, dysautonomia and neuropsychiatric manifestations. Symptoms in common with GBS, such as progressive muscle weakness and respiratory failure, highlight the importance of including AIP in your differential diagnosis in order to ensure early and adequate treatment.

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Neurological symptoms in a renal transplant patient: a case report

Gabriela Rodrigues Tomaz[1], Fernanda Lustosa Cabral Gomez[1], Lara Santos Machado[1], Luiz José Malta Gaia Ferreira Júnior[1], Larissa Braga Mendes[1], Fabiano Moulin de Moraes

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Gabriela Rodrigues Tomaz; Email: gabirodriguestomaz@gmail.com.

Case presentation: A 55-year-old male patient was admitted to the emergency room with a three-week history of apathy, difficulty in finding words, and performing daily activities. He had been on leave from work for two weeks due to worsening symptoms. He did not report falls, imbalance, or gait disturbances, remaining independent in basic daily activities. He denied fever, weight loss, and night sweats. The patient had a history of systemic arterial hypertension and diabetes mellitus and had undergone a kidney transplant from a living donor 24 years ago due to chronic kidney disease of unknown cause. He was taking Tacrolimus, Sodium Mycophenolate, Prednisone, Acetylsalicylic Acid, Atorvastatin, Sulfamethoxazole-trimethoprim, oral antidiabetics, and insulin. In the initial neurological examination, the patient was alert but inattentive, with temporal and spatial disorientation, bradyphrenia, ideomotor apraxia, and inexhaustible glabellar reflex. A cranial CT scan showed a lesion in the left frontal lobe with vasogenic edema. Cerebrospinal fluid examination revealed lymphomononuclear pleocytosis, high glucose, elevated lactate, and hyperproteinorachia. Treatment with Ceftriaxone and Metronidazole was started for a possible brain abscess. New CSF tests confirmed positive cryptococcal antigen and positive India ink, indicating cryptococcal infection. Due to suspected cryptococcoma, treatment with Amphotericin B and Fluconazole was initiated. Later, a blood culture revealed Actinomycete, and the treatment was changed to Cefepime and Clindamycin. The patient developed myoclonus, loss of speech ability, and a decrease in consciousness level, requiring intubation and being diagnosed with non-convulsive focal status epilepticus in the left frontotemporal region. After controlling the epileptic state and gradually tapering off sedation, the patient still did not wake up adequately. A repeated CT scan showed no changes from the initial image. With no clinical improvement after three weeks of treatment, a brain biopsy was performed, revealing diffuse large B-cell lymphoma, non-germinal center type, characterizing a post-transplant lymphoproliferative disorder (PTLD). **Discussion:** All lymphomas occurring in patients receiving post-transplant immunosuppression are considered PTLD, with an incidence of 1-20% in transplant recipients. Epstein-Barr virus positivity in the tumor is common (50-80%). Unlike classic lymphoma, extra-nodal involvement is much more frequent. Treatment involves reducing immunosuppression, rituximab, chemotherapy, and other drugs. **Final comments:** In transplanted patients with new neurological symptoms or mental status changes, consider primary CNS PTLD, which occurs in 10% of cases.

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Neurology activities in an academic league: an integral tool in medical formation

Luan Nascimento Pereira de Amorim[1], Rafael Hendrick Freitas de Paula[1], Luiz Carlos Figueiredo Filho[1], Francinaldo Lobato Gomes[2], Henrique da Costa Miranda[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil. [2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Luan Nascimento Pereira de Amorim; Email: luanamorim010@gmail.com.

Background: Academic leagues are crucial for enriching medical formation, promoting integration between students and experienced professionals. These student organizations play a fundamental role in complementing the formal curriculum, offering additional opportunities for learning, research, and extension. In this sense, the Academic League of Neurology and Neurosurgery of the State of Pará, founded in 2009, stands out for providing an environment conducive to theoretical and practical study in the area of neurology, which is essential for understanding the pathophysiological bases of neurological diseases and their clinical manifestations. Additionally, it not only enriches students' academic knowledge but also prepares them to face future professional challenges, contributing to the formation of more complete and qualified doctors. **Objective:** To report on the experience of this

academic league as an instrument for the integral learning of medical students and for the deepening of neurological knowledge in undergraduate courses. **Methods:** This is an experience report describing the practical and theoretical activities offered by an Academic League of Neurology and Neurosurgery in the State of Pará. **Results:** The Academic League of Neurology and Neurosurgery of Pará carries out theoretical and practical activities that enrich the training of students. The lectures, given by renowned specialists, cover everything from the development of neurological problems to diagnoses and treatments, providing a comprehensive and up-to-date understanding. Practical activities include weekly internships at a hospital in Belém, where students develop skills in identifying alterations in tests and understanding neurological problems. In addition, the league promotes outreach activities for the public, addressing topics such as headaches, neurodegenerative diseases and epilepsy. Scientific production is encouraged, with the preparation of articles, case reports and presentations at congresses, guided by experienced professionals. To consolidate knowledge, the league organizes courses and symposia on various neurological topics. These activities provide a comprehensive overview of the various cases that a future doctor may encounter in their clinical practice. **Conclusion:** The Academic League of Neurology and Neurosurgery of the State of Pará plays an essential role in preparing students for the medical profession. By offering a balanced combination of theoretical and practical activities, it promotes a significant deepening of knowledge and its application in real scenarios. It also contributes to scientific production and fosters valuable interaction between students and experienced professionals, providing comprehensive academic and professional development and preparing future doctors to face the challenges of neurological clinical practice with competence and confidence.

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Neurology first: a challenging case of early rheumatoid meningitis in a previously undiagnosed rheumatoid arthritis

Giovanna Reis Coelho[1], César Minoru Toita Koga[2], Guilherme Faria Cabrera[1], Camila Carneiro Ferreira[3], Catarina Dantas Corrêa[2], Caio César Diniz Disserol[2,3,4]

[1]Universidade Positivo, Curitiba PR, Brazil.
[2]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[3]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[4]Hospital Israelita Albert Einstein, Curitiba PR, Brazil.

Correspondence: César Minoru Toita Koga; Email: cesarmtkoga@gmail.com.

Case presentation: A 75-year-old man with a history of smoking and treated pulmonary tuberculosis presented to the emergency department with transient aphasia followed by headache and meningeal signs. He was admitted for investigation and, in the next day, experienced his first tonic-clonic seizure. Brain magnetic resonance imaging (MRI) showed bilateral frontoparietal pachymeningeal enhancement, and cerebral spinal fluid (CSF) analysis revealed high protein levels (57 mg/dL) and neutrophilic pleocytosis (30 cells/mm3; 83% neutrophils). Treatment with levetiracetam, ceftriaxone and acyclovir was started. One hour later, the patient developed fever, rash, tachycardia, and dyspnea, which resolved with anti-inflammatory medication and prednisone. Jarisch-Herxheimer reaction and neurosyphilis were suspected. Serum investigation revealed reactive serum treponemal test (IgG FTA-ABS), negative non-treponemal test (VDRL), and positive rheumatoid factor (56 UI/mL), with no history of prior syphilis treatment or joint symptoms. CSF viral panel was negative, and treatment was switched to 14 days of crystalline penicillin. Other infectious and rheumatological tests were negative, including serum IgG4 and interferon-gamma release assay. Later CSF tests for syphilis (IgM and IgG FTA-ABS and PCR) were negative, excluding neurosyphilis diagnosis. Follow-up brain MRI 2 months later was stable, but the patient continued to experience residual headache and developed symmetric peripheral polyarthritis and morning stiffness. Analysis of stored serum collected at admission revealed very high levels of anticyclic citrullinated peptide antibodies (anti-CCP: 500 U/mL; RR: < 17 U/mL), leading to the diagnosis of rheumatoid arthritis (RA). Treatment with corticosteroids and methotrexate resulted in complete resolution of the headache, no further seizures, and normalization of neuroimaging and CSF findings. **Discussion:** RA is a systemic inflammatory disease that can affect extra-articular regions. Among its rare manifestations is rheumatoid meningitis (RM), a challenging diagnosis that can present as focal neurological deficits, headache, neuropsychiatric symptoms, and seizures. Some patients, particularly males, may experience meningitis during the early stages of RA or as its initial manifestation, like in our case report. Diagnosis relies on laboratory and imaging results, including lymphocytic pleocytosis in CSF analysis and frontoparietal pachymeningeal enhancement on MRI. Biopsy may reveal rheumatoid nodules, lymphocytic infiltrate, or vasculitis. Treatment typically involves corticosteroids and immunosuppressive drugs, leading to a more favorable prognosis when started promptly. **Final comments:** Early RM diagnosis is challenging due to its rarity and nonspecific presentation, especially in patients with undiagnosed RA. Clinicians should consider RM in cases of meningoencephalitis with typical MRI findings to enhance patient outcomes and prevent complications.

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Neuromuscular ultrasound in non-5q spinal muscular atrophies

Pedro Henrique Marte de Arruda Sampaio[1], Rodrigo de Holanda Mendonca[1], Ana Lucila Moreira[1], Cristiane Araújo Martins Moreno[1], André Macedo Serafim da Silva[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Pedro Henrique Marte de Arruda Sampaio; Email: phsampaio@gmail.com.

Background: The non-5q spinal muscular atrophies (SMA) are a clinically and genetically heterogeneous group of hereditary disorders that primarily affect the lower motor neuron, leading to progressive muscle weakness and atrophy. Neuromuscular ultrasound is useful in the differential diagnosis and evaluation of neuromuscular diseases, and different patterns of muscle involvement can point to specific genes. **Objective:** Using a muscle ultrasound protocol, we evaluated patients with proximal and distal forms of non-5q SMA, along with genetic and clinical findings. **Methods:** We selected 21 patients from 19 families with clinical, electrophysiological and genetic findings consistent with non-5q SMA, classified according to the pattern of weakness (predominantly proximal or distal). We developed a neuromuscular ultrasound protocol that evaluated proximal and distal upper and lower limb muscles, axial muscles and diaphragm. Muscle echogenicity was evaluated by the Heckmatt scale, and muscles were observed for 30 seconds for the detection of fasciculations. Diaphragm thickness was measured at deep inspiration and at relaxed expiration, and diaphragm paralysis was defined as the ratio of diaphragm thickening less or equal to 1.2. We composed a heatmap with the muscle ultrasound findings, using the Heckmatt scale score for each muscle. **Results:** Patient's age ranged from 1 to 60 years old (median 33 years old), 15 were male and 6 were female, and the age of onset was congenital in 8 cases. Clinically, 12 patients were classified as proximal (Group 1), and 9 patients were classified as distal (Group 2). Variants were found in 11 patients, in the TRPV4, BICD2, DYNC1H1, VWA1, HINT1 and C19orf12 genes, whereas in 10 cases the genetics studies were inconclusive. The heatmap hierarchical clustering defined 3 patterns of muscle involvement that significantly corresponded (p=0.02) with clinical Group. The first (Pattern A), a diffuse pattern affecting proximal and distal limb muscles, muscles of scapular girdle and axial muscles, was observed in 9 patients, 8 of which were from Group 1 (proximal). The second (Pattern B), a pattern involving predominantly the lower limbs and hand muscles, but also deltoid, latissimus dorsi and rectus abdominis muscles, was observed in 6 patients, 5 of which were from Group 2 (distal). The third pattern (Pattern C) was a milder phenotype, with selective muscle involvement, notably distal lower limb muscles, and was equally represented in Groups 1 and 2. Muscle fasciculations were observed in 12 patients, and there was no statistically significant difference in the number of muscles affected and Group. Only one patient had unilateral diaphragm paralysis. **Conclusion:** The neuromuscular ultrasound protocol was useful in defining the phenotype and differentiating the proximal and distal forms of non-5q SMA. The findings also suggest that proximal and axial muscle involvement in distal SMA may be more common than initially thought.

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Neuromyelitis optica preventive treatment profile at a universitary hospital in Vitória, ES

Victória Pagani Samora Sousa[1], Daniel Mariani Favalessa[1], Igor Flavio Gonçalves Do Vale[1], Mackeyle Siqueira de Souza[1], Sávio Cesar Rodrigues Pestana[1], Vinícius Andreata Brandão[1], Paula Zago Melo Dias[1], Bruno Batitucci Castrillo[1], Carlos Alberto Magirius Peixoto[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Igor Flavio Gonçalves do Vale; Email: igor.f.vale@edu.ufes.br.

Background: Neuromyelitis optica (NMO) is a severe autoimmune disease of the central nervous system, characterized by inflammation of the optic nerves and spinal cord. Until recently, there was no treatment with randomized and controlled studies for the disease, and treatment was based on prescribing immunosuppressants based on retrospective studies. **Objective:** To conduct a cross-sectional evaluation of the maintenance treatment offered to patients and the rate of therapeutic failure. **Methods:** A retrospective study of patients diagnosed with NMO with positive anti-aquaporin 4 (AQP4) antibodies at HUCAM was conducted. Treatment refractoriness was defined as the need to discontinue or change treatment due to inefficacy or adverse effects. **Results:** In total, 37 AQP4+ patients were evaluated. Of these, 21 patients (56.76%) used azathioprine as the first choice of treatment. Among them, 9 patients (42.86%) experienced therapeutic failure, resulting in changes in the treatment regimen: 1 patient (4.76%) to mycophenolate; 6 (28.57%) to rituximab. Additionally, 11 patients (29.73%) of the total started treatment with rituximab, and only 1 (2.70%) began with mycophenolate. **Conclusion:** Azathioprine is an accessible therapeutic option for many NMO patients, but the observed therapeutic failure rate of 42.86%, close to that found in the literature, indicates the need for more effective therapeutic alternatives, such as rituximab or those recently approved for the treatment of NMO. Additional studies are needed to identify predictive factors of treatment response and to optimize NMO management.

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Neuromyelitis optica spectrum disorder (NMOSD) with negative aquaporin 4-igg in a patient with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP)

Lucas Toshio Uenishi[1], Daniel Delgado Seneor[1], Fernanda Lustosa Cabral Gomes[1], Gabriela Rodrigues Tomaz[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Leonardo Villaverde Buback Ferreira[1], Luiz José Malta Gaia Ferreira Júnior[1], Carolina Candeias da Silva[1], Henrique Ballalai Ferraz[1], Enedina Maria Lobato de Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Lucas Toshio Uenishi; Email:

Case presentation: A 45-year-old woman presented for evaluation with a 5-day history of progressive bilateral visual loss and orbital pain associated with numbness, tingling, and weakness of lower limbs, as well as urinary retention. Two years before, she was diagnosed with CIDP, considering a history of an insidious symmetric weakness with reduced tendon reflexes and sensory involvement of the four limbs, endorsed by characteristic findings in nerve conduction studies. She had been clinically stable after steroid courses. On examination, she could count fingers at two meters with the right eye, whereas her left eye had only light perception and a relative afferent pupillary defect. She had flaccid paraplegia with abolished reflexes and extensor plantar responses, as well as loss of touch, pain, and vibratory sensation below the level of T4. MRI was noticeable for bilateral optic neuritis involving the optic chiasm and a longitudinally extensive spinal cord lesion involving almost all segments except for the conus medullaris, with no evidence of nerve root involvement. Laboratory exams were normal, including serological, arboviral, and rheumatological tests. A lumbar puncture showed a CSF with 55 cells/mm3 (lymphocytic predominance but with 15% of neutrophils and 4% of eosinophils) and 98 mg/dL of protein, without other biochemical or microbiological findings (including herpesvirus and HTLV tests). Serum aquaporin 4-IgG and anti-myelin oligodendrocyte glycoprotein (MOG) were negative. She received a high-dose course of steroids and intravenous immunoglobulin. She was kept with a maintenance dose of prednisone, with good improvement of visual acuity but little of other symptoms. **Discussion:** To date, we have found only two cases in the literature of CIDP concomitantly with NMOSD. There was evidence of simultaneous central and peripheral inflammatory activity in both, whereas in our case, CIDP preceded optic neuritis and myelitis. Diagnosing NMOSD with a negative anti-AQP4 test requires the presence of at least two clinical syndromes (optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, acute diencephalic syndrome/symptomatic narcolepsy, or typical symptomatic cerebral syndrome), with at least one of the first three syndromes being present. Besides, there are additional MRI requirements, such as longitudinally extensive lesions on the spinal cord or optic nerve, and the need to exclude alternative diagnoses. In view of that, our patient fulfilled the criteria for NMOSD. **Final comments:** CIDP and NMOSD are inflammatory demyelinating diseases; the former is usually restricted to peripheral nerves and the latter to the central nervous system. However, no previous studies have determined a common underlying pathology to explain both presentations concomitantly. Despite the few cases reported so far, we should point out a possible autoimmune overlap of these two syndromes.

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Neuromyelitis optica spectrum disorder AQP4-AB positive and myasthenia gravis: a rare association of immune-mediated diseases

Thais Lavinas Moura Baptista Barroso[1], Marcella Turon Baran[1], Pedro Henrique da Silva Martins Cardoso[1], Marina Papais Alvarenga[1,2], Marcos Papais Alvarenga[2], Cleonice Alves de Melo Bento[3] , Regina Maria Papais Alvarenga[1,2]

[1]Universidade de Vassouras, Vassouras RJ, Brazil. [2]Hospital Federal da Lagoa, Rio de Janeiro RJ, Brazil. [2]Universidade Federal do Estado do Rio Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Regina Maria Papais Alvarenga; Email: reginaunirio2016@gmail.com.

Case presentation: A 43-year-old Caucasian woman presented in March 2021 with severe cervical pain and paresthesias affecting her entire body following a COVID-19 infection in February 2021. A neurologist suspected Transverse Myelitis and empirically treated her with oral prednisone. A magnetic resonance imaging (MRI) scan on March 15, 2021, revealed an elongated area of high signal intensity in the spinal cord at the C2 level on the right, and a focus in the left postcentral gyrus, both with contrast enhancement. Neurological examination confirmed altered superficial sensation. In her medical history, the patient reported episodes of eyelid ptosis and fluctuating muscle weakness since age 19 (1997), exacerbated during pregnancy, which remained undiagnosed despite consulting several neurologists. In 2001, after giving birth, her child had neonatal complications but recovered without treatment. Following discharge, she experienced arm weakness and sought another neurologist who suspected Myasthenia Gravis (MG), confirmed by anti-acetylcholine receptor antibody testing. She responded to oral anticholinesterase medication and thymectomy, after which the disease went into remission. Investigation for Transverse Myelitis and MG: Cerebrospinal fluid analysis and blood tests were normal; anti-acetylcholine receptor and anti-MUSK antibodies were negative; anti-aquaporin 4 antibody was positive, and anti-MOG was negative. Treatment with methylprednisolone improved her pain, with residual mild distal hypoesthesia (EDSS = 2.0). Prophylactic treatment with Rituximab was chosen and has been effective and safe. **Discussion:** This represents a rare case of AQP4-positive Transverse Myelitis currently classified as Neuromyelitis Optica Spectrum Disorder (NMOSD), despite not having extensive spinal cord involvement, associated with Myasthenia Gravis occurring 23 years earlier and stabilized post-thymectomy. This case shares similarities with findings in a multicenter study of 16 cases of MG-NMOSD overlap, where most initially presented with MG, and the interval to NMOSD onset varied (1 to 41 years). Thymic hyperplasia was frequent, with 70% undergoing surgery before NMOSD onset, resulting in MG remission for most. Genetic susceptibility studies indicate the DRB1*03:01 allele, associated with NMOSD, is also linked to other autoimmune diseases like Systemic Lupus Erythematosus and Sjogren's syndrome. While MG genetic susceptibility is heterogeneous, recent research suggests early-onset MG may be associated with the DRB1*03:01 allele, potentially linking these rare autoimmune conditions. **Final comments:** We describe the sole AQP4-positive NMOSD spectrum syndrome associated with MG in our cohort of 300 NMOSD cases in Rio de Janeiro.

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Neuromyelitis optica spectrum disorder with atypical presentation and evolution

Lucas Vergne Cheade Lins[1], Daniel Lordelo San Martin[1], Julia Cordeiro Braga[1], Adriana Alves Rapp[1], Brenda Francisca Cardoso Brito[1], Victor Xavier da Silva[1], Marcel Leal Ribeiro[1], Karla Oliveira Couto[1], Rubson Soares Rocha[1], Renan Carvalho Castello Branco[1]

[1]Hospital Santa Izabel, Salvador BA, Brazil.

Correspondence: Lucas Vergne Cheade Lins; Email: lucascheade@gmail.com.

Case presentation: Female patient, 45 years-old, white, previous hypothyroidism, reports itching for 45 days in the dorsal region with progression to allodynia up to the anterior thoracic region associated with burning. In a medical evaluation at the time was prescribed oral acyclovir. After a few days, sensory symptoms persisted and the patient developed reduced strength in the left lower limb, numbness in feet and urinary retention. There was clinical worsening requiring hospitalization. On examination, it was noted superficial hypoesthesia at T6 level and grade 4- strength in lower limbs, bilateral support for walking. CSF analysis: 320 cells/mm^3 (82% lymph), 50mg/dL protein and 61g/dL glucose. Magnetic Resonance of the brain and spine showed an inflammatory lesion with T2-T8 enhancing contrast. The initial possibility of infectious myelitis and demyelinating disease was raised, intravenous acyclovir and corticosteroids were started on 2º and 3º day of hospitalization, respectively. VDRL, hepatits viral, HIV serologies, vasculitis tests, and CSF viral panel (PCR for HSV 1/2, VZV, CMV) were negative. Vitamin B12 normal. Anti-AQP4 antibody: 1:10. Diagnosis of Neuromyelitis Optica Spectrum Disorder (NMOSD) was confirmed and it was decided to prescribe plasmapheresis, given little clinical improvement with corticosteroids. At the end of treatment, the patient developed significant improvements in urinary retention, gait and strength and sensory deficits **Discussion:** NMOSD is an inflammatory astrocytopathy of the CNS, common in women (10:1) with a mean age of 36 years. Its clinical course usually occurs lasting up to 30 days, typically with involvement of the optic nerve, extensive myelitis and area postrema syndrome. It's rare for the condition to progress for more than 4 weeks, 1-2% is considered secondary progression and only 1 case has been reported as chronic progressive course from onset. Cellularity in CSF may be > 50 cells/mm^3 in 13-35% and > 100 cells/mm^3 in 6% of cases. Acute treatment must be carried out immediately in order to reduce inflammatory activity. The options are: methylprednisolone, plasmapheresis or Human Immunoglobulin (IVIG). Complete recovery rates with the use of corticosteroids alone are low, studies indicate a percentage of 17%, often requiring associated plasmapheresis or IVIG. **Final comments:** Progressive cases are challenging in NMOSD, with difficult differentiation from other etiologies. The patient presented a prolonged evolution and CSF with significant pleocytosis, leading to the consideration of infectious etiology. However, these levels of pleocytosis and evolution of the clinical picture beyond four weeks may occur in a small proportion of NMOSD. The diagnostic exclusion or treatment of infectious agents favors earlier immunotherapy treatment, aiming for better functional recovery. The suspicion of NMOSD, even in atypical situations, is essential for the institution of early treatment at the first clinical manifestations

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Neuromyelitis optica spectrum disorders (NMOSD) with systemic lupus erythematosus and thrombotic thrombocytopenic purpura: an overlapping demyelinating syndrome with systemic disorders in a young male patient uncommon case report

Lucas Wilson Matos Gomes[1], Victor Hugo de Souza Silva Gomes[1], Gabriel Carvalho Alves[1], Gabriel Santos de Castro[1], Danyelle Rute Francisco e Silva[1], Letícia Leão Alvarenga[1], Luany Patrícia Liberato de Oliveira[1], Guilhermes Henrique Cavalcante[1], Denise Sisterolli Diniz[1], Delson José da Silva[1]

[1]Universidade Federal de Goiás, Goiânia GO, Brazil.

Correspondence: Lucas Wilson Matos Gomes; Email: lucaswilson95@hotmail.com.

Case presentation: A 19-year-old male, mixed race, previously healthy, began experiencing left-sided paresthesias accompanied by low-grade nocturnal fever, stiffness, and pelvic girdle arthralgia without signs of arthritis and worsening with movement. Within a month, he developed bilateral amaurosis, initially in the left eye (LE) progressing to the right eye (RE), accompanied by

intractable vomiting, epistaxis, and diffuse petechiae. He was hospitalized for investigation, evolving with symmetrical tetraparesis, a single seizure, and decreased consciousness. Complementary exams revealed pancytopenia, and neuroaxis magnetic resonance imaging (MRI) showed findings suggestive of demyelinating lesions with extensive bilateral optic neuritis up to the optic chiasm, rhombencephalitis, and extensive cervico-dorsal myelitis, with a probable diagnosis of neuromyelitis optica spectrum disorder (NMOSD), confirmed by the presence of anti-aquaporin IV antibody - AQP4-IgG - (1:40 in serum by CBA method). Additionally, he had a high titer of ADAMTS-13 in serum, ANA 1:640 multiple nuclear dot and centromeric pattern, and positive anti-SM, anti-RNP, and anti-RO. Therefore, he has a confirmed diagnosis of thrombotic thrombocytopenic purpura (TTP) secondary to probable Systemic Lupus Erythematosus (SLE) according to the 2019 EULAR/ACR criteria. He was started on pulse therapy with methylprednisolone 1g/day for 7 days, 21 sessions of plasmapheresis, and one cycle of cyclophosphamide 1g, with partial improvement. Rituximab (RTX) was chosen, with an attack dose of 500mg/week for 4 weeks, leading to stabilization and partial recovery of deficits. However, 3 months after the RTX cycle, he experienced a new decline in visual acuity, confirmed as a clinical relapse. New pulse therapy was administered, and a new high-efficacy and on-label disease-modifying therapy for AQP4-IgG-positive NMOSD with Ravulizumab, an anti-complement C5, was chosen, reducing the risk of new relapses by an estimated 98%. **Discussion:** Although rare, the overlapping of NMOSD with other autoimmune diseases, such as SLE and TTP, is relatively common, with an estimated association of 7%. Serum autoantibody positivity can be found in 38 to 45% of NMOSD patients, regardless of AQP4-IgG positivity. Chronic inflammation and immunological activation inherent to SLE can compromise the blood-brain barrier (BBB), facilitating the infiltration of AQP4-IgG into the CNS, precipitating complement activation with membrane attack complex formation and thus demyelinating lesions. Endothelial disturbances observed in TTP can exacerbate BBB permeability, corroborating the pathogenesis of NMOSD. **Final comments:** Currently, Ravulizumab is a new high-efficacy disease-modifying treatment option in cases of overlap between SLE and NMOSD due to the shared complement activation in their pathophysiologies, with this patient being one of the first in Brazil to receive this medication through the public health system (SUS).

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Neuromyelitis optica with ANTI-AQP4 antibodies associated with dermatomyositis: a case report

Luís Fernando Fernandes Miranda[1], José Guilherme Pinhatti Carrasco[1], Rodrigo Pernomian Cianca[1], Antonio Paulo Felci Rodrigues Leite[1], Matheus da Silva de Oliveira[1], Felipe Merchan Ferraz Grizzo[1], Juliana Yuri Sekiyama[1], Paulo Roberto Donadio[1], Guilherme Henrique Silva Fogaça[1], Gabriel Bortoli Ramos[1]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.

Correspondence: Luís Fernando Fernandes Miranda; Email: ra123503@uem.br.

Case presentation: A 20-year-old female,diagnosed with dermatomyositis (DM) and continuously using prednisone and hydroxychloroquine, required four pulse therapies with cyclophosphamide due to nonspecific interstitial pneumonia related to DM. After that, she sought medical care with fever (39°C), asthenia and severe ventilatory-dependent pain in the posterior cervical region. She developed urinary retention and tetraparesis and, on physical examination, had a sensory level at T2. Complementary tests were performed, revealing an erythrocyte sedimentation rate of 100mm, CRP <0.5, creatine kinase of 95 and cerebrospinal fluid analysis with 453 leukocytes/mm^3, hyperproteinorrhachia (718mg/dL), hypoglycorrhachia (23mg/dL) and lactate at 42.7mg/dL (reference <19.8). Bacterioscopy, Cryptococcus and VDRL were negative. Ceftriaxone 2g was administered every 12 hours. The patient was admitted to an intensive care unit due to hypoxemia. The patient's level of consciousness improved, however there was no improvement in the motor and sensory neurological deficit. Therefore, cefepime and vancomycin were started and a neuroaxis magnetic resonance imaging (MRI) was requested with cervical (C2-C7) and thoracic (T7-T11) myelitis was evident, affecting nearly the entire transverse diameter of the spinal cord with gadolinium enhancement. As a result, the patient received pulse therapy with methylprednisolone for seven days and was referred for plasmapheresis, in addition to the collection of Anti-aquaporin-4 (AQP4) and anti-MOG antibodies. The patient showed slight clinical improvement, gaining only one degree of strength in the upper limbs. It was decided to maintain 80mg/day of prednisone. The anti-AQP4 (CBA method) was reactive at a dilution of 1:40, confirming the diagnosis of neuromyelitis optica and the patient is currently awaiting rituximab administration. **Discussion:** NMO is an autoimmune disease that attacks astrocytes. In over 80% of cases, it is caused by IgG autoantibodies anti-AQP4, the most abundant water channel in the central nervous system, predominantly located at the astrocyte end-feet.It should be noted that 10-40% of patients with NMO have associated autoimmune diseases, with the most common being systemic lupus erythematosus, Sjogren's syndrome and thyroiditis. Cases of association between NMO and DM are rare,with only one case report in 2014 and the cause of this association is still unknown. Both diseases are autoimmune mediated by B cells, which suggests a possible role for specific biological therapy, such as the Rituximab. **Final comments:** The possibility of an association between NMO and rheumatological diseases should be considered in patients with neurological symptoms and previous rheumatological conditions. Complementary tests, such as MRI and anti-AQP4 antibodies, can help establish the diagnosis. The collaboration of neurologists and rheumatologists is essential for proper management of patients, optimizing the choice of immunosuppressive therapy.

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Neuronal intranuclear inclusion disease: a case report

Amanda Miranda de Mendonça[1], Matheus Alves da Silva[1], Ana Luisa Vieira de Faria[1], Eduardo Mesquita de Souza[1], Rafael Bragança Rodrigues Matias[1], Alex Araujo Fernandes[1], Matheus Luís Braga Munareto[1], Alex Machado Baeta[1], Carmen Lúcia Penteado Lancelotti[2] , Victor Hugo Rocha Marussi[1]

[1]Hospital Beneficência Portuguesa, São Paulo SP, Brazil.
[2]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.

Correspondence: Amanda Miranda de Mendonça; Email: amanda.miranda.dv@gmail.com.

Case presentation: A 63 years-old female patient presented a six month history of dementia, with prominent memory, visuospatial deficits and executive dysfunction. She was born in Brazil, but her family was from Japan. Four years before the onset of cognitive impairment, she presented with episodes of urinary retention of unexplained etiology. Cerebrospinal fluid analysis revealed a normal initial opening pressure, 2 cells, slightly elevated proteins (48mg/dL) and 57mg/dL of glucose. Her screening for infectious and autoimmune diseases was negative. During the investigation, she developed other episodic neurologic dysfunction, characterized by fever and acute confusional state after vaccination, with complementary investigation unremarkable. The same episode happened a year ago. On her brain MRI, the diffusion sequence revealed hyperintensities in the subcortical white matter of the cerebral hemispheres, predominating in the frontal lobes, without alteration in the ADC map. It was accompanied by signal changes on T2/Flair in both cerebellar hemispheres. These findings were suggestive of neuronal intranuclear inclusion disease (NIID). Suspecting it, a skin biopsy showed eosinophilic hyaline intranuclear inclusions on hematoxylin and eosin, compatible with NIID diagnosis. **Discussion:** NIID is a rare disease, with intranuclear eosinophilic inclusion bodies are the main characteristic pathological changes in the central and peripheral nervous systems and are caused by repeated GGC expansion in the NOTCH2NLC gene, especially in Asiatic patients. The most common feature of NIID is symptom heterogeneity. The clinical phenotypes are broad and include cognitive impairment, movement disorder, episodic events, autonomic dysfunction and neuromuscular disease. The most common in adults is the cognitive impairment subtype. The existence of intranuclear inclusions and symptoms in systemic organs may precede the onset of cognitive symptoms. The median onset of urinary symptoms is 55 years. Hyperintensity in the corticomedullary junction on diffusion-weighted imaging (DWI), focal cortical edema/enhancement, and white matter lesions involving the paravermis, middle cerebellar peduncle, and corpus callosum have been reported to be characteristic imaging for NIID. Currently, there is no treatment to cure or slow down the process of NIID, and the treatment is supportive for improving quality of life. **Final comments:** The high heterogeneity of clinical symptoms and signs of NIID increases the difficulty in the clinical diagnosis of NIID. The prevalence rate of adult-onset NIID may be higher than previously thought, and it may be underdiagnosed in the population. It must be considered during the differential diagnosis of dementia.

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Neuropalliative care in a tertiary-level hospital

Diego Belandrino Swerts[1], Ana Araújo[1], Polyana Vulcano[1], Bernard Prado[1], Caroline Miyake[1], Rafael Docema[1], Hye Hwang[1], Mario Fernando Prieto Peres[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Diego Belandrino Swerts; Email: diegobelandrino@gmail.com.

Palliative care in neurology is a recent speciality to improve the quality of life of patients with severe neurological diseases. This study aims to determine the frequency of neurological inpatients who had indication of palliative care, and evaluate the symptomatology, demographic profile, the need for supportive measures, advance directives for life and medical history of patients in a tertiary hospital in Brazil. This cross-sectional analytical study evaluated all patients admitted to the neurological semi-intensive care unit (ICU) with neurological conditions from February through August 2022. The Palliative Performance Scale, weight loss greater than 5% associated with body changes and a negative response to the question: "Would you be surprised if the patient died within 1 year?") was used to indicate palliative care. Patients were divided into three groups: patients with palliative care needs (group indication), patients without palliative care needs (group without indication) and patients who received at least one assessment of a palliative care team (group palliative). Demographic data were analysed using the Qui quadrado test for qualitative and Kruskal-Wallis test for quantitative variables. Of the 198 patients included in the study, 115 (58%) had palliative care needs. Only 6.9% received assessment by the palliative care team, and 9.56% had advance directives in their medical records. Patients in group indication had a higher prevalence of symptoms, such as fatigue, depression, shortness of breath and lack of appetite, and required more supportive measures, such as oxygen therapy, enteral/ parenteral nutrition,

admissions at ICU and days in hospital. Despite the high demand for palliative care in neurology, few patients receive this treatment, resulting in decreased quality of care. Therefore, greater integration and discussion of palliative care in neurology are needed.

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Neuropathy in V122I hereditary transthyretin amyloidosis: a multicenter retrospective cohort study

Anna Paula Paranhos Miranda Covaleski[1], Marcela C. M. Costa[2], Carolina Lavigne-Moreira[3], Diogo F. Santos[4], Marcondes C. França Junior[5], Jemima S. Batista[6], Osvaldo J. M. Nascimento[7], Eduardo B. U. Cavalcanti[8], Cleonisio R Leite[9], Wilson Marques Junior[3]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[3]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[4]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.
[5]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Campinas SP, Brazil.
[6]Universidade Federal do Vale do São Francisco, Petrolina PE, Brazil.
[7]Universidade Federal Fluminense, Niterói RJ, Brazil.
[8]Rede Sarah, Serviço de Reabilitação Neurológica, Brasília DF, Brazil.
[9]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Anna Paula Paranhos Miranda Covaleski; Email: paranhos.neurologia@hotmail.com.

Background: The V122I variant is typically associated to cardiomyopathy. Polyneuropathy is considered a minor manifestation. **Objective:** To evaluate the presence of neuropathy associated to the V122I variant in Brazilian referral centers and determine its demographic, clinical and neurophysiological characteristics among V122I carriers. **Methods:** A national, multicenter (10 referral centers), observational, retrospective cohort study. All V122I carriers underwent a demographic assessment. For clinical and EMG analysis we excluded: a compound heterozygosity in the TTR gene, the presence of comorbidities or other factors known to predispose a neuropathy, or patients who had not undergone EMG. The homozygous patients were analyzed separately. **Results:** We found 246 V122I carriers from 84 families: 240 heterozygous, 4 homozygous, and 2 compound heterozygous (V30M/V122I and I107V/V122I). Gender distribution was similar. Age:19-89 years, mean 53.4. Race: 26.7% White, 17.3% Black and 56% mixed. Individuals were born in all regions of Brazil, mainly in the states of Pernambuco (32.4%), Bahia (32.0%). Considering the 122 heterozygous submitted to clinical and electrophysiological analysis: 42.6% were symptomatic. Carpal tunnel syndrome (CTS) preceded the onset of polyneuropathy and/or cardiomyopathy in 69.2%. Onset manifestation excluding CTS: cardiomyopathy-52.9%, polyneuropathy-37.3% and mixed-9.8%. Mean age of disease onset: 64.9 years, 2.1 years earlier in homozygous. Follow-up: 14/27 cardiological patients developed polyneuropathy and 8/19 neurological patients developed cardiomyopathy. The mixed phenotype increased to 52.9%. Polyneuropathy: sensory/sensory-motor polyneuropathy-62.2%, small fiber neuropathy-32.4% and isolated dysautonomia-5,4%. Coutinho stage: stage1-92.3%, stage2-7.7%. Sensory symptoms-91.7%, weakness-44.4%, dysautonomia-14.3% to 54.3%, neuropathic pain- 52.8%. EMG was abnormal in 50%. Asymptomatic patients: CTS-20%, Radiculopathy-2.9%. Symptomatic patients: CTS-70.6%, Cubital tunnel syndrome-11.8%, sensory axonal polyneuropathy-13,7%, sensory-motor axonal polyneuropathy-35.3%, CIDP-like-2.0%, multiple mononeuropathy-3.9%, radiculopathy-11,8%. CTS was bilateral in 95.8%. SSR response was absent in 17.6%, never in isolation. **Conclusion:** The V122I variant was found in Blacks, mixed-race (Black/White), and White populations. The Northeast region is an endemic area, particularly Bahia and Pernambuco. Neuropathy is frequent at onset, and the only manifestation in 21·6% after follow-up. Symptoms related to dysautonomia, neuropathic pain and motor impairment were less common. CTS was the most frequent EMG finding, almost always bilateral. Cubital tunnel syndrome should be investigated regularly. An axonal polyneuropathy predominated; atypical patterns were also observed. Sympathetic skin response (SSR): absent in 17·6%, never occurring as an isolated change. CTS associated with an absence of SSR may be suggestive of amyloid neuropathy.

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Neurophobia and neuroaffinity: a study among medical students in a Brazilian federal Southeast University

Iuri Sena Melo[1], Roberto Pereira Santos[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Iuri Sena Melo; Email: iurisenamelo@gmail.com.

Background: With the increase in population longevity, there is a rise in the prevalence of neurological diseases. However, the training of specialists does not keep pace with the demand, leading to a potential shortage of neurologists. The scientific literature identifies a phenomenon known as neurophobia, characterized by a fear of neurological sciences and clinical Neurology, stemming from the inability to apply basic scientific knowledge to clinical situations. This topic has been evaluated in many countries and at various levels of medical education, prompting proposals for diverse strategies to prevent neurophobia throughout the academic trajectory. **Objective:** To identify the frequency of neurophobia among medical students and its possible causes. **Methods:** Cross-sectional study conducted with medical students divided into three subgroups: pre-clinical knowledge (2ºy of medical school), clinical knowledge (4ºy), and internship (6ºy). A standardized questionnaire was distributed to those who voluntarily agreed to participate, addressing levels of confidence, knowledge, interest, and ease in various medical specialties (scale 1-5). We created the Affinity Index (AI) based on the average values obtained - the lowest AI, the highest aversion to the speciality. We also enquired specifically for fear of Neurology and about possible causes for the negative perception of neuroscience knowledge (scale 1-4). **Results:** From 567 questionnaires sent, 58 responses were received. One was excluded due to inconsistent data. 42.1% reported fear of Neurology and/or neurosciences, but this percentage reached 56% in the last year of medical school. Neurology had the second-lowest value of AI (2.41), surpassed only by Rheumatology (2.07). The "ease in Neurology" had the lowest average value (1.98) compared to the other characteristics evaluated and also compared to the ease of other specialties, including Rheumatology (2.06). "Interest" in Neurology knowledge had the third-highest value (3.16), with only Nephrology (3.17) and Cardiology (3.29) scoring higher. Among the causes of fear of Neurology, the most associated factors were difficulties in neurophysiological knowledge, the complexity of diagnoses, insufficient class hours, and limited exposure to neurological patients. **Conclusion:** There is a high trend of aversion to Neurology, although in this small sample we found it less prevalent than in previous similar studies. Nonetheless, the high interest in Neurology compared to other areas may indicate that neurophobia does not reflect a disregard for its knowledge, being probably more associated with teaching failures. Among the possible causes for neurophobia, difficulty in "neurophysiology" was more prevalent, different from other studies who found it to be "neuroanatomy". In conclusion, this preliminary work points to the need for new medical education policies specifically aimed at teaching neurological sciences.

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Neurophysiologic data in a cohort of transthyretin familial amyloidosis patients: understanding where we stand on small fiber evaluation

Isadora Versiani[1], Eduardo Rodrigues Davidovich[1], Ligia Rocha Andrade[1], Salim Lazaro Balassiano[1], Robson Teixeira Vital[1], Clarissa Neves Spitz[1], Larissa Bittencourt de Carvalho Chaffin[1], João Vitor Ribeiro dos Santos[1], Livia de Almeida Afonso Abi-Haila[1], Marcia Rodrigues Jardim[1]

[1]Universidade do Estado do Rio de Janeiro, Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.

Correspondence: Isadora Versiani de Lemos; Email: isadora.versiani@gmail.com.

Background: Transthyretin familial amyloidosis (ATTRv) is a heterogeneous hereditary disease, endemic in our country, that gradually leads to significant disability. Recent advances in genetic research have allowed improvement in disease modifying treatment options. However, currently available therapeutic strategies show benefit when started early in the course of disease. It is therefore necessary to optimize diagnostic workup to ensure early detection. Neurophysiologic studies prove to be allies in managing these patients, and special techniques for evaluation of small nerve fibers are considered as possible tools for early diagnosis, since small fiber neuropathy is a common initial presentation of ATTRv. To contribute to future development of new diagnostic workup algorithms, we find it essential to debate where we stand regarding the use of neurophysiologic tests among these patients. **Objective:** Our goal is to analyze neurophysiologic data from our cohort of ATTRv patients, to learn where we stand and what we need to pursue to compare small fiber neurophysiologic studies, aiming to achieve earliest possible diagnosis. **Methods:** A cohort of 46 adult transthyretin mutation carriers was identified in our university neuromuscular outpatient clinic, from April/2019 to June/2024. Clinical and neurophysiologic data were collected via thorough chart review. Tests were done by our team on the same clinic. **Results:** Out of 46 transthyretin mutation carriers analyzed Val30Met mutations were detected on 18 patients, Val122Ile on 15 patients, Ile127Val and Phe84Leu on 5 patients each, and Glu74Lys on 1 patient. 2 patients had no documentation found of the specific mutation. 31 patients out of all 46 (67,4%) were symptomatic from a neurologic point of view. All symptomatic patients underwent routine nerve conduction studies, as well as 9 asymptomatic patients. Out of those tests, 14 came normal (35%), 20 showed sensorimotor polyneuropathy (50%), 1 showed pure sensory lower limb neuropathy (2,5%), and 5 showed carpal tunnel syndromes of varying degrees (12,5%). Only 6 patients (13%) underwent pain-related somatosensory evoked potentials, 1 with altered results. 17 patients underwent sympathetic skin response testing (42,5%), among which 9 were altered (52,9%). No patients had yet done R-R interval or electromyography silent period testing. **Conclusion:** Early accurate diagnosis defines ATTRv management and prognosis. Neurophysiologic studies focused on evaluation of small fibers may be an accessible, non-invasive and useful tool in this scenario. We found that although most of our mutation carriers underwent nerve conduction studies, only part of them had small fiber tests done. This shows the importance of bringing these tests and the need of standardization to light, and of continuing our search to compare different small fiber tests, aiming to identify whether one of them stands out as a potential standard.

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Neuroprotection and gliosis attenuation by memantine in a murine model of compressive spinal root lesion

Gabriel Gaspar Bíscaro[1], Arthur Ventura Martins Leão[1], Alexandre Leite Rodrigues de Oliveira[1], Luciana Politti Cartarozzi[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Gabriel Gaspar Bíscaro; Email: g.g.biscaro@gmail.com.

Nerve injuries frequently occur within the spinal canal, affecting the most proximal part of the peripheral nervous system, namely the spinal roots. This results in a longitudinal spinal cord injury, which is characterized by profound alterations in the spinal cord microenvironment. These include motoneuron apoptosis, chronic gliosis and synaptic pruning. Glutamate excitotoxicity, which is mediated by excessive NMDAr and AMPAr stimulation, plays an important role in its pathophysiology. Consequently, the use of NMDAr antagonists has been proposed as a promising treatment for central and peripheral nerve injuries. In this context, the present study aimed to investigate the putative neuroprotective effects of memantine, an NMDAr antagonist, after ventral root crush (VRC) in mice. Therefore, unilateral crushing of the ventral roots of L4, L5, and L6 was conducted in adult C57BL/6J mice, which were divided into four experimental groups: vehicle, and memantine at doses of 30 mg/kg, 45 mg/kg, and 60 mg/kg. The mice were administered memantine or vehicle solution via gavage daily for a period of 14 days following the injury. Twenty-eight days after the lesion, the mice were perfused, and the lumbar intumescences were collected and processed for histological analyses. The number of motoneurons was quantified in both sides of the respective spinal cord segments (following toluidine blue staining) in order to calculate the motoneuron survival rate. Astrogliosis and microglial reaction were assessed by immunohistochemistry for GFAP and Iba-1, respectively. Immunohistochemistry was further applied to synaptophysin and VGLUT-1 to evaluate synaptic pruning and plasticity. The protocol for the use and handling of animals was approved by the local ethical committee (CEUA protocol number 5740-1). The administration of memantine following VRC, at all studied doses, was associated with a significant increase in motoneuron survival. The 45 mg/kg group exhibited the most pronounced effect ($p<0.001$). Memantine also demonstrated the capacity to downregulate microgliosis in the doses of 45mg/kg and 60 mg/kg ($p<0.01$ and $p<0.05$, respectively), and astrogliosis in all treated groups in comparison to the vehicle ($p<0.01$). Moreover, the administration of memantine at a dose of 45mg/kg was found to result in a significant reduction of activated microglia ($p<0.05$ compared to vehicle group), as evidenced by morphological characterization. Memantine treatment was also associated with an increase in motoneuron synaptic coverage, which was observed in all doses ($p<0.001$). However, no statistically significant differences were observed in VGLUT-1 expression among the experimental groups ($p=0.08$). These findings suggest that memantine may have a significant impact on motoneuron survival, gliosis reduction, and inhibition of synaptic pruning, which could have implications for clinical applications.

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Neuroprotective effect of therapeutic hypothermia in critically ill patients with traumatic brain injury: a systematic review

Loyslene Maria de Paula Bonifácio[1], Leandro de Souza Cruz[1]

[1]Faculdade de Ciências Médicas e Saúde de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Loyslene Maria de Paula Bonifácio; Email: loyslene.med@gmail.com.

Background: Therapeutic hypothermia has emerged as the most extensively studied neuroprotective intervention in laboratory settings, demonstrating significant neuroprotective benefits across various neurological disorders. However, its clinical application remains limited due to challenges in routine medical practice and adaptation to specific pathological conditions. In recent years, therapeutic hypothermia has been recommended for traumatic brain injury with the aim of normalizing intracranial pressure while providing neuroprotective effects. Traumatic brain injury is a leading cause of mortality and morbidity globally. Clinical research indicates a negative correlation between elevated body temperature and outcomes in traumatic brain injury patients, suggesting that lowering the body temperature to around 35°C may reduce neurological damage. **Objective:** To evaluate the neuroprotective effect of therapeutic hypothermia on neurological function in critically ill patients with traumatic brain injury in an intensive care unit setting, through a systematic review. **Methods:** Randomized controlled trials published in English over the past decade were analyzed, focusing on adult human subjects (18 years and older). Data sources included the National Library of Medicine (MedLine) and the Brazilian Scientific Electronic Library Online (SciELO). The search utilized terms from the Health Sciences Descriptors (DeCS), specifically: induced hypothermia, traumatic brain injuries, and intensive care. Studies involving critically ill TBI patients treated with therapeutic hypothermia were included, while those with unclear methodologies, available only in abstract form, involving pediatric participants, or where therapeutic hypothermia was used in cardiac arrest were excluded. The PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) scale was used to enhance the reporting quality of this review. **Results:** Nine studies were analyzed, encompassing 1304 participants aged 19 years and older with traumatic brain injury. The effects of therapeutic hypothermia on mortality and neurological recovery during intensive care and subsequent months were examined. Statistical tests with a p-value <0.05 were considered significant. Despite the large body of preclinical and clinical research on therapeutic hypothermia, there is no robust evidence supporting its neurological benefits in the short and long term for critically ill traumatic brain injury patients. However, clinical trials indicate a reduction in mortality and neurological damage. **Conclusion:** The best available evidence supports the use of mild to moderate therapeutic hypothermia in traumatic brain injury patients to reduce mortality rates and enhance neurological recovery. Nevertheless, further randomized controlled trials are necessary to consolidate these findings

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Neuropsychiatric symptoms are related to CSF amyloid and tau in individuals with subjective cognitive decline and mild cognitive impairment

Ana Luiza Gonçalves Rochetti[1], Isadora Cristina Ribeiro[1], Ítalo Karmann Aventurato[1], Marjorie Cristina Rocha da Silva[1], Brenda Costa Gonçalves[1], Liara Rizzi[2], Marcio Luiz Figueredo Balthazar[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Laboratório de Imagem e Biomarcadores em Doenças Cognitivas, Campinas SP, Brazil.
[2]University of California San Francisco, Memory and Aging Center,

Correspondence: Gabriela Barbosa Rodrigues; Email: g242237@dac.unicamp.br.

Background: Alzheimer's disease (AD) is a neurodegenerative disorder that has become increasingly prevalent around the world and can be characterized in vivo by amyloid-beta peptide and hyperphosphorylated tau protein. Subjective cognitive decline (SCD) and mild cognitive impairment (MCI) are potential previous stages of AD dementia. Behavioral and psychological symptoms are common in SCD and MCI, but their biological basis is still not clarified. **Objective:** To investigate the potential correlations between neuropsychiatric symptoms and AD biomarkers in patients with SCD and MCI. **Methods:** 71 subjects were recruited for a medical and psychological evaluation in the Clinical Research Center of Universidade Estadual de Campinas, Brazil. They were diagnosed according to NIA-AA criteria and underwent neuropsychiatric scales, including the Mild Behavioral Impairment Checklist (MBI-C), a helpful questionnaire to measure behavioral and neuropsychiatric symptoms in the AD continuum. The subjects also underwent CSF collection to measure the biomarkers in the samples using immunoassay kits. We used Pearson correlation for the statistical analyses. **Results:** We found statistically relevant correlations between the MBI-C C domain and the CSF amyloid-beta ($p=0.026$, $r= -0.302$, $n=54$) and a correlation between the MBI-C E domain and the CSF total-tau protein ($p=0.026$, $r= 0.295$, $n=57$). **Conclusion:** The MBI-C C domain approaches symptoms of impulse dyscontrol, and the MBI-C E domain comes with symptoms of abnormal perception or thought content. We found that psychological symptoms imply an association of these symptoms with the neurodegenerative process due to the correlation with total-tau protein, a more specific biomarker.

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Neuropsychiatric systemic lupus erythematosus: rare manifestations in Brazilian Amazon, a case report

Frederico de Sousa Marinho Mendes Filho[1], Tales de Oliveira Junior[2], Marystela Batista Martins[2], Igor Menezes Martins[1], Julia Ágata Cardoso Barbosa[1], Eduarda Kei-lan Tavares Pierre[3], Fernando Luiz Westphal Filho[1], Paulo Roberto Moss Lopes[1], Jonathas Taumaturgo Soares[3], Vanise Campos Gomes Amaral[3]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Fundação Hospital Adriano Jorge, Manaus AM, Brazil.
[3]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Frederico de Sousa Marinho Mendes Filho; Email: frederico.smmf@gmail.com.

Case presentation: 34 years old female patient presented, in January 2024, with the symmetric, additive, and inflammatory course of poly arthritis, in symptomatic therapy at the moment without clinical benefit. In February, developed multiple episodes of emesis, asthenia, hyporexia, and mood disorder, with a history of 10 Kg weight loss. In April she developed alopecia, microvascular lesions in the skin, anemia, and worsening mood disorder, being admitted to Dr. João Lúcio Pereira Machado in a post-ictus state, after the first impaired awareness of tonic-clonic seizure event, and submitted to abortive therapy she developed proximal upper and lower limb par during the admissions, compromising her gait. After two days she presented symmetric lower limb paralysis and upper limb paresis, oropharyngeal dysphagia, and dyspnea, being transferred to Fundação Hospital Adriano Jorge for diagnostic investigation. At the time, she presented through a physical exam, psychomotor agitation, associated with disorientation in space and time, depressive affect, horizontal nystagmus, oral ulcers, symmetric +2 lower limb edema, +2 muscle weakness in the upper and lower limb, preserved deep tendon reflexes. She underwent several laboratory testings, highlighting anemia (6 hemoglobin), direct coombs test positive, renal impairment (4,5 creatinine), 24h proteinuria as 753,4 mg/24h (1.395 ml), as well as image exams such

as thoracic computed tomography (CT) scan revealing bilateral pleural infusion, cranial CT scan without signs of ischemia or bleeding, nuclear magnetic resonance without signs of vasculitis, renal ultrasound revealing bilateral nephritis and ascites. Due to central nervous system affection, worsening of agitation, and irritability, it was indicated a lumbar puncture and cerebrospinal fluid analysis to guide the treatment, presented no signs of infection, elevated protein levels, normal glucose, absence of cells, compatible with an inflammatory pattern. Autoantibody testing revealed a 1:640 mixed pattern anti-nuclear antibody, anti-Ro/SSA positive, and anti-LA/SSB negative. **Discussion:** The case presented evidentiate SLE patient, which is a chronic idiopathic autoimmune disease affecting multiple organs, including the central nervous system, presenting with several neurological syndromes such as seizure disorder and psychosis. The recurrent seizure events are a less common manifestation of Neuropsychiatric systemic lupus erythematosus (NPSLE) and psychosis presents rarely in less than 1%, presenting mood disorders and acute confusional state. Due to the presence of nephritis associated, she underwent pulse therapy with cyclophosphamide, followed by mycophenolate mofetil with significant clinical benefit. **Final comments:** Due to the unusual presentation, it is highlighted the importance of relating this case, the patient had severe clinical affection due to generalized disease activity with multiple organ damage associated with NPSLE, such as lupus nephritis.

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Neurorehabilitation of a patient with alpha-mannosidosis: a case report

Marcos Matheus da Silva Fonseca[1], Leonardo Honorato Cheng[1]

[1]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Marcos Matheus da Silva Fonseca; Email: marcosmatheus6@gmail.com.

Case presentation: A 44-year-old female patient, daughter of consanguineous parents, was admitted at the neurological rehabilitation clinic with history of delayed neuropsychomotor development, manifested by speaking and walking difficulties (tiptoe gait), frequent falls due to weakness in the lower limbs, poor school performance and hearing problems since childhood, requiring the use of a hearing aid. By the age of 20, she began to show behavioral changes and visual hallucinations, and was diagnosed with schizophrenia, currently stable. About 3 years ago, the patient developed progressively worse weakness in her lower limbs, requiring the use of a 4-point walker for walking at home and a wheelchair for locomotion in the community. On physical examination, she was lively, oriented in time and space, speech was slightly slurred, language was preserved, with no alterations in the cranial pairs; gait with support on a walker, with slowed speed and cadence, with an extensor pattern in the left hip due to a pain limitation, with no signs of imbalance or incoordination; overall motor strength was reduced, but sensitivity was preserved; index-index test with the presence of dysmetria and decomposition, no postural instability on the Romberg test; hypoactive osteotendinous reflexes overall; heel-knee test not performed due to pain in the hip joint, where there was a bilateral osteonecrosis of the femoral head, according to imaging tests. The diagnostic hypothesis of Alpha-mannosidosis was suggested by the patient's family history, who has a sister with the disease, and was confirmed by genetic testing (Alteration C.2696C > A P. (SER899*) in homozygosity in Exon 22 of the MAN2B1 gene). The therapies proposed were neurofunctional physiotherapy, aimed at balance and proprioception training; orthostatism and gait training with aids; global muscle strengthening, with emphasis on the lower limbs and antigravity muscle groups; stretching training; and occupational therapy, aimed at training activities of daily living, posture change, ergonomics and energy conservation training and routine enrichment measures. **Discussion:** Alpha-mannosidosis is a rare, progressive genetic disease caused by a deficiency of enzyme alpha-mannosidase. This deficiency leads to the accumulation of a substance called mannose oligosaccharide in various parts of the body, resulting in progressive damage to organs and tissues. The symptoms can vary in severity and include physical deformities, intellectual impairment, respiratory, skeletal and visual problems. There is no cure for alpha-mannosidosis, and treatment is mainly aimed at relieving symptoms and improving the patient's quality of life. Supportive therapies such as physiotherapy, occupational therapy and speech therapy may be recommended. **Final comments:** due to the complex nature of this condition, it is important that patients are followed up by a multidisciplinary medical team to ensure comprehensive and personalized treatment.

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Neurosarcoidosis and metastatic testicular seminoma: case report

José Victor Ribeiro Silva Gomes[1], Ligia Henriques Coronatto[2], Gabrielly Pereira Figueira[3], Kananda Oliveira Garcia Ruiz[1], Rafael Fonseca Drumond[1], Natália Silva Fernandes[2], Bruna Gutierrez Gambirasio[2], Adrialdo José Santos[1]

[1]Universidade Federal de São João do del-Rei, São João del Rei MG, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[3]Universidade São Judas Tadeu, Cubatão SP; Brazil.

Correspondence: José Victor Ribeiro Silva Gomes; Email: josevrsgomes@gmail.com.

Case presentation: A 34-year-old male patient presented with paresthesia and hemiparesis in the right limb. After 2 years, he presented with a progressive loss of fixed muscle strength in the left lower limb. Brain CT revealed a juxtacortical nodular formation in the right perirhinal region, associated with vasogenic edema of the adjacent parenchyma. Under the initial hypothesis of metastatic disease in the Central Nervous System (CNS), oncological screening was carried out and retroperitoneal lymph node enlargement and atrophic changes in the left testicle were found. The cortical lesion, investigated with brain MRI, revealed a "target" appearance, with central peripheral enhancement, and a dramatic response to treatment with dexamethasone. The patient developed severe abdominal pain, with radiological evidence of enlarged retroperitoneal lymph nodes in the periaortic and paracaval chains, with necrotic content. A lymph node biopsy revealed an infiltrative/metastatic connective tissue seminoma, treated with chemotherapy and currently in remission. The patient also developed epitheliopathy in the right eye and trigeminal neuralgia. The patient is still taking dexamethasone, with a sustained response to CNS lesions, and is clinically and neurologically stable. **Discussion:** Sarcoidosis, a chronic systemic granulomatous disease, has an unknown etiology and can affect the nervous system in approximately 5% of cases. In this condition, any part of the central or peripheral nervous system can be affected. In this case report, several corroborative findings support the clinical diagnosis, without confirmation by biopsy, of sarcoidosis: an inflammatory lesion in the CNS, highly responsive to corticotherapy; trigeminal neuralgia, and ocular epitheliopathy. In addition, the patient presented criteria for possible neurosarcoidosis, such as clinical manifestations and images suggestive of granulomatous inflammation, excluding other differentials, without pathological confirmation. Evolutionarily, the oncological picture of a seminomatous tumor supports the diagnostic hypothesis since previous data in the literature has already shown a correlation between this histological subtype and granulomatous disease. In most cases reported, the oncological diagnosis precedes or is simultaneous with the inflammatory diagnosis. From a pathological point of view, it is known that some seminoma-type tumors have granulomatous reactions (local, metastatic, or even in distant sites not affected by the cancer) **Final comments:** The synchronous occurrence of testicular seminoma and neurosarcoidosis is rare, although it has been reported in the literature. More studies are needed to understand the real pathophysiological correlation between the diagnoses presented here.

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Neurosensory anacusis after traumatic brain injury: a case report

Hélio Aquaroni Farão Gomes[1], Itamar Meireles Andrade Santos[1], Dominick Esthephanny Monteiro Silva[1], José Eduardo Trentim Longo[1], Caroline de Barros Viegas[1], Eduardo Abrão Spinola Rezk[1], Gabriela Rossini Dantas[1], Marcos Vinicius de Almeida Silva[1], Patrick Emanuell Mesquita Sousa Santos[1], Daniel Fabiano Barbosa dos Santos[1]

[1]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Hélio Aquaroni Farão Gomes; Email: heliofarao@hotmail.com.

Case presentation: AFP, 42 years old, with no previous comorbidities, suffered a car rollover accident presenting traumatic subarachnoid hemorrhage without the need for surgical intervention. During hospitalization, bilateral hearing loss was noted, and the patient was evaluated by the otolaryngology team, who performed auditory brainstem response (ABR) testing with normal latency values and interpeak values, normal tympanometry, and transient evoked otoacoustic emissions present in all frequency bands, leading to a referral to Neurology after 7 days due to worsening hearing. On evaluation, the patient reported more difficulty hearing low-pitched sounds, could hear the ringtone of a cellphone but had difficulty hearing people's speech. Additionally, the patient reported visual misalignments without diplopia and taste alterations. Neurological examination revealed no motor or sensory alterations, negative Romberg sign, preserved tandem gait, cerebellar tests showed dysmetria to the right, intact cranial nerves regarding ocular motricity or visual fields, preserved sensory and motor evaluations, Weber test was non-lateralizing, positive Rinne test, preserved taste sensation with glucose and salt, suggestive of a neurosensory disorder. Cerebrospinal fluid analysis showed 0 leukocytes, 62 proteins, lactate 1.2, with no infectious alterations, negative cultures, and MRI displayed hyperintense lesions in bilateral inferior colliculi, more prominent on the right, and increased density in the left vestibular nerve on SPIR and FLAIR sequences. The isolated lesion resulting from traumatic contusion was confirmed, not affecting other brain structures. Rehabilitation measures were indicated, and on a 2-month follow-up, the patient reported partial improvement in hearing, more noticeable on the left, continuing with biannual follow-ups for reassessment and rehabilitation. **Discussion:** Trauma-related neurosensory hearing loss commonly occurs with temporal bone fractures and is often associated with neurological symptoms like facial paralysis or fistulization, which were not present in this case. Traumatic cases usually affect the vestibular portion of the nerve, leading to vertigo or unilateral vestibular dysfunction with nystagmus. **Final comments:** The uniqueness of this case lies in the exclusive and bilateral auditory involvement with lesions in the inferior colliculi, an uncommon presentation in TBI, concussions, or brain contusions, or even for cerebrovascular injuries. The rehabilitation of bilateral anacusis is very complex due to impaired communication, so adaptations are necessary in comprehensive care and clinical improvement measures to allow the use of a hearing amplifier and return to usual activities.

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Neurosyphilis associated with uveitis and the many faces of an inflammatory disease: a case report

Danilo Cabral Pentagna[1], Leonardo Luiz Lino[1], Renato Cabral Pentagna[2], Cristiane Marcos Soares Dias Ferreira[3]

[1]FCMS, SUPREMA, Juiz de Fora MG, Brazil.
[2]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.
[3]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.

Correspondence: Leonardo Luiz Lino; Email: leonardolino5@gmail.com.

Case presentation: A 33-year-old woman presented to the emergency department with complaints of blurred vision and scotomas in oculus uterque (OU) for 20 days. Figure 1 illustrates colored retinography, red-free retinography and autofluorescence, and the latter denotes diffuse hypoautofluorescent spots in macular region in OU and superior temporal peripheral laser marks in left eye. Fluorescein angiography reveals bilateral diffuse hyperfluorescent macular spots, without extravasation, suggestive of bilateral acute macular chorioretinitis (Figure 2). These tests, combined with a VDRL titer of 1:64 and a positive treponemal test, corroborate the diagnosis of bilateral uveitis due to syphilis. The patient was hospitalized to receive crystalline penicillin for 14 days with improvement of visual symptoms on D1. Collected cerebrospinal fluid (CSF): clear; LDH 24; glucose: 54; proteins: 27; albumin: 0,7; lymphocytes: 100%; leucocytes: 2; chloride: 124; Cryptococcal antigen: negative; bacterioscopy: negative; FTA-ABS IgG: positive; VDRL: non-reactive. FAN and ANCA (non-reactive); Latex (< 15). On D14, after uneventful hospitalization and complete resolution of symptoms, the patient was discharged subsequent to 2.400.000 IU benzylpenicillin administration. She was advised to return in 3 months to VDRL and CSF (FTA-ABS IgG+) reevaluation, realize ophthalmological follow-up and the need of sexual partner assession. **Discussion:** Ocular syphilis can occur in any stage of syphilis and clinical manifestations include optic disc edema, uveitis, vitreitis, corioretinitis, vasculitis, retinal detachment and acute retinal necrosis, being uveitis the most common. All patients with syphilis presenting ocular symptons should have their cerebral spine fluid (CSF) tested due to co-ocurrence of neurosyphilis and evaluation of human immunodeficiency virus co-infection. A positive CSF-VDRL is the Gold standard for neurosyphilis diagnosis, with sensibility of 50-80% and high specificity. FTA-ABS, such as other treponemal tests present high sensibility but low specificity to diagnose neurosyphilis. Ocular syphilis and neurosyphilis treatment consist of 18-24 million units of intravenous aqueous crystalline penicillin G per day for 10-14 days. **Final comments:** There is a recrudescence of syphilis cases and its complications worldwide according to the Panamerican Health Organization. Uveitis may be the first symptom of the disease, so its promptly investigation is mandatory to provide early diagnosis, treatment and greater prognosis, preventing vision loss and in many cases, completely symptoms resolution.

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Neurosyphilis presenting as multiple cranial nerve palsy in an HIV-positive patient: a case report

Amanda Cipriano Torquato[1], Tainara Martins de Oliveira[2], Wanessa Caroline Ferreira[1], Maria Eduarda Oliveira Araújo[2], Paula Ravenna Sampaio Bastos[3], Arthur Pena Ferreira[3], Mariana Sotte da Silva[3], Mateus Nader Cunha[3], Grasielle Xavier[3]

[1]Universidade Vale do Rio Doce, Governador Valadares MG, Brazil.
[2]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[3]Hospital Municipal de Governador Valadares, Serviço de Neurologia, Governador Valadares MG, Brazil.

Correspondence: Amanda Cipriano Torquato; Email: amanda.cipritorquato@gmail.com.

Case presentation: Male patient, 35 years, HIV-seropositive for ten years in treatment (six months ago his viral load was undetectable), comes to the emergency service referred from primary care after one month with right peripheral facial palsy, partial hearing loss with related dizziness and diplopia that improves with closing the right eye. At the examination, the Skew test was positive. He presented an anterior positive rapid syphilis test and received two doses of intramuscular benzathine penicillin. The blood count, coagulogram, renal function and electrolytes were within the patterns; the serum venereal disease research laboratory (VDRL) was 1:32. It was hypothesized the diagnosis of neurosyphilis and the lumbar puncture and cranial magnetic resonance imaging (MRI) was performed. The cerebral spine fluid (CSF) analysis displayed a positive VDRL; glucose 49mg/dl (more than ⅔ of capillary glucose); white blood cell count 1074 cells/µl, 75% of neutrophils. Gram stain and culture were negative. Cranial MRI showed thickening after gadolinium administration in the intracanalicular portion of the VII cranial nerves bilaterally, being slightly evident on the right, probably related to neuritis. There was no evidence of ischemic attacks. The diagnosis was neurosyphilis with multiple cranial nerve palsy and the patient started on intravenous penicilin G. After a fourteen-day course, he exhibited clinical improvement. **Discussion:** Neurosyphilis associated with multiple cranial nerve syndrome is a rare presentation of neurosyphilis, representing an atypical form of the infection that can occur at any stage of tertiary syphilis. This condition is characterized by simultaneous involvement of multiple cranial nerves, which can lead to a variety of neurological symptoms, including facial weakness, hearing loss, diplopia, dysphagia, and facial pain. The pathogenesis involves inflammation and vasculitis of the cranial nerves and their nuclei, resulting in multifocal neurological deficits. Radiologic findings may reveal thickening of the affected cranial nerves. CSF analysis may show pleocytosis, increased protein, and a positive VDRL, although it may be negative in some cases. Contrast-enhanced magnetic resonance imaging may demonstrate enhancement of the cranial nerves. Intravenous penicillin G is the standard treatment, and corticosteroids may be considered to reduce inflammatory processes. **Final comments:** Neurosyphilis associated with multiple cranial nerve syndrome is an important clinical presentation to be considered in patients with multifocal neurological symptoms, especially those with a history of sexually transmitted infections or positive VDRL. Early diagnosis and correct treatment are essential for a good prognosis. In addition, VDRL testing and CSF analysis should be part of the evaluation in suspected patients, allowing a good therapeutic approach.

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Neurosyphilis with atypical presentation: case report

Sarah de Souza Chinelato[1], Guilherme Leonel Gomes da Cunha[1], Bruna Santos Silva Guzzardi[1], Rodrigo Batista Irikura[1], Anouar Sacca Colognesi[1], Gustavo Carvalho Viveiros[1], Isabella Brasileiro Quintella Migueis[1], Talles Camin Rajab[1], Fábio de Araujo Pereira[1], Luiz Henrique Soares Santos Stefano[1]

[1]Faculdade de Medicina de Marília, Marília SP, Brazil.

Correspondence: Rodrigo Batista Irikura; Email: rodrigoirikura@gmail.com.

Case presentation: Female patient, 35 years old, from Oscar Bressane - SP, referred for evaluation at the emergency department due to diplopia that started 1 month ago. The patient also complained of changes in ocular motility associated with a pulsatile right-sided hemicranial headache, starting in the periorbital region and radiating to the ipsilateral temporal region. She denied loss of visual acuity or other associated complaints. After ruling out ophthalmological causes, the patient was referred for evaluation by the Neurology team. Neurological examination revealed alterations in extrinsic eye movement, with paralysis of the right superior rectus and oblique muscles, reporting diplopia only when looking vertically upwards. Complementary imaging tests and cerebrospinal fluid analysis showed no significant changes. However, serological testing for syphilis was positive, with reactive treponemal test and VDRL result of 1:8. **Discussion:** The diagnosis of neurosyphilis can pose a challenge in medical practice, as infection of the central nervous system by Treponema pallidum can occur at any stage of the disease, although most patients with this condition have a negative VDRL in cerebrospinal fluid. In cases of atypical ocular presentations, such as in this patient, ophthalmoplegia due to cranial nerve III damage can be included in the differential diagnosis of any ocular inflammation in the context of syphilis infection. Therefore, it is important to include syphilis screening in laboratory tests for patients with these ophthalmological symptoms. **Final comments:** The presence of clinical signs consistent with cranial nerve III palsy associated with positive serological tests for syphilis led to the diagnosis of neurosyphilis. The patient was promptly treated with Crystalline Penicillin for 21 days and subsequently underwent corticosteroid therapy, with a good clinical response. Neurosyphilis occurs in approximately 25 to 35% of patients diagnosed with syphilis, but of these, only 10% present with cranial nerve abnormalities like the patient in this report, making this an atypical presentation of neurosyphilis.

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Neurosyphilis with ocular involvement

Jackson Batista da Silva[1], Mayara Romero Canal[1], Mariana Tonon Giovannone[1], Ketimila Aparecida Pereira[1], Fernando Pierini Costa[1], Mauro Gomes Araujo[1], William Thiago Almeida Leite[1], Natasha Cutolo[1], Eduardo de Almeida Guimarães Nogueira[1], Ana Luiza Vilela Sá Fortes[1]

[1]Santa Casa de Santos, Santos SP, Brazil.

Correspondence: Jackson Batista da Silva; Email: jacksonbs1992@gmail.com.

Case presentation: CLS, 54 years old, married, seeks care for asthenia, edema and joint pain associated with episodes of fever that have not been measured for 4 weeks. History of type 2 DM, under investigation for reactive arthritis. On admission, ophthalmology assessed hyperemia bilateral ocular, photophobia and edema and blurring of bilateral optic disc. We opted for hospitalization and investigation with serology for STIs, arboviruses, rheumatological screening, magnetic resonance imaging (MRI) of the brain/orbits and CSF collection. Observed rapid treponemal test FTA-ABS IgM reagents, CRP 5mg/dL; ESR 70mm/h; and CSF with glucose 70 mg/dL, proteins 135 mg/dL, VDRL reagent ¼. The diagnosis of neurosyphilis was made and treatment with crystalline penicillin G was started. **Discussion:** Syphilis is a curable STI, caused by the bacterium Treponema pallidum, transmitted through unprotected sexual intercourse with an infected person, blood transfusion and vertically, with various clinical manifestations and different stages. The diagnosis is challenging. There are increasing cases of ocular involvement caused by Treponema pallidum infection, which is why the diagnosis should be considered in all cases with unexplained neuro-ophthalmic findings, regardless of sexual history. Neurosyphilis is usually a late manifestation, but it can occur in the early stages of the disease, in immunocompromised individuals or in any infected individual.

According to the literature, treponemal neuroinvasion is seen in approximately 30-70% of patients with early syphilis, of which 80% have asymptomatic spontaneous recovery. The others evolve with different clinical syndromes, often with neuropsychiatric symptoms, cognitive disorders, autonomic dysfunctions, paresthetic syndromes, visual disorders, uveitis, neuroretinitis and even dementia when more advanced. Ocular manifestations can occur at any stage of the disease and target practically the entire ocular system (vessels, chambers, optic nerve, pupillomotor pathways and cranial nerves). Diagnosis is made through non-treponemal testing (indicators of disease activity) and confirmatory tests (FTAbs). Once reagents are associated with neurological symptoms or suspected neuroinvasion, CSF collection is performed. If positive, pleocytosis and hyperproteinorrhaquia are observed. Therefore, the case is now treated as neurosyphilis, regardless of whether or not there is positive serology in the sample. In this analysis, treatment with crystalline penicillin G was carried out 4 million units intravenously every 4 hours for 14 days. **Final comments:** Syphilis manifests itself in the body with the most diverse clinical syndromes, which is why it is called the great imitator. Eye involvement has been better and more observed, since for treatment purposes it must be treated as neurosyphilis. Highlighting that the probability of CNS and ocular syphilitic involvement increases in HIV co-infection, which is why screening for STIs is essential.

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Neurotoxic effects associated with anabolic androgenic steroids abuse: a systematic literature review

Willian Alves Costa[1], Gisele Rocha Lopes[1], Talita Alves Harrop[1], Juliana Alencar Isacksson Vieira[1], Caroline Lima de Freitas[1], Hanna Moraes dos Santos[1], Kauê Magalhães Castro dos Santos[1], Maria Eduarda Garcia de Azevedo[1], Hellen Karine dos Santos Almeida[1], Thiago da Costa Brito[2]

[1]Universidade Federal do Amapá, Macapá AP, Brazil.
[2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Willian Alves Costa; Email: willianac2021@gmail.com.

Background: Due to the widespread abuse of anabolic-androgenic steroids (AAS) among professional athletes and amateur sportspeople and their related health problems, determining the effects of these substances on the nervous system becomes vital for designing efficient preventive and educational measures. **Objective:** To investigate the main neurological effects associated with the abuse of anabolic-androgenic steroids. **Methods:** This systematic review was conducted through comprehensive searches of electronic databases, including PubMed, MedLine, Scopus, and Google Scholar (between 1980 and December 2023, in English), and also from citations in the selected articles. Keywords such as "anabolic-androgenic steroids", "AASs" and "neurological effects" were searched. Randomized clinical trials, prospective or retrospective human studies and experimental studies (in vivo) were included. **Results:** Twenty-one clinical and experimental articles were selected (12 for anabolic-androgenic steroids and 9 for adverse effects). The neurotoxic effects of AAS are associated with both the membrane androgen receptor (AR) and G protein-coupled receptors. Apoptosis plays a crucial role in brain damage, caused by high concentrations of methandienone and 17-a-methyltestosterone. These substances inhibit the maintenance of the neuritic network, leading to cell death by apoptosis and cleavage of protective chaperone proteins, as Hsp90. The apoptosis in brain areas of rats treated with nandrolone has been suggested to be associated with oxidative stress and NF-Kb signaling, affecting the hippocampus, striatum, and frontal cortex. In another study, prolonged oxidative stress due to AAS exposure was also related to lower cognitive functions, such as motor and executive performance. Dysregulation of miRNA may also contribute to AAS-related brain damage, with a significant increase in miR-34 and miR-132 observed in AAS users compared to cocaine abusers and the elderly. Neuroimaging studies have shown a reduction in global gray matter, cortical volume, putamen, cortex, and right amygdala volume, as well as a marked decrease in dACCgln/glu and Scyllo-inositol levels. Additionally, a chronic administration of EAA in high doses is linked to anxiety-like behavior through the corticotrophin-releasing factor, increasing gabaergic inhibitory effects from the central amygdala to the bed nucleus of the stria terminalis. Furthermore, abuse of EAA alters the phosphorylation of the NMDA receptor, with an increase in excitatory neurotransmission, affecting the control of aggression, which is intensified by a reduction observed in the orbitofrontal cortex in these cases. Finally, people who use AAS are more likely to be drug and alcohol abusers. **Conclusion:** The cognitive and neurodegenerative effects associated with EAA abuse have a wide range of mechanisms involving apoptosis, miRNA derangement, oxidative stress, and alterations in neurotransmitters and neurohormones.

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Neurotoxoplasmosis in a patient with a CD4 cell count above 500: a case report

Lara Mensato Döring[1], Afonso Henrique de Aragão[2], Louissa Srama Rosner Cidral[3], Crisslley Mulbauer[4], Igor Baroni Cardoso[1], Ayumi Oya Santos[4], Amanda Yumi Ikematu[4], Johnatan Neuls Pasta[5], Nátali Pimentel da Silva[6]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Santa Casa de Misericórdia de Curitiba, Curitiba PR, Brazil.
[3]Universidade Positivo, Curitiba PR, Brazil.
[4]Pontifícia Universidade Católica do Paraná, Curitiba PR, Brazil.
[5]Hospital da Cruz Vermelha Paraná, Curitiba PR, Brazil.
[6]Unifacear, Curitiba PR, Brazil

Correspondence: Lara Mensato Doring; Email: laradoring@ufpr.br.

Case presentation: A 63-year-old male patient was admitted to the hospital because his wife noticed behavioral and gait changes the previous day. According to her, he appeared distracted, disoriented, and stumbled several times during this period. He has HIV, hypertension, and heart disease as comorbidities, however adheres to his pharmacological treatments. Four days prior, the patient had been hospitalized in another city for acute respiratory failure of an unknown etiology, without the need for orotracheal intubation. During the second hospitalization, he reported a pulsing headache in the right hemicranium, triggered by coughing. Upon clinical examination, no other abnormalities were observed. During hospitalization, he became disoriented in time and space and began to desaturate. Additional examinations of cerebrospinal fluid, mucus, and blood ruled out the presence of acid-fast bacilli, fungi, Epstein-Barr virus, and bacteria; IgG and IgM antibodies for toxoplasmosis were non-reactive; HIV viral load was undetectable. A full blood count showed 15,500 leukocytes/mm^3 and an absence of basophils. Urea, aspartate aminotransferase, and C-reactive protein levels were elevated. The CD4 lymphocyte count was 629.7/μL. An MRI revealed a hemorrhagic lesion in the right temporoparietal corticosubcortical region, associated with vasogenic and cytotoxic edema, not typical of toxoplasmosis. A biopsy of the brain lesion exhibited round structures morphologically suggestive of tachyzoite forms of Toxoplasma sp., confirming the diagnosis through pathology. **Discussion:** Neurotoxoplasmosis typically presents with similar symptoms to other central nervous system infections, including headaches, motor disturbances, mental confusion, and sometimes ocular and pulmonary manifestations. Therefore, the diagnosis relies on clinical, radiological, serological, and histological examinations. Toxoplasma gondii rarely affects the nervous system of individuals with an adequate immune system. In HIV-positive patients, the risk of developing neurotoxoplasmosis is high when CD4 levels are below 100/μL and extremely rare in immunocompetent patients. Consequently, chemoprophylaxis in immunocompromised individuals with a positive IgG for toxoplasmosis is maintained until CD4 levels exceed 200/μL. **Final comments:** The effectiveness of recent treatments for HIV/AIDS has reduced the incidence and severity of opportunistic infections. Despite the correlation of neurotoxoplasmosis with HIV-positive patients, controlling the disease, indicated by an undetectable viral count and a CD4 lymphocyte count above 200/μL, makes the infection unlikely. Therefore, this clinical case shows an exception and underlines the importance of not disregarding neurotoxoplasmosis as an opportunistic infection of the nervous system in HIV-positive patients, even when controlled.

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Neurotoxoplasmosis induced by the use of multiple immunosuppressants in a patient with refractory dermatopolimyositis: case report

Maria Eduarda Vieira Fiber[1], Fernanda Kelly Moreira Almeida[1], Ana Paula Guadagnin[1], Bárbara Isabella Wentz Costa[1], Julia Mariana dos Santos Silva[1], Luana Mesquita de Lima Floriano[1]

[1]Faculdade Atenas, Sorriso MT, Brazil.

Correspondence: Maria Eduarda Vieira Fiber; Email: mdudavieira1@hotmail.com.

Case presentation: Female patient, 28 years old, with refractory dermatopolymyositis (DPM) taking prednisone, methotrexate, and azathioprine. She received an intravenous immunoglobulin infusion in August 2022, with paradoxical worsening observed in December 2022, manifested by the accentuation of pain, myasthenia, anasarca, nausea, and lipothymia. In June 2024, the patient presented with Toxoplasma gondii infection, with ocular involvement and bilateral amaurosis. She developed persistent fever, right hemiparesis, and ipsilateral rhyme deviation. The cranial tomography showed a hypodense lesion in the region of the capsular nucleus of the right cerebral hemisphere, midbrain, and medulla oblongata, with a hyperdense focus in between, associated with a mild mass effect, which was poorly absorbing to the contrast medium. There were no abnormalities on physical examination. Given the clinical picture and imaging findings, she was diagnosed with neurotoxoplasmosis. The treatment included sulfadiazine, pyrimethamine, and folinic acid. **Discussion:** DPM is a rare autoimmune disease belonging to the group of inflammatory myopathies. Treatment involves using potent immunosuppressants, such as glucocorticoids used by the patient in the reported case. Prolonged use of these immunosuppressants may predispose patients to the development of serious opportunistic infections, such as toxoplasmosis. Toxoplasma gondii infection in immunosuppressed patients can lead to serious complications, including neurotoxoplasmosis (NTX), which is the main cause of lesions with mass effect on the central nervous system (CNS), responsible for 50 to 70% of these cases. NTX is commonly found in individuals with the human immunodeficiency virus (HIV), unlike the case reported, where the possible cause of the infection was the treatment with multiple immunosuppressants. It progresses with focal brain lesions with mass effect, generating several sequelae that appear depending on the topography and number of brain lesions. Clinically, it presents as headache, fever, motor impairment, and focal neurological deficits. In this case, the patient presented right hemiparesis and lip rhyme deviation, symptoms suggestive of a focal brain lesion. The diagnosis of NTX is suggested by neuroimaging, which shows hypodense or hypointense lesions with ring enhancement after contrast administration, consistent with the patient's presentation. Standard treatment includes sulfadiazine and pyrimethamine, combined with folinic acid to prevent bone marrow

toxicity caused by pyrimethamine. **Final comments:** Opportunistic infections, such as neurotoxoplasmosis, represent serious challenges in immunosuppressed patients. Early diagnosis and appropriate treatment are crucial to improving clinical outcomes. This case highlights the importance of surveillance and integrated management in patients with complex autoimmune diseases and prolonged immunosuppressive therapy.

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Neurotuberculosis in a patient treated with rituximab for anti-synthetase syndrome

Geovane Brito Afonso[1], Maria Júlia Tabosa de Carvalho Galvão[1], Vitor Gustavo Leão Souto[1], Túlio Marlus Castro Lucena[2], José Alves da Costa Neto[1], Inah Araujo de Almeida Murta[1], Érika Santos Machado[2], Assis Porfirio Furtado Nogueira[2], Fernando Tenorio Gameleira[1]

[1]Universidade Federal de Alagoas, Hospital Universitário Professor Alberto Antunes, Maceió AL, Brazil.
[2]Hospital Geral do Estado, Secretaria de Estado da Saúde, Maceió AL, Brazil.

Correspondence: Maria Júlia Tabosa de Carvalho Galvão; Email: mariajuliatcgalvao@gmail.com.

Case presentation: A 48-year-old previously healthy woman began experiencing myalgia in 2016, associated with weakness in all four limbs, intermittent dysphagia, and Raynaud's phenomenon. Elevated CPK and aldolase levels, along with positive Anti-Jo1 and Anti-RO antibodies, and electromyography indicating muscle disease, supported the diagnosis of Anti-synthetase Syndrome. In 2019, she started treatment with rituximab for 2 years but subsequently developed interstitial lung disease and pulmonary tuberculosis. After treating tuberculosis, her condition stabilized clinically. In 2020, she developed a confusional syndrome and insidious cognitive deficit over 3 months. Cerebrospinal fluid analysis showed elevated protein levels, but rapid molecular tests and cultures were negative for tuberculosis. Brain MRI revealed right periventricular ring enhancement, suggestive of an abscess or cerebral tuberculoma. She underwent treatment for neurotuberculosis for 12 months, and post-treatment brain MRI showed diffuse brain volume reduction and gliosis with ischemic lacunae in both cerebral hemispheres. A new immunosuppressive regimen was initiated with prednisone 20 mg/day, methotrexate 10 mg/week, and monthly immunoglobulin. In 2024, she experienced generalized tonic-clonic seizures of likely structural origin and physical examination revealed bilateral palatal paresis, global muscle atrophy, abasia, and flaccid tetraparesis. The most recent imaging showed diffuse supratentorial hypoattenuating foci, diffuse brain atrophy, and ex-vacuo hydrocephalus. **Discussion:** Anti-synthetase syndrome is a type of idiopathic inflammatory myopathy that may progress with myositis, interstitial lung disease, arthritis, fever, and Raynaud's phenomenon. Anti-Jo1 is the most prevalent antibody. Treatment involves immunosuppressants, and patients using Anti-TNF-alpha agents like rituximab are at increased risk of developing tuberculosis. Neurotuberculosis, in the absence of other symptoms, can emerge as a rare infection in these patients, accounting for 5-10% of total cases. It progresses rapidly or insidiously and in its most common manifestation, tuberculous meningitis, can lead to complications such as hydrocephalus, stroke, tuberculoma, and cranial nerve involvement. More rarely, as reported in this case, some patients may present with tuberculoma without meningitis and with normal or slightly altered cerebrospinal fluid. Many patients are diagnosed at more advanced stages of the disease due to its paucibacillary nature. **Final comments:** Diagnosing neurotuberculosis in immunosuppressed patients undergoing treatment for Anti-synthetase syndrome is challenging and must be addressed to prevent the development of disabling neurological sequelae or fatal outcomes during the patients' productive years.

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Neurotuberculosis in Brazil: an analysis of the epidemiological profile related to the effectiveness of control strategies (2013-2023)

Vinícius Gabriel Nunes de Freitas[1], Juliana Alencar Isacksson Vieira[1], Arthur Andrade da Silva[1], Willian Alves Costa[1], Paulo de Oliveira Neto[1], Eliezer Paulo Ferreira Júnior[1], Thiago da Costa Brito[2]

[1]Universidade Federal do Amapá, Macapá AP, Brazil.
[2]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Juliana Alencar Isacksson Vieira; Email: julianaisacksson17@gmail.com.

Background: Tuberculosis (TB) is one of the oldest and most prevalent infectious díspares in history and primarily affects the lung tissue. However, when it invades the nervous system, it is called neurotuberculosis (NT). Therefore, the disease is a concern for healthcare services, once it has a difficult diagnosis and severe prognosis. Tuberculosis prevention and epidemiologic control are extremely important to public Healthcare system, which has measures against the disease like BCG vaccine and the National Program of Tuberculosis Control. Thus, understanding the epidemiological profile becomes in the context of tuberculosis control strategies. **Objective:** To describe the evolution of epidemiological and evaluate the effectiveness of brazilian neurotuberculosis control strategies in the period of 2013 until 2023. **Methods:** Data available from the Unified Health System Informatics Department and the Hospital Information System from 2013 to 2023 were used to conduct thus cross sectional descriptive study. The following informations were extracted: Mortality rate and absolute deaths specified by age group, sex, region of the country and year of notification. **Results:** Total number of deaths was 340. The Southeast Region was the most affected in absolute numbers, with 50% of the total (n=170) and mortality rate of 12.57. However, the North Region with 11.31 has the second highest mortality rate. Among the states from the region, São Paulo has the highest number of notifications, with 60,6% of the total (n=103). 2022 was the year with highest number of cases with 13,8% (n=47). In addition, the most affected age group was the one of 30-39 years old, with 20.6% of the cases (n=70), the male group has the highest number of cases with 64.4% (n=219) and the highest mortality rate prevail in the female group, with 11.1. The brown population has a higher number of deaths, 41.2% (n=140), in terms of mortality rate, indigenous people have the highest value with 13.33, followed by black people with 12.21. However, the data describing race/color have a large number of patients with no information, 23.5% (n=80). **Conclusion:** The epidemiological profile of neurotuberculosis in the period of time analyzed shows an relative increase in the number of deaths, with the peak in 2022. The Southeast Region is the one with highest absolute numbers, specially the state of São Paulo. Still, the northern region presents the second highest mortality rate despite its small population size. Thus, the data demonstrate the vulnerability of these populations, which are marginalized in healthcare services. The data also show that control strategies have been effective in reducing the incidence of neurotuberculosis cases in recent years, with an urgent need for greater focus on the most affected racial/ethnic groups and regions of the country.

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Neurotuberculosis in immunocompetent patients: atypical manifestations of a common disease

Maria Eduarda Slhessarenko Fraife Barreto[1], Gabriela Rodrigues Tomaz[1], Lucas Toshio Uenishi[1], Guilherme Antonio Silva Ribeiro[1], Lara Santos Machado[1], Luiz José Malta Gaia Ferreira Júnior[1], Enedina Maria Lobato de Oliveira[1], Henrique Ballalai Ferraz[1], Carolina Candeias da Silva[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Eduarda Slhessarenko Fraife Barreto; Email: madusfbarreto@gmail.com.

Case presentation: Case 1: A 41-year-old previously healthy man presented with acute behavioral change, mental confusion, and persistent fever during 3 days. He denied cough or weight loss but had a history of a car accident two years prior. On admission, he was inattentive, with right hemiparesis, and nuchal rigidity. Cranial CT revealed acute hydrocephalus and an urgent ventricular drainage was performed. Cerebrospinal fluid (CSF) analysis showed lymphomonocytic pleocytosis, low glucose, and elevated protein. CSF cultures, molecular tests for Mycobacterium tuberculosis and Histoplasma were negative, as were serologies for HIV, syphilis, and hepatitis. Chest CT was unremarkable. Empirical treatment with broad-spectrum antibiotics + tuberculosis regimen (RIPE) was initiated. Brain MRI demonstrated a left frontal heterogeneous hyperintensity lesion with irregular contrast enhancement. Clinically, he exhibited mild, progressive improvement. After 6 weeks, CSF culture confirmed Mycobacterium tuberculosis. He was discharged with a 1-year treatment plan. Case 2: A 28-year-old man developed continuous bilateral eyelid and lip myoclonus during three months. He had a history of adequately treated miliary tuberculosis five years ago. Brain MRI revealed multiple ring-enhancing nodular lesions with surrounding vasogenic edema in various brain regions. Some lesions exhibited a tree-in-bud appearance, with leptomeningeal enhancement. Laboratory investigations were unremarkable for immunodeficiency, and electroencephalogram was normal. Brain biopsy confirmed granuloma with molecular testing positive for Mycobacterium tuberculosis. The patient was discharged under neurotuberculosis treatment, with some radiological reduction in lesions noted, although involuntary movements persisted. **Discussion:** These two cases show atypical manifestations of Neurotuberculosis. Case 1 is remarkable due to an acute history, more compatible with bacterial agents. Case 2 presented with unusual cranial movement disorders. Neurotuberculosis typically presents as subacute/chronic meningitis with cranial nerve involvement, and may progress to hydrocephalus. Movement disorders are an unusual presentation. Usually, it disseminates through hematogeneous or lymphatic routes from another site. CSF often shows lymphomonocytic pleocytosis, decreased glucose, elevated adenosine deaminase (ADA), and specific microbiological tests. Direct bacillus detection is positive in 25-30% of culture-confirmed cases in the literature. Mycobacterial culture has <50% positivity in larger cohorts. Diagnosis can be reasonably made based on clinical data, CSF profile, and evidence of tuberculosis elsewhere, even if culture is negative. **Final comments:** CNS tuberculosis has a broad spectrum of clinical presentations. Definitive diagnosis is challenging and may require biopsy. Early empirical treatment should be started for suspected cases, after other diagnosis have been ruled out, particularly in low income countries, like Brazil.

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New generation sequencing in Hyper-CKemia evaluation

Lucas Marenga de Arruda Buarque[1], Nicolas Ruan dos Santos Cavalcante[1], Jose Pedro Soares Baima[1], Ian Felipe Barbosa Souza[1], Felipe Fiorelli Valenzuela[1], Andre Macedo Serafim da Silva[1], Rodrigo de Holanda Mendonça[1], Carlos Otto Heise[1], Cristiane de Araujo Martins Moreno[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Lucas Marenga de Arruda Buarque; Email: l.marengabuarque@gmail.com.

Background: Hyper-CKemia is a common complaint in neuromuscular clinics. Which is the best propaedeutic for its evaluation is still a matter of discussion. **Objective:** We describe the results of using new generation sequencing in patients with hyper-CKemia. **Methods:** All genetics tests made in the neuromuscular disorders division of HCFMUSP were revisited and only those with a history of rhabdomyolysis and hyperCKemia pauci- or asymptomatic were included. We considered hyper-CKemia as a creatine phosphokinase (CPK) level above 1,5 times above reference value. **Results:** Twenty three patients were selected, 21 were submitted to a gene panel for neuromuscular diseases and 2 to exome: 03 with rhabdomyolysis, 06 with asymptomatic hyper-CKemia and 14 with paucisymptomatic hyper-CKemia. Six (26%) were diagnosed by genetic test: 03 ANO5, 01 GPMPPB, 01 PYMG (all of which have paucisymptomatic hiper-CKemia) and 01 RYR1 (rhabdomyolysis). Of those 17 who did not reach a diagnosis, 01 had only one pathogenic variant in ANO5 gene (a recessive condition) and 04 had one or more VUS in genetic tests. The mean maximum CPK levels in the group that reached a diagnosis was 6.977U/L and in the group without genetic diagnosis was 5.851 U/L. **Conclusion:** The new generation sequencing has started a new era in neuromuscular disorders diagnostics. We report a efficacy of 26% of its use in hyper-CKemia evaluation and ANO5 gene was the most prevalent.

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New headache in heart transplant patient receiving tacrolimus

Renata Gomes Londero[1]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Renata Gomes Londero; Email: renataneuro-abn@yahoo.com.br.

Case presentation: We saw a 51-year-old patient, heart transplant recipient (October 2022), with recurrent headache since the transplant, which changed pattern at the end of September 2023. Patient wakes up at the end of the night or in the morning with a headache that is more intense than the previous one, throbbing, periorbital, with mild phonophobia and photophobia and no nausea. Headache improves spontaneously in the late morning or early afternoon. He also presents episodes of flashes in his vision. A new pattern emerged one week after a 50% increase in the previous dose of tacrolimus. Fundus of eye with bilateral grade 3 papilledema. Skull tomography with angiotomography showed left frontal ischemic sequelae, venous sinuses and patent superficial and deep veins. Brain MRI was unremarkable. Liquor with opening pressure 260mmH20, crystalline appearance, 1 leukocytes, proteins 45.9, glucose 58 (glycemia 110) | viral, bacteriological, bacterioscopic panel, PCR for mycobacterium tuberculosis, Syphilis, negative fungi and Cryptococcus research. He had a normal renal function, BMI of 21 and has not gained weight in the last year. The patient was managed with acetazolamide up to 1.5g and bilateral occipital nerve block (with good headache control). The papilledema was resolved within 1 month of his initial visit. As the patient has a long history of rejection, we did the option to not suspend the tacrolimus until now. But it is still in discussion. **Discussion:** Tacrolimus, a calcineurin inhibitor, is an immunosuppressant used globally to prevent rejection after organ transplantation. Posterior Reversible Encephalopathy syndrome, psychosis, intracranial hemorrhage has already been described as side effects of tacrolimus. We saw in literature one case of idiopathic intracranial hypertension (IIH), in a child submitted to a kidney transplantation and receiving tacrolimus. The elevated intracranial pressure and the response to acetazolamide makes us believe that it's one more case of IIH by tacrolimus. **Final comments:** With the growing number of people receiving transplantation it's very important to know the side effects of the immunosuppressant drugs used to manage those patients. Here we report a case of a 51 years old man, with ischemic cardiomyopathy that received a heart transplant and developed IHH secondary of tacrolimus, with good response to acetazolamide treatment.

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New-onset refractory status epilepticus (NORSE) after COVID-19 in a young adult with possible association of steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT)

Lorenzo Marinho Morais[1], Emanuel de Assis Bertulino Martins Gomes[2], Chiara Gübel Portugal[1], Antônio Alves Sobreira Neto[1], Samuel Cavalcante Marinho[1], Jose Wagner Leonel Tavares Junior[2], Manoel Alves Sobreira Neto[2], Samir Câmara Magalhães[2]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.
[2]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Lorenzo Marinho Morais; Email: lorenzo.morais@aluno.uece.br.

Case presentation: A 32-year-old previously healthy male patient presented low-grade fever and myalgias for 4 days. These symptoms subsided, but a week later, he presented persistent seizures even after initial drugs (diazepam and phenytoin), requiring the use of midazolam and orotracheal intubation. After sedation, the seizures returned with refractory status epilepticus. He underwent imaging and laboratory tests and lumbar puncture. The investigation only showed a positive test for COVID-19. He was transferred to the intensive care unit and sedated with midazolam and propofol. He underwent electroencephalography (EEG) monitoring, which still showed epileptiform activity. Levetiracetam was started with sedative drugs increased to achieve a suppression pattern. As it was a NORSE case, intravenous immunoglobulin (IVIG) and methylprednisolone (MPIV) were done for 5 days. EEG still disclosed epileptiform activity. A new burst-suppression was attempted for 48 hours, with association of ketamine, midazolam and propofol. The dose of Levetiracetam was increased, with clobazam and topiramate included. 30 days after admission, the patient woke up without seizures, with normal EEG activity and with rapid improvement in the level of consciousness. In the meantime, he received the results of some tests, with anti-tireoperoxidase and anti-thyroglobulin autoantibodies positive, in addition to anti-SSA. The presence of thyroid autoantibodies, in addition to fast clinical response after steroids, led us to raise the hypothesis of SREAT. **Discussion:** Physiopathology of these seizures due to COVID-19 could be the result of neuroinflammation, autoimmune response and viral invasion of the central nervous system (CNS). As far as we know, this is the first report of possible SREAT triggered by Covid-19. This is the first reported case of COVID-19 associated with NORSE in Brazil. Thyroid autoantibodies raised the hypothesis of SREAT. The treatment involves immunotherapy with methylprednisolone and/or immunoglobulin. The association between Covid-19 and SREAT may be due to the autoimmunity triggered by SARS-CoV-2. NORSE treatment involves antiseizure medication and anesthetic drugs. In addition, immunotherapy should always be considered. The present case showed improvement after the addition of ketamine to propofol and midazolam aiming at suppression burst with inter-surge intervals longer than 5 seconds, for 48 hours and after immunotherapy with IVIG and MPIV. This approach was very effective in our present case and the patient was discharged without sequels. **Final comments:** Here we present a case report of a patient who developed NORSE after COVID-19 with a possible SREAT. The patient showed a good therapeutic response to immunotherapy and improved the status epilepticus after burst suppression therapy. The present case serves as a basis for possible future similar cases and demonstrates the importance of correct investigation and early treatment.

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NFL levels significantly decrease in response to treatment with patisiran or vutrisiran in hattr amyloidosis with polyneuropathy

Michael Polydefkis[1], Emre Aldinc[2], Hans Nienhuis[3], Chafic Karam[4] , Ajroud-Driss[5], Yoshiki Sekijima[6], Marcia Waddington-Cruz[7], Jennifer Barnes[2], Paul Nioi[2], Simina Ticau[2]

[1]Johns Hopkins University, School of Medicine, Baltimore, MD, USA.
[2]Alnylam Pharmaceuticals, Cambridge, MA, USA.
[3]University of Groningen, University Medical Center Groningen Groningen, The Netherlands.
[4]University of Pennsylvania, Philadelphia, PA, USA.
[5]Northwestern University, Feinberg School of Medicine, Chicago, IL, USA.
[6]Shinshu University, School of Medicine, Asahi, Matsumoto, Japan.
[7]Universidade Federal do Rio de Janeiro, Centro de Paramiloidose Antônio Rodrigues de Mello, Rio de Janeiro RJ, Brazil.

Correspondence: Ana Elisa Rodrigues Marangon; Email: elisarois@yahoo.com.br.

Background: hATTR amyloidosis is a rare, progressive, and fatal disease. NfL is a potential biomarker of disease progression and treatment response in patients with hATTR amyloidosis with polyneuropathy. **Objective:** Examine neurofilament light chain (NfL) levels in patients with hereditary transthyretin-mediated (hATTR) amyloidosis with polyneuropathy from the Phase 3 APOLLO and HELIOS-A studies to assess the potential utility of NfL as a biomarker of disease progression. **Methods:** NfL levels were measured using the Quanterix® SimoaTM platform in healthy controls and patients with hATTR amyloidosis with polyneuropathy who participated in the APOLLO or HELIOS-A studies, gave consent, and had sufficient samples. The samples analyzed were from baseline, 21 days, 4 months, and 18 months in APOLLO, and baseline, 43 days, 4 months, 9 months, and 18 months in HELIOS-A. **Results:** At baseline, NfL levels were slightly higher in APOLLO than in HELIOS-A (69.4pg/mL and 58.2pg/mL, respectively) and did not differ significantly between treatment groups within each study. In the APOLLO placebo arm, NfL levels significantly increased vs baseline at 4 months (+19.0pg/mL, p<0.001) and at 18 months (+36.3pg/mL, p<0.001). In the APOLLO patisiran arm, NfL levels decreased significantly at 4 months and 18 months vs baseline (-20.4 pg/mL and -23.2pg/mL respectively; p<0.001 for both). In HELIOS-A, NfL levels in patisiran and vutrisiran groups significantly decreased vs baseline at 4 months (-9.7pg/mL and -11.0pg/mL, respectively; p<0.05 for both) and at 18 months (-16.4pg/mL and -19.9pg/mL, respectively; p<0.001 for both). **Conclusion:** NfL levels significantly decreased from baseline with patisiran and vutrisiran as early as 4 months after treatment initiation and decreased levels were maintained through 18 months. In contrast, NfL levels in untreated patients increased significantly. These results suggest that NfL is potentially useful for monitoring treatment response and disease progression in patients with hATTR amyloidosis with polyneuropathy.

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Niemann-Pick type C syndrome, epilepsy and obstructive sleep apnea in a pediatric patient: a case report

Helmut Kennedy Azevedo do Patrocínio[1], Tereza Cristina Batista Dias[1], Tiago Lameque de Sousa e Silva[1], Pedro Henrick Guimarães Carvalho[1], Cláudio de Souza Fernandes Júnior[1], Cláudio de Souza Fernandes[2], Fabricia dos Santos Almeida[1], Lucca Ferdinando Queiroz Fernandes[1], Matheus Araújo de Medeiros[1], Iury Hélder Santos Dantas[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[2]Universidade do Estado do Rio Grande do Norte, Mossoró RN, Brazil.

Correspondence: Helmut Kennedy Azevedo do Patrocinio; Email: hell.kenn@gmail.com.

Female patient, 4 years old, diagnosed with Niemann-Pick type C (NPC), Epilepsy, Severe Mental Retardation and Obstructive Sleep Apnea (OSA). The patient began presenting with splenomegaly at 10 months old. At 1.5 years, neurodevelopmental regression, especially in language and motor skills, was noted, which raised alarms for the investigation of neurodegenerative diseases. At 2 years and 2 months, NPC was diagnosed through the measurement of Chitotriosidase, Lyso-sphingomyelin, and clinical correlation; genetic studies were not performed. Six months later, the patient developed severe dysphagia, generalized hypotonia, inability to sustain head control, and lost her communicative ability, becoming bedridden and fully dependent. Furthermore, the presentation of OSA and epileptic seizures add another layer of complexity to the clinical picture. Despite treatment with anticonvulsant medications, the epileptic seizures persisted, impacting her quality of life. The patient's treatment involves a multidisciplinary approach, including anticonvulsant medications, continuous use of CPAP to correct problems related to OSA, and also Miglustat to reduce the progression of neurological deficits caused by Niemann-Pick disease. Niemann-Pick type C disease is an autosomal recessive disorder, which results in impaired transport of LDL and other macromolecules. It can present from the perinatal period to late adulthood. Clinical manifestations are highly heterogeneous, typically including cerebellar ataxia, vertical gaze palsy, cataplexy, dystonia, dysarthria, dysphagia, epileptic seizures, cognitive impairment, and hepatosplenomegaly. The patient is classified in the early infantile period of NPC. She exhibited isolated splenomegaly for months, associated with neurocognitive and psychomotor impairments classic to the disease, which guided the diagnosis. Additionally, she developed epileptic seizures refractory to standard treatment. Severe epilepsy is associated with a worse prognosis and significantly reduces patients' life expectancy and quality of life. OSA stands out in this patient as a non-motor symptom of NPC. It is believed that OSA associated with NPC results from impaired coordination and reduced tone of the respiratory muscles, despite limited research in this area, it is known to be associated with higher morbidity and mortality, as well as greater cognitive deficits. The patient's treatment is focused on improving her current quality of life and slowing the progression of neurological deficits, as NPC does not have a curative treatment. This case provides important information about NPC, a rare and complex medical condition, in a young patient. Sharing this case is essential to increase understanding about this condition, its symptoms, treatments, and prognoses. It also aims to spark scientific interest for future research related to correlations among the presented diseases to enhance comprehension and treatment of this condition.

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NOACs versus antiplatelet agents in ESUS with predictors of atrial fibrillation: a systematic review with meta-analysis

Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Luma Rodrigues da Silva[1], Ana Paula Lima Nascimento[1], Ester Vasconcelos Silva[1], Sabrina Ruthiele Santos de Carvalho[1], Samuel de Castro Campos[1], Lara Beatriz Alves Batista[1], José Henrique de Melo Feitosa[1], Pedro Henrique Ximenes Ramalho Barros[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Luma Rodrigues da Silva; Email: lumarodrigues@ufpi.edu.br.

Background: Embolic strokes of undetermined source (ESUS) account for approximately 17% of ischemic stroke cases. They are often considered to be caused by occult cardiac embolism from atrial cardiopathy, even without atrial fibrillation (AF). Thus, certain predictors of AF may determine potential advantages of anticoagulation with new oral anticoagulants (NOACs) over antiplatelet therapy. **Objective:** To compare the efficacy of NOACs with antiplatelet agents in relation to clinical outcomes in patients with ESUS and predictors of AF. **Methods:** A search was conducted in PubMed, Embase, and Cochrane Library up to January 2024, including randomized controlled trials (RCTs) and observational studies involving patients with ESUS exhibiting any predictor of AF. The study compared groups treated with anticoagulation versus antiplatelet therapy with aspirin, emphasizing the outcomes of stroke recurrence, major hemorrhagic events, and mortality. Statistical analysis was performed using R Studio 4.3.2 software. Heterogeneity was assessed using the I^2 method. **Results:** Three RCTs and one observational study were included, totaling 8,557 patients. Stroke recurrence showed a tendency to favor NOACs (RR=0.90; 95% CI 0.72 - 1.11, I^2=0%, p=0.32), although without statistical significance. No significant differences in major hemorrhagic events were observed between the groups (RR=1.02; 95% CI 0.33 - 3.14, I^2=0%, p=0.98), nor was there any disparity in mortality (RR=1.02; 95% CI 0.33 - 3.14, I^2=0%, p=0.54). **Conclusion:** Comparing NOACs with antiplatelet agents, NOACs did not demonstrate significant clinical superiority in patients with ESUS and predictors of AF.

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Nonconvulsive status epilepticus mimicking acute cerebrovascular syndrome: a case report

Ana Paula de Góes Louly Bustamante[1], Rodrigo Fernandes Cury[1], Regina Maria França Fernandes[1], Amanda Maieski[1], Caroline Darold Vieira[1], Gilberto Bento Magioni Junior[1], Lucas Fernando Chicheto Brancaglião[1], Luciano da Silva Pontes[1], Nadja Layane Gomes Santiago[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.

Correspondence: Ana Paula de Góes Louly Bustamante; Email: anapaulalouly@hotmail.com.

Case presentation: A.P.O.L., a 35-year-old female inpatient with chronic kidney disease, had cefepime recently introduced to treat a catheter-related bloodstream infection. One day before hemodialysis session, she suddenly presented with left hemiparesis, dysarthria and altered mental status. She was immediately submitted to a contrast-enhanced brain CT and CT angiography, neither of which showed signs of ischemia, hemorrhage or large vessel occlusion. Brain MRI was subsequently performed, and no areas of restricted diffusion were found. An electroencephalogram (EEG) was requested for differential diagnosis and it showed an asymmetric disorganized background with highly prevalent short bursts of lateralized intermittent rhythmic delta activity (LRDA) involving the left frontal region, lasting up to 10 seconds, with a frequency between 2.0 and 2.5 Hz, with superimposed sharp theta waves and no spatiotemporal evolution or time-locked clinical manifestations. Although not strictly falling into the ictal/interictal continuum (IIC) by definition (ref.), we hypothesized it was a case of possible nonconvulsive status epilepticus (NCSE). A therapeutic trial with 10mg/kg intravenous phenytoin was then carried-out, followed by significant clinical and electroencephalographic improvement, with complete recovery from previous neurologic deficits, suppression of the LRDA and appearance of a distinguishable posterior dominant rhythm. **Discussion:** In the context of impaired awareness with clinical/radiological dissociation, the presence of electroencephalographic rhythmic/periodic patterns (RPPs) with a frequency ≥0.5 Hz and up to 2.5 Hz could be considered as a possible NCSE/IIC. The criteria for IIC include: any periodic discharge or sharp wave >1.0 Hz and ≤2.5 Hz over 10 seconds; or any periodic discharge or sharp wave ≥0.5 Hz and ≤1.0 Hz over 10 seconds, with a "plus" modifier or fluctuation; or any LRDA >1.0 Hz over 10 seconds, with a "plus" modifier or fluctuation. Whether or not these criteria are strictly met, but clinical suspicion remains high, as was our case, a trial with intravenous anti-seizure medication (ASM) could be considered. Most cases of NCSE present with impairment of consciousness, and when motor symptoms are present, they are usually positive, such as subtle myoclonus or nystagmus. The hallmark of this case, however, were focal negative symptoms, including hemiparesis, resembling an acute stroke syndrome. **Final comments:** The main objective of this report is to alert clinicians for epilepsy-related etiology within the differential diagnosis of sudden onset neurologic deficits. This should be specially considered when the patient presents with altered mental status in the context of clinical/radiological dissociation. In some selected cases, even when EEG criteria for possible NCSE are not strictly met, a therapeutic trial with ASMs could be considered, depending on clinical suspicion.

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Non-epileptic seizures in vertebrobasilar occlusion: a case report

José Eduardo Trentim Longo[1], Ana Flávia Andrade Lemos[1], Gabriel Pinheiro Modolo[1], Rodrigo Bazan[1], Daniel Fabiano Barbosa Santos[1], Pedro Machry Pozzobon[1], Ana Beatriz Marangoni Baston[1]

[1]Universidade Estadual Paulista, Faculdade de Medicina de Botucatu, Botucatu SP, Brazil.

Correspondence: José Eduardo Trentim Longo; Email: jose.longo@unesp.br.

Case presentation: An 81-year-old female patient presented with sphincter release, decreased level of consciousness, left homonymous hemianopsia, and dysmetria, with symptoms persisting for over 24 hours. Brain CT and neck and brain angiography showed hypodensity in the right cerebellum and occipital lobe, and occlusion of the right distal vertebral artery. While in the stroke unit, the patient showed improvement in deficits but then experienced decreased level of consciousness and inexhaustible multidirectional nystagmus. Subsequent imaging showed improved flow in the vertebral artery. The patient maintained only hemianopsia but developed stereotyped limb movements. Repeated imaging identified artery patency. Etiological investigation revealed no epileptiform disturbances on EEG, and brain MRI demonstrated dissection and intramural hematoma in the vertebral artery with preserved flow in the 3D "time-of- flight" technique. Considering the sequence of events and the absence of epilepsy history, we hypothesize that brainstem hypoperfusion related to vertebrobasilar occlusion led to non-epileptic seizures. The interruption of inhibitory projections from the cortex to the medulla or brainstem is one proposed mechanism. Dual antiplatelet therapy was initiated as prophylaxis. **Discussion:** Vertebrobasilar occlusion can present in a manner indistinguishable from an epileptic seizure. If this indeed represents a true seizure, the mechanism may be related to ischemia of the occipital cortex or activation of subcortical epilepsy

generators, necessitating further studies on the pathophysiology. Atypical epilepsy presentations, such as basilar occlusion, represent a potentially fatal condition that can be treated if diagnosed in time. **Final comments:** This case highlights the diagnostic challenges and significant morbidity associated with posterior circulation ischemic strokes. The heterogeneity of signs and symptoms, such as decreased consciousness, cranial nerve abnormalities, tetraplegia, and locked-in syndrome, complicates the diagnosis and contributes to high morbidity and mortality rates. Understanding the nuances and mechanisms of atypical presentations, such as non-epileptic seizures due to vertebrobasilar occlusion, is crucial for timely and effective management.

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Non-motor and extracerebellar features in spinocerebellar ataxia type 6

Breno Kazuo Massuyama[1], Marcondes Cavalcante França Junior[2], Orlando Graziani Povoas Barsottini[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Breno Kazuo Massuyama; Email: breno.massuyama@gmail.com.

Background: Spinocerebellar ataxia type 6 (SCA6) is an autosomal dominant degenerative disease. SCA6 is described as the prototype of pure cerebellar ataxia. However, the calcium receptor subunit affected in SCA6 appears to be ubiquitous in neurons throughout the brain. **Objective:** To access the non-motor symptoms and extra-cerebellar signs of SCA6 patients. **Methods:** Eighteen clinically and molecularly proven SCA6 patients agreed to participate in this study. Our Institutional Ethics Committee approved this study. All patients provided written informed consent. To evaluate ataxia severity, patients were evaluated by the "Scale for the Assessment and Rate of Ataxia" (SARA) and "International Cooperative Ataxia Rating Scale". For evaluation of non-motor and extracerebellar features severity, we applied "Inventory of Non-ataxia symptoms" (INAS), REM Sleep Behavior Screening Questionnaire (RBDSQ), Epworth Sleepiness Scale (ESS), and Fatigue Severity Scale (FSS). INAS Count is a semiquantitative variable of extracerebellar involvement in SCA. RBDSQ is an n-point scale where values greater than five or more questions indicate RBD. ESS is an n-point scale where a cut-off of ten or greater was set for the diagnosis of excessive daytime sleepiness (EDS). FSS is an n-point scale where values greater than twenty-eight are indicators of the presence of fatigue. We performed Spearman's correlation test to calculate the strength of association between CAG repeat length ((CAG)n) and each of the following variables: SARA, ICARS, INAS Count, FSS, RBDSQ, and age at onset of symptoms (AOO). **Results:** Regarding the clinical characteristics of the patients with SCA6, disease duration in years (mean=13; SD=7.33), AOO (mean=53.33; SD=9.14), SARA (mean=13.30; SD=6.71), ICARS (mean=37; SD=17.13), INAS Count (mean=1.77; SD=1.06), FSS (mean-33.52; SD=13.23), RBDSQ (mean=4; SD=1.96), and ESS (mean=5.94; SD=4.00). Restless legs syndrome (RLS) was present in 28.57% (N= 4 out of 14), RBD was present in 23.52% (N= 4 out 17), and EDS was present in 23.52% (N= 4 out of 17). Fatigue was present in 64.70% (N= 11 out of 17). The (CAG)n at the expanded allele was strongly correlated with RBDSQ (cc:-0.609; p=0.046). In addition, (CAG)n was correlated with SARA (cc:0.300; p=0.342), ICARS (cc:0.325; p=0.301), INAS Count (cc:0.267; p=0.401), FSS (cc:-0.386; p=0.240) and AOO (cc:-0.226; p=0.479). We observed a high frequency of non-motor symptoms and other motor manifestation in SCA6 patients beyond cerebellar signs. SCA6 may represent a more widespread condition, affecting other regions of the nervous system besides the cerebellum and spinocerebellar pathways. RBD, RLS and EDS have been reported in SCA2 and SCA3. Considering that we observed comparable results in SCA6, it is feasible that similar pathophysiological mechanisms may also occur in SCA6. **Conclusion:** Our results support the possibility of revising the concept of SCA6 be a pure cerebellar disease.

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Non-motor epileptic seizures versus panic attacks: a diagnostic dilemma

Heron Alves Vale[1], Rraissa Josefa Pereira de Moura[1], Ana Caroline Paiva Simeao[1], Nereu Alves Lacerda[1], Ana Carolina Martins Fragoso Carneiro[1], Maria Luisa Roberto Lima Lisboa[1], Natália Nunes Tenório[1], Ana Beatriz Roberto Lima Borba[1], Laura Alícia Morais Lima Oliveira[2], Moema Peisino Pereira[1]

[1]Hospital Pelópidas Silveira, Recife PE, Brazil.
[2]Hospital Universitário Onofre Lopes, Natal RN, Brazil.

Correspondence: Heron Alves Vale; Email: heron_vale@hotmail.com.

Case presentation: A 69-year-old male patient with a prior diagnosis of major depressive disorder was admitted to the neurology emergency department, referred from psychiatry due to reports of transient perceptual behavioral crisis lasting less than 5 minutes over the past 6 months. Phenomenologically, he described these episodes as sudden sensations of "anguish" in the epigastric region, "cold/hot" sensations on his back, and a feeling of "imminent death", associated with sweating, tachycardia, and perioral and extremity tremors. These events progressed in intensity and frequency over three months, occurring up to four times a day. He experienced postictal amnesia and occasionally woke up at night with these symptoms. Concurrently, he demonstrated worsening of his pre-existing depressive symptoms, characterized by, anhedonia, frequent crying, avoidance behavior, and catastrophic thoughts. He sought help from several psychiatrists who considered these manifestations as psychiatric disorders, but treatment, including alprazolam 3mg/day, paroxetine 12.5mg, mirtazapine 45mg, clobazam 10mg, and aripiprazole 10mg, without improvement. On examination, he appeared generally well, conscious, oriented to person, place, and time, with a tearful facial expression consistent with his depressed mood. He had fluent speech, intact comprehension, repetition, and naming abilities. There were no motor or sensory deficits. Brain MRI showed only mild hippocampal rotation, and EEG revealed disorganized baseline activity. Further diagnostic evaluation included a SPECT scan to identify a possible epileptogenic functional lesion, although this had not yet been performed. Nevertheless, given the clinical context, a therapeutic trial with an anti-seizure medication (ASM), valproic acid, was initiated. The patient showed significant improvement with no seizures for weeks at present. **Discussion:** Epileptic seizures are characterized by transient signs and symptoms resulting from excessive and synchronous neuronal activity, with manifestations varying depending on the distribution of the discharges. Seizures originating in the temporal lobe typically present with focal onset and may manifest as sensory, autonomic, emotional, or purely subjective psychic phenomena such as nausea, malaise, fear, depression, anguish, déjà vu, jamais vu, forced thinking, gustatory, olfactory, auditory, somatosensory, and visual hallucinations. These seizure phenomenologies are commonly observed in adults with mesial temporal sclerosis (MTS), characterized by neuronal loss and gliosis primarily involving the hippocampus and amygdala. **Final comments:** Epileptic seizures can manifest in diverse and variable forms, ranging from typical motor presentations like tonic-clonic seizures to non-motor autonomic, sensory, emotional, or cognitive forms. Therefore, a thorough differential diagnosis with psychiatric disorders is essential.

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Non-myasthenic fluctuating muscle weakness

Ana Luísa Magalhães de Oliveira[1], Amanda Isabella de Souza[1], Gabriela Carneiro Neves de Carvalho[1], Sofia Silva Olsthoorn[1], Camilla Mannarino Calil[1], Danilo Jorge da Silva[1], Marcelo Maroco Cruzeiro[1]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Ana Luísa Magalhães de Oliveira; Email: analuisa.magalhaes@estudante.ufjf.br.

Case presentation: A 26-year-old woman with non-consanguineous parents began to experience fatigue and intermittent muscle weakness in her lower limbs (difficulty climbing stairs and standing up), weakness in her cervical region, and weakness in abducting her upper limbs at the age of 15. The weakness intensifies mainly after periods of food abuse, with the ingestion of fatty foods or foods containing milk. There was a progressive decline over 4 months, with a loss of 20 kg in 10 months, diarrhea (4 to 5 times a day), fatigue, dyspnea on medium exertion, pain in the right hypochondrium, progressive weakness, intense myalgia, postprandial fullness, and nausea relieved by vomiting. Grade 4 muscle strength was noted globally, with proximal predominance and global hyporeflexia, except for bilateral Achilles reflex arreflexia. Her brother died of fatty liver metaplasia with the same symptoms. The patient underwent a molecular study, identifying a pathogenic variant of the ETFDH gene. **Discussion:** Glutaric acidurias are part of organic acidurias and may be due to a deficiency of an enzyme in the mitochondria called glutaryl-CoA dehydrogenase (GCDH) (type I); a deficiency of one of the three molecules involved in electron transport in the mitochondria and in the metabolism of acetyl-CoA and glutaric acid: the alpha (ETFA gene) and beta (ETFB gene) subunits of electron transfer flavoprotein and electron transfer flavoprotein dehydrogenase (ETFDH gene) (type II); or glutaryl-CoA oxidase deficiency (type III). The patient has glutaric aciduria type II, also known as multiple acetyl-CoA Dehydrogenase Deficiency (MADD). This inheritance is autosomal recessive, causing impaired riboflavin transport. The genes involved are ETFDH, ETFA, and ETFB. The mutation in the ETFDH gene usually occurs in the late-onset form of the disease and has a myopathic phenotype, rarely causing metabolic acidosis. The most common clinical manifestations are muscle weakness, liver disease, and respiratory disorders. Family history and symptoms are characteristic. It is noteworthy that it occurs more often in men. Early diagnosis can reduce morbidity and mortality by reducing the risk of cardiovascular disease and hepatic steatosis. Treatment includes a low-protein, fatty acid-restricted, high-carbohydrate diet. Meals should be frequent, prolonged fasting should be avoided (this would activate mitochondrial ß-oxidation of fatty acids to obtain energy), and Riboflavin should be administered. **Final comments:** MADD causes impaired mitochondrial metabolism associated with the oxidation of fatty acids of autosomal recessive inheritance, whose diagnosis should be considered in the presence of breathing difficulties, liver problems, kidney and/or heart problems, hypoglycemia, and weakness. Early identification followed by treatment can potentially reduce morbidity and mortality.

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Non-oclusive mesenteric ischemia during the management of subarachnoid hemorrhage: case report

Márcio Assis Messias Filho[1] , Ana Flávia Amitti Alvarez[2], Nícolas de Sousa Belonato Terra[2], Raysa Fontes Martins[2], Georgia Piredda Fernandes[2]

[1]Hospital São José do Avaí, Itaperuna RJ, Brazil.
[2]Universidade Iguaçu, Itaperuna RJ, Brazil.

Correspondence: Raysa Fontes Martins; Email: 190062471@aluno.unig.edu.br.

Case presentation: A 32-year-old male patient, with moderate intellectual disability, was admitted to the emergency department in a comatose state for approximately 20 minutes (Hunt-Hess V), with third-party reports of severe headache for 3 days. Non-contrast computed tomography of the skull performed on admission showed hyperdense collections in brain grooves and cisterns, characterizing Subarachnoid Hemorrhage (Fisher IV). After diagnosis, the patient was treated according to SAH management protocols. Cerebral arteriography was performed showing the presence of saccular aneurysmal formation in the topography of the anterior communicating artery, and the aneurysm was embolized. Seven days after the ictus, there was a worsening of the hemodynamic state and shock, accompanied by abdominal distension and gastrointestinal tract failure demonstrated by an increase in gastric residue. Abdominal computed tomography was performed showing the dilated small intestine, with signs of intestinal pneumatosis, intestinal dilation, portal venous gas and aerobilia. Abdominal CT angiography was performed, showing the superior mesenteric artery with preserved caliber, contours and flows, without signs of thrombus that would justify occlusion. The diagnosis of Non-Occlusive Mesenteric Ischemia (NOMI) is then concluded. Over the next few days of hospitalization, the patient continued to show increasing clinical and radiological severity, associated with severe acidosis, shock refractory to treatment with vasoactive amines and volume replacement. He died on the 7th day of hospitalization. **Discussion:** Non-Occlusive Mesenteric Ischemia is associated with vasoconstriction of the mesenteric arteries, without evident occlusion, which causes intestinal hypoperfusion. NOMI as a complication of SAH is secondary to vasospasm, which generally occurs 3 to 14 days after aneurysm rupture. NOMI does not present characteristic symptoms, therefore the diagnosis usually occurs during the progression to a severe stage of the disease, the disease is associated with high mortality rates. The correlation of the diseases in question are barely described in the literature due to the rarity in which they present themselves, however, it has been concluded in previous research that one of the main preponderant factors for their prognostic establishment is how late their diagnostic finding and subsequent installation occur. therapy, with the speed of its execution being essential to minimize the deleterious effects of the disease. **Final comments:** Therefore, it is described how important the need for timely diagnosis of Non-occlusive Mesenteric Ischemia as a complication resulting from Subarachnoid Hemorrhage, not only for Neurosurgeons, but also for Intensive Care physicians, aiming for its identification and subsequent management quickly and effectively, expanding the patient's chances of recovery, as this complication is defined as atypical in current medical-scientific circles.

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Not all dropped heads are Parkinson's disease: an atypical manifestation of myasthenia gravis

Paulo Ricardo Sampaio de Melo[1], Brenda Francisca Cardoso Brito[1], Adriana Alves Rapp[1], Julia Cordeiro Braga[1], Camila Maria Barreto Ribeiro[1], Victor Xavier da Silva[1], Lucas Vergne Cheade Lins[1], Marcel Leal Ribeiro[1]

[1]Hospital Santa Izabel, Salvador BA, Brazil.

Correspondence: Brenda Francisca Cardoso Brito; Email: brendaf.cardosob@gmail.com.

Case presentation: Female, 78 years old, from Salvador, Bahia; Medical history: Parkinson disease with 8 years from the diagnosis, using the initial dose of levodopa/benserazide. Relatives described that, 15 days before the admission, the patient presented with a suddenly dropped head, nonspecific generalized discomfort and weakness leading to the inability to walk. At the admission, the patient presented dyspnea and worsening of the malaise, then submitted for orotracheal intubation due to respiratory insufficiency. In the following days, the patient proceeds with failure on the extubation process. Physical examination: cervical, proximal superior and inferior limbs flaccid muscular weakness, however with hyperreflexia, and cervical rigidity. Investigation: Electroneuromyography revealed neuromuscular junction dysfunction in proximal superior limb muscles and bulbar region with decremental pattern. Nonspecific rheumatologic tests, antinuclear antibody (ANA), perinuclear autoantibody (p-ANCA), and thyrotropin receptor antibody (TRAb), were positive. The antibody anti acetylcholine receptor is under analysis until the submission of this report. Diagnosis: probable myasthenia gravis. Treatment: intravenous immunoglobulin for five days, glucocorticoids, and pyridostigmine. Outcome: the patient experienced gradual and significant improvement in muscle strength and was removed from mechanical ventilation after the treatment. **Discussion:** Myasthenia gravis is an antibody mediated autoimmune disease that targets proteins of the postsynaptic membrane of the neuromuscular junction. The diagnosis is challenging, mainly in atypical cases. It is an uncommon condition that occurs in the early second and third decades in females, and in the sixth to eight decades in males. The typical clinical manifestations are ocular with ptosis and diplopia (50%), bulbar with dysarthria dysphagia, and expressionless appearance (15%), proximal limb weakness (<5%), isolated neck (uncommon), isolated respiratory (rare), and distal limb weakness (rare), usually followed by areflexia. Several steps can be performed in order to confirm this condition such as clinical (ice pack and pharmacological testing), serologic (acetylcholine receptor, MuSK, and LRP4 antibodies), and electrophysiologic testing. However, in most scenarios in Brazil, the standard tests are not covered by the public health or insurance services, so the patient must pay individually. These tests are very expensive, which can hamper the diagnosis. Different reports suggest an association of myasthenia gravis and neuromyelitis optica, autoimmune thyroid disease, systemic lupus erythematosus, and rheumatoid arthritis. **Final comments:** Efforts should be applied to disseminate knowledge about the prevalence, epidemiology, and clinical manifestations of myasthenia gravis and to enable the most important tests on the diagnosis, such as electroneuromyography and the autoantibodies involved in the disease.

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Novel ERLIN2 variant expands the phenotype of spastic paraplegia 18: a case report of four affected individuals from an isolated community of Northeastern Brazil

Guilherme Carvalho de Souza[1], Maria Carolina Malta[1], Mirele Raíssa Silva Santos[1], Marshall ítalo Barros Fontes[2], Juliana Lopes de Sousa Anjos[3], Diego Patrício Ribeiro[3], Fernando Kok[4], Thalita Figueiredo[1]

[1]Universidade Federal de Alagoas, Faculdade de Medicina, Setor de Genética Médica, Maceió AL, Brazil.
[2]Universidade Federal de Alagoas, Faculdade de Medicina, Hospital Universitário, Setor de Genética Médica, Serviço de Genética Clínica, Maceió AL, Brazil.
[3]Universidade Federal de Alagoas, Faculdade de Medicina, Hospital Universitário, Setor de Oftalmologia, Maceió AL, Brazil.
[4]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, Serviço de Neurologia Infantil, São Paulo SP, Brazil.

Correspondence: Guilherme Carvalho de Souza; Email: guicadesouza@gmail.com.

Case presentation: As part of a larger project investigating neurodevelopmental disorders in highly inbred regions of Northeastern Brazil, we evaluated four individuals from the same family (two siblings and their cousins), aged between 11 and 20 years, that presented early onset neuropsychomotor development delay, characterized by progressive spasticity, predominantly in the lower limbs, and lower limb muscle weakness. In addition, they had intellectual disability and speech impairment. We performed the genetic investigation, and the Whole Exome Sequencing (WES) showed a mutation with autosomal recessive inheritance in exon 8 of the ERLIN2 gene: c.47_48delinsAA, identified as the potentially deleterious variant, which was selected for segregation analysis through Sanger Sequencing involving both the affected and unaffected family members. A previously unreported homozygous variant in the ERLIN2 gene, associated with hereditary spastic paraplegia type 18 (SPG18), was identified in all four patients. Furthermore, all patients were diagnosed with childhood cataract, a condition that was not found in any other family member. **Discussion:** Hereditary spastic paraplegias (SPGs) are a group of neurodegenerative diseases with heterogeneous clinical manifestations and genetic etiologies. Mutations in ERLIN2 gene have been associated with SPG18, and its symptoms include contractures, progressive global muscle weakness, muscle atrophy, delayed walking, abnormal gait, spasticity, extensor plantar responses, hyperreflexia, speech impairment, intellectual disability, seizures, scoliosis and kyphosis. To date, all of these patients with different types of SPG18 were from Asia and Europe, and none of them from the American continent. Additionally, childhood cataract were not reported in any other SPG18 patients described in the literature before this study. Here, we present two affected siblings and their two cousins with a novel variant in the ERLIN2 gene. It's important to notice that the children's fathers are brothers and their mothers are cousins to each other, all from an isolated region in the Brazilian northeast. Once the same homozygous variant was found in all four patients, it possibly suggests that they share a common ancestor. Also, all patients displayed childhood cataract, expanding the known clinical spectrum of SPG18. **Final comments:** This research marks the first documented case of SPG18 in the Americas, and the findings highlight a consistent presentation of the complicated form of Hereditary Spastic Paraplegia (HSP) and a notable association with childhood cataracts, suggesting a phenotypic expansion of SPG18. Importantly, it introduces a novel variant in the ERLIN2 gene, contributing to our evolving understanding of the genetic factors underlying SPG18. The approach of combining field investigation in highly inbred regions of Brazil, where clusters of genetic disorders are sought, with advanced molecular techniques has proven to be successful.

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Novel homozygous mutation on TRIM32 gene causing autosomal recessive limb-girdle muscular dystrophy 8 (LGMDR8): a case report

Enzo Stinghel Pellacani[1], Alzira Alves de Siqueira Carvalho[1], Ana Marina Dutra Ferreira da Silva Pontes[1], Igor Dias Brockhausen[1], Alessandra Lima Nogueira Tolentino[1], Natalia Merten Athayde[1], Mariah Gomes de Lima[1], Danilo Marinho Pereira[1], Isaías Mendes da Silva Júnior[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Enzo Stinghel Pellacani; Email:

Case presentation: A 50-year-old Brazilian male presents at the neurology department with progressive muscular weakness primarily noticed at 15 years old, when he first

experienced difficulties with running, gradually worsening over time. By 42, he struggled with climbing stairs, initially relying on unilateral support and later evolving to bilateral support. The patient had a medical history of consanguinity (parents were cousins). He had four siblings: two deceased in childhood of unknown causes and two alive and healthy. Neurological examination revealed shoulder girdle muscle atrophy, a waddling gait, symmetrical lower extremities weakness that was worse for hip flexion, hip abduction, hip extension, and knee flexion. The deep tendon reflexes were normal. Laboratory tests showed elevated CPK levels (970U/L; Normal Value: <170U/L). Myography showed a myopathic pattern. Lower limbs MRI revealed fatty infiltration and diffuse atrophy of both thighs and legs, especially afflicting the posterior leg muscles. The echocardiogram was unremarkable, and pulmonary function testing revealed a restrictive pattern. The muscle biopsy revealed a primary dystrophic muscular disorder with protein clusters and cytoplasmic vacuolization. A genetic myopathy panel was performed, identifying a homozygous nonsense variant c.1885C>T (p.Gln629*) in the TRIM32 gene, coding a premature stop codon, leading to a shortened transcript. This variant is currently categorized as of uncertain significance due to the lack of reported data on its clinical impact. Based on clinical and laboratory characteristics, we established the diagnosis of LGMDR8. **Discussion:** TRIM32 acts as an E3 ubiquitin ligase in the ubiquitin-proteasome system and plays multifunctional roles in maintaining skeletal muscle. Pathological homozygotic variants in the TRIM32 gene are known to be associated with protein aggregates and vacuolization in skeletal muscle biopsy. When associated with symptoms of slow progressive limb-girdle weakness (predominantly the posterior compartments of lower limbs), such findings lead to the diagnosis of LGMDR8. To our knowledge, it is the first time this variant is described in association with a phenotypical pattern of LGMDR8. **Final comments:** This report aims to apprise a novel variant associated with LGMDR8. The association between homozigotic variant c.1885C>T (p.Gln629*) in the TRIM32 gene and the muscle biopsy findings safely point toward this diagnosis. Such a variant has not yet been described in the current databases and should be considered as potentially pathogenic.

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Novel variants in PLA2G6 gene: a case report in a young adult

Aline Oliveira dos Santos Meneses[1], Rhaná Carolina Santos[1], Lorenna Lima Regueira Costa[1], Camila Matzenbacher Bittar[2], Carlos Eduardo Mantese[1,3]

[1]Universidade do Vale do Rio dos Sinos, São Leopoldo RS, Brazil.
[2]Hospital Moinhos de Vento, Porto Alegre RS, Brazil.
[3]Hospital Mãe de Deus, Porto Alegre RS, Brazil.

Correspondence: Rhaná Carolina Santos; Email: santoscrhana@gmail.com.

Case presentation: 24 years old female patient, previously healthy. Family members report falls and motor difficulties since the age of 6, worsening in adulthood; no cognitive decline. She denies any family history of similar cases or parental consanguinity. Neurological examination revealed dysarthria, ataxia, and spasticity of all four limbs. No signs of parkinsonism or chorea were detected. MRI showed cerebellar atrophy and iron accumulation in the globus pallidus and substantia nigra. Complete blood count, renal function, electrolytes, liver function tests, serum copper, ceruloplasmin, urinary copper and ferritin were unaltered. Ophthalmologic assessment excluded Kayser-Fleisher rings. Genetic evaluation was carried out through germline exome analysis, identifying two variants in the PLA2G6 gene: c.2032A>G p.(Lys678Glu) and c.1525A>G p.(Lys509Glu), both of undetermined clinical significance. The first has been previously described and the second is absent in the available databases. Based on clinical manifestations, imaging findings and genetic results, PLA2G6-associated neurodegeneration was diagnosed. **Discussion:** PLA2G6-associated neurodegeneration is an autosomal recessive disorder with various phenotypes and distinct clinical features: Infantile Neuroaxonal Dystrophy (INAD), Neuroaxonal Dystrophy (NAD) and PLA2G6-related Dystonia-Parkinsonism. INAD begins between six months and three years of age, characterized by psychomotor delay, hypotonia and progressive spastic tetraparesis, with rapid evolution and high mortality. NAD presents greater phenotypic variability, including gait instability, ataxia or speech delay, as well as neuropsychiatric disorders such as impulsivity, inattention, hyperactivity and emotional lability, which can be stable or progress to encephalopathy. PLA2G6-related Dystonia-Parkinsonism manifests in adulthood with gait disturbances, dystonia, and parkinsonism, possibly accompanied by rapid cognitive decline. There is extensive documentation of phenotypes and genotypes related to PLA2G6, especially in its adult manifestation. The patient's condition suggests an intermediate phenotype, with imaging findings highly suggestive of neurodegeneration with iron accumulation in the globus pallidus and substantia nigra. Exome sequencing confirmed two variants of uncertain significance in the PLA2G6 gene. Segregation study in the parents identified that the two variants are in trans, one on each allele of the PLA2G6 gene, consistent with autosomal recessive inheritance. Possibly, different variants and their combinations are responsible for the wide range of phenotypes associated with PLA2G6-related neurodegeneration. **Final comments:** The case presented is notable as it demonstrates the atypical progression of a rare case with novel germline variants of the PLA2G6 gene.

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Number of deaths by cerebral vascular accident on a population between 20 to 29 years in the state of São Paulo: a timeline comparison from 2014 to 2023

Aline Marques Barreto Scalioni[1], Ísis Larissa de Brito Dichtl[2], Gabriela Fonseca Póvoas[3], FernandoÉriton Aguiar Moita[4], Michely Laiany Vieira Moura[5]

[1]Universidade Nove de Julho, Guarulhos SP, Brazil.
[2]Universidade Estadual do Piauí, Teresina PI, Brazil.
[3]Universidade Santo Amaro, São Paulo SP, Brazil.
[4]Universidade Federal do Ceara, Fortaleza CE, Brazil.
[5]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Aline Marques Barreto Scalioni; Email: alinemscalioni@gmail.com.

Background: Cerebral Vascular Accident (CVA) is the main cause of death in adults in Brasil. This neurological syndrome is characterized by the sudden obstruction of the blood flow to the brain, and it is categorized into two types: ischemic stroke and hemorrhagic stroke. Around 40 to 50% of stroke victims die after 6 months, and it is estimated that around 70% of patients that survive don't completely recover, making it a highly disabling illness. **Objective:** To analyze the deaths by CVA in the state of São Paulo, between the years of 2019 to 2023, and comparing them with the timeline from 2014 to 2018. **Methods:** Ecological, retrospective and descriptive study using data from the Hospital Information System (SIH/SUS) through the platform Department of Informatics of the Unified Healthy System (DATASUS). It analyzed the number of deaths in São Paulo between the ages of 20 to 29 years old, comparing 2014 to 2018 with the years of 2019 to 2023. It excluded remaining filters that were not previously mentioned. **Results:** After reviewing data from 2014 to 2018, the number of deaths by CVA between the ages of 20 to 29 years old in the state of São Paulo was 88 in total, being 2016 the highest, with 55 deaths. When analyzing the years of 2019 to 2023, it was noted an increase in deaths, with a total of 124 deaths, of which 31 happened only in 2022. This increase in deaths by CVA in a young population can be related to the increased exposure to risk factors like smoking, emotional conditions, stress, obesity, a sedentary lifestyle and diabetes. **Conclusion:** There was an alarming increase of almost 50% in deaths when comparing both timelines. Following the results, it is important to raise awareness among young adults between the ages of 20 and 29 about the risk factors of Cerebral Vascular Accident and how to cultivate healthy habits such as avoiding alcohol, eating healthier and exercising.

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Oblique decubitus in bed and its relation with spatial neglect

Tiago César Costa da Silva[1], Camilla Vanessa Araújo Soares[1], Felipe Nunes de Araújo[2], Luciano Gonçalves do Nascimento Júnior[2], Ana Luisa Castelo Branco Gomes[2], Gilmar Leite Pessoa Filho[2], Adriana M. T. Nepomuceno[3], Alex Tiburtino Meira[4]

[1]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[2]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[3]Universidade Federal da Paraíba, Programa de Mestrado Profissional em Gerontologia, João Pessoa PB, Brazil.
[4]Universidade Federal da Paraíba, Departamento de Medicina Interna, Serviço de Neurologia, João Pessoa PB, Brazil.

Correspondence: Tiago César Costa da Silva; Email: tiagocesarcs@hotmail.com.

Background: Stroke is the second cause of mortality in the general population, and its incidence tends to increase because of population aging. The blood vessel territory most affected in the stroke is the middle cerebral artery (MCA). A frequent complication of stroke in the right MCA territory is the hemispatial neglect (HN), which can be defined as a disorder characterized by an unconscious attention deficit expressed in the patient's behaviors in relation to the space contralateral to the lesions. The most common presentation is unilateral neglect, related to lesions in the right cerebral hemisphere with deficits in spatial perception on the left. HN correlates with poor prognosis, longer hospitalization and greater acute-phase complications. From clinical practice, some patients who suffered a stroke tend to lie in an oblique decubitus (OD) in the stretcher (diagonal position on the stretcher). **Objective:** Herein, we attempt to relate OD in the stretcher with HN. **Methods:** We performed a clinical prospective cohort study. The sample consisted of 17 individuals and the participants were recruited from the Dom José Maria Pires Hospital Metropolitano, in Paraíba, Brazil. The sample was divided into two groups: case (12) and control (5). The instruments used for data analysis were Star Cancellation Task (SCT), Bcos Apples Cancellation Task (BACT) and Catherine Bergego Scale (CBS) itens 2,5,6,7,9,10 and a Sociodemographic Questionnaire was used to identify the patient's profile. **Results:** 17 individuals were included in the study. Out of 17, 10 (59%) were female, 70% had a right MCA stroke. The average age was 54.8. All participants were right-handed and in the acute phase of the stroke; 29% were in OD; 23% drink; 35.2% smoke; 58% have high blood pressure; 23% have diabetes. The mean NIHSS score was 7; 23% had HN on physical examination and 41% had HN on SCT, of which 28% were in OD. In the BACT 58% presented HN, with 20% in OD. On CBS, 29% presented HN, with 60% found in OD. A correlation was found between right MCA stroke and SCT (p value 0.02) and a positive correlation between CBS items and SCT (p value 0.01). There was no correlation between the other variables. **Conclusion:** Data analysis showed no correlation between OD and HN. However, there was a good result between CBS and SCT, indicating a good choice for testing HN in the acute phase of stroke. Due to the low sample number, further study is needed in the future. This information contributes to neurological semiology when seeking to confirm OD related to HN in patients presented with acute stroke considering that this type of information does not exist in the current literature.

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Obstruction of the fourth ventricle decompensated by anabolic steroid use: a case report

Brenno Tavares de Vasconcelos Brandao[1], Ana Carolina Leite[1], Barbara Pina Aiello[1], Marcos Martins da Silva[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Brenno Tavares de Vasconcelos Brandao; Email: btvbrandao@gmail.com.

A young male, 24 years old, seeks emergency care with a two-week history of progressively worsening headache, associated with visual blurring and photophobia, which worsens when lying down. He was diagnosed with intracranial hypertension. At 10 months old, he had hydrocephalus and required a ventriculoperitoneal shunt (VPS), but he does not know the cause. He had no complaints during childhood or adolescence after the procedure. He reports using thyroid hormone, vasodilator, veterinary beta agonist, and anabolic steroids (both intramuscular and oral) for aesthetic purposes for six months, stopped after the onset of symptoms. During the investigation of intracranial hypertension, fundoscopy showed no abnormalities, but cranial MRI revealed disproportionate tetraventricular hydrocephalus, with the third and fourth ventricles larger than the lateral ventricles. The VPS catheter was in a normal position. Lumbar puncture demonstrated elevated opening pressure (30 cmH20), protein of 129 mg/dL, leukocytes of 8/mm3 (lymphomononuclear), and glucose of 50 mg/dL. There was partial symptom relief after cerebrospinal fluid removal. Treatment with a carbonic anhydrase inhibitor was started but had an unsatisfactory clinical response and caused gastric intolerance, leading to surgical intervention. An endoscopic third ventriculostomy was performed, with postoperative symptom improvement and reduction in the size of the third and fourth ventricles on follow-up imaging. Hydrocephalus can be obstructive or communicating. The obstructive form occurs due to obstructions of the foramen of Monro or the aqueduct of Sylvius. Obstructions of the fourth ventricle outlet, at the foramina of Magendie and Luschka (FVOO), are rare and generally secondary to infections, hemorrhages, or congenital malformations. FVOO is associated with disorders such as Dandy-Walker and Arnold-Chiari malformations in childhood. Other causes include trauma, intracranial hemorrhages, and infections like meningitis or arachnoiditis. However, presentation in adults is rarely described. Obstruction usually manifests in childhood, as in the case of this patient, who remained asymptomatic after VPS insertion. However, there was an acute decompensation of the cerebrospinal fluid drainage system, likely secondary to systemic changes related to anabolic steroid use, with the onset of typical FVOO symptoms such as headache, nausea, and vomiting. Therefore, in cases of hydrocephalus, it is essential to perform imaging evaluation focusing on the foramina of Luschka and Magendie to check for obstructions. This allows for the most appropriate therapeutic proposal, yielding good results in these cases, such as endoscopic third ventriculostomy.

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Obstructive sleep apnea treatment in stroke patients: a randomized clinical trial

Millene Rodrigues Camilo[1], Alan Luiz Eckeli[1], Aline Marques Franco[1], Fernando Stelzer[1], Arthur Veras[1], Ícaro Araújo[1], Yago Fernandes do Nascimento[1], Marina Foresti dos Santos[1], Ana Paula Camargo[1], Octávio Marques Pontes Neto[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Millene Rodrigues Camilo; Email: millene.camilo@gmail.com.

Background: Obstructive sleep apnea (OSA) is present in more than 70% of stroke patients and has been associated with worse short- and long-term outcomes. Therefore, studies that evaluate the benefit of early OSA treatment in this context are essential. The treatment of choice for OSA is the use of CPAP (continuous positive airway pressure). Previous results regarding the feasibility and safety of using CPAP in post-stroke patients are encouraging. However, efficacy studies are scarce and controversial. Evidence of the benefit of treating OSA in patients with acute stroke will drastically change daily clinical practice. Primary **Objective:** To assess whether early OSA treatment with CPAP in patients with ischemic stroke has a favorable effect on functional recovery. Secondary Objectives - to evaluate whether the use of CPAP will be associated with (1) lower rate of early neurological deterioration; (2) smaller final volume of the cerebral infarction; (3) improvement in cognition and quality of life; (4) lower rate of depressive episodes; (5) reduction of excessive daytime sleepiness and sleep-related complaints; (6) lower serum levels of biomarkers of vascular endothelial injury; and (7) lower rate of recurrence of cardiovascular events and mortality. **Methods:** This is a prospective, multicenter, randomized study, blinded to outcomes (PROBE design), which will include 214 patients diagnosed with ischemic stroke and severe OSA, who will be randomized in a 1:1 ratio (CPAP+ vs. CPAP-). The study will be divided into two stages: (1) Acute Phase: up to 3 months after the ictus and (2) Chronic Phase: from 3 months to 2 years. A multidisciplinary team (nurses, psychologists, physiotherapists, and doctors) will monitor all patients during these stages. **Results:** At the end of this study, we are expected to contribute with answers to gaps in the literature. **Conclusion:** Once the expected benefit is demonstrated, a new perspective will be opened regarding treating OSA in post-stroke patients, enabling a change in daily clinical practice.

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Occult neuroendocrine cancer presenting as Isaacs syndrome

Letícia da Silva Alves[1], Wender Rodrigues Teodoro[1], Matheus Cunha Nunes[1], Diogo Fernandes dos Santos[1], Iago Resende Carvalho[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Letícia da Silva Alves; Email: draleticiasilvaalves@gmail.com.

Case presentation: A 61-year-old female patient, previously healthy, who presented with sudden onset muscle weakness and ambulation difficulty. Subsequently, she complained muscle cramps and slow relaxation worsened by voluntary muscle contraction. Symptoms progressively worsened, resulting in marked functional impairment and difficulty walking. She also reported distal sensory loss, intense fatigue, and exercise intolerance, in addition to intense sweating, weight loss, and muscle stiffness. On neurological examination, sustained involuntary muscle contractions were noted, with intense difficulty in relaxation. These included abnormal postures on the face, feet, and hands, with a predilection for the distal segments. The stiffness and difficulty in relaxation actually worsened with voluntary muscular activity, confirming the presence of pseudomyotonia. Strength and deep tendon reflexes were normal. An extensive laboratory evaluation was conducted, including metabolic, hormonal, serological, creatinine kinase levels, and inflammatory markers. She underwent electroneuromyography, which showed the presence of afterdischarges in the study of motor conduction and evaluation of F waves, in addition to the presence of high-frequency discharges, affecting the face, paravertebral region, and distal muscles of the limbs, with worsening after voluntary contraction, compatible with neuromyotonic discharges, confirming the diagnosis of a hyperexcitability syndrome. Brain magnetic resonance imaging and cerebrospinal fluid showed no abnormalities. Furthermore, given the possibility of a paraneoplastic condition, a neoplasm investigation was carried out, and a rectal polyp was found on a colonoscopy. Histopathological analysis was compatible with a well-differentiated neuroendocrine tumor. She was treated with Intravenous immunoglobulin (IVIg), presenting a significant improvement in her symptoms. **Discussion:** Isaacs syndrome has been reported in patients with malignancy, suggesting that tumor antigens trigger an autoimmune response and result in antibodies that cross-react with neuronal voltage-gated ion channels. This is consistent with the increased incidence of autoimmune neurological paraneoplastic disorders in patients with Isaacs syndrome. Several neoplasms have already been described in this association, including thymoma, lung cancer, Hodgkin lymphoma, plasmacytoma, lymphoblastic lymphoma, hemangioblastoma, ovarian and bladder cancer. **Final comments:** Our patient demonstrates that Isaacs' syndrome could be the initial and only neuromuscular manifestation of neuroendocrine cancer.

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Occurrence of pelvic obliquity and scoliosis in 5q-spinal muscular atrophy

Graziela Jorge Polido[1], Eduardo Vital de Carvalho[1], Rodrigo de Holanda Mendonça[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.

Correspondence: Eduardo Vital de Carvalho; Email: duvital1983@gmail.com.

Background: Spinal muscular atrophy linked to chromosome 5 (5q-SMA) is a progressive genetic disease caused by disease-causing variants in the survival motor neuron 1 (SMN1) gene. The modifying therapies have changed the natural history of 5q-SMA, but the scoliosis is still a problem and can present a rapid progression and cause important function impact. **Objective:** To show the frequency of pelvic obliquity in patients with 5q-SMA and occurrence of the scoliosis associated with pelvic obliquity. **Methods:** This retrospective observational study evaluated all patients who developed only pelvic obliquity, pelvic obliquity and scoliosis or only scoliosis. **Results:** 16 patients with 5q-SMA, type 3 (n=3), type 2 (n=6) and type 1 (n=7), were included. All of them were using any disease-modifying therapies. From the type 3 patients, one had pelvic obliquity and was walker. No one had scoliosis. From the six patients with type 2, all had pelvic obliquity, one had spinal surgery and three had unstructured scoliosis. In addition, two had therapeutic walking and one community walking. From the seven patients with type 1, all had pelvic obliquity and five had scoliosis. In addition, all sat without support and four were able to stand with support using upper limbs. **Conclusion:** The pelvic obliquity is an important signal preceding the development of scoliosis and it needs to be treated carefully to avoid the early onset of scoliosis.

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Ocular amyloidosis: a rare differential diagnosis of orbital pseudotumor

Monique Evelyn Mendonça do Nascimento[1], Marcelo Ataíde de Lima[1], Mariana Holanda Gameleira[1], Edna Lúcia de Freitas Silva[1], Raiane Raianne Silva Lima[2], Ândrea Virgínia Ferreira Chaves[3], Vanessa de Souza Coutinho[1], Brenda Letícia Lopes Batista[1], Rosevaldo Alves Junior[1], Bárbara Borges de Oliveira[1]

[1]Hospital da Restauração, Recife PE, Brazil.

[2]Fundação Altino Ventura, Recife PE, Brazil.
[3]Hospital Agamenon Magalhães, Recife PE, Brazil.

Correspondence: Monique Evelyn Mendonça do Nascimento; Email: moniqueemnascimento@gmail.com.

Case presentation: A 57-year-old female patient presented with visual loss and proptosis in her right eye. Initial imaging revealed thickening of the extrinsic muscles of the right eye, and subsequent MRI confirmed this, showing thickening and enhancement in the superior rectus muscle and its tendon and insertion. Cerebral angiography ruled out arterial abnormalities. The patient received pulse therapy with methylprednisolone, resulting in partial improvement. Initial hypotheses of orbital pseudotumor, granulomatous diseases (IGG4 disease), and thyroid disease were considered but ultimately ruled out. The patient was discharged on corticosteroid therapy, but two weeks later, she returned with worsening symptoms despite the treatment. She did not experience pain. An ophthalmology re-evaluation was requested, and an ocular biopsy was carried out, with results compatible with ocular amyloidosis (apple-green birefringence to cone-red colouration). **Discussion:** Amyloidosis is a group of diseases caused by the deposition of insoluble fibrils composed of poorly aggregated proteins, which form insoluble protein aggregates. The clinical manifestations and sites affected vary depending on the clinical manifestations and sites. Ocular amyloidosis, or amyloidosis of the ocular appendages, is uncommon and may be related to both the systemic and localized forms of the disease. There is a predominance of females, with an average age of 55 years. In most cases, orbital involvement is asymptomatic and indolent. The differential diagnosis of amyloidosis should include idiopathic orbital pseudotumor, lymphoma, leukaemia, sarcoidosis, and other granulomatous. A biopsy with a finding of birefringence under polarized light after staining with Congo red is the gold standard for a diagnosis. Transthyretin amyloidosis, or ATTR, is one of the subtypes most associated with ocular involvement. Neuropathies, renal and cardiac dysfunction are included in the clinical picture, in addition to ophthalmopathy. The treatment of the ophthalmic manifestations of amyloidosis can be conservative with the use of tafamidis, radiotherapy, or a surgical approach. **Final comments:** Amyloidosis is a heterogeneous spectrum of disease that can affect the eye and the periocular region. The most prevalent ophthalmic manifestations of amyloidosis are proptosis, hyperaemia, and ophthalmoparesis. The gold standard for diagnosis includes optical microscopy after biopsy, with typical features. It is important to consider the disease within the differential diagnoses, as it can mimic many other pathologies. Following the diagnosis, the patient was monitored by the neurophthalmology department and scheduled for radiotherapy.

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Ocular dipping in a severe traumatic brain injury patient after being buried: a case presentation

Yves Henrique Ramos Mansano[1], Pamella Francisquini Gardin[1], Rodrigo Pernomian Cianca[1], Ana Letícia Manso Assakawa[1], José Guilherme Pinhatti Carrasco[1], Isabela Kirach Gaio[1], Felipe Bento Michelato[1], Antônio Paulo Felice Rodrigues Leite[1], Antônio Marcos Reissureição Galindo[1], Gabriel Bortoli Ramos[1]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Yves Henrique Ramos Mansano; Email: ra124955@uem.br.

Case presentation: Male, 35 years old, victim of burial beneath a mudslide, was transported to the hospital via air ambulance in trauma protocol. At the time of admission, he had 4 points on the Glasgow Coma Scale and was being ventilated through Guedel and high-flow mask. The patient presented with periorbital ecchymosis, anisocoria and otorrhagia from the right ear. Head computed tomography revealed complex skull fracture, pneumocephalus and discrete intraparenchymal hemorrhagic foci, suggesting traumatic subarachnoid hemorrhage and severe diffuse axonal injury. Neurological examination demonstrated brainstem reflexes present, hyperreflexia in lower limbs and decerebrate posturing in response to painful stimuli. In addition, the patient presented an abnormal spontaneous vertical eye movement with slow phase down, followed by a quick return to the original position. **Discussion:** Ocular dipping consists in cyclical and spontaneous eye movements, which are characterized by a slow deviation of the gaze downward, followed by a delay at the lower extreme of the gaze and a quick return to the intermediate position. Most cases occur in the context of severe anoxic brain injury with associated diffuse cortical and lenticular pathology, but it has also been described in prolonged status epilepticus and Creutzfeldt-Jakob disease. Despite generally being present in severe cases, the movement is not a good prognostic marker, as there are reports in the literature of substantial recovery after an ocular dipping episode. It is assumed that a mechanism for ocular dipping involves dysfunction of cortical/lenticular connectivity - with the centers of vertical gaze located in the rostral portion of the brainstem - and ascending arousal system. The neurological examination is essential for the correct topographic diagnosis and request for complementary exams, especially because there are several ocular movement abnormalities that can be confused with dipping. Reverse ocular dipping is a variant that was described in the context of metabolic encephalopathy or encephalitis, which is characterized by a slow upward deviation with a fast descending phase to the primary position. Ocular bobbing involves quick eye movements downward and a slow return to the intermediate position, usually occurring in cases of pontine lesions. Moreover, dipping and bobbing are differentiated from vertical nystagmus by their irregularity, brief pause between phase changes, and facilitation by harmful stimuli. Prognosis for patients with ocular dipping is variable and, interestingly, a return of consciousness or the use of pentobarbital can abolish the movement. **Final comments:** Neurological examination in critical care patients is crucial for differential diagnosis of brain lesions. Ocular dipping is strongly associated with anoxic brain injuries in critical patients. However, the appearance of ocular dipping cannot be used as a prognostic marker in coma.

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Oculomotor nerve paralysis by varicella zoster after dengue fever: case report

Iury Venâncio Pinheiro[1], José Elias Makaron Neto[1], Leonardo de Oliveira Moreno[1], Thire Baggio Machado Marazzi[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Iury Venâncio Pinheiro; Email: iuryvenancio145@gmail.com.

Case presentation: A previously healthy 47-year-old woman reported myalgia, headache and fever with a diagnosis of dengue confirmed by positive serum NS1. After 5 days, she presented with a new complaint of horizontal diplopia on the lateralization of the gaze to the left, in addition to eyelid ptosis and pain in eye movement. Left eye asymptomatic. On physical examination, she had right eyelid ptosis, mydriatic and bradiareactive right pupil, mild exotropia, restriction of adduction, and infraversion of the right eye gaze. She has no evident skin lesions. Campimetry unchanged. Complementary tests showed thrombocytopenia, increased transaminases, leukopenia, and lymphocytosis. CFS analysis showed no pleocytosis or protein abnormalities, and Dengue DNA PCR was negative. However, a DNA PCR of the varicella-zoster virus (VZV) in CSF was detected. Brain MR showed enhancement in the cisternal path of the right III nerve, suggestive of neuritis, in addition to diffuse thickening of the pachymeninges. The treatment proposed was acyclovir for 10 days. The patient had progressively improved but maintained only mild eyelid ptosis. **Discussion:** Reactivation of VZV classically occurs in situations of immune depletion, with a skin rash respecting the territory of dermatomes and neuralgia. Cranial polyneuritis and ocular inflammatory manifestations induced by reactivation of VZV without rash have been previously described. However, isolated oculomotor nerve palsy, in the absence of skin lesions, is a rare finding and represents a challenging diagnosis in the presence of other viral infections in immunocompetent patients, as in the present case, in which the dengue virus can also cause neurological dysfunctions of self-limited course. Findings of normal CSF biochemistry with dengue negativity reinforce the diagnosis of reactivation and VZV-induced neurological damage. **Final comments:** The reported case is a rare isolated involvement of III nerve by VZV, in the absence of cutaneous lesions, in an immunocompetent patient with dengue. Viral coinfections are uncommon but should be part of the differential diagnosis of cranial neuritis.

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Odontogenic cerebral abscess presenting as thunderclap headache and sensory aphasia

Leonardo Villaverde Buback Ferreira[1], Sofia Mônaco Gama[1], Lucas Toshio Uenishi[1], Fernanda Lustosa Cabral Gomez[1], Lara Santos Machado[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Daniel Delgado Seneor[1], Gabriel Lopes da Silva[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Leonardo Villaverde Buback Ferreira; Email: leonardo.buback@gmail.com.

A 28 years-old female was admitted to the emergency department after the sudden onset of a headache described as a pressure feeling, reaching its maximal intensity in a few seconds and associated with "difficulty of speech", all of it starting a few minutes after waking up, 4 days before admission. Her relevant medical history included a diagnosis of Noonan syndrome, and an interatrial communication surgically repaired at 6 years-old. At the moment of the examination, she was afebrile, awake and unable to attend to verbal commands, repeat sentences or name objects, but kept the ability to reproduce gestures and her speech was fluent, thus characterizing a sensory aphasia, without other abnormalities on physical evaluation. Her head CT-scan demonstrated a hypodense lesion in the left temporal lobe, with ring-enhancing aspect after the administration of intravenous contrast, suggesting a cerebral abscess. After the introduction of intravenous antibiotic therapy, using ceftriaxone, metronidazole and linezolid, she was submitted to the surgical evacuation of the lesion which revealed the presence of purulent material, subsequently sent to microbiologic and molecular studies, the latter represented by the metagenomic next-generation sequencing (mNGS) which identified Staphylococcus intermedius. The other microbiological tests (blood and abscess cultures) were all negative, and systemic investigations (transesophageal echocardiogram, thorax and abdomen CT) also. Given the association of this microorganism with odontogenic infections, a facial CT was acquired, demonstrating a dental abscess in the right mandibular region, then submitted to surgical treatment. In the outpatient follow-up 2 months after discharge, she was asymptomatic. The clinical triad of headache, focal deficits and fever is found in about 25% of cases, being fever absent of over 50%. The prevalence of headache reaches

70%, though the most common presentation is a subacute headache, with thunderclap being an uncommon pattern mNGS is a method to directly detect DNA, RNA or transcriptional products in a sample. Its major importance is evident in cases where certain pathogens might not be suspected only by clinical data, and when microbiology tests do not show positive results. In cerebral abscesses, the identification of these rely hugely on cultures, negative in over 63% of cases. The low sensibility can be partially explained by the early introduction of antibiotics and by the delay in culture execution after sample acquisition, which are less important aspects when using mNGS. The present case illustrates an atypical presentation of a common disease, where the molecular identification of the causative agent by mNGS was crucial in the correct investigation and treatment of a primary focus responsible for a cerebral abscess.

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Olfactory changes in Parkinson's disease patients undergoing deep brain stimulation in the subthalamic nucleus: a systematic review and meta-analysis

Pedro Ferreira de Souza Evangelista[1], Maria Antônia Oliveira Machado Pereira[2], Bernardo Caetano Novaes[3], Myleyde Dayane Pereira da Silva[4], Monyque Alves Loiola[5], Artur Menegaz de Almeida[6], Ítalo Barros Andrade[1], Lilianne Rodrigues Fernandes[7], Michele Kreuz[8]

[1]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[2]Universidade Estadual do Piauí, Teresina PI, Brazil.
[3]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.
[4]Universidade Federal do Cariri, Barbalha CE, Brazil.
[5]Universidad Nacional de Rosario, Santa Fe, Argentina.
[6]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[7]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.
[8]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Pedro Ferreira de Souza Evangelista; Email: pedro.evvan@gmail.com.

Background: Deep brain stimulation in the subthalamic nucleus (STN-DBS) is a common therapeutic intervention for patients with Parkinson's disease (PD), primarily aimed at improving motor symptoms. However, patients with PD often experience non-motor symptoms such as anosmia. Therefore, the impact of STN-DBS on olfaction remains poorly understood. **Objective:** We performed a systematic review and single-arm meta-analysis to evaluate and synthesize the evidence on olfactory changes in Parkinson's patients undergoing STN-DBS. **Methods:** PubMed, Scopus, and Web of Science databases were systematically searched until May 13, 2024. Eligible studies included observational studies with ≥4 patients reporting the effects of STN-DBS on olfactory function in PD patients. A random-effects model was used to calculate the Proportion analysis with 95% confidence intervals (CIs). RStudio version 4.3.2 was used for statistical analysis. **Results:** A total of six studies, encompassing 179 PD patients, were included in our analysis. Of them, 58.6% were men. Six endpoints were evaluated and all presented statistically significant improvements tending towards the post-STN DBS state: Odor discrimination (OD) (mean difference 2.03; 95% CI: 1.25 to 2.82; p<0.000001; I^2=2%), Odor identification (OI) (mean difference 2.38; 95% CI: 1.43 to 3.33; p<0.000001; I^2=16%), Odor thresholds (OT) (mean difference 1.47; 95% CI: -0.36 to 3.30; p<0.000001; I^2=87%), and the TDI (composite score derived from the sum of OD, OI, and OT results) (mean difference 5.46; 95% CI: 3.39 to 7.54; p<0.000001; I^2=0%). In contrast, the Unified Parkinson's Disease Rating Scale part III (UPDRS III) (mean difference -20.67; 95% CI: -34.47 to -6.86; p=0.003340; I^2=93%) and the University of Pennsylvania Smell Identification Test (UPSIT) (mean difference -1.20; 95% CI: -4.23 to 1.83; p=0.438727; I^2=0%) tended towards the pre-DBS state, with UPDRS III being statistically significant and UPSIT not statistically significant. **Conclusion:** In this single-arm meta-analysis, consistent results suggest that STN-DBS is associated with improved olfactory function in PD patients.

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One sided chorea associated with lupus anticoagulant with no other abnormalities: case report

Letícia Klabinske Marques Monteiro[1], Eduardo Sousa de Melo[1], Mariana de Almeida Cruz Silva[2], Gabriel Farina Giovani[2], Aimê de Paula Santos[2], Evelliny Gomes da Silva[1], Allefy Beltrão Albano[1], Fabrício Martins Veras[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Leticia Klabinske Marques Monteiro; Email: leticiaklabinske@gmail.com.

Case presentation: An 80-year-old woman reported symptoms of hyperkinetic and unpredictable involuntary movements in the left side, daytime sleepiness, morning headaches and snoring for 8 months, with worsening of the condition in situations of anxiety. Neurological assessment revealed left hemichorea, with no other alterations. Laboratory screening for chorea was carried out and polysomnography and brain MRI were requested. Negative anti-beta-2 and anticardiolipin antibodies and positive lupus anticoagulant were stated by coagulometric method, but there was no association with Antiphospholipid Syndrome (APS). Brain MRI didn't reveal any findings. She is taking risperidone 2 mg, and is awaiting polysomnography to better elucidate this condition. **Discussion:** Chorea correlated with autoimmune diseases has been reported, as well as in the context of the presence of high plasmatic titers of Anticardiolipin Antibodies, anti-β2-GPI and Lupus Anticoagulant, even in the absence of APS. These recent clinical reports go in contrast to the past belief that this autoimmune chorea occurred only in the setting of fully developed APS. That said, case reports of this condition are uncommon, and the physiopathological subtractes of chorea related to antiphospholipids antibodies (aPL) remain unclear. Two mechanisms have been proposed to clarify the neurological impairment to the Central Nervous System caused by aPL. One of them suggests that the high affinity of aPL to the phospholipids in specific neurons causes damage in the dopaminergic nigrostriatal pathway, which may explain abnormal motor symptoms. The other states that the tropism of aPL for the endothelium of cerebral vessels disturbes its permeability, resulting in a leaked Blood-Brain-Barrier and contributing to thrombosis and hypercoagulation. An older theory used to associate chorea and other movement disorders to the occlusion of lenticule-striate arteries, producing basal ganglia ischemia. **Final comments:** Even though chorea is a recognized but rare manifestation of immune dysfunction related to aPL, the mechanisms of this disturbance are still to be explained, as well as the natural history of the condition. Case reports such as this, in which the clinical presentation of chorea with no other abnormalities besides Lupus Anticoagulant presence are exposed, must be relevant to that discussion.

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Oneiric stupor: a clue for immune-mediated encephalopathy?

Carolina Souza Sanchez[1], João Henrique Fregadolli Ferreira[1], Mauricio Elias Nunes[2], Andrey Soares[2], Maira Honorato[2], Lívia Almeida Dutra[1]

[1]Instituto do Cérebro, São Paulo SP, Brazil.
[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Carolina Souza Sanchez; Email: carol.souza.sanchez@gmail.com.

Case presentation: An 83-year-old man was admitted for investigation after a 7-day history of temporal disorientation, memory complaints, and excessive daytime sleepiness. He was diagnosed with lung carcinoma with brain metastasis in 2018, and underwent chemotherapy and pembrolizumab and continued at 21-day intervals until 2022. He was a prior heavy smoker, had hypertension, and total colectomy in 2011. Neurological examination revealed daytime drowsiness and hypersomnolence. During sleep, he exhibited purposeful, stereotyped masticatory and hand movements, responding to verbal stimuli but sustaining vigilance for less than ten seconds. He was temporally and spatially disoriented, with dysarthric speech. Positive and negative myoclonic jerks in the upper limbs and limb rigidity were also observed. Strength, sensory, cerebellar functions and cranial nerves were normal. Brain MRI showed late post-operative resection in the left frontal lobe with global atrophy. EEG showed mild disorganization. Polysomnography revealed disorganized brain activity, with excess slow waves and lack of N1 and REM stages, with wake-like movements during arousals, increased muscle tone in both limbs, and excessive hand movements consistent with oneiric stupor (OS). CSF analysis revealed < 1 cell/mm^3, protein 32 mg/dL, low beta amyloid 42 mg/dL, with normal pTau and total Tau levels. Cultures and viral panel were negative, as well as antineuronal antibodies for cell surface antigens and high-risk antibodies. PET-CT scan did not show hypermetabolic lesions. He was treated with intravenous immunoglobulin and showed complete improvement. **Discussion:** OS is one of the hallmarks of Agrypnia excitata (AE), an overactivation syndrome with persistent insomnia and autonomic sympathetic activation due to disruption of thalamolimbic circuits that regulates the sleep-wake cycle. AE is classically associated with Fatal Familial Insomnia, Delirium Tremens and Morvan syndrome. However, our patient did not meet diagnostic criteria for any of those diagnosis. The clinical presentation and extensive investigation ruled out autoimmune encephalitis and paraneoplastic syndromes. This report indicates that other conditions may present with OS. Although we could not demonstrate an inflammatory etiology, we believe that it may have an immune-mediated mechanism, due to the prompt response to IVIG and prior use of pembrolizumab. **Final comments:** OS is a rare and complex condition often linked to severe underlying neurological or systemic diseases. This case suggests that OS can occur in other clinical conditions, with a presumed immune-mediated mechanism, emphasizing the need for clinician awareness of OS symptoms and atypical presentations.

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On-line speech-language assessment in children with autism spectrum disorder

Camila Pimentel Piemonte[1], Maria Eduarda Godinho Freire[1], Letícia Segeren[1], Fernanda Dreux Miranda Fernandes[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Camila Pimentel Piemonte; Email: camila.piemonte@usp.br.

Background: Autism spectrum disorder (ASD) is characterized by deficits in verbal communication, social interaction, and cognitive flexibility, often resulting in repetitive behaviors. Speech therapy interventions have

shown positive outcomes in improving social communication and adaptive behaviors. Many services, including speech therapy, have transitioned to online delivery. Teletherapy offers several benefits but also presents challenges. **Objective:** Assess the efficacy of an online pragmatic language assessment procedure to determine the social-communication profiles of children with ASD. **Methods:** Fifteen children aged 4 to 13 years undergoing online speech-language pathology intervention at the speech-language research laboratory in ASD at the School of Medicine, University of Sao Paulo, participated in this study. This specialized service for children with ASD provides supervised practice for undergraduate students. Parents were instructed to record their child's behavior during various activities. These videos were analyzed using the «Functional Communication Profile – Checklist» (FCPC), which evaluates communicative functions to determine each child's profile, abilities, and difficulties. **Results:** The video analysis revealed difficulties in asynchronous evaluation by parents. Despite initial recommendations, video durations varied widely (mean 1.32 minutes, range 25 seconds to 10 minutes 24 seconds). Parents often failed to select appropriate behaviors to record. Most videos showed guardian-dominated interactions, which are not typical of face-to-face assessments and may not adequately capture the child's communicative intent. A minority of videos showed challenging situations with predominant children's communication. Frequently observed communicative functions included recognition of the other, exhibition, naming, performative, and exclamatory. Assessment difficulties were due to unclear instructions to parents on what situations to record. With better instructions, parents could provide more focused videos on the communicative functions each child could express, allowing better comparison with face-to-face assessments. Nonetheless, the data obtained about family dynamics and the child's social communication abilities in a natural environment provided valuable insights that could complement therapist evaluations. **Conclusion:** Online speech-language assessments are feasible with clear protocols guiding caregivers on what and how to record. These assessments should ideally be supplemented by face-to-face sessions conducted by speech-language therapists. Home environment videos and caregiver reports can significantly contribute to understanding a child's communication profile in telepractice.

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Onset of occipital seizures over 20 years after temporal lobectomy: case report

Márcio Pinheiro Lima[1], Jeana Torres Corso Duarte[1], Adriel Rêgo Barbosa[1], Gabriel Pinheiro Martins de Almeida e Souza[1], João Victor Cabral Correia Férrer[1], André Felipe Candêas Amorim[1], Rafael Tuzino Leite Neves Maffei[1], Ruan Gambardella Rosalina de Azevedo[1], Victor Cardoso de Faria[1], Pedro Henrique Almeida Fraiman[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Márcio Pinheiro Lima; Email: mplima1997@gmail.com.

Case presentation: A 43-year-old woman was admitted to the emergency department with a complaint of "changes in vision" for four days. She noticed these changes upon waking and they consisted of metamorphopsia (she described objects as being bigger or smaller than they actually were, besides seeing faces as distorted and "monstrous"), difficulty in recognizing objects, and visualizing simple geometric shapes and "stars". These complaints were fluctuating, lasting up to four hours. The patient had a past medical history of a traumatic brain injury at five years old and epilepsy starting at seven years old, with seizures characterized by episodes of behavioural arrest with occasional progression to bilateral tonic-clonic seizures. She was treated with a right temporal lobectomy at 20 years old. After the surgery she had four seizures, similar to those she had previously. At admission, she was using extended-release valproic acid (500 mg/day). Neurological examination revealed left homonymous hemianopia with preserved visual acuity, though she reported she couldn't distinguish details of the examiner's face. General biochemical tests did not show any noteworthy alterations. A 3-tesla magnetic resonance imaging (MRI) of the brain was performed, showing an area of restricted diffusion in the right occipital cortex. An electroencephalogram (EEG) was performed, finding six electroclinical seizures and lateralized delta rhythmic activity (LDRA) in the right temporal and occipital regions. The antiseizure drugs were gradually optimized to valproic acid (1000 mg every 12 hours), clobazam (10 mg every 8 hours), and phenytoin (100 mg every 8 hours), resulting in clinical normalization. **Discussion:** The prevalence of epilepsy is around 1% of the population. Of those, up to 40% have drug-resistant epilepsy. Up to 70% of the patients who undergo surgery for temporal lobe epilepsy achieve a seizure-free outcome. In patients who do not have this outcome, the mechanisms for maintaining seizures are diverse and may involve incomplete resection of the epileptogenic area, mirror foci, other structural brain lesions, or epileptogenic activity in the surgical scar. In these patients, approximately 70% of seizures begin in the hemisphere ipsilateral to the surgical resection. Quantitative evaluations with MRI show cortical changes in the temporal lobe, hemispherical, and bilateral changes postoperatively. In cases of new extratemporal seizures, it is possible that other cortical epileptogenic areas, after the temporal surgical resection, assume the role of generating the seizures. Changes in the propagation pattern of the seizures may also occur. **Final comments:** Epilepsy surgery is an effective procedure for seizure control, but certain patients do not become seizure-free. This may occur due to various mechanisms, and it is necessary to consider the possibility of the appearance of new semiological patterns in these patients.

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Ophtalmoparesis with areflexic mydriasis as an atypical presentation of anti-GT1a IgG syndrome

Laura Araújo Paulino[1], Camila Caroso Lobo[1], Kaio Cezar Ferreira[1], Rafael Guimarães Kanda[1], Mayko Nascimento Merscher[1], Luiz Fernando Urzeda[1], Péricles Maranhão Filho[2], Laura Silveira Moriyama[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Laura Araújo Paulino; Email: laurapaulino16.2@bahiana.edu.br.

Case presentation: A 21-year-old male patient with sickle cell anemia developed diplopia and mydriasis 12 days after a gastrointestinal infection. Two days later hematological assessment revealed 60% HbS and two exchange transfusions and transfusion of a packed red blood cell was performed with reported discreet improvement of mydriasis. Two days later he was seen by neurology with ophthalmoplegia characterized by limitation in abduction and adduction bilaterally, worse in the right eye. Additionally, bilateral areflexic mydriasis with a larger pupil diameter on the right, slight ptosis on the left, horizontal nystagmus with rapid phase ipsilateral to lateral gaze, and mild imbalance in tandem gait were noted. Brain magnetic resonance imaging was performed to rule out associated intracranial lesions, revealing subtle enhancement of the III, IV, and V cranial nerves on the right side. Cerebrospinal fluid (CSF) testing was unremarkable including protein electrophoresis and oligoclonal bands. Considering atypical Miller Fisher syndrome (MFS) serum anti-ganglioside testing was performed and detected positive anti-GT1a IgG antibodies (with negative IgM) in the cerebrospinal fluid, without other abnormal findings. Treatment with intravenous human immunoglobulin was administered, resulting in incomplete but significant improvement of both external and internal ophthalmoplegia, as well as improvement in imbalance. **Discussion:** Anti-GT1a IgG antibodies are commonly found in MFS, cases of the pharyngeal-cervical-brachial variant of Guillain-Barré Syndrome (GBS), or in overlaps of GBS spectrum syndromes, whose clinical syndromes commonly involve ophthalmoplegia, weakness in arms and legs, ataxia, and drowsiness. In a retrospective cohort of 140 patients with positive anti-GT1a developed in 2004, 9 has negative anti-GQ1bb and 131 with concomitant positive anti-GQ1b; in the group with activity of anti-GQ1b, external ophthalmoparesis was present in 89 patients; in the group without positive anti-GQ1b, 22 had external ophthalmoplegia; internal opththalmoplegia was only described in those with concomitant anti-GT1a and anti-GT1b IgG detected (40 patients). Among patients with positive anti-GT1a without anti-GQ1b positivity, diarrhea was the most common antecedent, and diplopia was the second most common initial manifestation. Internal ophthalmoplegia is often more described in association with anti-GQ1b antibodies, which is the most common auto-antibodies in Miller Fisher syndrome. **Final comments:** To our knowledge this is the first description of internal opththalmoplegia in atypical MSF with positive anti-GT1a without GT1b.

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Opsoclonus-myoclonus-ataxia syndrome associated with dengue virus infection

Nathalye Fernanda Pedroso Dircksen[1], Marcelo Simplicio Carvalho[1], Letícia de Barros Dias[1], Lorena Fernandes Kronbauer[1], Fábio Gonçalves[1], Felipe Leonardo Semensati[1], Mônica Marcos de Souza[1], Damacio Ramon Kaimen Maciel[1]

[1]Irmandade da Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Nathalye Fernanda Pedroso Dircksen; Email: n.athalye@hotmail.com.

Case presentation: A 36-year-old woman diagnosed with dengue, on the thirteenth day after the onset of symptoms presented with mental confusion, involuntary eye movements, vertigo and difficulty walking. When evaluated by the neurology team, she appeared confused, with movement eye chaotic, involuntary, multidirectional and hyperkinetic. Non-rhythmic and spontaneous appendicular myoclonic movements, trunk ataxia, consistent with opsoclonus-myoclonus-ataxia syndrome (OMAS). The cerebrospinal fluid showed predominant pleocytosis. Magnetic resonance of the neuroaxis was unremarkable. Dengue serology was reactive and other serology and neoplastic screening tests were normal. During hospitalization, the patient presented a worsening level of consciousness, emotional lability, dysarthria and dysphagia. Due to clinical worsening, it was decide to start pulse therapy with intravenous methylprednisolone at a dose of 1g/d for 5 days, followed by an infusion of immunoglobulin human intravenous 400mg/kg/weight per 5 days. Clonazepam 2 mg every 12 hours was also administered to control symptoms with good response. There was complete improvement after about two weeks. **Discussion:** Dengue is a systemic viral infection, transmitted between humans by the Aedes aegypti mosquito, and can manifest itself in different ways, including neurological involvement like meningitis, encephalitis, transverse myelitis, Guillain-Barré Syndrome, optic neuromyelitis, optic neuritis, and rarely OMAS. Movement disorders as a consequence of dengue virus infection are rare and there are few cases reported in the literature. OMAS was described in 1962 by Marcel Kinsbourne and is also known as "dancing eyes-dancing feet" or "Kinsbourne syndrome". It is associated with ocular, motor, behavioral, sleep and language disorders. In most cases it is a consequence of

neuroblastoma in children and paraneoplastic syndrome in adults. Neurological complications in dengue infection occur through some mechanisms: neurotropism, immune-mediated or antibody-dependent reaction, being more likely the OMAS secondary to encephalitis and post-infectious immunological damage, due to the delay in the onset of symptoms and the response to corticosteroid therapy. Although the pathogenesis from the illness he comes from the dysfunction of pontine neurons, skull MRI does not demonstrate lesions in the parenchyma, whereas CSF with pleocytosis suggests encephalitis. Depending on the patient's clinical condition, expectant treatment or immunotherapy (corticosteroids, intravenous immunoglobulin and/or plasmapheresis) can be chosen. Symptomatic treatment with clonazepam can also be performed. **Final comments:** There are few cases in the literature of OMAS secondary to dengue infection. Therefore, there are no evidence-based guidelines or consensus available to guide treatment, which is carried out empirically. We hope to contribute to the literature by demonstrating the case and outcome after empirical therapy.

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Opsoclonus-myoclonus-ataxia syndrome: a case report

Igor Baroni Cardoso[1], Louissa Srama Rosner Cidral[2], Lara Mensato Döring[1], Amanda Yumi Ikematu[3], Ayumi Oya Santos[3], Crislley Mulbauer[3], Johnatan Neuls Pasta[4], Nátali Pimentel da Silva[5], Ana Carolina Andrade[6]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Universidade Positivo, Curitiba PR, Brazil.
[3]Pontifícia Universidade Católica do Paraná, Curitiba PR, Brazil.
[4]Hospital da Cruz Vermelha Brasileira, Curitiba PR, Brazil.
[5]Unifacear, Araucária PR, Brazil.
[6]NeuroDOC, Curitiba PR, Brazil.

Correspondence: Igor Baroni Cardoso; Email: igorbaronic@gmail.com.

Case presentation: A 28-year-old man was admitted to hospital with complaints of involuntary eye movements and motor coordination difficulties. Three weeks prior, the patient had experienced a self-limiting viral febrile episode. Clinical examination revealed bilateral, intrusive saccadic eye movements, exacerbated both during gaze fixation at rest and upon light exposure, consistent with opsoclonus. The patient exhibited mild ataxia in all four limbs, more pronounced on the left side, and truncal ataxia. A subsequent MRI of the neuraxis showed no abnormalities. Cerebrospinal fluid (CSF) analysis revealed a leukocyte count of 40 (98% lymphocytes), protein level of 56 mg/dL, and glucose level of 64 mg/dL. A paraneoplastic screening with a tomography of the chest, abdomen, and skin, along with testicular ultrasound, showed no abnormalities. CSF antibody testing returned negative results for Anti-Hu, Anti-Yo, Anti-Ri, Anti-Amphiphysin, Anti-CV2, Anti-Ma2, Anti-MGT-30, Anti-AGNA, and Anti-Recoverin. Given the syndromic diagnosis of Opsoclonus-Myoclonus-Ataxia, treatment was initiated with methylprednisolone pulse therapy (1g/day) for five days, followed by intravenous human immunoglobulin (2g/kg) for another five days. The patient was discharged from hospital after significant symptom improvement. **Discussion:** Opsoclonus-Myoclonus-Ataxia Syndrome is a rare neurological syndrome, scarcely described in recent literature, requiring precise diagnosis and appropriate therapeutic approach. This case highlights a potential presentation of the syndrome, with intrusive eye movements, significant ataxia, and neuroimaging plus the cerebrospinal fluid studies aided in the exclusion of other possible etiologies. The management of the patient with methylprednisolone followed by intravenous human immunoglobulin, resulting in symptom improvement, suggests a potential therapeutic regimen for Opsoclonus-Myoclonus-Ataxia Syndrome, despite the absence of confirmed autoimmune or paraneoplastic etiology. **Final comments:** The described case illustrates Opsoclonus-Myoclonus-Ataxia syndrome with a positive response to treatment with methylprednisolone and intravenous human immunoglobulin. This underlines the importance of disseminating information about the presentation of this syndrome and its therapeutic possibilities. Furthermore, it exemplifies the significance of considering paraneoplastic and autoimmune hypotheses in the diagnostic and therapeutic approach to this clinical condition. Documenting such cases is crucial for improving the clinical and therapeutic approaches to rare neurological conditions.

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Optic neuritis secondary to bartonellosis (cat scratch disease by bartonella quintana): a case report

Luiz Felipe Mota Freitas[1], Iasmim Portela Maifrede[1], Rafael Barbosa Mokdeci Surerus[2], Daniel Sabino de Oliveira[1], Leopoldo Antonio Pires[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Ebserh, Juiz de Fora MG, Brazil.
[2]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Luiz Felipe Mota Freitas; Email: luizfelipemotafreitas@gmail.com.

Case presentation: A 19-year-old male was admitted in January 2023 due to right optic neuritis without diagnostic elucidation at that time, but treated with methylprednisolone (1g/day for 5 days) with improvement. After discharge, he used prednisone for a month but discontinued on his own, resulting in visual loss in the left eye, which only improved after resuming corticosteroid therapy. In April 2023, he developed focal motor seizures on the left side with secondary bilateral dissemination, requiring intensive support hospitalization. He evolved with moderate, lobulated, exudative pleural effusion with nonspecific inflammatory characteristics, indicating serositis, and also experienced alopecia. Empirical treatment with acyclovir was initiated due to suspected viral encephalitis, even though brain MRI and lumbar puncture investigations showed no changes. During this period, bartonellosis was suspected, and serology showed positive for Bartonella quintana IgG 1:640 and negative IgM. After bartonellosis diagnosis, he was treated with doxycycline for 28 days, showing good progress with the treatment. **Discussion:** Bartonellosis is a vector-borne zoonosis with worldwide distribution, capable of infecting a wide variety of mammals, including humans. The most commonly related species to cat scratch disease (CSD) is Bartonella henselae, with cats as the main reservoir. This case illustrates a less common etiological agent, Bartonella quintana. Clinical presentations of bartonellosis can be self-limiting or severe, such as uveitis, neuroretinitis, and endocarditis. Transmission between animals and humans occurs through flea feces inoculation, scratches, bites, blood transfusions, and needles. Optic neuropathy associated with CSD is relatively rare, often characterized by sudden, painless visual loss, usually unilateral, and associated with macular star formation. Visual complications occur in about 10% of cases, including optic neuritis. The condition is self-limiting with complete or near-complete recovery of visual acuity in immunocompetent patients. Diagnosis is clinical-laboratory based, confirmed by serological testing. The antibiotic therapy used in this case was appropriate for eliminating the infection and shortening the visual recovery time. **Final comments:** Such cases should be reported to clarify the importance of early diagnosis and treatment to reduce complications, especially ocular/visual, in patients with bartonellosis. This case is particularly significant due to the less common etiological agent presented.

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Optical coherence tomography in early-onset parkinsonism

Felipe Thomas Braz[1], Max Victor Peter Schramm[1], Larissa Schneider[1], Emanuel Cassou dos Santos[1], Matheus Gomes Ferreira[1], Mario Teruo Sato[1], Naoye Shiokawa[2], Renata Cristine Alves[1], Hélio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Clínica Retina e Vítreo, Curitiba PR, Brazil.

Correspondence: Renata Cristine Alves; Email: renata_cristinealves@hotmail.com.

Background: Parkinson's disease (PD) is the main etiology of Parkinsonism and was. originally described as a motor disease. However, in recent decades, there has been better understood its non-motor component and its relevance. Among the non-motor signs and symptoms are changes in the visual processing, resulting from the loss of amacrine cells, dopaminergic neurons, in the inner retina. Thus, some studies have been demonstrated a reduction in thickness of the inner layers of the retina, where amacrine cells (dopaminergic neurons) are located. Optical Coherence Tomography (OCT) has been studied as a preclinical diagnostic tool in patients with PD, but there are still no studies on patients with Early-Onset Parkinsonism (EOP). **Objective:** To describe the retinal changes in patients with Early-Onset Parkinsonism (EOP) using Optical Coherence Tomography (OCT), to compare the thickness of retinal layers between the case and control groups, and to evaluate the correlations with disease duration, patient age, and disease severity. **Methods:** Sixteen patients with EOP had retinal layer thickness measured in the macular and peripapillary areas. The inner plexiform layer (IPL), ganglion cell layer (GCL), retinal nerve fiber layer (RNFL), total retina, and minimum rim width (MRW) were all assessed. All patients underwent neurological assessment, and the severity of symptoms classified using Part III of the UPDRS scale. **Results:** There was an increase in RNFL thickness in the Inferior Temporal sector (148.6μm vs. 133.71μm; p<0.001) in the peripapillary retina, while in the macula, there was a significant reduction of IPL in the Nasal (41.4μm vs. 48.5μm; p<0.001) and Temporal sectors (41.8μm vs. 49.9μm; p<0.001), and in the GCL in the Superior (52.7μm vs. 56.9μm; p=0.030), Nasal (52.7μm vs. 58.2μm; p=0.009) and Lower sectors (52μm vs. 58μm; p=0.002). An inverse correlation between the peripapillary MRW in the nasal sector and the severity of symptoms, evaluated by Part III of UPDRS (Spearman correlation = -0.67; p=0.033). **Conclusion:** OCT can detect the neurodegeneration present in EOP by measuring the reduction in thickness of the IPL and GCL in the macula. Furthermore, the value of nasal peripapillary MRW may be related to clinical severity of the disease. However, longitudinal studies with larger samples are necessary to establish macular measurements as diagnostic and disease progression biomarkers.

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Optimizing diagnostic pathways for myopathies with exercise intolerance: a random forest model approach

Thales Pardini Fagundes[1], Lucas Gondim Briand Vieira[1], Karolina de Brito Barbosa Trindade[1], Lucas Ravagnani Silva[1], Giuliano Roberto Scarpellini[1], Ana Marina Dutra Ferreira da Silva[1], Eduardo Yukio Nakamura[1], Raissa Gontijo Cabral de Souza[1], Pedro José Tomaselli[1], Cláudia Ferreira da Rosa Sobreira[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Thales Pardini Fagundes; Email: pardinithales@gmail.com.

Background: Myopathies encompass a heterogeneous group of skeletal muscle diseases with diverse clinical manifestations, including weakness, fatigue, myalgia, atrophy, hypertrophy, contractures, cramps, and myotonia. While some diseases in this group may present with relatively stereotypical clinical pictures, the spectrum of manifestations can include relatively nonspecific phenotypes, especially when fatigue and myalgia predominate. Metabolic myopathies often manifest with exercise intolerance, fatigue, and myalgia related to physical activity, whereas structural myopathies typically lead to muscle weakness. However, a pseudometabolic presentation of muscular dystrophies is well recognized and poses a diagnostic challenge. Despite advances in medical diagnostics, accurately identifying specific myopathies remains challenging, particularly when patients present with oligosymptomatic profiles. **Objective:** To develop a machine learning model to assist in the differential diagnosis of metabolic and structural myopathies in patients with exercise intolerance. **Methods:** The study cohort included 97 patients with myopathies, comprising 58 patients with structural myopathy and 39 with metabolic myopathy. Data were preprocessed, with missing values imputed by the median and features standardized. SMOTE was applied to balance the classes. A Random Forest model was trained on the balanced data and evaluated on the test set. **Results:** The Random Forest model achieved an accuracy of 72%, a precision of 86%, a sensitivity of 73%, and an F1-score of 74% on the test set. The confusion matrix revealed high precision in identifying structural myopathies, while precision was lower for metabolic myopathies. The most important variables for classification were maximum lactate concentrations, minimum creatine phosphokinase (CPK) concentrations, and the initial type of myalgia presented by the patients. The learning curve indicated good model performance and a low risk of overfitting. Analysis of raw data revealed that patients with metabolic myopathies had higher concentrations of lactate and CPK compared to patients with structural myopathies. **Conclusion:** The Random Forest model showed potential in assisting the differential diagnosis of metabolic and structural myopathies. Lactate and CPK concentrations, along with the initial type of myalgia, were the main predictors.

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Optimizing epilepsy surgery outcomes: the impact of accurate epileptogenic zone resection on clinical course and survival

Gabriel Chagas Lutfala Paulino[1], Raphael Fernandes Casseb[1], Brunno Machado de Campos[1], Marina Alvim[1], Enrico Ghizoni[1], Clarissa Lin Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Gabriel Chagas Lutfala Paulino; Email: lutfalagabriel1@gmail.com.

Background: Epilepsy is a debilitating condition characterized by recurrent, unprovoked seizures, affecting 0.5-1% of the world population. Resective surgery of the epileptogenic zone is the treatment of choice in focal epilepsies refractory to antiseizure medications. Nevertheless, the varied rates of good surgery outcomes elicit uncertainties regarding the definition of the target regions to be removed. In this study, we investigated whether cases in which regions with changes identified using EEG-fMRI and which were in fact removed during surgery presented better clinical and survival results over time. **Objective:** To characterize the clinical condition and survival curve of patients undergoing resective surgery throughout time. **Methods:** Thirty-six epilepsy patients (16 women) underwent a 3.0 T MRI scan (Philips Achieva) at Unicamp's Hospital. The anatomical postoperative T1 images were visually inspected for artifacts and then, the resective lacuna was manually segmented using MRIcron (version 1.0.20190902). To assess the correspondence between epileptiform activity and the surgical lacuna, we calculated the distance of the blood-oxygen-level-dependent EEG-fMRI activation clusters to the centroid of the lacuna mask that was segmented manually. In addition, the mean value of the distances was assessed in relation to the Engel score. We divided patients into two groups (Engel I-II and III-IV) and compared their survival curves over time. **Results:** The distances between the EEG-fMRI clusters and lacuna ranged from 27.1mm to 117.2 mm. In the group classified as Engel I-II, patients had significantly ($p<0.05$) smaller mean distances ($\bar{x}$=85.3mm, σ=17.7mm) compared to those classified as Engel III-IV ($\bar{x}$=95.4mm, σ=12.6mm). Furthermore, a cutoff distance of 90 mm revealed that patients with distances ≤90 mm had a higher percentage of Engel I-II (68.75%) than those with distances >90 mm. At 85 mm, this percentage increased to 81.81%, and at 80 mm, it rose to 83.33%. Notably, Engel I-II patients exhibited higher survival rates over 2 years when compared to Engel III-IV patients, with rates of 100% and 83.33%, respectively. **Conclusion:** The enduring correlation between reduced distances from epileptiform activity activation clusters to the surgical lacuna centroid and better Engel scores emphasizes the importance of accurately removing the epileptogenic zone for favorable surgical outcomes. This association also persists when assessing the survival curve over time, with better outcomes in patients with smaller activation-lacuna distances. However, further studies with larger sample sizes are necessary to validate these findings conclusively.

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Orthostatic headache as a presentation of jugulotympanic paraganglioma in the cerebellopontine angle: a case report

Luan Yudi Prando Kasuga[1], Melissa Caroline Godoi Prestes[1], Ádria Rodriges da Silva[1], Bruna Beppler[1], Caroline Zapelini[1], Lia Maki Hatisuka Imai[1], Gabriel Rodrigues Espelho Rossi[1], Aline Vitali da Silva[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Melissa Caroline Godoi Prestes; Email: melissa.caroline@outlook.com.

Case presentation: An 82-year-old hypertensive female patient presented with a history of progressive orthostatic headache for 3 months, which became incapacitating over the past month. She reported intense, pulsatile left hemicranial headache radiating to the left side of her face, associated with nausea, vomiting, hypoacusis, dysphagia, and decreased visual acuity. The symptoms worsened in the orthostatic position, during movement, or when lying on the left side. The initial cranial computed tomography (CT) scan was performed when the patient experienced a headache and revealed a hypodense nodular formation with contrast enhancement in the left pontocerebellar cistern and jugular foramen, associated with venous dilation of the sigmoid sinus. Due to the progression of symptoms, the patient required hospitalization. A cranial magnetic resonance imaging (MRI) scan was conducted, revealing an infiltrative lesion in the left mastoid and petroclival junction, invading the sigmoid sinus/jugular bulb and the posterior fossa, compressing the left cerebellar hemisphere and involving the left vertebral artery (measuring 4.8x 3.3 x 4.1 cm), suggestive of a Paraganglioma or Glomus Jugulotympanicum. The patient was treated with analgesics, anti-inflammatory drugs, and carbamazepine, initially responding well but later experiencing episodes of delirium and irritability. During her hospitalization, she developed multiple respiratory infections, sepsis, and clinical instability. With surgical intervention being unfeasible, priority was given to comfort measures. Palliative care was initiated, and the patient passed away one month later. **Discussion:** Paragangliomas are rare, well-vascularized, slow-growing neuroendocrine tumors that originate from the chromaffin cells of the paraganglia. These tumors can involve the skull base, commonly affecting the jugular foramen and the paraganglia associated with the tympanic branch of the glossopharyngeal nerve. They are locally invasive and can affect the lower petrous and sigmoid sinuses, the cerebellopontine angle, and the Eustachian tube. The clinical presentation varies depending on the affected region. Involvement of the jugular foramen leads to symptoms such as dysphagia, dysarthria, hypophonia, and weakness of the trapezius and sternocleidomastoid muscles. In contrast, involvement of the tympanic branch results in otalgia, hearing loss, pulsatile tinnitus, and aural fullness. Although headache is not common, it was the primary symptom in this case. The diagnosis involves correlating clinical symptoms with imaging studies: CT allows for the assessment of structural damage to the jugular foramen, while MRI typically demonstrates a "salt and pepper" appearance. **Final comments:** This challenging case of jugulotympanic paraganglioma illustrates the complexity associated with the diagnosis and management of rare and invasive neurological tumors, along with an atypical presentation characterized by secondary headache.

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Orthostatic tremor and leukoencephalopathy secondary to chronic toluene poisoning

Brenda Albuquerque de Araújo Marinho[1], Brenda Letícia Lopes Batista[1], Marcelo Ataíde de Lima[1], Mariana Holanda Gameleira[1], Larissa Albuquerque Paes de Araújo[2], Élida Cristina Nascimento do Rêgo[1], Maria Luísa Gonçalves Torres Inácio[1], Bárbara Martins de Carvalho[1], Mateus de Melo Andrade[1], Leandro Correia Gonçalves de Souza[1]

[1]Hospital da Restauração, Recife PE, Brazil. [2]Faculdade de Ciências Médicas da Paraíba, João Pessoa PB, Brazil.

Correspondence: Brenda Albuquerque de Araújo Marinho; Email: brenda.albuquerque65@gmail.com.

A 44-year-old female patient presents with ambulation problems due to tremors over the past three months, accompanied by double vision for the last seven days. Tremors have persisted for over 20 years, but recently, walking difficulties and double vision have worsened. She denies other neurological symptoms, current or past alcoholism, and smoking. She reports long-term use of "glue" (an illicit substance containing toluene) for 30 years. Neurological examination is normal except for tremors in the face and trunk while sitting, appendicular tremors in upright posture, and no tremors when supine. Several complementary tests were conducted, including MRI of the skull revealing diffuse white matter signal alterations in the corona radiata, centrum semiovale, splenium of the corpus callosum, middle cerebellar peduncles, and bilateral dentate nucleus of the cerebellum, with findings of undetermined nature. Given the history of solvent abuse, toluene-related leukoencephalopathy is considered. Electroneuromyography (ENMG) demonstrated postural action tremor with a frequency of approximately 13 Hz in the left tibial muscle, absent in supine and sitting positions. Peripheral nervous system abnormalities were not observed on ENMG. The diagnosis of orthostatic tremor and leukoencephalopathy secondary to chronic toluene poisoning was confirmed. The treatment plan includes discontinuation of "glue" use, psychiatric monitoring, psychotherapy, motor physiotherapy, and pharmacological therapy (Benzodiazepine + Amantadine), with a follow-up in 30 days. Tremor is the most common movement disorder, characterized by rhythmic oscillation due to contractions of antagonistic muscles, typically confirmed by electromyography (EMG). Long-term toluene inhalation can lead to neurological symptoms including cerebellar, cognitive, and pyramidal dysfunctions, as well as optic atrophy, visible on radiological studies. Benzodiazepines have shown efficacy in managing toluene-induced tremors. The main feature of toluene-induced leukoencephalopathy is progressive dementia, correlating with the extent of white matter damage. Additional central nervous

system deficits may include cognitive, cerebellar, and cranial nerve dysfunctions.

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Osteitis due to neurosyphilis: a case report

Luiz Felipe Mota Freitas[1], Fernanda Vieira[2], Rafael Barbosa Mokdeci Surerus[3], Daniel Sabino de Oliveira[1], Leopoldo Antonio Pires[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Hospital Universitário, Ebserh, Juiz de Fora MG, Brazil.
[2]Centro Universitário de Volta Redonda, Volta Redonda RJ, Brazil.
[3]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Luiz Felipe Mota Freitas; Email: luizfelipemotafreitas@gmail.com.

Case presentation: A 21-year-old female patient with a history of chronic tension-type headaches with irregular medical follow-up presented with an elevated and bulging lesion in the frontal region of the skull associated with more intense headache that became refractory to her usual analgesics. She denied any local trauma. Examination revealed a painless, mobile and fibroelastic cervical lymphadenopathy in the posterior chain on the left side. There were no lesions in the oral cavity or skin neither neurological abnormalities or neck stiffness. Brain MRI showed a lesion involving the left frontal bone marrow. Bone scintigraphy revealed a focal increase in osteogenic activity of significant intensity in the frontal region of the skull. CSF analysis showed 6 cells/mm3, with lymphocyte predominance, glucose 51mg/dL (serum: 89mg/dL), proteins 9.53, and a non-reactive VDRL in CSF. Cytopathological examination was normal. Serum tests showed VDRL 1:256 with reactive serology for Treponema pallidum IgM and IgG. Serologies for viral hepatitis and HIV were negative. As CSF showed a slight increase in cells with a lymphomonocytic predominance, we diagnosed an asymptomatic neurosyphilis and treated with intravenous crystalline penicillin for 14 days. There were no complications, maintaining clinical stability throughout hospitalization and the clinical picture improved. Primary bone lesions were ruled out, and the lesion was associated with syphilitic osteitis. Post-treatment tests: serum VDRL 1:2; serology for Treponema pallidum IgM NR, IgG reactive. Post-treatment MRI: near-complete resolution of the permeative lesion involving the left frontal bone marrow. **Discussion:** Treponema pallidum, a gram-negative spirochete bacterium, has an affinity for bone tissue, most commonly affecting long bones and the skull. Neurosyphilis can develop by contiguity from bone lesions of the calvaria and should be ruled out when close to the meninges. Syphilis cases have increased worldwide, with higher incidence among men who have sex with men and HIV patients. Bone involvement is uncommon in primary and secondary syphilis, with a prevalence of 0.15 to 0.23%. The presence of mucocutaneous findings and lymphadenopathy raise suspicion of syphilitic osteitis. There is no consensus on the treatment of secondary syphilis osteitis cases, as they are rare. Symptoms resolve after therapy, but bone lesions may persist for up to 7-11 months. **Final comments:** Syphilitic osteitis is more frequently associated with tertiary or congenital syphilis. It should be considered in the differential diagnosis of bone lesions when accompanied by positive serology for syphilis.

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Outcomes in pediatric surgery for drug-resistant epilepsy: a systematic review

Leonardo Portugal Lima[1], Beatriz Caroline Pazzini de Almeida[1], Gabriel Barreto Vinhas Abreu[2]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Leonardo Portugal Lima; Email: leonardo_mali@hotmail.com.

Background: Epilepsy is a chronic nervous system disease characterized by fluctuations between moments of stability and instability within nerve cells, which presents in forms of epileptic seizures. Most cases of epilepsy are controlled by Anti-seizure medication (ASM), but up to 30% can be drug-resistant, posing a higher risk of psychological and neurological impairment in children. In such cases, surgery presents itself as a therapeutic possibility. **Objective:** To conduct a systematic review on current literature of surgical outcomes in epileptic pediatric patients, with focus on efficacy, safety and long-term follow-up. **Methods:** This is a systematic literature review based on articles indexed in the United States National Library of Medicine (PubMed) published between 2019 and 2024. Were used "epilepsy", "surgery" and "pediatric" as a descriptor. Initially, 57 articles were found of which 15 were used after applying an inclusion and exclusion criteria. **Results:** From the articles analyzed, all being systematic review and/or meta-analysis, were mainly composed of long-term follow up of patients as long as 10 years post-operatively, the full-scale intelligent quotient (FSIQ) and neurocognitive academic abilities assessment in pre and post procedure and comparisons with non-surgical groups. **Conclusion:** The review demonstrated that surgical intervention shows significant efficiency in improving quality of life and reducing seizure frequency in pediatric drug resistant epilepsy, with high percentage remaining seizure-free in a long-term follow-up. Among the patients, seizure-freedom was even more strongly correlated with shorter epilepsy duration, highlighting the importance of considering surgery early in those patient's treatment process. Moreover, it was documented that after the procedure, patients mostly surpassed or maintained their neurocognitive functions, with a minority experiencing slight neurologic deficit, most of which were transient. Furthermore, there was no observed decline in core academic skills and in the FSIQ evaluation, and individuals were able to keep their preoperative performances, underscoring the safety implications of the procedure. By consolidating the current literature, it illustrates valuable insights and establishes a solid foundation for the significance of early referral to surgery and a personalized treatment approach in pediatric patients with drug-resistant epilepsy.

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Outlining the epileptogenic zone with cortical stimulation in SEEG

William Alves Martins[1], Claude Steriade[2], Imad Najm[3], Juan Bulacio[3]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre SC, Brazil.
[2]New York University, Langone Health, New York, USA.
[3]Cleveland Clinic, Ohio, USA.

Correspondence: William Alves Martins; Email: walvesm.br@gmail.com.

Background: Mapping of neurological function by cortical stimulation is an essential step in the evaluation of epilepsy patients undergoing invasive EEG for epilepsy surgery. Yet, controversy still exists regarding its role as a tool for delineating epileptogenic cortex. **Objective:** To evaluate the impact of electroclinical patterns of cortico-stimulation induced seizures (CIS) in stereo-electroencephalography (SEEG) and removal of CIS electrodes in seizure freedom. **Methods:** We reviewed all 230 consecutive patients who underwent SEEG and cortical stimulation from 2013 to 2016. CIS were induced in 79 (30.4%) patients, of whom 49 (21.2%) underwent surgical resection and entered the final analysis. Demographic, epilepsy and pathological data were collected. In addition, a detailed analysis of cortical stimulation and Stereo-EEG was performed. For the purpose of our study, we created a simple way to classify CIS based on semiological (complete, partial and atypical patterns), and EEG similarity to spontaneous seizures. Seizure freedom was used to calculate the predictive value of CIS with complete semiology and identical EEG (complete electroclinical picture). **Results:** Seventy-one (45.8%) patients were seizure-free with a mean follow up of 1.42±0.9 years. Twenty-six (52%) patients in the CIS group were seizure-free compared to 45 (42.9%) non-CIS patients (p=0.286). Complete seizure semiology was obtained in 22 (42.9%) patients and resection of electrodes involved in CIS led to seizure freedom in 73.7% compared to 45.9% of the stimulation cohort (p=0.021). Conversely, resection of electrodes involved in partial seizure semiology provided a rate of seizure-freedom of only 20% (p=0.021). Resection of electrodes involved in CIS identical to spontaneous seizures (both semiology and EEG) were significantly associated with seizure-freedom (80% x 42.4%; p=0.006, OR 5.4, 95% CI 1.46-20.0) and largely represented by FCD type 2 (42.9%; Fisher's exact test, p=0.006). Complete electroclinical correlation of CIS displayed sensitivity of 16.9% (95% CI, 9.05-27.66%), specificity of 96.4% (95% CI, 89.8-99.25%) and positive predictive value of 80% (95% CI, 54.03-93.16%) for accurate localization of epileptogenic zone. **Conclusion:** Electroclinical characteristics of CIS are a valuable tool to localize the epileptogenic zone and craft a successful surgical strategy in SEEG. Moreover, it may represent a marker of underlying FCD type II.

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Overdiagnosis of normal pressure hydrocephalus: evidence of clinical-radiological dissociation

Ana Carolina Gomes[1], Sephora Sabrina Candido de Almeida[1], Diogo Haddad Santos[1], Yngrid Dieguez Ferreira[1], Alexandre Shoji[1], Isabela Martins de Freitas[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Ana Carolina Gomes; Email: gomesac90@gmail.com.

Background: Normal pressure hydrocephalus (NPH) was described in 1965 by Hakim and Adams and is characterized by the classic triad of cognitive decline, gait apraxia, and urinary incontinence associated with ventriculomegaly on neuroimaging, with normal opening cerebrospinal fluid (CSF) pressure. Its diagnosis is challenging due to its similarity with other neurodegenerative diseases, such as Alzheimer's and vascular dementia. Additionally, there is a noted clinical-radiological dissociation, often leading to the overdiagnosis of the condition. In this context, there has been an increase in the referral of patients with suspected NPH to the Centro de Cognição Paulista (CECOP). **Objective:** This study measures the dissociation between clinical and radiological findings in patients evaluated between January 2022 and December 2023, as well as the overdiagnosis of NPH and the need for revision of diagnostic criteria. **Methods:** A retrospective analysis was conducted on 46 patients referred to CECOP with suspected NPH based on neuroimaging. The presence of imaging findings compatible with NPH, age, gender, clinical symptoms, and final outcome were evaluated. **Results:** Of the 46 patients, 25 had a clinical presentation compatible with NPH (12 men and 13 women, mean age 75 years) but without characteristic neuroimaging. Of these, 14 were diagnosed with Alzheimer's, 5 with vascular dementia, 3 with frontotemporal dementia, and 3 remain under investigation. Only 7 patients (5 men and 2 women, mean age 72 years) had both clinical and imaging findings consistent with NPH and were referred for a tap test. Another 7 patients, despite having clinical

and neuroimaging suggestive of NPH, had more probable other diagnoses. **Conclusion:** The analysis revealed a significant clinical-radiological dissociation, with only 7 patients (15.22%) presenting both clinical and imaging findings compatible with NPH. Of the 25 patients with neuroimaging alterations, 54.3% did not have clinical indications of the condition and received other diagnoses. These findings highlight the complexity of diagnosing NPH and the importance of rigorous criteria. Although the tap test is an important step in evaluation, the latest Japanese consensus considers that patients over 60 years old with compatible clinical and neuroimaging findings can be diagnosed with NPH without the need for a tap test. Furthermore, correlation with CSF biomarkers can help differentiate NPH from other dementias, as NPH typically shows low levels of Aβ42 and pTau in the CSF, contrasting with patients with Alzheimer's. The use of the Radscale Score, which quantifies the severity of neuroradiological changes, can increase diagnostic accuracy of NPH and be an ally in practice. Thus, there is a need for an amplified diagnostic approach: detailed clinical evaluation, CSF biomarkers, and standardized radiological criteria to improve the accuracy of NPH diagnosis and differentiate it from other forms of dementia.

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Overdrainage syndrome after idiopatic intracranial hypertension: a case report

Thaís dos Santos Scheid[1], Breno Rampeloti[1], Giuliana Moro[1], Caio de Lima Ferreira[1], Mayara Tszesnioski Maçaneiro[1], Laise Koenig de Lima[2], Natália Cardoso Darabas[2], Thabata Emanuelle Martins Nunes[2], Felipe Ibiapina dos Reis[1]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Thais dos Santos Scheid; Email: scheidthais@gmail.com.

Case presentation: A female patient, 47 years old, was admitted to the hospital with progressive right-sided amaurosis and severe visual loss. Physical examination showed a hyperemic optic disc with blurred edges, microhemorrhages and peripapillary exudates associated with macular edema from intraretinal fluid with attenuation of the foveal depression. The neuroimaging showed a partially empty sella and dilated optic nerve membranes, suggestive of idiopathic intracranial hypertension, as well as cerebrospinal fluid (CSF) opening pressure examination in spinal cord above 400mm/H2O. A lumboperitoneal shunt with a high-pressure device was performed 5 days after hospital admission, with significant visual improvement without complications. One month later, the patient was readmitted due to a history of falling from her own height, headache and persistent vomiting for 1 week. The examination revealed drowsiness, dysarthria, paresis of the lower limbs, esotropia in the right eye, papilledema and bilateral retinal detachment. Computed tomography showed erasure of cerebral cisterns, hyperdensity in the subarachnoid space and hypodensity in the bilateral cerebellum and pons. The patient was taken to the operating room, where was found ribbon ventricles, a posterior fossa with tonsillar herniation and a functioning shunt on digital pressure and test puncture. These findings are compatible with probable overdrainage despite the high-pressure shunt and it was decided to close the emergency valve. After the surgical approach, symptoms progressively improved and without loss of vision. **Discussion:** Idiopathic intracranial hypertension is a rare condition and without known cause that mainly affects young obese women. Clinical manifestations include chronic migraine-like headaches, progressive visual deterioration, and pulsatile tinnitus. The diagnosis must meet all five criteria: papilledema; no neurological deficit; no cause shown on imaging; normal cerebrospinal fluid; and elevated lumbar puncture opening pressure (greater than 250 mm/H2O) under proper puncture conditions. Imaging studies can suggest intracranial hypertension, but never used alone for diagnosis. Treatment should be aimed at treating the underlying cause, protecting vision and reducing headache morbidity. Overdrainage is an excessive drainage of cerebrospinal fluid, usually associated with valved shunt systems. In terms of management, replacement or closure of the valve usually resolves the clinical presentation. **Final comments:** The idiopathic intracranial hypertension presented by the patient in this report was well managed clinically and surgically in this case according to recent scientific recommendations and guidelines. The development of overdrainage syndrome, a complication of lumboperitoneal shunting, can occur especially in cases where the surgical approach was an emergency, as in the present report.

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Overlap of Guillain-Barré syndrome variants: a case report

Marco Antônio Delazeri[1], Carollina Danezi Felin[1], Guilherme Vanik Pinto[1], Eduarda Rosa Fernandes[1], Carolina Heck Haubrich[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Marco Antônio Delazeri; Email: marco.delazeri@outlook.com.

Case presentation: A 63-year-old man, previously diabetic, hypertensive and with coronary artery disease, presents a subacute condition of paresthesias with an ascending and progressive onset, distal to proximal, grossly symmetrical, accompanied by paresis of the same characteristic. Evolving concomitantly with dysphagia, dysarthria and hypogeusia. The neurological examination confirmed the aforementioned condition, also showing facial paralysis with a peripheral pattern on the right, global areflexia and gait ataxia. Normal sensitivity. Computed tomography and magnetic resonance imaging of the brain are normal. Liquor: albuminocytologic dissociation. Electroneuromyographically compatible with demyelinating sensorimotor polyneuropathy in all four limbs, suggestive of Guillain-Barré Syndrome. The patient progresses with partial and spontaneous resolution of the condition, leaving only subsequent paresthesias and facial paresis. **Discussion:** Guillain-Barré syndrome or acute inflammatory polyradiculoneuropathy is a disease of the peripheral nervous system, mostly monophasic and predominantly preceded by vaccination or infectious conditions. Its clinical presentation is vast, however it classically presents as bilateral and symmetrical ascending flaccid paresis, distal to proximal, reaching its symptomatic nadir in two weeks. Considering its clinical variability, variants of the same disease were determined. In the report above we present a curious intersection of variants: acute inflammatory desmyelinating polyneuropathy (AIDP) classic and Miller-Fisher. It has a classic triad: gait ataxia, global arephexia and ophthalmoparesis. In this report, antibodies such as GTA1 and GQ1B could help to differentiate and classify it more precisely. Typically, it is treated with plasma exchange or immunoglobulin. It is important to assess respiratory dysfunction, with the option to use the Erasmus GBS Respiratory Insufficiency Score (EGRIS) to predict the probability of respiratory failure within the first week of admission in patients with the syndrome. **Final comments:** Guillain-Barré syndrome is a disease with a wide clinical presentation and potentially unfavorable outcomes if not promptly recognized and treated. Its variations are not always pure and can intersect with others, which is why we report this case.

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Overlap of neuroectodermal syndromes: Sturge-Weber syndrome, Klippel-Trenaunay syndrome, and pigmentary vascular phakomatosis in a pediatric patient - a case report

Lourdes Helena Rabelo Dias[1], Luiz Augusto Fernandes da Silva[1], Cecília Secchin de Jesus[1], João Pedro Cuelhar Rodrigues[1], Marcela Lopes de Almeida[1], Bruno Antunes Contrucci[2], Ursula Thomé[2], Maria Karolina Arantes Assis Castro[1], Igor Mota Andrade[1], Maria Fernanda Alli Ahmad[1]

[1]Centro Universitário Estácio IDOMED, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, Hospital das Clinicas de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Luiz Augusto Fernandes da Silva; Email: fernandeslaugusto@gmail.com.

Case presentation: Female, four years old, non-consanguineous parents, uneventful pregnancy and perinatal period, was referred due the hypothesis of Sturge-Weber syndrome (SWS). Since birth, body spots have been observed, evolving with focal epilepsy starting at five months of age, with daily seizures despite regular use antiepileptic medication (carbamazepine, phenobarbital and lamotrigine). Left hemiface with "port-wine stain" and violet spots scattered in the right frontal region, right hemibody, upper left hemibody, posterior region, genitals, and lower limbs; slight soft tissue enlargement on the right; eyebrow thinning; bilateral congenital glaucoma and developmental delay was presented. Cranial CT scan at 12 months showed right parietal volume increasing suggestive of hematoma. EEG at two years revealed diffuse asymmetric encephalopathy on the left cerebral hemisphere and focal base epilepsy. MRI and angiotomography suggested SWS, with atrophy and probable pial angiomas, cortical-subcortical calcifications on the left, choroid plexus prominent, and bilateral deep venous drainage, especially on the left, diagnostic hypothesis of Pigmentary Vascular Phakomatosis, SWS and Klippel-Trenaunay syndrome (KTS). **Discussion:** The overlap of neuroectodermal syndromes as observed in this case, involves multiple complex anomalies. SWS is the third most common phakomatosis, characterized as neurocutaneous disease with angiomas on the face, choroid, and leptomeninges due to somatic or mosaic mutations in GNAQ gene. Pathophysiology involves deficiency of superficial cortical venous circulation, leading to stasis and ischemic changes. Manifestations include various epileptic seizures, plegias, intellectual disability, glaucoma, and dermal capillary malformations resulting in port-wine stain. KTS is rare congenital mesodermal disease of undefined etiology, characterized by port-wine stain, venous malformations and hypertrophy of bones and soft tissues, usually affecting a single lower limb but can involve trunk and face. The overlap of SWS and KTS increases the phlebitis, bleeding, deep vein thrombosis, pulmonary embolism, hemoperitoneum, hemothorax, and chronic venous insufficiency risks. Hemangiomas and varices are usually presented at birth and become more prominent during adolescence. Venous abnormalities include agenesis, hypoplasia, atresia, valvular incompetence, and deep venous system occlusion due to fibrosis. Pigmentary Vascular Phakomatosis Cesio-Marmorata, vascular malformation with systemic manifestations, affects vision associated with various systemic and cutaneous abnormalities, including Sturge-Weber and Klippel-Trenaunay syndromes. The overlap of these neuroectodermal syndromes highlights the need of deeper studies for better etiological diagnosis and viable therapies. **Final comments:** Our case contributes to scientific knowledge of these rare neurocutaneous diseases, aiding in management and definition.

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Overlap of neuromyelitis optica spectrum disorder and systemic lupus erythematosus: a case report

Alexandre Baldissera[1], Marcos Madeira de Lima[1], Eduardo Drews Amorim[1], Alessandro Finkelsztejn[1], Renata Gomes Londero[1]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Eduardo Drews Amorim; Email: eduardodrewsamorim@gmail.com.

Case presentation: A 34-year-old woman with a history of Systemic Lupus Erythematosus (SLE) and popliteal vein thrombosis on warfarin presented with paraparesis, hypoesthesia, and urinary incontinence for about a week. One day before arriving at the emergency department, her condition progressed to paraplegia. Cervical and thoracic spine MRI demonstrated longitudinally extensive myelitis from C7 to D11. There was no evidence of infection, and pulse therapy with methylprednisolone 1g was initiated. After two days of pulse therapy, the patient developed a fever spike and productive cough. A lumbar puncture performed two days after pulse therapy (previously contraindicated due to coumarin intoxication) diagnosed probable meningoencephalitis associated with autoimmune myelitis. Antibiotic therapy was initiated for pulmonary and neurological coverage, and pulse therapy was suspended (immunosuppression with methylprednisolone 1mg/kg was maintained). An autoantibody test revealed positive anti-aquaporin-4 (anti-AQP4), diagnosing Neuromyelitis Optica Spectrum Disorder (NMOSD) with positive anti-AQP4. After controlling the infectious condition, plasmapheresis was performed along with the resumption of pulse therapy, resulting in the return of tactile sensation in the lower limbs. The patient continued treatment with Rituximab as an immunomodulatory therapy. She was diagnosed with longitudinally extensive myelitis due to the overlap syndrome of NMOSD and SLE complicated by meningoencephalitis. **Discussion:** The patient presented a complex clinical picture of progressive paraparesis that evolved into paraplegia, associated with active SLE. MRI revealed longitudinally extensive myelitis, a distinctive feature of NMOSD. The presence of anti-AQP4 antibodies was crucial for diagnosing NMOSD, a condition that often overlaps with other autoimmune diseases such as SLE. The overlap syndrome of NMOSD and SLE presents a significant challenge in diagnosis and treatment. The co-occurrence of these diseases can result in atypical clinical presentations and additional complications, such as secondary infections and variable responses to immunosuppressive treatment. The therapeutic approach for patients with this overlap syndrome must be carefully planned to address both autoimmune activity and associated complications. **Final comments:** This case highlights the importance of considering complex differential diagnoses in patients with SLE and severe neurological manifestations. The overlap syndrome of NMOSD and SLE requires a multidisciplinary approach for effective management. Early identification and appropriate treatment, including the use of intensive immunomodulatory therapies and management of infectious complications, are essential to improving the prognosis and quality of life of patients.

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Overlap syndrome: systemic lupus erythematosus and dermatopolymyositis

Yasmin de Freitas Santos[1], Luis Fernando Guimarães[1], Marianna Lessa Coelho Vieira de Quadros[2], Rebecca Alves Marinho[2], Esther Alves Marinho[2], Júlia Rodrigues de Senna Mendonça[2], Osvaldo Pantoja de Oliveira[1], Marina Brasileiro Cesar Leitão[1], Luiz Guilherme Gerhardt Fernandes[1], Maria Luiza Levindo Coelho Martinis[1]

[1]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.
[2]Universidade Estadual de Montes Claros, Montes Claros MG, Brazil.

Correspondence: Luis Fernando Guimarães; Email: luisguimaraesneuro@gmail.com.

Case presentation: A female patient, 14 years old, presented prostration and asthenia, fifty days prior to admission to the hospital . She evolved with night fever (38.3), weight loss of 06 kg, diffuse alopecia,proximal weakness of the upper and lower limbs and hyperemic, non-pruritic papules on the limbs and trunk that have evolved into plaques. These lesions were not influenced by sun exposure. The patient has no previous history of comorbidities or rheumatic diseases in the family. Her neurological examination had the following findings: predominantly proximal tetraparesis in the upper (4/5) and lower (5/5) limbs; arthritis in wrists, ankles and knees; heliotrope bilaterally; erythematous plaques in the trunk region, hypophonetic sounds on cardiac auscultation and mild splenomegaly. Laboratory examination indicated high muscle injury markers, positive antibodies, among others (Table 01); Doppler echocardiogram with signs of pericarditis and an electroneuromyography with a myopathic pattern. The muscle biopsy showed skeletal striated tissue with atrophy, edema, discrete mononuclear infiltrate, and the presence of associated focal necrosis. The diagnosis of SLE Overlap and DPM was considered, and then prednisone, hydroxychloroquine and methotrexate were started, with significant clinical improvement. The patient remains under outpatient rheumatology follow-up. **Discussion:** The overlap syndrome (Overlap) is characterized by the presence of two or more autoimmune diseases in the same patient, following the formal criteria for the individual diagnosis of each one. Overlap in Systemic Lupus Erythematosus (SLE) and Dermatopolymyositis (DPM) is rare, there no data mentioning its incidence in adolescents. Cases reports of patients with SLE and Myositis, regardless of the presence of dermatological alterations are described with an incidence between 4% and 16%, without analyzis of a specific age group. Overlap of SLE and DPM will be reported in a 14-year-old patient. **Final comments:** The Overlap between SLE and DPM is rare and the context of overlapping of juvenile DPM and SLE is even rarer. The importance of electroneuromyography is emphasized as an important complementary test to confirm myopathic impairment with classic findings of the syndrome on examination.

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Overview of admissions and deaths due to meningococcal infection in patients aged 0 to 14 in the Southeast between 2018 to 2023

Maria Luiza Leme Camargo[1], Maria Clara Caparroz Cassioti[1], Danielli Casonato da Silva[1], Marcela dos Santos de Deus[1], Mariana Aires Silva[1], Luah da Silva Ishikawa Manhani[1], Jéssika Yumi Taba Ono[1], Luana Fornetti Castilho Cazani[1], Pedro Keemper Rocha Alpino[1], Maiara Silva Tramonte[1]

[1]Universidade Nove de Julho, Bauru SP, Brazil.

Correspondence: Maria Luiza Leme Camargo; Email: maria.luiza.camargo@uni9.edu.br.

Background: Acute bacterial meningitis is defined as an infectious process affecting the leptomeninges and the subarachnoid space. It is primarily caused by the agent Neisseria meningitidis, transmitted through prolonged and close contact with the patient's nasopharyngeal secretions. In Brazil, this infection presents a 20% mortality rate in confirmed cases. Given its high mortality and morbidity, coupled with childhood vaccination schedules, recognizing symptoms and mapping the epidemiological profile from 0 to 14 years old in the Southeast becomes crucial for better directing prevention efforts and public health policies. **Objective:** Analyzing the epidemiological profile of patients aged 0 to 14 with meningitis in the Southeast. **Methods:** This is a descriptive, retrospective, and quantitative epidemiological study, analyzing the period from 2018 to 2023 in the Southeast region of Brazil. Data were obtained from the Hospital Information System (SIH), accessible through the Department of Informatics of the Unified Health System (DATASUS). The analysis considered variables such as sex, age range from 0 to 14 years, number of hospitalizations, and number of deaths related to meningococcal infection in childhood. Data collection was conducted in April 2024. **Results:** A total of 807 hospitalizations due to meningococcal infection were reported during the analyzed period and location. Hospitalizations showed a decreasing trend from 2018 (211) to 2021 (70), with an increase in 2022 (96) and 2023 (138). Males accounted for 54.52% of notifications. Regarding age group, children aged 1 to 4 years were most affected (290), followed by those under 1 year (249), 5 to 9 years ranked third (170), and the least affected among the age groups analyzed were those aged 10 to 14 years (98). There were 69 deaths from 2018 to 2023 in the 0 to 14 age group, with infants under 1 year being the most affected (24), followed by those aged 1 to 4 years (22). **Conclusion:** A declining trend in hospitalization rates due to meningococcal infection in the 0 to 14 age group was observed until 2021. However, from this point onwards, there was a significant increase in 2022, followed by an even more pronounced increase in 2023. This pattern shift suggests the urgent need for review of prevention and control strategies, especially in the Southeast region. The implementation of effective measures, such as vaccination programs, becomes crucial to reverse this worrying trend and mitigate the impact of meningococcal infection on the population in question.

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Overview of bacterial meningitis in brazil: analysis of variables in the last five years

Geovanna Barros Rocha[1], Sávio Roberto Silva Costa[1], Lucas Miquilini de Arruda Farias[1], Douglas Ferreira Pereira[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Geovanna Barros Rocha; Email: geovannaroccha@outlook.com.

Background: Bacterial meningitis is defined as an inflammation of the meninges of infectious origin, commonly caused by respiratory secretions, however, whose access routes to the meninges are hematogenous, due to contiguity or direct access. The etiological agent is linked to factors such as age, vaccination status, site of infection, host susceptibility and other mechanisms, however, Neisseria meningitidis, Haemophilus influenzae and Streptococcus pneumoniae are most commonly found. It is an acute and serious notifiable disease that, despite the lower incidence following advances in immunizations, maintains high rates of morbidity and mortality and motor, sensory and cognitive sequelae. **Objective:** To analyze trends in bacterial meningitis in Brazil and associated factors, from 2019 to 2023, to guide better prevention and treatment strategies. **Methods:** This study adopts an observational, cross-sectional and descriptive approach, using quantitative data obtained from the Diseases and Notifiable Diseases system (SINAN), made available by the Department of Informatics of the Unified Health System (DATASUS). The variables considered include sex, race, age group, region of residence, etiology and clinical evolution. Data analysis was performed using Biostat 5.3 software. **Results:** During the observed period, there were 59,787 confirmed cases of meningitis in Brazil, with the majority of cases occurring in the southeast region, with 52.2%, while the central-west region reported fewer cases (4.3%). Regarding gender, 58.4% were male. In relation to ethnicity, the most affected ethnicity was white, with 47.31% of cases, while the least affected ethnicity was indigenous, with 222 cases. Regarding age, the most affected age group was between 20 and 59 years old, with 36.3%. Regarding the type of meningitis, the majority of registered cases were viral meningitis, with 43.9%, followed by unspecified meningitis, with 18.2%. The vast majority

of cases evolved with the patient's discharge, with 75.6%, while there were around 10% of deaths due to meningitis in the period studied. **Conclusion:** It was observed that, in the last 5 years, the majority of meningitis cases occurred in the southeast region, with males being slightly more affected. The most affected ethnic group is white. Regarding age, the most affected group was between 20 and 59 years old. The majority of cases were viral meningitis, with the vast majority of cases being discharged from hospital. Therefore, it is important that prevalent epidemiological factors are taken into consideration in order to plan strategic and effective actions to reduce the occurrence of meningitis in the country.

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Overview of childhood stroke by regions of Brazil in the pre- and post-COVID-19 pandemic periods

Paula Cely da Silva Torres[1], Karollyne de Castro Santos[1], Alexandre David do Nascimento Santos[1], Ester Costa Lima[1], Fabrício Rodrigues Barbosa[1], Victor Alex Purificação de Castro Neves[1], Paulo Vitor Silva Santos[1], Bruna Rafaela Bastos Freite Quixabeira[1], Jéssica de Andrade Ribeiro Lima[1], Lucas Granja Kaupert[1]

[1]Universidade do Estado da Bahia, Salvador BA, Brazil.

Correspondence: Ester Costa Lima; Email: ester.enfe@gmail.com.

Background: Stroke in the pediatric population is a rare neurological disorder, however, it is associated with more severe outcomes, being among the ten main causes of long-term neurological, motor and cognitive impairments, contributing to significant morbidity and mortality. In the context of the COVID-19 pandemic, it has been suggested that coronavirus infection is linked to an increased risk of stroke in childhood, leading to substantial economic costs for society. **Objective:** To evaluate the epidemiological profile and costs of hospitalizations for stroke in children aged 0 to 19 years in Brazil, in the pre- and post-COVID-19 pandemic periods. **Methods:** Ecological study using SIH/DATASUS data on ischemic and hemorrhagic stroke. The period analyzed was from January 2018 to December 2023. Data were collected on the number of hospitalizations, type of care, and total costs according to place of residence. Microsoft Excel® 2016 software was used for data analysis. The variables analyzed were: year of hospitalization, sex, color/race, age group, mortality rate, and regions of the federation. **Results:** During the period analyzed, there were 4621 hospitalizations in Brazil. Among the regions, the Northeast stood out with the highest number of hospitalizations (36.36%), followed by the Southeast (34.75%) and South (13.44%). It was observed that the years with the most cases were 2022 (18.18%) and 2023 (18.03%), while the years 2020 and 2018 obtained the lowest values (13.78% vs. 15.75%), highlighting a considerable variation, probably linked to the post- and pre-pandemic context. Of the services listed, most were emergency (92.21%). There was no significant discrepancy regarding gender, with females being only slightly higher than males (50.36% vs. 49.64%). Regarding color/race and age group, brown individuals (48.17%), between 15 and 19 years old (51.72%) were the ones with the highest number of hospitalizations. The total value of the services provided in the period was R$9,150,002.66. Among the regions of the country, the Southeast was the one with the highest expenses (R$3,569,781.11) and the year with the highest costs was 2023 (R$1,777,286.28). The national mortality rate corresponded to 7.88%, with its peak observed in 2021 (8.63%) and 2022 (8.21%) and the lowest value found in 2018 (7.14%), in the pre-pandemic period, mainly affecting children under 1 year of age (13.31%). **Conclusion:** It is noted that the profile of children hospitalized for stroke in the pre- and post-pandemic period has undergone considerable variations, with an increase in cases registered in the post-pandemic period, especially in the Northeast and Southeast regions, in individuals aged 15-19 years. In addition, there was an increase in costs during the period, and in the mortality rate in the years 2021 and 2022, reflecting the possible increase in notifications and identification of suspected cases, concomitantly with the decline of the pandemic.

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Overview of Parkinson's hospitalizations in Brazil from 2019 to 2023

Aline Teixeira[1], Ana Clara Lacerda Freitas[1], Laysa de Souza Maia[2], Andressa Gonçalves Vicente[3], Ana Alice Lemos Lima[4], Gabriela Hernandez Dumani[5], Ana Cristina Carvalho e Silva[3], Estefany Moreno de Souza[6], Geovana Franklim Gomes e Silva[7], Juliana Braga Rodrigies de Castro[8]

[1]Centro Universitário Funorte, Montes Claros MG, Brazil.
[2]Faculdades Integradas Aparício Carvalho, Jaru RO, Brazil.
[3]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[5]Universidade Franciscana, Santa Maria RS, Brazil.
[6]Universidade Aquino de Bolivia, Santa Cruz de La Sierra, Bolivia.
[7]Centro Universitário de Patos de Minas, Patos de Minas MG, Brazil.
[8]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Laysa de Souza Maia; Email: laysamaia1503@gmail.com.

Background: Parkinson's disease is a neurological disorder of poorly understood and multifactorial etiology, characterized by the degeneration of dopaminergic neurons located in the substantia nigra of the brain. These cells are responsible for the production of the neurotransmitter dopamine, which is essential for the conduction of nerve impulses throughout the body. Deficiency or reduction of dopamine compromises bodily movements, resulting in tremors, bradykinesia, muscle rigidity, imbalance, and changes in speech and writing. **Objective:** To analyze the profile of hospital admissions for the treatment of Parkinson's disease throughout Brazil over the past five years. **Methods:** This is an observational, descriptive, and cross-sectional study using secondary data on Parkinson's disease available from DATASUS - the Hospital Information System of the Brazilian Unified Health System (SUS), covering the period from January 2019 to December 2023. **Results:** Between 2019 and 2023, there were 4,399 hospital admissions for Parkinson's disease. The highest number of cases was recorded in 2023, with 1,099 cases, while the lowest occurrence was in 2021, with 747 cases. In 2020, there was one admission per 100,000 inhabitants, possibly influenced by the COVID-19 pandemic, suggesting potential underreporting. During the study period, 345 people died due to Parkinson's disease. The highest incidence rates were observed in 2023, 2019, and 2022, with 0.47, 0.54, and 0.46 respectively. The highest incidence rates were recorded in the Southeast and South regions of the country. The average incidence rate was 0.43% and the mortality rate was 0.03%. The case fatality rate was 7.87%. The Southeast region recorded the highest incidence rate of 0.98% and mortality rate of 0.07%. The highest case fatality rate was in the Northeast region (9.62%), while the South region had the lowest values. Regarding gender, males were the most affected, accounting for 2,646 admissions (60.15%). The ethnicity with the highest self-reported hospital admissions was White, with 2,273 admissions (51.67%). The majority of hospital admissions were among individuals aged 60 to 69 years, responsible for 2,760 admissions (26.82%), followed by those aged 70 to 79 years, with 2,716 admissions (26.32%). **Conclusion:** Thus, it can be stated that hospital admissions for Parkinson's disease among the male population can be considered a public health issue that requires action from the government and Brazilian society as a whole. In the context of Parkinson's disease, the demand for encouraging studies based on collected data is significant, indicating a high contingency in evaluating data related to deaths due to Parkinson's disease. Therefore, it is pertinent to understand and suggest that new studies addressing this issue should be conducted periodically, to expand data visualization, thereby enhancing the technical-scientific field and the implementation of public policies.

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Overview of the number of pediatric hospitalizations due to bacterial meningitis and vaccination coverage in Brazil from 2013 to 2022

Lívia Burim Cerreti[1], Cecília Rioja Gomes[1], Luana Almeida Verçosa[1], Amanda Almeida Verçosa[1], Tainan Gomes Ferreira[1], Djalma de Campos Gonçalves Júnior[1], Igor Bueno Garrido[1], Carlos Roberto Massella Junior[2] , Cezar Kabbach Calaça Prigenzi[3], Caio Cesar Nuto Leite França[3]

[1]Universidade Nove de Julho, Sao Paulo SP, Brazill.
[2]Hospital Militar de Área de São Paulo, Sao Paulo SP, Brazill.
[3]Conjunto Hospitalar do Mandaqui, Sao Paulo SP, Brazill.

Correspondence: Tainan Gomes Ferreira; Email: tainanferreira@uni9.edu.br.

Background: Meningitis is characterized by an inflammation of the leptomeninges (arachnoid and pia mater) and cerebrospinal fluid (CSF) that often occurs due to hematogenous dissemination. It can be caused by viruses, fungi and bacteria, including H. influenzae (Hib), N. meningitidis and S. pneumoniae. The preventive measures implemented by the Unified Health System (SUS) include the administration of the meningococcal C vaccine, targeting serotype C of Neisseria meningitidis, the 10-valent pneumococcal conjugate vaccine (PCV10), which encompasses 10 serotypes of Streptococcus pneumoniae, and the pentavalent vaccine (DTPw-HB/Hib), which combines the diphtheria, tetanus, pertussis (DTPw), hepatitis B (HB), and Haemophilus influenzae type B (Hib) conjugate vaccines, that prevents Hib meningitis. **Objective:** To establish an epidemiological overview of the number of pediatric hospitalizations due to bacterial meningitis in Brazil, relating it to meningococcal C, pneumococcal-10, and pentavalent vaccines coverage in the country between 2013 and 2022. **Methods:** The following research consists of an epidemiological, descriptive and cross-sectional study. Data were obtained from the DATASUS database (TABNET) from 2013 to 2022 in all regions of Brazil. The analysis focused on the number of pediatric hospitalizations in the age groups from 0 to 9 years and bacterial meningitis vaccines available in Basic Health Units (UBS) coverage, in addition to the main causes of the disease over the years in the country. **Results:** According to the data, bacterial etiology was the second leading cause of meningitis in the pediatric population between 2013 and 2022, accounting for 13,6% of cases. In 2013, there were 28 children hospitalized due to bacterial meningitis, a number that grew to 678 the following year. Hospitalizations increased progressively over the years, reaching a peak of 1510 in 2019. There was a decrease in 2020 and 2021, with 877 hospitalizations. However, in the last two years this number has risen again, reaching a record of 1546 in 2022. Simultaneously, the vaccination coverage for meningitis has been decreasing over the years across all regions of the country. The meningococcal C vaccine, which had a coverage of 99.7% in 2013, decreased to 72.17% in 2021 and had a slight increase in 2022 (78,63%). Pneumococcal-10 coverage was 93.57% in 2013 and decreased to 74.84% in 2021, rising again in 2022 (81.51%). On the other hand, the

pentavalent vaccine had a coverage of 95.8% in 2013, which gradually decreased to 70.7% in 2019, and increased in the subsequent years, reaching 77.2% in 2022. **Conclusion:** The period from 2013 to 2022 was marked by an increase in pediatric hospitalizations due to bacterial meningitis, simultaneously with a decrease in vaccination coverage in Brazil, thus highlighting the importance of vaccines in order to eradicate diseases. A decline in hospitalizations was observed in the years 2020 and 2021.

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Overview of traumatic brain injury in young adults

Tainá Reis Martins[1], Wívian Maria Pires Figueredo[1], Esther Pinto Veloso Mendes[1], Karina Andrade de Prince[1]

[1]Centro Universitário FIPMoc, Montes Claros MG, Brazil.

Correspondence: Tainá Reis Martins; Email: taina2002@hotmail.com.

Background: Traumatic brain injury (TBI) is defined as an injury due to external biomechanical forces, which generate anatomical or functional brain impairment. TBI is considered the leading cause of disability and death in the population under 45 years of age worldwide. In the Brazilian scenario it represents an important public health issue, due to the high cost of resources for the treatment of its victims. **Objective:** Analyzing the aspects related to traumatic brain injury in young adults in the last 10 years. **Methods:** This is an ecological time series study that used the database of the Health Information System (TABNET), provided by the SUS Department of Informatics (DATASUS). In the SUS Hospital Information System (SIH/SUS), information on TBI in the age group between 20 and 59 was collected from January 2014 to December 2023. Gender, hospital cost, nature of care, average hospitalization rate and deaths were analyzed. **Results:** In the period analyzed, a total of 579,321 hospitalizations with the highest number of cases were reported in 2014 (61,292). There was a decrease of 10.3% (6,342) in hospitalizations between 2014 and 2020 and an increase of 7.9% (4,375) in the period from 2020 to 2023. The region with the most hospitalizations was in the Southeast, with 38.5% (223,162). The highest number of cases occurred in male patients, which represented 82.7% (479,643) of hospitalizations, being the age group most affected by TBI, common to both genders, from 20 to 29 years old, accounting for 29.1% (168,912). In contrast, the mortality rate decreased 11.9% (9.4 to 8.3), in turn the highest mortality rate occurred between 50 and 59 years of age, with an index of 11.2. The region with the highest mortality rate was Northeast, with a rate of 10. In character, 85.5% (495,318) of the visits were emergency. The highest total number of hospitalizations was in 2023 (R$ 145,001,068.62), with an increase from 2014 to 2023 of 46.8% (R$ 46,254,166.74). Male patients comprise 86.6% (R$ 997,181,140.64) of total expenses. The average value of hospitalization increased by 51.7% (R$ 833.09). **Conclusion:** TBI hospitalizations in young adults predominated in men aged 20 to 29, with high mortality between 50 and 59 years of age. The reasons for the increase in the number of hospitalizations in the last 3 years should be analyzed, as well as the justifications for the increase in the necessary financial contribution over the last few years, in order to reduce, respectively, morbidity and mortality and health system workload.

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Oxidative stress and mitochondrial dysfunction contributes to postoperative cognitive dysfunction in elderly rats dependent on NLRP3 activation

Edgar Pereira[1], Sandra Bonfantea[1], Lucinéia Gainski Danielskia[1], Nathalya Camila Cruz Guerra Correa[1], Khiany Mathias[1], Solange Stork[1], Richard Simon Machado[1], Naila Maciel Andrade[1], Maria Amélia[1], Fabricia Petronilho[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Edgar Pereira; Email: roserp8564@gmail.com.

Postoperative cognitive dysfunction (POCD), a complication following procedures such as orthopedic surgery and is associated with a worsened prognosis, especially in the elderly population. Several mechanisms have been proposed for communication between the immune system and the brain after surgery. In an experimental tibial fracture (TF) model, we aimed to understand the role of the NLR family pyrin domain containing 3 (NLRP3) on oxidative stress and mitochondrial dysfunction as mechanisms underlying POCD in aged and adult rats. Adult or aged male Wistar rats were subjected to the TF model and received intracerebroventricular saline or MCC950 (140 ng/kg), a specific small-molecule inhibitor that selectively blocks activation of the NLRP3 inflammasome. We followed the control (sham) and TF groups treated with MCC950 or saline for seven days to determine cognitive function and survival. The prefrontal cortex and hippocampus were isolated for NLRP3 evaluation, cytokine analysis, oxidative stress measurements, myeloperoxidase activity, nitric oxide formation, mitochondrial respiratory chain enzymes, and succinate dehydrogenase (SDH) activity. Seven days after TF induction, NLRP3 levels increased in the hippocampus and prefrontal cortex in both ages, showed an enhancement in aged rats compared to adults, and experienced a reversal with MCC950 administration. The administration of MCC950 restored memory, IL-1β and IL-10 levels, nitrite/nitrate, lipid peroxidation in the hippocampus and prefrontal cortex, and preserved catalase activity in the prefrontal cortex in aged rats. At the same age, the complex I activity alteration in both regions and complex II, IV, and SDH in the prefrontal cortex were reversed. In conclusion, brain oxidative stress and mitochondrial dysfunction after orthopedic surgery led to POCD, which is linked to higher NLRP3 activation, mostly in elderly rats.

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P2Y12 inhibitors plus aspirin versus aspirin in patients with stroke or transient ischemic attack: an updated meta-analysis of randomized controlled trials

Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Hilaria Saugo Faria[2], Victor Gonçalves Soares[3], Rian Barreto Arrais Rodrigues de Morais[4], João Lucas de Magalhães Leal Moreira[5], Elísio Bulhões[6], Lucas Cael Azevedo Ramos Bendaham[7], Marianna Leite[8], Márcio Yuri Ferreira[9], Yafell Serulle[9]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.
[3]Federal University of the Jequitinhonha and Mucuri Valleys, Diamantina MG, Brazil.
[4]Centro Universitário São Lucas, Porto Velho RO, Brazil.
[5]Universidade Estadual de Feira de Santana, Feira de Santana BA, Brazil.
[6]College of Higher Education of the United Amazon, Redenção PA, Brazil.
[7]Federal University of Roraima, Boa Vista RR, Brazil.
[8]School of Medicine, Santa Marcelina College, São Paulo SP, Brazil.
[9]Lenox Hill Hospital, Northwell Health, Department of Neurosurgery, New York, USA.

Correspondence: Ocílio de Deus da Rocha Ribeiro Gonçalves; Email: ocilio41@gmail.com.

Background: Several randomized controlled trials (RCTs) have shown that combining P2Y12 inhibitors with aspirin may improve National Institutes of Health Stroke Scale (NIHSS) and reduce recurrent strokes. However, their efficacy remains unclear. Therefore, we conducted an updated systematic review and meta-analysis to compare these drugs with aspirin alone. **Methods:** We systematically searched PubMed, Embase, and Cochrane Central for RCTs comparing P2Y12 inhibitors plus aspirin with aspirin alone in patients with stroke or transient ischemic attack (TIA). Primary outcomes were all-cause mortality, major bleeding, and stroke recurrence. We performed a subgroup analysis to stratify patients according to their NIHSS. A random effects model was used to calculate the risk ratios (RRs) with 95% confidence intervals. Statistical analysis was performed using R (version 4.3.2). **Results:** Fifteen studies were included, comprising 51,209 patients, of whom 12,416 (25.3%) received clopidogrel, and 5,523 (11.2%) received ticagrelor. Follow-up ranged from 7 days to 3 years. P2Y12 inhibitors plus aspirin significantly reduced stroke recurrence (RR 0.78; 95% CI = 0.71-0.87; p<0.05; I2=0%), but increased the incidence of all-cause mortality (RR 1.38; 95% CI = 1.11-1.72; p< 0.05; I2=0%) and major bleeding (RR 2.47; 95% CI = 1.21-5.07; p>0.05; I2=0%). There was no difference between groups in all-cause mortality in patients with NIHSS ≤3 (RR 1.27; 95% CI = 0.88-1.82; p>0.05; I2=0%) or NIHSS ≤10 (RR 1.26; 95% CI = 0.91-1.75; p>0.05; I2=0%). **Conclusion:** In patients with stroke or TIA, the use of P2Y12 inhibitors plus aspirin increased the incidence of all-cause mortality and major bleeding. However, the drug combination reduced stroke recurrence. There was no difference between groups in all-cause mortality in patients with mild and moderate NIHSS.

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P3 event-related potentials before and after lumbar puncture as auxiliary tool in the diagnosis of idiopathic normal pressure hydrocephalus: preliminary results

Liliane Aparecida Fagundes Silva[1], Lucas Pinto Mielle[1], Nayara Pereira Santos[1], Heloise Cazangi Borges[2], Helena Alessi[3], Fernando Vieira Pereira[3], Renan Barros Domingues[3], Carlos Alberto Salzedas Giafferi[3], Sandro Luíz de Andrade Matas[4], Carla Gentile Matas[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.
[2]Centro Universitário Nossa Senhora do Patrocínio, Itu SP, Brazil
[3]Laboratório Senne Liquor de São Paulo, São Paulo SP, Brazil.
[4]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Sandro Luiz de Andrade Matas; Email: sandro.matas@gmail.com.

Background: Idiopathic Normal Pressure Hydrocephalus (iNPH) is characterized by an abnormal enlargement of the cerebral ventricles with increased cerebrospinal fluid (CSF) in the elderly population. For the diagnosis, the CSF Tap-test (TT) is indicated, in which a neuropsychological and gait assessment is carried out before and after a CSF lumbar puncture, and the clinical improvement, whether in gait or cognitive performance, indicates a positive predictive result for iNPH. Despite this, CSF TT remains with low clinical sensibility, requiring complementary evaluation methods to improve the diagnosis accuracy. The P3 is a cognitive potential that has emerged as a neurophysiological biomarker in several other pathologies. Thus, it is of scientific and clinical interest to evaluate its applicability as an additional method during CSF TT, minimizing risks to the individual and complementing the diagnosis of iNPH. **Objective:** This study investigates whether patients with a positive predictive value of the CSF TT for iNPH exhibit improved P3 auditory responses after lumbar puncture. **Methods:** Clinical study approved by the Research Projects Appraisal Ethics Committee

institutional, under number 5.964.351. Forty elderly people suspected of iNPH were included in the study, 20 had a negative, and 20 had a positive predictive value of the CSF TT, both sexes, aged between 62 and 86 years old. Besides gait and neuropsychological assessment, auditory P3 responses were recorded with an oddball paradigm before and after lumbar puncture with the removal of 30 to 40 ml of CSF. The changes in P3 auditory responses were compared between the groups by ANOVA test. **Results:** Subjects with a positive predictive value of the CSF TT had a significant decrease in the P3 latency in the right ear, around 27.65 ms, after lumbar puncture compared to subjects with a negative predictive value of the CSF TT. The auditory P3 response is considered a cognitive component and reflects the attentional response to the stimulus, related to the integrity of cognitive functions, providing information on auditory abilities, such as attention and auditory discrimination, and temporal processing. No increase in amplitude was observed in subjects with a positive CSF TT predictive result. **Conclusion:** The decrease in latency of the P3 auditory response after lumbar puncture was associated with the positive predictive value of CSF. Thus, the latency analysis of this component has shown promising for supporting the CSF TT outcome of subjects with suspected iNPH.

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Pain and fatigue in amyotrophic lateral sclerosis: a multi-method study

Tatiana Souza Ribeiro[1], Stephano Tomaz da Silva[1], Letícia Maria de Queiroz Aquino[1], Diogo Neres Aires[1], Aline Alves de Souza[1], Luciana Protásio de Melo[1], Lorenna Raquel Dantas de Macedo Borges[1], Ricardo Alexsandro de Medeiros Valentim[1], Ana Raquel Rodrigues Lindquist[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Tatiana Souza Ribeiro; Email: tatiana.ribeiro@ufrn.br.

Background: Pain and fatigue are frequently reported symptoms by individuals with Amyotrophic Lateral Sclerosis (ALS). However, the presentation of these symptoms in populations with the condition remains uncertain. Additionally, the literature is controversial regarding the efficacy of physiotherapy practices in improving these symptoms. **Objective:** To analyze the profile of pain and fatigue symptoms in a Brazilian sample of individuals with ALS, as well as to identify evidence-based physiotherapy practices for treating these symptoms. **Methods:** This is a multi-method study composed of a prospective cross-sectional design and a systematic retrospective design. Individuals with ALS were recruited intentionally, using a non-probabilistic sampling method. Upon agreeing to participate in the research, they were assessed using a specific evaluation form, and data on pain and fatigue levels were collected. The measurement instruments used were the Fatigue Severity Scale, the Visual Analogue Scale, and a body diagram. Data analysis was descriptive, and we also categorized fatigue (presence/absence). The study searches were conducted in four databases: MEDLINE, CENTRAL, EMBASE, and PEDro. The risk of bias and the quality of evidence were assessed using the PEDro scale, the Downs and Black scale, and the Scottish Intercollegiate Guidelines Network, respectively. **Results:** Data were collected from 72 individuals with ALS. The sample had an average of 4.31 points on the Visual Analogue Scale and 37.2 points on the Fatigue Severity Scale, indicating moderate levels of symptoms. The systematic retrospective design, after the searches, yielded 16,747 references. Following the selection process, 13 included studies identified various physiotherapy practices for treating pain and fatigue; however, the evidence analysis pointed to uncertainty regarding the effects. **Conclusion:** The prospective analysis of the pain and fatigue profile in individuals with ALS in Brazil revealed moderate levels of both symptoms. The systematic retrospective analysis, in turn, indicated uncertainty about the effect of physiotherapy on the studied symptoms.

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Pain-related evoked potentials uncovers small fiber damage in hereditary transthyretin amyloidosis with polyneuropathy

Renan Flavio de França Nunes[1], Maria Paula Carvalho Azevedo[1], Alberto Rolim Muro Martinez[1], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Renan Flavio de Franca Nunes; Email: renanunes@uol.com.br.

Background: Pain-related evoked potentials (PrEP) are a valuable neurophysiological tool for screening small fiber neuropathy. They have proven useful in diagnosing various neuropathies; however, no study has yet used PrEP to investigate small fiber damage in ATTRv-PN. **Objective:** To determine the damage pattern using a non-invasive approach. **Methods:** We evaluated 30 genetically confirmed ATTRv-PN patients, dividing them into two groups: 17 symptomatic individuals, identified based on symptoms consistent with the diagnosis, and 13 pre-symptomatic individuals, along with 23 age- and sex-matched controls. The subjects underwent detailed clinical examinations and PrEP evaluations on the sensory nerves of the arms (median) and legs (superficial fibular). Subsequently, we compared PrEP latencies (negative peak) and the frequency of absent PrEP responses in nerves with abnormal sensory function. **Results:** The mean age was 45 years for symptomatic and 45 for pre-symptomatic ans symptomatic patients. The main mutation was V30M (26 out of 30 cases). PrEP N1 latency was significantly prolonged in the symptomatic group compared to the pre-symptomatic group in arms (180.46+/-23.72 vs. 139.83+/-13.6ms, Student's t-test p<0.01) and controls (180.46 +/-23.72 vs. 132.2 +/- 15.79ms, Student's t-test p<0.01). The proportion of absent PrEP responses was also higher in the symptomatic group compared to the pre-symptomatic group in legs (Fisher's p<0.01) and controls (Fisher's p<0.01). The comparison of N1 latencies between pre-symptomatic individuals and controls did not reveal statistically significant differences in either the upper limbs (139.83±13.06ms vs. 127.67±22.7ms, Student's t-test p=0.12) or the lower limbs (166.25±32.82ms vs. 151.45±29.74ms, Student's t-test p=0.27). **Conclusion:** Although preliminary, these results suggest that pain-related evoked potentials (PrEP) may be a valuable tool for assessing small fiber damage in patients with ATTRv-PN. PrEP could potentially offer a non-invasive method to identify and monitor neuropathic changes in these individuals, providing important insights into disease progression and aiding in early diagnosis. However, further studies with larger sample sizes and comprehensive methodologies are necessary to validate these findings and establish standardized protocols for PrEP use in clinical practice.

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PAR-2 monoclonal antibody as a treatment for migraine: a systematic review

Ryan Lucas Barbosa Oliveira[1], Mariana Araújo Oliveira Barbosa Rocha[1], Maria Beatriz Cavalcante Brêda[1], Laís Souza da Silva Vitorino[1], Maria Danyelle Farias de Oliveira[1], William Correia da Silva Filho[1], Maryanne Maria Simplício Santos[1], Taíssa Kelly Novais Veiga[1], Valdirene Maria de Barros Barbosa[2]

[1]Universidade Federal de Alagoas, Arapiraca AL, Brazil.
[3]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Ryan Lucas Barbosa Oliveira; Email: ryan.oliveira@arapiraca.ufal.br.

Background: Migraine is a highly debilitating pathology capable of causing loss of ability to work and social life. Despite the recent introduction of monoclonal antibodies targeting the calcitonin gene-related peptide for migraine treatment, not all patients respond well to these drugs. Recently, a new class of drugs, the protease-activated receptor 2 (PAR-2) monoclonal antibodies, has emerged as a potential alternative treatment. PAR-2 is based on blocking meningeal nociceptors, which represents a new therapeutic target. **Objective:** To evaluate advances in migraine treatment using monoclonal antibodies targeting PAR-2. **Methods:** This systematic review analyzed studies from Pubmed, Lilacs, BVS, Embase, and Scielo databases, published between 2018 and 2024. The descriptors used were "monoclonal antibodies, migraine disorders, and PAR-2 receptor", using Boolean operators "and" and "or". Systematic reviews, in vitro studies and animal studies on PAR-2 monoclonal antibodies were included. Studies that did not address its use for migraine were excluded. Two authors independently assessed the methodological quality of the studies. **Results:** A total of 314 articles were found, of which 15 were duplicates. 8 articles were eligible for full reading. Among the articles analyzed, a phase 1 study of MEDI0618, a PAR-2 monoclonal antibody, aimed to evaluate its safety and tolerability in healthy volunteers. Initial findings showed it was well-tolerated with no serious adverse effects; common mild side effects were injection site reactions, mild headache, and transient fatigue. No other changes were observed, suggesting a favorable safety profile. Another antibody, PAR650097, demonstrated significant efficacy in inhibiting PAR-2 in human and mouse cells, preventing cutaneous allodynia induced by PAR-2 agonists or CGRP in mice. Selective activation of PAR-2 in the dura mater triggered headache-related responses in normal mice, but not in PAR-2-deficient mice, highlighting its potential as a therapeutic target for migraine. Studies showed that PAR-2 activation in trigeminal neurons and dural fibroblasts could induce nociceptive behaviors, which were attenuated by the antibody C391 and by the sumatriptan. PAR-2 also emerged as a candidate for modulating nitric oxide sensitivity in the meninges by activating its donor, glyceryl trinitrate (GTN). PAR-2 is also a key mediator in chronic inflammatory conditions, including arthritis, asthma, inflammatory bowel disease, pruritus, and pain. In osteoarthritis and rheumatoid arthritis, PAR-2 contributes to several pathological processes, including synovial hyperplasia, cartilage destruction and pain. The involvement of PAR-2 in these conditions highlights its potential. **Conclusion:** PAR-2 antagonism appears to be a promising approach for treating migraines, particularly for patients with comorbid conditions such as asthma and arthritis. Further research is needed to evaluate its efficacy in human migraine treatment.

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Paradoxical worsening of gait due to l-dopa induced dystonia: a case report

Matheus Alves da Silva[1], Caetano Ternes Coimbra[1], Bruno Sanches Lima[1], Aurea Soriano-Vargas[1], Rickson Coelho Mesquita[1], Laura Silveira Moriyama[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Matheus Alves da Silva; Email: matheus.alves.123@outlook.com.

Response to levodopa (l-dopa) is characteristic of Parkinson´s disease (PD), but patients often complain of worsening of symptoms after l-dopa intake. Reasons include worsening of non-motor features (dysautonomia, gastrointestinal intolerance, sleepiness, etc) but also motor phenomena. We present the case of a 34 year male patient complaining of paradoxical worsening with l-dopa who

presented with l-dopa induced dystonia on a levodopa challenge test. **Case presentation:** A 34 year male patient living with Parkinson disease for six years presented to clinical evaluation due to a complaint of gait worsening after levodopa usage. He previously worked in an agricultural industry, he had no history of parkinson disease or other neurological disorders in his family, his parents were not consanguineous and were born in different states of Brazil. A broad investigation for secondary parkinsonism was made and his magnetic resonance imaging was normal. He had no genetic testing. He is the middle son of two other healthy male brothers. His cognitive profile was unremarkable, besides a cognitive worsening after a short trial of anticholinergic therapy previously, with improvement after the drug suspension. He was using a total dosage of 800 mg of levodopa divided four times daily. Because of the clinical complaint, we started amantadine 100 mg BID and scheduled a levodopa testing. His UPDRS-III score was 62 before levodopa and he showed a clinical improvement to 30. But presented torsional phasic and tonic spasms in the left lower limb affecting the gait, compatible with dystonia. We then adjusted the dosage of amantadine for clinical improvement. **Discussion:** Levodopa is the mainstay of PD treatment, and in patients with young onset Parkinson disease, like in our case, it is well-known that motor fluctuations and dyskinesias may present earlier in the course of disease and with lower dosages. Besides dyskinesias, other conditions, like worsening of dysautonomia, may be a cause of worsening after levodopa. Paradoxical gait response with dopa worsening dystonia is a previously described phenomenology of levodopa induced dyskinesia, with some author suggesting relation to mutations in the Parkin gene. Amantadine is an anti-dyskinetic drug for PD and could be tried in such cases. **Final comments:** The understanding of young onset parkinson disease brought us previously the knowledge of the premature motor fluctuations and dyskinesias with lower doses of levodopa. Dopa worsening dystonia is one of the phenomenologies recently reintroduced in the literature and we present this case to highlight the importance of recognizing this semiological clue for the adequate management of young patients with parkinson disease.

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Paraneoplastic cerebellar degeneration syndrome associated with breast cancer: a case report

Glauco Kody Nagata[1], Paôla Barbosa Schmitt Mendonça[2] , Letícia Bassani Devens[1], Tatiane Morgana da Silva[1], Lucas P. Schilling[1], Cristiano S. Aguzzoli[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Universidade Feevale, Novo Hamburgo RS, Brazil.

Correspondence: Glauco Kody Nagata; Email: glauconagata@gmail.com.

Case presentation: We present a female patient of 66 years of age who was admitted to the emergency department with persistent subacute vertigo for 3 weeks, associated with 15kg of weight loss. She had a history of rheumatoid arthritis, type 2 diabetes mellitus, gastroesophageal reflux disease, systemic arterial hypertension, anxiety, and depression, all controlled. She was discharged from the emergency department and was referred to the neurology outpatient clinic. In the 6-month follow-up neurological examination, she presented with opsoclonus, tongue fasciculations, dysmetria with bilateral intentional tremor in the index-nose test, tactile hypoesthesia in the limbs, and significant axial and appendicular ataxia. At this time, she was hospitalized and underwent extensive diagnostic workup. Magnetic resonance imaging revealed mild microangiopathy (Fazekas I) and cerebellar atrophy. Cerebrospinal fluid showed oligoclonal bands and an IgG index of 6,2, but otherwise normal. Pulse therapy with methylprednisolone was performed, but no improvement in symptoms. Chest tomography revealed a left axillary lymph node measuring 1.4 cm x 1.4 cm, and the breast magnetic resonance imaging was classified as BI-RADS 5. A whole-body PET-CT showed hypermetabolic activity in the left lymph node, which supported further pathological investigation. The patient underwent left lymph node biopsy with immunohistochemical study, which showed a positive Automated CK 7 (SP 52); negative CK 20 (Ks 20.8); positive GATA-3 (L50-823); ER (SP1): weakly positive in about 2% of neoplastic cells; negative PR (1E2). C-erbB-2 (4B5): indeterminate (score 2). Ki-67 (30-9): estimated proliferative index of 50% in the neoplasm. The immunohistochemical profile and histopathological aspects of the biopsy confirmed metastasis from primary breast carcinoma. The rapidly progressive cerebellar syndrome associated with a positive pathological study supported the diagnosis of a paraneoplastic syndrome with cerebellar degeneration secondary to breast cancer. **Discussion:** Paraneoplastic neurological syndromes manifest because of autoimmune responses triggered by the presence of a primary tumor or its metastases. Clinical manifestations may appear months or years before or after the diagnosis of the neoplasm. The diagnosis of the disease is based on three pillars: type of clinical phenotype, presence or absence of neuronal antibodies, and presence or absence of cancer. Afterward, a Care Score is established, which is divided into three diagnostic levels: definite, probable, or possible paraneoplastic neurology syndrome. **Final comments:** Early recognition and treatment of paraneoplastic neurological syndromes are crucial not only to alleviate the patient's neurological symptoms but also to identify and effectively address the underlying cancer. Highlighting the importance of carrying out a physical examination and anamnesis, aiming at the differential diagnosis of neurological syndromes.

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Paraneoplastic polyradiculopathy secondary to papillary renal cell carcinoma with detection of anti-MGT30: a case report

Laura Dias Borges[1], Maria Laura Repache Vitti[1], Mateus dos Santos Renesto[1], Maria Eduarda Melo Mendes[1], Marcos Dieyson Rodrigues Mendes[1], Ana Karen de Medeiros[1], Joyce Yuri Silvestre Yamamoto[2], Giuliano Roberto Scarpellini[2], Jorge Alberto Martins Pentiado Junior[2], Marcell Maduro Barbosa[1]

[1]Centro Universitário Barão de Mauá, Ribeirão Preto SP, Brazil.
[2]Hospital São Lucas, Departamento de Neurologia, Ribeirão Preto SP, Brazil.

Correspondence: Mateus dos Santos Renesto; Email: mateusrenesto@hotmail.com.

Case presentation: A 63-year-old woman presented with progressive gait difficulty over a year, abnormal weight loss, hypoesthesia, burning sensation, and paresthesias in her right leg. Physical exam revealed weakness in right foot for extension, inversion, and eversion (MRC grade 2), tactile and painful anesthesia in L5/S1 dermatomes on the right side, absent patellar and Achilles tendon reflexes bilaterally. ENMG showed signs of subacute, chronic and progressively severe denervation from L1 to S1 bilaterally, with acute activity signs. ESR was 91 and CRP was normal, negative serology tests (including herpes and Lyme disease), as well as extensive negative rheumatologic, metabolic, and endocrine screenings. Brain, cervical and thoracic spine MRI were normal, but the lumbar-sacral revealed periradicular thickening with contrast enhancement in nearly all roots and thickening in the cauda equina. Cerebrospinal fluid (CSF) showed 21 leukocytes (98% lymphocytes), protein level of 185, negative cultures and viral PCRs, positive oligoclonal bands, and IgG index of 1.43. Neoplastic screening identified a right renal nodule (Bosniak II) with PET-CT scan showing increased glycolytic hypermetabolism in the right renal nodule with no signs of sarcoidosis. ACE levels, neuronal antibodies, and anti-neuronal surface antibodies were negative (both blood and CSF), except for positive anti-MGT30 in CSF. Partial nephrectomy and pulse therapy were performed, in addition to plasmapheresis; discharged after overwall improvement with oral prednisone, tapered over 2 months (with minimal residual hypoesthesia on the dorsum and lateral aspect of the right foot, with palsy remission after physiotherapy). Pathological examination of the nephrectomy specimen revealed papillary carcinoma without perineural or vascular invasion, confirmed by immunohistochemistry. Months later, there was recurrence after herpes zoster vaccination, managed with new pulse therapy and rituximab (resulting in remission). Subsequent testing for neuronal and surface neuronal antibodies (both blood and CSF) was negative, with normal nerve biopsy and a negative neuropathies genetic panel. **Discussion:** It is an inflammatory polyradiculopathy, related to early diagnosed renal tumor, with an antibody related to thymoma/myasthenia gravis (despite of common ones, such as anti-CV2/CRMP5, amphiphysin and PCA-2). Responsiveness to corticosteroids and the MRI pattern resembled sarcoidosis, which was ruled out by PET and laboratory testing. Recurrence suggested immune dysregulation, with subsequent negative anti-MGT30 testing in the absence of the tumor. **Final comments:** The case illustrates the relationship between inflammatory disorders of the nervous system without obvious etiology and antibodies produced by neoplasms. Further studies are needed to better understand the exact relationship between these conditions, although clinical signs and complementary tests sometimes may guide treatment.

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Paraneoplastic subacute cerebellar degeneration associated with invasive ductal breast carcinoma: a case report

Antônio Lourenço Pires Neto[1], Amanda Selvátici dos Santos Dias[1], Humberto Ramos Crispim[1], Lucas do Rego Barros Correa Martins[1], Marcos Matheus da Silva Fonseca[2], Guilherme Vedovato Vilela de Salis[1], Matheus Hisamitsu Facine[1], Amanda Leme da Silva Campinas[1], Fabio de Nazaré Oliveira[1], Andressa Regina de Mello Galego[1]

[1]Fundação Faculdade de Medicina de Rio Preto, São José do Rio Preto SP, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Antonio Lourenco Pires Neto; Email: ntto.cbb@hotmail.com.

Case presentation: A 36-year-old woman presented 6 months before admission a sudden and progressive tremor, gait instability and incoordination. She was first diagnosed with meningitis in another hospital with little improvement on her symptoms after therapy. About 4 months before, she had a new clinical worsening with the same symptoms associated with diplopia and was then referred to a quaternary hospital. Her history included hypothyroidism and cholecystectomy. She denied infectious symptoms, weight loss, and sweating. At the exam, she presented appendicular and axial ataxia, gait incoordination and skew deviation. Her laboratorial exams and serologies were normal. Imaging with Head CT confirmed an arachnoid cyst and subcutaneous nodules of indeterminate etiology on the scalp. The cerebrospinal fluid was normal. We proceed the investigation of a subacute global ataxia syndrome with Magnetic Resonance (MRI) that confirmed the findings of CT exams, with no other alterations. Rheumatologic exams (Anti RO, Anti La, Anti microssomal, Anti GAD, Anti TPO, Antitransglutaminase IGG, IGM, IGA) was normal. The patient was lately full reexamined and was found a nodule on her right breast. The ultrasound confirmed the nodule, and posterior mammography indicated a BIRADS 5 score. She underwent biopsy that confirmed an Invasive ductal breast

cancer (hormonal receptor positive, HER2 Negative, Ki 67 50%). She was diagnosed with subacute cerebellar degeneration secondary to paraneoplastic and was treated with Methylprednisolone 1 gram for 5 days, followed by every other day of plasmapheresis for 5 days. A lumpectomy was also made, and the patient started to improve after the 3rd section of plasmapheresis, with significant improvement in ataxia, walking alone, with no tremors and diplopia after the 5th session. The patient was discharged for follow-up with mastology and oncology to continue staging the disease and was also referred to the genetics clinic. **Discussion:** Cerebellar degeneration secondary to paraneoplastic is a rare but well-known complication of cancer. This case shows a woman with subacute onset, that had a great response and recovery with the corticosteroids and plasmapheresis, reinforcing the needing of fast diagnostic and treatment. **Final comments:** The diagnosis of paraneoplastic syndromes is crucial for the early diagnosis of cancer, as well as it is important to remember the paraneoplastic causes of subacute ataxia in young people, especially those with a positive family history, since early diagnosis has a close influence on the prognosis of the disease.

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Paraproteomic polyneuropathy linked to antimyelin antibody: a case report

Eduardo Mendonça Werneck da Silva[1], Caio Gonçalves Gomes[1], Maycon Melo Lopes[1], Jessica Daniele Marques[1]

[1]Hospital das Clinicas Samuel Libaneo, Pouso Alegre MG, Brazil.

Correspondence: Eduardo Mendonça Werneck da Silva; Email: eduardomwerneck@gmail.com.

Case presentation: A 67-year-old male patient with a symmetrical paretic condition that began in 2021 with distal predominance and progressive evolution in the meantime, evolving with hypoaesthesic changes and gait ataxia on an outpatient basis, with an investigation being carried out for the condition with evidence of cytological protein dissociation and electroneuromyographic examination with findings of sensory-motor polyneuropathy with primarily demyelinating motor predominance, presented concomitantly with a monoclonal peak of IGM in protein electrophoresis and urinary immunofixation in follow-up examinations, he evaluated by a hematologist and the hypotheses of POEMS, myeloma and amyloidosis were ruled out at that time. The investigation continued into the case in which the Igm fraction of the antimyelin antibody was present, confirming the diagnosis. **Discussion:** In cases of polyneuropathy, one of the initial screening tests is electrophoresis, given the multiple possible differences linked to antibody clones. The paraproteomic polyneuropathies must Always be considered in such cases, especially if presence of sensory ataxia, demyelinating neuropathy or anti-myelin antibodies. **Final comments:** Due to the wide possibility of diagnosing hupothesis regarding polyneuropathies, one should always search in the anamnesis and physical examination, as well as in routine complementary exams, for cardinal conditions suggestive of specific pathologies, in order to accelerate the diagnosis and thus institute early and effective treatment to reduce disease-related morbidity and mortality.

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Parechovirus encephalitis in a Brazilian adult: an extremely rare case report in the world

Yan da Silva Raposo[1], Ilana Werneck Augsten[1], Laryssa Garcia de Almeida[1], Hiago Antunis Silva[1], Patrícia Marques Mendes[1], Igor Pereira Matos de Oliveira[1], Eduardo Mendonça Werneck da Silva[1], Caio Gonçalves Gomes[1], Matheus de Paula Batista[1], Vitor Hugo Machado Silva[1]

[1]Hospital das Clinicas Samuel Libaneo, Pouso Alegre MG, Brazil.

Correspondence: Yan da Silva Raposo; Email: yanraposo@yahoo.com.br.

Case presentation: This case portrays a 50-year-old Brazilian caucasian man, intellectually disabled, chronic alcoholic, epileptic, and history of pellagra who on January 29th, 2024 presented with seizures. He progressed to refractory status epilepticus and required continuous intravenous midazolam. He had been complaining of otalgia for a few days. The computed tomography (CT) scan of the skull showed no significant brain abnormalities, but there was evidence of unilateral mastoiditis. The CT of the chest revealed broncho-aspiration. Ceftriaxone 2 grams twice a day and Clindamycin 600mg four times a day intravenously were started. The cerebrospinal fluid (CSF) showed pleocytosis (13 cells), hyperproteinorraquia (67mg/dL), glucose normal (85mg/dL), and elevated lumbar opening pressure (28cm H2O). Acyclovir 10mg/kg endovenous was empirically associated. The Allplex Meningitis V2 test detected the Parechovirus. It evolved with a mastoid focus and the antimicrobial spectrum was expanded. The second CSF sample was collected on day 7 of treatment and parameters were normal and the meningitis/encephalitis panel PCR CSF was negative. Sedation was gradually reduced and extubation was performed after clinical stability and electroencephalogram without status epilepticus. On March 21st he was awake, alert, and oriented temporally, spatially, and personally, with preserved strength in the upper and lower limbs (MRC scale 5/5) and bilateral patellar (3+/4+) and Achilles (4+/4+) hyperreflexia, and bilateral Babinsky sign. Magnetic resonance imaging of the brain on February 16th revealed a hyperintense lesion on T2-weighted and FLAIR imaging in the right external capsule. **Discussion:** Parechovirus is the Picornaviridae family, comprising 16 genotypes, the most common being HPeV 1, 3, and 6. It presents gastrointestinal, respiratory, and central nervous system tropism (HPeV 3) and transmission is fecal-oral. HPeV is a significant etiological agent of viral meningoencephalitis and sepsis in infants. However, infection in adults is rare, with few cases having been described. **Final comments:** Searching in PubMed, Virtual Health Library, and Cochrane for terms such as meningoencephalitis, encephalitis, and Parechovirus, we found a single report depicting an Australian patient, also a male, chronic alcoholic who began the condition with status epilepticus.

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PARK ON - proposal for a computational decision support system for assessing the eligibility of patients with Parkinson's disease for functional neurosurgery, including dbs system implantation and ablative procedures

Renê Vasconcelos Silva[1], Jorge Dornellys da Silva Lapa[1], Myllena Maria Santos Santana[1], Wellington Costa da Silva Filho[1], Gabriel Ribeiro Andrade Neto[1], Helton Benevides Santana de Oliveira[1], Roberto César Pereira Prado[1]

[1]Universidade Federal de Sergipe, Aracaju SE, Brazil.

Correspondence: Helton Benevides Santana de Oliveira; Email: heltonbenevides@vidaampla.com.br.

Parkinson's disease (PD) is the second most prevalent neurodegenerative disease in the world, second only to Alzheimer's disease. Between 1990 and 2015, the number of cases increased by 118%. This implies a growing demand for advanced therapies to manage motor symptoms and preserve the quality of life as the disease progresses. In response to this need, we developed PARK ON, a computational solution aimed at the efficient screening of PD patients for functional neurosurgical treatments, such as deep brain stimulation (DBS) and ablative procedures. The relevance of PARK ON is amplified by the limited availability of specialized PD teams in Brazil, which requires a more accurate selection of patients to be referred, and the lack of similar tools in Portuguese. PARK ON was developed by the Movement Disorders Group of the Neurology Service at the University Hospital of the Federal University of Sergipe using the open-source programming language PHP - Hypertext Preprocessor. PHP is widely used in other computational systems and has low maintenance and hosting costs. The system uses a decision algorithm created by our group based on international standards, including CAPSIT-PD and the Multinational European Delphi Panel. PARK ON is available on a webpage and can be used on any device with a web browser, making it widely accessible. This tool provides non-specialist physicians in movement disorders with a robust initial assessment, optimizing referrals to specialized centers. It is important to emphasize that PARK ON acts as a screening tool and does not replace detailed evaluation by multidisciplinary teams. We hope to optimize patient referrals for the evaluation of neurosurgical therapies, improving clinical outcomes and the quality of life for patients. PARK ON represents a significant advance in the management of PD in Brazil, promoting an accessible and efficient initial approach for selecting potential candidates for advanced surgical therapies. This work paves the way for future improvements and adaptations of the tool, as well as its validation studies, aligned with the specific needs of the Brazilian population and technological advancements in healthcare.

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Parkinson's disease in Brazil and its hospital morbidity in 1 decade

Ana Carolina Ribeiro de Araujo e Araujo[1], Aeriberto Carlos Lindoso de Souza[1], Consuelo Penha Castro Marques[1], José Carlos Gomes Patriota Neto[1], Tassya Jordana Coqueiro Batalha[1]

[1]Universidade Federal do Maranhão, Pinheiro MA, Brazil.

Correspondence: Ana Carolina Ribeiro de Araujo e Araujo; Email: anacarolinaraa@hotmail.com.

Background: Parkinson's disease (PD) is a chronic condition, characterized by nigrostriatal degeneration, causing a reduction in dopamine. It is the second most common neurodegenerative disease among the elderly, with age being the most important risk factor. It manifests itself through motor symptoms, such as bradykinesia, tremors (unilateral, resting and asymmetric), rigidity (cogwheel phenomenon) and postural instability; and non-motor, such as hyposmia, psychosis, sleep and mood disorders, considerably impairing the patient's quality of life. The diagnosis is made through a clinical history and detailed physical examination, which can be supported by a good response to pharmacological therapy with levodopa. **Objective:** to study the hospital morbidity profile of patients with Parkinson's Disease in Brazil in the period between 2013 and 2022. **Methods:** This is a descriptive, epidemiological, quantitative study on hospital morbidity due to Parkinson's in Brazil, with secondary data from the System of Hospital Information of the SUS-SIH/SUS, available through the Information Technology Department of the Unified Health System (TABNET/DATASUS), in the period between 2013 and 2022. The variables used were: region, age group, sex and color/race. **Results:** n=9,414 hospitalizations for PD were reported in Brazil between 2013 and 2022, the majority concentrated in the Southeast region, which represented 44.06% of the country, followed by the South region with 27.6%. Furthermore, age groups over 50 years old have a higher incidence of hospital

morbidity, with 89.4% of all hospitalizations due to the disease. Furthermore, over the years, the disease is more prevalent in males, with higher numbers (58.2%) compared to females (41.8%), in addition to affecting mainly individuals of color/ white race (n=4,578), followed by mixed race (n=2,133), highlighting the large amount of hospital morbidity without information regarding color/ race (n=2,321). **Conclusion:** In view of the above, it can be concluded that Parkinson's disease affects, in a potentially serious way, a clinical profile that is based on the male sex, the white race and the age group over 50 years old, highlighting a greater occurrence for this population. of hospital morbidity. That said, clinical monitoring of individuals with this profile must be motivated early, in order to reduce hospitalizations as much as possible, and if so, guarantee improvements in the quality of life of this population. This study has the limitation of being an epidemiological study with secondary data of a descriptive nature and does not allow causality to be inferred, but further studies with primary data must be carried out to better understand this hospital morbidity.

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Parkinson's disease: a baseline tremor (electrocardiographic change in Parkinson's disease)

Nadja Layane Gomes Santiago[1], Maria Eduarda Messias Vassoler[2], Ayla Morais Vitório Guerra[2], Gabriel Souza Medrado Nunes[2], Fernanda Garcia Silva Pereira Nascimento[2], Ricardo Jose Menezes Barberino Mendes[1], Luiz Pereira de Magalhães[1], Roberta Borges Gomes Kauark[1]

[1]Universidade Federal da Bahia, Hospital Universitário Professor Edgar Santos, Salvador BA, Brazil.
[2]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Maria Eduarda Messias Vassoler; Email: maria.vassoler@ufba.br.

Electrocardiogram (EKG) is a simple and non-invasive test that allows clinicians to evaluate heart rhythm. There are some features that can compromise the reliability of the test, such as neurostimulation, psychomotor agitation, fever and, in regards to Parkinson disease (PD), tremor. Such tremors can mimic arrhythmias, thus, if these artifacts are not recognized, they may lead to misdiagnosis. To warn the possibility of misdiagnosis and iatrogenic damage in patients with movement disorders. **Case presenation:** An 80-year-old male patient presented to the neurology department with progressive resting tremor, initially observed in his right arm, and bradykinesia persisting for one year. The patient has a medical history significant for an ischemic stroke affecting the right nucleocapsular region. Neurological examination revealed globally preserved muscle strength, facial hypomimia, parkinsonian gait, bilateral bradykinesia, plastic hypertonia, and resting tremor. The patient was on a regimen of Levodopa/BD 100/25mg, administered four times daily. For secondary cerebrovascular prophylaxis, the patient was advised to continue taking acetylsalicylic acid and a statin, and an EKG was recommended to further investigate cardiovascular status. The subsequent EKG indicated atrial flutter (AFL) with a heart rate of 68 bpm. However, a 24-hour Holter monitor demonstrated a sinus rhythm with an average heart rate of 67 bpm and rare atrial or ventricular extrasystoles. **Discussion:** A multidisciplinary discussion with cardiology and arrhythmology services suggested that the motor artifact, likely baseline tremor, might be responsible for the observed alterations in the electrocardiographic pattern. Approximately one year later, the patient experienced another ischemic event and sought care at a different public health facility, where AFL was again diagnosed. The patient underwent electrical cardioversion and was placed on continuous anticoagulation therapy with Rivaroxaban. Upon returning to our follow-up clinic, both cardiovascular and neurological examinations remained unchanged from the initial assessment. A new EKG showed the same findings as the initial examination. Consequently, anticoagulation therapy was discontinued, and the patient was referred to the arrhythmology service at our hospital for further evaluation. **Final comments:** This case underscores the critical importance of vigilant and comprehensive assessments, particularly in elderly patients presenting with movement disorders. The overlapping symptoms of neurological and cardiovascular conditions, such as tremors potentially mimicking arrhythmias in EKG readings, highlight the necessity for multidisciplinary collaboration, advocating to ensure accurate diagnosis and appropriate management. This case also emphasizes the need for continuous re-evaluation and close monitoring, as initial findings may evolve, necessitating adjustments in therapeutic approaches.

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Parkinsonism in bipolar mood disorder: a new look at an old problem

Lorena Dias Resende[1], Leonardo Ferreira Caixeta[1]

[1]Universidade Federal de Goiás, Goiânia GO, Brazil.

Correspondence: Lorena Dias Resende; Email: resende2@discente.ufg.br.

Background: Bipolar Spectrum Disorder is a mood disorder characterized by episodes of depression and mania that usually occur for no apparent reason. When treating Bipolar Disorder with neuroleptics, it is common for patients to be diagnosed with parkinsonism in the medium and long term. This generic term is a movement disorder that presents a blockage of nigrostriatal dopaminergic transmission and its prevalence is increasing. Motor symptoms and functional disorders such as tardive dyskinesia and parkinsonism induced by pharmaceutical agents are commonly found in clinical practice, highlighting the convergence of neuro-psychiatric diseases with the motor system. Most patients with Bipolar Disorder who are treated with antipsychotics develop a degree of facial inexpressiveness, action tremor, rigidity or bradykinesia. **Objective:** Systematic review of studies published in the last 25 years on parkinsonism in Bipolar Disorder. **Methods:** We conducted a systematic review guided by the PRISMA guidelines of prospective studies evaluating the association of Bipolar Disorder with a subsequent diagnosis of idiopathic Parkinson's Disease, with a special focus on motor symptoms and executive dysfunction. 128 original articles indexed between 2000 and 2024 were selected from the Cochrane, Lilacs, Medline, Pubmed, Springerlinks and Psyclnfo databases using the terms Parkinson's Disease, Bipolar Mood Disorder, mania, depression, executive dysfunction, motor alterations and quality of life. The inclusion criteria were impact on the literature and methodological quality. The exclusion criteria were not meeting the inclusion criteria. **Results:** 60 articles were fully evaluated and 35 articles made up the results of this systematic review. The information provided by this study promotes reflection on the association between mood and motor fluctuations, pointing out that tardive dyskinesia and drug-induced parkinsonism are the movement disorders most commonly identified in neuro-psychiatric practice. Motor disorders are relevant symptoms in the assessment of neuro-psychiatric patients, as they are related to variables such as the severity of cognitive and functional impairment, and can contribute to accurate differential diagnoses. **Conclusion:** The causes of parkinsonism in Bipolar Disorder have not yet been fully elucidated, as the combination of medication is a complicating factor for an objective assessment. A variable percentage of patients with drug-induced parkinsonism have previous disorders in the nigrostriatal system, which suggests that in these cases the drug reveals a previous neurodegenerative disease. Parkinsonism has a significant morbidity for patients, so it is important to recognize it at an early stage in order to limit its clinical impact. Further research is needed.

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Paroxysmal kinesigenic dyskinesia associated with a pathogenic variant in the PRRT2 gene: a case report

Felipe Thiago Bandeira Serpa[1], Artur Menegaz de Almeida[1], Delcio Francisco Chagas Netto[1], Gabriel Henrique Penha Custódio[1], Ricardo de Oliveira[1], Fernando Augustus de Paula Barreto Garcia[2], Fernando Augustus Bignardi Garcia[1]

[1]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[2]Universidade Anhembi Morumbi, São Paulo SP, Brazil.

Correspondence: Artur Menegaz de Almeida; Email: arturomenegaz@gmail.com.

Case presentation: A 16-year-old male presented with a 1-year history of involuntary movement attacks in his right members that last up to 15 seconds and occur 20 to 30 times daily. He denied having loss of sensitivity, sphincter relaxation, or tongue biting during episodes. Pain was not reported, and conscious level remains unaltered. Systems inquiry was unrevealing. He denied a family history of similar symptoms but his mother was diagnosed in adolescence with migraine with aura. The patient had no previous diagnosed pathology, comorbidities, previous surgeries, or is taking any medications. His vaccines were up-to-date. Upon evaluation, he was found to have dystonic findings triggered by sudden muscle movements. MRI of the brain was normal. He was treated with 2mg trihexyphenidyl once daily, after which there was a partial improvement of dystonia, but no change was seen in attack frequency. A karyotype revealed a mutation in the PRRT2 gene, suggesting a diagnosis of paroxysmal kinesigenic dyskinesia. Then, he initiated 200mg carbamazepine once daily, resulting in a complete reduction of attacks and dystonia. **Discussion:** Paroxysmal kinesigenic dyskinesia is a rare movement disorder characterized by sudden involuntary movements affecting one side of the body (unilateral) or both sides (bilateral). Diagnostic criteria require a (1) triggering factor, (2) attacks during less than a minute, (3) unaltered consciousness and no pain during episodes, (4) age of onset of symptoms between 1 and 20 years of age, and (5) clinical response to anticonvulsants. The karyotype was suggested because of PRRT2 heterozygous mutations being associated both with migraine and paroxysmal movement disorders. The identification of the mutation was characterized by a dominant pattern, that is, the allele probably was inherited from the patient's mother, whose partial presence originated the migraine with aura. **Final comments:** This case report contributes to the understanding of the correlation between paroxysmal kinesigenic dyskinesia (PKD) and migraine with aura. The identification of a heterozygous mutation in the PRRT2 gene in the patient, along with the maternal history of migraine with aura, suggests a potential genetic link between these two neurological conditions. These finding underscores the importance of genetic research in the diagnosis and treatment of neurological disorders, enabling a more precise approach and faster diagnosis.

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Paroxysmal kinesigenic dystonia: a misdiagnosed condition

Natália da Costa Chiote Pinheiro[1], Luiz Felipe Vasconcellos[1], Bruna Andrade de Oliveira[1], Carolina Rocha da Silveira Brugger Cardoso[1], Gloria Maria Cardoso de Andrade Penque[1]

[1]Universidade Federal do Rio de Janeiro, Instituto de Neurologia Deolindo Couto, Rio de Janeiro RJ, Brazil.

Correspondence: Natália da Costa Chiote Pinheiro; Email: nataliacchiote@hotmail.com.

Case presentation: A 36-year-old man presented with brief attacks of dystonia on his left upper and lower limb triggered by sudden movements from rest, such as starting to walk, with onset in his childhood. The symptoms lasted less than 5 minutes and subsided spontaneously, occurring at least 3 times a week. Despite their short duration, the symptoms had a major impact on the patient's quality of life, interfering with his job and physical activity. The patient has always been in good health and did not take any medications. His family history was negative for involuntary movements. However, his sister presented with absence seizures in her childhood, with complete remission after a few years. Neurological exam and neuroimaging findings were normal. Genetic testing confirmed a PRRT2 mutation in the patient. It was prescribed Carbamazepine (200 mg/day), leading to a significant reduction in the frequency of attacks. **Discussion:** Paroxysmal kinesigenic dyskinesia is a rare movement disorder characterized by episodic and recurrent attacks of uni or bilateral dystonia, chorea, ballistic, and athetoid movement induced by abrupt movement or an increase of speed. Most cases are idiopathic, although familial cases occur and usually have an autosomal dominant inheritance pattern. It can also be secondary to brain lesions, such as multiple sclerosis or stroke. The onset is usually in childhood, and the attack has a short duration without loss of consciousness. The diagnosis is based on history and clinical observation, and in genetic cases, it can be confirmed by genetic testing, showing PRRT2 mutation, the major causative gene for paroxysmal dyskinesia and also related to self-limited familial infantile epilepsy. Neuroimaging is useful for differential diagnosis. The attacks usually have a dramatic response to antiepileptic drugs, being carbamazepine and oxcarbazepine the first-line choices. This case report illustrates a typical presentation of paroxysmal kinesigenic dystonia. Although his sister has not been genetically tested, it is possible that she has the gene due to her history of benign epilepsy in childhood. Despite the typical presentation, it took more than 20 years to get to the correct clinical diagnosis, being confused with epilepsy and psychogenic disorders, showing that this diagnosis is often a challenge. It is important to recognize the presenting features of this condition to facilitate a timely diagnosis and initiate appropriate treatment. **Final comments:** Paroxysmal dyskinesias are a group of rare movement disorders that are underdiagnosed and often misdiagnosed. It is important that physicians are aware of its clinical presentation in order not to dismiss it as a psychosomatic or psychiatric illness, leading to the use of an unnecessary or inappropriate medication. The disease has a good response to antiepileptic drugs and patients should be informed about the benign course and the tendency to have decreased attack frequency with age.

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Paroxysmal kinesiogenic dyskinesia: a case report

Bibiana Mayer[1], Chadi Emil Adamo[2]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Bibiana Mayer; Email: bibiana_mayer@hotmail.com.

C.R.S, 19 years old, male, computer science student. During a neurological consultation, he complained of multifocal involuntary sudden movements since he was 7 years old, with a maximum duration of 20 seconds and starting only at the beginning of a movement, without any signs and symptoms in the interictal periods. The condition was exacerbated in anxious contexts. There was no perception or associated pain. These episodes were not triggered by strenuous physical exercise or caffeine-based foods. There was no exacerbation in a post-viral infectious context. He reported that when getting off the bus, he noticed imbalance as his legs were 'chcrooked' and sometimes he fell, as well as letting objects fall from his hands - according to information collected. He did not present developmental changes, as well as any other changes related to cortical functions and psychiatric morbidities. During his youth, the patient had already been investigated for a series of extrapyramidal diseases, such as Wilson's Disease, Hungtinton's Disease, neurodegenerative diseases related to iron accumulation (NBIA), myopathies, among others. On neurological examination, he showed no changes in his cranial nerves, cerebellar, motor or sensory tests. Deep cutaneous reflexes were largely preserved and there were no extrapyramidal changes. Given the above situation, he was diagnosed with Paroxysmal Kinesiogenic Dyskinesia Not Associated with Exercise. Treatment with carbamazepine 200mg/day was started, showing rapid and satisfactory improvement after 1 week with low doses, as expected for this condition. Attacks of Paroxysmal Kinesiogenic Dyskinesias (PKD) consist of any combination of dystonic postures, chorea, athetosis, and ballismus. They can be unilateral – always on one side or the other – or bilateral. Unilateral episodes may be followed by a bilateral episode. The attacks are brief, usually lasting only seconds, but rarely can last up to 5 minutes. They are precipitated by a sudden movement or a startle, usually after the patient has sat silently for some time. Attacks can be severe enough to cause a fall. There can be up to 100 attacks per day. After an attack, there is usually a short refractory period before another attack can occur. Speech may be affected, with inability to speak due to dystonia, but there is never any change in consciousness. Attacks can sometimes be stopped if the patient stops moving or warms up slowly. Patients often report variable sensations at the onset of paroxysms. These may consist of paresthesias, a feeling of stiffness, crawling sensations, or a feeling of tension. Equivalent to PKD are equally brief bilateral attacks that are not precipitated by sudden movement or fright. Because the duration and therapeutic response are the same as in PKD, they are listed here under the heading PKD and not in a separate category. Seizures respond well to treatment with anticonvulsants such as phenytoin and carbamazepine.

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Paroxysmal tonic upward gaze mimicking occipital epilepsy

Lohana Guimarães Souza[1], Samir Andres Ramirez Ojeda[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Samir Andres Ramirez Ojeda; Email: samir.ramirez@edu.pucrs.br.

Case presentation: A 1-year and 6-month-old boy, born full-term at 39 weeks and 2 days via elective cesarean section, with an Apgar score of 10/10, and appropriate neuropsychomotor development for his age, presented with no family history of epilepsy or other relevant personal and family medical history. At 1 year and 5 months, he began experiencing episodes of upward eye deviation lasting 10 seconds each, occurring more than 100 times per day, followed by a spontaneous return to normal eye movements. During these episodes, his consciousness was preserved, body posture was maintained, and he exhibited downward head movements. After a neurological evaluation, valproic acid was prescribed, showing an initial good response. However, after a few days, the seizures, identical in semiology to the previous ones, resumed. Levetiracetam was then prescribed but discontinued due to side effects of irritability. He was on valproic acid, 2 capsules per day, with adjustments being made to transition to carbamazepine, and clobazam 7.5 mg. A Video-EEG showed an organized baseline rhythm in the posterior regions (theta-alpha 7-9Hz, irregular and symmetrical, with attenuation upon bilateral eye opening) and rare slowing over the left parieto-occipital region. Interictal activity showed rare epileptiform paroxysms in the centrotemporal and left occipital regions. Several episodes of upward eye deviation and slight head drop were recorded with no convening EEG changes. After the VEEG, antiepileptics were gradually discontinued, and the episodes of upward eye deviation persisted for another 3 weeks, completely disappearing afterwards. He remains with no further episodes and with normal development two years later. **Discussion:** The concept of paroxysmal tonic upward gaze (PTU) was proposed by Ouvrier and Billson in 1988. It is a non-epileptic disorder that begins in childhood, characterized by episodes of sustained conjugate upward eye deviation, neck flexion, downward saccadic eye movements to look in that direction, with preserved horizontal eye movements and consciousness. One-third of patients suspected of having seizures suffer from non-epileptic disorders upon VEEG evaluation. Although most patients with non-epileptic events have normal VEEGs, occasional point discharges between attacks can occur. In such cases, non-epileptic events might be mistakenly diagnosed as seizures. The symptoms of PTU are like focal or absence seizures in childhood. Despite being uncommon, epileptiform paroxysms were observed in the present case's VEEG; however, the discharges were interictal. The causes of PTU are unknown, and most patients recover within a year after onset. Additional symptoms, such as developmental delays or ataxia, can predict worse prognoses. **Final comments:** PTU is an important confounder of epilepsy, especially when interictal electroencephalographic discharges are present. Therefore, distinguishing evidence between seizures and non-epileptic events needs to be clarified.

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Pathogenic mtDNA variants at low-level heteroplasmy may induce mitochondrial dysfunction and cause exercise intolerance

Margleice Marinho Vieria Rocha[1], Raíssa Gontijo Cabral de Souza[1], Silvia Helena Adrião Escarso[1], Claudia Ferreira da Rosa Sobreira[1]

[1]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Margleice Marinho Vieira Rocha; Email: margleice@gmail.com.

Background: Exercise intolerance (EI) is an inability to perform a certain level of physical activity expected for that person, accompanied by myalgia and fatigue. **Objective:** To identify the frequency of pathogenic mtDNA variants in patients with IE and to evaluate whether the cumulative effect of pathogenic variants at low levels of heteroplasmy is capable of inducing mitochondrial dysfunction. **Methods:** Medical records, laboratory tests, muscle biopsy and mtDNA sequencing using the NGS technique of 30 patients with IE were retrospectively reviewed. We evaluate the correlation between molecular changes in mtDNA and mitochondrial dysfunction. Mitochondrial dysfunction was defined based on the criteria of Bernier et.al. and the patients were divided into two groups: group A (with dysfunction), group B (without dysfunction), which were compared. **Results:** 30 patients with IE, 1:2 male:female, aged 23 to 69 years, were included. The average age at onset of symptoms was 28.8+10.25 years and on average. Pure IE was observed in 53.3% of patients; and 46,7% showed mild signs of other systems. All the patients presented at least one probably or definitely pathogenic mtDNA variant and 73.3% of patients presented at least one definitely pathogenic variant. The most prevalent variant in this population was m.3271T>C. The percentage of mutants ranged from 1% to 4%, and only one patient showed 100% mutant. The MT-ND6, MT-ND5, MT-ND1 and MT-TL1 genes were hotspots for pathogenic variants. Patients with CIII or CIV deficiency presented a higher number of pathogenic variants and a higher accumulated percentage of mutants, especially those located in genes encoding tRNAs. We demonstrated a correlation between the number of pathogenic mtDNA

variants in low heteroplasmy and the accumulated percentage of mutants with laboratory signs of mitochondrial dysfunction (increased resting lactate and reduced CIV activity). The group A and the group B was composed by 37% and 63% of the patients, respectively. Resting lactate levels and pathogenic variants were higher in group A, but there was no difference between the number of definitively pathogenic variants and CK levels between the two groups. From muscle biopsy findings, group B had more signs of denervation, fibers with subsarcolemmal accumulation of SDH and COX and failures in NADH activity. Group A had a higher prevalence of deficiencies of all complexes. Psychiatric disorders were common in carriers of the m.3394T> C variant in this series. **Conclusion:** Our group of patients with IE have a high prevalence of mtDNA variants with low heteroplasmy in skeletal muscle, significantly higher than the prevalence observed in the general population. The percentage of mutant of our results suggest that, although in low heteroplasmy, those pathogenic variants are contributing to mitochondrial dysfunctions and possibly in IE of ours patients.

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Pathological jealousy as an initial symptom of Alzheimer's disease: anatomy clinical study of a case

Paulo Roberto de Brito Marques[1], Janaina Mariana de Araujo Miranda Brito Marques[1], Maria Luiza Vasconcelos Montenegro[1]

[1]Universidade de Pernambuco, Recife PE, Brazil.

Correspondence: Janaina Mariana de Araujo Miranda Brito Marques; Email: marianaaraujo6@gmail.com.

M, 74 years old, higher education. Until the 54-year-old, her husband had never spoken or shown any jealousy that attracted attention. From that age onwards, the patient began to become progressively pathologically jealous of his wife, so that no man could have access to her. Even old employees do housework such as bricklayers, plumbers, electricians, friends, and neighbors. She didn't want to give him money for shopping, avoiding contact with men on the street. His wife consulted a lawyer to initiate separation proceedings, but due to the children, his wife withdrew on the judge's advice. The patient continued to be jealous this time of his youngest son, who denied being his, and when he was shown his identity card the patient said it was easy to put anyone's name there. After 10 years, he progressively stopped being jealous, and not giving much importance to his wife, he lost his language and gradually became dependent and without autonomy for civil activities. MEM=00. Pfeffer=30/30. He lived in bed for 2 years. His maternal grandmother, mother, and 4 cousins died of dementia, and he has 1 demented cousin. Structural and functional imaging studies were carried out. An autopsy confirmed the diagnosis of Alzheimer's disease. Othello syndrome (OS) has already been described in the literature, initially in a theater (Shakespeare, 1596-1598), and a film (The Merchant of Venice, 2004). Its dissemination in the medical literature covers various clinical conditions in psychiatry, neurology, and clinical medicine, including medications like dopaminergic agonists. Psychiatric disorders reach 22%, neurological disorders more than 30%, drug use 19%, etc. However, there is no case in the medical literature of OS whose etiology is Alzheimer's disease with pathological confirmation. We conclude with the statement that Othello Syndrome caused by Alzheimer's disease is a rare entity in neurology.

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Pathophysiology of olfactory disorders related to COVID-19 infection

Caroline Lima de Freitas[1], Bruno Augusto Machado Tavares[1], Julia Azevedo Fleury da Fonseca[1], Pedro Lucas de Oliveira Ramos[1], Luis Alexandre Lemos Costa[1]

[1]Universidade Federal do Amapá, Macapá AP, Brazil.

Correspondence: Caroline Lima de Freitas; Email: carolfreitasm7@gmail.com.

Background: The COVID-19 pandemic was declared in March 2020 by the World Health Organization (WHO). The disease is transmitted through close contact with infected respiratory droplets, and symptoms range from cough and fever to severe pneumonia with respiratory failure. Olfactory disorders such as hyposmia, anosmia, and phantosmia are also common. Understanding the pathophysiological mechanisms of these chemosensory alterations is important for formulating effective treatments and for early detection of symptoms and, consequently, the infection itself. **Objective:** To elucidate the pathophysiology of olfactory disorders observed in some cases of COVID-19. **Methods:** This is a systematic literature review, conducted through a PubMed search using the keywords "COVID-19," "Olfactory manifestations," and "Pathophysiology" in October 2023, which yielded 59 results. After applying filters for publication date (2020-2023) and free full-text availability, 53 studies were selected. Titles and abstracts were read, and inclusion and exclusion criteria were applied, resulting in 20 articles that were fully analyzed and used in this study. **Results:** Anosmia was highlighted as a common symptom of COVID-19, and studies revealed it to be a useful neurological marker for early diagnosis. The olfactory epithelium (OE) is composed of various cells that, when infected by SARS-CoV-2, suffer damage, leading to olfactory alterations. The virus's spike protein interacts with the angiotensin-converting enzyme 2 (ACE2), facilitating viral entry into host cells and causing inflammation and cellular damage. Initially, olfactory dysfunction was believed to indicate neuroinvasion, but later studies showed that the virus primarily affects the support cells of olfactory receptor neurons (ORNs), not the neurons themselves. The resulting inflammation can block the olfactory clefts, impeding airflow and olfactory perception. Anosmia resolution varies from an average of 10 days in mild cases to longer periods in some patients. In severe cases, recovery depends on the recruitment of stem cells from the OE. The potential for viral neuroinvasion, which could lead to central nervous system damage, was also explored, but no significant correlation was found between anosmia and other severe neurological manifestations. The spontaneous remission of olfactory dysfunction suggests a localized inflammatory pathogenesis in the olfactory epithelium. **Conclusion:** COVID-19 presented neurological challenges, including olfactory disorders. Studies indicate that SARS-CoV-2 infection of OE support cells via the ACE2 receptor leads to ORN dysregulation and inflammatory alterations, resulting in anosmia. Understanding the mechanisms of how the virus affects chemical senses can aid in developing effective treatments and prophylactic measures.

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Patient with hereditary diffuse leukoencephalopathy axonal with spheroids requiring gastrostomy

Bianca Pinheiro Cabral[1], Aléxia de Rezende Gonçalves[1], Uandy Dias Terras[1]

[1]Universidade de Ribeirão Preto, Guarujá SP, Brazil.

Correspondence: Bianca Pinheiro Cabral; Email: biancapinheirocabral@gmail.com.

Case presentation: Diffuse Hereditary Leukoencephalopathy with Axonal Spheroids (HDLS) is a rare, inherited, autosomal dominant neurodegenerative disease first identified in 1984. The initial symptoms manifest around the age of 45, with a variable life expectancy of 6 years. In the pathology, structural brain alterations in the white matter are observed, characterized by demyelination and irregular axonal swellings (spheroids), resulting in motor, cognitive, and personality changes. We present a case report of Diffuse Hereditary Leukoencephalopathy with Axonal Spheroids in a 60-year-old female patient who was admitted with dysphagia, requiring Gastrostomy for nutritional support. She remained hospitalized in the clinic ward for 7 days. **Discussion:** Brazilian female, 60 years old, residing in Santos - São Paulo, presented to the hospital with progressive dysphagia over the past month. Prior to this period, she maintained a diet of liquids and soft foods, but due to worsening dysphagia, this diet has become unfeasible. Gastrostomy is necessary for maintenance of nutritional support. During the anamnesis, it was reported that she was previously diagnosed with Hereditary Diffuse Leukoencephalopathy with Axonal Spheroids (HDLS) since 2016. The diagnostic hypothesis was confirmed by Single Photon Emission Computed Tomography (SPECT) of the brain, which showed moderate decreased perfusion and/or brain activity in the projection of the bilateral frontal lobes, more pronounced on the left side, as well as mild hypoperfusion symmetrically in the temporal lobes. For diagnostic confirmation, Whole Exome Sequencing was utilized, revealing a likely pathogenic heterozygous variant in the CSF1R gene associated with Hereditary Diffuse Leukoencephalopathy with Axonal Spheroids. Following the anamnesis, it was reported that initial symptoms included cognitive impairment (predominantly visuospatial and executive), progressing to bilateral spastic hemiparesis, confining her to bed, anarthria, and dysphagia. Additionally, she presents with pseudobulbar affect and impairment of language and previously learned tasks. During this hospitalization, there has been noted progression of symptoms, with significant dysphagia being the reason for admission.On June 10, 2024, surgery was performed to place a gastrostomy tube for nutritional support, with a good surgical recovery. And on June 12, 2024, she was discharged from the hospital. Currently, the patient is residing in a nursing home for necessary care and treatment in light of the progression of the neurodegenerative symptoms. **Final comments:** Diffuse Hereditary Leukoencephalopathy with Axonal Spheroids (HDLS) is a rare neurodegenerative disease within the group of inherited, autosomal dominant conditions. With a high degree of neurological impairment and clinical repercussions, measures for patient comfort and support are necessary, as presented in this case. With an unfavorable evolution and prognosis.

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Patterns of response to antiseizure medication over decades in a cohort of patients with mesial temporal lobe epilepsy with hipocampal sclerosis

Aline de Fátima Dias[1], Pamela Regina Henning[1], William Souza Martins Ferreira[1], Marina Koutsodontis Machado Alvim[1], Luciana Ramalho Pimentel da Silva[1], Clarissa Lin Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Aline de Fátima Dias; Email: alinedef.dias@yahoo.com.br.

Background: Mesial temporal lobe epilepsy (mTLE) is the most common cause of focal epilepsy in adults and hippocampal sclerosis (HS) is its major cause. Even though the natural history of this type of epilepsy is well studied, little is known about the pattern of seizures and outcomes over decades of follow up. **Objective:** To evaluate outcomes of seizures frequency in patients under pharmacological treatment. **Methods:** We analyzed 275 patients with mTLE-HS under pharmacological treatment

followed at Unicamp. We collected data related to seizures frequency, age of epilepsy onset, duration of epilepsy, dosages, and types of antiseizures medications used during treatment. We divided the patients into 3 groups: pharmacoresponsive (no seizures in the last two years or more of follow-up), fluctuating course (seizure-freedom periods of one year or longer) or pharmacoresistant (never had a seizure-free period of more than one year). Data related to patients who underwent surgery were analyzed up to the year prior to the surgical intervention. **Results:** The mean age was 49.57 years (SD 14.13) and the mean follow-up was 15.27 years (SD 11.2). 50.5% (n=139) had left m-TLE, 38.9% patients (n=107) right m-TLE and 10.5% (n=29) bilateral m-TLE. 60 were pharmacoresponsive (21.84%), 67 had fluctuating course (24.36%) and 148 were pharmacoresistnat (53.8%). Age of onset was lower in the pharmacoresistant group when compared to pharmacoresponsive group (p=0.001). There were no significant differences related to epilepsy duration or family history of epilepsy (p≥0.094). In the pharmacoresponsive group, the mean epilepsy duration before they became seizure-free was 33.7 years (SD 16.64). In 88.3% of these patients it took more than 10 years after the diagnosis of epilepsy until they became seizure free. The number of ASMs used along the follow-up were higher in the fluctuating group when compared to the other groups (p<0.028). The number of ASMs used in the last follow-up appointment were lower in the pharmacoresponsive group when compared to the other groups (p<0.033). There were no differences related to the use of carbamazepine (p=0.306). The most common ASM was carbamazepine. **Conclusion:** Younger age at the epilepsy onset was related to worse seizure control, as demonstrated in other studies. The pattern of response to ASM was not static in our cohort of mTLE-HS, and up to 21% achieved seizure freedom at some point in life; however, this took 10 years or longer for most of them, and in many, more than 30 years. This reinforces that early surgical treatment of mTLE-HS is essential to guarantee better quality of life. As expected, the number of ASMs used was lower in the pharmacoresponsive group. Our study suggests that a subgroup of patients with mTLE-HS begins to experience good seizure control after decades of refractory epilepsy. It is still unclear whether this improvement is linked to older age or other factors.

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Pediatric Guillain-Barré syndrome in a 27-year cohort

Mario Emílio Teixeira Dourado Junior[1], Daiane de França Falcão Leal[1], Isadora de Albuquerque Falcão Feitosa[1], Tarcila Fontes Lucena[1], Henrique de Paula Bedaque[1], Selma Maria Bezerra Jerônimo[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Mario Emilio Teixeira Dourado Junior; Email: medourado03@gmail.com.

Background: Guillain-Barré syndrome (GBS) is the most common cause of acute flaccid paralysis (AFP) in childhood after eradication of Polio in many countries. Various clinical variants and divergent pathogenic mechanisms that lead to axonal, demyelinating, or mixed findings of GBS observed on electrodiagnostic studies have been described. However, compared to adults, recognition of GBS is sometimes delayed in young children. **Objective:** To examine epidemiological and clinical characteristics aspects of pediatric GBS over the past 27 years in n the state of Rio Grande do Norte, Brazil **Methods:** Data collected prospectively from a cohort of 132 children (<18 years) with GBS, between June 1995 through December 2022 were assessed. **Results:** median age 8 (1- 17); 22% were under 3 years of age. Antecedent events were found in 71.2% patients (56% respiratory infections; 15.2 % Diarrhea). Clinical variants included: 36.6% motor-sensory, 20.4% pure motor, 7.6% overlap GBS-Miller-Fisher, 3% sensitive pure, 1.5% Miller-Fisher syndrome, 4.6% others. Electrophysiological subtypes were: AIDP in 64.2% and axonal in 30.1% patients. 23.2% needed mechanical ventilation and 5.3% died. Pain was present in 70.5% and meningeal irritation was observed in 48.8% of the patients. The presence of pain was associated with elevated protein in cerebrospinal fluid. At 6 months, 93% recover the walking ability. Pediatric GBS had shorter nadir, more frequently presented pain/meningeal irritation, and had a better prognosis. **Conclusion:** The favorable evolution of the clinical outcome is evident, however, GBS can be severe in some patients, progressing to the need for mechanical ventilation and even death. Therefore, considering the particularities of the clinical picture of the pediatric population it is important to have GBS as differential diagnosis for paralysis and early and appropriate treatment be started timewise.

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Pediatric ischemic stroke in a sickle cell patient with patent foramen ovale and Moya-Moya syndrome: the challenges of selecting a secondary prevention strategy

Ednaldo França de Oliveira[1], Michel Ferreira Machado[1]

[1]Hospital Santa Marcelina, São Paulo SP, Brazil.

Correspondence: Ednaldo França de Oliveira; Email: ednaldoliveira1@hotmail.com.

Case presentation: Strokes occur in the pediatric population, although usually considered an adult's disease. Delayed treatment and misdiagnosis are common due to the unfamiliarity and uncommon occurrence. Moyamoya disease is increasingly diagnosed worldwide and represents a significant cause of childhood stroke in Western countries. Similar angiographic features are common in children with other medical conditions, such as sickle cell disease. In these cases, the term "moyamoya syndrome" is used. A patent foramen ovale (PFO) is the most frequent incidental communication diagnosis between the two atria during echocardiography in otherwise healthy children. No further follow-up or intervention is necessary for most healthy children with an isolated patent foramen ovale. Here, we report a case of a 7-year-old male child with sickle cell anemia (SCA) who was admitted to the hospital with a motor deficit in the left limb, noticed upon waking. The previous day, he had been prostrated and feverish. The blood count suggested significant anemia (Hb:6.7 g/dl). **Discussion:** Empirical antimicrobial treatment for incipient meningitis and blood transfusions were administered. After an etiological investigation, the symptoms were attributed to an ischemic stroke, and two potential disease mechanisms were identified: patent foramen ovale and Moya-Moya syndrome. During hospitalization, the child presented bradycardia, hypertension, and a right axis deviation in the electrocardiogram. He completed antibiotic therapy, ventilatory, and transfusion support and was discharged with a complete improvement based on the Pediatric National Institute of Health Stroke Scale (PEDNIHSS) under a scale of 0. Exchange transfusions due to sickle cell anemia were maintained, and he was referred to outpatient cardiology for patent foramen ovale follow-up. **Final comments:** In the absence of large randomized clinical trials in pediatrics, much of the management of pediatric stroke is adapted from adult guidelines. The challenge and the decision-making process for secondary prevention therapy are critical when a child with stroke has rare diseases simultaneously.

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Perception of quality of life and social support of patients with amiotrophic lateral sclerosis followed up at a tertiary outpatient in Fortaleza

Tamires Ferreira Do Carmo Macedo[1], Lara Albuquerque de Brito[1], Cleonísio Leite Rodrigues[1], Hermany Capistrano Freitas[1], Eduardo Braga de Oliveira[1], Monica Maria Costa do Rego Alvares[1]

[1]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Tamires Ferreira do Carmo Macedo; Email: tamisfcarmo@gmail.com.

Background: Amyotrophic lateral sclerosis is a neurodegenerative disease that affects upper and lower motor neurons. This neuropathological process is responsible for the clinical presentation, which includes progressive muscle weakness, atrophy, fasciculations, spasticity, dysarthria, dysphagia, among other symptoms. **Objective:** To evaluate the perception of quality of life and satisfaction with social support of patients with amyotrophic lateral sclerosis who are followed up at the neuromuscular diseases outpatient clinic of the General Hospital of Fortaleza. **Methods:** This is a cross-sectional, quantitative, and descriptive study in which interviews were conducted using a sociodemographic and quality of life questionnaire, answered by 26 patients diagnosed with amyotrophic lateral sclerosis according to the revised El Escorial criteria. **Results:** Among the 26 individuals, 10 (38.4%) were female and 16 (61.5%) were male. The average age was 51 years, ranging from 30 to 79 years. Regarding the perception of quality of life, 5 patients reported being very satisfied, 6 satisfied, 5 neither satisfied nor dissatisfied, 7 dissatisfied, and 2 very dissatisfied. Regarding social support, all 26 patients responded that they were satisfied or very satisfied with family support, and 20 patients were satisfied or very satisfied with support from friends. When asked about the importance of family and friends in coping with the disease, 96% of the patients declared that family members were important or very important, and 65% considered friends to be important or very important. **Conclusion:** Among the participants, 42.3% had a good perception of quality of life despite the severity of the disease and its consequent loss of functionality. The social support provided by family and friends was markedly important in coping with the disease and was recognized by the patients.

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Performance of a Brazilian population with mild cognitive impairment and Alzheimer's disease in the three words: three shapes (3W3S) test

Carla Karnoski[1], Wellington da Cruz Souza[2], Claudia Lopes Carvalho[2], Ariella Fornachari Ribeiro Belan[2], Orestes Vicente Forlenza[2], Márcia Radanovic[2]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, Departamento e Instituto de Psiquiatria, Laboratório de Neurociências (LIM-27), São Paulo SP, Brazil.

Correspondence: Carla Karnoski; Email: carla.karnoski@hotmail.com.

Background: The three words - three shapes (3W3S) test enables differentiation between aspects of learning and memory, characterizing incidental recall (IR), delayed recall (DR), and recognition (REC), thereby allowing assessment of both verbal and non-verbal memory. The test's simplicity and its form of assessment can be useful in diagnosing amnestic and non-amnestic mild cognitive impairment (a/naMCI), and Alzheimer's disease (AD). **Objective:** To verify the applicability and discriminatory capacity of the 3W3S test in patients with aMCI, naMCI and AD within a Brazilian population. **Methods:** We evaluated 107 elderly individuals divided into four diagnostic groups: normal control; (n=33), aMCI (n=20), naMCI (n=42), and

mild AD (n=12) and their performance throughout different stages of the 3W3S test were compared. **Results:** In the non-verbal task, the AD group exhibited the poorest performance across all stages; in IR, the naMCI group surpassed the AD group; both MCI groups (except by AD) were able to match controls after learning and in recognition memory; the MCI group performed worse than the controls in DR (all p-values <0.001). In the verbal task, the AD group demonstrated the worst performance in all stages; the aMCI group performed better than naMCI in IR (all p-values <0.001). Patients with aMCI and naMCI achieved similar scores in all stages of the tasks (all p-values >0.005). **Conclusion:** The 3W3S test demonstrated effectiveness in clinical diagnosis between AD, MCI, and normal controls, revealing score discrepancies. There was no significant difference in scores between the naMCI and aMCI groups.

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Performance of a cost-effective olfactory test to evaluate hyposmia in Parkinson's disease patients

Maria Fernanda de Abreu Rodrigues[1], Josevânia Fulgêncio de Lima Arruda[1], Liene Duarte Silva[1], Rodrigo Tavares Brisson[1], Gabriel de Castro Micheli[1], Ana Lucia Zuma de Rosso[1], Marco Antônio Sales Dantas de Lima[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Maria Fernanda de Abreu Rodrigues; Email: maferodriguesufrj@gmail.com.

Background: Parkinson's disease (PD) is a neurodegenerative condition with its main clinical signs being motor. However, hyposmia is evidenced in more than 90% of patients. Olfactory analysis is conducted using tests such as the Sniffin' Sticks test® (SS-12®) and the Connecticut Chemosensory Clinical Research Center (CCCRC), which has recently been validated for olfactory assessment in Brazilians. The CCCRC has not yet been tested in Parkinsonian patients. Objectives To verify the cost and the ease of manufacturing the CCCRC test and to compare its performance with the SS-12® in the evaluation of hyposmia in PD. Methods A cross-sectional case-control study conducted at the outpatient Neurological clinic of a university hospital. The PD group (PDG) consisted of individuals aged up to 85 years with PD diagnosis of at least 2 years and no dementia diagnosis. The control group (CG) comprised volunteers without neurodegenerative conditions, aged between 60 and 85 years. The H&Y and the UPDRS-III were used to evaluate PDG. Cognitive function was evaluated using the MoCA for all participants. Clinical data and complaints of hyposmia were collected. Exclusion criteria included a history of head trauma, symptoms of rhinosinusitis, or previous nasal surgery. The groups underwent olfactory testing using SS-12® and CCCRC. Mann-Whitney test was employed to compare scores obtained in the two olfactory tests. Correlations used Spearman's rank correlation coefficient (rho). Results SS-12® cost US$200.00 with validity of 1 year use; CCCRC cost US$5.00 and could be used for up to 3 months, being easily manufactured. PDG (n=34) and CG (n=38) had similar age, gender and educational level. CG scored higher on MoCA (21.3+4.0 vs 17.1+6.2 points; p=0.003). Most part of PDG was in H&Y stages I to III (85%). CG scored higher in olfactory tests: 7.76±2 hits in SS-12® and 4.63 ±2.1 hits in the CCCRC vs 4.70±2.3 hits and 1.17±1.6 hits for the PDG (p<0.0001). The results of both tests showed a moderate correlation (rho=0.65; p<0.0001) and a non-significant tendency toward a negative correlation with age for both groups together (SS-12® rho=-0.05; CCCRC rho=-0.13). There was a positive correlation between MoCA scores and odor results on both tests. There was no correlation between motor performance or disease duration and olfactory test scores. In CG, 21.5% scored normally in SS-12® and 44.7% in CCCRC. In the PDG, only 2.9% had normal scores in SS-12® and 0% in CCCRC. We found that, among participants classified as having hyposmia, 96.6% in CG and 30.3% in PDG denied it. **Conclusion:** The CCCRC test showed similar good performance to SS-12® in assessing olfaction. The ease of manufacturing and low cost of the CCCRC olfactory test may facilitate its use in neurological practice, particularly in medium to low-income countries in the future.

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Performance of a healthcare line stroke and its impact on stroke mortality in the Brazil's central arid Northeast countryside

Alan Alves de Lima Cidrão[1], Deborah Moreira Rangel[2], Mara Cibelly da Silva Piheiro[1], Wesley Soares de Melo[2], Nicole Evelly Andrade Alves[1]

[1]Hospital Regional do Sertão Central, Quixeramobim CE, Brazil.
[2]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Alan Alves de Lima Cidrão; Email: alancidrao@hotmail.com.

Background: Stroke is a major national health problem in Brazil and, in 2022, was the main cause of mortality in Brazil. The hospitalization of patients in an organized Stroke Unit Care (SUC) during acute phase improves survival, functional outcomes and decreases length of hospital stay when compared to patients admitted elsewhere. **Objective:** The purpose of this study is to show the performance and describe the impact of SUC on stroke mortality in the Brazil's Central Arid Northeast countryside, during the first five years of the service's operation. **Methods:** Data on consecutive stroke admissions to the SUC were prospectively collected from 31 July 2018 to 31 July 2023. The SUC is composed by multidisciplinary team, has 10 monitorized hospital beds and is reference to 20 near cities. The SUC performance was evaluated by time number of thrombolysis, time door-CT, time door-needle, median days of hospitalization, Rankin modified scale on admission and discharge and NIH scale on admission and discharge. Stroke mortality in SUC was monitored internally and mortality in the macro-region was analyzed according to the DATASUS registry. Continuous variables were presented as mean and standard deviation (SD), or median, and categorical variables as frequency and percentage. SPSS version 21 was used for all analyses. **Results:** There were 2375 cases of ischemic stroke/transient ischemic attack, 313 of hemorrhagic stroke and 25 of cerebral venous thrombosis. 440 intravenous thrombolysis were performed (42 in year 1, 100 in year 2, 114 in year 3, 93 in year 4 and 91 in year 5). These numbers represent, in relation to all cases of ischemic stroke each year, respectively, 10.5% - 19.8% - 24.1% - 19.6% - 17.2%. The median door-CT (min) was, respectively, 14 - 11 - 9.6 - 10 - 8. The median door-needle (min) was, respectively, 39.5 - 22 - 17 - 21 - 21. Hospitalization median was 10.5 days, 9.4, 8.3, 9.1 and 8.6 days on year 1 to 5, respectively. There was also a clear improvement in the NIH and Rankin modified scales at hospital discharge compared to admission. By the end of third year, stroke unit hospital mortality was 9.05%. On the last year, this data went down to 5%. When compared the mortality in local macro-region between years before the stroke unit and after its implantation, we can see a clear and sustained decrease since 2019. The year of 2021 had decrease of 24.5% in relation to 2011. **Conclusion:** The SUC performance was better than worldwide average described previously. As for door-needle time, the unit has 80.2% of thrombolysis until 30 minutes, accordingly with desired by American Heart Association, which goal is 50% until 30 minutes. The numbers of hospital mortality are comparable with developed countries services. The impact on local macro-region mortality rates by the stroke unit implementation is one of the main data of our research, showing the importance of SUC on countryside regions.

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Peripheral facial paralysis post-viral dengue infection: a case report

Luiza Rodrigues Simões[1], Evelyn Paradzinski Alves[1], Nina Vieira Russo[1], Gabriel Frota Salbego[1], Juliana Oliveira Freitas Silveira[2], Fernando Von Bock Bolli[3], Kelly de Oliveira Harada[1], Fabio Pascotto de Oliveira[1]

[1]Universidade Franciscana, Santa Maria RS, Brazil.
[2]Hospital Universitário de Santa Maria, Santa Maria RS, Brazil.
[3]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.

Correspondence: Juliana Oliveira Freitas Silveira; Email: julianaofs@yahoo.com.br.

Case presentation: A 48-year-old previously healthy woman presented to the emergency department with myalgia, headache, retroauricular pain and fever for 6 days. Laboratory tests confirmed dengue infection. She was evaluated and discharged with symptomatic treatment. The next day, she returned with a worsening headache associated with right hemiface paresis and paresthesia. Physical examination revealed leftward deviation of the labial commissure and paresis of the right orbicularis oculi muscle, consistent with Bell's sign. Cranial computed tomography (CT) showed no abnormalities. Laboratory tests indicated elevated transaminases and tests for Hepatitis, HIV and syphilis were negative. The patient was admitted, received analgesics, hydration, and started on corticosteroids. She also reported tinnitus, altered taste and hearing acuity. Contrast-enhanced cranial magnetic resonance imaging (MRI) revealed significant enhancement of the canalicular and labyrinthine segments of the right seventh cranial nerve. Partial improvement in facial mimicry was achieved with physiotherapy. Three months later, she developed right hemifacial spasm and was prescribed clonazepam. The patient was referred for botulinum toxin injection. **Discussion:** Bell's palsy is the most common type of spontaneous peripheral facial paralysis. Its etiology remains unknown, but it is suspected to occur due to viral reactivation of herpes simplex type 1 in most cases. Other viruses, such as cytomegalovirus, herpes zoster, and Epstein-Barr, may also be associated with Bell's palsy. Dengue is an arboviral disease caused by one of the dengue viruses, transmitted by Aedes aegypti or Aedes albopictus mosquitoes. There are four serotypes: DENV1, DENV2, DENV3, and DENV4. DENV2 and DENV3 have a bigger affinity for the central nervous system. Dengue progresses through three phases: febrile, critical, and recovery. During the febrile phase, patients experience fever, myalgia, headache, ocular and joint pain, nausea and vomiting, lasting 3 to 7 days. Afterward, most patients advance to the recovery phase without entering the critical phase. Dengue should be suspected in patients with the typical clinical picture and a history of exposure to the virus. Diagnosis can be clinical or confirmed through laboratory testing, including serology with IgM detection. **Final comments:** Dengue is a notifiable arbovirus disease with an increasing number of cases in Rio Grande do Sul, becoming a public health problem. Acute infection rarely causes neurological symptoms. The exact mechanism of the virus's action in the nervous system is unknown, leading to cases of encephalitis (the most common presentation), Guillain-Barré syndrome, vascular lesions, myelitis, and myositis. There are some reports of cranial nerve involvement. However, the epidemiology of the association between dengue and Bell's palsy is not well-established yet, making this case extremely important.

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Persistent hypoglossal artery stroke presenting as alternating hemiparesis with fourth nerve palsy, internuclear ophthalmoplegia, and dissociated torsional-vertical nystagmus

Carolina Oldoni[1], Marcelo Somma Tessari[1], Gabriel Paulo Mantovani[1], Ana Leticia Amorim de Albuquerque[1], Arthur Pille[1], Rosane Brondani[1], Sheila Cristina Ouriques Martins[1]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Carolina Oldoni; Email: carololdoni15@gmail.com.

Case presentation: An 81-year-old woman presented with the sudden onset of left eye ptosis, binocular diplopia, nausea, and vertigo. The neurologic examination revealed an NIHSS score of 6, which included left eye hypertropia, minor right facial palsy, right arm drift, mild hypoesthesia of the right arm, and mild dysarthria. Additionally, she exhibited internuclear ophthalmoplegia and dissociated torsional-vertical nystagmus. There were no pupil abnormalities. Initially, she had partial ptosis of the left eye, which days later improved. A head CT scan didn't show any new ischemic or hemorrhagic areas. CT angiography revealed a persistent hypoglossal artery arising from the right internal carotid artery (ICA) and passing through the hypoglossal canal to form the basilar artery. The vertebral arteries were hypoplastic. These arteries showed moderate to severe atherosclerosis and an occlusion in the distal right P1 segment with distal recanalization via the anterior circulation. Brain MRI revealed a new ischemic stroke in the medial portion of the left inferior colliculus, left paramedian midbrain, and left posterior thalamus. **Discussion:** This case presents dual rarities in neuroanatomy and clinical presentation. First, the persistent hypoglossal artery (PHA), which is an embryonic carotid-vertebrobasilar artery anastomosis that arises from the ICA, usually between the C1 and C2 vertebral levels, and traverses the hypoglossal canal to join the vertebrobasilar system. It has an estimated angiographic incidence of 0.03-0.09% and represents a failure of regression of the presegmental arteries. Bilateral vertebral arteries can be hypoplastic in these cases, making the PHA the main feeder supplying the vertebrobasilar territory. Second, this patient presented with a rare form of nystagmusdissociated torsional-vertical nystagmus (the left eye exhibited ipsiversive torsional nystagmus and the right eye an upbeat nystagmus, a seesaw pattern). This can be explained by the lesion of the excitatory fibers from the anterior canal. These fibers cross the midline in the lower pons and ascend in the medial longitudinal fasciculus (MLF) to innervate the ipsilateral superior rectus and contralateral inferior oblique muscles. Damage to this pathway in the MLF from the contralateral anterior canal causes a downward deviation of the contralesional eye and intorsion of the ipsilesional eye, resulting in corrective quick phases of nystagmus with upbeating in the contralesional eye and extorsion in the ipsilesional eye. It is worth noting that this presentation is reminiscent of Weber's syndrome; however, in this case, there was a trochlear nerve lesion (intra midbrain trajectory) at the left inferior colliculus and a MLF lesion without involvement of the cerebral peduncle, as first described by Hermann Weber. **Final comments:** In the literature, there are no previous reports combining these two features.

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Persistent MRI contrast enhancement in calcified neurocysticercosis and its association with refractory epilepsy: a case report

Rafael Guimarães Kanda[1], Fernando Cendes[1], Felipe Guimarães Kanda[2], Leslie Ortega Hernandez[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade de Taubaté, Taubaté SP, Brazil.

Correspondence: Rafael Guimarães Kanda; Email: kanda@unicamp.br.

Case presentation: Neurocysticercosis (NCC), caused by the larval stage of Taenia solium, is a significant cause of seizures in endemic regions. We report the case of a 48-year-old woman with recurrent seizures since age 21. Initial cranial CT in 1999 revealed multiple active NCC lesions, accompanied by papilledema and abnormal cerebrospinal fluid findings. Despite multiple treatments with albendazole (1999, 2000, 2005), praziquantel (2004), and corticosteroids (multiple courses of prednisone), her seizures remained uncontrolled. Four cranial MRIs from 2020 to 2024 showed multiple persistent contrast-enhancing lesions. **Discussion:** This case highlights the dynamic nature of calcified NCC lesions and their potential role in refractory seizure disorders. Previous studies have demonstrated the association between perilesional edema and seizure activity, but these were often based on single-timepoint MRI data. Our longitudinal perspective, showing persistent contrast enhancement over a four-year period, underscores the need for comprehensive surveillance and specialized treatment strategies. Histopathological findings indicate that inflammation and immune response play a crucial role in perilesional edema, suggesting that calcified NCC lesions are not entirely inert. This supports the evidence that targeted anti-inflammatory treatments could be beneficial for managing refractory epilepsy in patients with calcified NCC. **Final comments:** Persistent MRI contrast enhancement in calcified NCC lesions may be linked to severe, difficult-to-control epilepsy. This case emphasizes the need for ongoing monitoring and tailored therapeutic interventions, including potential anti-inflammatory treatments, to manage complex epilepsy cases effectively. By providing a longitudinal perspective on MRI findings and their correlation with seizure activity, our report contributes to a deeper understanding of NCC's pathophysiology and management, potentially leading to improved patient outcomes and quality of life.

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Pharmaceutical and non-pharmaceutical treatments for migraine: a systematic review of efficacy and safety

Mônica Bagnara[1], Danielly Machado Iaroszeski[1]

[1]Universidade Comunitária da Região de Chapecó, Chapecó SC, Brazil.

Correspondence: Mônica Bagnara; Email: monica.bagnara.eq@gmail.com.

Backgound: Migraine is a chronic neurological condition marked by episodic attacks of headache, usually unilateral, pulsating in quality, and of moderate to severe intensity. Migraine pathophysiology involves complex interactions between genetic, environmental, and neurological factors. Triptans, acetaminophen, nonsteroidal anti-inflammatory drugs, dihydroergotamine, calcitonin gene-related peptide antagonists, lasmiditan, and certain nonpharmacologic treatments have been associated with improved pain relief and function. However, the evidence for many other interventions, such as opioids, is limited, and adverse side effects from pharmacological treatments cause people with migraine to delay or avoid taking medication. **Objective:** To investigate pharmaceutical and non pharmaceutical treatment for migraine and their efficiency to control the symptoms. **Methods:** A systematic review was conducted in the Pubmed database using expressions such as 'migraine non pharmacological' and 'migraine pharmacological'. The filters used were, '5 years,' 'full text,' 'clinical trial' and 'Meta-analysis.' A total of 277 papers were found related and 161 were chosen to be relevant for the research. **Results:** Migraine medications include triptans, beta-blockers, anticonvulsants, and CGRP inhibitors. Triptans, such as sumatriptan and rizatriptan, are often used to relieve acute migraine attacks but can cause side effects like dizziness, drowsiness, and nausea. Beta-blockers, like propranolol and metoprolol, are commonly prescribed for migraine prevention, though they may lead to fatigue, cold extremities, and weight gain. Anticonvulsants, including topiramate and valproate, help reduce the frequency of migraines but can result in cognitive impairment, weight changes, and hair loss. CGRP inhibitors, such as erenumab and fremanezumab, are a newer class of preventive medications with side effects that may include injection site reactions and constipation. Although non-pharmacological treatments offer benefits without the side effects of medications, some disadvantages can limit their effectiveness and accessibility for some people with migraines. Neuromodulation appears as a promising treatment for migraine, causing moderate pain relief. Other non-pharmaceutical prophylaxis treatments include emiketo diet and cognitive behavioral therapy. Overall, however, non-drug treatments have proven to be moderately effective for treating pain and very effective as a preventive measure. Recent drugs, such as erenumab, are safe and capable of alleviating migraine attacks. **Conclusion:** It is concluded that classic medications used in migraine prophylaxis, such as beta-blockers, anticonvulsants, and triptans, are good, effective, and easily accessible. However, new medications and non-pharmacological interventions should be considered, especially in refractory and difficult-to-control cases.

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Phelan-McDermid syndrome, autism spectrum disorder and intellectual disability in a pediatric patient: a case report

Lucas Medeiros Dunga[1], Matheus Procópio Guimarães[1], Tiago Lameque de Sousa e Silva[1], Helmut Kennedy Azevedo do Patrocínio[1], Pedro Henrick Guimarães Carvalho[1], Cláudio de Souza Fernandes Júnior[1], Artur Rebouças de Souza[1], Diógenes Emanuel Dantas da Silva[1], Iury Hélder Santos Dantas[1], Cláudio de Souza Fernandes[2]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[2]Universidade do Estado do Rio Grande do Norte, Mossoró RN, Brazil

Correspondence: Lucas Medeiros Dunga; Email: lucasdunga2000@gmail.com.

P.G.G.B, 7 years old, is diagnosed with Phelan-McDermid Syndrome (PMS), Autism Spectrum Disorder (ASD), and Intellectual Disability (ID). The initial presentation included strabismus, muscle hypotonia, and deficits in neurodevelopmental. Despite consultations with various pediatricians, no causes for these alterations were found. After contact with pediatric neurologists, a more thorough investigation was conducted, including a cranial MRI that demonstrated anatomical brain alterations, such as thinning of the corpus callosum, a prolonged EEG with normal results and a genetic study revealing alterations in chromosome 22. Currently, the patient presents deficits in social and communicative skills, repetitive patterns and restricted interests, flapping, motor stereotypies, significant delays in speech and language. Based on these characteristics, he is diagnosed not only with the syndrome but also with ASD and intellectual disability. He has a history of three epileptic seizures that were considered febrile by pediatric neurologists. His current treatment involves a multidisciplinary approach with physiotherapist, occupational therapy, speech therapy, psychopedagogy, and the use of risperidone. Phelan-McDermid syndrome is a rare condition of unknown prevalence.. It is a genetic disorder caused by SHANK3 haploinsufficiency due to a 22q13.3 chromosomal deletion. Individuals with PMS present a wide range of systemic and neurodevelopmental manifestations, including global developmental delay/intellectual disability, impaired/limited language, autism spectrum disorder, hypotonia, epilepsy, congenital heart defects, gastrointestinal (GI) disturbances, urogenital malformations, and short stature. The patient initially presented with neonatal hypotonia and strabismus, which should serve as early warning signs for the diagnosis of

the syndrome. Subsequently, he developed neurodevelopmental delay, impaired social and communicative skills, associated with stereotyped, repetitive movements and restricted interests, leading to an additional diagnosis of ASD. The overlap of PMS and ASD symptoms adds complexity to the clinical picture, but the genetic confirmation of the 22q13.3 deletion is decisive for diagnosing PMS. Although there is a history of three febrile seizures and a normal EEG, the occurrence of seizures must be carefully monitored, as it is a common aspect associated with the syndrome. The current therapeutic approach for PMS involves a multidisciplinary treatment plan with interventions tailored to the patient's specific needs, maximizing developmental potential and improving the patient's quality of life. Reports like that of P.G.G.B. contribute to the early identification and treatment of PMS, highlighting early warning signs and the clinical overlap with other conditions, such as ASD. The multidisciplinary therapeutic approach emphasized in the report provides a treatment model that can be adapted and applied to other patients with PMS.

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Phenotypes associated with autosomal recessive polg in adults: description of a series of cases

Alessandra Lima Nogueira Tolentino[1], Ana Marina Dutra[1], Igor Dias Brockhausen[1], Pedro Nogueira Fontana[1], Roseli Corazzini[1], Alzira Alves de Siqueira Carvalho[1]

[1]Centro Universitário Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Alessandra Lima Nogueira Tolentino; Email: ale.tolentino1994@hotmail.com.

Variants in the DNA polymerase gamma gene (POLG) are the most common cause of autosomal inheritance mitochondrial disease, affecting around 2% of the population. The following is a description of 7 cases. **Case presentation:** Patients 1/2: Two male siblings, 60 and 51 years old, with a history of epilepsy, sensory ataxia, psychiatric symptoms, ptosis, chronic progressive external ophthalmoplegia (CPEO) with 21 and 15 years of evolution respectively. Nerve conduction study and myography (NCSM) revealed sensory neuronopathy and myopathic motor unit potentials (MUP). Muscle biopsy showed ragged red fiber in modified Gomori's Trichrome and COX-negative fibers. Brain MRI indicated cortical atrophy and foci of hypersignal T2/FLAIR in white matter. NGS study showed a homozygotic variant in POLG, c.1399G> A (p. Ala467Thr) in both. Patient 3: A 43-year-old female with a history of epilepsy, ataxia, CPEO initiated in her 30's. Presents family history. NCSM was compatible with sensory neuronopathy. NGS panel showed c.1399G> A (p. Ala467Thr) in homozygosis in the POLG. Patients 4/5: Two brothers, 67 and 59 years old. The first one presents CPEO and sensory ataxia since the age of 30 and, last two years, initiated choreic movements and myoclonus. Brain MRI showed cortical and cerebellar atrophy with hypersignal T2/FLAIR of basal nuclei. The second brother has had CPEO and ataxia since he was 50. NCSM showed sensory neuropathy and myopathic MUP. NGS revealed c.752C> T (p. Thr251Ile) in homozygosis in the POLG in both siblings. Patient 6: 46-year-old male with a history of CPEO since the age of 33, evolving with cognitive dysfunction and Parkinsonism. The muscle biopsy was similar to that of patient 1. Brain MRI revealed diffuse cortical atrophy. NGS panel revealed c.1198G> A (p. Val400Met) in homozygosis in the POLG. Patient 7: 26-year-old female, initiated at 19 recurrent stroke-like episodes and status epilepticus. Brain MRI showed foci of hypersignal T2/FLAIR in white matter. Trio exome sequencing showed a compound heterozygous variant: c.1433+1G> A (her father) / c.1880G> A (p. Arg627Gln) (her mother). **Discussion:** Five patients presented the SANDO phenotype (sensory ataxia, neuropathy, dysarthria, ophthalmoplegia). Two families have the p. Ala467Thr, and another has the p. Thr251Ile. Both variants were described as associated with the SANDO phenotype. One patient presents early-onset Parkinsonism and CPEO associated with the variant p. Val400Met. This variant was cited in a Chinese cohort as a cause of early-onset Parkinsonism. The last patient presented the MELAS phenotype (mitochondrial encephalomyopathy, lactic acidosis, stroke-like episodes), with compound heterozygous variant in the POLG. The p. Arg627Gln variant was reported in a patient with MELAS phenotype; c.1433+1G> A was related to POLG-related disorders. **Final comments:** Autosomal recessive POLG gene variants can manifest in adults with epilepsy, SANDO, early-onset Parkinsonism, and MELAS.

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Phenotypic and genotypic profile of Brazilian patients with hereditary spastic paraplegia type 76

Isadora Maria Castilho da Silva Ferreira[1], Marcondes Cavalcante França Junior[1], Carelis Del Valle González Salazar[1], Alberto Rolim Muro Martinez[1], Fabrício Diniz de Lima[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Isadora Maria Castilho da Silva Ferreira; Email: isacast01@gmail.com.

Background: Hereditary spastic paraplegia (SPG) is a heterogeneous group of monogenic conditions sharing lower limb weakness and spasticity as the key features. There are multiple genes and modes of transmission. Biallelic variants in the CAPN1 gene were recently associated to this condition and account for SPG type 76 (SPG76). Little is known about the phenotypic and genotypic spectrum of this condition, particularly in Brazil. **Objective:** Hence, we designed the current study to address this issue. **Methods:** All patients with molecular confirmation of SPG76 under regular follow-up at a tertiary neurogenetics reference center between 2021 and 2024 were enrolled. We collected genotypic data from each individual. Detailed phenotypic information was also recorded, including age, sex, age at onset, neurological manifestations and estimated rate of progression, based on the Spastic paraplegia rating scale. **Results:** There were 12 patients (9 men:3 women). Mean age and age at onset were 49.5±10.4 and 32.5±7.0, respectively. None of them had pediatric onset. Eleven out of 12 patients had a complicated phenotype. Associated manifestations were ataxia (n=9), bulbar symptoms (n=7) and cognitive decline (n=2). One patient had comorbid myotonic dystrophy type 1. The estimated rate of progression was 1.6±2.5 points per year. Parental consanguinity was reported by 8 patients. We found 7 distinct variants in our cohort. 11/12 patients had homozygous CAPN1 variants whereas 1/12 compound heterozygous. The CAPN1 variant c.1176G>A was the most prevalent, found in 10 cases. **Conclusion:** SPG76 is an adult-onset condition that presents most frequently as a slowly progressive spastic paraplegia. The variant CAPN1 c.1176G>A seems to be frequent in Brazilian patients, possibly due to a founder effect.

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Physical disabilities in leprosy patients in Brazil: twenty years of analysis

Luan Victor Silva Ribeiro[1], José Antônio Solano Silva[1], Hálisson Ferreira Freitas Filho[2], Laisa de Paula da Costa Lobato[1], Leandro Gabriel Costa Macedo[1], Bárbara Alessandra Solano Silva[3], Bárbara Coutinho Morgado[3], Dirceu Jose de Freitas Cardoso[1], Ricardo Ferreira Silva[1], Leonardo Kadu Tavares Veloso[1]

[1]Afya Faculdade de Ciências Médicas, Abaetetuba PA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.
[3]Centro Universitário do Pará, Belém PA, Brazil.

Correspondence: Luan Victor Silva Ribeiro; Email: luanvictorsilvaribeiro21@gmail.com.

Background: Leprosy is a chronic infectious disease of significant public health concern in Brazil, primarily affecting individuals in disadvantaged socio-economic conditions. This disease is caused by the bacillus Mycobacterium leprae, which primarily affects epithelial and neural cells, thereby conferring a high potential for disability to the patient. Patients with leprosy are categorized based on skin lesion count, bacterial load, and peripheral nerve involvement. Those without neural impairment are classified as grade 0 for physical disability. Grade 1 disability denotes decreased sensitivity or loss in the eyes, hands, and feet, while grade 2 applies to severe lesions in these areas. **Objective:** To evaluate the epidemiological profile of leprosy patients according to the degree of physical disability from 2002 to 2022 in Brazil. **Methods:** This is a descriptive study, prepared with secondary data obtained from the Notifiable Diseases Information System (SINAN), provided by the Department of Informatics of the Ministry of Health (DATASUS). Notifications of physical disability associated with leprosy were evaluated according to the degree of impairment during the period from 2002 to 2022, in each region of the country. **Results:** During the analyzed period, 254,273 cases of physical disability due to leprosy were reported in the country, with 189,357 cases classified as grade 1 physical disability and 64,916 as grade 2 physical disability. Regarding the number of physical disability cases reported per year, the highest number of notifications was observed in 2007, with 15,899 cases, and the lowest in 2020, with 8,489 cases. In terms of physical disability episodes reported by region, the Northeast region leads with 92,115 cases, of which 68,725 are grade 1 disabilities and 23,390 are grade 2. Next, the Southeast recorded 53,355 cases, including 37,882 grade 1 physical disabilities and 15,473 grade 2 disabilities. The North ranks third with 49,995 notifications, of which 37,671 are grade 1 disabilities and 12,324 are grade 2. The Central-West is fourth with 45,258 cases, including 35,450 grade 1 physical disabilities and 9,808 grade 2 disabilities. The South is fifth with 13,550 cases, including 9,629 grade 1 physical disabilities and 3,921 grade 2. **Conclusion:** There was a high number of notifications of grade 1 physical disabilities due to leprosy in Brazil, with the Northeast region standing out as having the highest number of cases in the country, possibly due to socio-economic conditions, limited access to health services, and climate. Followed by the Southeast, explained by higher population density, social inequality, and greater access to diagnostic methods. To achieve goals aimed at reducing disability levels associated with this disease, it is necessary to improve national health surveillance, access to quality public services, and combat the long-standing stigma associated with leprosy.

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Physiotherapeutic treatment with motor imagery and tdcs on essential tremor: a case presentation

Tatiana de Paula Oliveira[1], Ricardo Galhardoni[2]

[1]Glia Fisioterapia, Barueri SP, Brazil.
[2]Universidade Cidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Tatiana de Paula Oliveira; Email: tatidepaula@gmail.com.

Case presentation: MEP, female, 69 years old, essential tremor (ET) for five years. Previous use of pramipexole, levodopa, and cannabidiol without effect. She is currently taking escitalopram oxalate 10mg and medication for hypertension and heart disease. On assessment: bilateral

tremor in the upper limbs at rest, worsening during movement, making it impossible to eat independently, doing bimanual activities, and carrying objects; pain in the neck and shoulders (VAS 6) and difficulty sleeping; 35.8 on the Tremor Rating Scale Fahn Tolosa Marin (FTM); 34 sec on the nine-hole peg test (NHPT) in the dominant hand; 4 on the cup test. A mental training protocol of motor imagery (MI) with biofeedback, using a brain-machine interface (BMI), was carried out with training in the following tasks: holding hands still at rest, handling cutlery, bringing food to the mouth, bringing a glass to the mouth and handling a cell phone, followed by physical practice (PP) of these activities associated with non-invasive neuromodulation (tdcs: cerebellar cathodal; F3 and F4 anodal stimulation,4mA for 25 minutes). The treatment consisted of 5 weeks of intervention: 10 sessions of MI/BMI +PP/tdcs in the first two weeks and six sessions of PP/tdcs for three weeks, plus 30 minutes a day of MI+PP at home throughout the intervention period. After treatment: 19.23 in the FTM; 25 sec in the NHPT; 3 in the cup test, and a perceived improvement of 80% in keeping hands still at rest, 80% when handling cutlery, 80% when bringing food to the mouth, 50% when getting a glass to the mouth and 80% when handling a cell phone. She began to eat independently and sleep through the night, and her pain completely improved. After five months, she was reassessed: 15,38 in FTM, and 25 sec in the NHPT, maintaining functional gains. **Discussion:** Evidence reports the increased activity of the cerebellar cortex and deep cerebellar nuclei in ET patients. The mental training of motor imagery (MI) can lead to plastic changes in the brain and, when associated with BMI can provide real-time feedback on the person's mental attempts and is considered a promising tool in the rehabilitation of movement disorders. Tdcs applied to the cerebellar region has been shown to cause inhibition of the primary motor cortex by regulating dentate-thalamo-cortical synaptic excitability. Some case reports show positive results with cerebellar and prefrontal tdcs of ET. In this case, we combined these resources to promote neuroplasticity while training functional activities affected by ET. **Final comments:** In this case of ET with significant impairment of activities of daily living (ADL) and no response to medication, physiotherapeutic treatment with motor imagery plus biofeedback and neuromodulation during ADL training promoted improvement of the condition, with gains maintained over five months. This report could be instructive for long-term randomized controlled trial protocols with a larger number of patients.

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Pisa syndrome induced by brexpiprazole: a case report

Ilana Cwajgenberg[1], Orsini Neves[2], Wanderson Alves Ribeiro[3], Daniel Antunes Pereira[2]

[1]Universidade do Estado do Rio de Janeiro, Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.
[2]Universidade Iguaçu, Itaperuna RJ, Brazil.
[3]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Ilana Cwajgenberg; Email: ilanacwajg@gmail.com.

Case presentation: A 58 year old female, with a medical history of high blood pressure and depression, presented dystonic movement of the trunk after initiating Brexpiprazole 2mg per day. The drug dose was reduced until it was interrupted and, after twelve days of drug suspension, the dystonic movements ceased. **Discussion:** Drug induced Pisa syndrome (PS) was initially considered to be a rare acute ou tardive collateral effect of typical antipsychotics. Nowadays, however, it has been linked to various classes of drugs, including atypical antipsychotics. However, Bexpripazole has been described as inducing PS in only one previous report. **Final comments:** PS, a dystonic posture of the trunk, may be idiopathic or drug induced, most frequently associated with typical antipsychotic drugs but has already been linked with the use of atypical antipsychotic drugs and other classes of drugs. This case report describes the second case of Bexpripazole induced PS. To better understand this condition and its real prevalence, further studies are necessary.

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Pituitary tumor simulating cluster headache

Amauri Pereira da Silva Filho[1], Danielle Cortellazzi Colonna Romano[2], Gabriela Yumi Nakamura[2], Melissa Fernandes Vilela de Freitas[2], Sabrina Borges Batista[2], Natália Rebeca Karpejany[3], Taianara Sampaio Reis[3]

[1]Hospital de Emergência e Trauma Dom Luiz Gonzaga Fernandes, Campina Grande PB, Brazil.
[2]Universidade Nove de Julho, São Paulo SP, Brazil.
[3]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Amauri Pereira da Silva Filho; Email: amauri.filho@yahoo.com.br.

Case presentation: Patient, male, 42 years old, with periocular pain radiating to the right temporal region, associated with malaise and vomiting. He had intermittent headaches lasting a few minutes several times a day. These headaches always occur at similar times and partial drooping of the right eyelid and tearing, which have occurred in several previous episodes. After the emergency room (ER), he had partial relief with intravenous dipyrone and nasal oxygen (O2), and a probable diagnosis of cluster headache was made. Due to the high frequency of headaches (7 days per month), a decision was made to initiate prophylactic treatment with verapamil in addition to abortive treatment. Additionally, he presented with a visual field defect (bitemporal hemianopsia). The MRI with and without contrast showed an expansive sellar and suprasellar process with slight extension into the cavernous sinus. The diagnosis was revised to a pituitary macroadenoma, and the patient was referred for surgical treatment. **Discussion:** Pituitary tumors account for approximately 17% of all intracranial neoplasms with the majority being pituitary adenomas. They have unique clinical features that are determined by the secreted hormone gene product. Often, these are found incidentally during a workup for headache, in special the trigeminal autonomic cephalalgias (TACs), a group of primary headache syndromes all marked by unilateral headache and ipsilateral cranial autonomic features. The TACs include cluster headache, paroxysmal hemicrania, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing, and hemicrania continua. Pathophysiology includes the trigeminal pain system, autonomic system, hypothalamus, and more recently an identified role for the vagus nerve. Diagnosis is made after looking at headache frequency, duration, and accompanying symptoms. Each TAC has its own unique treatment, which is discussed in depth. Cluster headache (CH) is a TAC with a prevalence of 0.1-0.2% and it is not caused by any known underlying structural pathology. It is often considered to be one of the most painful conditions known to mankind and can be mimicked by symptomatic causes in a small minority of patients, for example tumours, dissections and infections, but a causal relationship between the underlying lesion and the headache is difficult to determine in many cases. There are several reports in the literature suggesting that CH can be mimicked by pituitary disease. The proper diagnostic evaluation of cluster headache is an issue unresolved. **Final comments:** A pituitary tumor presenting with cluster headache is challenging to diagnose due to the variability of symptoms and the need to accurately identify the headache type. The main challenge is to create a treatment plan that effectively addresses both conditions, minimizes side effects, and improves the patient's quality of life through multidisciplinary care.

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Plasma Aβ42/p-Tau ratio predicts csf amyloid status in subjective cognitive decline and mild cognitive impairment

Marcio L. F. Balthazar[1], Isadora Cristina Ribeiro[1], Marjorie Cristina da Rocha Silva[1], Ítalo Karmann Aventurato[1], Liara Rizzi[1], Luis Eduardo da Silva Santos[2], Gustavo Bruniera Peres Fernandes[3], Fernanda Guarino de Felice[4]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Instituto D'Or de Pesquisa e Ensino, Rio de Janeiro RJ, Brazil.
[3]Hospital Israelita Albert Einstein, São Paulo, São Paulo SP, Brazil.
[4]Queen's University, Ontario, Canadá.

Correspondence: Marcio L. F. Balthazar; Email: mbalth@unicamp.br.

Background: Detection of amyloid status in the central nervous system, based on a non-invasive blood test, is essential, especially in patients in potentially pre-dementia stages and candidates for receiving new anti-amyloid drugs. **Objective:** To evaluate if plasma measurements of Aβ42/Aβ40, Aβ42/p-Tau181, and Aβ42/T-tau ratios can predict cerebrospinal fluid (CSF) amyloid alteration in patients with Subjective Cognitive Decline (SCD) and Mild Cognitive Impairment (MCI). **Methods:** We evaluated 51 patients (31 women) diagnosed with SCD and MCI (NIA/AA criteria). All participants underwent neuropsychological assessment, magnetic resonance imaging, blood and cerebrospinal fluid tests. Roche's Elecsys immunoassay measured the quantification of CSF Aβ42 peptide (normal > 1000 pg/mL). Plasma biomarkers Aβ42, Aβ40, p-Tau 181, and t-Tau were measured on an automated SIMOA HD-X immunoassay equipment. Logistic regression models considering Aβ42/Aβ40, Aβ42/p-Tau181and Aβ42/t-Tau ratios in plasma, age, and sex were performed, as well as ROC curve analysis to evaluate sensitivity and specificity. **Results:** 29 subjects had altered Aβ42 in the CSF. The full model was significant, $\chi 2$ (5.51) =11.53, p=0.042, R2=0.272. Only Aβ42/p-Tau181 contributed significantly to the model (B=8.09, p=0.022). At the threshold of 0.368, the model achieved a sensitivity of 75.8% and specificity of 54.5%, with an AUC of 66.6%, 95% CI: 0.52-0.79), p = 0.033 **Conclusion:** The plasmatic Aβ42/p-Tau181 ratio could predict CSF amyloid alteration with good sensitivity and low specificity in subjects at high dementia risk. As far as we know, this is the first study in a Brazilian sample to evaluate plasmatic Alzheimer's biomarkers in potential pre-dementia subjects using gold-standard diagnostic methods like Elecsys and SIMOA.

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Plasma miRNAs correlate with structural brain and cardiac damage in Friedreich's ataxia

Alexandre Hilário Berenguer de Matos[1], Thiago M. Peluzzo[1], André S. Vieira[2], Cynthia Silveira[1], Mariana Martin[1], Otávio R. C. Filho[1], Thiago J. R. Rezende[1], Alberto R. M. Martinez[1], Marcondes C. França Jr[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Departamento de Neurologia, Campinas SP, Brazil.
[2]Universidade Estadual de Campinas, Instituto de Biologia, Departamento de Biologia Estrutural e Funcional, Campinas SP, Brazil.

Correspondence: Alexandre Hilário Berenguer de Matos; Email: alexandrehbm@gmail.com.

Friedreich's ataxia (FRDA) is the most common autosomal recessive ataxia worldwide and is caused by biallelic

unstable intronic GAA expansions at FXN. In recent years, miRNAs have emerged as promising biomarkers in various medical fields such as oncology, cardiology, and epilepsy. Cell-free plasmatic miRNAs have potential advantages as biomarkers because of their size, stability against blood RNases, relative ease of obtaining (from peripheral blood), storage (-80°C), and qRT-PCR measurement. However, there is a limited number of studies looking at plasmatic miRNA expression in FRDA. In this study, we attempted to characterize the plasma miRNA signature (using RNA-Seq followed by real time PCR) and its clinical correlates in a cohort of Brazilian patients with FRDA. We found two abnormally expressed miRNAs, the miR-26a-5p overexpressed and the miR-15a-5p underexpressed. The first was correlated with age at onset, cerebellum volume, spinal cord cross-sectional area (C2-CSA) and the left cardiac ventricle mass (LV_Mass). For the miR-15a-5p, significant correlations were found with the cerebellum volume, spinal cord eccentricity and LV_Mass. Although the specific biological pathways these miRNAs act are not yet fully elucidated, both have BDNF as their target and, hypothetically, modulate its expression. The reduced expression of this gene leads to neuronal loss, both in the cerebellum and sensory pathways, explaining the ataxic phenotype and our results reinforce this hypothesis. Both miRs were correlated with the LV_Mass, but the miR-26a-5p was already associated with cardiomyocyte hypertrophy through the increased inflammasome NLRP3 activity, which is indirectly linked with cardiac hypertrophy. Considering that, we propose these miRNAs as possible prognostic biomarkers for FRDA. However, we should consider that this is a cross-sectional study, so, longitudinal studies are still needed to validate the clinical use of these miRNAs.

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PLP-1 spectrum disease: two female cases of a X-linked inheritance

Catarina Dantas Corrêa[1], Renata Cristine Alves[1], Luis Eduardo Borges de Macedo Zubko[1], Daniel Fortes Amarante[1], Francisco Manoel Branco Germiniani[1], Max Rainer Rosado Novaes[1], Luiz Eduardo Novis de Farias[1], Helio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Renata Cristine Alves; Email: renata_cristinealves@hotmail.com.

Case presentation: A female patient began experiencing right-hand tremor and dysarthria at age 15. Seven years later, she developed progressive lower limb spasticity and ataxia, leading to frequent falls. By age 38, she presented with urinary incontinence and tonic-clonic seizures. Neurological examination revealed mild dysarthria, dysmetria in the upper limbs with right-hand kinetic tremor, mild lower limb paresis with spastic tonus, global hyperreflexia, and bilateral Hoffmann and Babinski signs. She was born into a family of German descent with no history of consanguinity. Her mother was diagnosed with spastic paraparesis and cognitive decline in her fourth decade. Her brother experienced delayed neuropsychomotor development and frequent falls and died at 29 years old after status epilepticus. Her younger sister began experiencing upper limb polymyoclonus, nystagmus, and cervical and laryngeal dystonia at age 13. Both sisters had brain MRIs showing diffuse white matter increased T2 signal intensity. These patients underwent whole exome sequencing (WES), which revealed the same PLP1gene mutation: heterozygous variant c.449del. This mutation causes a frameshift starting at asparagine 150, resulting in a stop codon 10 amino acids later, confirming the diagnosis of Pelizaeus-Merzbacher disease or Spastic Paraplegia type 2 (SPG2). **Discussion:** Pelizaeus-Merzbacher disease (PMD) is a rare X-linked inherited demyelinating leukodystrophy of the CNS, typically manifesting in early childhood with impaired neuropsychomotor development, nystagmus, and hypotonia, and progressing with spasticity and ataxia. Epilepsy is rare in this condition. PMD results from loss or gain of function mutations in the proteolipid protein 1 (PLP1) gene, located on the long arm of the X chromosome (Xq22), which is crucial for myelin synthesis. These mutations lead to abnormal protein production and toxicity to oligodendrocytes. As an X-linked disease, only 14 cases have been reported in women, who typically presente with later onset and milder symptoms. **Final comments:** As far as we know, this is the first reported case of PMD with epilepsy in a female patient.

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Poliomyelitis sequelae in Brazil: an analysis of hospitalizations and deaths according to age groups in the eradication era

Alisson Rafael de Oliveira Pereira[1], Ana Júlia Alves de Oliveira Andrade[1], Lucas Abreu Dias[1], Rodrigo Ferreira de Moura[1]

[1]Universidade Federal de Lavras, Lavras MG, Brazil.

Correspondence: Alisson Rafael de Oliveira Pereira; Email: alissonropereira@gmail.com.

Background: Oral poliomyelitis vaccine (OPV) was introduced in Brazil in 1961, initiating a process of interrupting the circulation of the virus. Officially, the last case of a wild poliovirus infection occurred in 1989. Despite the eradication status of acute poliomyelitis, hospitalization for polio sequelae continued to be recorded, mainly due to three mechanisms: patients whom had contact with poliovirus prior to eradication, and therefore may have developed disease at an earlier age; patients who received the oral attenuated virus vaccine, and due to immunodeficiency, developed a vaccine reaction; and patients who had contact with the wild virus in another country, importing the disease. **Objective:** To evaluate the distribution of hospitalizations and deaths due to poliomyelitis sequelae according to age group and investigate changes in trends between the period 2008-2023. **Methods:** A retrospective study was conducted using data recorded in the SIH available in DATASUS. Groups according to age were evaluated using joinpoint regression analysis and a generalized linear model (GLM). **Results:** Overall, hospitalization rates for polio sequelae decreased (AAPC -8.1; 95%CI: -10.4 to -5.7; p<0.01), with a change in trends. The significant reduction up to 2015 (APC: -15.1; p<0.01) was replaced by a stationary trend with a non-significant decrease (APC: -1.6; p=0.73). Regarding death rates, a non-significant increase was identified (AAPC: 1.4; 95%CI: -3.7 to 6.1; p=0.52) composed of a joinpoint with trend reversal. In 2018, the previous reduction effect (APC: -11.4; p<0.01) was replaced by a significant increase that has been maintained to date (APC: 33.1; p<0.01). Analysis of the subgroups according to age group showed that the overall trend of reduction in hospitalizations was similar between those over 30 years of age who had contact with the wild virus and those who received at least one dose of OPV (AAPCw: -7.9; APCv: -8.5; p<0.01). The GLM showed an important difference between the groups (B= 0.13; 95%CI: 0.08 to 0.16; p<0.001), with the hospitalization rate being 0.13 units higher for those who had contact with the wild virus. Similarly, in absolute numbers, hospitalizations were 3.6 times higher than those in the vaccinated group after eradication. The analysis of death rates showed similar results (B=0.004; 95%CI: 0.003-0.005; p<0.001), with the number of deaths 25.6 times lower in the group after eradication. **Conclusion:** A similar trend of reduction in hospitalizations between groups demonstrates the collective effect of OPV in interrupting the transmission of the virus, while the higher concentration of cases in the >30 years age group indicates that those who were presumed to have had contact with the wild virus before eradication accounted for the majority of hospitalizations for polio sequelae. Further investigations are needed to determine the abrupt upward trend in deaths that began in 2018.

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Polyneuropathy and folate deficiency: a case report

Natália Cardoso Darabas[1], Gustavo Cardoso da Silva[2], Caroline Cardoso da Silva[3]

[1]Hospital Municipal São José, Joinville SC, Brazil. [2]Hospital São João Batista, Criciúma SC, Brazil. [3]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Natália Cardoso Darabas; Email: natiii_cardoso@hotmail.com.

Case presentation: Female patient, 22 years old, underwent bariatric surgery 4 months ago. She began with a deficit in strength and sensitivity in the lower limbs, later progressing to intense shock-like pain. An investigation was carried out with an orthopedist, with spinal resonance showing no changes. Admitted to the neurology service due to severe pain. On physical examination, she presented a subtle strength deficit in the lower limbs and reduced tactile, painful and proprioceptive sensitivity in the lower limbs, worse distally, with abolished deep reflexes. No changes to the upper limb examination. A new neuraxial resonance was performed during hospitalization, with no changes, normal liquor, electroneuromyography with predominantly motor axonal injury, acute denervation in progress. Laboratory tests showed vitamin B12 1.159 pg/mL, vitamin B1 92,5 mcg/L and folic acid 1.4ng/mL. Diagnosed with peripheral polyneuropathy with subacute onset due to folic acid deficiency. The patient began treatment with appropriate replacement and resulted in complete improvement of symptoms. **Discussion:** Peripheral neuropathy is a term given to indicate a disorder of the peripheral nervous system. Characterized by symmetrical sensory symptoms, such as numbness, paraesthesia, pain, and muscle weakness, which usually begins in the hands and feet and may progress to the arms and legs. The most common causes of polyneuropathy are diabetes, excessive alcohol consumption and deficiencies in certain vitamins, but about 25% are considered idiopathic. About the deficiencies, is much more common among patients with vitamin B12 deficiency. The nerve cells may have some ability to accumulate folate, the risk of neuropathy in those with folate deficiency is meant to be much rarer than that of vitamin B12 deficiency. However, patients with folate deficiency can develop peripheral neuropathy, because of the essential role of folate in the methylation of homocysteine to methionine, a precursor to S-adenosyl methionine, which is important for myelination of nerve cells. Importantly, the risk of peripheral neuropathy increased as serum folate decreased. The study carried in 1978 was the first characterization of a pure folate deficiency neuropathy confirmed. Even after several years since it became known that folic acid deficiency can cause polyneuropathy, there are still few studies on the subject. The relationship between folate deficiency and neurological symptoms it's harder, since most patients suffer from nutritional disorders or excessive alcohol intake, resulting in deficiency of nutrients other than folate. **Final comments:** This report demonstrates the importance of research and quantification of folic acid in the investigation of polyneuropathies. Furthermore, in the absence of large randomised controlled trial evidence, the association between serum folate and peripheral neuropathy should be examined in large longitudinal studies.

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Polyneuropathy as an initial manifestation of adrenal neoplasia: case report

Bruna Santos Silva Guzzardi[1], Gustavo Carvalho Viveiros[1], Rodrigo Batista Irikura[1], Anouar Sacca Colognesi[1], Isabella Brasileiro Quintella Migueis[1], Sarah de Souza Chinelato[1], Guilherme Leonel Gomes da Cunha[1], Talles Camin Rajab[1]

[1]Faculdade de Medicina de Marília, Marília SP, Brazil.

Correspondence: Gustavo Carvalho Viveiros; Email: gustavocarvalhoviveiros@outlook.com.

Case presentation: A 62-year-old male patient was referred via cross for evaluation by the institution's neurology service. He was admitted to the emergency room on April 4, 2024, with the report that about 15 days ago he began to suffer from pain in his right shoulder, which progressed slightly to paresis, followed by progressive evolution of the paresis to the ipsilateral lower limb, and in less than 5 days the patient already had tetraparesis. As soon as tetraparesis was established, it was associated with excruciating muscle pain of a generalized nature, dysphagia for liquids, psychomotor alterations (with hallucinatory experiences: visual and auditory). CSF was collected and showed xanthochromic CSF, no cells, protein 505 mg/dl, red blood cells 32/mm3, glucose 26mg/dl. As a result of the CSF, human immunoglobulin was started for 5 days, with no response to treatment. Myositis was confirmed with CPK 1445. Magnetic resonance imaging showed infiltrative lesions in the brain and cervical spinal cord, causing ascending edema at the level of the lesion. A CT scan of the abdomen showed a heterogeneous nodule with contrast enhancement and areas suggestive of cystic degeneration/necrosis in the topography of the left adrenal gland, measuring 6.7 x 8.3 x 8.0 cm. Due to the patient's weakness, the neurosurgery and urology specialties decided not to biopsy the lesion and, together with the neurology and oncology teams, a probable diagnosis of malignant neoplasm of the left adrenal gland with brain and spinal cord metastasis was reached. **Discussion:** Paraneoplastic syndromes represent clinical disorders that are often not directly attributed to the physical effects of the primary tumor or metastasis. Paraneoplastic alterations can develop from the production of substances released by tumor cells, as well as being triggered by the depletion of substances commonly present in the body, whose deficit leads to the onset of symptoms, or even as a result of the body's immune response to the tumor. It's important to remember that the patient's symptoms may be synchronous with the onset of the tumor, but they may be a late symptom that appears after the tumor is established or even precede the tumor. **Final comments:** polyneuropathy can manifest as one of the first symptoms of neoplasms, and in this case it was related to malignant adrenal neoplasia. Unfortunately, however, the time of presentation of the tetraparesis did not coincide with the initial appearance of the tumor, as brain and spinal cord metastases had already set in at the time of diagnosis. It is important to note that human immunoglobulin was not effective in resolving the patient's paretic symptoms, despite being the treatment used in polyneuropathies. However, it would be more likely that the symptoms of the paraneoplastic syndrome would be resolved by treating the tumor lesion with chemo and radiotherapy, which in turn was not eligible for this patient.

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Polyneuropathy secondary to Rocky Mountain spotted fever: a case report

Sarah de Souza Chinelato[1], Bruna Santos Silva Guzzardi[1], Rodrigo Batista Irikura[1], Anouar Sacca Colognesi[1], Isabella Brasileiro Quintella Migueis[1], Gustavo Carvalho Viveiros[1], Talles Camin Rajab[1], Guilherme Leonel Gomes da Cunha[1], Fábio de Araujo Pereira[1], Luiz Henrique Soares Santos Stefano[1]

[1]Faculdade de Medicina de Marília, Marília SP, Brazil.

Correspondence: Sarah de Souza Chinelato; Email: sarahchinelato@hotmail.com.

Case presentation: Male patient, 47 years old, presented to the emergency room reporting the presence of four ticks on his skin after returning home from a fishing trip. The following week, he developed fever, malaise, myalgia, and headache. He was diagnosed with Rocky Mountain spotted fever based on clinical and epidemiological criteria. Eighteen days after symptom onset, he experienced worsening symptoms, including seizures, progressing to toxemia and sepsis, along with flaccid tetraparesis in the limbs, with overall muscle strength grade II. During hospitalization, cranial and total spine MRI showed no abnormalities, and electromyography of all four limbs diagnosed severe axonal motor polyneuropathy predominantly affecting the lower limbs. Cerebrospinal fluid collected at admission showed 10 cells (67% neutrophils) and 115 proteins; a repeat procedure by the Neurology Team after 16 days showed normalization. Serology for Rocky Mountain spotted fever was positive (02 samples >1/256). Throughout the hospital stay, the patient developed respiratory failure requiring orotracheal intubation and sedation. He received various antibiotics, including doxycycline, and after 40 days showed favorable clinical improvement. **Discussion:** Rocky Mountain spotted fever is an acute febrile infectious disease with variable clinical presentation and severity, potentially lethal. It is caused by the obligate intracellular gram-negative bacteria Rickettsia rickettsii and Rickettsia parkeri, primarily transmitted by the Lone Star tick, Amblyomma americanum. The disease mainly affects men aged 20 to 49 who have been exposed to ticks or frequented areas such as waterfalls, rivers, or forests. Most cases occur between June and November when young tick stages are abundant, with these arachnids remaining infected throughout their lifespan. Initially, the disease may present with nonspecific symptoms such as high fever, myalgia, diarrhea, vomiting, and headache. A maculopapular rash may appear after several days. The clinical course can progress with neurological complications such as encephalitis, polyneuropathy, and meningitis. Polyneuropathy associated with Rocky Mountain spotted fever is characterized by diffuse damage to peripheral nerves, resulting in muscle weakness, pain, and paresthesia. Treatment with doxycycline is recommended as first-line therapy, while chloramphenicol is the second-line therapeutic option, prioritized in severe cases. **Final comments:** This study is relevant due to the low prevalence of reported cases regarding neurological complications of Rocky Mountain spotted fever, particularly polyneuropathies. Clinical and epidemiological detection is essential for early diagnosis and treatment, thereby improving prognosis with reduced mortality and severe complications.

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Polyradiculoneuritis due to HSV-2 mimicking miller Fisher syndrome: a case report

Pedro Vinicius Brito Alves[1], Riccardo Casarotti[1], Luiz Gustavo Breinneisen Santos[1], Aida Maria Negrao Esteves[1]

[1]Prevent Senior Brasil, São Paulo, São Paulo SP, Brazil.

Correspondence: Pedro Vinicius Brito Alves; Email: pvbalves@gmail.com.

Case presentation: We report the case of a 75-year-old man who, 4 days before admission, presented with paresthesias in hands and feet associated with postural instability, urinary retention, and pain on radicular stretching maneuvers. Cerebrospinal fluid analysis revealed lymphocytic pleocytosis (7 cells) and elevated protein levels (86mg/dl), with a positive PCR for HSV-2. During hospitalization, despite treatment with intravenous acyclovir for 14 days and pulse therapy with methylprednisolone 1g/day for 5 days, the patient experienced further neurological deficits with loss of axial strength and worsening paresthesia and radicular pain. He also developed areflexia, sensory and cerebellar ataxia, complex ophthalmoparesis and facial diparesis. Given the triad of complex ophthalmoparesis, areflexia and ataxia, immunotherapy was escalated to plasmapheresis for the hypothesis of Miller Fisher Syndrome (MFS). The patient showed subtotal reversal of cranial nerve symptoms and gained independent walking. Good control of neuropathic pain symptoms was achieved with low dose opioids and gabapentinoids. **Discussion:** Herpes Simplex II infection is generally unrecognized, with up to 90% of infections going undiagnosed. When active, it is usually oligosymptomatic and associated with milder neurological conditions, such as aseptic meningitis and Mollaret's meningitis. The patient presented with an acute sensory-motor symptoms, with urinary retention, asymmetrical areflexia and radicular pain, which could represent a HSV-2 associated radiculitis. However as facial diparesis, ophthalmoparesis, sensory and cerebellar ataxia developed, we leaned towards a Miller Fisher syndrome (MFS) clinical picture and approach. HSV-2 infection can affect the entire nervous system, leading to potential necrotizing encephalitis, cranial neuropathies, myelitis, and involvement of roots and peripheral nerves. The recommended treatment consists of intravenous acyclovir for 14 days, with some authors advocating for extending treatment to 21 days based on persistently positive PCR results. Intravenous corticosteroids may hasten recovery in single cases. The clinical progression despite acyclovir and methylprednisolone pulse therapy and improvement with plasmapheresis highlights the anti-body and complement mediated natures of the auto-immune response and aligns with the treatment guidelines for Guillain-Barré syndrome and its variants. **Final comments:** This case highlights heterogeneous clinical spectrum of the HSV-2 infection in the central nervous system, granting it a mimicker fame. Furthermore, touches on the presence of more complex immune-mediated responses to viral infections. Understanding which component of the immunity one wishes to address can help them focus on more effective and tailored therapies, obtaining better functionality outcomes.

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Polyradiculoneuropathy and multisystem complications in a Chikungunya-infected patient: a case report

Natalia Rebeca Alves de Araujo Karpejany[1], Taianara Sampaio Reis[1], Vinicius Araujo do Vale[1], Amanda Melo Barradas[2], Maria Clara Andrade Alvim Vieira[2], Priscila de Sousa Barros Lima[2], Danielle Cortellazzi Colonna Romano[3], Melissa Fernandes Vilela de Freitas[3], Sabrina Borges Batista[3], Gabriela Lustosa Said Uchoa[1]

[1]Universidade Estadual do Piauí, Hospital Universitário, Teresina PI, Brazil.
[2]Universidade Estadual do Piauí, Teresina PI, Brazil.
[3]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Natália Rebeca Alves de Araújo Karpejany; Email: nataliarebeca2607@gmail.com.

Case presentation: A 43-year-old man began experiencing, about 5 months ago, joint pain and muscle weakness in his right lower limb, which progressed to his left lower limb, upper limbs, accompanied by lower back pain. Subsequently, muscle weakness worsens, progressing to inability to walk. Concurrently, he experienced severe, asymmetric, migratory polyarthralgia associated with local heat and edema. He was then referred to the Neurology department of a University Hospital for further investigation. Electroneuromyography showed sensory-motor impairment in the nerves of all limbs, primarily axonal in nature, relatively symmetrical, with diffuse distribution, worse in the lower limbs, and of marked degree, suggesting polyradiculoneuropathy. During hospitalization, he developed sepsis of pulmonary origin, rapidly progressing to respiratory failure, requiring mechanical ventilation in the ICU. After stabilization, he returned to the Internal Medicine ward for further investigation. CT and MRI scans of the CNS showed no abnormalities, but cerebrospinal fluid analysis revealed 88 mg/dl of protein

and 4 cells. Serologies for arboviruses were requested from serum, cerebrospinal fluid, and urine samples, with Chikungunya IgM in the serum sample, confirming the infection. During hospitalization, the patient underwent an echocardiogram which showed an LVEF of 22% associated with diffuse hypokinesia. Given that the patient was previously healthy, myocarditis secondary to arbovirus infection was hypothesized, and treatment with inotropes was initiated. However, to date, the patient is awaiting a cardiac MRI for better etiological definition. At the time of writing this report, the patient is undergoing treatment with Prednisone, Methotrexate, Folic Acid, Gabapentin, and Carvedilol, in addition to physical therapy sessions, showing improvement in motor function, reduction in pain, and better hemodynamic stability. **Discussion:** Chikungunya is caused by an alphavirus transmitted by Aedes aegypti mosquitoes, presenting with acute febrile polyarthralgia, inflammatory arthritis, skin eruptions, and other systemic symptoms. Although rare, severe complications have been reported during chikungunya outbreaks. These include respiratory failure, cardiovascular decompensation, myocarditis, renal failure, hemorrhage, and neurological involvement such as Guillain-Barré syndrome. These complications are more frequently in patients over 65 years old and with underlying comorbidities. This case is unusual due to occurring in a previously healthy 43-year-old male, with respiratory, cardiovascular, and neurological complications. **Final comments:** The case is notable for the numerous severe complications associated with Chikungunya. Early awareness and recognition of these complications are essential for effective management. Continued surveillance and research are needed to comprehensively understand the diverse manifestations and impact of Chikungunya across various patient groups.

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Polysomnographic findings in patients with Parkinson's disease

Emillaine Alves Noronha[1], Diego Monteiro de Carvalho[2], Marcus Vinicius Della Coletta[3], Alvaro Siqueira da Silva[2], Raissa Costa Said[1], Marcello Facundo do Valle Filho[2], Agatha Lopes Azedo[1], Mariana Carvalho Ferreira[1], Yenly Gonzalez Perez[2], Carlos Maurício Oliveira de Almeida[2]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Agatha Lopes Azedo; Email: agathalopesazedo@gmail.com.

Background: Parkinson's disease (PD) is the primary cause of parkinsonism in the elderly, affecting 1-3% of individuals in this age group and about 1% of the global population. PD is associated with poor sleep quality and various sleep disorders such as obstructive sleep apnoea (OSA), insomnia, Restless Legs Syndrome (RLS), and REM Sleep Behaviour Disorder. Polysomnography (PSG) is the gold standard for the objective assessment of sleep disorders. **Objective:** To analyse the polysomnographic findings in patients with Parkinson's Disease. **Methods:** This is a cross-sectional observational study with a duration of 12 months, conducted at a referral hospital in Manaus, Amazonas. The study was approved by the local Ethics Committee under CAAE No. 58684622.2.0000.0007. The sample consisted of patients with PD, diagnosed according to the criteria of the London Brain Bank and classified as stage 1-3 on the Hoehn-Yahr scale. Exclusion criteria included a diagnosis of dementia, atypical parkinsonism, and language disorders. Patients were invited to spend one night in the sleep laboratory. A digital polygraph was used: BW-III, with 45 channels (Neurovirtual, Brazil), including EEG (10-20 system), bilateral oculogram, ECG, tibial, flexor, and mentalis EMG, airflow sensors and cannula, respiratory effort, body position, O2 saturation, and synchronised video (Intelbras, Brazil). Sleep staging was performed according to the criteria of the American Academy of Sleep Medicine (AASM) by trained and blinded physicians. **Results:** After meeting all the study requirements, eighteen patients underwent PSG over 12 months. The total sleep time averaged 447.25 minutes. REM sleep latency had a mean of 234.2 minutes with a standard deviation (SD) of 123.42. Sleep onset latency (N1) had a mean of 13.74 minutes and an SD of 18.77. The mean O2 saturation during total sleep time was 95.33%, while the mean desaturation index (DI) was 10.44. Regarding diagnoses, 7 patients (38.9%) were diagnosed with OSA, 10 patients (55.6%) with primary snoring, and 2 patients (11.1%) with REM Sleep Behaviour Disorder. The mean periodic limb movement index (PLMI) was 1.17 with a standard deviation of 3.51. **Conclusion:** This study evidenced a high prevalence of sleep disorders in patients with PD, with 38.9% diagnosed with OSA and 55.6% with primary snoring. Sleep fragmentation and prolonged REM sleep latency are common, reinforcing the need for polysomnographic evaluation. Proper assessment and treatment of sleep disorders can significantly improve the quality of life for these patients, although further studies are necessary.

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Polysomnography and simplified sleep apnea test devices: mixing water and oil in the cup of the new cfm resolution

Karla Carlos[1], Lucas Martins Teixeira[1], Ligia Geminiani[1], Carla de Oliveira Carletti[1], Douglas Sousa Soares[1], Ana Flavia Machado Pitz[1], Letícia Lambert[1], Gilmar Fernandes do Prado[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, Departamento de Neurologia e Neurocirurgia, São Paulo SP, Brazil.

Correspondence: Karla Carlos; Email: ka.carlos1@hotmail.com.

Background: Simplified sleep apnea test devices (SSATD) can be used to diagnose obstructive sleep apnea (OSA) in patients with a high probability of having the disease. Supposedly valid in contexts of limited resources, they have notable limitations, such as the inability to record EEG and define the sleep state during the test. **Objective:** Assessing how manufacturers ethically address the limitations of these devices in institutional disclosures. **Methods:** We used the SCOPER criteria to assess SSATD registrations with ANVISA, looking at official and fantasy names, descriptions, purposes, official and advertising data, compliance with SCOPER, and adherence to CFM standards. **Results:** We have identified 5 SSATDs, one of which has been discontinued. They have the following technical versus promotional names: Pulse Oximeter (OP) vs Nocturnal Oximetry; Sleep Monitoring System (SMS) vs Home Sleep Test; Sleep Apnea Syndrome Testing Device (DTSAS) vs Sleep Exam & Snoring and Sleep Apnea Diagnosis; Plethysmograph (PG) vs Sleep Exam. One device is a sleep study type III, and 3 type IV, whose SCOPER classifications are: OP- S0C4O1xP0E0R0; SMS- S0C4O1xP0E0R2; DTSAS- S0C4O1xP0E0R0; PG-S3C4O1xP0E0R0). Discussion. Only OP conducts marketing in line with its ANVISA description. Others take advantage of the PSG's established position in diagnosing sleep apnea by referring to their devices as PSGs. The numbering of these devices, such as type IV PSG, may lead people to assume they are a more advanced version. The ads are superlative, exaggerating both the qualities and advantages of SSATD and the disadvantages of PSG. The ads sometimes include doctors endorsing the technology, complicating them in false advertising, and subject to the penalties provided for in CFM resolution 2336/2023 (articles 9 and 11, items II). The official international guidelines specify the naming conventions, limitations, and conditions of use for SSATD. A doctor must evaluate the test result with the patient in the light of clinical data. Justice and morality are at the mercy of poor oversight, impunity, massification, insufficient rational exercise, and deluded consumption of the magical and unprecedented. This new Giges' ring (The Republic, Plato) allows the visible face to show concern about sleep apnea and the invisible one to hide the imperfections of SSATD. Medical power (Foucault: The Birth of the Clinic and The History of Sexuality) is not meant to exploit people (mostly the fragile), making them mere means, depriving them of the Kantian autonomy that we promote to this day as the categorical imperative (Foundations of the Metaphysics of Customs), obstructing their freedom to decide together with the doctor. **Conclusion:** SSATDs can help manage OSAS, but patients and professionals need to be aware of the limitations in diagnostic accuracy and the risks of not detecting a severe disease. They should not be promoted as PSGs to avoid misleading use and ethical issues.

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Pompe disease diagnosed in a neurology clinic in the interior of the Amazon: first adult case in the state of Pará

Marcos Manoel Honorato[1], Eliane Batista Silva[1], Lucas Henrique Silva Coelho[1], Gabriel Nascimento Borges[1], Leandro Picanço de Carvalho[1], Kelson Vinicius Dallabrida Padilha[1], Samuel Oliveira de Amorim[1], Matheus da Silva Ferreira[1], Ana Flávia Ribeiro Nascimento[2], Átila Barros Magalhães[1]

[1]Universidade do Estado do Pará, Santarém PA, Brazil.
[2]Santa Casa de Misericórdia de Barretos, Barretos SP, Brazil.

Correspondence: Marcos Manoel Honorato; Email: marcos.honorato.dr@gmail.com.

Case presentation: A 45-year-old female patient visited the neurology clinic complaining of right lower limb weakness and back pain for approximately 10 years. The condition progressed to the left lower limb, accompanied by numbness and warmth in the legs and feet, and later to the upper limbs, with headache and burning sensation in the mouth. Family history: brother with similar symptoms, progressive dyspnea, cardiorespiratory arrest leading to death without established diagnosis. Consanguineous grandparents. Physical examination: atypical gait, tetraparesis with muscle strength 3 in lower limbs. Reduced deep tendon reflexes. Bilateral plantar reflex in flexion, preserved sensitivity. Complementary tests: CPK: 932 U/L and aldolase 11 U/L. Lumbar spine MRI showed spondylodiscoarthrosis of L4 and L5. Electroneuromyography revealed normal sensory and motor conduction. Enzyme screening for Pompe disease showed reduced activity of the acid alpha-glucosidase (GAA) enzyme of 0.42 mmol/L/h, the genetic panel identified a compound heterozygous mutation in the GAA gene, pathogenic variants c.-32-13T>G and c.925G>A, findings consistent with glycogen storage disease type II. The measurement of acid alpha-glucosidase enzyme activity in leukocytes was low (0.25 nmol/mg PTN), confirming the diagnosis. Enzyme replacement therapy through periodic intravenous infusion was indicated. **Discussion:** Pompe disease is a rare autosomal recessive disorder resulting from a mutation in the gene encoding the GAA enzyme. It participates in glycogenolysis, and its deficiency results in glycogen accumulation in various tissues, particularly cardiac and skeletal muscles. The most common presentation occurs in infancy during the first months of life, but in some cases, it can begin in adulthood. The typical clinical pattern of skeletal muscle involvement, exercise intolerance, progressive girdle weakness, and back pain are notable in the patient in question and are characteristic of the late-onset form. The initial hyperCKemia observed in the case, along with muscular complaints, constitutes a substantial finding of the presymptomatic stage of the disease. Elevated serum levels of AST and LDH often accompany CK levels, which can misleadingly suggest liver disease and contribute to

diagnostic delays. **Final comments:** Despite being a rare, difficult-to-diagnose, and lethal disease in cases like the patient's brother, early initiation of enzyme replacement therapy is crucial for better prognosis, as only then is it possible to effectively alter the clinical progression.

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Porphyria variegata: a case report

Lucas Gonçalves Coelho[1], Victoria Nunes Alvares[1], Clara Ribeiro Rizzo[1], Isabella Navarro Diaz Horta[1], Maria Eduarda Braz Ferreira Leão Caldas dos Santos[1], Bernardo Leal Cupello[1], Vanessa Conceição Saar Mautone Lima[2], Ricardo Cézare Araújo[2], Deborah Santos Sales[2], Vivian Pinto de Almeida[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gafrée e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: Lucas Gonçalves Coelho; Email: lucasgcoelho@edu.unirio.br.

Case presentation: A 38-year-old female patient presented with a history of hospitalization for urinary tract infection, lower back pain on the right and reddish urine six months prior to arriving at our service. During this hospitalization, she also reported epigastralgia, nausea, generalized paresis and paresthesia. The patient mentioned a similar case of polyradiculoneuropathy 5 years ago, in which the investigation led to the main hypothesis of Guillain-Barré Syndrome. Therefore, she was treated with intravenous IgG, showing partial improvement after six months. Currently, the patient presents distal paresis and atrophy of the upper limbs, global areflexia and proximal changes in sensitivity of the forearms and tights, being restricted to a wheelchair. Laboratory investigation revealed elevated levels of porphobilinogen (PBG), but normal levels of delta-aminolevulinic acid (ALA) in urine samples. Molecular genetic testing for Acute Porphyria was performed and identified a heterozygous pathogenic variant of the PPOX gene in the patient, confirming the diagnosis of Variegate Porphyria. **Discussion:** Porphyrias are a group of rare manifestations caused by enzyme deficiencies in the heme group synthesis pathway. Among the subtypes of this disease is the Variegate Porphyria, which results from a pathogenic variant of the PPOX gene, derived from an autosomal dominant inheritance that leads to decreased activity of the protoporphyrinogen oxidase enzyme. This rare disease has a wide spectrum or non-specific manifestations, ranging from cutaneous symptoms to abdominal and neuropsychiatric acute conditions. In the presented case, the absence of pathognomonic symptoms, associated with the rarity of the disease, hinders both the diagnosis and the beginning of the appropriate treatment. **Final comments:** The inclusion of porphyria as a differential diagnosis of acute neurovisceral cases is essential to ease the early identification of these conditions. Therefore, specific laboratory and molecular genetic testing are essential for the accurate diagnosis. With the proper diagnosis and treatment, it is possible to control the progression of the disease, in addition to significantly improving the quality of life and reducing mortality associated with Porphyria Variegata.

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Positive outcome of natalizumab therapy in multiple sclerosis with leptomeningeal lesion: a case report

Gustavo Silva Schafascheck[1], Laísa Fernandes Bonatto[1], Roberto Navarro Rocha Filho[1], Luiza Lima Oliveira[1], Viviane Tavares Carvalho Crelier[1], Gutemberg Augusto Cruz dos Santos[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Gustavo Silva Schafascheck; Email: gustavo.med2016@gmail.com.

Case presentation: A 60-year-old patient was diagnosed with Multiple Sclerosis (MS) in September 2022 and started treatment with Copaxone. In January 2023, a new lesion was identified, and by July, a positive test for latent tuberculosis required additional treatment. In November 2023, a subsequent MRI scan revealed an occipitotemporal leptomeningeal lesion, a rare occurrence in MS. Due to the lack of improvement with the initial treatment, the medical team decided to initiate treatment with Natalizumab, an immunomodulator, in November 2023. After two months of Natalizumab therapy, a follow-up MRI showed a substantial reduction in the leptomeningeal lesion, accompanied by significant clinical improvements, including reduced fatigue, improved vision, and enhanced walking stability. **Discussion:** This case underscores the importance of early and personalized intervention in MS, especially in atypical presentations such as occipitotemporal leptomeningeal involvement. Recent literature highlights the association between leptomeningeal involvement and rapid disease progression, reinforcing the need to tailor therapeutic strategies to individual patient profiles. The use of Natalizumab, as demonstrated in this case, offers a promising alternative for challenging cases where conventional treatments may not suffice. The patient's positive response suggests that the choice of targeted immunomodulators can be crucial in managing complex disease activities and improving patients' quality of life. **Final comments:** The response to Natalizumab in this patient with an atypical presentation of MS emphasizes the need to re-evaluate conventional treatments and consider personalized therapeutic options. This case reinforces the importance of individualized strategies, which are crucial for addressing the complexities of MS and achieving better clinical outcomes. Further studies are needed to optimize outcomes in complex MS presentations and to validate the effectiveness of specific therapies in patient subgroups with distinct disease characteristics.

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Possible co-occurrence of CNS demyelination and subdural haematoma caused by TNF-alpha inhibitors

Alberto Pereira Firmino Filho[1], Maria Weryca de Souza Belo Silva[1], Rodrigo Alencar e Silva[1], Joanisson Rubens Gomes Diniz[1], Paulo Santiago Morais Brito[1], Heitor Caetano Santos[1], Miguel Inacio Silva Gomes[1], Fatima de Menezes Dantas[1], Felipe Olobardi Freire[1], Nathalia Lindsay Pereira de Queiroz[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Alberto Pereira Firmino Filho; Email: firminofilho.ap@outlook.com.

Case presentation: A 77-year-old man with psoriatic arthritis was treated with adalimumab for twelve months. He presented with a sudden state of confusion, tetraparesis, and urinary and fecal incontinence. Neurological examination revealed mild asymmetric tetraparesis. There was no history of head trauma or laboratory abnormalities. Magnetic ressonance imaging of the brain and cervical spine revealed right spontaneous subdural hematoma and cervical demyelinating event. **Discussion:** Rare side effects associated with TNF-α blocker therapy have been reported in the literature, including cerebrovascular disease and demyelinating disorders. To our knowledge, no simultaneous injuries of a different nature caused by the use of adalimumab have been described. **Final comments:** We must be attentive and maintain close neurological surveillance of patients undergoing treatment with anti-TNF-alpha therapies to be able to suspect and diagnose possible complications arising from their use, even the atypical occurrence of simultaneous side effects.

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Post radiculomedullary trauma intracranial hypertension

Mayhara Rosany da Silva Santiago[1], Ana Caroline Paiva Simeão[1], Raissa Josefa Pereira de Moura[1], Nereu Alves Lacerda[1], Maria Luisa Roberto Lima Lisboa[1], Natália Nunes Tenório[1], Ana Beatriz Roberto Lima Borba[1], Ana Carolina Martins Fragoso Carneiro[1], Heloisy Maria Nunes Galvão[1], Bárbara Matos Almeida Queiroz[1]

[1]Hospital Pelópidas Silveira, Recife PE, Brazil.

Correspondence: Mayhara Rosany da Silva Santiago; Email: santiago.mayhara@gmail.com.

Case presentation: A 46 years old man presented with reduced bilateral visual acuity, described as progressive, within a month of initial perception, and worsening of symptoms in the last 3 days preceding the hospital admission, associated with bilateral ear tinnitus. The patient mentioned a fall from about 4 meters high 5 months before the actual admission, and was seen to have a vertebral lesion, in T12, and opted for conservative treatment. On neurologic examination the low visual acuity was confirmed with 20:200 in the right eye and 20:100 in the left eye, in the fundoscopy was seen bilateral papilledema, and in the amsler grade the patient described tunnel vision. Initial evaluation with neuroimage with head CT scan and magnetic resonance had no signs of mechanical obstruction of liquorice flux, and showed indirects signs of intracranial hypertension such as the empty sella sign. The liquorice puncion confirmed the major hypothesis, with an opening pressure of 60 cmH2O. The laboratorial liquorice evaluation was negative to infections and other inflammatory disorders. The investigation progressed with an arteriography that showed a complex dural fistula, with multiple origins, more prominent in the occipital and vertebral arteries, promoting local venous hypertension and secondary intracranial hypertension. The patient was transferred to the neurosurgery sector, where the fistula embolization was programmed through a hemodynamic approach. **Discussion:** Dural arteriovenous fistulas are a vascular abnormality characterized by connections between dural arteries and venous sinus, resulting in intracranial hypertension. Multiple etiology are possible, being the most common, in adults, venous sinus thrombosis, but is also possible as a result of thrombophilias and head trauma. **Final comments:** When confronted with a case of severe refractory cranial hypertension, the active search of past head trauma and investigation of dural fistulas are essential and may be a key factor for the diagnosis and adequate treatment and, consequently, the prognosis.

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Post-endoscopy bilateral cortical infarction presenting as a stroke chameleon: a case report

Raíssa Gomes Castro e Souza[1], Luis Felippe Barros e Silva Pereira Ribeiro[1], Marcus Vinícius Buratti Leal[1], Maria Paula Oiticica de Jesus[1], Sabrina Vechini Gouvêa[1], Izabel Crystine Pereira Barbosa[1], Wagner Mauad Avelar[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Raíssa Gomes Castro e Souza; Email: gcsraissa@gmail.com.

Case presentation: A 66 year-old woman presented with a history of sudden onset rightward gaze preference, quadriplegia, spasticity in inferior limbs and right arm as well as global hyperreflexia with Babinski, Trömner and Hoffmann's signs elicited bilaterally. She also displayed myoclonic spasms in both legs and right arm. Vital signs upon arrival consisted of blood pressure of 132/70mmHg, heart rate 100bpm and peripheral oxygen saturation of 95%. The neurological signs had arisen after a routine endoscopy, as soon as the patient roused from anaesthesia, having been under conscious sedation with IV midazolam (3.5mg), fentanil (30mcg) and propofol (70mg). Prior medical history consisted of glaucoma, dyslipidemia, an ex-smoking habit and esophagectomy for esophageal cancer two months before. Urgent head CT scan showed no abnormalities. Further evaluation with diffusion-weighted MRI revealed acute cortical infarct in precentral gyrus regions in both cerebral hemispheres. Relevant laboratory values included haemoglobin of 7.8g/dl (baseline 10.4g/dl two months prior). Poststroke assessment with electrocardiogram, echocardiogram, transcranial doppler ultrasound and computed tomography angiography was normal. Given the fact that additional investigations yielded no unwonted findings, the most likely diagnosis in this patient remained a hypoxic-ischaemic cerebrovascular injury. She was followed in the outpatient setting and experienced gradual improvement on both motor and speech functions. **Discussion:** Cerebrovascular events following endoscopy have been reported as a consequence of the cardiopulmonary effects, which include periods of hypotension, arrhythmias and hypoxemia. Hypoxic-ischaemic injury to the CNS is typically bilateral and symmetrical, and the damage is most prominent in grey matter structures such as cerebral cortex, basal ganglia, thalami and hippocampi. This type of injury is most commonly seen as a result of cardiac arrest, drowning or asphyxiation but can also occur as a complication of invasive procedures, especially in the presence of predisposing features such as older age and anaemia. As to the clinical aspects, this patient displayed an unusual stroke presentation - mainly related to the motor deficits which encompassed all of the four limbs, but also positive signs such as spasticity and myoclonia. For this reason upon admission the hypothesis of a stroke remained questionable, until proven otherwise with the aid of MRI. **Final comments:** This case has been reported in account of its unique clinical features as well as its presumed aetiology. Being aware of atypical stroke presentations is a resourceful perspective in acute clinical settings, for it might lead to prompt evaluation and timely intervention in dubious cases. Furthermore, although rarely reported, hypoxia-induced stroke can occur during procedures, and discussing means to prevent it is advisable.

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Posterior reversible encephalopathy syndrome (PRES) in an infant: case report

Rodrigo Ibañez Tiago[1], João Ormond de Miranda[2], Jéssica Feltraco[2], Karina Galli Cardoso Mello Roberto[2], Isabela de Carvalho Florêncio[3], Gabriel Pereira Braga[3], Glauco Almeida[3]

[1]Universidade Federal de Mato Grosso do Sul, Faculdade de Medicina, Campo Grande MS, Brazil. [2]Hospital São Julião, Campo Grande MS, Brazil. [3]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.

Correspondence: Rodrigo Ibañez Tiago; Email: rodrigoibaneztiago@gmail.com.

Case presentation: An 8-year-old female with a three-day exacerbation of asthma developed severe headache and projectile vomiting, without fever. Initial laboratory tests were normal, and cranial computed tomography revealed mucosal thickening of the left maxillary sinus without intracranial abnormalities. After 12 days, she experienced a seizure, progressing to right-sided hemiparesis and bilateral visual impairment. Cerebrospinal fluid analysis showed normal cell count and other parameters. Ophthalmological evaluation revealed visual acuity of 20/25 in both eyes, intraocular pressure of 12 mmHg bilaterally, normal anterior segment examination, and blurred optic disc margins on fundoscopy. Visual field examination (24-2) showed diffuse sensitivity loss in the right eye, particularly in the temporal region, and nasal hemifield loss in the left eye. Optical coherence tomography (OCT) demonstrated increased nerve fiber layer thickness and anterior displacement of the RPE-Bruch's membrane complex, suggesting increased intracranial pressure. The autoimmune panel revealed fine speckled pattern ANA 1:128 and weakly reactive anti-cardiolipin IgG. Brain magnetic resonance imaging showed vasogenic edema affecting bilateral frontal, parietal, and occipital lobes, with a pattern consistent with vascular boundary zones and mass effect, with no abnormalities on angiography. Clinical evaluation indicated elevated blood pressure (170/90 mmHg). Captopril was initiated with subsequent improvement in clinical and neurological status. Further rheumatological investigation revealed a nuclear fine speckled pattern ANA 1:160. Follow-up showed resolution of papilledema and significant improvement in visual field defects approaching normal parameters within 4 months, confirming the diagnosis of Posterior Reversible Encephalopathy Syndrome (PRES). **Discussion:** PRES is a clinical and radiological syndrome characterized by acute encephalopathy with vasogenic edema, often associated with elevated blood pressure, preeclampsia, autoimmune, and infectious diseases. The pathophysiology involves cerebral autoregulatory dysfunction and increased blood-brain barrier permeability, leading to cerebral edema. The association with positive autoimmune markers may suggest an underlying autoimmune predisposition, although causality remains unclear. **Final comments:** This case emphasizes the importance of considering PRES in diagnosing children with seizures and acute neurological symptoms. Effective management includes strict blood pressure control. Recovery is generally complete with appropriate intervention, as observed in this patient. Investigating underlying diseases is crucial to prevent recurrence and ensure a favorable prognosis.

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Post-irradiation internal carotid artery vasculopathy: a case report

Samia Talise El Horr de Moraes[1], Fernanda Michely Nicoli[1], Bárbara Akemy Barbosa Cruz[1], Juliano André Muzzio[1], Claudia Suemi Kamoi Kay[1]

[1]Universidade Federal do Paraná, Complexo do Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Samia Talise El Horr de Moraes; Email: samiatalise1@gmail.com.

Radiotherapy used to treat various neoplasms can result in acute, subacute, and late clinical syndromes. Cranial blood vessels may be affected by radiation, resulting in late structural changes. Knowledge of the clinical picture, the time interval from radiation therapy, and additional tests will assist doctors in diagnosis and treatment. A 17 years-old female patient presented with a changed-pattern headache. She had a previous history of migraine without aura and right ocular rhabdomyosarcoma treated with resection, chemotherapy, and radiotherapy at 4 years-old, using monthly combined injectable contraceptive. Physical and neurological examinations were unremarkable, as well as further investigation with laboratory tests, cerebrospinal fluid, and cranial tomography. However, cerebral angio-MRI showed signs of probable inflammatory process affecting the right internal carotid artery with significant hypoflow of the ipsilateral middle cerebral artery. Also, transcranial Doppler was performed presenting significant decrease in flow velocity and pulsatility index in the right middle cerebral artery, reversal of flow direction in the right anterior cerebral artery, intracranial right internal carotid artery with significant reduction in pulsatility index (15/0.35). These findings were compatible with the diagnosis of internal carotid artery stenosis after radiotherapy, and treatment with cilostazol was initiated. It is important to remind general neurologists that vasculopathy is a possible complication after radiotherapy. The prevalence of carotid stenosis in patients with head and neck neoplasia varies from 10% to 50% after radiotherapy and can lead to TIAs or infarction. Children are more sensitive to vascular injuries and can develop moyamoya syndrome and vascular malformations. The average interval from the end of irradiation to the onset of ischemic symptoms is 5.4 years in children. Post-treatment follow-up of these patients is essential to have an early diagnosis and approach of these kind of complication, minimizing permanent neurological deficits.

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Post-pump chorea: a rare and persistent neurological complication of cardiopulmonary bypass

Cândida Driemeyer[1], Renato Dumbá Monteiro de Castro[1], Ana Letícia Amorim de Albuquerque[2], Daniel Teixeira dos Santos[2], Arlete Hilbig[1], Carlos Roberto de Mello Rieder[1]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil. [2]Santa Casa de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Cândida Driemeyer; Email: candidaplena@gmail.com.

Case presentation: A 42-year-old woman was referred to the neurology outpatient clinic due to involuntary movements. The symptoms started 1 day after thromboendarterectomy with cardiopulmonary bypass support due to thromboembolism and secondary pulmonary hypertension when she was 34 years old. Before thromboembolism, the patient had no comorbidities. On neurological examination, the patient presented with chorea affecting the face, tongue, neck, trunk and limbs. Hemogram, electrolytes, renal, hepatic and thyroid function were normal. Investigating the procedure, it needed 248 minutes of extracorporeal circulation with aortic clamping and intermittent circulatory interruption. Hypothermia at 18 degrees Celsius was required for 144 minutes. Computed tomography of the skull at the time showed hypodensity with a sequelae appearance of 0.4cm in the lentiform nucleus on the left and investigation by magnetic resonance imaging two months after the opening of the neurological condition showed hyperintensity on the T1 sequence bilaterally in the pale globus, suggestive of metabolic alteration. Initial rheumatological investigation returned normal. The recently performed MRI shows minimal sequelae in the globus pallidus, more evident on the left, without hypersignal and without restriction to diffusion. Currently using haloperidol 1 mg once a day with stability of symptoms, because of low tolerance to the adverse effects of atypical neuroleptics. **Discussion:** Suggested risk factors for neurological sequelae such as chorea after cardiopulmonary bypass are age of onset, latency of onset, underlying disease, response to treatment and surgical characteristics (minimum body temperature, time of total circulatory arrest and control of gaseous microembolism). There are several pathophysiological mechanisms that can explain the emergence of this symptom post extracorporeal circulation, although the exact pathogenesis has not yet been revealed: microembolic events affecting basal ganglia, lactic acidosis induced by hypothermia, increased blood viscosity with resultant thrombi formation and also the circulation of inflammatory agents and hemolysis products that occur in the stages of the circuit. The higher frequency observed in children remains unclear. **Final comments:** The appearance of chorea in an adult patient requires extensive investigation, which includes rheumatological diseases, toxic-metabolic insults, use of continuous medications,

neoplasia, cerebrovascular and autoimmune diseases. In our case, the abrupt appearance of chorea 1 day after pulmonary thromboendarterectomy and the absence of laboratory changes leads us to the probable diagnosis of chorea after cardiopulmonary bypass, or "post pump chorea". The detailed description of the occurrences of neurological sequelae secondary to extracorporeal circulation has an important contribution to clarifying and differentiating the multiple causal factors.

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Post-stroke delirium in younger is associated with worse long term functional outcome

Gabriel Souza Suzart[1], Iva Taiuan Fialho Silva[1], Joao Pedro[1], Vinicius Mendez[1], Luiz Paulo Oliveira Martins[1], Marivaldo Yslan Silveira Leal[1], Luísa Dias Moreno Aravena[1], Thais Barbosa de Oliveira[1], Gabriel Souza Medrado Nunes[1], Pedro Antonio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Luiz Paulo Oliveira Martins; Email: luizpom2000@gmail.com.

Background: Although post-stroke delirium is a common condition in the elderly, younger patients are less prone to have it and are recognized for favorables outcomes. **Objective:** To assess the impact of delirium on functional outcomes of stroke patients in different age groups. **Methods:** Prospective cohort of stroke patients admitted in a stroke unit from 2017 to 2023. Delirium was diagnosed using the CAM-ICU. Patients with delirium were divided into ≥60 years or <60 years and compared to non-delirium patients. The main outcome was mRs>2 in 90 days post-stroke. **Results:** A total of 585 patients were enrolled. Delirium occurred in 115 patients (19.7%), of which 81 (70.4%) were elderly and 34 (29.6%) were younger. Even though they had lower Charlson Comorbidity Index (median, 3 vs 5 points; p<0.001), younger patients affected by delirium had higher NIHSS scores than elders and no-delirium patients [12 (±5) vs 10 (±6) vs 8 (±5); p<0.001] and lower ASPECTS scores [IQR, 8 (4-9) vs 8.5 (6-10) vs 9 (8-10) points; p<0.001]. Delirium in younger people showed higher odds of long term functional dependence than in elderly (OR [IC95%], 2.4 [1.8-3.1; p<0.001] vs 2.0 [1.6-2.5; p<0.001], compared to no-delirium patients. **Conclusion:** Although delirium is less frequent in younger people, this group may have a poorer prognosis. Delirium in younger patients may be predisposed by larger lesions and higher neurological impairment. This is the first study to investigate the influence of age on post-stroke delirium prognostic value.

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Post-traumatic epilepsy: biological and social description

Giulianna Luzia Pereira[1], Livia Nogueira de Godoi[1], Helena Rubini Nogueira[1]

[1]Universidade Municipal de São Caetano do Sul, São Caetano do Sul SP, Brazil.

Correspondence: Giulianna Luzia Pereira; Email: giuliannaluzia2013@gmail.com.

Background: Post-traumatic epilepsy (PTE) refers to recurrent and spontaneous seizures caused by complex and chronic repercussions resulting from traumatic brain injury (TBI). Primary injuries are those that occur seconds to minutes after the trauma, while secondary injuries involve physiological mechanisms and progressive damage, as is the case with PTE. In addition, this epileptic process tends to be resistant to the anticonvulsant drugs in use, with 30% of patients suffering from the disease refractory to treatment. **Objective:** To understand the basic biological process of post-traumatic epileptogenesis and the social consequences of this condition. **Methods:** This is an integrative review made with PUBMED as a database, using the descriptors "Epilepsy" AND "Brain Injuries, Traumatic"; and "Epilepsy, Post-Traumatic". The inclusion criteria were: publication date within the last 5 years and free access to the full text. The exclusion criteria chosen were: not having post-traumatic epilepsy as the main theme, being written in chinese and repeated texts. **Results:** 34 articles were selected, of which 25 explained the pathophysiology and 12 the impact that its occurrence would have on the patient's life. With regard to etiopathogenesis, mechanisms such as the breakdown of the blood-brain barrier, oxidative damage and the release of chemokines are fundamental to epileptogenesis. At the same time, the social impact of Post Traumatic Epilepsy can be seen both for the patients themselves and for life in society, since they can suffer disorders that disable them from executive activities. **Conclusion:** Post-traumatic epilepsy is a highly complex condition that requires specialized care and which repercussions have significant potential for chronicity and incapacity in the patient's activities of daily living. Given the biopsychosocial and economic impact involved in this condition, we highlight the importance of further studies to promote collective policies that specifically address the Brazilian population.

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Post-vaccinal dengue encephalitis and dengue shock syndrome: case report

Lucas Oliveira Mourão[1,2], Cíntia Silveira[1], Sônia Maria Dozzi Brucki[2], Fernando de Mendonça Cardoso[1]

[1]Hospital Glória D'or, Rio de Janeiro RJ, Brazil. [2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Lucas Oliveira Mourão; Email: lucas.omourao@gmail.com.

Case presentation: A 87-years-old male, Rio de Janeiro resident, hypertensive, diabetic, with non-dialytic chronic renal disease, diabetic polyneuropathy, and normal pressure hydrocephalus (NPH), walking with aid, received his first vaccination of a live attenuated dengue tetravalent vaccine (QDenga®) on May 16th 2024. After 6 days (22nd of May) he became wheelchair-dependent. On the 23rd of May he had headache. One day later, he lost consciousness for 10 minutes and was admitted to the Intensive Care Unit for a syncope workup: he was oriented on space and time and didn't present any neurological focal deficit. Brain MRI was unremarkable for acute lesions, displaying ventriculomegaly and a callosal angle of 82 degrees compatible with previous NPH diagnosis. However, the following day he had nausea and progressed to torpor, hemodynamic shock and acute renal failure with need for norepinephrine and continuous hemodialysis. Over the next 24 hours he became comatose with need for orotracheal intubation. On May 27th he underwent lumbar puncture and Dengue virus was identified through PCR on cerebrospinal fluid (CSF). Other CSF findings: initial pressure of 33 cm H2O; 1 cell/mm3 (rare lymphocytes); protein 24 mg/dl; glucose 60 mg/dl. **Discussion:** An elder received the first vaccination of QDenga® (drug TAK-003), a live attenuated dengue tetravalent vaccine licensed for dengue prevention in Brazil, for people up to 60 years of age. Phase 3 double-blind, randomized, placebo-controlled studies assessed QDenga®'s efficacy on 702 children from 4 to 16 years living in endemic areas of Asia and Latin America (DEN-301) and 379 adults from 18 to 60 years living in parts of the United States considered non-endemic for dengue (DEN-304). Open-label, phase 3 study DEN-307: 174 United States adults (18 to 60 years); sepsis was reported 2 days after second dose and wasn't considered drug-related. QDenga® studies reported mild neurological adverse events (headache, somnolence and dizziness), but it is known that vaccines may cause severe neurological adverse events and that dengue can cause a wide range of neurological manifestations: encephalopathy, acute encephalitis, chronic progressive panencephalitis, stroke cranial neuropathies, acute demyelinating encephalomyelitis (ADEM), acute cerebellitis, Guillain-Barré Syndrome, acute transverse myelitis and acute myositis. Dengue encephalitis diagnosis was made due to impaired consciousness and dengue virus PCR detection on CSF. In a dengue-endemic region dengue is the main cause of encephalitis with normal CSF cellularity, as observed. He also had dengue shock syndrome: hemodynamic shock and organ failure (acute kidney injury). **Final comments:** to our knowledge we report the first case of post-vaccinal dengue encephalitis and dengue shock syndrome. Possibly a result of the elder's frail immune system response after receiving a live attenuated dengue tetravalent vaccine. Patient's age group wasn't studied.

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Potentials and limitations of genetic testing in autism spectrum disorder

Luisa Maria Resende Morais[1], Gleison Carlos Arantes Filho[1], Clara Oelze Cardoso[1], Gustavo Lucas Viana dos Santos[1], Sanny Kemelly Miquelante Yoshida[1], Leonardo Brandão Barreto[1]

[1]Universidade Federal de Ouro Preto, Ouro Preto MG, Brazil.

Correspondence: Luisa Maria Resende Morais; Email: luisamresendem@gmail.com.

Background: The diagnosis of Autism Spectrum Disorder (ASD) is challenging due to considerable clinical and genetic heterogeneity. However, with the development of molecular technologies, it has been understood that genetic findings such as copy number variations (CNV), single nucleotide variations (SNV), and small insertions/deletions are associated with the disorder, enabling the use of genetic testing for etiological investigation. **Objective:** To systematize the current evidence in the literature on the potentials and limitations of genetic tests in ASD. **Methods:** A systematic review of the databases: PubMed, LILACS, SciELO, and Web of Science using the descriptors: "Autism Spectrum Disorder" and "Genetic Testing". Inclusion criteria were articles published in the last 5 years, and exclusion criteria were systematic review papers or those not addressing the studied proposal. **Results:** A total of 762 studies were found, and after applying the criteria, 21 were selected. For the family, the main reported benefits were providing information for reproductive planning (n= 7 studies) and understanding the disease's etiology, which led both to a change in parents' perceptions of ASD, with a tendency to blame their children and themselves less for the presence of this condition (n=2 studies), and to an improvement in their ability as caregivers (n=1 study). For the patient, early and definitive diagnosis stood out (n= 6 studies), considering that many neurodevelopmental disorders have overlapping clinical presentations, making diagnosis based solely on symptom investigation difficult. Additionally, 5 articles highlighted that genetic tests provide prognostic information, aiding in directing earlier and more personalized interventions, as some genetic variations are associated with specific comorbidities and clinical presentations. However, 2 of these articles emphasized that genetic testing cannot definitively predict prognosis, as the same genetic variation can lead to different phenotypic profiles. Another important limitation is the current low diagnostic yield, such as exome sequencing, with a yield of 17.1% (11%-25%) for ASD, generating many negative results that can be false negatives and do not rule out a genetic cause (n=7 studies). Finally, the difficulty of interpreting results by non-geneticist doctors, in addition to the high

cost for families and the health system, was pointed out (n=2 studies). **Conclusion:** Many medical societies recommend the use of genetic tests, which, despite having several potentials, still have limitations to be overcome. It is expected that, with more studies directed at the genetic architecture of ASD, these tests will become more accurate and advantageous. It is essential that healthcare professionals inform families about these difficulties to align expectations regarding genetic tests.

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Precision medicine in the FGF12 gene related to refractory epilepsy responsive to phenytoin: a case report

Joana Laura Aihara Barbosa[1], Lucas Henrique de Moura Rogerio Garcia[1], Matheus Rodrigues Nogueira[1], Ahmad Kassem El Zein[1], Josenice Gomes Trindade Seidel[1], Lucas D'Andrea Pereira Souza[1], Luiz Gustavo Brenneisen dos Santos[1], Paula Fiuza Rodrigues de Medeiros Lacerda[1], Eliana Garzon[1], Luciana Midori Inuzuka[1]

[1]Hospital Sírio-Libanês, São Paulo SP, Brazil.

Correspondence: Joana Laura Aihara Barbosa; Email: joana.lab@gmail.com.

Case presentation: 12-year-old boy, unrelated parents, uncomplicated pregnancy, full term, no relevant family and neonatal history, no dysmorphic features. The first afebrile seizure was at 3 months (hypotonic, cyanotic, hypoventilation, less than a minute), neurological exam, cerebrospinal fluid, brain MRI and electroencephalogram (EEG) were normal. After a week he had recurred focal motor seizures. Carbamazepine was prescribed. Until 3 years old, the seizures remained every 6 months, and an episode of status epilepticus occurred. The patient stayed seizure-free for two years and antiseizure medication (ASM) was withdrawn. After one month seizures had recurred, and levetiracetam, pyridoxine, clobazam, and sodium valproate were administered unsuccessfully. EEG demonstrated normal background and sporadic centrotemporal, frontocentral, bifrontal, central epileptiform discharges. The genetic panel at this time (2017) had no pathogenic variants. In the following two years, although the patient had a normal neuropsychological test and no changes in neurodevelopment, the seizures remained bilateral, tonic-clonic, generalized, and pharmacoresistant. Exome revealed a pathogenic variant of the FGF12 gene, missense, resulting in a substitution of glycine-to-serine [(Chr3: 192.335.441 C>T, p(Gly112Ser ENST 00000454309)]. After this finding we prescribed phenytoin. The patient remains seizure-free to date. **Discussion:** Recent studies have identified mutations in the fibroblast growth factor 12 (FGF12) gene as a potential genetic cause of epilepsy and neurodevelopmental disorders. The gene encodes a cytosolic binding protein that enhances the voltage-dependent fast inactivation of neuronal sodium channels, modifying their excitability. Pathogenic variants in FGF12 have few published case reports, with most other cases with neuropsycomotor development disorder and refractory epilepsy (Developmental and Epileptic Encephalopathy 47). In our patient, neurodevelopment was normal and revealed an expansion of phenotype. Genetic panel had identified no pathogenic variants, at this time FGF12 gene had not yet been related to epilepsy. Due to the persistence of the seizures, an exome was requested and revealed a pathogenic variant of the FGF12 gene. Phenytoin was utilized in other published cases of refractory epilepsy with this gene mutation, owing to its molecular mechanism, which stabilizes neuronal membranes by blocking voltage-gated sodium channels, thereby inhibiting excessive neuronal firing. New generation sequencing is essential in refractory epilepsy guiding ASM strategies for better outcomes. **Final comments:** Genetic causes of epilepsy, particularly FGF12 gene mutations, highlight the relevance of precision medicine strategies for treatment selection.

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Predictors of azathioprine therapeutic success in patients with neuromyelitis optica spectrum disorder

Ana Carolina Amaral de Andrade[1], Gabriel de Deus Vieira[1], Mariana Moreira Soares de Sá[1], Alfredo Damasceno[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Ana Carolina Amaral de Andrade; Email: anaca_andrade@yahoo.com.br.

Background: Approximately 44.9% of patients using azathioprine (AZA) for the treatment of Neuromyelitis optica spectrum disorder (NMOSD) have therapeutic failure. **Objective:** To evaluate prognostic factors of treatment failure of AZA in patients with NMOSD. **Methods:** Demographic and clinical aspects of 49 patients with NMOSD, according to fulfilling INPD 2015 diagnostic criteria, were analyzed in regards the therapeutic success of AZA 2 to 3 mg/kg/day for 6 months, 79,6% of these patients, were reagents for serum anti-aquaporin 4 antibodies (AQP4-IgG) using the cell based essay (CBA) methodology. We consider therapeutic failure to be the replacement of AZA due to relapse or adverse effects. Severe relapse were defined as those that progressed without improvement after plasmapheresis; bilateral visual impairment or worse than 20/200; muscle strength less than grade III with gait or sphincter disorder. Univariate and multivariate logistic regression were performed to evaluate factors associated with treatment failure (gender, ethnicity, AQP4-IgG positivity, ages at the first outbreak and at the onset of AZA, severity and type of the first outbreak, number of pre-AZA relapse, and time between the first relapse and the onset of AZA). **Results:** The group of treatment failure with AZA had 90.9% of female subjects, 59.1% non-white, who were between 18 and 40 years old in the first outbreak (63.6%) and at the beginning of AZA (40.9%), positive for AQP4-IgG (77.27%), 27.3% had at least 1 severe pre-AZA relapse, with severe optic neuritis being the most common type of first relapse (40.9%), 81.8% started AZA as their first treatment, 1 to 5 years after the first relapse (45%) and after the occurrence of 2 to 4 relapse (68.2%), with an annualized relapse rate (AAR) pre AZA between 1 and 5 (50%). After 180 days of AZA use, 22.27% of the patients took up to 6 months to present the first therapeutic failure and 50% of them had AAR post-AZA of up to 1. After logistic regression analysis, the factors associated with failure with AZA in the total sample were: white ethnicity (p=0.023; OR=0.183, 95%CI 0.025 - 0.763), age of onset AZA (p=0.017; OR=0.892, 95% CI 0.812 - 0.980) and number of relapses before the onset of AZA (p=0.049; OR=1.675, 95%CI 1.003 - 2.795). Similar results were observed in logistic regression analyses in patients who tested positive for AQP4-IgG: white ethnicity (p=0.037; OR=0.026), age at first relapse (p=0.039; OR=0.809) and number of relapse before AZA (p=0.028; OR=3.606). AZA is considered a first-line therapy for NMOSD in emerging countries, in which monoclonal antibodies are in the process of being incorporated into health services, however, almost 45% of patients using AZA have therapeutic failure in the management of NMOSD. **Conclusion:** Knowledge of predictors of treatment failure with AZA may contribute to determining which patients should start treatment with monoclonal antibodies.

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Predictors of death during internmnent in patients with hemorrhagic stroke and high risk of obstructive sleep apnea: a transversal study

Aurea Maria Lago Novais Castello Branco[1], Juliana Cerqueira Gomes[1], Marina Soledad Benitez Bitencourt[1], Thaise Braga de Oliveira[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Renan Carvalho Castello Branco; Email: castellorenan@yahoo.com.br.

Obstructive sleep apnea is a disease of upper airways, with great association with stroke through diverse mechanisms. Although, it still lacks in literature tha relation with hemorrhagic subtype. This study aims to stablish the predictors of death in patients with hemorrhagic stroke and high risk of obstructive sleep apnea. We performed a transversal study, part of a cohort of stroke patients. They were evaluated about risk of obstructive sleep apnea with SOS and STOP-Bang scores. Data of previous comorbidities and stroke profile were collected. Comparison between groups was done with T test/chi-schare/Mann-Whitney, considering p< 0.1, and multivariate analysis with logistic regression, considering p< 0.05, to outcome of death during internment. We obtained 81 patients, of which 64.2% were classified as high risk with STOP-Bang (19 hypertensive, 18 traumatic, 6 aneurysmatic) and 52% with SOS (16 hypertensive, 14 traumatic, 4 aneurysmatic). Considering high risk of STOP-Bang, there was higher prevalence of obesity (p=0.02), hypertension (p=0.02) and atrial fibrillation in low risk (p=0.05). Considering high risk of SOS, we obtained obesity (p=0.03) and sleep disorder (p=0.09). Patients with higher risk of OSA evolved more to death (p=0.08). There were no independent factors related to death during internment. The difference in obesity and hypertension was due to their character of comorbidity with obstructive sleep apnea. The stroke profile did not vary between groups, denoting a less preponderant role of obstructive sleep apnea in hemorrhagic stroke, although higher risk evolved more to death, demonstrating subjacent mechanisms of worst outcomes, not necessarily related to stroke. Further studies are needed to achieve a higher sample.

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Predictors of fast visual recovery in idiopathic intracranial hypertension: a cohort study

Ana Carolina Montouro Storarri[1], Annelise Akemi Higa Lee[1], Lorrane Campidelli Arthuzo[1], Luis Armando Sawada[1], Beatriz Marques dos Santos[1], Thassia Cavalcante de Oliveira Moraes Marques[1], Bruno Pissolati Mattos Gonzaga[1], Renan Barros Domingues[1]

[1]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Ana Carolina Storarri; Email: anastorarri@gmail.com.

Background: Idiopathic intracranial hypertension (IIH) is characterized by increased intracranial pressure without a discernible cause, often leading to visual impairment and papilledema. Understanding the factors influencing visual outcomes in IIH patients is crucial for improving management strategies. This study aimed to identify predictors of satisfactory visual outcome (SATISF) defined as the resolution of reduced visual acuity and/or disappearance of papilledema. **Objective:** To evaluate the factors associated with satisfactory visual outcomes in female patients with IIH. **Methods:** This retrospective cohort study included female patients diagnosed with IIH. The potential predictors of a satisfactory visual outcome evaluated were age, initial body mass index (BMI), cerebrospinal fluid (CSF) opening pressure (Pi), duration of symptoms, and the need to discontinue acetazolamide. Visual outcomes were categorized as SATISF or non-satisfactory (NAOSATISF). Independent sample t-tests were used for comparing means, and chi-square tests were employed for comparing proportions. The average interval between the first and second visual assessments was

55.4±48.4 days. Results The study involved the following comparative **Results:**

- Age: NAOSATISF 32.2±12 years, SATISF 35.9±18.8 years, P=0.3
- BMI: NAOSATISF 33.4±4.3, SATISF 33.7±4.3, P=0.8
- Pi: NAOSATISF 41.7±12.5 cmH2O, SATISF 39.3±13.2 cmH2O, P=0.5
- Duration of symptoms: NAOSATISF 352.25±566.5 days, SATISF 317.2±815.7 days, P=0.9
- Acetazolamide discontinuation: NAOSATISF 4.7%, SATISF 15.7%, P=0.24

None of the evaluated factors showed a statistically significant association with satisfactory visual outcomes. **Conclusion:** This study did not identify any significant predictors of satisfactory visual outcomes in IIH patients. The results suggest that age, BMI, CSF opening pressure, duration of symptoms, and the need for acetazolamide discontinuation do not significantly impact visual recovery. Further research is needed to better understand the factors contributing to the rapid neuro-ophthalmological recovery in IIH patients.

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Predictors of hemorragic transformation in patients with stroke and high risk of obstructive sleep apnea: a transversal study

Thaise Braga de Oliveira[1], Aurea Maria Lago Novais Castello Branco[1], Juliana Cerqueira Gomes[1], Marina Soledad Benitez Bitencourt[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Renan Carvalho Castello Branco; Email: castellorenan@yahoo.com.br.

Obstructive sleep apnea (OSA) is a disease of Upper airways associated with stroke through diverse mechanisms, but there is no clear association with hemorrhagic transformation. This study's aim is to determine the predictors of hemorrhagic transformation in patients with high risk of OSA. We performed a transversal study, part of a cohort of stroke patients. They were evaluated about risk of obstructive sleep apnea with SOS and STOP-Bang scores. Data of previous comorbidities and stroke profile were collected. Comparison between groups was done with T test/chi-schare/Mann-Whitney, considering p<0.1, and multivariate analysis with logistic regression, considering p<0.05, to outcomes of hemorrhagic transformation and its type (ECASS). We obtained 326 patients, 71% high risk with STOP-Bang and 51% with SOS (27% cardioembolic, 24% atherothrombotic, 11% de small vessels, 6% other causes e 31% undetermined; p=0.068), of which 12% evolved to hemorrhagic transformation. High risk had higher prevalence of male sex, hypertension (p<0.001), diabetes (p<0.001), ASPECTS (p=0.02), also they evolved more to death (p=0.03). Considering SOS, etiology of stroke (OR=1.6; IC=95% 1.1-2.4; p=0.013) and ASPECTS (OR=1.347; IC=95% 1.1-1.7; p=0.006) were independently associated to hemorrhagic transformation. Obesity was independently associated to ECASS I and II (p<0.001). Cardioembolic etiology is a determinant of higher risk of hemorrhagic transformation, due to the mechanism well described in literature of increased risk of hemorrhage in infarcted area with emboli and its inherent friable characteristic. In patients with high risk of obstructive sleep apnea, better admitional ASPECTS can be relevant to further worst outcomes, leading to a improvement of care in patients that appear to have better images.

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Predictors of quality of life in patients with myasthenia gravis treated in the unified health system

Antonio Lázaro Santos Soares Junior[1], Luísa Dias Moreno Aravena[1], Marivaldo Yslan Silveira Leal[1], Marcel Leal Ribeiro[2], Bruno Bacellar Pedreira[2], Alice Silva Oliveira[2], Jamary Oliveira Filho[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Antonio Lázaro Santos Soares Junior; Email: antoniolssj@ufba.br.

Background: Myasthenia Gravis (MG) is an autoimmune disease caused by antibodies targeting structures at the neuromuscular junction, leading to skeletal muscle weakness characterized by fatigability and fluctuating strength. These clinical manifestations create various difficulties for sufferers, compromising their quality of life. **Objective:** To evaluate predictors of quality of life in individuals with MG treated by the Unified Health System. **Methods:** This single-center cross-sectional study involved patients over 18 diagnosed with MG at a Unified Health System reference center. Current symptoms and quality of life were assessed using the "MG-Activities of Daily Living" (MG-ADL) and "MG-Quality of Life-15 revised" (MG-QoLr) scales, respectively. Relationships between variables were analyzed using univariate linear regression. Variables with a p-value less than 0.1 were included in the final model to identify factors independently affecting quality of life. The final multivariate analysis was conducted using multiple linear regression, with a significance level of 0.05. **Results:** Fifty-four individuals were recruited, predominantly female (72.2%), brown (46.3%), and high school graduates (35.2%). The average age was 44.7 (±14.5) years, with an onset age of 31.3 (±15.5) years and illness duration of 13.4 (±9.5) years. The predominant clinical form was generalized, affecting 37 (68.5%) participants, and the serological profile was anti-AchR positive in 26 (48.1%). The median MG-ADL score was 3 (1-6.25). Comorbidities were present in 39 (72.2%) participants. The median time to diagnosis was 7 (2-22.25) months, with 18 (33.3%) having a late diagnosis (over 12 months between symptom onset and diagnosis). Azathioprine was used by 51.9% and corticosteroids by 48.1%. At the time of the survey, 45 (83.3%) did not have private health insurance, and 32 (59.3%) reported difficulty accessing the public health network, with a median difficulty perception score of 6 (0-8) on a scale of 0 to 10. Of the total sample, 27 (50%) stopped working and/or studying after diagnosis. The median MG-QoLr score was 10.5 (3.75-15.25). The following variables with a p-value < 0.1 in simple linear regression were included in the multiple linear regression model: MG-ADL, age at disease onset, late diagnosis, use of azathioprine, corticosteroid therapy, interruption of work and/or study activities, and self-perceived difficulty accessing Unified Health System. MG-ADL (B=1.47; CI: 0.95 - 1.99) and late diagnosis (B=3.12; CI: 0.31 - 5.93) correlated with MG-QoLr in the final model, with all p-values < 0.05. No significant correlation was found with other variables. **Conclusion:** Higher MG-ADL scores and late diagnosis are predictors of poorer quality of life in patients with MG.

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Preliminary analysis of clinical and neurophysiological data of patients with Charcot-Marie-Tooth disease

Rafael Oliveira Vidon[1], Clarissa Neves Spitz[2], Izabela Jardim Rodrigues Pitta[2], Larissa Bittencourt de Carvalho Chaffin[2], Robson Vital[2], Ligia Rocha Andrade[2], Salim Balassiano[2], Eduardo Davidovich[2], Márcia Rodrigues Jardim[2]

[1]Universidade Federal do Estado do Rio de Janeiro, Hospital Universitário Gaffrée e Guinle, Rio de Janeiro RJ, Brazil.
[2]Universidade do Estado do Rio de Janeiro, Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.

Correspondence: Rafael Oliveira Vidon; Email: rafaelvidon@hotmail.com.

Background: Hereditary Sensory and Motor Neuropathy (HSMN), also known as Charcot-Marie-Tooth disease (CMT), is part of a heterogeneous group of genetic diseases where the peripheral nerve is primarily affected. CMT affects about 1 in every 2,500 people worldwide. Despite its prevalence, patients with the disease may take many years to have their diagnosis confirmed. More than 90 causative genes have been identified for this neuropathy, with diverse phenotypic manifestations. It can start as early as at birth or as late as the fourth or fifth decade of life. In terms of severity, some patients may need early assistance with walking, while others may have only mild symptoms. Etiological diagnosis of neuropathy avoids unnecessary, sometimes costly and harmful treatments. It allows for genetic counseling, guidance, and anticipation of possible morbidities, as well as identifying potential therapeutic options. **Objective:** The study aims to achieve a better understanding of the clinical and neurophysiological history characteristics in a population treated at Pedro Ernesto University Hospital (HUPE), State University of Rio de Janeiro. **Methods:** The study consists of a retrospective and prospective analysis through a clinical and neurophysiological database of patients with the syndromic diagnosis of sensory-motor polyneuropathy, who are treated at the neuromuscular disease outpatient clinic at HUPE and who have undergone genetic testing to confirm the disease. **Results:** Thirty-four patients with suspected hereditary neuropathy, seen between 2018 and June 2024, are being followed. Results Of the total, 32 patients from 27 families had a diagnosis of CMT, with the genetic mutation confirmed. The most frequent was the duplication of the PMP22 gene (43.7%, 14/32), followed by the MPZ gene (18.7%, 6/32), deletion of PMP22 (12.5%, 4/32), and MFN2 (6.25%, 2/32). The observed pattern was predominantly distal involvement in the lower limbs (96%, 28/29), with a slow and progressive course and initial manifestation of loss of strength (82.7%, 24/29), often associated with foot deformities (80%, 20/25). The age of onset of symptoms ranged from 0 to 53 years, with a median of 14.5 years. Most manifestations occur in the first two decades of life. Patients underwent neurological examination and the scales used to measure the degree of neurological disability were the Neuropathy Impairment Score - Lower Limbs (NIS-LL) and CMT neuropathy score (second version). The electromyography data of 25 patients were analyzed, with analysis of conduction velocities, amplitudes, and morphologies of motor and sensory potentials. In one patient with late onset and predominant sensory symptoms, a mutation in the BAG3 gene, a gene recently associated with axonal polyneuropathy, was identified. **Conclusion:** The study describes phenotypes associated with mutations causing CMT and contributes to expanding the spectrum of diagnostic possibilities in patients with peripheral neuropathy.

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Preliminary assessment of the use of the virtual brain in the study of patients with brain gliomas: a descriptive approach

Thalles Olivi de Almeida[1], Jose Reynaldo Walther de Almeida[2], Ana Carolina Campos[1], Brigida S. de Oliveira[1], Marcos Vinicius Calfat Maldaun[1]

[1]Hospital Sirio Libanes, São Paulo SP, Brazil.
[2]Instituto de Neurociências de Araraquara e de Barretos, Araraquara SP, Brazil.

Correspondence: Thalles Olivi de Almeida; Email: almeida.thalles@outlook.com.

Background: Brain gliomas represent a significant challenge for both diagnosis and treatment due to their heterogeneous nature and their ability to alter the functionality of surrounding brain regions and at a distance. Mathematical models can contribute to the prior understanding of

possible postoperative complications. However, effective parceling of MRI images containing gliomas is complicated by the presence of the tumor, which can displace or infiltrate functional areas, making it difficult to accurately identify healthy and affected regions. MRI with Diffusion Tensor Imaging (DTI) and fMRI offer the possibility to study the different structural connectivities both visually and through validated models. The Virtual Brain (TVB) emerges as a potentially powerful tool, enabling the simulation of brain activity and analysis of the impact of gliomas on connectivity and neural function. **Objective:** To preliminarily explore in 3 glioma patients and 3 normals the application and difficulties of TVB in analyzing the influence of gliomas on brain structure and function. **Methods:** We conducted the parceling of T1 and DTI MRI of available images. The process was supervised until the result was in accordance with the neurosurgeon's understanding using the Glasser atlas, which includes 180 cortical regions. We applied the Wong-Wang model and carried out simulations in the TVB environment and the statistics in the R platform, using local and global efficiency, and MPR complexity, in an exploratory search for insights into the organization and efficiency of the brain tumor regions. **Results:** Evaluation of the pipeline - The parceling of MRI images in patients with tumors revealed the complexity of identifying functional areas in brains with gliomas. Regions of necrosis need to be adequately differentiated from tumor areas with functional potential. Evaluation of peritumoral structural connectivities in relation to the total connected areas in both hemispheres - The mean MPR complexity in tumor regions was 0.207 compared to 0.373 in contralateral homologue regions ($p<0.05$). The means of global and local efficiency were also significantly different between tumoral (0.20) and contralateral homologue regions (0.36) ($p<0.05$). In the control group, there were no differences ($p>0.05$). **Conclusion:** The implementation of The Virtual Brain (TVB) in the analysis of gliomas can be an exceptionally valuable tool, offering new perspectives on the structural and functional modifications caused by these tumors in the brain. The ability to identify and differentiate regions affected by necrosis from active tumor tissue, although challenging, highlights the importance of a detailed approach and specialized knowledge in the interpretation of magnetic resonance images.

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Preliminary baseline characteristics from evoke and evoke+: two phase 3 randomized placebo-controlled trials of oral semaglutide in patients with early Alzheimer's disease

Philip Scheltens[1], Alireza Atri[2], Howard H. Feldman[3], Oskar Hansson[4], Filip K. Knop[5], Mary Sano[6], Claus Dethlefsen[7], Peter Johannsen[7], Teresa León Colombo[7], Jeffrey Cummings[8], Wylliams Borelli[9,10]

[1]VU University Medical Center Amsterdam, Alzheimer Centre, Amsterdam, Netherlands.
[2]Banner Sun Health Research Institute, Sun City, AZ, USA.
[3]University of California San Diego, Department of Neurosciences, Alzheimer's Disease Cooperative Study, La Jolla, CA, USA.
[4]Lund University, Clinical Memory Research Unit, Department of Clinical Sciences Malmö, Lund, Sweden.
[5]University of Copenhagen, Gentofte Hospital, Center for Clinical Metabolic Research, Hellerup, Denmark.
[6]Cahn School of Medicine at Mount Sinai, New York, NY, USA.
[7]Novo Nordisk A/S, Søborg, Denmark.
[8]University College London, Institute of Neurology, Queen Square, London, UK.
[9]Universidade Federal do Rio Grande do Sul, Departamento de Ciências Morfológicas, Porto Alegre RS, Brazil.
[10]Hospital Moinhos de Vento, Centro da Memória, Porto Alegre RS, Brazil.

Correspondence: Luiz Eduardo Novis de Faria; Email: luizeduardonovis@hotmail.com

Background: Current treatment options for early Alzheimer's disease (AD) are limited to recently approved amyloid-lowering monoclonal antibodies, necessitating the exploration of additional therapeutic avenues due to the complexity of AD. Semaglutide, a glucagon-like peptide-1 receptor agonist (GLP-1RA) approved for type 2 diabetes and obesity, is now being investigated for early AD. **Methods:** The ongoing phase 3 evoke (NCT04777396) and evoke+ (NCT04777409) trials are multicenter, randomized, double-blind, placebo-controlled studies being conducted in 40 countries. Enrollment was completed on September 8, 2023. Participants, aged 55-85 years, were amyloid-positive, with mild cognitive impairment due to AD or mild AD dementia. The primary endpoint is the change in CDR-SB score from baseline to week 104. A cerebrospinal fluid (CSF) sub-study is being conducted in 15 countries to explore the effect of semaglutide on CSF biomarkers of AD and neuroinflammation. Baseline characteristics were compiled after enrollment completion and without knowledge of randomization assignments. evoke and evoke+ enrolled 1855 and 1953 participants, respectively. **Results:** Baseline demographics and clinical parameters were similar in the two trials. Additionally, 2.8% of participants in evoke+ exhibited concurrent significant small vessel pathology. The proportion of participants on an AD medication was 59.7% and 56.0% in evoke and evoke+, respectively. Type 2 diabetes was present in 11.8% of participants in evoke and 15.6% in evoke+. **Conclusion:** The evoke and evoke+ trials represent the first large-scale phase 3 investigations into the disease-modifying potential of GLP-1RAs in individuals with early AD. Both trials recruited a very similar population of individuals with early AD. The primary read-out of the trials is expected in the second half of 2025.

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Preliminary data on the validation of the comprehensive coordination scale for individuals with Parkinson's disease

Clara Rezende Rocha[1], Maria Eduarda Salum Aveiro[1], Caroline Rodrigues Osawa[1], Pedro Henrique Souza de Andrade[1], Luciane Aparecida Pascucci Sande de Souza[1], Gustavo José Luvizutto[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Clara Rezende Rocha; Email: d202010477@uftm.edu.br.

Background: The motor coordination refers to the recruitment and organization of muscles and joints to produce precise movements. The inefficiency of this aspects may reduce the quality of life and daily activities execution. The Parkinson's disease (PD) is a neurodegenerative condition wich symptoms as tremor, badykinesia and muscle stiffness, reduce motor coordination in PD patients. **Objective:** To validate the Comprehensive Coordination Scale (CCS) as a evaluation instrument of motor coordination in PD. **Methods:** It is a cross sectional study with PD patients. Was assess 20 individuals: Parkinson's Group (PG; n=13) and Control Group (CG; n=7), neurological healthy older adults. For the PD evaluation, beside CCS, was applied the Hoehn and Yard Scale disability states scale (H&Y) and the motor evaluation of the Unified Rating Scale of Parkinson's Disease adapted by Movement Disorder Society (MDS-UPDRS). Both groups were evaluated by CCS, a qualitative scale with specifical tests that measure the precision, performance and quality of the movements of each task (finger-to-nose; arm-trunk coordination; finger opposition test; interlimb coordination test - bilateral upper limb movements; lower extremity motor coordination – lower limber coordination; interlimb coordination – bilateral upper and lower limb movements). The scale score is divided in bilateral and unilateral for lower and upper limbs to assess the left and right hemibody, it totalizes 69 point. The evaluation was recorded and assessed by five evaluators. The data comparation of CCS between groups was assessed by the Mann-Whitney test and the correlation between CCS, H&Y and UPDRS was measured by the Spearman correlation test. **Results:** In PG the H&Y mean was of 1.8 which demonstrated a group with more unilateral commitment with or without axial deficit. In the MDS-UPDRS the mean was 29,61 which demonstrated a higher presence of individuals with PD dominant tremor in relation to akinetic ones. In regarding of CCS there was a statistical difference between both groups on the domains: upper limb ($p<0.001$), lower limb ($p=0.006$), bilateral ($p=0.015$) and total ($p<0.001$). There was negative and moderate correlation of the total and subdomains of CCS and MDS-UPDRS, the same happened between CCS and H&Y. **Conclusion:** This study demonstrated that there is a moderate correlation between the subitens of CCS, MDS-UPDRS and H&Y. It is important to highlight that this scales presents different domains which makes necessary the investigation of CCS and other motor coordination tests. Furthermore it is important the modification of some evaluation itens of CCS to make the scale more sensitive to PD symptoms.

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Preliminary study evaluating the structures of medial temporal lobe in epilepsy with hippocampal sclerosis using transcranial sonography

Bárbara Pina Aiello[1], Ingrid Pereira Marques[1], Fernando de Oliveira Morais Nacur[1], Sophia Keiko Inuzuka[1], Claudia Cecilia da Silva Rego[1], Valeria Coelho Santa Rita Pereira[1], Soniza Vieira Alves Leon[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Bárbara Pina Aiello; Email: drabarbarapina@gmail.com.

Background: Mesial temporal lobe epilepsy (MTLE) has a high prevalence, accounting for 40% of epilepsy cases. In 80% of MTLE, the crisis begins in the hippocampus, with Hippocampal Sclerosis (HE) being the etiology found in 50-70% of patients with MTLE refractory to clinical treatment, with surgical treatment as an option. **Objective:** Considering the prevalence and burden of this morbidity, we aim to evaluate the feasibility of transcranial sonography (TCS), a cost-effective and harmless neuroimaging method in detecting structural changes in ELTM-EH and to compare it to the already validated method, Magnetic Resonance Imaging (MRI), in patients with MTLE-EH. **Methods:** Preliminary prospective cross-sectional study. Patients were recruited from the epilepsy outpatient clinics of a university hospital with a diagnosis of MTLE-EH with MRI performed <1 year ago. The TCS was performed with HD11XE equipment (Phillips, Netherlands) and a 2-4 MHz sector probe positioned in the temporal bone window in axial and coronal planes. The same measurements were obtained by MRI and TCS bilaterally - choroidal fissure (CF), temporal pole of the lateral ventricle (TP) and height of the hippocampus (HIPh). With MRI, images of the hippocampal formations were obtained in T1 sequences in the sagittal plane with 1mm slices, in addition to FLAIR and T2 sequences angled for the hippocampus without the administration of gadolinium. Measurements were taken at the largest vertical diameter of the structure, regardless of its position in relation to the hippocampus. The mesial temporal lobe atrophy index (MTA=CF+TP) was calculated for both methods. **Results:** Four patients were analyzed, 3 men and 1 woman, aged 24 to 50 years and disease duration between 48 and 420 months. The comparison of the measurements taken by the 2 exams showed a good correlation in all patients, especially the

MTA, with an agreement of 74%. The worst measurements were those of TP, with an agreement of 43%. Only 1 patient had no bone window for analysis by TCS on the left side. The definition of the laterality of hippocampal atrophy between the 2 exams was 100%. **Conclusion:** To the best of our knowledge, this is the first study to evaluate patients with MTLE-EH diagnosed by MRI with TCS, a practical, harmless and relatively low-cost method. The structures of the mesial temporal lobe were easily visualized with TCS and, in this preliminary study, the measures obtained showed good agreement with MRI and offered the correct identification of the laterality of hippocampal sclerosis. The disadvantages of the method are its dependency of a permissible bone window, which usually occurs in 85-90% of individuals, and of an experienced operator. Studies with larger participant groups are highly desirable to prove TCS accuracy in clinical neurological practice as a neuroimaging modality in epilepsy patients.

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Pre-surgical hippocampus and amygdala volumes do not present relationship with seizure freedom after surgery in temporal lobe epilepsy patients

Marco Tulio Medina[1], Lucas Scárdua-Silva[1], Brunno Machado de Campos[1], Marina Koutsodontis Machado Alvim[1], Fabio Rogerio[1], Enrico Ghizoni[1], Helder Tedeschi[1], Fernando Cendes[1], Clarissa Lin Yasuda[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Lucas Scárdua Silva; Email: scardua@unicamp.br.

Background: Temporal Lobe Epilepsy (TLE) is the most common cause of pharmacoresistant epilepsy in adults, and many neuroimaging studies have presented microstructural alterations in the brains of these patients. However, it is still a research theme if pre-surgical neuroimaging can predict surgical outcomes. **Objective:** To compare bilateral amygdalae and hippocampi volumes (before surgery) of surgical-responsive and surgical-non-responsive TLE patients - as evidence of possible neuroimaging-base prediction. **Methods:** We acquired 3T T1-weighted images (Achieva Phillips) of 86 TLE patients before surgery and divided them into two groups: the surgical-responsive (56 subjects who achieved seizure freedom until the last visit after surgery) and the surgical-non-responsive (30 subjects without seizure freedom until the last visit after surgery). We used MATLAB 2019/SPM12/CAT12 to preprocess and perform a Voxel-Based Morphometry (VBM) calculation. After this, we utilized MATLAB2019 to perform the normality test (Kolmogorov-Smirnov, the samples were statistically normal, p>0.05) and the two sample t-tests. **Results:** The two-sample t-tests yielded negative results, in which the left hippocampus, right hippocampus, left amygdala, and right amygdala volumes did not show a statistical difference in surgical-responsive and surgical-non-responsive patients (p>0.05). Although there was no statistical difference, surgical-responsive patients exhibited bigger volume means and medians in all four structures. **Conclusion:** While our study did not find a statistical difference in the volumes of amygdalae and hippocampi between surgical-responsive and surgical-non-responsive patients, it is noteworthy that the surgical-responsive group exhibited higher means and medians for all four pre-surgical volumes (left hippocampus, right hippocampus, left amygdala, right amygdala). These findings, though not conclusive, could potentially guide future studies in this area.

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Prevalence of comorbidities in patients admitted to a stroke care unit in a reference hospital in the state of Pernambuco

Caio Roberto Alexandre Cavalcante dos Santos[1], Ana Dolores Firmino Santos do Nascimento[2], Déborah Emmily de Carvalho[1], Rafael Cabral de Carli[1], Janaina Gomes Pereira de Oliveira[1], Maria Eduarda Moreira Cardoso[2], Ricardo Martins Silva[2], Monique Evelyn Mendonça do Nascimento[2], Noelle Ventura Jordão[2]

[1]Universidade de Pernambuco, Recife PE, Brazil. [2]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Caio Roberto Alexandre Cavalcante dos Santos; Email: caiosantosupe@gmail.com.

Background: Stroke is a focal disorder of vascular origin. Approximately 87% of total stroke cases are ischemic, mainly caused by atherosclerosis of large arteries, occlusion of small vessels, and cardioembolism, often associated with atrial fibrillation. Etiological factors are known to be associated with various comorbidities that increase the risk of stroke and interfere with recovery after the acute event, with some of these being modifiable and others non-modifiable. Among hemorrhagic strokes, hypertension, excessive alcohol consumption, and smoking are known risk factors. **Objective:** To analyze the prevalence of comorbidities as risk factors in patients admitted to the stroke unit of a high-complexity hospital in Northeast Brazil. **Methods:** This is a retrospective, cross-sectional study analyzing medical records of stroke patients from May 2023 to May 2024 at a neurology reference hospital in the state of Pernambuco, including patients with ischemic stroke, hemorrhagic stroke and transient ischemic attack. Statistical analysis was performed using Excel software, with appropriate statistical tests applied to the selected variables. **Results:** A total of 2391 patients were included in the study. Of the total sample, 1228 patients (51.35%) were previously diagnosed with Systemic Arterial Hypertension. Regarding other comorbidities, Diabetes Mellitus was observed in 614 (25.67%) patients, previous stroke in 272 (11.37%), and previous Acute Myocardial Infarction in 77 (3.22%). Among the modifiable risk factors, 143 (5.98%) patients had been active smokers for more than ten years, and 112 (4.68%) had a history of alcohol abuse. **Conclusion:** There is a high prevalence of comorbidities in the studied population. Considering stroke is a rapidly evolving pathology with a high mortality rate, it is essential to track the main risk factors associated with its progression. Therefore, preventing and treating such comorbidities is crucial to reducing the incidence of this neurological vascular condition and improving outcomes.

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Prevalence of gastrointestinal symptoms in patients with autism spectrum disorder: a systematic review

Milena Fernandes de Oliveira[1], Sofia Sampaio Paixão[1], Thais Barbosa de Oliveira[1], Cibele Dantas Ferreira Marques[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Milena Fernandes de Oliveira; Email: milenafer.oliv@gmail.com.

Background: Children with ASD have a higher frequency of concomitant conditions, such as gastrointestinal symptoms, epilepsy, sleep disorders, autoimmune and psychiatric diseases. The presence of gastrointestinal symptoms in children with ASD is associated with worsening repetitive behaviors, worsening sleep habits, difficulty in social reciprocity and increased anxiety. **Objective:** To identify whether there is a higher prevalence of gastrointestinal symptoms in the population of children with ASD compared to the population of neurotypical children. **Methods:** A systematic literature review with meta-analysis was carried out. Cross-sectional case-control observational articles from 2008 onwards, in English, Portuguese and Spanish, were considered eligible. The case group consisted of individuals with ASD, diagnosed by the DSM-V, DSM IV, ICD-10 criteria, or by the ADOS and ADI-R tests, and the control group consisted of children without any developmental disorder, both groups younger than 18 years. The search was carried out in four databases: PubMed, Embase, Lilacs and Scielo. The studies needed to provide information on at least one of the following gastrointestinal symptoms: constipation, diarrhea, vomiting, abdominal pain and excessive flatulence. **Results:** Of the 803 studies, 194 duplicates were deleted. Thus, 609 articles were selected. The selection was carried out by two researchers independently and any disagreements were decided between the two. Of the 609 articles, 9 articles were selected for this study. Afterwards, the risk of bias was assessed using a specific questionnaire for prevalence articles and all studies had acceptable risk to be included in this review. There was a higher risk of constipation (RR 2.62; 95%CI 2.17-3.17; I^2=38%), diarrhea (RR 4.22; 95%CI 2.58-6.91; I^2=59%), abdominal pain (RR 1.90; 95%CI 1.29-2.82 ; I^2=44%), excessive flatulence (RR 3.51; 95%CI 1.84-6.69; I^2=71%) and vomiting (RR 3.09; 95%CI 1.61-5.92; I^2=49%), that is, a higher prevalence of symptoms gastrointestinal disorders in individuals with ASD compared to children without developmental disorders. **Conclusion:** Gastrointestinal symptoms are more present in individuals with ASD and this may be due to greater difficulty in eating due to the restrictive diet pattern or genetic and intestinal microbiota findings.

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Prevalence of hypertension and its impact on NIHSS scores in ischemic stroke patients at a tertiary hospital

Maria Eduarda Vieira de Oliveira[1], Maria Thayana Barros Bandeira[2], Noelle Ventura Jordão[3], Maria Eduarda Moreira Cardoso[3], Ana Dolores Firmino Santos do Nascimento[3], Maria Luiza Vasconcelos Montenegro[1], Letícia Gomes Fonseca[4], Lívia Barbosa Cavalcanti[1], Mariana Lucena Loureiro[1]

[1]Universidade de Pernambuco, Recife PE, Brazil. [2]Faculdade de Medicina de Olinda, Olinda PE, Brazil. [3]Hospital da Restauração, Recife PE, Brazil. [4]Faculdade Pernambucana de Saúde, Recife PE, Brazil.

Correspondence: Maria Eduarda Vieira de Oliveira; Email: eduardaoliveira0194@gmail.com.

Background: Ischemic stroke is a leading cause of morbidity and mortality worldwide, with hypertension being a significant risk factor. The National Institutes of Health Stroke Scale (NIHSS) is a critical tool for assessing the severity of stroke and predicting outcomes. Understanding the relationship between hypertension and stroke is crucial for developing targeted interventions to improve patient outcomes. **Objective:** To determine the prevalence of hypertension in patients admitted with ischemic stroke and its impact on NIHSS scores at admission and discharge in a tertiary hospital in Northeastern Brazil, compared with non-hypertensive patients. **Methods:** A retrospective cohort study was conducted in a stroke unit at a tertiary hospital from May 2023 to May 2024. Data were meticulously collected from medical records. Statistical analysis included descriptive statistics to outline patient demographics and health status, and the Student's t-test to assess the association between hypertension and NIHSS scores at discharge. **Results:** The sample consisted of 1100 ischemic stroke patients, 923 (83.91%) of whom had hypertension. The mean NIHSS score at discharge was analyzed for both groups, with and without hypertension. Among hypertensive patients, the mean NIHSS score at discharge was significantly higher compared to non-hypertensive patients, indicating worse neurological outcomes. The Student's t-test showed a statistically significant difference between the two groups at discharge (t=2.178, p=0.030). At admission, however,

the NIHSS scores did not show a statistically significant difference between hypertensive and non-hypertensive patients (t=0.175, p=0.861). The average NIHSS score at discharge for hypertensive patients was 10.57 (SD 8.66), while for non-hypertensive patients it was 7.72 (SD 9.41). Despite the lack of significant difference at admission, the higher NIHSS scores at discharge among hypertensive patients suggest that hypertension may contribute to poorer recovery or greater complications during hospitalization. **Conclusion:** Hypertension is highly prevalent among ischemic stroke patients and is associated with worse neurological outcomes, as indicated by higher NIHSS scores at discharge. The statistically significant difference observed at discharge underscores the importance of effective hypertension management in improving stroke recovery. Further studies with larger samples and more robust methodologies are needed to confirm these findings and develop effective management strategies for hypertension in stroke patients.

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Prevalence of incidental findings in the brains of non-sedated nonclinical population of full-term neonates undergoing MRI scans

Larisssa Melo Marques de Sousa[1], Célia Maria de Araújo[1], Gabriel Ronatty Tavares[1], Lucas Pinto Ribeiro[1], Maret Rand[1], Aline Camargo Ramos[1], Ana Carolina Milani[1], Ivaldo Silva[1], Nitamar Abdala[1], Andrea Parolin Jackowski[1]

[1]Pontifícia Universidade Católica de São Paulo, Sorocaba SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Larissa Melo Marques de Sousa; Email: marq.lari@gmail.com.

Background: Magnetic resonance imaging (MRI) is considered safe for neonates, but incidental findings (IF) in healthy populations pose a challenge due to limited data. These findings can guide early intervention or monitoring. **Objective:** To investigate the prevalence IFs in a nonclinical population of full-term neonates participating in the Healthy MINDs study. **Methods:** MRI were performed at the Diagnostic Imaging Department of the Federal University of São Paulo in a Siemens 3T (IRB:1200/2017). Before the MRI scan, the neonates were examined by a pediatrician and after having their ears protected and falling asleep naturally, they conducted a brain MRI. Structural analysis included cerebral myelination, anatomical variants or malformations, brain parenchyma lesions, ventricular system changes, and intra- or extracranial hematomas or hemorrhages, always evaluated by the same neuroradiologist. **Results:** Out of 301 neonates (mean age = 32.13 ± 9.018 days; 153 males), 32 (10.63%) had IFs and 77 (25.58%) had anatomical variants like cavum of septum pellucidum (CSP). The IFs included 4 subgaleal hematomas, 1 subdural hematoma, 1 cortical laminar necrosis, 7 arachnoid cysts, 4 ventricular anomalies, 2 vascular anomalies, 3 volumetric reductions of the left cerebral hemisphere, 1 lipoma, 3 cerebellar and intracranial hemorrhagic components, 1 subcortical signal alteration/hemoglobin degradation product, 1 brain parenchyma signal alteration, 2 Virchow-Robin perivascular space increases, 1 benign enlargement of the subarachnoid space, and 1 thickening of the mucous lining of the ethmoidal cells bilaterally. All altered exams were communicated to the mothers, and infants were referred for follow-up as needed. **Conclusion:** The prevalence of IFs was 10.63% and CSP was 25.58%. Arachnoid cysts were present in 21.7% of IFs, higher than previous studies. A 9.3% prevalence of left cerebral hemisphere volumetric reduction was noted, contrasting with other studies. More research is needed on IFs in nonclinical full-term neonates to inform early intervention and follow-up practices.

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Prevalence of leprosy in the state of Maranhão from 2013 to 2023

Maria Regina Saddi Guerra[1], Yasmim Melo Costa de Azevedo[2], Marcelly Silva dos Santos Brito[3], João Pedro Rosa Barroncas[4], Raul Pereira Bassotto Franco[5], Cayara Mattos Costa[6]

[1]Universidade Católica de Brasília, Brasília DF, Brazil.
[2]Universidade Nove de Julho, São Paulo SP, Brazil.
[3]Universidade Federal de Pernambuco, Recife PE, Brazil
[4]Universidade Federal do Amazonas, Manaus AM
[5]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.
[6]Universidade Federal do Maranhão, São Luís MA, Brazil

Correspondence: Maria Regina Saddi Guerra; Email: mreginasaddi@gmail.com.

Background: Leprosy is a notifiable disease, with high infectivity and low pathogenicity. Initially, it affects the peripheral nervous system (PNS) and, later, the skin (non-contagious, paucibacillary form). This disease is associated with factors such as social inequality and unfavorable socioeconomic conditions and can cause physical disability, being neglected. Although it has been eliminated in many countries, it persists in developing nations, such as Brazil, especially in the state of Maranhão, which has the highest number of new cases. **Objective:** Analyze the epidemiological profile of Leprosy in the state of Maranhão from 2013 to 2023. **Methods:** This was an ecological study using data of leprosy cases in the Brazilian state of Maranhão, diagnosed between 2013-2023 from the Notifiable Health Conditions Information System (SINAN). The variables investigated were: total frequency of notifications per year, sex, age group, color/race, level of education, operational class, smear microscopy and number of lesions. **Results:** During the period from 2013 to 2023, 40,327 cases of leprosy were reported in the state of Maranhão. The disease predominantly affected men (51.9%), of mixed race (67.4%). The age group with the highest notification was between 30 and 39 years old (17.7%), followed by the age group of 40 to 49 (16.4%) notifications, and in children under 15 years old there was the lowest notification (8.3%). Notifications were predominant in people with low education, with 43% of reported cases not having completed primary education and 14.6% being illiterate. The multibacillary classification was predominant in 80.45%, the paucibacillary in 19.5%. The predominant bacilloscopy was negative (27.3%), however 28.5% had an inconclusive result and in 25.9% the test was not performed. According to the number of injuries, 35.8% had more than 5 injuries, 28.7% between 2 and 5 and 22.3% had single injuries. During the period analyzed, it was noticed that notifications of Maranhão disease are in constant decline, with the highest number of notifications in 2013 (11.9%) and with the lowest number in 2023 (2.27%), this reduction could be attributed to the actions of the Family Health Strategy and its teams in the fight against leprosy. **Conclusion:** The epidemiological profile of leprosy in Maranhão is characterized by a higher prevalence among men of mixed race, with multibacillary classification, aged 30 to 39 years, and who have an education level of up to elementary school. Even with the reduction in cases in the period, public policies are still essential to eradicate them.

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Prevalence of meningitis by case evolution in Brazil between the years 2013 and 2023

Luan Victor Silva Ribeiro[1], José Antônio Solano Silva[1], Laisa de Paula da Costa Lobato[1], Hálisson Ferreira Freitas Filho[2], Leandro Gabriel Costa Macedo[1], Bárbara Coutinho Morgado[3], Ricardo Ferreira Silva[1], Bárbara Alessandra Solano Silva[3], Dirceu Jose de Freitas Cardoso[1], Leonardo Kadu Tavares Veloso[1]

[1]Afya Faculdade de Ciências Médicas, Abaetetuba PA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.
[3]Centro Universitário do Pará, Belém PA, Brazil.

Correspondence: José Antônio Solano Silva; Email: jsjosesolano108@gmail.com.

Background: Meningitis is an acute inflammation of the membranes that cover the brain and spinal cord, with significant potential to cause neurological and systemic damage, sometimes irreversible, and is important for notification systems, directly impacting public health. Thus, understanding these data is essential for developing effective public health policies that can reduce incidence and improve outcomes for affected patients. **Objective:** To investigate the epidemiological data related to the evolution of meningitis cases from 2013 to 2023 in Brazil. **Methods:** This is a cross-sectional, descriptive epidemiological study with a temporal trend design and quantitative focus, using secondary data collected from the Notification of Injury Information System (SINAN), provided by the Department of Informatics of the Ministry of Health (DATASUS). Confirmed cases of meningitis were reviewed according to their etiology: Meningococcemia (MCC), Meningococcal Meningitis (MM), Meningococcal Meningitis + Meningococcemia (MM+MCC), Tuberculous Meningitis (MTBC), Bacterial Meningitis (MB), Unspecified Meningitis (UM), Viral Meningitis (VM), Meningitis of other etiology (MOE), Haemophilus Meningitis (HM), Pneumococcal Meningitis (PM), and by their clinical evolution, whether discharge, followed by deaths due to the disease or deaths due to another associated cause, from January 2013 to December 2023, in each region of the country. **Results:** Between 2013 and 2023, there were 149,475 confirmed cases of meningitis, including 3,053 of MCC, 4,425 of MM, 3,022 of MM+MCC, 3,609 of MTBC, 24,496 of MB, 23,129 of UM, 70,003 of VM, 7,035 of MOE, 1,185 of HM, and 9,518 of PM. Thus, a rapid progression of the disease is noted, respectively, in VM (96.24%), MM (84.72%), UM (81.39%), HM (80.50%), MM+MCC (78.25%), MB (76.78%), MTBC (65.41%), PM (64.79%), MOE (60.98%), and MCC (56.66%). On the other hand, mortality from this disease is highest in MCC (39.79%), followed by PM (32.01%), MOE (21.76%), MM+MCC (21.24%), MTBC (20.42%), HM (17.21%), MB (14.68%), MM (14.30%), UM (11.92%), and VM (1.61%). Finally, deaths due to other causes in patients with meningitis follow the order of MOE (17.25%), MTBC (14.15%), MB (8.53%), UM (6.63%), MCC (3.53%), PM (3.19%), HM (2.27%), VM (2.13%), MM (0.97%), and MM+MCC (0.49%). **Conclusion:** The findings presented highlight a high number of VM cases, which accounted for the majority of confirmed cases between 2013 and 2023, followed by MB and UM. VM, followed by MM and UM, were the most prevalent concerning rapid disease progression, with high mortality in cases of MCC, PM, and MOE. Additionally, deaths due to other causes associated with the pathology are also alarming, such as in MOE, MT, and MB, which were the most predominant. In this context, the data emphasize the need for preventive measures and strategies tailored to the specificities of each meningitis etiology, aiming to reduce mortality and disease burden in the country.

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Prevalence of zika virus in pregnant women as a risk factor for microcephaly in babies: an epidemiological analysis of the Northern region of Brazil

Luan Victor Silva Ribeiro[1], Leandro Gabriel Costa Macedo[1], José Antônio Solano Silva[1], Hálisson Ferreira Freitas Filho[2], Laisa de Paula da Costa Lobato[1], Bárbara Alessandra Solano Silva[3],

Ricardo Ferreira Silva[1], Bárbara Coutinho Morgado[3], Dirceu Jose de Freitas Cardoso[1], Leonardo Kadu Tavares Veloso[1]

[1]Afya Faculdade de Ciências Médicas, Abaetetuba PA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.
[3]Centro Universitário do Pará, Belém PA, Brazil.

Correspondence: Leandro Gabriel Costa Macedo; Email: leandrogabrielzinho11@gmail.com.

Background: For a long time, the possibility of viral infections interfering with pregnancy and causing congenital anomalies in newborns (NB) has been studied. Currently, this interference is well established in the literature, with Zika virus (ZIKV) being a major focus of study in this field. Numerous studies indicate that the presence of ZIKV RNA in amniotic fluid can lead to malformations in NB, such as neonatal microcephaly, a condition where the baby is born with a smaller than expected head, leading to problems in neurological development. **Objective:** To present evidence related to comparative findings between ZIKV infection during pregnancy and the occurrence of microcephaly in NB. **Methods:** This is a descriptive, analytical, and retrospective study with a quantitative approach, using secondary data from the Notification of Injury Information System (SINAN) and the Live Births Information System (SINASC). We evaluated cases of ZIKV among pregnant women, trimester of pregnancy, year of notification, and analyzed the frequency per Live Birth Certificate with microcephaly, by year of birth, from January 2016 to December 2022, in the Northern region of Brazil. **Results:** In 2016, an alarming number of ZIKV cases among pregnant women were observed (3,356), with 920, 1,298, and 1,138 cases in the first, second, and third trimesters of pregnancy, respectively. In 2017, there were 1,304 cases, with 371, 509, and 424 in the first, second, and third trimesters. In 2018, there were 598 cases of ZIKV among pregnant women, with 187, 224, and 187 occurring in the first, second, and third trimesters. In 2019, there were 344 cases, with 97, 129, and 118 in the first, second, and third trimesters. In 2020, 175 cases, divided as follows: 56, 67, and 52 in the first, second, and third trimesters. In 2021, there were 149 cases, with 27, 46, and 45 in the first, second, and third trimesters. As for the numbers of NB with microcephaly from 2016 to 2021, the following values were obtained respectively: 181, 62, 42, 32, 21, and 22 cases. Finally, in 2022, unlike previous years, there was no decrease trend, with 242 cases of ZIKV among pregnant women, including 54, 100, and 76 in the first, second, and third trimesters, and 27 cases of microcephaly in babies. **Conclusion:** In 2016, a ZIKV epidemic spread across Brazil, resulting in a higher number of pregnant women testing positive for ZIKV and more babies born with microcephaly that year. In 2017, there was a significant decline in these cases, possibly due to a combination of acquired immunity in the population, vector control efforts, and reduced virus exposure; a trend that continued in the following years. However, in 2022, there was an increase in ZIKV rates among pregnant women and microcephaly in NB. Thus, measures should be implemented in the country, considering transplacental transmission and the complexity of the pathophysiological mechanisms of Zika Congenital syndrome.

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Primary amebic meningoencephalitis caused by free-living amoeba: report of 2 fatal cases in Natal/RN, Brazil

Yasmin Luíse Lucena e Silva[1], Lênia Simone Fernandes de Lucena[2], Nelly Luchiari de Lucena[2], Ana Maria de Oliveira Ramos[2], Miguel Angel Sicolo[3], Vanessa Pache da Rosa[4], Micássio Fernandes de Andrade[1], Samila Marissa Pinheiro Gomes[1]

[1]Universidade Federal Rural do Semi-Árido, Mossoró RN, Brazil.
[2]Serviço de Verificação de Óbito, Natal RN, Brazil.
[3]Casa de Saúde São Lucas, Natal RN, Brazil.
[4]Hospital Universitário Onofre Lopes, Natal RN, Brazil.

Correspondence: Yasmin Luíse Lucena e Silva; Email: yasmin.silva43227@alunos.ufersa.edu.br.

Case presentation: Primary amebic meningoencephalitis (PAM) is a rare, highly lethal infection caused by the free-living amoeba (FLA) Naegleria fowleri. This pathogen penetrates the central nervous system through the cribriform plate following exposure to contaminated water, causing fulminant hemorrhagic brain necrosis. Diagnosing PAM is challenging and there is no widely standardized treatment. This report describes two PAM cases in Northeast Brazil. Case 1: A 34-year-old male with systemic arterial hypertension and hypothyroidism, working in oil refinery maintenance, was admitted with intermittent headache and fever 40 days after cleaning a water tank. He received a clinical diagnosis of meningoencephalitis. Cerebrospinal fluid (CSF) and serological analyses didn't detect the pathogen. A month later, he developed intense occipito-vertebral pain and vomiting, leading to intensive care admission. Treated with antifungals and an extensive combination of antibiotics, he showed slight improvement but then developed neck stiffness, underwent external ventricular drainage (EVD) and suffered a subarachnoid hemorrhage; brain death was declared. Autopsy revealed brain edema, ventriculitis and hemorrhagic necrosis in the right temporal lobe and olfactory tract, with microscopy confirming the diagnosis of PAM associated with fungal infection. Case 2: A 7-year-old male student with no comorbidities experienced vomiting, headache and diplopia within two days of visiting a water park. Fundoscopy revealed papilledema and an expansive brain lesion was detected by neuroimaging. Biopsy ruled out neoplasm. He then had headaches, vomiting and seizures. Increasing intracranial pressure led to a craniectomy and EVD. The patient remained under mechanical ventilation, hemorrhagic CSF was drained and new imaging revealed extensive intraparenchymal hemorrhage. Laboratorial tests did not detect the parasite. Clinical suspicion of PAM led to treatment for FLA, as well as for bacteria and other protozoa, but the patient died 16 days after symptom onset. Autopsy revealed extensive hemorrhagic necrosis of the right temporal lobe and ventral surface of the brain. Microscopy confirmed PAM. **Discussion:** This report reinforces the importance of clinical history in suspecting PAM, as prior contact with potentially contaminated water and fulminant disease progression are characteristic. Lesions in the right temporal lobe were common to both cases and fungal coinfection is noteworthy. Despite initiating treatment for PAM in one of the cases, the outcome was still fatal, consistent with the documented 98% lethality. Both diagnoses were only confirmed post-mortem, as in most cases of PAM, highlighting the need for rapid and widely available diagnostic methods. Underreporting remains significant in Brazil and worldwide. **Final comments:** Although rare, PAM is highly lethal, justifying efforts for its control. Broad access to rapid diagnostic methods and new therapeutic plans are urgent.

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Primary central nervous system lymphoma presenting as demyelinating disease

Patrick Emanuell Mesquita Sousa-Santos[1], Pedro Machry Pozzobon[1], Itamar Meireles Andrade Santos[1], Helio Aquaroni Farao Gomes[1], Paula de Almeida Souza Santos da Costa[1], Daniela Cristina dos Santos Souza[1], Laura Cardia Gomes Lopes[1], Fernando Coronetti Gomes da Rocha[1]

[1]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Patrick Emanuell Mesquita Sousa Santos; Email: patrickemanuell@gmail.com.

Case presentation: Male, 32-year-old, previously healthy and HIV-negative was diagnosed with COVID-19 in January/2022 and two weeks later present progressive vertigo, gait instability, binocular diplopia and right hemifacial palsy. Only in April he was sent for investigation in our hospital. MRI showed a lesion with FLAIR hypersignal in the pontomesencephalic transition with gadolinium enhancement. Cerebrospinal fluid (CSF) examination showed protein levels of 83mg/dL, 2 leukocytes, glucose of 79mg/dL (serum glucose 115mg/dL), and lactate level of 1.2. Oligoclonal band testing in CSF was negative. CSF cytology did not reveal neoplastic cells. Levels of anti-AQP4 and anti-MOG antibodies were also negative. In this moment, considering the infectious antecedent and MRI characteristics, he has been diagnosed with ADEM and submitted to pulse therapy with methylprednisolone, showing improve of symptoms. In outpatient follow-up he presents with worsening of symptoms. In November he was hospitalized again, MRI show new lesions on medulla and cerebellum. Submitted to pulse therapy followed by plasma exchange, with no improve. In December he had a fall with head trauma and CT show hyperdense lesion on pontomesencephalic transition with perilesional edema. New MRI presents with infiltrative lesion without gadolinium enhancement. CSF analysis show 48 cells (98% lymphocytes), no evidence of neoplastic cells. In this moment the diagnosis was reviewed to Primary CNS Lymphoma (PCNSL), confirmed on biopsy showing atypical lymphoid infiltrate with B-Cells predominance. Initiated chemotherapy, but unfortunately patient die in consequence of neutropenia. **Discussion:** PCNSL is an uncommon neoplasm and rarely affects brainstem. When it occurs the prognosis is poor, nearly half of patients died within 1 year after diagnosis. Glucocorticoids could promote tumor shrinkage. Neuroimaging characteristics could be confounding with infections, demyelinating lesions, and gliomas. **Final comments:** Our case illustrates a patient initially diagnosed with ADEM and submitted to corticotherapy with good response, however he worse in months later and the diagnosed reviewed to PCNSL. This shows the importance of considering atypical PCNSL in differential diagnosis of demyelinating diseases.

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Primary presentation of demyelinating polyneuropathy in POEMS syndrome: a case report

Tainá Reis Martins[1], Wívian Maria Pires Figueredo[1], Ana Luisa Colares Ribeiro[1], Érica Guimarães Castro[2]

[1]Centro Universitário FIPMoc, Montes Claros MG, Brazil.
[2]Hospital Santa Casa de Montes Claros, Montes Claros MG, Brazil.

Correspondence: Tainá Reis Martins; Email: taina2002@hotmail.com.

Case presentation: Patient, 42 years, male, referred to neurology due to chronic low back pain for three years, evolving with hypoesthesia and progressive weakness in the lower limbs (LL) after one year. At physical examination, she presented tetraparesis, with chilling, dysesthesia and hypoesthesia of the LL. He performed electromyography with evidence of sensory-motor polyradiculopathy, predominant myelin, chronic. He was referred for hospitalization due to suspicion of chronic demyelinating inflammatory polyneuropathy (CDIP) and performed pulsotherapy for five days, without improvement of the picture. After discharge, he returns with disabling neuropathic pain and dysesthesia, using prednisone and gabapentin, presenting cutaneous rash, flaccid tetraparesis, intense dysesthesia and Raynaud in feet. Requested further investigation for secondary diagnoses of CDIP, as well as prescribed duloxetine and tramadol. It returns with complete improvement of the pains, but the weakness of the LL remains. It presents electrophoresis proteins with presence of monoclonal band in the region of gamma globulins and computed tomography (CT) of

spine and lumbosacral with diffuse sclerotic bone lesions in vertebrae and pelvis. Requested new propaedeutics, in which presented immunofixation in blood and urine with presence of monoclonal protein IGG/lambda. Chest and abdomen CT showed a numerical increase in lymph nodes in several sites, as well as mild bilateral gynecomastia and hepatosplenomegaly. Referred to hematology with suspected POEMS syndrome (PS), after hospitalization myelogram was performed with inconclusive result, and lymph node biopsy that demonstrated chronic lymphadenopathy with Plasmocyte proliferation, suggestive of Castleman's disease (CD). The presence of another criterion defined the PS diagnosis and made it possible to perform treatment with ritoxumab and the patient evolved with exponential improvement of symptoms and mobility. **Discussion:** PS is a paraneoplastic disease and its acronym represents its main manifestations: polyneuropathy; visceromegaly; endocrinopathy; monoclonal gamopathy and dermatological changes. The diagnosis tends to be late because it is a rare syndrome, which is commonly confused with CDIP, due to its symptoms and electrophysiological findings. The diagnosis of PS is made with three of the major criteria, being peripheral neuropathy and monoclonal gamopathy mandatory, and at least one of the minor criteria. The patient presented the mandatory criteria, in addition to the larger criteria of sclerotic bone lesions and CD and smaller ones: visceromegaly, cutaneous rash and gynecomastia. **Final comments:** The PS represents an important differential diagnosis in the face of polyneuropathy, although it represents a great challenge. Therefore, there is an urgent need for early recognition in order to ensure adequate treatment and support measures, consequently improving prognosis and quality of life.

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Probable bickerstaff encephalitis due to dengue fever: a case report

Marcelle Janaina Baldez do Amaral[1], Camila Albuquerque Pavão[1], Beatriz Passinho Heinle[1], Myllena Batista Ribeiro[1], Thuane dos Santos Leske[1], Pedro Gil Reis de Sá[2], Luiza Joaquina Botton Reginatto[1], Fabio Pascotto de Oliveira[1], Thiego Teixeira Cavalheiro[1], Juliana Oliveira Freitas Silveira[3]

[1]Universidade Franciscana, Santa Maria RS, Brazil.
[2]Universidade Federal de Santa Maria, Santa Maria RS, Brazil.
[3]Hospital Universitário de Santa Maria, Santa Maria RS, Brazil.

Correspondence: Juliana Oliveira Freitas Silveira; Email: julianaofs@yahoo.com.br.

Case presentation: Woman, 64 years old, hypertensive and diabetic, is admitted to a tertiary hospital due to reduced level of consciousness and ophthalmoplegia. Two days before admission, she started with asthenia, nausea, vomiting and fever. She then developed ataxia, mental confusion and obnubilation. On exam, she presented spontaneous eye opening, with incomprehensible sounds and mobilized the left upper limb to painful stimuli, with bilateral sixth nerve paresis, right hemiparesis (grade IV), bilateral Babinski sign. Ataxia could not be assessed due to disturbance of consciousness. She also presented dysautonomic signs with fluctuations in blood pressure and tachycardia. Laboratory tests showed thrombocytopenia. Serologies for HIV, syphilis and hepatitis B and C were non-reactive. ANA, ANCA and complement levels C3 and C4 were normal. CSF examination showed proteinorrhaquia with proteino-cytological dissociation and there was no growth of microorganisms. Research for Cryptococcus neoformans, herpes types 1, 2 and 6, also negative. Brain MRI showed focal areas of FLAIR hypersignal in the white matter of both hemispheres, in addition to the corpus callosum, mesial region of the thalami, pons and midbrain, suggesting encephalitis. Anti-GQ1b was also negative, but it was only available after 30 days of the onset of symptoms. After the patient's family members started showing symptoms of acute febrile syndrome with positive NS1 antigen, the patient's dengue serology was requested, with positive IgG and IgM, suggesting recent dengue infection. **Discussion:** Bickerstaff brainstem encephalitis (BEE) is a rare neurological condition, a variant of Guillain-Barré syndrome (GBS), as well as Miller-Fisher syndrome (MFS). It is associated with a combination of bilateral symmetric ophthalmoplegia, ataxia, muscle weakness and sensory changes, in addition to brain stem involvement, with pyramidal signs. CSF examination may demonstrate proteino-cytological dissociation. Anti-GQ1b antibodies may be presente but is not necessary for the diagnosis of MFS and BEE. In Brazil, studies point to dengue as the main cause of encephalitis and viral meningitis. Patients with dengue encephalitis may experience decreased consciousness, headache, seizures and behavioral symptoms. GBS has been described in reported cases of patients who manifested a variety of neurological symptoms after being infected with the dengue virus. **Final comments:** Dengue should be considered in the differential diagnosis of acute febrile illnesses with neurological symptoms, especially in endemic regions and in patients with a history of recent travel to such areas. Many neurological manifestations of dengue have been recorded, given the virus's tropism for the nervous system. Dengue encephalitis is a common complication, however, BEE represents a rare neurological syndrome, being an atypical neurological manifestation of dengue, making this report important in the academic and scientific sphere.

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Problematic of neurocysticercosis in Angola: a systematic review

Adalberto Cassucussuco[1], Job Monteiro C. Jama António[2]

[1]Universidade José Eduardo dos Santos, HMZ, Luanda, Angola.
[2]Universidade Agostinho Neto, Luanda, Angola.

Correspondence: Adalberto Moisés João Cassucussuco; Email: adalbertomoises017@gmail.com.

Background: Neurocysticercosis is one of the leading preventable causes of epilepsy worldwide. It is estimated to contribute to up to 30% of epilepsy cases in areas where the disease is endemic. The total number of people with symptomatic or asymptomatic neurocysticercosis is estimated to be between 2.56 and 8.30 million, according to available data on the prevalence of epilepsy. However, these numbers may be underestimated due to the limited access of the most at-risk groups to diagnostic tests. **Objective:** To describe neurocysticercosis in Angola, through a systematic review of the literature, to highlight the relevant clinical, epidemiological, and social aspects. **Methods:** Systematic review that searched works and literature articles with available text, published between 2008 and 2023. Through the descriptors: "Neurocysticercosi", "Diseases of the Nervous System" and "Angola". On the following platforms: Angolan Journal of Health Sciences, Science Direct, SciELO, Google Scholar, UFV Repository, and FM-UAN Repository. In addition, studies that did not address the research proposal, as well as repeated or duplicate studies, were excluded. **Results:** A total of 3 studies were reviewed, and the frequency of neurocysticercosis was 3.3%, and the prevalence was considered high, which is 55.0%. Social impact was estimated at 208 years lived with disability. 51.6% were female, and the annual economic impact was estimated at 246.94 USD=106,133.38 Kwanzas, per affected individual. Most of the patients came from urban areas. There was a significant variation in age from 25 to 75 years. The most frequent symptoms were seizures and headache. The diagnosis was made by computerized tomography and magnetic resonance imaging, with calcifications being the most relevant imaging finding. **Conclusion:** Neurocysticercosis is a common disease in Angola, with high prevalence, and high economic and social impact. It mostly affects individuals from urban areas, especially females, with seizures and headaches being the most frequent clinical manifestations.

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Professor, how is your mental health?

Ana Beatriz Bataglia Sperandio[1], Eliana Meire Melhado[1], Liara Ruiz Lima[1], Laura Beatriz Maciel Comisso[1], Julia Lucato Fontes[1], André Soares Rodrigues[1], Jadde Inhã Lisboa de Souza[1], Bruno Seabra Queiroz[1]

[1]Centro Universitário Padre Albino, Faculdade de Medicina de Catanduva, Catanduva SP, Brazil.

Correspondence: Ana Beatriz Bataglia Sperandio; Email: anabsperandio@gmail.com.

Background: Quality of life at work consists of the dynamic management of physical, technological and socio-psychological factors that affect culture and renew the organizational climate, reflecting on the well-being of workers. The promotion and protection of mental health at work is an area of growing interest and can be supported through legislation and regulation, organizational strategies, manager training and interventions for workers. With this in mind, the mental health of university professors is an area of growing concern, especially in the academic context of Medicine, since teachers in this area have to reconcile the demand for academic excellence with factors such as financial pressure and working conditions. Thus, this work sought to determine the mental health status and quality of life of university medical professors, along with symptoms related to mental health, using a cross-sectional observational approach. **Objective:** To determine the mental health status and quality of life of university medical professors, in addition to the symptoms presented related to mental health. **Methods:** The present work is a cross-sectional observational study carried out in a single interaction using a form on health and quality of life with a focus on mental health, carried out on Google Forms with 46 responses. **Results:** The results presented demonstrate the analysis of association between different categorical variables using the chi-square test of independence. When comparing mental health with physical health, there is a significant association between the two, with another words, there is an unequal distribution between the categories of mental and physical health, indicating that one influences the other's mental and physical health. A significant association was found between physical health and anxiety classification (χ^2=17.64, p=0.040). The results suggest that anxiety can negatively influence physical health. The analysis revealed a significant association between anxiety classification and depression classification (χ^2=60.78, p<0.001). This indicates that different levels of anxiety are associated with different levels of depression. Statistical analysis revealed that different levels of anxiety and depression are correlated with different mental health states, indicating the complexity of these interactions. **Conclusion:** These results provide solid evidence that mental health is significantly associated with physical health, frequency of perceived lack of money, anxiety rating, and depression rating. This can be crucial for guiding mental health policies and interventions, showing how these factors are interrelated and highlighting the importance of integrated approaches to promoting overall well-being.

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Profile and outcome post-rehabilitation of patients with vertigo in a clinic in Southern Santa Catarina

Tatiana Pizzolotto Bruch[1], Camila Pezente Dal Toe[1], Letícia Salvaro Fernandes[1], Emanuela Hannoff Pilon[1], Vitor Benincá Fernandes[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Tatiana Pizzolotto Bruch; Email: tatibruch@yahoo.com.br.

Vertigo is the sensation of spinning or rotation of the environment. The demand for medical attention due to these complaints has been increasing, making it important to understand the profile of patients presenting with vertigo, its primary causes, comorbidities, associated factors, and their response to rehabilitation. A cross-sectional secondary data analysis examined the epidemiological profile of patients treated for vertigo at a specialized clinic in Southern Santa Catarina, along with the outcomes of physiotherapeutic rehabilitation. A questionnaire developed by the authors was used to evaluate 195 patients, with an average age of 51.18 years and 62.6% being female. Benign Paroxysmal Positional Vertigo (BPPV) was the most common cause (55.9%), often accompanied by symptoms such as tinnitus (30.8%), depression, and/or anxiety (29.2%). The majority of patients responded positively to vestibular rehabilitation, with 58% showing improvement as early as after the first session. Thus, it was identified that vertigo predominantly affects women over 50 years old, with BPPV being the main cause often associated with psychological symptoms. Vestibular rehabilitation proved effective in improving symptoms in a high proportion of patients. The population over 50 years old is most affected by vertigo, predominantly women, with BPPV as the primary cause and associated symptoms such as tinnitus and psychiatric disorders.

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Profile of deaths and hospitalizations of elderly patients affected by stroke in Northeastern Brazil: analysis of a 12-year epidemiological overview

Marconny Alexandre Oliveira de Medeiros Cavalcante[1], Thiago Luís Marques Lopes[1], Mateus Dutra Balsells[1], Hildel Freire Leite Filho[1], Matheus Lima de Oliveira[1], Oliver Reiks Miyajima[1], Lorenzo Marinho Morais[1], Daniel Pereira dos Santos[1], Chiara Gübel Portugal[1], Késia Sindy Alves Ferreira Pereira[1]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Marconny Alexandre Oliveira de Medeiros Cavalcante; Email: alexandremarc1912@gmail.com.

Background: Stroke is an acute illness caused mainly by an abrupt rupture of the cerebral vasculature or by a blockage of blood vessels (hemorrhagic and ischemic stroke, respectively), leading to tissue damage to this organ. The incidence of stroke and associated mortality increase with age, especially in elderly individuals. As life expectancy increases, the occurrence of strokes is expected to more than double in the next 30 years. Even with its relevance, this topic needs better elucidation, given the lack of publications related to stroke in this age group. Thus, the rationale for this study is to better understand this epidemiological profile in northeastern Brazil. **Objective:** To analyze the relationship between deaths and hospitalizations caused by stroke in the northeastern region of Brazil, using a twelve-year epidemiological overview. **Methods:** As this is an epidemiological study of secondary data, of the observational, descriptive and analytical type, the "TabNet" platform was accessed, where the axis "Epidemiological and Morbidity" and the topic "Hospital Morbidity of the SUS (SIH/SUS)" were chosen. Next, we selected "General, by place of hospitalization - from 2008 onwards" and "Brazil by Region and Federation Unit". Subsequently, in order to obtain the data for this study, the line "Age group 1", the column "Year of processing" the period "Jan/2012-Dec/2023" and the contents "deaths" and "hospitalizations" were adopted. Finally, from the available selections, "Cerebral vascular acid, ische, transit and correl syndrome" and "2 Northeast Region" were chosen in the ICD-10 Region and Morb List, respectively. **Results:** In the period from January 2012 to December 2023, 87,512 deaths from strokes were recorded, of which 67,920 (77.6%) occurred in elderly people. By separating this period into four three-year intervals, it is possible to perceive, comparing the 2012-2014 interval (14,333 deaths) with that of 2021-2023 (18,575 deaths), an increase of more than 25% in the number of deaths of people over 60 years of age. With regard to hospitalizations, during the 12-year period previously defined, 517,599 occurrences were recorded, of which the majority, 373,541 (72.2%), were associated with elderly people. Comparing the last three years (104,857 hospitalizations) with the oldest three-year period under analysis (77,912 hospitalizations), an increase of 34.6% is seen. **Conclusion:** Therefore, it is valid to infer that, along with the increase in the number of elderly individuals due to strokes, there was an increase in the number of deaths due to this type of episode in the population considered. Thus, due to the increasing relevance of this conjuncture, there is a need for more and more studies that evaluate the epidemiological circumstances linked to stroke in the elderly, in order to facilitate the understanding and, consequently, the attenuation of this phenomenon.

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Profile of hospitalizations by traumatic brain injury in elderly in Brazil from 2014 to 2023

Marcela Cristina de Souza Lemus[1], Anna Beatriz Dias Bertozzi[2], Ana Lígia Tiemi Soares Nishimura[3], Gabriela Silva Toro[4], Lívia Maria Oliveira Franco Vieira[5], Lucas Fernando Silva Moreira[6], Vitória Stefanny Souza Barbosa[7], Pedro Lucas Gomes Rodrigues[8], Yasmim Melo Costa de Azevedo[9], Cayara Mattos Costa[10]

[1]Universidade Federal do Rio de Janeiro, Macaé RJ, Brazil.
[2]Faculdade de Minas, Belo Horizonte MG, Brazil
[3]Universidade de Gurupi, Gurupi TO, Brazil.
[4]União Metropolitana de Educação e Cultuta, Lauro de Freitas BA, Brazil.
[5]Centro Universitário Inta.
[6]Centro Universitário FIPMoc, Montes Claros MG, Brazil.
[7]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.
[8]Universidade de Ribeirão Preto, Ribeiro Preto SP, Brazil.
[9]Universidade Nove de Julho, São Paulo SP, Brazil.
[10]Universidade Federal do Maranhão, São Luís MA, Brazil

Correspondence: Marcela Cristina de Souza Lemus; Email: marcelalemus47190@gmail.com.

Background: The traumatic brain injury (TBI) is a harm that occurs when there was a head impact, resulting in damages to the skull and the brain. In Brazil, occurred, in the last years, 131,014.83 hospitalizations because of TBI, showing up a theme of huge importance due to the high incidence and morbimortality. In the elderly, the TBI could be specifically worrisome due to aging of the central neurologic system and the fragility of the bones, increasing the risk of grave complications and revealing a problem of public health. **Objective:** Describe the profile of hospitalizations by traumatic brain injury in elderly in Brazil from 2014 to 2023. **Methods:** Ecological and descriptive study with the utilization of quantitative data extracted by the Department of Information and Informatics of the Unified Health System, of Hospitals Informations System of the Unified Health System, according to the International Classification of Diseases (CID-10). The hospitalizations analyzed covered the period of 2014 January to 2023 December, with analysis of the variables: hospitalizations per region, year and age group over 60 years. The data search was organized in spreadsheets of Microsoft Excel. **Results:** In the period analyzed, there were registered a total of 258,078 hospitalizations of elderly patients with traumatic brain injury in Brazil. Among the data analyzed, there are a higher incidence in the Southeast region, with 131,189 (50.8%) of the cases; followed by Northeast with 52,295 (20.2%); South with 45,327 (17.6%); Midwest with 17,813 (6.9%); and North with 11,454 (4.4%). In this period, there was an increasing number of hospitalizations, with a minimum in 2014 (20,953 cases) and a peak in 2023 (34,096 cases), in exception of 2020 (25,321), where it was a decrease in relation to 2019 (26,151). **Conclusion:** It was observed that the number of hospitalizations was concentrated in the most developed region of the country (Southeast) while the less developed region (North) presents the lower number of hospitalizations. In addition, there was a progressive increase in the numbers of hospitalizations per year. This study highlights the necessity of public actions aimed at prevention, especially in the elderly.

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Profile of hospitalizations for viral encephalitis in children and teenagers under 14 years old in the Southern region of Brazil from 2013 to 2023

Marina Ottmann Boff[1], Alessandro Batista Soares[1], Eloísa Bortolini[1], Eduarda Alexander Hilgert[1], Eduarda Gandolfi Horst[1], Júlia Supptitz[1], Natalie da Silveira Donida[1], Taís Michele Werle[1], Frederico Orlando Friedrich[1], Magda Lahorgue Nunes[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Marina Ottmann Boff; Email: mottmannboff@gmail.com.

Background: Viral encephalitis is a severe and rare condition characterized by inflammation of the brain parenchymal, associated with neurological dysfunction. The incidence of encephalitis in children at the age of two is about 16 per 100,000, and at fifteen years old, it is 1 per 100,000, highlighting its high morbidity rates. Hence, the importance of identifying the profile of children affected by this disease is emphasized, given its severity. **Objective:** Analyze hospitalizations for viral encephalitis in children and teenagers under 14 years old in the Southern Region of Brazil from 2013 to 2023. **Methods:** An ecological study with a quantitative approach was conducted in April 2024, using data from the Hospital Information System of SUS (SIH/SUS), linked to the Department of Informatics of the Unified Health System (DATASUS-TABNET). The study examined variables such as hospitalizations, year of service, Southern Region, state, sex, age group, color/race, deaths, and mortality rate. The study period was from January 2013 to December 2023. The data were organized into Microsoft Excel 365 spreadsheets and analyzed using descriptive statistics. **Results:** During the investigated period, the Southern Region recorded a total of 1,786 hospitalizations due to viral encephalitis. The distribution of hospitalizations was uneven, with the years 2013 and 2014 registering the highest percentages of cases (11.2% and 9.9%, respectively) and the years 2020 and 2021 registering the lowest (5.4% and 6.8%, respectively). From 2013-2015, the frequency of hospitalizations was 30.4%, from 2016-2018 it was 29.5%, and from 2019-2023 it was 40%. Paraná accounted for 51.2% of the cases, Rio Grande do Sul 30%, and Santa Catarina 18.8%. Males were more affected (58.5%) than females (41.5%). Regarding age groups, hospitalizations were distributed as follows: 14.4% in children under one year old, 37.5% in children aged 1 to 4 years, 27.2% in children aged 5 to 9 years, and 20.7% in children aged 10 to 14 years. In terms of color/race, 57.4% were white, 32.4% had no race information, 7.2% were mixed race, 2% were black, 0.7% were Asian, and 0.16% were indigenous. There were a total of 20 deaths from 2013 to 2023, with the year 2014 having the highest number of deaths (5) and the highest mortality rate (2.81/100,000). **Conclusion:** There is a close

relationship between hospitalizations for viral encephalitis in the Southern Region from 2013 to 2023, particularly in children aged 1 to 4 years, males, white children, and those from the state of Paraná, with the most negative outcomes in 2014. Additionally, the lower frequency of hospitalizations between 2020 and 2021 may be correlated with the SARS-CoV-2 pandemic. Therefore, public health measures that reinforce integrated care, especially for the youngest age groups, are essential.

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Profile of hospitalizations of patients affected by multiple sclerosis in Northeastern Brazil: a 12-year epidemiological overview

Marconny Alexandre Oliveira de Medeiros Cavalcante[1], Thiago Luís Marques Lopes[1], Mateus Dutra Balsells[1], Hildel Freire Leite Filho[1], Matheus Lima de Oliveira[1], Oliver Reiks Miyajima[1], Lorenzo Marinho Morais[1], Daniel Pereira dos Santos[1], Chiara Gübel Portugal[1], Késia Sindy Alves Ferreira Pereira[1]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Marconny Alexandre Oliveira de Medeiros Cavalcante; Email: alexandremarc1912@gmail.com.

Background: Multiple sclerosis (MS) is an idiopathic inflammatory demyelinating disease. One of the main suspicions regarding the etiology of MS is that it is associated with Epstein-Barr virus infections. This disorder is the most common neurodegenerative disease in young adults, affecting the performance of various daily activities of these individuals with the progression of the disorder and, thus, interfering with their quality of life. Despite such relevance, there is a lack of research on the occurrence of this disease in northeastern Brazil. Thus, based on the need for a better understanding of this scenario, the justification of this study is given by the intention to better understand this situation, analyzing its epidemiological profile in the referred region. **Objective:** To analyze the progression of hospitalizations caused by MS in the northeastern region of Brazil, using a twelve-year epidemiological overview. **Methods:** As this is an epidemiological study of secondary data, of the observational, descriptive and analytical type, the "TabNet" platform was accessed, where the axis "Epidemiological and Morbidity" and the topic "Hospital Morbidity of the SUS (SIH/SUS)" were chosen. Next, we selected "General, by place of hospitalization - from 2008 onwards" and "Brazil by Region and Federation Unit". Subsequently, in order to obtain the data for this study, the line "Age group 1", the column "Year of processing", including the period "Jan/2012- Dec/2023" and the content "hospitalizations" were adopted. Finally, in the available selections, "2 Northeast Region" and "Multiple Sclerosis" were chosen in the ICD-10 Region and Morb List, respectively. **Results:** From 2012 to 2023, 3,840 hospitalizations caused by MS were recorded in the Northeast of Brazil. Dividing this time interval into four three-year periods, it is noted that, comparing the 2021-2023 interval (1,813 hospitalizations) with the 2012-2014 interval (553 hospitalizations), there was an increase of more than 227.8% in the number of admissions to health services for this reason. It can also be observed that the majority of hospitalized people, 2811 (73.2%), belong to the age group ranging from 20 to 49 years and that young adults, represented here by individuals aged 20 to 29 years, represent 25.5% of the total hospitalizations in the Northeast, with 978 hospitalized. **Conclusion:** Therefore, it is correct to understand that, according to the literature, there is a very significant occurrence of MS in the young adult population. It can also be inferred that, in the period analyzed, there was a very strong increase in hospitalizations due to this neurodegenerative disorder, confirming the need for more and more studies to be developed to help in a deeper understanding of the epidemiological scenario in question, in order to promote greater precision in the identification of these cases of MS and, thus, improve the quality of life of the affected group.

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Profile of hospitalizations, deaths and mortality rate in Brazil due to malignant brain neoplasms from 2008 to 2023

Luís Henrique dos Santos Sousa[1], Vitor Fernando Bordin Miola[2], Maria Fernanda de ávila Reis[3], Rafael Alvarenga de Oliveira Pereira[4], Nicolas Minavia Delgadillo[4]

[1]Universidade de Pernambuco, Serra Talhada PE, Brazil.
[2]Universidade de Marília, Marília SP, Brazil.
[3]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[4]Universidade Metropolitana de Santos, Santos SP, Brazil.

Correspondence: Rafael Alvarenga de Oliveira Pereira; Email: rafael.oliva192@gmail.com.

Background: Malignant brain neoplasms, although less common than other types of cancer, represent a significant impact on global morbidity and mortality, being responsible for serious clinical outcomes and high costs for health systems. In Brazil, these neoplasms represent a particular challenge due to regional disparities in access to early diagnosis and specialized treatments, in addition to the aging population. Healthcare infrastructure, availability of medical resources, and unequal distribution of services can significantly influence patients' clinical outcomes. Therefore, understanding the profile of hospitalizations, deaths and the mortality rate associated with these conditions is essential to inform public health policies and direct resources effectively. **Objective:** To describe the number of hospitalizations, deaths and mortality rate due to malignant neoplasia of the brain in each region of Brazil, analyzing the period from 2008 to 2023. **Methods:** This is a quantitative, observational and descriptive study based on secondary data from the Department of Informatics of the Unified Health System (DATASUS), of the Ministry of Health, on cases of hospitalizations for malignant neoplasia of the brain in the last 16 years (2008 to 2023). **Results:** In Brazil, during the period observed, there were 215,386 hospitalizations for brain cancer. Of these, 43.6% were from the Southeast region, 23.5% from the South, 20.3% from the Northeast, 8.6% from the Central-West and 4.0% from the North. Of these hospitalizations, 76.5% were urgent, with an average length of stay of 10.7 days, with a mortality rate of 13.66%, which corresponded to 29,411 deaths, 46.5% of which were in the Southeast region, 21.7% from the South and 19.5% from the Northeast. **Conclusion:** A high incidence of brain neoplasms is noted in the Southeast when compared to other regions, representing a significant part of Brazilian health costs. Furthermore, it demonstrates regional disparities in the incidence of pathology or diagnostic capacity. Most hospitalizations occurred urgently and with a significant average length of stay, in addition to a significant mortality rate, which highlights the severity of the condition and the need for improvements to enable early diagnoses. Furthermore, more in-depth studies are needed to understand the causes of hospitalizations and develop strategies to avoid hospitalizations and deaths due to brain neoplasia.

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Profile of meningitis mortality in Brazil from 2010 to 2022: an ecological study

Barbara Zorzi Sanfins[1], Lucas Charuri de Andrade Castello Branco[1], Francisca Isabelly Leite de Almeida[2]

[1]Faculdade São Leopoldo Mandic, Campinas SP, Brazil.
[2]Centro Universitário Unifacisa, Campina Grande PB, Brazil.

Correspondence: Barbara Zorzi Sanfins; Email: basanfins@gmail.com.

Background: Meningitis is a disease characterized by inflammation of the meninges, caused mainly by viruses and bacteria. Despite the fact that vaccination is only available for some serogroups of bacterial etiology, 895 deaths from meningitis were reported in 2022, which constitutes a public health problem. To date, there is no literature analyzing the mortality profile in the selected period. **Objective:** To describe the epidemiological profile of meningitis deaths in Brazil between 2010 and 2022. **Methods:** Ecological study with secondary data collection on deaths from meningitis (ICD G03.9) in Brazil between 2010 and 2022. The data was collected from the Mortality Information System (SIM), managed by the Health Surveillance Secretariat in conjunction with the state and municipal health secretariats, available on the website of the SUS Information Technology Department - DATASUS. Descriptive statistics were carried out and the following variables were used: age, gender and region of notification. **Results:** A total of 12,066 deaths from meningitis were recorded in Brazil during the period analyzed. There was a pattern of decreasing deaths from 2010 to 2021, with an increase in 2022 (n=895) compared to the previous year (n=574), representing an increase of 55.92%. Deaths predominated in the 50-59 age group, totaling 15.5% (n=1,871). In addition, men were more affected, accounting for 59.18% (n=7,140) of deaths. The North and Midwest regions showed the lowest number of deaths in the period, despite the increase of 25.64% and 52.5% respectively, comparing 2022 with 2021. Meanwhile, the Southeast showed the highest mortality in all years, with an increase of 86.49% from 2021 to 2022. **Conclusion:** The epidemiological profile of deaths found in the period studied was: men, aged 50-59, with the highest numbers in the Southeast and the lowest in the North and Midwest. The analysis shows that there was an increase in deaths from meningitis from 2021 to 2022 in all regions. Therefore, greater incentive and public policy actions are essential to control and reduce the disease throughout the country, especially in the regions with the most deaths.

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Profile of mortality due to viral meningitis in Brazil between 2014 and 2023

Kaike Eduardo da Silva Lobo[1], Felipe Moraes Costa[1], Marcus Paulo Oliveira Lopes[1], Leonardo Bastos Santos[1], João Lucas Lalor Tavares[1], Flavio Lobo Altieri[1], Arthur Moraes dos Santos[1], Arley Nikson Oliveira Barreto[1], Ruan Pablo da Silva Gomes[1], Leonardo Aguiar dos Santos[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Marcus Paulo Oliveira Lopes; Email: marcus.polopes@aluno.uepa.br.

Background: Meningitis is an inflammatory process of the meninges, protective membranes surrounding the brain and spinal cord. Viral Meningitis, usually a self-limited condition with a favorable prognosis, has become more common than bacterial meningitis, which has declined due to successful vaccination efforts. Enteroviruses, herpes simplex virus, varicella-zoster and Epstein-Barr virus account for about 95% of Viral Meningitis cases. Despite its generally good prognosis, Viral Meningitis still contributes to mortality, particularly in vulnerable populations and in cases where timely medical intervention is not received. **Objective:** Evaluate the epidemiological profile of mortality due to Viral Meningitis in Brazil from 2014 to 2023. **Methods:** This is a descriptive ecological study, using secondary data in the public domain, accessed

from the Hospital Information System (SIH-SUS) in the database of the Information Technology Department of the Unified Health System (DATASUS). The variables analyzed were: number of hospitalizations, geographic region, sex, age and race. Descriptive statistics were performed using absolute (n) and relative (%) frequencies. **Results:** During the ten years analyzed, there were 886 deaths due to Viral Meningitis in Brazil, with a decrease from 113 deaths in 2014 to 95 in 2023. The year with the highest number of deaths was 2015 with 118, while 2020 had the lowest number with 61 records. In the distribution by region of the country, it was observed that 40.2% (356) occurred in the Southeast, 23.8% (211) in the Northeast, 21.6% (191) in the South, 8.1% (72) in the North, and 6.3% (56) in the Midwest. In addition, it is possible to note that mortality was higher in males, accounting for 56% (496), while females accounted for 44% (390) of the deaths. The age groups most affected by mortality due to viral meningitis were as follows: those aged 30 to 39 years, comprising 16.2% (144); those aged 40 to 49 years, comprising 14.2% (126); and those aged 50 to 59 years, comprising 12.3% of deaths (109). Furthermore, mortality was higher among individuals of brown race, with 34.6% (307) of the deaths, followed by those of white race, with 33.5% (297). **Conclusion:** Despite a slight reduction in mortality over the ten-year analysis, it remains high, predominantly affecting individuals in the Southeast region, males, those aged 30-39, and those of brown race. These findings underscore the need for targeted public health interventions and improved health management strategies, particularly in the most affected regions and demographic groups, to reduce the burden of Viral Meningitis in Brazil.

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Profile of multiple sclerosis in Brazil over the last 12 years in an epidemiological and cost analysis: an underestimated disease ?

Lorenzo Marinho Morais[1], Thiago Luís Marques Lopes[1], Oliver Reiks Miyajima[1], Chiara Gübel Portugal[1], Marconny Alexandre Oliveira de Medeiros Cavalcante[1]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Lorenzo Marinho Morais; Email: lorenzo.morais@aluno.uece.br.

Background: Multiple sclerosis (MS) is the leading cause of non-traumatic disabilities around the world, mainly affecting young adults. It is characterized by a relapsing-remitting stage and a possible secondary progressive stage. In Brazil, MS affects approximately 40,000 people. This study becomes relevant due to the increase in hospitalizations in the country over the years, raising questions about possible causes of this raise and how the government has reacted to economically manage it through the Universal Health System (SUS). **Objective:** To analyze hospitalizations and SUS expenses for MS in Brazil over the last 12 years. **Methods:** This is an analytical epidemiological study. The "TabNet" platform was used. Under the section "epidemiological and morbidities", in the topic "Hospital Morbidity of the SUS (SIH/SUS)", the subtopic "General, by place of hospitalization - from 2008" was selected, defining "Brazil by Region and Federation Unit" as the data territory. Then, the cell content "Internment" was selected. The cause "Multiple Sclerosis", the period "2012-2023", and the variables of "age group", "sex" and "region" were analyzed. "Total value" was accessed, in which the annual cost was analyzed before and after monetary correction, through the website of the Central Bank of Brazil. **Results:** From 2012 to 2023, there were 1,806, 1,793, 2,021, 2,078, 3,168, 4,134, 4,963, 4,374, 3,972, 5,456, 7,299 and 6,485 cases, totaling 47,549 hospitalizations and an increase of 259.08% between first and last year. Dividing this period quadrennially, it is inferred that from the first (7,698 cases) to the second (16,639 cases) there was an increase of 116.15% and from the second to the third (23,312 cases) there was an increase of 39.50%. All regions had an increase, with the Northeast having the highest percentage increase, followed by the Southeast, Central-West, North and South. Females had more hospitalizations, with a ratio of 2.3 women to 1 man, and the most affected age group was 20 to 49 years old, representing 74.23% of all cases. As for expenses, R$27,583,582.83 was spent by the SUS. Comparing the values of 2012 (R$1,394,753.77) and 2023 (R$3,649,310.85), there was an increase of 161.14%. After monetary correction, this increase drops to 17.96%. Finally, in 2012, R$1,712.94 was spent per hospitalization, while in 2023 this value was R$562.73. **Conclusion:** Hospitalizations due to MS have increased significantly over the years. However, the amounts spent by the SUS have not increased proportionally. This should worry the government, since the disease mainly affects economically active adults. Therefore, more studies should be conducted to understand the reasons for the increase in hospitalizations, evaluating the impact of new diagnostic methods, as well as to understand why the government has not been able to reduce this increase with the amounts currently expended on hospitalization, evaluating the efficiency of SUS treatments.

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Profile of neurological complications in patients treated with CAR-T cell therapy at a Nove de Julho Hospital in são Paulo

Augusto Bragança Reis Rosa[1], Lucas Bruno Rezende[1], Gustavo Ramos de Lima[1], Ana Carolina Zetehaku[1], Antonio Francisco de Araújo Netto[1], Carolina Novaes Perniqueli[1], Giancarlo Fatobene[1], Celso Arrais[1], Vanderson Geraldo Rocha[1], Rafael Paternò Castello Dias Carneiro[1]

[1]Hospital Nove de Julho, São Paulo SP, Brazil.

Correspondence: Augusto Bragança Reis Rosa; Email: augustobraganca@gmail.com.

Chimeric Antigen Receptor (CAR) T cell therapy is an emerging therapeutic modality used in patients with hematopoietic and lymphoproliferative tumors, with the expectation of increased availability in the future, including for the treatment of solid tumors. The most common complications associated with its use include Cytokine Release Syndrome (CRS) and Immune Effector Cell-Associated Neurotoxicity Syndrome (ICANS), which can range in severity from mild to fatal. ICANS, may present with a variety of neuropsychiatric symptoms such as encephalopathy, behavioral changes, visual and auditory hallucinations, linguistic and speech disturbances, headache, fatigue, tremor, dysgraphia, clinical or subclinical seizures, coma with cerebral edema, and death. Onset is usually three to ten days after immunotherapy infusion, more frequently following previous CRS. Daily screening for neurological symptoms along with bedside tools such as the Immune Effector Cell-Associated Encephalopathy (ICE) score, aid in identifying and monitoring symptoms. Additionally, electroencephalogram, cranial MRI, and lumbar puncture help to understand the greater involvement of the central nervous system. Patients at higher risk for ICANS include younger individuals, those with preexisting neurological conditions and medical comorbidities, higher malignancy burden, severe lymphocyte depletion and other cytopenias, and those with severe and early-onset CRS. In our center CAR-T cell therapy has been in use since March, 2023. Six patients have been treated thus far, all males. Median age at treatment was 65 years old (24 - 82), and the most common therapeutic indication was Diffuse Large B-Cell Lymphoma. Therapeutic cells used were axicabtagene ciloleucel (Yescarta®) and tisagenlecleucel (Kymriah®). Herein, we describe the clinical and radiological profile of patients undergoing CAR T cell therapy at Nove de Julho Hospital, as well as the occurrence of complications such as CRS and ICANS associated with this treatment.

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Profile of patients affected by movement disorders in subspecialized outpatient clinics of a tertiary hospital in Southern Brazil

Eduardo Drews Amorim[1], Daniel Teixeira dos Santos[1], Thalya Osmilda Alves de Carvalho[2], Patricia Ribeiro Rigo[2], Jullivan Käfer Pasin[2], Isabella Fonseca Benati[2], Manoella Guatimuzim Testa da Silva[2], Gabriela Magalhães Pereira[2], Artur Francisco Schumacher Schuh[1,2]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Eduardo Drews Amorim; Email: eduardodrewsamorim@gmail.com.

Background: The distribution of movement disorders in outpatient settings is key to planning and resource allocation, but data is lacking, particularly in southern Brazil. Profile of patients affected by movement disorders in subspecialized outpatient clinics of a tertiary hospital in Southern Brazil **Objective:** To delineate the epidemiological and clinical characteristics of movement disorders (MD) in patients attending subspecialty clinics at a tertiary hospital in Southern Brazil. **Methods:** This cross-sectional study analyzed medical records from patients with MD who consulted in 2023 at specialized outpatient clinics of a public academic hospital. It included patients who had at least one visit that year and used ANOVA, Kruskal-Wallis, and Chi-square tests to examine demographic and clinical differences across MD syndromes. **Results:** Patient distribution across outpatient clinics are as follows: 307 in dystonia, 289 in parkinsonism, 90 in the DBS clinic, and 246 in the other MD clinic. Significant age and sex differences existed across clinics ($p<0.0001$). Comorbidities profiles among clinics are also detailed. Regarding MD syndromes distributed across all clinics, parkinsonism (362 cases), dystonia (330), ataxia (128), and tremor (59) were the most common. Significant age and sex differences were noted ($p<0.0001$). Dystonia was more prevalent in females (69.7%), while tremor and parkinsonism cases were older (mean ages: 68.0±13.2, 65.8±10.2 years). Chorea and ataxia patients showed younger onset ages (13.6±14.4, 32.9±16.2 years). The syndromes with the most diagnostic delay were other movement disorders (paroxysmal dyskinesias, tics and myoclonus), tremor, and ataxia, averaging 16.0±21.0, 6.0±5.3, and 4.7±5.6 years, respectively. The most common etiology for each syndrome are the following: Parkinson's disease (87.8%) in parkinsonism; primary dystonia (85.2%) in dystonia, hereditary ataxia (70.3%) in ataxia, essential tremor (62.7%) in tremor, and Huntington's disease (75%) in chorea. Notably, a considerable number of tremor and ataxia patients (33.9% and 23.4%, respectively) lacked a definitive etiological diagnosis. **Conclusion:** This study at a Southern Brazilian tertiary hospital shows a varied profile of MD, with significant age, sex, age at onset and diagnostic delay differences. The high number of undiagnosed cases indicates limited access to advanced genetic testing in Brazil's public health system.

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Profile of patients who underwent intravenous thrombolysis after ischemic stroke at a stroke care unit

Luis Bandeira Alves Neto[1], Maria Eduarda Moreira Cardoso[2], Noelle Ventura Jordão[2], Maria Luiza Vasconcelos Montenegro[1], Isaias Henrique Martins de Vasconcelos Norte Bezerra[1], Lívia Barbosa Cavalcanti[1], Caio Peixoto Parizi[1], Ricardo Martins Silva[2], Monique Evelyn Mendonça do Nascimento[2], Ana Dolores Firmino Santos do Nascimento[2]

[1]Universidade de Pernambuco, Recife PE, Brazil.
[2]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Luis Bandeira Alves Neto; Email: luis.bandeira@upe.br.

Background: Ischemic stroke is associated with high rates of functional disability and, consequently, high health costs, corresponding to a severe global public health problem. This disease accounts for 80% of the cases, and its early diagnosis is essential to the possibility of thrombolytic therapy, which can improve neurological recovery rates and reduce rates of incapability. The analysis of variables in patients undergoing this therapy has the potential to enhance the outcomes in these patients. **Objective:** To study the profile of patients who underwent intravenous thrombolysis with alteplase in a stroke care unit in northeastern Brazil. **Methods:** This retrospective study considers medical records from January 2023 to May 2024. **Results:** In the study population, the mean age was 65 years old, 55% male. The most common comorbidities in patients were hypertension (76%), diabetes mellitus (31%), previous stroke (10.6%) and smoking (6.8%). Atrial Fibrillation (AF) was present in 9.3% of the patients, and 4% of them had a previous heart attack. The mean time from ictus to the hospital was 152 minutes. The mean time from arrival to CT scan was 18.2 minutes, and the mean door-to-needle time was 48.2 minutes. The mean National Institutes of Health Stroke Scale (NIHSS) score was 14.45 (±7.64) at admission and 11 (±12.07) at discharge, with difference between them of 3.45, no statistically significant (95% CI: -1.73, 8.63; p=0.18). All intracranial hemorrhage (including symptomatic and non-symptomatic) was seen in 12.3% of patients. The overall in-hospital mortality was 3%. **Conclusion:** As prevention strategies struggle to reduce stroke incidence, the literature reports outcome enhancement by thrombolysis. Lower ictus-to-hospital, door-to-needle, and ictus-to-needle time have been linked to fewer deficits and higher functionality post-treatment. This study is vital once it can improve the understanding of the patients with acute ischemic stroke, allowing the development of strategies to achieve better outcomes and fewer thrombolytics adverse effects.

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Profile of patients with chronic demyelinating polyneuropathy (CIDP) according to the 2021 diagnostic criteria in a reference center: a series of cases

Maria Eduarda Moreira Cardoso[1], Bruno Paulo Teles Chaves[1], Lidiane Alves de Macedo[1], Monique Evelyn Mendonça do Nascimento[1], Maria Íris Morais Machado[1], Vanessa Cristina Fragoso Cassiano Alencar[1], Alvaro José Porto Moreira[1], Marcelo Ataíde de Lima[1], Maria Lúcia Brito Ferreira[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Maria Eduarda Moreira Cardoso; Email: dudacardoso1145@gmail.com.

Background: Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated disease whose diagnostic criteria have been updated recently. Identification of the disease, although challenging, is crucial because it can be functionally debilitating if not diagnosed and treated correctly. **Objective:** To describe the socio-demographic, laboratory characteristics, and treatments performed on patients. **Methods:** Retrospective study with information from medical records followed up at a reference center for neuroimmunological diseases, at Hospital da Restauração, from 1999 to 2024. **Results:** The sample consisted of 25 patients, 56% female. The average age at onset of symptoms was 34.79 years (from 2 to 66 years), with a mean time to diagnosis of 31.42 months. There was a high prevalence of pediatric patients, with 32% under 16 years of age, with a mean time to diagnosis of 51.28 months. Regarding ethnicity, we identified that 13% of patients were white, 65.2% were mixed race, and 21.7% did not specify their ethnicity. All individuals underwent electroneuromyography, and 81.81% were compatible with chronic demyelination. The mean cerebrospinal fluid (CSF) cellularity was 6.85 (from 0 to 93), and the mean CSF protein was 111.86 (from 24 to 293). In the study population, 32% did not present comorbidities, and among these, the most common were diabetes mellitus (20%), systemic arterial hypertension (16%) and hypothyroidism (12%). Other less common ones included Chikungunya, Dengue, non-Hodgkin's lymphoma type B, Hashimoto's thyroiditis, papillary thyroid tumor, plasmacytoma, and POEMS syndrome, among others. In 12% of patients, there was a report of recent vaccination. Relative to CIDP subtypes, 26.7% of patients presented the typical form, including 46.7% with an acute initial presentation, mimicking Guillan-Barré Syndrome. Regarding treatment, 77.3% of patients received induction therapy with intravenous immunoglobulin. Additionally, 40.9% received pulse therapy with methylprednisolone, 4.5% received high-dose oral prednisone, and 4.5% underwent plasmapheresis. Two patients received induction treatments with methylprednisolone followed by intravenous immunoglobulin. Concerning maintenance therapy, 56.3% of patients received monthly intravenous immunoglobulin, 18.8% received monthly pulse therapy with methylprednisolone, 12.5% received daily oral corticosteroids, and 12.5% received azathioprine. Of the 17 patients with treatment information available, only 17.6% showed a complete response to treatment. **Conclusion:** Considering CIDP is a rare disease with significant morbidity, with evolving diagnostic and treatment concepts, it is vital to establish a patient profile for differential diagnosis and treatment. In light of discoveries and reclassifications, especially considering autoimmune nodo-paranodophaties, there must be a review of these cases considering red flags for possible nosological review.

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Profile of renal function in stroke patients with high risk of obstructive sleep apnea: a transversal study

Aurea Maria Lago Novais Castello Branco[1], Juliana Cerqueira Gomes[1], Marina Soledad Benitez Bitencourt[1], Thaise Braga de Oliveira[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Renan Carvalho Castello Branco; Email: castellorenan@yahoo.com.br.

Renal impairment is known as risk fator to worst outcomes in stroke patients. Recently, stroke has been associated to obstructive sleep apnea (OSA), but the link between OSA-stroke-renal impairment is not yet known, neither the renal function is related. This study objectives to describe renal function of stroke patients with diferente risks of obstructive sleep apnea. We performed a transversal study, part of a cohort of stroke patients. Individuals were evaluted to risk of OSA with STOP-Bang and SOS scores. Renal function (creatinine and urea) at admission,peak,and discharge was assessed, described with dispersion measures and compared between groups with T test/Mann-Whitney, depending on normality (Kolmogorov-Smirnov test). We obtained 430 patients (69% classified as high risk with STOP-Bang and 50% with SOS). In high risk with STOP-Bang: creatinine of admission median 1 (0.89-1.31), peak 1.7 (1.02-1.62), discharge median 1.1 (0.9-1.3) and clearance median 55.5 (39.4-76). Urea of admission mean 35 (27-48), peak mean 49 (36-83) and discharge mean 38 (28-50). In low risk with STOP-Bang: creatinine of admission mean 0.89 (0.74-1.1), peak 1.08 (0.89-1.28), discharge mean 0.97 (0.81-1.13) and clearance median 59.6 (43-80.3). Urea of admission mean 33 (26-38), peak mean 42 (32-58) and discharge mean 32 (25-45). There was significant difference between groups with all the renal measures (p<0.02), considering STOP-Bang. With SOS score, discharge urea was significant (p=0.02). This is the first study to demonstrate significant diferences between renal function in stroke patients with different OSA risks. It can lead us to hypothetise about a connection between OSA, renal impairment and stroke. Patients are admited with similar measures, but the difference is significant, which reafirm the relation between obstructive sleep apnea and kidney impairment. Individuals are admitioned with similar measures (althought significant), but those with higher risk are discharged with worst renal function, suggesting that stroke triggers acute renal injury in patients with high risk of OSA. Further studies are needed to clarify subjacent mechanisms.

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Progressive encephalomyelitis with rigidity and myoclonus (PERM) associated with anti-GAD antibodies

Luiz Eduardo Betti Paiva[1], João Henrique Fregadolli Ferreira[1], Lívia Almeida Dutra[1], Caio César Diniz Disserol[2,3,4]

[1]Hospital Israelita Albert Einstein, Instituto do Cérebro, São Paulo SP, Brazil.
[2]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[3]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[4]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Luiz Eduardo Betti Paiva; Email: luizeduardobettipaiva@gmail.com.

Case presentation: A 32-year-old woman presented, in the last month, severe lumbar pain, acute spasms of the abdominal, dorsal, and thoracic musculature lasting minutes to hours and triggered by tactile stimuli, sound, or movement, with significant impact on daily activities. She also developed hyperhidrosis, hypertension, tachycardia and episodes of anxiety when climbing the stairs, bathing, and loud noises. Within two weeks she developed focal seizures evolving to bilateral tonic-clonic seizures. After ten days, she was admitted to a hospital due to decreased level of consciousness and worsening of the spasms and myoclonus. Her physical examination showed a rigid abdomen, which led her to undergo an exploratory laparotomy to rule out abdominal life-threatening causes. She remained in the ICU for 40 days, lost 30kg, and required tracheostomy and gastrostomy. After slight clinical improvement, the patient was discharged without a diagnosis. Two weeks later, neurologic examination revealed hypertonic gait, severe hypertonia of abdominal and lumbar muscles and hyperreflexia. Brain and spinal MRI were normal, antineuronal antibodies were tested in serum and CSF, both of which revealed anti-GAD antibodies, confirming the diagnosis of anti-GAD-associated PERM. The patient was started on IVIg, with partial response, and she was switched to rituximab. **Discussion:** PERM is a clinical syndrome characterized by stiffness, muscle spasms, reflex myoclonus, encephalopathy, symptoms affecting the brainstem and spinal cord, specific-task phobias, and dysautonomia. PERM is mainly associated with anti-GAD and anti-glycine (GlyR) antibodies. Pathophysiology involves autoimmune responses leading to neuroinflammation, which contributes to neuronal damage and loss of inhibitory neurotransmission such as glycinergic and GABAergic signaling, resulting in neuronal hyperexcitability. PERM is considered part of the stiff-person syndrome spectrum and differs from stiff-person itself by the occurrence of encephalopathy, dysautonomia and brain stem involvement. Management involves symptom-alleviating, and combinations of corticosteroids, IVIg, and plasma exchange have shown sustained improvement, with rituximab in refractory cases. **Final comments:** This report aims to alert the medical community regarding PERM, in order to prevent diagnostic delay and consolidate

knowledge about the most associated antibodies and the importance of testing them, as well as to outline the main recommended treatment approaches for this rare and complex autoimmune disorder.

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Progressive multifocal leukoencephalopathy as a differential diagnosis for rhombencephalitis in an HIV-positive patient: a case report

Caique Alberto Dosualdo[1,2], Lucas do Rego Barros Correa Martins[1,2], Matheus Hisamitsu Facine[1,2], Humberto Ramos Crispim[1,2]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.
[2]Fundação Faculdade de Medicina de Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Caique Alberto Dosualdo; Email: caiquedosualdo100@gmail.com.

Case presentation: A 39-year-old male patient presented with a progressive 3-month history of diplopia, associated with hypoesthesia on the right side of his face, progressing to motor incoordination and imbalance, as well as gait ataxia and unspecified limb weakness during the period. Initially investigated elsewhere, cerebrospinal fluid showed mild pleocytosis with lymphomonocytic predominance and slightly elevated protein levels. Brain MRI revealed hyperintense T2/FLAIR lesions involving the pons, middle cerebellar peduncles, dentate nuclei, and cerebellar white matter, without contrast enhancement. Initially treated with 5 days of pulse therapy for suspected demyelinating disease, the patient was later evaluated in our service with positive HIV serology and CD4 count of 55. Further investigation for listeriosis via CSF culture and PCR, as well as molecular tests for herpes family viruses, yielded negative results. The patient received empirical therapy with acyclovir, ampicillin, and gentamicin. A cerebral PET/CT scan showed diffuse hypermetabolism in the brainstem and bilateral mesial temporal regions. Polyomavirus testing in urine was positive. Despite therapy, the patient clinically deteriorated, prompting a stereotactic brain biopsy in the right cerebellar peduncle region, which histopathological analysis revealed inclusions staining positive for SV-40, confirming the diagnosis of progressive multifocal leukoencephalopathy. **Discussion:** The investigation of rhombencephalitis poses a significant challenge for neurologists and general practitioners, encompassing various etiologies such as autoimmune, infectious, neoplastic, and metabolic causes. Early diagnosis and treatment are crucial for reducing morbidity and mortality. In this case, stereotactic biopsy provided definitive diagnosis confirmation; however, in most cases, this is not feasible, necessitating investigation through CSF profile, imaging patterns, clinical progression, and confirmatory tests in serum or plasma. **Final comments:** Attention to clinical history and results from complementary exams should guide the next steps in the investigation and empirical treatment of rhombencephalitis, aiming to reduce morbidity and mortality.

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Progressive proximal weakness in an adult female: a case of spinal muscular atrophy type III with p.Ala2Gly variant

Maria Clara Motta Barbosa Valente[1], Rafael Chaves Claudino de Queiroga[1], Letícia Machado Dumont[1], Camila Lopes Figueiredo[1], Filipe Brito Ferraz da Silveira[1], Ana Clara Teixeira Vaz[1], Renan Brandão Rambaldi Cavalheiro[1], Ana Luisa Vilela Sá Fortes[1], Francisco Tomaz Meneses de Oliveira[1], Melissa Nunes Polaro[1]

[1]Santa Casa de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Clara Motta Barbosa Valente; Email: mariaclarambvalente@gmail.com.

Case presentation: A 42-year-old female presented for the first time to our service with a history of progressive motor weakness starting at the age of 17. The weakness initially affected her lower limbs, predominantly proximally, and gradually progressed over the years. In 2003, she was evaluated by a neurologist and diagnosed with multiple sclerosis (MS), but she did not have any medical records from that time and denied taking any medication. The symptoms continued to progress over the years and eventually involved her upper limbs (also predominantly proximally) and were associated with significant muscle atrophy. The patient was currently wheelchair-bound. She denied any other comorbidities. The patient had two brothers who were also diagnosed with MS and died from the disease in adulthood. She did not know of any other family history and denied consanguinity of her parents. The patient was admitted for further investigation. Initial laboratory tests were within normal limits, with no abnormalities suggestive of muscle disease or other metabolic etiologies. Magnetic resonance imaging (MRI) of the brain, cervical spine, and thoracic spine ruled out lesions in the central nervous system but showed significant liposubstitution in the paraspinal muscles. MRI of the thigh revealed diffuse atrophy and liposubstitution in the pelvic and thigh muscles, as well as edema in the gluteus maximus, suggesting chronic myopathy. Based on the findings of the neurological examination and MRI, myopathy was suspected. However, electroneuromyography (ENMG) showed anterior horn involvement of the spinal cord, without signs of current denervation, suggesting the diagnosis of Spinal Muscular Atrophy (SMA). The definitive diagnosis was confirmed by genetic testing, which identified the p.Ala2Gly variant in the SMN1 gene, in addition to two copies of the SNM2 gene. **Discussion:** The clinical presentation and course of the case are consistent with SMA type III, with onset in adolescence, gradual progression of muscle weakness, and proximal motor involvement in both the upper and lower extremities. The c.5C> G (p.Ala2Gly) variant in the SMN1 gene is located in exon 1 and has been reported in patients with SMA type II and III. In some specific mutations in SMA cases, there is a correlation between clinical phenotype and the number of SMN2 copies, but the c.5C> G variant generally determines a milder phenotype regardless of SMN2 copies. Genetic testing in this context was essential for the confirmation of the diagnosis. **Final comments:** SMA type III is a rare disease, but with proper diagnosis and treatment, disease progression can be slowed and patients' quality of life can be significantly improved. This case highlights the importance of recognizing the symptoms of SMA, especially in the adult population, and emphasizes the importance of genetic testing for definitive diagnosis.

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Progressive supranuclear palsy with predominant cerebellar ataxia: clinical and neuroimaging manifestations of two cases

Olívia Gameiro de Souza[1], Jano Alves de Souza[2], Anderson Ribeiro de Sales[3,4], Johnatan Felipe Ferreira da Conceição[1], Nicole Mancen Freire Rosa[1], Laísa Fernandes Bonatto[1], Ana Luísa Utrine Pimentel[1], Sofia Gameiro de Souza[2], José Antônio Vieira da Silva Calsoni[2], Rodrigo Amorim Jorge[5]

[1]Universidade Federal Fluminense, Hospital Universitário Antônio Pedro - EBSERH, Niterói RJ, Brazil.
[2]Universidade Federal Fluminense, Niterói RJ, Brazil.
[3]IRSA Diagnóstico Por Imagem, Niterói RJ, Brazil.
[4]Hospital Universitário Clementino Fraga Filho, Rio de Janeiro RJ, Brazil.
[5]Universidade Unigranrio Afya, Duque de Caxias RJ, Brazil.

Correspondence: Olívia Gameiro de Souza; Email: gameiroolivia@gmail.com.

Case presentation: Case 1: 89 years old white woman. The condition began 7 years ago with unexplained falls, imbalance, retropulsion when trying to get up, fatigue, and mild cognitive impairment. At that time, the neurological examination was normal; mini-mental = 25/30. Brain MRI showed enlargement of sulci and fissures compatible with the age; mild microangiopathy (FAZEKAS 1); MTA 1 hippocampi; normal spectroscopy, brainstem and basal nuclei unaltered. Current appointment revealed tendency to fall, mainly backwards, dysphagia. Physical examination showed hypomimia, procerus sign, absence of Romberg's sign, positive pull test, base enlargement, imbalance when turning around own axis, axial hypertonia, bilateral dysdiadochokinesia, unchanged sensitivity and reflexes, supranuclear vertical ophthalmoplegia, square waves, applause sign (perseveration in clapping), dysarthria (scanning speech). No symptoms or signs of dysautonomia (no urinary incontinence or postural hypotension). A new MRI of the brain revealed changes in the upper contour and volumetric reduction of the mesencephalon, usually associated with progressive supranuclear palsy; high parkinsonism index: 10.3 (version 1.0) and 2.3 (version 2.0). No signs suggestive of olivopontocerebellar atrophy were found. Case 2: 78 years old white woman. Onset one year ago with dysarthria, cognitive impairment, emotional lability. Followed by mood depressive disorder and imbalance. Neurological examination: bradykinesia, hypomimia, dystonic neck posture, eyelid retraction, procerus sign, reduced automatic arm movements, axial and upper limb rigidity, dysdiadochokinesia, dysarthria (scanning speech), pseudobulbar affect (involuntary crying and laughter), bradyphrenia. Neuropsychological assessment revealed CDR 1. No symptoms or signs of dysautonomia (no postural hypotension, effort urinary incontinence). MRI of the brain showed changes in the upper contour and volumetric reduction of the mesencephalon; parkinsonism index 15.0 (version 1.0) and 5.1 (version 2.0). No signs suggestive of olivopontocerebellar atrophy were found. **Discussion:** Progressive Supranuclear Palsy is a neurodegenerative disease with different phenotypes. The predominantly cerebellar form is rare and can be confused with cerebellar form of Multiple System Atrophy. An important distinction between the two conditions is the absence of dysautonomia in Progressive Supranuclear Palsy, which is, on the other hand, one of the diagnostic criteria for Multiple System Atrophy. Two cases with clinical presentation and imaging findings compatible with Progressive supranuclear palsy with predominant cerebellar ataxia have been reported. **Final comments:** Progressive supranuclear palsy should be considered in the differential diagnosis of late-onset cerebellar ataxia. Detailed medical history, repeated neurological examinations at each appointment, and imaging studies contribute to establishing the diagnosis and differentiation from other neurodegenerative diseases.

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Prospective randomized study to evaluate the efficacy of additional dopaminergic replacement in cociceptive pain in patients with Parkinson's disease

Darah Fontes da Silva Assunção[1], Marina Buldrini Filogonio Seraidarian[1], Isabella Nascimento Guedes[2], Alessandra Fonseca[1], Clarice Listik[1], Manoel Jacobsen Teixeira[1], Egberto Reis Barbosa[1], Rubens Gisbert Cury[1], Gabriel Taricani Kubota[1], Daniel Ciampi de Andrade[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[2] Universidade Estácio de Sá, Rio de Janeiro RJ, Brazil.

Correspondence: Isabella Nascimento Guedes;
Email: isabellaguedes21@icloud.com.

Background: Pain is a common non-motor symptom of Parkinson's disease (PD) that disables and significantly compromises patients' quality of life. However, there is little data on the best treatment for this symptom. Although clinical experience and evidence from observational studies suggest a role for additional dopaminergic replacement therapy (DRT) in its treatment, clinical trials are needed to confirm this hypothesis. **Objective:** To evaluate DRT efficacy on nociceptive pain in PD patients. **Methods:** The present study is a randomized, double-blind, placebo-controlled, monocentric clinical trial that is currently ongoing. The data collection is expected to be completed in January 2025, with subsequent data analysis and results disseminated. 90 patients with PD will be selected, aged 18 to 85 years, with nociceptive pain related to PD according to the Parkinson's Disease Pain Classification System (PD-PCS), with pain intensity ≥4 out of 10 on the numeric rating scale (NRS). Participants will be divided into 3 groups of 30 people each (A, B and C). Group A will receive placebo, group B an DRT of 10% of the levodopa equivalent daily dose (LED), and group C an DRT of 20% of the LED. The treatment intervention will last 14 days. The DRT will be performed in all groups with levodopa/benserazide tablets and the placebo will be composed of tablets similar to those of the active medication. During the study, patients will be assessed in person at baseline (D0) and by videoconference at the end of treatment (D14) according to: numeric rating scale (NRS), PD-PCS, Brief Pain Inventory, 8-item Parkinson Disease Questionnaire, Hospital Anxiety and Depression Scale, Global Impression of Change and the third item of the MDS Unified Parkinson's Disease Rating Scale. Efficacy analyzes will be carried out by intention to treat, and the primary outcome will be the variation in pain intensity in NRS between D0 and D14. The level of statistical significance will be $p<0.05$. The number needed to treat (NNT) will be calculated to reduce NRS by ≥30% and ≥50%.

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Pro-thrombotic antibodies in patients with post-infection or post-COVID-19 vaccine stroke

Marta Rodrigues de Carvalho[1], Felipe Von Glehn Silva[2], Lucas Luiz Vieira[3], Márcia Silva Santos Neiva[2], Maria Lucia Brito Ferreira[4], Jurandy Junior Ferraz de Magalhães[5], Clarice Neuenschwander Lins de Morais[6], Cristiane Campello Bresani Salvi[6], Maria de Fatmia Pessoa Militão de Albuquerque[7], Maria Cynthia Braga[7]

[1]Hospital de Base, Brasília DF, Brazil.
[2]Hospital Universitário de Brasília, Brasília DF, Brazil.
[3]Laboratório Central de Saúde Pública, Brasília DF, Brazil.
[4]Hospital da Restauração, Recife PE, Brazil.
[5]Laboratório Central de Saúde Pública, Recife PE, Brazil.
[6]Fundação Oswaldo Cruz, Instituto Aggeu Magalhães, Laboratório de Virologia e Terapia Experimental, Recife PE, Brazil.
[7]Fundação Oswaldo Cruz, Instituto Aggeu Magalhães, Recife PE, Brazil.

Correspondence: Marta Rodrigues de Carvalho;
Email: martinhabhs@gmail.com.

Background: Stroke is one of the main causes of morbidity and mortality in the world. With the advancement of the SARS-CoV-2 pandemic, cases of stroke in patients hospitalized with COVID-19 have accumulated in the scientific literature. **Objective:** To investigate the association between exposure to SARS-CoV-2 and the presence of pro-thrombotic factors in stroke patients treated in public neurology services of the SUS. **Methods:** This is a prospective, hospital-based study. The study is nested within the multicenter case-control titled NeuroCOVID: Association of SARS-CoV-2 with the occurrence, prognosis and pathogenesis of cerebrovascular diseases and other neurological manifestations, funded by the Inova Fiocruz Program (Fundação Oswaldo Cruz - Fundo Emergencial de Combate to COVID-19: VPPCB-005-FIO-20-2-22). **Results:** 148 patients were enrolled for this study at a single center. Among the participants, there was a predominance of female individuals (51.4%, n=76), white ethnicity (29.1%, n=43) and specifically the economically active population (43.9%, n=65). The majority of admitted patients had acute/subacute exposure to SARS-CoV-2 (72.3%, n=107), most of whom were previously vaccinated (80.4%, n=86). Ischemic troke constituted the majority of stroke cases in this study (77%, n=114). The average age was 55.3 years, with a median of 55.5. Fifteen patients (10.1%) showed expression of some prothrombotic antibody (IgM-B2 glycoprotein, IgA-B2 glycoprotein, IgM Anticardiolipin, IgA-Anticardiolipin or Anti-PF4). It is clear that the majority of patients who were positive for any antibody were in the acute/subacute phase of exposure classification. **Conclusion:** This study is one of the pioneers in demonstrating clinical-epidemiological and laboratory characteristics of patients investigated with stroke. The studied sample presents characteristics similar to those described in other clinical studies.

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Protocols for analyzing effective connectivity in patients with refractory frontal and temporal lobe epilepsies

Helka Fabbri Broggian Ozelo[1], Benito Pereira Damasceno[2], Fernando Cendes[2], Andréa Alessio[2]

[1]Unviersidade Federal de São Carlos, Araras SP, Brazil.
[2]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Helka Fabbri Broggian Ozelo;
Email: helka@ufscar.br.

A growing literature has corroborated the understanding of focal epilepsy as a network disease, which has led to an increasing interest in neuroimaging techniques that allow for studying in vivo neuronal networks. However, there is still a need for more research investigating the default mode network (DMN) effective connectivity in patients with refractory frontal and temporal lobe epilepsies (FLE and TLE) as compared to normal controls, by means of functional magnetic resonance imaging (fMRI). Therefore, the first step of this study was to develop appropriated protocols for data analysis, having in mind different hypotheses about neuronal connectivity to be quantified in six distinct groups. The sample comprised: 15 normal controls; 23 patients with FLE related to unilateral frontal focal cortical dysplasia (FCD), 10 right- and 13 left-sided; 21 patients with mesial TLE (MTLE) associated with unilateral hippocampal sclerosis (HS), 11 right- and 10 left-sided; and 9 patients with TLE related to unilateral left temporal FCD, in the absence of HS. All subjects underwent structural and fMRI with resting-state (RS) paradigm. The effective connectivity analyses were performed using the spectral dynamic causal modeling (spDCM) and considering four DMN nodes: the ventromedial prefrontal cortex (vMPFC), the right/left inferior parietal cortex (RIPC/LIPC) and the posterior cingulate cortex (PCC). Soon afterwards, group comparisons were made by means of the parametric empirical Bayes (PEB) model. The data analysis of the four nodes revealed positive bidirectional effective connectivity between: (1) LIPC and RIPC in the six groups; and (2) vMPFC and PCC in control, right-FLE, left-FLE, right-MTLE and left-TLE. Positive unidirectional connectivity was found from: (1) LIPC to vMPFC in in all groups; (2) LIPC to PCC in control, right-FLE, left-FLE, right-MTLE and left-MTLE; and (3) PCC to vMPFC only in left-MTLE. A single negative connection was observed from PCC to RIPC in right-MTLE group. In general lines, the protocols developed for data analysis revealed rs-DMN effective connectivity preponderance in left cerebral hemisphere, which is dominant for language in most people, including the four investigated areas listed in the current literature, as well as some relevant modifications in both left-MTLE and left-TLE groups. Based on that, it will be possible not only to add more regions of interest to the basic model but also to apply the same final protocols for analyzing the subsequently acquired cognitive data.

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Pseudo-INO in Steinert myotonic dystrophy: a case series

Pedro Lucas Grangeiro de Sá Barreto Lima[1], Antônio Edvan Camelo-Filho[1], Pedro Helder de Oliveira Junior[1], Rebeca Bessa Maurício[2], Mariana Michiles Santos Ramos[1], Jorge Luiz de Brito de Souza[2], Julia Viana de Castro[1], Avelino Missialdes Dutra Junior[1], Pedro Braga-Neto[1], Paulo Ribeiro Nóbrega[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Pedro Lucas Grangeiro de Sá Barreto Lima; Email: pedro.lucas@alu.ufc.br.

Background: Myotonic Dystrophy Type 1 (Steinert Myotonic Dystrophy or DM1) is the most frequent muscular dystrophy in adults. This disorder is a monoallelic autosomal disorder caused by pathogenic expansion of CTG trinucleotides in DMPK. DM1 is associated with weakness, muscular atrophy and myotonia. These clinical manifestations may vary from mild to severe. The disease often causes ocular impairment, mostly in the form of posterior subcapsular cataracts, ptosis and slow saccades. The current literature supports the hypothesis of relative sparring of the extrinsic ocular musculature in DM1. However, most recently ophthalmoparesis in DM1 has been increasingly reported in the literature. **Objective:** To report a series of patients with DM1 presenting with ocular mobility impairment. **Methods:** This was a retrospective observational study including patients with DM1. Patients were identified through an active search in the medical records of the neurology department of our service. **Results:** Three unrelated patients with previous diagnosis of Myotonic Dystrophy Type 1 (Steinert's disease or DM1) presented with progressive impairment of ocular mobility associated with ptosis. All patients had medical history marked by progressive muscular weakness, diffuse myalgia, frequent falls, postural instability and posterior cataracts compatible with the diagnosis of DM1. They had the genetic confirmation of the diagnosis by the study of were tested for CTG expansion in DMPK Southern Blot analysis. Upon general inspection, the patients had frontal alopecia, "hatchet" facies and bilateral ptosis. Neurological examination revealed diffuse muscular hypotrophy, diffuse muscular weakness, global hyporeflexia and myotonic phenomenon in both hands. No fasciculations or tremors were noticed in these patients. The three patients presented exotropia and bilateral ophthalmoparesis for the 9 cardinal positions of gaze. The ophthalmoparesis was more markedly observed during ocular abduction, impairing the gaze convergence. This specific pattern of ocular paresis resembled an internuclear ophthalmoplegia (INO), with the absence of nystagmus in the abducted eye during the pursuit test. Interestingly, this pattern of impairment suggests a peripherical-muscular cause. We defined this combination of findings as pseudo-INO, as it is similar to an "incomplete" internuclear ophthalmoplegia. This pattern of ophthalmoparesis is a rare manifestation of DM1 and is reported consistently in 3 patients in literature to

the best of our knowledge. Those 3 patients in the literature all had very similar presentations compared to ours. **Conclusion:** We presented a series of three patients with Steinert's disease presenting with an uncommon pattern of ocular impairment. This series provides insights in disease manifestations and may help to identify future DM1 cases. Further studies are necessary to better comprehend the observed phenomenon and to characterize the ocular findings in DM1.

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Pseudotumoral lesion mimicking glioblastoma in a patient with multiple sclerosis

Luana Gomes Dias Pimentel[1], Francyane Peixoto Ramos de Abreu[1], Marina Papais Alvarenga[1], Regina Maria Papais Alvarenga[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.

Correspondence: Luana Gomes Dias Pimentel; Email: luanagpimentel@hotmail.com.

Case presentation: On December 3rd, 2018, a 62-year-old white female doctor presented with altered sensitivity in her right hand, which progressed to incoordination of the ipsilateral upper and lower limbs due to proprioceptive alteration. She was admitted to a private hospital in Rio de Janeiro, and between December 4th and 6th she underwent serial MRI scans of the skull with spectroscopy, showing a nodular lesion with parietal contrast enhancement, measuring 3.0x2.3x2.4 cm, in the left parietal region, with extensive perilesional edema, suggestive of glioblastoma, and foci of hypersignal in T2 and FLAIR in the left anterior commissure. A CT scan of the chest and abdomen was carried out to rule out another primary site of neoplasia or metastases, which revealed no alterations. On December 7th, she underwent microsurgery with partial resection of the lesion. Post-operatively, she underwent an MRI scan which showed persistence of the foci of hypersignal. She also had weakness in the right dimidium and incomplete Gerstmann's syndrome with right-left confusion and dyscalculia. After discharge, she developed visual clouding and a CSF test was carried out, which was negative for infections and oligoclonal bands. According to the brain biopsy histopathology report, a demyelinating lesion was suggested, and corticosteroid therapy was started. In September, 2019, she went to a neurologist specialized in demyelinating diseases, who indicated prevention with Rituximab. In the week scheduled for the infusion, she experienced vertigo, and the neurological examination confirmed cerebellar dysfunction. A new MRI was requested which showed recent demyelinating lesions in T2 and FLAIR. In November, she underwent pulse therapy with methylprednisolone and the first infusion of Rituximab, showing clinical and radiological improvement. She continued to be treated every six months, and in 2022, switched to Ocrelizumab, and since then she has had no recurrences. **Discussion:** The pseudotumoral lesion is a rare form of presentation of demyelinating diseases, with a prevalence of 1-3 per 1000 cases of multiple sclerosis. It has an acute evolution and is usually solitary, larger than 2 cm, hyperintense, in which the clinical picture and MRI pattern mimic CNS neoplasms. In most cases, the pseudotumoral lesion manifests in patients previously diagnosed with MS. In this case, the tumefactive lesion was the first presentation of the disease. Diagnosis was confirmed by histopathology. The suspicion of MS was raised by the T2 and FLAIR foci identified from the first MRI scans, and concluded after a new neurological outbreak. **Final comments:** In this report, the manifestation of the pseudotumoral demyelinating lesion before the diagnosis of MS, which is rare, and the MRI report suggestive of glioblastoma led to diagnostic confusion, in which unnecessary surgery caused neurological sequelae, thus highlighting the need for further investigation before defining surgery.

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Pseudotumoral manifestation of primary central nervous system vasculitis: a case report

Maria Clara Motta Barbosa Valente[1], Camila Lopes Figueiredo[1], Filipe Brito Ferraz da Silveira[1], Cícero Rodrigues Veloso[1], Vitor Colpo Paes[1], Angelo Chelotti Duarte[1], Carmen Lucia Penteado Lancellotti[1], Antonio José da Rocha[1], Vivian Dias Baptista Gagliardi[1], Rubens José Gagliardi[1]

[1]Santa Casa de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Clara Motta Barbosa Valente; Email: mariaclarambvalente@gmail.com.

Case presentation: A 34-year-old male patient was admitted to the emergency department with recurrent generalized tonic-clonic seizures. The initial neurological examination was unremarkable. A cranial CT scan revealed a frontal right expansive lesion of undetermined etiology, associated with significant vasogenic edema and midline shift. Further investigation with brain MRI showed extensive involvement of the right hemisphere, with focal periventricular lesions with ring enhancement on gadolinium, as well as vasogenic edema. The perivascular enhancement and calcifications favored the diagnosis of an inflammatory vascular process, particularly inflammatory vasculopathy (primary vasculitis) with a pseudotumoral pattern. Laboratory tests were normal and serologies were negative. Lumbar puncture was not performed due to significant midline shift. A biopsy of the lesion was performed, and pathological evaluation revealed extensive necrosis with perivasculitis, no evidence of neoplasia in the material studied was observed. Ziehl-Neelsen (BAAR) and fungal (PAS and GROCOTT) stains were negative. Immunohistochemistry (with antigen testing for toxoplasma, BAAR, BCG, CMV, CD3, and CD68) revealed CD3 immunoexpression, demonstrating perivascular and scattered CD3-positive T-lymphocytes, along with abundant CD68-positive macrophages. Considering the observed inflammatory response, immunocompromise was questioned. Further investigation ruled out other underlying diseases - especially infectious and immune diseases - allowing treatment with steroids. **Discussion:** Primary central nervous system (CNS) vasculitis is a rare inflammatory disease involving small and medium-sized vessels. The reported case demonstrates one of the rare forms of manifestation of CNS vasculitis. The pseudotumoral form presents a diagnostic challenge as it allows for the differential diagnosis of a series of other expansive processes, including neoplastic ones. Early diagnosis is essential for treatment initiation, as early initiation implies better outcomes given the severity of the condition and the progression to death if untreated. In this context, the importance of biopsy and pathological evaluation for definitive diagnosis is emphasized. **Final comments:** Primary central nervous system vasculitis has a variable clinical presentation, and among the imaging findings, the pseudotumoral form presents a diagnostic challenge as it can be interpreted as other expansive lesions, including neoplastic ones. In these cases, biopsy plays an essential role in definitive diagnosis and should be performed to allow for prompt initiation of appropriate treatment.

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Psychiatric comorbidity in Moyamoya disease: a case presentation

Ana Luísa Castelo Branco Gomes[1], Gilmar Leite Pessoa Filho[1], Davi Telécio Firmino[2], Rafael Gonçalves Duarte Cunha[1], Luciano Gonçalves do Nascimento Júnior[1], Felipe Nunes de Araújo[1], Juliana Magalhães Leite[1], Francisco Anderson de Sá Carvalho[1], Paulo Antonio Farias Lucena[1], Rafael de Souza Andrade[1]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[2]Universidade Federal da Paraíba, João Pessoa PB, Brazil.

Correspondence: Ana Luísa Castelo Branco Gomes; Email: analuisa_gomes@hotmail.com.

Case presentation: F.S, 26-year-old female, in late puerperium, with a previous diagnosis of hypertension, presented with disorientation in time-space, weakness in the left hand, psychomotor agitation and recent memory deficit four days prior to the admission. She has a history of ischemic stroke 9 months ago. The neurological examination revealed impaired comprehension and short-term memory loss, along with left hemiparesis. Brain MRI revealed a vascular pattern compatible with Moyamoya disease (MMD), as well as bilateral chronic ischemic lesions in the frontal lobe and temporal lobe on the right. Cerebral angiography revealed bilateral carotid occlusion in its supraclinoid portion, confirming the diagnosis of MMD. Laboratory results for investigation of lupus and antiphospholipid syndrome without abnormalities, as well as electrocardiogram and echocardiogram. Due to the clinical picture of personality disorder, the patient was monitored by psychiatry during hospitalization. She was treated with anticonvulsant, antipsychotics, antiplatelet drugs and was discharged to follow-up in the neurology and psychiatry outpatient clinics. **Discussion:** MMD is a chronic occlusive cerebrovascular disease. It is characterized by endothelial hyperplasia and fibrosis of the intracranial portion of the carotid artery and its branches, leading to progressive stenosis and cerebral hypoperfusion, causing multiple and recurrent ischemic events. The chronic hypoperfusion to the brain induces the formation of collateral vessels that can rupture and cause intracranial hemorrhage. It can occur at any age, but is most common in children aged 5 to 10 and adults aged 25 to 50. The initial manifestations include transient ischemic attack, ischemic stroke, hemorrhage, headache, seizures, cognitive impairment, and movement disorders. The diagnosis relies on both anatomical and functional aspects, with angiography serving as the gold standard for anatomically assessing the disease. The cornerstones of the clinical approach to MMD are prophylactic and generic symptomatic treatment, such as the use of antiplatelet drugs, anticonvulsant drugs, and pain management, Surgical revascularization is the treatment of choice in symptomatic patients. Several studies have linked MMD with psychiatric sequelae; however, no controlled trials have definitively established causality or investigated the effectiveness of specific treatments for managing associated psychiatric symptoms. **Final comments:** In the present case, we report a patient with MMD and psychiatric comorbidity, a condition that might result from the progression of the disease. Therefore, as it is a complication that may have a negative impact on quality of life, disability, and prognosis, a team-based multidisciplinary approach is highly recommended in the MMD, with collaboration between psychiatric and neurology early diagnosis.

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Psychiatric symptoms and cognitive impairment in patients with idiopathic cervical dystonia

Denise Maria Cury Meneses Portela[1], Sarah Texeira Camargo[2], Francisco Eduardo Costa Cardoso[2], Gustavo Sousa Noleto[3], Luana Larisse de Sousa Araújo[4], Felipe Augusto de Oliveira Dantas[5]

[1]Centro Universitário UniNovafapi,Teresina PI, Brazil.
[2]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.
[3]Universidade Federal do Piauí, Hospital Universitário,Teresina PI, Brazil.
[4]Universidade Estadual do Piauí, Teresina PI, Brazil.
[5]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Luana Larisse de Sousa Araújo; Email: luanalarissedesaraujo@aluno.uespi.br.

Background: Dystonia is defined as a movement disorder characterized by sustained or intermittent muscle contractions, which cause abnormal and often repetitive movements and postures. Despite the definition exclusively related to motor aspects, recent studies show non-motor manifestations in patients diagnosed with dystonia. There has been great emphasis on aspects related to psychiatric disorders, cognitive impairment, pain, sleep and abnormalities in sensory perception and their importance as predictors of quality of life. **Objective:** Investigation of psychiatric symptoms and cognitive impairment in patients with idiopathic cervical dystonia in a tertiary center. **Methods:** Cross-sectional, descriptive and analytical study including 31 patients diagnosed with isolated idiopathic cervical dystonia and 31 healthy control subjects matched by sex, age and education. The sample was evaluated by standardized scales and interviews to obtain data on psychiatric symptoms and cognitive disorders. The instruments used were Mental State Mini Exam, Brief Cognitive Test Battery, Digit symbol substitution test, Hayling Test, Facial Emotion Identification Test, Frontal Evaluation Test Battery, Mini International Neuropsychiatric Interview, Yale Brown Obsessive Compulsive Scale and Toronto Western Spasmodic Torticollis Rating Scale. **Results:** The psychiatric symptoms assessed in this study showed statistically significant differences in the prevalence of current depressive episode (38.7% and p=0.020) and depressive episode with melancholic symptoms (32.3% and p=0.003) among idiopathic cervical dystonia patients compared to the prevalence of 12.7% and 3.2% respectively among control group participants. Regarding cognitive aspects, no statistically significant difference was found in this study for results of tests applied to assess cognitive domains. A statistically significant difference was observed between patients with cervical and idiopathic dystonia and healthy control subjects to identify anger and neutral emotions (p<0.05) on the emotion recognition test. **Conclusion:** Our study showed no difference between cervical dystonia and control subjects in tests of visual and verbal episodic memory, attention, visuo-constructive skills and executive function. However, both the higher prevalence of depression and the facial emotion recognition test were capable of distinguishing patients with cervical dystonia in this study.

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Psychiatric-onset multiple sclerosis: case report

Daniel Lacerda da Costa[1], Rafael Augusto Rosalem[1], Gabriela Abrahão Allioni[1], Guilherme Diogo Silva[1], Samira Luisa dos Apostolos Pereira[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Daniel Lacerda da Costa; Email: dlacerdadacosta@gmail.com.

Case presentation: H.V.P, 31 years old, has had a history of epilepsy since he was 4 months old. When he was 4 months old, he began to have afebrile seizures controlled with phenitoin. At age 10, he had his first psychotic break. In 2010, he was diagnosed with bipolar affective disorder and started on lithium and carbamazepine. In 2016, after a suicide attempt with the medications he was already taking, he developed severe apathy, affective dullness, progressive loss of functionality, in addition to worsening gait. He was admitted to our institution in April 2022, after presenting with acute strabismus. On neurological examination Vigilance with impaired attention, executive dysfunction with echolalia and echopraxia. He presented significant rigidity and waxy flexibility, with resting tremors in the upper limbs, in addition to internuclear ophthalmoplegia on the right. On MRI, he presented global atrophy, with a capturing lesion in the territory of the medial longitudinal fasciculus on the right. He was admitted with suspicion of a demyelizing event and catatonia. He underwent treatment with lorazepam, risperidone and electroconvulsive therapy due to refractoriness. We chose to undergo pulse therapy with corticosteroids and natalizumab for hospital discharge. **Discussion:** 95% of people with multiple sclerosis (MS) have some psychiatric comorbidity. These include depression, anxiety and psychosis. Bipolar affective disorder is twice as common as in the population without MS. The prevalence of "psychiatric-onset MS" was initially studied by Lyoo et al, when they assessed MS in a psychiatric population who met criteria for MS by T2 MRI. Other studies have come out and seen an average of 1% of the population with MS. There are reports of up to 9% of cases in study by Skegg of MS with psychiatric onset. Berna et al, evaluated that patients who presented relapses with purely psychiatric symptoms, had a delay in diagnosis of 7 years. Identifying warning signs are important to increase the suspicion of MS in psychiatric patients, such as: onset of the disorder at an atypical age, family history of MS, absence of psychiatric illness in family members, neurological evaluation in search of neurological signs, absence or minimal response to standard therapy and cognitive decline. All of these signs were described in the patient, and the need for imaging is important for diagnosis. Other signs that point to an already chronic condition are the significant atrophy in the patient's image, in addition to the loss of functionality upon arrival at the hospital. **Final comments:** MS with psychiatric onset is a rare entity and difficult to diagnose. It is known that delayed diagnosis of multiple sclerosis leads to a worsening of the prognosis. The recognition of alarm signals and the importance of neurological examination are relevant keys to organic diagnosis in an endogenous context.

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Psychosocial impact of narcolepsy on the health-related quality of life of late-diagnosed patients: a systematic literature review of the last 10 years

Nicoli Lindissey Zuim[1], Anna Júlia Silveira Freitas[1], Tamiris Romão Pereira da Costa[1], Anna Beatriz Popi e Souza[1], Clara Molinari Ferraresi Lopes[1], Giovanna do Nascimento[1], Fabrício de Mira Vieira[1], Ana Julia Fragoso Dias Rodrigues[1], Ana Beatriz Melo de Andrade[1], Thais Aiala Silva[1]

[1]Universidade Nove de Julho, Bauru SP, Brazil.

Correspondence: Nicoli Lindissey Zuim; Email: nicolind14@gmail.com.

Background: Narcolepsy is a chronic and rare neurodegenerative disorder characterized by excessive daytime sleepiness and REM sleep dissociation manifestations, such as: cataplexy, sleep paralysis, hypnagogic hallucinations, and early REM sleep. With an incidence of 25-50 patients per 100,000 inhabitants, the disease is commonly associated with psychological and social dysfunctions, leading to considerable damage to daytime life. The impact of this disorder is reflected in the impairment of health-related quality of life, which results in occupational and academic difficulties. **Objective:** To evaluate the impact of late diagnosis of narcolepsy on psychological and social aspects of health-related quality of life. **Methods:** Articles containing the descriptors "Narcolepsy, Quality-Adjusted Life Expectancy, Delayed Diagnosis" were searched through the following databases: PUBMED, LILACS, and GOOGLE SCHOLAR. Inclusion criteria: articles in Portuguese and English, published between 2013 and 2023, addressing the theme proposed for this research, and available in full. After analysis, of the 37 selected articles, 16 were retained for reading and data collection. **Results:** Data analysis revealed the following statements: a) the symptoms type 1 narcolepsy pentad (especially excessive daytime sleepiness, cataplexy, and interrupted nocturnal sleep) have a critical impact on health-related quality of life, both in self-care and in habitual activities; b) the functional impairment of the disease directly implies an increase in pain and discomfort, as well as anxiety and depression; c) health-related quality of life is affected due to the constancy of sleepiness and the unpredictability of symptoms, with greater impairment in underdiagnosed cases; d) patients with late-diagnosed narcolepsy have greater impairment in physical, emotional, cognitive, and social health compared to patients with early diagnosis and follow-up. Data collection included 303 people interviewed. **Conclusion:** The results suggest that there is a substantial impairment of health-related quality of life in patients with narcolepsy, especially those with late diagnosis. Important components of the individual's social life, such as physical and mental well-being, are persistently compromised due to excessive daytime sleepiness, cataplexy, and interrupted nocturnal sleep, characteristics more common to type I narcolepsy. In the meantime, the patient presents greater episodes of pain and discomfort, as well as psychiatric disorders, such as anxiety and depression, due to functional impairment and impact on quality of life. In addition, there is a negative public perception of the disease due to constant sleepiness and the unpredictability of the associated symptoms. Knowing that late diagnosis leads to greater impairment in the individual's physical, emotional, cognitive, and social health, it is essential to make it as skillfully as possible.

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Puerperium as a risk factor for immunocompetent patients with neurotuberculosis?

Mariana Tonon Giovannone[1], Fernando Pierini Costa[1], William Thiago Almeida Leite[1], Mauro Gomes de Araújo[1], Evelyn Chevitarese Nastari Saia[1], Simone Pincer[1], Maria Leíza Vinhadelli Ribeiro[1], Jackson Batista da Silva[1], Natasha Cutolo[1], Eduardo Nogueira[1],

[1]Santa Casa de Santos, Santos SP, Brazil.

Correspondence: Mariana Tonon Giovannone; Email: mariana.tonon@yahoo.com.

Case presentation: A 33 year old female patient, postpartum, without comorbidities, started in December 2023 with a 3 week history of weight loss, progressing to encephalopathic syndrome, decreased level of consciousness, and intracranial hypertension syndrome. Initially treated at the Emergency Care Unit, a cranial CT scan was performed, showing areas of nonspecific frontal hypodensity on the left and cerebellar hypodensity on the right, associated with vasogenic edema. During hospitalization, laboratory and imaging tests were performed, including cerebrospinal fluid collection with a positive GeneXpert® result, diagnosed with Neurotuberculosis, in addition to serology collection, all negative. The patient had an unfavorable evolution due to extensive vasogenic edema, requiring EVD due to intracranial hypertension, remaining in the ICU for approximately 30 days. Treatment was initiated with rifampicin, isoniazid, pyrazinamide and ethambutol, corticosteroid therapy and anti-seizure medication, in addition to antibiotic therapy for sepsis due to VAP and hemodynamic control with vasoactive drugs. During clinical stabilization, a cranial MRI was performed, showing nodular leptomeningeal thickening in the frontal and base of the skull and tuberculoma nodules in the right cerebellum and in the left MCA. After starting treatment with the RIPE regimen, the patient's neurological condition improved and he was discharged after 3 months of hospitalization. **Discussion:** Tuberculosis is an infectious disease caused by Mycobacterium tuberculosis and its most severe extrapulmonary form is Neurotuberculosis. Its occurrence increases in patients with the HIV, making it a rare diagnosis in immunocompetent individuals. The infection is acquired by inhalation of bacilli, which produce granulomas in the CNS, which may become inactive;

The decrease in immunity causes its rupture, forming a serofibrinous exudate containing blood vessels and areas of caseous necrosis. The symptoms are not specific and may present with headache, irritability and anorexia. The diagnosis is made through clinical, laboratory and imaging evaluation, with GeneXpert® being one of the most sensitive and specific for diagnosis. Its treatment is based on 2 months of RIPE, which can be extended to 18 months. The puerperium is the period from six to eight weeks after childbirth, when the woman suffers from significant immunological changes; studies show that despite the increase in interleukins, promoting a pro-inflammatory state and decreased immunity, there is no increase in the occurrence of Neurotuberculosis during this period. **Final comments:** The case shows that, although rare, Neurotuberculosis in immunocompetent patients is a disease present in our clinical practice. Early diagnosis helps the patient's progress. Furthermore, there is still no evidence that the postpartum period increases the risk of contamination.

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Pulmonary arteriovenous microfistulas as an uncommon cause of stroke in a young adult

Vinícius Canhestro Roedel[1], Maíra Cardoso Aspahan[1], Rachel Laguardia Rego[1], Aretha Miranda Silva[1], Bárbara Cristina Silveira de Oliveira[1], Verônica Moreira Belo[1], Samuel Inácio Oliveira Araújo[1], Larissa Cruvinel Leite[1], Arthur Braga Pereira[1], Paolla Giovanna Rossito de Magalhaes[1]

[1]Hospital Madre Teresa, Belo Horizonte MG, Brazil.

Correspondence: Vinícius Canhestro Roedel; Email: roedelvinicius@gmail.com.

Case presentation: A 24-year-old man with a history of focal epilepsy and mild cognitive impairment due to a childhood traumatic brain injury experienced sudden weakness and numbness in his fourth and fifth fingers of the right hand. Over the following days, he developed right hemibody paresthesia and abnormal movements in his right upper limb, which were challenging to precisely characterize since they weren't seen by the staff. Upon examination, mild weakness was noted in his right upper limb, and a brain computed tomography revealed a new area of hipodensity in the left postcentral gyrus. Considering his history of focal seizures originating in the same limb and the lack of traditional stroke risk factors, focal status epilepticus was suspected. A phenytoin attack was prescribed, with no effect on symptoms. Subsequent electroencephalogram and angiotomography scans were inconclusive. A brain magnetic resonance imaging then indicated restricted water diffusion in the left postcentral gyrus. Transesophageal echocardiogram revealed a right-to-left shunt with microbubbles crossing, though no patent foramen ovale was found. Doppler ultrasound of the lower limbs showed no signs of thrombosis. Cardiac arteriography uncovered a high-flow right-to-left shunt in the left lung, but no correctable fistulas were visualized. A diagnosis of paradoxical embolism due to pulmonary arteriovenous microfistulas was considered. The patient was discharged with instructions to take aspirin and undergo rehabilitation physiotherapy. **Discussion:** Pulmonary arteriovenous fistulas are vascular abnormalities, both congenital and acquired, that can lead to various neurological issues such as strokes, seizures, and migraines. They are frequently linked with hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber disease), which is characterized by recurrent bleeding and paradoxical brain emboli. While most reported cases of symptomatic fistulas involve patients with the syndrome, our patient did not present with it. Some patients may benefit from embolization therapy, a course of action not feasible in this case due to the size of the fistula, which was not large enough to be visualized. As a stroke prophylaxis measure, we opted for the use of single antiplatelet therapy, though more research is needed to determine the optimal antithrombotic treatment in these cases. **Final comments:** Pulmonary arteriovenous fistulas are a rarely considered cause of stroke in young individuals and should be part of the differential diagnosis for neurologists investigating stroke etiology. Right cardiac arteriography remains the gold standard for identifying this condition. Best practices for managing patients with microfistulas are not yet defined, underscoring the need for additional research to establish optimal antithrombotic regimens for secondary prophylaxis

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Pure ataxia related to CACNA1A: report of a non-episodic and non-migrainous case

Lucas do Rego Barros Correa Martins[1], Laura A. M. Montouro[1], Maria da Penha Morita[1], Érica Nogueira Coelho[1], Fábio de Nazaré Oliveira[1], Charles Marques Lourenço[1], Humberto Ramos Crispim[1], Matheus Hisamitsu Facine[1], Caique Alberto Dosualdo[1], Eduardo de Paula Estephan[1,2]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Lucas do Rego Barros Correa Martins; Email: lucasrbcmartins@gmail.com.

CACNA1A gene has long been a focus of expert studies due to its clinical relevance. The range of phenotypic changes that emerge from mutations on that gene usually correlates with the abnormal functioning of the voltage-dependent Ca^2+ channels. This entails neurologic syndromes such as epilepsy, migraine and different forms of ataxia. This case presents a rare association between a mutation in CACNA1A and pure congenital non progressive ataxia, reinforcing the vastness of clinical features that can be expressed by mutations in this gene. **Case presentation:** A 37-year-old woman, with persistent motor difficulties since childhood due to notable imbalance, such as the inability to ride a bicycle and climb stairs unaided. Delay in motor development was reported, having delayed ability to crawl and walk. No notable headaches or abrupt worsening of coordination during her life was reported. On physical examination, the patient exhibited dysarthric speech, as well as a difficulty to perform the Tandem Walk test. There were no signs of pyramidal tract lesions and no dysdiadochokinesia. Her daughter and one of her brothers also had a history of symptoms such as incoordination and speech alterations, respectively, suggesting a possible hereditary with autosomal dominat fashion of the disorder. On magnetic resonance imaging cerebellar vermis atrophy was revealed. Genetic panel for movement disorders identified a pathogenic variant on the CACNA1A gene: c.4034G> A (p.Arg1345Gln; chr19:13.262.789 C> T on GRCh38 genome version). **Discussion:** Pathogenic variants in CACNA1A are associated with a range of neurological phenotypes, such as developmental and epileptic encephalopathy, spinocerebellar ataxia (SCA6), episodic ataxia type 2 and familial hemiplegic migraine. More rarely, it has been linked to autosomal dominant nonprogressive congenital ataxia, in which the phenotype of our clinical report fits. Cases of congenital ataxia have been described in patients with CACNA1A missense mutations, but usually associated with migraine or epilepsy. The prognosis of such presentation is usually good, with mild/moderate symptoms throughout life. **Final comments:** This case report highlights the importance of considering CACNA1A among ataxic patients, even in the absence of episodic attacks, migraine and epilepsy. Moreover, it also illustrates the possibility of phenotype variability with the same pathogenic variant.

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Pure transverse myelitis secondary to multiple sclerosis: an unusual case

Carolina Flores Welker[1], Larissa Lucas Schloicka[1], Nícolas Philippe Balduíno Nogueira[1]

[1]Instituto Hospital de Base do Distrito Federal, Brasília DF, Brazil

Correspondence: Carolina Flores Welker; Email: carolinafwelker@gmail.com.

DAS, 37 years old, male, previously healthy, developed paresis and paresthesia in all four limbs on May 2022, one day after receiving the influenza vaccine. There was a partial improvement in symptoms over the days, but the patient still complained of spasms and paresthesias in the right upper limb, in addition to shocks along the spine during cervical flexion. In June 2022, MRI of the cervical spine without contrast showed centromedullary hypersignal on T2/STIR at C2, with a slight swelling effect. On August 2022, the lesion had larger dimensions and also captured contrast. The other neuraxial MRIs showed no changes and the infectious, neoplastic and rheumatological investigations were normal. The CSF analysis in September 2022 revealed the presence of BOC type 2 and serum levels of Anti – AQP 4 (CBA) and Anti MOG (CBA) were non-reactive on October of the same year. After discussion with the attending physician, the patient received methylprednisolone 1 gram for 5 days on November 2022. He remained asymptomatic for 20 days after the end of corticosteroid therapy and, afterwards, he presented brief episodes (lasting less than 24 hours) of paresthesias in right arm and shocks along the spine. A control imaging examination was performed in May 2023, which showed the same centromedullary lesion at the C1-C2 level, with stable dimensions compared to the examination on June 2022. Considering that the patient met the McDonald 2017 criteria for diagnosis of MS, in August 2023, the attending neurologist decided to start fingolimod and, since then, the patient has remained asymptomatic. During follow-up, a neuraxial MRI was performed on May 2024, which indicated that the centromedullary lesion at the level of C1-C2 captured contrast and that two small left lateral lesions have appeared at the level of C4 and C6, both with slight contrast enhancement. Considering the radiological progression of the lesions, the exchange of fingolimod for natalizumab was indicated. Multiple sclerosis (MS) is an immune-mediated, inflammatory, demyelinating and neurodegenerative disease that involves the white and gray matter of the central nervous system (CNS). Its etiology is not yet well understood and involves genetic and environmental factors that are not yet well defined. It predominantly affects individuals between 20 and 50 years of age; it is twice as common in females and has a lower incidence in the Afro-descendant, oriental and indigenous population. The structures most affected by MS are the optic nerve, brain stem, cerebellum and spinal cord. Transverse myelitis lesions secondary to MS are usually partial, short and eccentric. Sensory symptoms tend to be more exuberant than motor symptoms; the clinical picture is asymmetric and the evolution is usually subacute. The presence of a neurogenic bladder is also common. This report describes a case of MS with pure spinal cord involvement, which is considered uncommon.

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Pyridostigmine in the management of dropped head syndrome following botulinum toxin injection: case report

Gabriel Felipe Almeida de Barros[1], Eduardo Sousa de Melo[1], Leticia Klabinske Marques Monteiro[2], Evelliny Gomes da Silva[2], Allefy Beltrão Albano[2], Fabrício Martins Veras[2], Carla Bianca Alves Leite Santos[1], Giovanna da Silva Gamba Dias[1], Aimê de Paula Santos[1]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil
[2]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil

Correspondence: Leticia Klabinske Marques Monteiro; Email: leticiaklabinske@gmail.com.

Case presentation: A 66-year-old man, diagnosed with Parkinson's disease (PD), presented a significant anteriorization of the cephalic and dorsal regions with contractions and significant pain, in addition to a lateralization of the neck to the right. It was recognized as camptocormia, which has been frequently associated with PD. The medical team opted to administer botulinum toxin in the affected area. However, two days after the application, the patient developed Dropped Head Syndrome, a relatively rare condition. Thus, pyridostigmine was prescribed to enhance symptom management, along with physical therapy sessions. After about a month of use, the treatment proved to be effective. **Discussion:** Dropped head syndrome (DHS) is characterized by severe weakness of the cervical paraspinal muscles, resulting in gradual sagging of the head and usually culminating in the chin-on-chest deformity. It is a relatively rare condition with various differential diagnoses that can be categorized into neurological, neuromuscular, muscular, and other causes. This deformity has significant implications on the quality of life of affected individuals, resulting in considerable restrictions to daily activities. Botulinum toxin injection has been reported to produce measurable weakness at both adjacent and distant sites after injection, by systemic spread and retrograde axonal transport. However, this application can cause adverse effects, such as DHS, as in this clinical report. The management of this symptom can include physical therapy, orthotic devices (including cervical collars), and even, in severe cases, surgical intervention. For cases of DHS caused by myasthenia gravis, pyridostigmine has been used for symptom reversion. This drug is a reverse acetylcholinesterase inhibitor, which works by preventing the breakdown of acetylcholine in the neuromuscular junction. Therefore, given that the mechanism of action of both myasthenia gravis and botulinum toxin are similar, since the two work by reducing the amount of acetylcholine in the synaptic cleft, it was decided to use pyridostigmine to reverse the DHS caused by botox. The acetylcholinesterase inhibitor's mechanism is such that it may reduce the effect of BTX by potentiating neuromuscular transmission, decreasing muscle flaccidity, and thus promoting the resolution of the case. **Final comments:** Although pyridostigmine is not commonly used to reverse the head drop syndrome caused by BTX, its effects are known for DHS treatment in myasthenia gravis. Therefore, considering the similarities in mechanism of action of both myasthenia gravis and botulinum toxin and the safety and efficacy of this medication, pyridostigmine should be considered to treat severe sequelae of botulinum toxin injection in selected patients.

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Quality of life of patients with amyotrophic lateral sclerosis (ALS) and satisfaction with the care and treatment received in a specialized service for rare diseases

Maria Rodrigues Lima de Souza Pessoa[1], Ariele Amaral Alves[1], Laís Brandão Mota[1], Stefania Zingone Andrade Carvalho[1], Luna Souza Silva Vilanova[1], Luana Emanuelly Sinhori Lopes[2], Debora Borges dos Santos Pereira[2], Marilaine Menezes de Matos Ferreira[1], Marcela Camara Machado Costa[1], Têmis Maria Félix[3],

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Mapes Solutions, São Paulo SP, Brazil.
[3]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Maria Rodrigues Lima de Souza Pessoa; Email: mariapessoa20.1@bahiana.edu.br.

Background: The JAV-Raras study is an initiative integrated in the National Network of Rare Diseases project and aims to measure the Journey of Value (JAV) of patients with rare diseases in Brazil. Among the diseases under analysis, there is Amyotrophic Lateral Sclerosis (ALS). **Objective:** To understand the quality of life of patients diagnosed with ALS and to evaluate their satisfaction with the care and treatment received at a specialized in Rare Diseases service. **Methods:** Data was collected from medical records and clinical and quality-of-life questionnaires were applied. The EUROQOL questionnaire was used to assess quality of life, which consists of the EQ-5D descriptive system and the EQ-VAS scale. The EQ-5D is a multiple-choice questionnaire that assesses health-related quality of life in five dimensions: mobility, self-care, daily activities, pain/discomfort and anxiety/depression. The EQ-VAS uses a visual analog scale ranging from 0 to 100 to measure quality of life. In addition, the Net Promoter Score (NPS) was used as an indicator of patient satisfaction with the care team and treatment received. **Results:** The sample consisted of 30 patients followed up in a specialized service, 40% female and 60% male. The quality of life results assessed by the EQ-5D-3L show that the median score is 0.238, with a minimum score of -0.131 and a maximum score of 0.47. Regarding satisfaction with the care team: 50% of patients rated the team in the "excellent zone", which indicates a high level of satisfaction. However, 26.7% rated the team in the "critical zone" or "improvement zone", suggesting that there are areas where the quality of care can be improved. With regard to satisfaction with the treatment: 60% rated the treatment as excellent, which shows a high level of satisfaction with the effectiveness and quality of the treatment received. **Conclusion:** The median EQ-5D-3L score indicates a low health-related quality of life, suggesting that, in general, these patients have significant problems in the multiple dimensions assessed, such as mobility, personal care, usual activities, pain/discomfort and anxiety/depression. This low score reflects the negative impact that ALS has on the quality of life of these individuals. These results reinforce the necessity for a multidisciplinary approach to the management of the disease, aimed not only at clinical treatment, but also at improving the quality of life of these patients. With regard to the NPS, the results indicate that, in general, patients have a good level of satisfaction with both the care team and the treatment received.

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Quality of life of patients with ATTRV amyloidosis and satisfaction with the care and treatment received in a specialized rare disease service

Maria Rodrigues Lima de Souza Pessoa[1], Ariele Amaral Alves[1], Marie Lidio dos Santos Galvão Ribeiro[1], Stefania Zingone Andrade Carvalho[1], Luna Souza Silva Vilanova[1], Luana Emanuelly Sinhori Lopes[2], Thiago Godoy de Oliveira[2], Marilaine Menezes de Matos Ferreira[1], Marcela Camara Machado Costa[1], Têmis Maria Félix[3]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Mapes Solutions, São Paulo SP, Brazil.
[3]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Maria Rodrigues Lima de Souza Pessoa; Email: mariapessoa20.1@bahiana.edu.br.

Background: The JAV-Raras study is an initiative integrated in the National Network of Rare Diseases project and aims to measure the Journey of Value (JAV) of patients with rare diseases in Brazil. Among them is the transthyretin-related familial amyloidosis (ATTRv). **Objective:** To measure the quality of life of patients diagnosed with ATTRv amyloidosis and to assess their satisfaction with the care and treatment received in a specialized service. **Methods:** Data was collected from medical records, clinical and quality-of-life questionnaires. The EUROQOL questionnaire was used to assess quality of life. This instrument consists of the EQ-5D descriptive system and the EQ-VAS scale. The EQ-5D is a multiple-choice questionnaire that assesses health-related quality of life in five dimensions: mobility, self-care, activities of daily living, pain/discomfort, and anxiety/depression. The EQ-VAS consists of a visual analog scale with a score from 0 to 100 to assess quality of life. In addition, the Net Promoter Score (NPS) is an indicator of patient satisfaction with the care team and treatment. **Results:** 33 patients were included, 46% female and 54% male. All responded to the Quality-of-Life Questionnaire and the results assessed by the EQ-5D-3L show that the median score is 0.582, with a minimum value of -0.131 and a maximum value of 1. Of this sample, 32 patients responded to the questionnaire assessing satisfaction with the care team: 40.6% rated the team in the "critical zone", meaning a high level of dissatisfaction. Another 40.6% rated the team in the "zone of excellence" indicating satisfaction with the care received. Only 18.8% rated the team in the intermediate zones of "improvement" and "quality". In terms of satisfaction with care, almost all of them (93.7%), rated the care received in the "excellence" zone, indicating a high level of satisfaction with the effectiveness and quality of care. Only 6.7% rated it in the "quality zone", with no negative evaluations. **Conclusion:** The median score of 0.582 on the EQ-5D indicates that the sample's quality of life is significantly reduced compared to the quality of life of those outides of the sample. This is probably due to symptoms, functional limitations, disability and reduced general well-being. This highlights the need to implement care strategies that include more effective therapeutic approaches, psychosocial support and measures to minimize the impact on quality of life. Regarding the NPS, data suggest that there is dissatisfaction with the care team on the part of patients, although the same percentage is very satisfied with the care. This points to the need for improvements in communication, empathy and quality of care provided by the healthcare professionals involved.

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Quantifying ataxia in RFC1-related disorder using wearable sensor technology

Camila Caroso Lobo[1], Isabella Colombani[1], Nadson Bruno Serra Santos[1], Rachel Paes Guimarães[1], Fabrício Diniz Lima[1], Paula Camila Alves de Assis Pereira Matos[2], Alberto Rolim Muro Martinez[1], José Luiz Pedroso[2], Orlando Graziani Povoas Barsottini[2], Marcondes Cavalcante França Jr[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Camila Caroso Lobo; Email: camilaloboc@gmail.com.

Background: RFC1-related ataxia is a neurological disorder characterized by progressive impairment of motor coordination, affecting standing balance and gait. Quantitative assessment of ataxia-specific gait impairments using wearable technology holds promise for providing sensitive outcome measures. However, the application of motion sensors to evaluate these impairments in patients with RFC1-related ataxia remains unexplored. **Objective:** This study aimed to identify specific standing balance conditions and digital postural sway measures that differentiate between patients with RFC1-related ataxia patients and healthy controls. Additionally, the study sought to evaluate the correlation between motion sensor parameters and the severity of ataxia. **Methods:** In this study, seventeen patients with RFC1-related ataxia and nine healthy controls were equipped with six inertial sensors to assess parameters, including gait during a 2-minute natural pace walking task, postural sway, turn times, and sit-to-stand times. The severity of ataxia was assessed using the Scale for the Assessment and Rating of Ataxia (SARA), administered consistently by the same

experienced neurologist across all patients. The median values of these parameters were compared between the groups using the Mann-Whitney test. Spearman's correlation coefficient was employed to examine the relationships between these measures and the severity of ataxia. **Results:** Significant differences were observed between RFC1 mutation patients and healthy controls in postural and gait parameters. Mann-Whitney tests showed higher mean postural sway velocity (p=0.030), larger sway area (angles and acceleration, p=0.030), and greater turn angle (p=0.004) in RFC1 patients. Spearman's correlations indicated significant associations between the SARA scale and various parameters: increased double support duration (R=0.634, p=0.015 left, R=0.664, p=0.010 right) and decreased turn velocity (R=-0.633, p=0.009). Strong correlations were also found between postural sway with eyes closed and feet apart, particularly mean velocity (R=0.718, p=0.003), reflecting impaired postural control in RFC1 patients. **Conclusion:** Wearable sensors can effectively differentiate motor impairments in RFC1-related ataxia and correlate well with the severity of the condition. These technologies may offer sensitive measures for clinical assessment and therapeutic strategies in ataxia management in these patients.

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Quantifying myelin loss with [11C]PIB PET and MTR: predicting cognitive and functional decline in MS

Kenia Repiso Campanholo[1], Milena Sales Pitombeira[1], Graziella Aguiar Santos Faria[1], Fabio Luis de Souza Duran[1], Samira Luisa dos Apostolos Pereira[1], Carolina Rimkus[1], Dagoberto Callegaro[1], Daniele de Paula Faria[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Graziella Aguiar Santos Faria; Email: graziasf@hotmail.com.

Background: Cognitive decline is a significant concern in Multiple Sclerosis (MS) and is closely associated with myelin loss. Various neuroimaging methods have been employed to quantify myelin and correlate it with cognitive dysfunctions. Magnetization Transfer Ratio (MTR) and Positron Emission Tomography (PET) with [11C]PIB have yielded promising results in cross-sectional studies; however, longitudinal follow-up investigations remain underexplored. **Objective:** This study aims to measure myelin content using PET and MTR and evaluate their potential in predicting cognitive decline and functional capacity over a one-year follow-up period in patients with MS (pwMS). **Methods:** Fifty-one pwMS underwent clinical and neuropsychological assessments, as well as MTR and [11C]PIB-PET imaging. After one year, 39 of these patients were re-evaluated clinically and neuropsychologically. Myelin content was measured in white matter (WM), normal-appearing white matter (NAWM), and lesions. **Results:** The clinical follow-up evaluations did not show significant differences overall, but when divided by phenotype, the Expanded Disability Status Scale (EDSS) was significantly higher one year later for progressive MS (p=0.025). The cognitive profile remained similar between both assessments. Lower MTR values were associated with lower cognitive z-scores in the follow-up evaluation. Specifically, in NAWM, lower MTR values correlated with lower scores on the Selective Reminding Test (SRT) delayed recall (r^2=0.079, p=0.042), Paced Auditory Serial Addition Test (PASAT) (r^2=0.127, p=0.019), Word List Generation (WLG) (r^2=0.076, p=0.036), and EDSS (r^2=0.792, p=0.020). Similarly, in WM, lower MTR values were associated with lower scores on the SRT delayed recall (r^2=0.088, p=0.035), PASAT (r^2=0.117, p=0.023), WLG (r^2=0.061, p=0.048), Symbol Digit Modalities Test (SDMT) (r^2=0.136, p=0.011), and EDSS (r^2=0.792, p=0.021). Lower [11C]PIB uptake was associated with lower z-scores one year later in NAWM (PASAT r^2=0.071, p=0.050) and WM (PASAT r^2=0.094, p=0.030; SDMT r^2=0.073, p=0.041). There was no significant relationship between MTR and [11C]PIB uptake in lesions and cognitive variables. **Conclusion:** Although there were no significant overall differences in cognitive and disability measures between the initial assessment and the follow-up, lower MTR and [11C]PIB uptake were associated with worse cognitive performance and higher disability scores one year later. This suggests that MTR and [11C]PIB uptake could be valuable in predicting cognitive decline and increased disability in pwMS over time.

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Quantitative study of hospital admissions for Alzheimer's disease in Brazil from 2019 to 2023

Rynele Almeida de Fonseca[1], Deborah Araujo Silva[1], Annayza de Oliveira Bessa[1], Larissa Bianca Borsone[1], Amanda Oliva Spaziani[1], João Carlos Bizinotto Leal de Lima[1], Vanessa Maria Gonçalves de Souza[1], Luis Carlos Spaziani[2], Gustavo Henrique da Silva[3], Francieli Gomes Lima[4]

[1]Universidade Brasil, Fernandópolis SP, Brazil.
[2]Fundação Educacional de Fernandópolis, Fernandópolis SP, Brazil.
[3]UPA Santa Fé do Sul, Santa Fé do Sul SP, Brazil.
[4]AME Fernandópolis, Fernandópolis SP, Brazil.

Correspondence: Rynele Almeida de Fonseca; Email: rynelef_@hotmail.com.

Background: Alzheimer's disease (AD) is a neurodegenerative disorder, an irreversible and progressive brain pathology. Etiological hypotheses suggest that genetic risk factors and environmental factors contribute to the onset of AD. It is characterized by the formation of senile plaques and neurofibrillary tangles through amyloid fibrillar deposits and hyperphosphorylation of Tau protein, which cause lesions in brain areas. The incidence of neurodegenerative diseases, including Alzheimer's Disease, increases with age, impairing memory and cognitive functions. Symptoms begin insidiously and progress, with subtle memory losses, motor weakness, and loss of ability to perform tasks, eventually leading to incapacitation. **Objective:** To evaluate the profile of hospital admissions for AD in Brazil between 2019 and 2023. **Methods:** Data collection was carried out using information available from the Hospital Information System of SUS (SIH/SUS) of the Ministry of Health, which is publicly accessible on Tabnet/DATASUS, between January 15 and May 15, 2024. This retrospective, longitudinal study utilized a quantitative, descriptive design. Data were grouped by Brazilian macroregions, type of care, sex, age group, and race/ethnicity. Statistical inference was performed using BioEstat 5.3 software, employing the paired T-test for two related samples to compare groups, with data presented through descriptive statistics, measures of central tendency, and dispersion. **Results:** During the period from 2019 to 2023, there were 7,547 hospital admissions for AD in Brazil. The majority of these admissions occurred among females, accounting for 65.76% (n=4,963) (p<0.0001), and 50.02% (n=3,775) among whites (p<0.0001). The population aged over 80 years was proportionately more affected, comprising 59.36% (n=4,480; p<0.0001) of admissions, primarily concentrated in the Southeast region (n=3,549), representing 47.03% (p=0.0005) of the total, with an average annual cost of R$ 2,634,689.11 for care. Deaths were more frequent in the Southeast, with about 58.01% (n=1,105) of cases (p=0.0006), and the average mortality rate was 50.21 (p=0.0043). The number of admissions peaked in 2023, accounting for 28.10% (n=2,121) of the total, compared to 16.06% (n=1,212) in 2020. Most admissions were classified as urgent, representing 83.32% (n=6,288; p<0.0001) of the cases. **Conclusion:** Hospital admissions for AD are concentrated in the Southeast region of Brazil, with higher proportionality of deaths and care costs during the period. The majority of cases involved females over 80 years old of white race, with urgent care being the most prevalent type of admission.

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Racemose neurocysticercosis and cerebral venous thrombosis: case report

Lucas Vieira Pinto[1], Alexandre Motta Mecê[1], Alessa de Andrade Santana[1], Matheus Oliveira Povoa[1], Júlia Domingues Gatti[1], Leonardo Grande de Almeida[1], Rafael Shiniti Yabiku[1], Kevin Yoshimi Nakaie[1], Gabriel Costa dos Reis[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Lucas Vieira Pinto; Email: lucasvip2803@gmail.com.

Case presentation: A 29-year-old female, with no significant past medical history, first presented with an altered level of consciousness preceded by vomiting, and behavioral changes. On first evaluation in her hometown, the patient obtained a computer tomography (CT) scan revealing hydrocephalus with evidence of intracranial hypertension (ICH), being referred to a tertiary service for further evaluation. Upon admission, the patient presented with a Glasgow Coma Scale of 13, being empirically treated for acute meningoencephalitis with vancomycin, acyclovir and ceftriaxone, also presenting spontaneous improvement with orthostatic posture. A magnetic resonance imaging (MRI) scan was performed, revealing signs of ICH and a nodular cystic lesion located in the fourth ventricle, partially obstructing the foramina of Magendie and Luschka bilaterally, suggestive of neurocysticercosis. The MRI also showed indirect signs of cerebral venous thrombosis of the inferior sagittal, transverse and sigmoid sinuses, later confirmed in vascular exams. The patient was anticoagulated with Rivaroxaban for 30 days, with evidence of thrombosis recanalization observed on a follow-up image exam, along with preoperative corticosteroid use. The diagnosis was confirmed through histopathological examination when the patient underwent cystectomy in the fourth ventricle, revealing Cysticercus cellulosae (with scolex and membrane). The patient is currently stable, undergoing outpatient follow-up, corticosteroid tapering, and proposed anticoagulation for 6 months. **Discussion:** Neurocysticercosis can manifest in the parenchymal form (an important cause of epilepsy) and the racemose form (obstructing the ventricular system and causing hydrocephalus), making high clinical suspicion essential. The association with cerebral venous thrombosis is, however, a rare event in the literature. It is believed that the increase in procoagulant factors secondary to the local infectious process, combined with the proximity of the venous sinuses, is primarily responsible for the thrombus formation. We highlight the case reported, as the first one reported in literature, presenting this rare association of events with a remarkable response to anticoagulation treatment, corticosteroid therapy, and surgical approach. **Final comments:** Neurocysticercosis is the most common helminthic infection of the central nervous system, accounting for 30% of epilepsy cases in endemic countries. The racemose form is a severe presentation often causing hydrocephalus and intracranial hypertension. This case report presents a favorable outcome resulting from proper treatment of the disease and its complications, supporting the significance of being vigilant towards the suspect of the disease and the possibility of neurovascular complications, which, although rare, have significant implications for clinical management.

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Radiation therapy-induced plexopathy: a case report

Ana Paula de Almeida Campos[1], Ketimila Aparecida Pereira[1], Maria Clara Motta Barbosa Valente[2], Fernando Pierini Costa[1], Mauro Gomes Araujo[1], William Thiago Almeida Leite[1], Ana Luisa Vilela Sá Fortes[1]

[1]Santa Casa de Misericórdia de Santos, Santos SP, Brazil.
[2]Santa Casa de São Paulo, São Paulo SP, Brazil.

Correspondence: Ana Paula de Almeida Campos; Email: anapac187187@gmail.com.

Case presentatio: An 88-year-old woman presented with a one-month history of ascending paresthesia, hypoesthesia, and paresis in all four limbs, more pronounced in the lower limbs. She had a history of breast cancer treated with radiotherapy (RT) and chemotherapy (CT) in 2019 and 2020 for disease recurrence. Physical examination revealed bilateral ulnar nerve hypoesthesia, symmetric weakness in all limbs, and hypoactive reflexes. Magnetic resonance imaging (MRI) of the spine and laboratory tests were normal. Electromyography (EMG) showed axonal peripheral polyneuropathy in all four limbs associated with bilateral lumbosacral plexopathy and signs of current denervation (fibrillation and positive sharp wave) at rest. Considering the clinical findings, EMG, and reported history, a diagnosis of RT-induced plexopathy was raised, although secondary involvement due to CT could not be ruled out, especially since CT is a risk factor for plexopathy. **Discussion:** This case report presents a patient who developed axonal peripheral polyneuropathy in all four limbs associated with bilateral lumbosacral plexopathy after treatment for breast cancer. These electromyographic findings can be seen in both chemotherapy-induced neuropathy and radiation-induced plexopathy, although they are more commonly secondary to radiation therapy. The clinical presentation can be early or late onset, usually more than six months after radiation therapy. The exact mechanism of injury is not well established, but it is known that there is direct damage to axons due to toxic effects, as well as microvascular damage, and that concomitant chemotherapy has a synergistic effect, increasing the risk of plexopathy. Symptoms include sensory and motor changes, as well as neuropathic pain. An important differential diagnosis is tumor recurrence or metastasis. EMG plays an essential role in assessing the severity and prognosis of the disease. Nerve conduction velocities help to detect blockages in the damaged nerve. In severe cases, EMG shows fibrillation and absence of motor unit potentials in the affected muscle group supplied by the plexus. Treatment consists of supportive therapies due to the irreversible nature of the lesion. **Conclusion:** Radiation-induced neuropathy is a rare condition that is an important differential diagnosis for tumor recurrence (depending on the site) or metastatic disease. It is also a differential diagnosis for purely axonal polyneuropathies without plexus involvement. There is no curative treatment, and supportive therapy for symptom control is of paramount importance for patient quality of life.

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Radiation-induced mesial temporal and frontobasal lesions mimicking viral encephalitis

Gustavo Lopes de Freitas Honório[1], Natália da Costa Chiote Pinheiro[2], Carolina Rouanet[1], Ingrid Pereira Marques[1], Marco Antônio Sales Dantas de Lima[1]

[1]Universidade Federal do Rio de Janeiro, Hospital Universitário Clementino Fraga Filho, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Rio de Janeiro, Instituto de Neurologia Deolindo Couto, Rio de Janeiro RJ, Brazil.

Correspondence: Gustavo Lopes de Freitas Honorio; Email: gugahonorio@hotmail.com.

Case presentation: A 53-year-old male diagnosed with peripheral non-Hodgkin T lymphoma involving the right frontal, right ethmoid and left maxillary sinuses presented with tonic-clonic seizures and altered mental status. Antiepileptic drugs (phenobarbital 200/day and levetiracetam 1000 mg/day) were started, leading to control of seizures. He was previously treated with one session of gemcitabine-oxaliplatin and one session of dexamethasone, methotrexate, ifosfamide, L-asparaginase and etoposide without response. Two months before the neurological symptoms, he underwent 20Gy local radiotherapy, divided into 10 sessions of 2Gy under intensity-modulated radiotherapy (IMRT). Facial Magnetic Resonance Imaging (MRI) showed an expansive contrast-enhancing lesion in the lower portion of the nose extending to the anterior portion of the maxilla and the palate, while cranial MRI T2/FLAIR revealed hyperintensity on cortico-subcortical in the right frontobasal, insular, medial temporal region and frontobasal left cortex, without contrast enhancement. Initially, lymphoma progression and viral encephalitis was suspected but cerebrospinal fluid analysis findings were normal, including neoplastic cells, HSV-1 and HSV-2 PCR. However, despite any specific treatment, a control brain MRI revealed reductions of the parenchymal lesions, leading to the diagnosis of radiation-induced brain injury. Although in treatment, the patient had systemic progression, being referred to palliative care. **Discussion:** Radiotherapy is widely used in the treatment of neoplasms, generating direct double-stranded DNA breakage and cell death, vasculopathy causing tissue hypoxia, and even inducing increased inflammatory activity with microglial activation. The effect on healthy brain tissue can be long-lasting, remaining for years after initial exposure. Brain injuries associated with radiation can be divided into acute, occurring within two months after radiation therapy; early-delayed, up to 6 months; and delayed, that develops 6 months after the treatment. Acute brain lesions are associated with high total doses, few fractions and high brain volume exposure to radiation. The clinical features can range from altered mental status, seizures and focal signs, as in the presented case. Additionally, when dealing with aggressive systemic lymphomas, it is important to consider progression to the central nervous system as differential diagnosis. In this case, involving an immunosuppressed patient and given the initial imaging pattern with bilateral mesial temporal and insular involvement, it was also necessary to exclude herpetic encephalitis. **Final comments:** As radiotherapy is widely used in medical practice, neurologists must be aware of the entire spectrum of clinical manifestations and the neurological complications that a patient exposed to radiotherapy may have. Moreover, in patients with aggressive cancer, it is important to consider progression of the underlying disease to the nervous system.

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RAINBOWFISH: primary efficacy and safety data in risdiplam-treated infants with presymptomatic spinal muscular atrophy (SMA)

Edmar Zanoteli[1], Richard S. Finkel[2], Michelle A. Farrar[3], Laurent Servais[4], Dmitry Vlodavets[5], Mohammad Al-muhaizea[6], Alexandra Prufer[7], Leslie Nelson[8], Carolyn Fisher[9], Birgit Jaber[10]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[2]St Jude Children's Research Hospital, Memphis, TN, USA.
[3]University of New South Wales Sydney, Sydney, Australia.
[4]University of Oxford, Oxford, UK.
[5]Pirogov Russian National Research Medical University, Moscow, Russia.
[6]Research Center-Riyadh, Riyadh, Kingdom of Saudi Arabia.
[7]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[8]University of Texas Southwestern Medical Center, Dallas, TX, USA.
[9]Roche Products Ltd, Welwyn Garden City, UK.
[10]Pharma Development, Safety, F. Hoffmann-La Roche Ltd, Basel, Switzerland.

Correspondence: Edmar Zanoteli; Email: edmar.zanoteli@usp.br.

Background: Risdiplam (EVRYSDI®) is a centrally and peripherally distributed, oral survival of motor neuron 2 (SMN2) pre mRNA splicing modifier that has been approved in more than 90 countries worldwide. **Methods:** RAINBOWFISH (NCT03779334) is an open-label, single-arm, multicenter study assessing the efficacy, safety and pharmacokinetics/pharmacodynamics (PK/PD) of risdiplam in infants with genetically diagnosed and presymptomatic spinal muscular atrophy (SMA) from birth-6 weeks of age (at first dose), regardless of SMN2 copy number. The primary endpoint is the proportion of infants, with two SMN2 copies and baseline compound muscle action potential (CMAP) amplitude ≥1.5mV, who are able to sit without support for ≥5 seconds (assessed by Item 22 of the Gross Motor Scale of the Bayley Scales of Infant and Toddler Development, third edition). Secondary endpoints include: the development of clinically manifested SMA; survival and permanent ventilation; motor milestone achievement; motor function; growth measures; nutritional status; CMAP; PK/PD; and safety monitoring. **Results:** The median age at first dose was 26.5 days (range: 16-40 days) for the first 18 enrolled infants (data cut-off: 1 July 2021). No treatment-related serious adverse events were reported in infants treated for ≤22.8 months. A preliminary analysis of seven infants treated for ≥12 months demonstrated that most reached near maximum scores on the Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders scale by 4-5 months of age and achieved motor milestones within World Health Organization windows for healthy children. All seven infants treated for ≥12 months were alive without permanent ventilation, maintained swallowing and feeding abilities, and had not required hospitalization. **Conclusion:** Here we report for the first-time results from the RAINBOWFISH primary analysis conducted at Month 12.

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Rapidly progressive and reversible dementia: an abnormal finding in MRI not to be missed for dural arteriovenous fistula diagnosis

Rafael Amaral de Matos Albuquerque[1], Marcus Vinicius Cristino Albuquerque[2], Ananda Vieira de Lima Almeida[1], Alexandre Maia Barros Drayton[2], Saulo Pimenta Lacerda[2], Renato D'Oliveira Vieira, Silas Santos Santana[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital São Rafael, Salvador BA, Brazil.

Correspondence: Marcus Vinicius Cristino Albuquerque; Email: drmarcusalbuquerque@gmail.com.

Case presentation: 72-year-old woman presented with depressive symptoms 8 months ago. After 2 months, she experienced memory impairment, language difficulties, perseverative speech, and hypokinesia. After 3 months, presented with acute worsening of speech, psychomotor slowing, drowsiness, and lower limb weakness. She was hospitalized, aphasic for 3 days. She was discharged with a presumptive diagnosis of lacunar ischemic stroke and prescribed dual antiplatelet therapy and high-dose statin. The following month, she was readmitted to another facility for further investigation of rapidly progressive dementia syndrome associated with an ictal event, initially suspected to be encephalopathy. PMH: osteogenesis

imperfecta, hypertension, dyslipidemia. On examination: normal speech and language; grade V muscle strength, normal reflexes; no movement disorders or crisis stigmata; axial facial signs present; no sensory, coordination, or cranial nerve abnormalities. MMSE: 14, MoCA: 6 points. Laboratory tests were normal (complete blood count, sodium, calcium, TSH, free T4, VDRL, vitamin B12, anti-TPO, anti-GAD). MRI showed no ischemic changes. PET scan: moderate glycolytic hypometabolism in the frontal lobes extending to the anterior cingulate, associated with slight hypometabolism of the right striatum. Thoracic CT: normal, abdominal CT: ovarian cyst; CSF: 1 cell, protein 86, no oligoclonal bands; normal EEG. She was readmitted the following month due to worsening ability to retain and recall information and temporal disorientation. MRI displayed FLAIR and DWI hypersignal in the semioval centers in the venous drainage topography. Angiography revealed a fast-flow dural fistula in the torcula region along with signs of chronic venous sinus thrombosis with cortical venous stasis and contralateral venous reflux. She underwent embolization of an extensive dural fistula in the right superior sagittal and transverse-sigmoid sinuses with an 80% reduction of the diseased dural segment. Two months after the procedure, the patient showed cognitive and radiological improvement. MoCA: 10 (previously 6). MMSE: 18 (previously 14). Cranial MRI: improvement of FLAIR hypersignal in semioval centers. **Discussion:** Dural fistulas comprise 10 to 15% of intracranial vascular malformations, with 35% located in the posterior fossa. They are more common in females and in the sixth decade of life. Symptoms are usually benign: bruit, tinnitus, headache, dizziness, conjunctival bruising, and/or orbital congestion. Occasionally, more aggressive manifestations can occur: intracranial hemorrhages, dementia, encephalopathy, epilepsy, cranial nerve deficits, intracranial hypertension, focal/transient deficits. **Final comments:** This pathology should be considered in all cases of rapidly progressive dementia and recurrent ictal events. Chronic venous sinus thrombosis with venous congestion should also be considered whenever FLAIR and DWI hypersignal are present in the semioval centers.

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Rapidly progressive dementia with focal deficits and seizures in a 35-year-old male: from behavioral changes to CNS vasculitis diagnosis amidst HIV infection

Samia Talise El Horr de Moraes[1], Kemily Parenti Perotto[1], Caroline Link[1], Lucas Wagner Gortz[1], Maurício de Carvalho[1], Cláudia Suemi Kamoi Kay[1], Rosana Herminia Scola[1], Helio Afonso Ghizoni Teive[1], Livia Almeida Dutra[2], Caio Cesar Diniz Disserol[1]

[1]Universidade Federal do Paraná, Complexo Hospital de Clínicas, Curitiba PR, Brazil.
[2]Instituto do Cérebro do Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Samia Talise El Horr de Moraes; Email: samiatalise1@gmail.com.

A 35-year-old man with a history of hypertension and gout presented with a one-month history of infantile behavior, memory loss and repetitive speech, followed by sudden left arm weakness and focal seizures. Neurological examination revealed dysarthria, apathy, infantile behavior, slowed speech, temporo-spatial disorientation, executive function decline, left lower limb weakness with hyperreflexia, Babinski sign, and clonus, and hypopalesthesia. A monoparetic gait was observed, with no cranial nerve or meningeal abnormalities. Brain MRI showed extensive cortico-subcortical T2/FLAIR hyperintensities with scattered extensive microhemorrhages and gyriform gadolinium enhancement in the mesial portions of the frontal lobes, left frontal and parietal lobe, right insula, and the temporo-occipital transition, apparently respecting multiple vascular territories. The patient tested positive for HIV, and antiretroviral therapy was initiated (dolutegravir, lamivudine, and tenofovir). CSF analysis showed 4.3 leukocytes/mm^3, glucose 72 mg/dL (serum glucose: 99 mg/dL), protein 111.3 mg/dL, lactate 2.20 mmol/L, and VDRL, viral panel, bacteria and fungi tests were negative. The EEG showed diffuse cerebral dysfunction, worse in the left temporal lobe. Although the viral panel was negative, he was empirically treated with acyclovir. There was no improvement, but neurological stabilization. The patient's condition worsened over some weeks, developing drowsiness, left hemiparesis, and worsened memory, behavior, and seizures. Further MRI and a brain biopsy indicated chronic lymphocytic vasculitis. Treatment included intravenous methylprednisolone and cyclophosphamide monthly for 3 months, followed by an oral taper of prednisone. New brain MRI showed involution of the previous lesions. Despite treatment, the patient remained with severe cognitive and motor impairments. Brain biopsy metagenomics ruled out other pathogens, concluding HIV-associated CNS vasculitis. HIV-related CNS angiitis is rare and usually associated with other pathogens (such CMV, EBV, and others). The differential diagnosis between this condition and primary CNS vasculitis is difficult in this case. Due to the recent unavailability in our service of CSF viral load and genotyping testing, we opted for antiretroviral therapy characteristically with high CNS penetration and, concurrently, started immunosuppressive therapy with pulse therapy with methylprednisolone for 5 days, monthly cyclophosphamide, and prednisone for gradual corticosteroid tapering. With the appropriate stabilization of the condition, prednisone remains in gradual tapering, and cyclophosphamide has already been discontinued. This case report is an important way to remind neurologists about these two rare conditions, and how the appropriate diagnosis can make the difference in the patient's prognosis.

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Rapsyn-associated congenital myasthenic syndrome: distinctive phenotype mimicking limb-girdle dystrophy

Gabriella Corrêa Dousseau[1], Mariana Hiromi Oku[1], Gabriel Saboia de Araújo Torres[1], Filipe Tupinambá Di Pace[1], André Macedo Serafim Silva[1], Beatriz Carneiro Godim Silva[1], Antonio Alberto Zambon[1], Eduardo de Paula Estephan[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Gabriella Corrêa Dousseau; Email: gabrielladousseau@gmail.com.

Case presentation: A 51-year-old woman has experienced fatigue after any physical effort since childhood. During her teenage years, she noticed slower movement compared to peers, difficulty running, climbing stairs, and supporting her neck. In adulthood, her condition worsened, resulting in difficulty rising from a chair without support, frequent falls, increased neck weakness, bilateral semiptosis, and episodes of intense shortness of breath, particularly after infections. On a few occasions, she required hospitalization with mechanical ventilation. Her family history revealed no consanguinity between her parents and noted that 2 of her 5 siblings have similar weakness. Physical examination showed eyelid ptosis, proximal limb and girdle weakness, axial neck weakness, and fatigue with sustained muscle effort. General laboratory tests and CK were all normal. Electromyography (ENMG) demonstrated a decrease greater than 10% with repetitive stimulation and a myopathic pattern. Genetic sequencing revealed two pathogenic variants in compound heterozygosity in the RAPSN gene: c.264C> A (p.Asn88Lys) + c.490C> G (p.Arg164Lys). She was gradually put on pyridostigmine, and salbutamol, result in increased independence in walking, greater resistance for daily activities and no more hospitalizations. On periods of intensification of symptoms, low-dose oral ephedrine provided additional benefit. **Discussion:** Congenital myasthenic syndromes (CMS) are a heterogeneous group characterized by abnormal synaptic transmission at the neuromuscular junction. Pathogenic variants in the RAPSN gene result in a deficiency of the end-plate acetylcholine receptor because rapsyn stabilizes this receptor to the postsynaptic membrane and subsynaptic cytoskeleton. Most RAPSN patients exhibit myasthenic symptoms at birth or in childhood, though some present in the second or third decade of life. Common symptoms include eyelid ptosis, but only 25% have ophthalmoparesis. Other manifestations may include facial, cervical, and bulbar weakness. Respiratory infections or febrile illnesses can exacerbate weakness and respiratory distress. Rarely, RAPSN cases may present proximal weakness, as in our patient. Limb-girdle weakness phenotypes are more frequent with other CMS genes, such as DOK7, GFPT1, MUSK, COLQ, GMPPB, and AGRN. The long-term prognosis for RAPSN-related CMS is generally favorable. The clinical course is typically stable, with intermittent weakness triggered by infections or fever. Respiratory crises decrease in frequency and severity and are rare in adulthood. However, exacerbations characterized by cervical weakness, bulbar symptoms, and ptosis may persist throughout adulthood. **Final comments:** Limb and girdle weakness should not preclude suspicion of congenital myasthenic syndrome. In RAPSN cases, patients typically present with mild to moderate myasthenic symptoms and respond favorably to anticholinesterase treatment, with beta-agonists also showing effectiveness.

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Rare association between CADASIL and multiple system atrophy: a report of three Brazilian cases and literature review

Vanderson Carvalho Neri[1], Ralph Lettieri[2], Karoline Lobo[2], Saulo Machado[3], Leonardo Vandesteen[2], Cristiane Cardoso[4], Ana Luisa Goes[5]

[1]Hospital da Lagoa, Departamento de Neurologia, Rio de Janeiro RJ, Brazil.
[2]Faculdade de Medicina de Campos, Campos dos Goytacazes RJ, Brazil.
[3]Universidade Estadual do Norte Fluminense Darcy Ribeiro, Campos dos Goytacazes RJ, Brazil.
[4]Hospital Unimed, Campos dos Goytacazes RJ, Brazil.
[5]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Vanderson Carvalho Neri; Email: vandersoncn@yahoo.com.br.

Case presentation: Report of 3 cases of patients with association between CADASIL and MSA-C. Case 1: Woman, 58 years old, symptoms for 18 years, clinical presentation of imbalance, dysarthria and left crural hemiplegia. Progressing to urinary incontinence, short-term memory and language impairment, as well as dysphagia. Severe dependence on daily activities. Case 2: A 75-year-old woman, with pathology for 8 years, has been presenting with imbalance, short-term memory changes, left crural hemiparesis. 1 year ago the condition progressed to cognitive decline (moderate bradypsychism), urinary incontinence and dysphagia. Moderate dependence on daily activities. Case 3: Male, 55 years old. Clinical condition for 5 years, characterized by imbalance, bradykinesia, generalized decrease in muscle strength, bradypsychism and urinary incontinence. Intense paresthesia in the upper limbs and tongue region, denies dysphagia. Independent living. For all cases, secondary causes were ruled out and neuroimaging studies demonstrated typical CADASIL changes, as well as the "sign of the cross" in the pons and atrophy of the cerebellum and peduncles. NOTCH3 mutation gene was found in all cases. **Discussion:** CADASIL (Autosomal

Dominant Cerebral Arteriopathy with Subcortical Infarcts and Leukoencephalopathy) is an autosomal dominant pathology caused by mutations in the NOTCH 3 gene. Multiple System Atrophy - MSA is a group of neurodegenerative syndromes characterized by cerebellar ataxia, parkinsonism and dysautonomia. In this work, 3 cases of association of these 2 chronic pathologies were reported. All individuals were followed up on an outpatient basis according to radiological criteria compatible with the 2 pathologies, CADASIL and MSA-C (cerebellar form). MRIs were performed to assess the progression of structural degeneration and leukoencephalopathy. **Final comments:** This is a very rare association between CADASIL and MSA. Currently, only one post-mortem case has been described in the literature. All three forms had cerebellar involvement (MSA-c). The three cases presented cognitive impairment in several domains, with little response to the proposed medications. The association between CADASIL and MSA may be associated with some types of mutations in the NOTCH3 gene.

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Rare early onset CSF-1R-related leukodystrophy

Renata Cristine Alves[1], Luis Eduardo Borges de Macedo Zubko[1], Camila Lorenzini Tessaro[1], Luis Daniel Silva Pilatti[1], Daniel Fortes Amarante[1], Talita Aparecida Conte[1], Caio Cesar Diniz Disserol[1], Francisco Manoel Branco Germiniani[1], Helio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Renata Cristine Alves; Email: renata_cristinealves@hotmail.com.

Case presentation: A 20 years-old man, born to non-consanguineous parents, presented with a history of epilepsy starting at 8 months of age and unilateral amaurosis at 4 years of age, associated with short stature and thoracic disproportion. Conversely, he had normal neuropsychomotor and academic development. He later developed right hemiparesis, initially responsive to corticosteroids, rapidly progressing to spasticity, dysarthria, and dystonia in the right upper limb. He was diagnosed as having multiple sclerosis, but showed inadequate response to therapy. A few months later, he had further significant functional deterioration, with dysphagia leading to multiple hospitalizations for bronchopneumonia and became completely dependent for activities of daily living. He also had a second epileptic seizure, which was adequately controlled with phenobarbital. Additionally, on physical examination there was psychomotor slowing, executive dysfunction, bilateral pyramidal release signs, dystonia, dysarthria, and other signs of appendicular ataxia, as well as reduced visual acuity with unilateral amaurosis and optic nerve papillary atrophy. Cranial MRI showed progressive symmetric subcortical white matter lesions with water diffusion restriction, suggestive of leukodystrophy. Following a comprehensive diagnostic work-up that ruled-out other relevant abnormalities, genetic analysis confirmed a diagnosis of genetic leukodystrophy related to the CSF-1R gene (c.1441C> T variant, p.Gln481Ter). **Discussion:** Leukoencephalopathy with spheroids, a hereditary disease linked to CSF-1R gene mutations on chromosome 5, typically manifests in the fourth decade of life with neuropsychiatric symptoms, cognitive decline, parkinsonism, pyramidal signs, bulbar manifestations, and ataxia. This case reports an unusually early onset, suggesting possible childhood manifestations, which is rather rare, particularly in males. **Final comments:** The possibility of CSF-1R disease with an early presentation should be considered in young patients presenting with the specific phenotype, as a more accurate and earlier diagnosis better correlates with a potential positive response to therapy, especially, bone marrow transplantation.

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Rare presentation of giant cell arteritis following COVID-19 infection

Bruna Pereira Correia[1], Thire Baggio Machado Marazzi[1], Ellen Silva de Carvalho[1], Soraia Fábio Ramos Cabette[1], Maria Lúcia Habib Simão[1], Francisco Antonio Coletto[1], Renato Ramon da Cruz[1]

[1]Hospital Unimed, Ribeirão Preto SP, Brazil.

Correspondence: Bruna Pereira Correia; Email: brunapereirac@hotmail.com.

Case presentation: A 75-year-old woman was admitted to a private tertiary hospital with two days of bilateral consecutive visual impairment, headache, and mandibular pain during feeding. She tested for COVID-19 (Elisa method) for 30 days because of cough and rhinorrhea without support from respiratory necessity. Her past medical history included obesity, arterial systemic hypertension, and dyslipidemia. An ophthalmologic exam described an altitudinal inferior defect in the left eye (LE) and perception of hand movement in the right eye (RE). Fundoscopy showed a papilledema and retinal pallor. Retinal angiography suggested central retinal artery occlusion RE. Doppler US of temporal arteritis demonstrated halo signal bilateral-perivascular hypoechogenicity and a severe thickening vessel wall. MR brain was expected, but orbital MR showed an actual diffusion restriction and SWI hypersocial bilateral intrabulbar optic nerve. Hemo Sedimentation velocity was 72 mmol-seg(ref 15), and no alteration was detected in CFS, including viral pain. The diagnosis of GCA was performed. Screening for other infectious diseases (respiratory, urinary, and sexuality) resulted in negative. Five days of corticoids pelotherapy were performed following rituximab. **Discussion:** Recent concerns have arisen regarding GCA post-COVID-19 vaccination and infections, previously perceived as rare with an incidence of less than 1 in 1,000,000 cases. The median duration from initial symptom onset to the presentation was approximately seven days, with a range spanning from 2 to 25 days, signifying the rapid progression of this condition. This underscores the critical need for healthcare practitioners to remain vigilant, particularly post-vaccination or in infectious contexts when assessing elderly patients with similar symptoms. Given the potential for irreversible vision loss, if diagnosis and treatment are delayed, a multidisciplinary team approach involving ophthalmologists, neurologists, and rheumatologists remains paramount in effectively managing cases like these. **Final comments:** The possibility of GCA following COVID-19 infection is rare but consistent in European literature. No other rares were detected in Brazilian reports.

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Rare simultaneous bilateral thalamic hemorrhage with the presence of the Austregésilo-Esposel sign: a case report and literature review

Leticia Bragalia Passarella[1], Hilton Mariano da Silva Júnior[2], Pedro Neves Fortunato[3]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[2]Pontifícia Universidade Católica de Campinas, Depatamento de Neurologia, Campinas SP, Brazil.
[3]Hospital Mário Gatti, Departamento de Neurologia e Neurocirurgia, Campinas SP, Brazil.

Correspondence: Leticia Bragalia Passarella; Email: leticiabpassarella@gmail.com.

Case presentation: A 46-year-old man was admitted with loss of consciousness, unresponsiveness, and a blood pressure of 220/105 mmHg. He had bilateral hemiplegia and plantar extension after left sole stimulation, indicating a positive Babinski sign on the left. Hallux extension after stimulating the anterior left thigh characterized the Austregésilo-Esposel sign. Brain tomography revealed bilateral thalamic hyperdensity. Findings suggest a rare hemorrhagic stroke, with pyramidal tract involvement. The Austregésilo-Esposel sign, eliciting the same reflex as the Babinski sign, can be used alternatively. However, its literature description is scarce. **Discussion:** Antonio Austregésilo Rodrigues Lima, a Brazilian neurology pioneer, first described the Austregésilo-Esposel sign in 1912. Squeezing the anterior thigh triggers hallux extension. Mentioned in "The Babinski Sign: A Centenary" by Van Gijn in 1996, this rare sign is clinically relevant but underused due to unfamiliarity. Plantar extension reflexes, like the Babinski sign, indicate pyramidal system dysfunctions and help detect lesion levels. Described in 1896, Babinski established the link between this reflex and pyramidal tract injuries. Notable substitutes include Chaddock's, Oppenheim's, and Gordon's signs, which are beneficial when traditional methods are unsuitable, ensuring a complete examination, especially when patients are sensitive to sole stimulation. This case is remarkable not only for the rare Austregésilo-Esposel sign but also for the presentation of simultaneous bilateral intracranial hemorrhage. Typically associated with hypertension, spontaneous intracranial hemorrhages often affect the thalamus and putamen due to artery degeneration from untreated chronic hypertension. The simultaneous occurrence of bilateral hemorrhages is exceptionally rare and scarcely mentioned in the literature. This suggests symmetrically vulnerable vessels, highlighting the need for heightened clinical awareness. Recognizing symmetrically vulnerable vessels can alert clinicians to the risk of such events, prompting vigilant monitoring of patients with hypertension. This case underscores the importance of comprehensive diagnostic evaluations and tailored treatment plans to address these rare but critical conditions effectively. **Final comments:** Neurological semiology evolves continually, and understanding diverse signs is crucial for accurate diagnoses. The Austregésilo-Esposel sign, though scarce, emphasizes the need for knowledge of various neurological assessments. Babinski sign substitutes improve examination accuracy and patient compliance. Recognizing simultaneous bilateral hemorrhage enriches clinical understanding. The medical community must familiarize itself with both conventional and alternative tests to enhance patient assessments, particularly for pyramidal system dysfunctions. This case underscores the complexity and rarity of semiological signs and clinical presentations in medical practice.

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Realistic simulation training in stroke care: a systematic review

Maria Clara Motta Barbosa Valente[1], Vivian Dias Baptista Gagliardi[1], Rubens José Gagliardi[1]

[1]Santa Casa de São Paulo, São Paulo SP, Brazil.

Correspondence: Maria Clara Motta Barbosa Valente; Email: mariaclarambvalente@gmail.com.

Background: The safety of patients and improvement in medical care to minimize the occurrence of adverse events is a widely discussed topic. In neurological emergencies, stroke stands out as a major concern. The time from recognition of the event to the initiation of treatment is crucial for outcomes, considering morbidity and mortality. Due to the emergency environment, there is a higher propensity for adverse events. Therefore, continuous education for team training is of utmost importance. For adults, the learning process differs from children, as they need to understand the reasons for their learning, motivated by problem-solving and active involvement in the learning process; in this aspect, realistic simulation becomes relevant. **Objective:** To evaluate the effectiveness of Realistic Simulation as a teaching and training method for stroke care teams. **Methods:** For the conduction of this review,

the methodology proposed by the Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) was used. Experimental studies of the type Clinical Trial, Quasi-Experimental, Cohort Studies and Case-Control studies were included; the study population was health professionals and the intervention was the use of Realistic Simulation as a training method in acute stroke care. The databases used were MEDLINE (PubMed), LILACS and Cochrane. The search was carried out in October 2023, by two independent evaluators, using the following search strategy: ("Realistic Simulation" OR "Simulation Training") AND ("Acute Stroke" OR Stroke). **Results:** A total of 159 studies were identified, of which 90 were selected and 17 included. Among the 17 studies evaluated, seven had the primary outcome of Door-to-Needle Time before and after team training through realistic simulation. All studies observed a reduction in Door-to-Needle Time, with some findings being statistically significant. Two studies assessed knowledge on the topic through pre and post-testing, both demonstrating an increase in score on the evaluation after training with Realistic Simulation. Another important concept addressed was Patient Safety, improvement in non-technical skills such as Leadership and Team Communication, which were also proven. Additionally, there was an improvement in personal perception of training capacity, greater learning of the topic, increased confidence in decision-making, and indication for thrombolysis. One of the limitations of this review was that, due to the diversity of study designs and primary outcomes, a meta-analysis was not performed. **Conclusion:** The results obtained through this systematic review allow us to conclude that training through Realistic Simulation for stroke patient care is associated with improved personal learning perception, acquisition of technical and non-technical skills, directly influencing important markers in acute stroke care, such as Door-to-Needle Time and higher thrombolysis rates.

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Real-world experience with Fabry disease: case series in a neuromuscular center

Patrícia Marques Mendes[1], Renan Brandão Rambaldi Cavalheiro[1], João Paulo Barile[1], Kaliny Oliveira Peixoto[1], Wladimir Bocca Vieira de Rezende Pinto[1], Paulo Victor Sgobbi de Souza[1], Paulo de Lima Serrano[1], Igor Braga Farias[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Patrícia Marques Mendes; Email: patricia.marques.11@hotmail.com.

Case presentation: In order to investigate the clinical presentations of Fabry disease, 12 medical records were reviewed in a neuromuscular outpatient clinic. Of these, 7 were female, 5 were male and 8 of them had a similar family history. Patients had an average onset of neurological symptoms at 21 years of age with an interval of 17 years until diagnostic definition. The initial neuromuscular manifestations were burning pain (58%), shock pain (16%) and dysesthesia (25%), 66% of which were in the upper and lower limbs and 33% in the lower limbs. The pattern of involvement was mostly small fiber neuropathy (83%), and less than 10% manifested as ganglionopathy or sensorimotor polyneuropathy. The other associated disorders were fatigue (100%), neuropsychiatric disorders (91%), changes in intestinal habit (66%), migraines (66%), sleep changes (58%), abdominal pain (58%), angiokeratomas (51%), structural heart disease (41%), nephropathy (50%), cornea verticillata (50%), arthralgias (50%), anhidrosis or hyperhidrosis (41%), systemic arterial hypertension (33%), urinary incontinence (30 %), gastroparesis (25%), mild sensorineural deafness (25%), arrhythmias (25%), fever (25%), ischemic stroke (16%). Electroneuromyography was normal in the majority, but 25% had axonal sensory-motor polyneuropathy and less than 10% had sensory polyneuropathy. The findings on brain resonance were leukoencephalopathy (41%) and basilar doliectasis (16%). The mutations found in the alpha-galactosidase (GLA) gene were p.Asn34Asp (33%), p.Asp25fs* and p.Arg118Cys (16%), and p.His186Pro (8%). **Discussion:** Fabry disease is an X-linked sphingolipidosis caused by mutations in GLA that manifests predominantly in men from childhood onwards. Its morbidity is generated by the accumulation of ceramide-trihexoside in the walls of nerves and vessels, triggering systemic damage, which can then culminate in vascular diseases in addition to small fiber neuropathy. The diagnostic investigation begins with the measurement of the alpha-galactosidase enzyme in leukocyte cultures, which is reduced and is confirmed by genetic testing, and electroneuromyography may show normal or reduced sensory potentials. **Final comments:** In contrast to the literature, our patient sample consisted mainly of women, and the most prevalent manifestations were non-specific, such as fatigue, neuropsychiatric disorders and small fiber neuropathies, leading to a diagnostic delay of almost two decades. It is noted that specific findings such as angiokeratoma and cornea verticillata were present in only half of the patients. Since enzyme replacement treatment with agalactosidase-beta can offer clinical improvement and reduce morbidities when started early, we must pay attention to atypical presentations and optimize genetic investigation in small fiber neuropathies.

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Reasons for ketogenic diet therapy discontinuation in pharmacoresistant adult epilepsy patients

Raque Alencastro Veiga Domingues Carneiro[1], Maiara Cristina de Lima[1], Mariana dos Santos Lunardi[1], Fernando Netto Zanette[1], Rosa Mendes Cardoso[1], Maria Teresa Salvador[1], Leticia Carina Ribeiro da Silva[1], Júlia Dubois Moreira[1], Débora Kurrle Rieger Venske[1], Katia Lin[1]

[1]Universidade Federal de Santa Catarina, Florianópolis SC, Brazil.

Correspondence: Raquel Alencastro Veiga Domingues Carneiro; Email: raquelalencastrov@gmail.com.

Background: Ketogenic diet therapy (KDT) is a well-established and effective nonpharmacologic therapeutic option for adult patients with pharmacoresistant epilepsy. However, this treatment may present challenges to compliance and adherence which need to be better understood to improve KDT protocols and develop new ways to aid patients in KDT. **Objective:** To describe the main reasons adult patients with pharmacoresistant epilepsy discontinued or did not initiate KDT. **Methods:** Adult patients with non-surgical amenable pharmacoresistant epilepsy were offered to initiate KDT protocol between 2018 and 2024 and were followed-up by a multidisciplinary team (Neurologists, Nutritionists, and Psychologists). Patients could discontinue the diet at any time, and the reasons for discontinuation were recorded. Patients who discontinued the diet before two years of completion or failed to initiate it were analyzed. The reasons for discontinuation were categorized into six groups: (1) Implementation (failure to maintain diet due to its restrictive nature and desire to eat forbidden foods); (2) Social (patients felt the diet negatively impacted their social activities); (3). Financial (diet was too costly to maintain); (4). Weight loss (patients were losing weight excessively); (5). Adverse events (ie, increase in cholesterol levels), 6. Unknown reasons (loss of follow-up). **Results:** A total of 21 participants (F = 10, Mean age = 36 years) were analyzed from a pool of 29 patients who were offered to initiate KDT between 2018 to 2024. Among total, only 2 patients (9.5%) did not start KDT and 5 patients (23.8%) interrupted the diet due to loss of follow-up (group 6). Most patients (33.3%) lost adherence due to the diet's restrictive nature (group 1), often reporting the desire to eat carbohydrates in greater amounts, difficulties to match the daily fat intake or the wish to drink alcoholic beverages. Even though the diet was specifically designed for each patient, some (14.3%) experienced excessive weight loss (group 2), and one patient (4.8%) presented a negative impact on cholesterol levels (group 5), raising concerns about his cardiovascular health. As to socio-economic impacts, one (4.8%) patient did not continue KDT because of social limitations (group 4) caused by the diet, especially at work over not being able to participate in collective meals, for example. A total of 3 patients (9.5%) reported the main adversity being financial reasons (group 3), in which the diet became too expensive to maintain. None of the patients discontinued the diet due to lack of effectiveness for seizure control. **Conclusion:** People with pharmacoresistant epilepsy undergoing KDT face challenges in treatment compliance mainly due to the diet's restrictive nature and cost, as well as its impact on social activities and work environments. Understanding these parameters can aid in the development of new strategies and alternatives so patients can better cope with KDT challenges.

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Reccurent epilepsy seizures in carriers of a heterozygous variant of the PIGN gene

Lorena Emanuele da Silva Castro Oliveira[1], Sophia Calábria da Silveira[1], Fernanda Franklin Pedroso Moreira[1], Marilena Regina de Oliveira Vaz[1], Daniele Soares Vidal[1], Pietro Soares Motta[1], Iasmin Portela Maifrede[1], Bruno Jhonatan Costa Bordest Lima[2], Marcelo Maroco Cruzeiro[1], Danilo Jorge da Silva[1]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[2]Universidade Federal do Mato Grosso, Cuiabá MT, Brazil.

Correspondence: Danilo Jorge da Silva; Email: danilomachadosilva@gmail.com.

Case presentation: Male child, born after uncomplicated pregnancy and cesarean section. No consanguinity history in the family. Patient presented with profound developmental delay. Initial symptoms were noticed at 2 weeks old, characterized by global hypotonia, seizures, and dysphagia. Seizure semiology included multiple events per day of head turning and tonic posture. Additionally, physical examination revealed shortened metacarpals, facial dysmorphisms, and horseshoe-shaped epiglottis on videolaryngoscopy. Initial investigation with cranial magnetic resonance imaging (MRI), transfontanelle ultrasound, and screening for inborn errors of metabolism showed no alterations. A variant classified as likely pathogenic in the PIGN gene c.283C> T and two mutations of uncertain significance in the CILK1 and CCSNK2A1 genes, all in heterozygosity, were detected. At his last evaluation, currently at 3 years old, patient had flaccid tetraparesis, significant cognitive impairment, and unmanageable epilepsy. New cranial MRI showed hypoplasia of the cerebellar vermis, hippocampus, and predominantly frontotemporal cerebral hemispheres. The hypothesis of Multiple Congenital Anomalies-Hypotonia-Seizures Syndrome type 1 (MCAHS1) was raised. **Discussion:** MCAHS1 is a rare genetic condition (1: 1,000, 000) associated with mutations in the PIGN gene, involved in glycosylphosphatidylinositol (GPI) anchor biosynthesis. The syndrome manifests with severe hypotonia, uncontrollable epilepsy, developmental delay, and varied congenital anomalies. The clinical extent of the disease depends on the severity of the PIGN mutation, with possible gastrointestinal malformations (including severe gastroesophageal reflux), global cerebral and cerebellar hypoplasia, diverse genitourinary and cardiac malformations, and facial dysmorphisms such as micrognathia and depressed nasal bridge. Diagnosis is confirmed by identifying mutations in the PIGN gene, typically in homozygosity, through genetic testing. However, there are reports in scientific literature of MCAHS1 associated with compound heterozygous mutations, considered of

uncertain significance in some databases but reported as possibly pathogenic in literature. **Final comments:** MCAHS1 is a underdiagnosed cause of epilepsy, which can lead to delayed diagnosis and initiation of appropriate support for affected individuals. It should be considered in cases of hypotonia and multi-organ/system malformations. This case report demonstrates a particularly atypical case with a clinical picture consistent with MCAHS1, extensive investigation excluding other syndromes, yet revealing a heterozygous mutation in the PIGN gene. Despite this unexpected genetic finding, diagnosis should not be dismissed. Presence of deep intronic variants, repetitive elements in the gene of interest, allelic dropout, and imprinting disorders are some examples of mechanisms that could explain the phenotypic manifestation in the absence of a clear molecular diagnosis.

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Recurrence of epilepsia partialis continua on the opposite side after hemispherectomy: an uncommon yet severe outcome of focal cortical dysplasia

Samir Andres Ramirez Ojeda[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Samir Andres Ramirez Ojeda; Email: samir.ramirez@edu.pucrs.br.

Case presentation: At three months old, a girl began experiencing clonic spasms originating in her left hand, spreading to her left arm, face, and leg, sometimes accompanied by eye deviation to the left. Antiseizure drugs failed to control these spasms, which increased in frequency and occasionally progressed to TC seizures. EEG findings initially showed slowing in the right hemisphere, with frequent multifocal discharges evolving into a hemi-hypsarrhythmic pattern. MRI revealed mild hemispheric asymmetry with significant right-sided atrophy. By the age of two, she presented to our epilepsy surgery service with inability to speak and dependence on support for sitting. Physical examination indicated noticeable weakness on the left side. Subsequent MRI revealed multifocal cortical dysplasia and marked atrophy in the right hemisphere. EEG continued to depict intense epileptic activity predominantly in the right hemisphere, confirmed by PET-SCAN findings of reduced metabolism in that area. A decision was made to proceed with right hemispherectomy, initially resulting in partial control of seizure frequency. However, shortly after surgery, seizures recurred in her right arm and face. EEG demonstrated disorganized activity with a slow rhythm on the right and bilateral epileptiform discharges, more frequent in the left hemisphere. Histological examination confirmed focal cortical dysplasia. Despite adjustments in ASDs, her seizures persisted, highlighting ongoing challenges in managing her condition post-hemispherectomy. **Discussion:** Contralateral epilepsia partialis continua following hemispherectomy is a rare and complex clinical phenomenon. Hemispherectomy involves partial or complete removal of one cerebral hemisphere, typically performed for severe, medically refractory epilepsy originating from a single hemisphere. The goal is to eliminate the epileptogenic zone and achieve seizure control. However, in some cases, seizures may reappear in the contralateral hemisphere, presenting as contralateral EPC. This condition poses diagnostic challenges as it may mimic or coexist with residual or new-onset epilepsy following hemispherectomy. Diagnosis relies on comprehensive clinical evaluation, Video-EEG, and neuroimaging to assess post-surgical brain anatomy and identify any residual abnormalities. Long-term outcomes vary widely based on factors such as age at surgery, underlying epilepsy etiology, completeness of resection, and cortical reorganization extent. They range from successful seizure control to persistent epilepsy necessitating ongoing management. **Final comments:** Contralateral EPC following hemispherectomy underscores the complexity of epilepsy surgery and the potential for intricate post-operative seizure patterns. Continued research into mechanisms of contralateral epileptogenesis, along with advancements in surgical techniques and adjunctive therapies, is crucial to optimizing outcomes for patients with refractory epilepsy undergoing hemispherectomy.

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Recurrent anti-LGI1 limbic encephalitis: a case report

Wender Rodrigues Teodoro[1], Arthur Costa Nascimento[1], Lucas Figueira Vieira[1], Letícia da Silva Alves[1], Matheus Cunha Nunes[1], Amanda Ribeiro França[1], Vicente José da Silva Filho[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Wender Rodrigues Teodoro; Email: wenderteodoro@gmail.com.

Case presentation: A 71-year-old male patient began presenting in 2021 with mental confusion, psychosis, episodic and work-related amnesia, and epileptic seizures with brachiofacial dystonic presentation. He was hospitalized and underwent a lumbar puncture that showed normal cellularity (1 cell/mm3), hyperproteinorrhachia (108 mg/dl), non-reactive VDRL, culture for fungi and bacteria without alterations, negative oligoclonal bands, positive IgG index, and antibody panel with positive LGI1. Brain MRI showed signs of supratentorial microangiopathy. ECG showed diffuse disorganization of the basal activity with the presence of triphasic waves. He underwent pulse therapy with methylprednisolone, presenting partial improvement of symptoms, maintaining episodic amnesia and visuospatial disorientation. Given the incomplete response, it was decided to start pulse therapy with monthly cyclophosphamide for 3 months, with partial response. During hospitalization, neoplastic screening was performed, with no findings. Approximately 1 year after the onset of the first symptoms, the patient began to have nocturnal awakenings accompanied by visuospatial disorientation, mental confusion and epileptic seizures with the same initial pattern. Upon admission, the patient was disoriented in time and space and had high-frequency, low-amplitude distal tremor. During the initial laboratory investigation, hypokalemia was identified, which was corrected by enteral route. A cerebrospinal fluid puncture was performed, the result of which showed hyperproteinorrhachia (139.5 mg/dl), glucose (64 mg/dl) and 5 cells. After ruling out infectious causes, a new cycle of pulse therapy with methylprednisolone for 5 days was chosen, with significant improvement in symptoms. The patient is being monitored on an outpatient basis using immunosuppressive therapy with rituximab, with no recurrence of epileptic seizures, but with cognitive impairment. **Discussion:** Anti-LGI1 encephalitis is an autoimmune condition that usually presents with confusion, amnesia, cognitive decline, brachiofacial dystonia, and seizures with low response to anticonvulsants, with the possibility of progressive symptomatic onset with no signs in the cerebrospinal fluid or imaging. The LGI1 protein acts as a ligand for epileptic proteins such as ADAM22 and ADAM23. When the antibody binds to LGI1, it triggers a state of neuronal hyperexcitability, manifesting symptoms. Therefore, in most patients, immunomodulatory therapy tends to be effective. Approximately one-third of patients present relapses, which are more common in the first six months after therapy, predicting poor outcome. Most patients maintain cognitive deficit despite therapy. **Final comments:** The reported case presents the challenge of identifying and treating recurrent anti-LGI1 encephalitis after approximately 1 year of therapy. In addition, it portrays therapeutic management with improvement of the condition but with maintenance of cognitive dysfunction.

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Recurrent encephalopathy with extensive brain lesions associated with mog antibody: a case report

Pedro Ferreira Bastos[1], Rafael Bastos Delgado[1], Regina Maria Papais Alvarenga[1], Marina Papais Alvarenga[1], Camille Freitas de Araujo[1], Gabriel Lisbôa Pereira[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil.

Correspondence: Pedro Ferreira Bastos; Email: pedroferreirabastos1@gmail.com.

Case presentation: Girl, 11 years old, brown, suddenly started aphasia and distal paresis in the right upper limb in February 2020 without previous neurologic episodes. Magnetic resonance (MRI) of the brain showed hypersignal on FLAIR and T2-weighted images with a digitiform edema pattern in the subcortical white matter of the frontal regions, more to the left, with slight hypersignal diffusion and areas of contrast enhancement with a perivascular/perivenular impregnation pattern. There were also small areas of hypersignal on T2-weighted and FLAIR sequences in the subcortical white matter of the parietal and temporal regions, with no hypersignal diffusion and no contrast enhancement, suggestive of ADEM. CSF unaltered. The patient progressed in 15 days with progressive improvement of aphasia. The examination was repeated after 2 months, demonstrating significant improvement of the lesions. Still, it showed a new lesion on the right parietal surface and a more extensive lesion involving the right temporooccipital subcortical white matter, consistent with new active lesions. Signs of optic neuritis were also observed on the left, with no further contrast uptake, and on MRI of the cervical and thoracic cords, no demyelinating plaques were identified. Pulse therapy (PST) was performed with methylprednisolone in a decreasing regimen (3 days in the first month, 2 days in the second month, and 1 day in the third month) in June, July, and August, with complete remission of symptoms. In 2021, without preventive treatment, she presented a focal seizure, went to the emergency room, and was released without treatment. In February 2022, the patient started a generalized seizure with a postictal state that evolved with somnolence and Brady psychism. After 1 week, she was admitted to three days of treatment with methylprednisolone with total improvement. She was seen for the first time at the referral center in Rio de Janeiro in June 2022, with no new neurological manifestations. She was investigated for the biological markers AQP4 and MOG. The result indicated negative AQP4 and positive MOG. The tests were repeated in December of the same year, confirming the findings. After this period, the patient again presented distal paresis of the right upper limb, alteration of coordination, and dysarthria. In 2023, she showed partial recovery of the deficit, without dysarthria and seizures, but maintained paresis in the right upper limb. **Discussion:** MOG-Ab antibodies were identified in NMO and ADEM negative for AQP4-Ab in 2011. Encephalopathy MOG-AB was described after 2017. In 2023, international criteria for the diagnosis of MOGAD were proposed for the first time, considering that the spectrum of the syndrome was broadening. **Final comments:** The presenting case met the diagnostic criteria for MOGAD. The core syndrome was encephalopathy with polyfocal deficit with an additional criterion of multiple lesions on MRI.

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Recurrent hemorrhagic stroke with associated use of lurasidone: a case report

Isabella Beserra Ramos[1], Rebeca Talita de Souza Siqueira[1], Pedro Thiago Simões Ferreira[1], Neila Clediane de Sousa Menezes[1], Raíra Yana Lima Barbosa[2], Rita Di Cássia Oliveira Ângelo[2], Américo Danuzio Pereira de Oliveira[1,2]

[1]Hospital Eduardo Campos, Serra Talhada PE, Brazil. [2]Universidade de Pernambuco, Serra Talhada PE, Brazil.

Correspondence: Isabella Beserra Ramos; Email: isabellabeserramos@gmail.com.

A 45-year-old woman presented to the emergency department with a sudden and severe headache, accompanied by nausea, vomiting and dizziness. Despite the intense pain, she was fully conscious, alert, and showed no signs of neurological impairment. Her previous medical history included generalized anxiety disorder, scleroderma, obstructive sleep apnea syndrome. She was taking Desvenlafaxine and Alprazolam in previous consult. A week earlier before the symptoms it was prescribed Lurasidone because of the worst of the anxiety symptoms. A cranium CT scan revealed a dense area in the right parietal region. AngioCT of the intracranial vases was negative for aneurysm, arterial or venous malformation and thrombosis. She received conservative treatment, and her symptoms improved over a 5-day period, allowing her to reasume her regular medications. Three days after being discharged, she returned with a recurrence of similar headache symptoms and a new dense area on the opposite side, along with evidence of subarachnoid hemorrhage. Magnetic resonance angiography showed narrowing of the bilateral middle cerebral arteries. Analysis of cerebrospinal fluid indicated elevated red blood cells with normal glucose and protein levels. The onset of symptoms coincided with the recent introduction of lurasidone. Treatment with nimodipine provided pain relief, and lurasidone was discontinued upon discharge. Follow-up angioMRI after 60 days showed normal vascular findings. Reversible cerebral vasoconstriction syndrome (RCVS) is characterized by sudden, severe thunderclap headaches and focal narrowing of cerebral arteries. The global epidemiology of RCVS remains uncertain, likely underestimated, and predominantly affects middle-aged women. It can occur spontaneously or secondary to factors such as childbirth and medications affecting vascular tone. While the direct vasoactive effects of lurasidone are not well-established, antipsychotics like lurasidone can potentially influence vascular regulation through modulation of neurotransmitters. Lurasidone primarily antagonizes dopamine and serotonin receptors, with serotonin playing a role in regulating cerebral vascular tone, potentially contributing to vasoconstriction. Diagnosing RCVS requires differentiation from conditions like aneurysmal subarachnoid hemorrhage, necessitating a high index of suspicion. Treatment focuses on managing symptoms and preventing cerebrovascular complications. This case underscores the diagnostic challenges of RCVS and emphasizes the importance of investigating potential triggers, including psychotropic medications. Further studies are needed to fully elucidate any causal association between lurasidone and RCVS, demonstrating the need for a multidisciplinary approach to patients with complex neurological presentations.

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Recurrent Takotsubo cardiomyopathy associated with recurrent status epilepticus: a clinical case report

Julia Costa Nacif Santos[1], Luna Lila Fernandes de Faria[1], Mariana Di Paula Rocha Rodrigues[1], Camila Mendes dos Passos[1], Rodolpho Poeys Lima[1], Lucas Vilas Boas Magalhães[1]

[1]Universidade Federal de Viçosa, Viçosa MG, Brazil.

Correspondence: Lucas Vilas Bôas Magalhães; Email: lvbmagalhaes@ufv.br.

Case presentation: A 63-year-old woman, diagnosed with epilepsy in 2002, was admitted to the emergency room (ER) in 2016 with prolonged tonic-clonic status following an episode of significant emotional distress. The patient went into respiratory depression, and an electrocardiogram was performed, showing diffuse ST-segment elevation. She was admitted in the ICU and underwent cardiac catheterization, which was suggestive of Takotsubo Cardiomyopathy. Sequential echocardiograms showed ventricles with normal systolic function and just a grade 1 diastolic dysfunction after 4 months from the episode. Patient's history revealed a similar episode of cardiogenic shock following status epilepticus in 2011. Catheterization performed in this first event showed left ventricle with antero-lateral-apical akinesia and increase in end-systolic and end-diastolic volumes, with no signs of arterial obstruction, which leads us to believe that was, in fact, her first episode of Takotsubo syndrome (TTS). In 2017, a new admission to the ER by TTS following a status epilepticus and emotional distress occurred, although early diagnosis and adequate treatment led to better evolution of the condition. **Discussion:** Takotsubo syndrome consists of an acute transient left-ventricular systolic dysfunction, typically triggered by a physical and/or emotional stressor. According to an International TTS Registry, neurological triggers account for 6.7% of TTS's admissions and seizures have been characterized as the second most common of those. Albeit the incomplete understanding of Takotsubo Cardiomyopathy, the sympathetic nervous system seems to play a pivotal role in its pathophysiology, and catecholamine release after seizures may explain the correlation between these two conditions. TTS occuring after seizures is more frequent in younger and male patients, they suffer less from chest pain, have a more serious course and have higher recurrence than other conditions. Furthermore, recent data from an international cohort on TTS showed an overall recurrence rate of 4.7%. Out of the 1402 patients, only 5 had more than one recurrent episode, and all of these had associated psychiatric and/or neurological conditions. Despite that, we haven't found, in our research, a description of these multiple recurrences or their triggers. This case describes the rare presentation of multiple recurrent episodes of Takotsubo syndrome, all triggered by both emotional and tonic-clonic status epilepticus. **Final comments:** There are very few multiple recurrent episodes of Takotsubo syndrome documented in literature. Neurologic comorbidities might be a predisposing condition for recurrence, and better characterization of neurologic triggers in patients presenting with multiple episodes of TTS might contribute to elucidate this association.

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Red flags in dermatomyositis diagnosis: a pilot study

Pedro Nogueira Fontana[1,2], Alessandra Lima Nogueita Tolentino[1], Roseli Corazzini[1], Carolina da Cunha Correia[2], Nathalia Merten[1], Ana Marina Dutra[1], Igor Brockhausen[1], David Feder[1], Alzira Alves de Siqueira Carvalho[1]

[1]Centro Universitário Faculdade de Medicina do ABC, Santo André SP, Brazil.
[2]Hospital Universitário Oswaldo Cruz, Recife PE, Brazil.

Correspondence: Alessandra Lima Nogueira Tolentino; Email: ale.tolentino1994@hotmail.com.

Background: Dermatomyositis (DM) is classically defined as an inflammatory myopathy associated with typical rash, elevated creatine phosphokinase (CK) and characteristic patterns on muscle biopsy, such as inflammatory infiltrate, perifascicular atrophy (PFA) and positive immunohistochemistry (IHC) for myxovirus resistance protein A (MXA). However, some cases may be difficult to diagnose because they may present some atypical characteristics, such as normal CK levels and absence of inflammation or PFA on muscle biopsy. **Objective:** We present a series of cases with atypical morphological findings, in which it was possible to conclude a diagnosis of DM through MXA immunostaining. **Methods:** All cases presented proximal muscle weakness, DM skin lesions and positivity for MXA at IHC. All underwent serum CK measurement. Muscle biopsy was performed using previously described techniques; analysis included IHC for CD4, CD8, CD20, CD68, C5b9 and MXA. Immunostaining for MXA was performed in cases with unspecific findings on muscle biopsy and/or negative immunostaining for CD4, CD8, CD68 and C5b9 and/or normal CK. Typical cases of DM in which MXA was also positive were used as positive controls. **Results:** We identified 6 patients with these redflags, being 3 women, with a mean age of 37±8.85 years old. Median CK was 146.5 IU/L, ranging from 77 to 15.220 IU/L; 5 cases had normal CK. 4 cases were using immunossupressive treatment at biopsy time (mean time: 2.8 months). At muscle biopsy, 2 cases had no PFA, 4 cases had no CD4 and 5 cases had no CD8 inflammatory cells, 2 cases had negative immunostaining for MHC-I, 3 cases had no expression of C5b9. In the comparison group, there were 5 women and 1 men, with a mean age of 51±16 years old. Median CK was 2402, ranging from 390 to 7000 IU/L; all cases were using immunossupressive treatment at biopsy moment (mean time: 5.9 months). At muscle biopsy, 2 cases had no PFA and 1 case had no CD4 inflammatory cell. All cases had MHC-I, CD8 and C5b9 expression. In both groups, all cases were positive for MXA immunostaining. **Conclusion:** We demonstrate that, in the absence of classical pathological criteria and normal CK values, IHC for MXA can be a valuable diagnostic tool due to its high specificity for DM (98-100%), despite a reported sensitivity of around 70%. Imunossupressive treatment does not seem to influence MXA immunostaining. In the absence of such characteristics, MXA immunostaining should be included to confirm DM diagnosis. More studies are necessary to confirm our results.

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Refractory chronic inflamatory demyelinating polyneurophaty (CIDP) syndrome in a pregnancy patient: a case report

Pedro Henrique Queiroz de Oliveira[1], Guilherme Menezes Mescolotte[1], Maria Teresa Fernandes Castilho Garcia[1], Ana Caroline Kuniochi[1], Elaine Fernanda Dornelas de Souza[1], Guilherme Eugênio Gil[2], Mayara Zavatini Cansini[1]

[1]Universidade do Oeste Paulista, Presidente Prudente SP, Brazil.
[2]Universidade de Marília, Marília SP, Brazil.

Correspondence: Pedro Henrique Queiroz de Oliveira; Email: pedroqueiroz@unoeste.edu.br.

Case presentation: Female, 22 years old, presented to the Emergency Care Unit in July 2023 with paresthesias and loss of strength in the lower and upper limbs, with caudo cranial evolution, areflexia and frequent falls. No comorbidities were related. She was referred to a tertiary service with normal cranial tomography. Cerebral spinal fluid had 5/mm^3 lymphomononuclear cells, and 140mg/dL of proteins. The hypothesis of Guillain Barré Syndrome was made, she was treated with 2g/kg of immunoglobulin in the Intensive Care Unit, with improvement, and was discharged and returned to outpatient care. She returned in 2 months with sudden tetraparesis and difficulty swallowing and walking. During this hospitalization, electroneuromyography showed an active demyelinating process suggestive of CIDP. Methylprednisolone therapy (1g/day) was carried out for 5 days with improvement, the patient was discharged and prescribed continuous use of prednisone 20mg/day, until her return. In January 2024, she returned with complaints of tetraparesis and difficulty walking. Due to the pregnancy, it was decided with obstetrics to carry out treatment with immunoglobulin, however, it was not released. In two days the patient presented a progressive worsening of motor symptoms, and was decided to start methylprednisolone for 5 days. She improved and received prednisone 60 mg/day orally,

with almost no more symptoms. **Discussion:** CIDP is an autoimmune neuropathy that affects peripheral nerves and nerve roots, with a remitting-recurrent or progressive course of symmetrical weakness of proximal and distal muscles. It is identified by electrodiagnostic and/or pathological features of demyelination. For untreated patients with progressive worsening, treatment with intravenous immunoglobulin (IVIG), plasmapheresis or glucocorticoids is recommended. Glucocorticoids can be administered intravenously or orally and have been shown to be effective, being reduced once clinical stabilization is achieved. However, there is still little data regarding the course of CIDP in pregnant women and how to manage cases during pregnancy. **Final comments:** CIDP is a progressive, rare and sometimes disabling disease which, although it mainly affects men in adulthood, in the case presented, we observed a pregnant woman suffering from the same condition. There is still a lack of evidence as to the best course of action in these cases. This study makes a scientific contribution by presenting a rare and little-reported condition of CIDP during pregnancy, so that health professionals can better guide cases that present in the same way in their daily work.

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Refractory intracranial hypertension associated with the use of all-trans-retinoic acid: a case report

Oliver Reiks Miyajima[1], João Vitor Moreira Nogueira[2], Matheus Henrique Brustolim Marcucci[3], Bruna Barbosa Nobre[4], Paulo Ribeiro Nóbrega[5], Nádia Gurgel Alves[6], Tayenne Nélly de Lucena Viana[7], Yan Bruno Colares Botêlho[6], Viviany de Oliveira Viana[6], Carolina de Figueiredo Santos[6]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.
[2]Unichristus, Fortaleza CE, Brazil.
[3]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[4]UC, Fortaleza CE, Brazil.
[5]Hospital Universitário Walter Cantídio, Fortaleza CE, Brazil.
[6]Hospital Infantil Albert Sabin, Fortaleza CE, Brazil.
[7]Universidade de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Oliver Reiks Miyajima; Email: oliver.miyajima@aluno.uece.br.

Case presentation: A 14-year-old female diagnosed with (AML) began treatment with all-trans-retinoic acid (ATRA), presenting with headache and vision loss a few days later. Lumbar puncture revealed a cerebrospinal fluid (CSF) opening pressure of 52 cmH2O and no other abnormalities, confirming the diagnosis of intracranial hypertension . ATRA was stopped and acetazolamide was started. Magnetic resonance imaging (MRI) showed no abnormalities.There was initial improvement in pain intensity, but the patient had severe metabolic acidosis. Acetazolamide was switched to topiramate with reduction of headache intensity. Two weeks later ATRA was reintroduced and headache recurred with increased intensity. She was treated with opioids with little response and repeated LP revealing a persistently increased CSF pressure of 41 cmH2O. Despite very high CSF pressure, fundoscopy did not reveal papilledema. She completed 3 cycles of ATRA and received most of the fourth cycle before the medication was suspended to control the refractory intracranial hypertension syndrome. One month later, the patient reported improvement of headache and CSF opening pressure decreased to 26 cmH2O. She was able to achieve remission of AML. **Discussion:** Idiopathic intracranial hypertension (IIH) is defined as intracranial hypertension without changes in neuroimaging or CSF composition. This patient presented with IIH during treatment with ATRA and acetazolamide was administered in an attempt to control intracranial hypertension, without significant improvement. Only once ATRA was discontinued was IIH controlled. Despite very elevated intracranial pressure, papilledema was not observed. IIH is manageable and early intervention is essential in preventing persistent visual loss and other complications. Acute promyelocytic leukaemia (APL) is an aggressive subtype of acute myeloid leukaemia with the presence of retinoic acid receptors and a high response to ATRA, achieving remission rates of 100%. The pathophysiology of ATRA-induced intracranial hypertension is unclear. ATRA is thought to elevate intracranial pressure by altering choroid plexus and arachnoid villi composition, leading to enhanced CSF production and impaired absorption. In the context of ATRA-induced intracranial hypertension, withdrawal of ATRA therapy and reintroduction at lower dose alongside prophylactic administration of acetazolamide is a useful treatment approach, but this approach was attempted in this case and failed. Using topiramate and low dose ATRA, the patient was able to achieve remission. **Final comments:** Intracranial hypertension is a significant complication in Acute Myeloid Leukaemia patients in treatment with all-trans-retinoic acid. Early treatment can prevent long-term complications, such as persistent visual loss. Risks and benefits must be weighed when deciding whether to continue treatment with all-trans-retinoic acid in patients with intracranial hypertension.

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Refractory post-herpetic anti-NMDA receptor autoimmune encephalitis: case report

Ellen Silva de Carvalho[1], Bruna Pereira Correia[1], Thire Baggio Machado Marazzi[1], Renato Ramon da Cruz[1], Soraia Fábio Ramos Cabette[1], Francisco Antonio Coletto[1]

[1]Hospital Unimed, Ribeirão Preto SP, Brazil.

Correspondence: Bruna Pereira Correia; Email: brunapereirac@hotmail.com.

Case presentation: An 18-year-old woman started experiencing irritability, progressing to headache, fever, and language alteration. Herpes Simplex Virus type 1 (HSV1) encephalitis was confirmed, and treatment with acyclovir was initiated in the hospital and continued on an outpatient basis. On the 8th day of treatment, she developed vomiting, focal epileptic seizure, and language alteration, leading to readmission and treatment for left temporal and insular focal structural epilepsy. During the investigation, left middle cerebral artery arteritis was identified, and pulse therapy with methylprednisolone 1g/day for five days was administered. Two weeks later, she was readmitted due to dysautonomia and behavioral and consciousness level changes. Fludrocortisone was started, resulting in improvement. However, she was readmitted a week later with mutism and oromasticatory movements, suspected of post-herpetic autoimmune encephalitis. Cerebrospinal fluid analysis revealed positive anti-NMDA antibodies. Pulse therapy was administered with 1g/day methylprednisolone for five days and human immunoglobulin infusion at 2g/kg over five days. Due to poor clinical response, rituximab 1g was given, repeated in 15 days. At discharge, she followed simple commands, moved all four limbs spontaneously (more on the left), and spoke single words. Ambulatory motor rehabilitation therapy was initiated, resulting in improvement, with fluent and mildly dysarthric speech, active limb movement, and return to previous instrumental activities. **Discussion:** Herpetic encephalitis is uncommon, with an estimated annual incidence of 2-4 cases per million. Clinical deterioration after herpetic encephalitis can occur due to viral invasion and autoimmune mechanisms. Anti-NMDA autoimmune encephalitis, though rare, is the most common autoimmune etiology in these cases, with HSV1 being identified as the primary associated virus. Managing these conditions requires a detailed approach considering both viral and autoimmune aspects, reflecting the evolving trends and diagnostic complexities in neuroinflammatory disorders. First-line treatment includes pulse therapy with methylprednisolone and intravenous human immunoglobulin or plasmapheresis. Second-line treatment with rituximab or cyclophosphamide is indicated for refractory cases. **Final comments:** The case report discusses the rare co-occurrence of HSV1 encephalitis and anti-NMDA receptor encephalitis, presenting a challenging clinical scenario. A positive clinical outcome was achieved through tailored treatment and collaborative interdisciplinary efforts, emphasizing the importance of ongoing surveillance and research in infectious and autoimmune encephalitis. This case underscores the intricate epidemiology and clinical challenges in post-herpetic anti-NMDA receptor autoimmune encephalitis, emphasizing the importance of ongoing research and vigilant management in infectious and autoimmune encephalitis conditions.

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Regional epidemiological analysis and costs associated with admissions for headache in the unified Brazilian health system: notifications from 2019 to 2023

Ana Clara Lacerda Freitas[1], Giovanna Edméa Rosário Luthy Carletto[2], Jeová Fernandes Pereira[3], Guilherme de Andrade Ruela[4]

[1]Centro Universitário Funorte, Montes Claros MG, Brazil.
[2]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[3]Universidade Estadual do Tocantins, Augustinópolis TO, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Ana Clara Lacerda Freitas; Email: anaclf15@yahoo.com.br.

Background: Primary headaches are one of the most disabling and common neurological conditions around the world. Although general lifestyle factors such as poor diet, excessive stress and bad posture are known causes of headaches, other aspects of modern life such as smartphone use also affect headaches, contributing to the disorder and lack of focus that characterize presenteeism in the workplace. That way, it is important to understand headaches and their associated costs, as well as to define the most affected population profile. **Objective:** To describe the profile and hospital costs of hospitalizations for headache in each region of Brazil from 2019 to 2023. **Methods:** Ecological study carried out using data extracted from the SUS Hospital Information System (SIH/SUS) through the Department of Informatics of the Unified Health System (DATASUS). A descriptive analysis was carried out in Microsoft Excel, covering the variables number of hospitalizations, total cost spent, gender, race and region, from 2019 to 2023. **Results:** The data obtained indicate that 51,257 hospitalizations for headache were recorded in Brazil, with an accumulated cost of R$28,033,387.80. The Northeast Region recorded the highest number of hospitalizations, 30.9% of cases, and the highest volume of costs, R$11,081,078.73 (39.5%). In contrast, the North Region, with 8.3% of cases, had costs of around R$2,864,694.80 (10.2%), not representing the lowest costs despite the smaller number of cases. The Central-West Region had 5.8% of hospitalizations, with a cost of R$777,869.80 (2.8%), presenting the lowest costs and the smallest number of cases. It was observed that women, with 65.5% of hospitalizations, and the mixed race, with 44% of cases, were the most affected. Furthermore, there was a 13.6% reduction in hospitalizations in 2020 compared to 2019, followed by a recovery in subsequent years. Thus, the Northeast Region has the highest prevalence and highest costs associated with hospitalizations for headache. However, there are variations in terms of number of cases and costs between different regions, as well as racial disparities. **Conclusion:** The results obtained

in this study reveal a heterogeneous distribution of hospitalizations for headache in Brazil between the years 2019 and 2023, with a decrease of 13.6% in 2020, possibly due to the restrictions of the COVID-19 pandemic and changes in practices seeking medical care. These data indicate that there are regional differences in the profile of hospitalizations, access and treatment practices. The importance of public health policies that promote effective therapeutic practices and guarantee equitable access to health services is highlighted, with a focus on primary care, which plays a crucial role in preventing hospitalizations and improving quality of life.

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Relapse comparison between parenchymal and non-parenchymal forms of Neuro-Behçet

Gabriel Samir Martins de Souza[1], Gabriela Abrahão Allioni[1], Guilherme Diogo Silva[1], Marcia Rubia Rodriges Gonçalves[1], Dagoberto Callegaro[1], Leandro T. Lucato[1], Rafael Alves Cordeiro[1], Gisela Tinone[1], Henrique Ayres Mayrink Giardini[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Gabriela Abrahão Allioni; Email: gabriela.allioni@hc.fm.usp.br.

Background: Neuro-Behçet (NB) refers to neurological manifestations associated with Behest disease, encompassing two main clinical and radiological presentations: parenchymal (e.g., brainstem encephalitis) and non-parenchymal (e.g., cerebral venous thrombosis) forms. Although NB is recognized as either monophasic or recurrent, there is limited data on relapse frequency in Latin-American patients and how distinct clinical and radiological presentations influence this risk. **Objective:** To determine the frequency of relapses in NB and assess whether the initial presentation in parenchymal or non-parenchymal form impacts this outcome. **Methods:** This is a retrospective observational study conducted at the tertiary academic referral outpatient clinic specializing in Behçets disease in São Paulo, Brazil. We reviewed medical records of patients diagnosed with NB from 2010 to 2021, according to the International Consensus of NB criteria. Data on relapse occurrence and clinical-radiological characteristics were collected to classify the initial NB presentation into parenchymal and non-parenchymal forms. Time to relapse was illustrated using Kaplan-Meier curves, and the log-rank test compared time to relapse between non-parenchymal and parenchymal presentations. **Results:** Thirty-one patients were included (48% male; 24 (77%) with parenchymal disease), with 20 (65%) experiencing recurrence during follow-up. All participants with non-parenchymal disease (n=7) had cerebral venous thrombosis. While half of NB patients experienced relapse within the first five years after presentation, the timing varied from one to ten years. Participants with non-parenchymal disease had a median time to relapse of one year compared to a median time to relapse of six years in those with parenchymal disease (p=0.043). **Conclusion:** Recurrence was observed in 65% of NB patients, time to relapse ranged from one to ten years, and occurred earlier in patients with non-parenchymal disease, particularly cerebral venous thrombosis. This study is ongoing.

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Relapse frequency and predictors in immune-mediated hypertrophic pachymeningitis

Amanda Angulo Soares[1], Tarso Adoni[2], Guilherme Diogo Silva[2], Luiz Henrique Martins Castro[2]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Amanda Angulo Soares; Email: anguloamanda@gmail.com.

Background: Immune-mediated hypertrophic pachymeningitis (IHP) is an inflammatory condition characterized by dura mater thickening, headaches, cranial nerve palsies, seizures, and focal deficits. Previous studies showed different recurrence rates of the disease, and little is known about prognostic factors for relapse. A deeper understanding of recurrence is necessary to guide optimal immunosuppressant strategies in IHP. **Objective:** To evaluate the frequency and time to the first relapse, and to identify potential clinical, radiological, and laboratory predictors of relapse in this patient population. **Methods:** This observational retrospective case series was carried out in a dedicated outpatient clinic for IHP in São Paulo, Brazil, followed from 2002 to 2024. We included consecutive patients with radiological or pathological evidence of pachymeningitis due to probable immune-mediated etiology, after excluding infection, neoplasm, and intracranial hypotension. Patients were divided into relapse (R) and non-relapse (NR) groups, according to occurrence of relapse until the last follow-up. We collected clinical, neuroimaging, and laboratory features from the initial or first available clinical presentation. Time to relapse was calculated using Kaplan-Meier curves. Variables that were significantly different between R and NR groups were compared to identify prognostic factors for relapse. **Results:** We included 78 patients, of whom ten (13%) had IgG4-related disease, 15 (19%) probable or definitive neurosarcoidosis, two (3%), systemic vasculitis, and one (1%), amyloidosis. Idiopathic pachymeningitis corresponded to 51 (65%) of cases. Relapse occurred in 43 (55%) patients. Median time to relapse was six years (range 0 to 17 years). Seizures at disease onset and C-reactive protein levels above 10mg/dL in presentation were associated with shorter time to release (p<0.05). **Conclusion:** In this consecutive single center series of IHP relapse was common, and associated with seizures and elevated C-reactive protein levels on initial presentation. Future studies should evaluate immunosuppressant use strategies, especially for those identified as higher relapse risk.

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Relapse of neuromyelitis optica with atypical presentation of painful tonic spasms and area postrema syndrome: a case report

Juliana Magalhães Leite[1], Thaís Magalhães Lima Leite[2], Lucas Silva Barbosa Bezerra[2], Lívia Gonçalves Santos Costa[2], Ana Lívia de Andrade Rangel[2], Bianca Etelvina Santos de Oliveira[3], Matheus Saraiva Gurgel[4], Rafael de Sousa Andrade[4]

[1]Faculdade de Medicina Nova Esperança, João Pessoa PB, Brazil.
[2]Centro Universitário de João Pessoa, João Pessoa PB, Brazil.
[3]Fundação Centro Integrado de Apoio à Pessoa com Deficiência, João Pessoa PB, Brazil.
[4]Hospital Metropolitano Dom José Maria Pires, Santa Rita PB, Brazil.

Correspondence: Thaís Magalhães Lima Leite; Email: thaismlleite@gmail.com.

Case presentation: A 37-year-old female patient, diagnosed with anti-aquaporin-4 antibody positive Neuromyelitis Optica Spectrum Disorder (NMOSD) for 4 years, was on prednisone and azathioprine. She had sequelae from previous relapses, including grade IV+ paraparesis of the lower limbs and visual acuity loss in the left eye due to optic neuritis. Ten days prior to hospitalization, she began experiencing recurrent painful contractions in the left upper limb, described as mild internal shoulder rotation, forearm pronation, and wrist flexion lasting a few seconds with spontaneous improvement, without loss of consciousness. Three days later, she developed intractable nausea and vomiting, exacerbated postprandially. A cranial MRI revealed subtle signal alterations affecting the deep white matter of the corona radiata, posterior limbs of the internal capsules, and the periaqueductal region, showing hyperintensity in T2/FLAIR sequences without diffusion restriction or contrast enhancement. Additionally, a localized alteration was noted on the posterior surface of the medulla in the right median/paramedian area corresponding to the area postrema. Extensive cervical hyperintensity in T2/FLAIR sequences extended from C3-C4 to C6-C7, and another lesion with similar characteristics from D4-D5 measuring 2.8 cm without contrast enhancement. The patient underwent immunoglobulin therapy and was started on carbamazepine and antiemetic medications. She showed partial improvement in painful spasms and complete resolution of nausea and vomiting. **Discussion:** The presence of intense pain associated with tonic limb posture characterizes painful tonic spasms (PTS) or paroxysmal dystonia, common in NMOSD patients. Additionally, the area postrema syndrome, presenting with intractable nausea and vomiting, is a clinical syndrome that can occur in NMOSD. However, the onset of a relapse with the acute association of area postrema syndrome and painful spasms is atypical. Thus, recognizing this presentation is crucial for the early treatment of NMOSD relapses, preventing sequelae, and improving the patient's quality of life. **Final comments:** The onset of a relapse with the acute association of area postrema syndrome and painful spasms is atypical and rare. Therefore, recognizing this presentation is essential for the early identification and treatment of NMOSD relapses, preventing sequelae, and enhancing the patient's quality of life.

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Relapse rate and associated factors in multiple sclerosis: a Brazilian cross-sectional study

Nara Samara Silva dos Santos[1], Júlia Xavier Oliveira[1], Alex de Cerqueira Silveira Figueiredo[1], Maria Eduarda Messias Vassoler[1], Milena Fernandes de Oliveira[1], Luana Araujo Passos[1], Caio Fontes do Nascimento[1], Marcos Paulo Rodrigues Oliveira[1], Bruno Bacellar Pedreira[2]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Hospital Universitário Professor Edgard Santos, Salvador BA, Brazil.

Correspondence: Nara Samara Silva dos Santos; Email: nara.samara@ufba.br.

Background: Multiple sclerosis (MS) relapses are episodic exacerbations of neurological signs and symptoms lasting at least 24 hours in the absence of infection or fever. Although relapses help form part of the diagnostic criteria for MS, their incidence increases the chances of worse outcomes throughout the disease course. Therefore, identifying factors related to the occurrence of more relapses provides an opportunity to enhance disease management and improve functional outcomes. **Objective:** To investigate risk factors associated with relapse incidence in MS patients. **Methods:** This cross-sectional study was conducted on patients primarily diagnosed with RRMS based on the McDonald criteria, carried out from July 2021 to May 2024. Clinical and demographic characteristics were obtained through interviews and medical records review at a reference center in Bahia, Brazil. The relapse rate was stratified based on quartiles in three different groups: individuals experiencing a single relapse (0-50), those with 2 or 3 relapses (50-75), and those with 4 or more relapses (75-100). After checking the prerequisites, ordinal regression was performed to analyze the association

between sex, race, comorbidities, illness duration, visual and sensory symptoms at first relapse, diagnosis delay (>3 months), and the incidence of relapses before and after the MS diagnosis. **Results:** A total of 270 individuals were selected, of whom 87 (32.2%) were female. The median age at admission was 37.50 (29-46.25) and at disease onset was 30 (22-38). Of the patients included in the analysis, 73 (27.0%) had only one relapse, 117 (43.3%) had 2 or 3 relapses and 80 (29.6%) had more than 3 relapses. As for the clinical course, 268 had relapsing-remitting MS (RRMS) and 2 had secondary progressive MS (SPMS). In the ordinal regression, illness duration (OR 1.008 [95% CI 1.005-1.012], p=0.000) and visual symptoms at disease onset (OR 1.946 [95% CI 1.203-3.148], p=0.007) were the factors associated to a higher incidence of relapses, while younger age at onset (OR 0.957 [95% CI 0.935-0.979]) was related to fewer relapses (p=0.000). **Conclusion:** In this study, a longer time of illness was an important risk factor in the incidence of relapses, which is understandable, given the greater exposure to pathological activity. The impact of initial visual symptoms as a factor related to a higher relapse rate may point to optic neuritis as an indicator of a worse disease course, highlighting the importance of not neglecting it in clinical assessments and the need for more targeted studies to better understand this relation.

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Relation between childcare centers and neuropsychomotor development: an experience report from the "conviver" project

Amanda Mendes Santos[1], Elzilaine Domingues Mendes[1]

[1]Universidade Federal de Catalão, Catalão GO, Brazil.

Correspondence: Amanda Mendes Santos; Email: amanda.mendes@discente.ufcat.edu.br.

Background: Neuropsychomotor development is the result of various factors, including environmental ones. In this context, childcare centers promote socialization and are a space where children are introduced to language. At daycare, children make significant progress in their cognitive, emotional and motor development, as long as they are properly stimulated. There are, however, restrictions on the presence of health professionals in these spaces, who could help with early detection of risks of developing diseases, such as neurological ones. **Objective:** To report the experiences and observations made in a daycare center and the impact on children's development. **Methods:** In a daycare in a Brazilian municipality, weekly visits were made, lasting 2 to 3 hours during one semester. The classrooms observed were pre-school (children aged 2 to 4), and kindergarten (children aged 4 to 6). The actions belong to the "Conviver" extension project, which aims to detect developmental problems and promote children's health. **Results:** The project allowed the observation and contact with educators and children, in a space where they are far from their guardians. In each classroom, there was a teacher/caregiver and an assistant with incomplete secondary education. However, due to the large number of students, the caregivers were overworked and consequently imposed too many rules and limits, as well as being exclusively concerned with essential biological functions such as hygiene, feeding and sleep. There were no educational activities or games that could stimulate the children at an early age, mainly due to the teachers' working conditions. Children's interaction with their peers was also not encouraged, nor were group activities. In this sense, limitations in locomotion, socialization difficulties and deficits in language acquisition were noted among the children. It is worth noting that in all the classrooms, at least one child needed special care or was suspected of having neurodevelopmental disorders, such as autism spectrum disorder. In addition, another factor that possibly influenced the behavior and hindered the neuropsychomotor development of the children was the constant presence of the television on, exceeding the recommended daily hours for each age group. This meant that the children often stopped interacting and playing in order to watch TV programs. **Conclusion:** There was an excessive use of screens and a lack of stimuli, due to the overload of work by the staff, which leads to difficulties in socialization and neuropsychomotor development. Therefore, it can be concluded that the daycare space is extremely important for the child's development and could be improved, including hiring more teachers, in order to contribute to the child's integral health. The observations made in these places are fundamental for screening for signs and symptoms of possible psychopathologies and deficits in neuropsychomotor development, thus enabling early interventions.

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Relationship between anxiety and stroke: experience report from the world stroke campaing

Fernanda Liborio Pereira de Paula[1], Ana Carolina Chagas Alves da Silva[1], Euclair Benedito Cremasco[1], Sinesio Grace Duarte[1], Marina Rodrigues de Oliveira[1]

[1]Universidade de Franca, Franca SP, Brazil.

Correspondence: Fernanda Liborio Pereira de Paula; Email: fer2004fernandafer@hotmail.com.

Background: Stroke occurs when cerebral vessels become obstructed or ruptured, depriving brain tissue of nourishment. This is associated with various risk factors, including atherosclerosis, smoking, and hypertension, which in turn is correlated with generalized anxiety disorder making them related to stroke either directly or indirectly. **Objective:** To draw a parallel between generalized anxiety disorder and stroke in order to deepen research on such prevalent topics in today's world. **Methods:** From 2021 to 2023, in the month of October, stroke prevention campaigns were conducted. During these campaigns, risk factors, blood pressure, and glucose levels were assessed. The data was stored and analyzed to establish correlations. Additionally, searches were conducted in the PubMed, BVS, Elsevier ScienceDirect, and Scielo databases using the terms "high blood pressure and anxiety," "high blood pressure and stroke," and "stroke vs anxiety". **Results:** Anxiety disorder was found to be directly associated with stroke. Articles that relate anxiety to the uncontrolled management of chronic diseases such as hypertension, a factor already proven to be a risk for stroke, were found. Furthermore, during the campaign, blood pressure values were found to be above the reference value, already classified as stage 3 hypertension. **Conclusion:** Anxiety is a factor for the uncontrolled management of blood pressure, which leads to stroke and indirectly is an important risk factor. Moreover, as reviewed in the literature, this factor can directly result in stroke.

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Relationship between motor and respiratory function in Duchenne muscular dystrophy patients

Karlla Danielle Ferreira Lima, Leandro Augusto de Almeida[1], Tatiana Ribeiro Fernandes[1], Marco Antônio Veloso Albuquerque[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Karlla Danielle Ferreira Lima; Email: karlla.dfl@gmail.com.

Background: Duchenne muscular dystrophy (DMD) is a genetic disease caused by a mutation in the DMD gene, leading to the inability to produce dystrophin in skeletal muscle. As a result, there is a progressive loss of skeletal muscles, leading to progressive muscle weakness that begins in early childhood in boys, progressing to severe disability and loss of gait in adolescence, progressive cardiomyopathy and respiratory failure. Motor scales are important assessments to monitor the deterioration of motor function in these patients, with the MFM-32 being one of the most used scales. The MFM-32 assesses proximal, distal and axial motor dysfunctions, across the broad spectrum of neuromuscular diseases. The percentage of correct answers on this scale progressively reduces with the loss of motor function and progression of the disease. Respiratory failure, a later manifestation of the disease, is assessed mainly with forced vital capacity (FVC) and progressively reduces as the disease progresses, beginning a decline around 10 years of age. **Methods:** This is a single-center prospective observational study, carried out at the Muscle Clinic of a reference center, from January 2022 to July 2023. Twenty-four DMD patients between 0-18 years of age were evaluated for the correlation between lung function and motor function. All patients underwent spirometry to quantify FVC% and for the application of the MFM-32, that consists of 32 items divided into 3 domains (D1, D2, D3). **Results:** Among 24 evaluated DMD patients, 14 were able to walk. The FVC mean was 95% for the walking patients and 78% for the non-walking. MFM-32 motor scale scores were 28% (± 33.4) on D1, 84.9% (± 19.7) on D2, 86.1% (± 13.2) on D3 and 60.8 % (± 20.4) in total domain. Patients in the non-walking group had lower scores in all domains of the scale, but the age had statistical significance correlation only in domain D1 (p=0.006). When comparing lung function with age, we observed a statistically significant correlation between age and FVC% (p=0.026, r=-0.45), demonstrating that the older the patient, the lower is FVC%. The results of the correlation between the assessment of motor tests using the MFM-32 scale (D1, D2, D3 and total D) with pulmonary function (FVC) showed statistical significance only between the categories D1 (p=0.005), D2 (p=0.031) and total D (p=0.011) with FVC. **Conclusion:** This study shows that the group of non-walking DMD patients had worse results in MFM and FVC%. Only D1, D2 and D total of MFM32 had a significant correlation with FVC. These findings confirm the close relationship between worsening motor function and progression of the ventilatory pattern, which is in line with other studies in the literature.

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Relationship between the presence and subtypes of auras and extrahippocampal cortical and subcortical atrophy in patients with temporal lobe epilepsy associated with hippocampal sclerosis

Giovanna Gobbi Obalhe[1], Fernando Cendes[1], Luciana Ramalho Pimentel da Silva[1], Clarissa Lin Yasuda[1], Marina Koutsodontis Machado Alvim[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Giovanna Gobbi Obalhe; Email: g235613@dac.unicamp.br.

Background: Mesial temporal lobe epilepsy (MTLE), the most common form of focal epilepsy in adults, primarily stems from hippocampal sclerosis and affects the amygdala, thalamus, and entorhinal cortex. Seizures can initiate with brief focal perceptual events known as auras. **Objective:** This study explores the relationship between hippocampal and extra-hippocampal structural changes in MTLE patients with and without auras. **Methods:** 129 MTLE patients with unilateral hippocampal atrophy on Magnetic Resonance Imaging (MRI) were categorized into: (G1) exclusively autonomic auras, (G2) other auras (cognitive, emotional, sensory) with or without

autonomic auras, and (G3) no auras. Controls included 205 healthy individuals. MRIs were acquired using a 3T Achieva scanner with high-resolution 3D T1-weighted sequences and processed via Freesurfer 6.0 for subcortical volumes and cortical thickness in temporal regions and insula. We used SPSS Statistics V-26 for statistical analyses. We used the Chi-square test for categorical data and a generalized linear model with the Sidak post hoc test to analyze continuous data. Subcortical volumes included total intracranial volume as a covariate. We included age in the cortical thickness analysis based on the Akaike Information Criterion. **Results:** There were no significant age or sex distribution differences between patients and controls. G3 had 7.96±2.98 more years of epilepsy duration than G2. Subcortical volumetry showed atrophy in the thalamus ipsilateral to the seizure focus on all patients, with G3 showing further reduction than G1 and G2. The contralateral thalamus was reduced in G1 and G2 compared to controls and G3 compared to G2. The ipsilateral hippocampus was reduced in all groups compared to controls, with G3 additionally reduced compared to G1. The ipsilateral amygdala was reduced in G3 vs. controls and G2. The contralateral hippocampus and amygdala did not differ between patients and controls. Cortical thickness revealed ipsilateral middle temporal gyrus reduction in all groups, with G1 also showing contralateral reduction compared to controls. The ipsilateral superior temporal gyrus was reduced in all groups, with G3 also showing contralateral reduction vs. controls. Thickness reductions were observed only in the ipsilateral side: inferior temporal gyrus (G3 vs. controls), parahippocampal gyrus (G3 vs. controls and G1), and insula (G3 vs. controls). **Conclusion:** We identified group-specific structural changes. Only G2 and G3 had atrophy of the ipsilateral amygdala. All groups had atrophy of the ipsilateral hippocampus, thalamus, and the superior and middle temporal gyri, but it was worse in G3. In addition, G3 had more widespread atrophy, including the ipsilateral parahippocampal gyrus, insula, inferior and middle temporal gyri, contralateral thalamus, and superior temporal gyrus. In conclusion, MTLE patients with no auras had more pronounced and extensive extrahippocampal damage than those who reported auras.

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Reliability of perfusion measured by non-contrast magnetic resonance imaging (arterial spin labeling) in patients with ischemic stroke: a systematic review and meta-analysis

Jéssica Borges Kroth[1], Suzete Nascimento Farias da Guarda[2,3,4], Adriana Bastos Conforto[5]

[1]Universidade de São Paulo, São Paulo SP, Brazil.
[2]Harvard Medical School, Massachusetts General Hospital, Boston, USA.
[3]D'Or Institute for Research and Education (IDOR).
[4]Universidade Federal da Bahia, Salvador BA, Brazil
[5]Universidade de São Paulo, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Jéssica Borges Kroth; Email: jessicakroth@usp.br.

Background: Magnetic resonance imaging (MRI) with pseudocontinuous arterial spin labeling (PcASL) is a non-invasive technique that quantifies cerebral blood flow (CBF) and other hemodynamic parameters as hypoperfusion volume, diffusion/perfusion mismatch volume and mismatch ratio. It is possible to monitor these parameters in hours or days, without use of radiotracers or contrast, compared to dynamic susceptibility contrast (DSC), computed tomography (CT) perfusion and positron emission tomography (PET-Scan). **Objective:** This systematic review aims to verify the test-retest reliability of the pCASL technique in measuring CBF and to compare hemodynamic parameters measurements with those obtained with PET-Scan, CT perfusion and DSC in patients with ischemic stroke. **Methods:** This study was carrie out according to the PRISMA protocol and registered on PROSPERO platform (CRD42024539509). Three databases were systematically consulted by two independent reviewers and 245 records were accessed. The risk of bias and methodological quality was evaluated by Quality Appraisal for Reliability Studies (QAREL) in test-retest studies and Quality Assessment of Diagnostic Accuracy Studies (QUADAS-2) in studies comparing PcASL and the other imaging tests. **Results:** Test-retest reliability was measured in one study, PcASL was evaluated against DSC in eight studies and PcASL was contrasted with CT perfusion in two articles. Test-retest reliability study was classified as low quality with a high risk of bias. Excellent reliability of CBF was found across all assessed cerebral regions, particularly in white matter. Most studies comparing DSC and PcASL showed low risk of bias and low concerns regarding applicability. A meta-analysis of relative CBF (rCBF) and hypoperfusion volume is ongoing; data synthesis was not possible for other hemodynamic parameters due to insufficient reported values. All articles that compared CT perfusion with PcASL had low risk of bias and low applicability concerns. CBF was the only outcome analyzed in both included studies, but missing data prevented summarization. The other hemodynamic parameters were analyzed in only one study, indicating excellent correlation between CT perfusion and PcASL using rCBF thresholds of less than 35 to 45% for hypoperfusion volume and less than 40% for mismatch volume. **Conclusion:** There was good to excellent test-retest reliability of PcASL technique in measuring CBF in ischemic stroke patients and correlation between PcASL and CT perfusion values for hypoperfusion volume and mismatch volume. However, more studies are needed to confirm this result. Comparison between rCBF and hypoperfusion volume obtained with PcASL and DSC is still under verification through meta-analysis.

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REM sleep: definition and relevance

Gabriel Dalves Lauretti Betez[1], Antonio Augusto Faria Castro[1], Eduarda Antunes Giacometti[1], Nidia Ledur Muller[2], Catherine Arcaro Coghi[1], Larissa Rossetti Davanso[1], Isabela Moura de Oliveira[1], Enrico Saccomani Marcondes[1], Gabriela Lara[1], Maria Eduarda Zorer Meneghetti[1]

[1]São Leopoldo Mandic Araras, Araras SP, Brazil.
[2]Universidade de Rio Verde, Luziânia GO, Brazil.

Correspondence: Gabriel Dalves Lauretti Betez; Email: gabriel.betez@icloud.com.

Background: The stage of sleep known as REM is characterized by rapid eye movements, brain activity similar to wakefulness, and hypotonia. Since its discovery, it has been established that this phase is not merely a passive stage but is extremely important for memory consolidation, especially procedural and emotional memory. Moreover, it is strongly associated with emotions and brain development. Therefore, understanding what REM sleep is, its functions, and related areas of the nervous system is extremely important for medical practice to develop greater importance to a significant pillar of health: sleep. **Objective:** To review the evidence on the role of REM sleep in memory consolidation, emotional regulation, and physical health, highlighting the main brain structures involved and the impacts of deprivation of this sleep phase. **Methods:** This study is a narrative review. Research was conducted in the PubMed database, covering the period from 1969 to 2024. Keywords "REM sleep" and "memory" were used. Inclusion criteria involved studies addressing the role of REM sleep in memory consolidation, emotional regulation, and physical health.**Results:** REM sleep, or paradoxical sleep, is a crucial phase of the sleep cycle characterized by rapid eye movements, intense brain activity, and almost complete muscle atonia. Discovered in 1953, REM sleep revolutionized the understanding of sleep, showing it to be an active state with important functions for the brain and body. During this phase, the brain exhibits activity similar to wakefulness, essential for the consolidation of emotional and procedural memories. Deprivation of this phase can impair memory and the ability to learn new skills, increase irritability and anxiety. In the first months of life, REM sleep is predominant and vital for brain development, stimulating growth and plasticity. In adults, it plays an important role in creativity and problem-solving, facilitating the formation of new connections and insights. REM sleep is also associated with cellular recovery and tissue repair, being crucial for physical health. It regulates hormone production (cortisol). Furthermore, during REM sleep, the brain reorganizes information acquired during the day, facilitating learning and adaptation. Structures such as the brainstem, thalamus, and limbic system are activated during REM. Increased activity in various regions of the cerebral cortex during this phase explains vivid dreams and intense mental activity. **Conclusion:** Understanding the relevance of REM sleep is crucial for medical practice. This sleep phase is vital for memory, emotional regulation, brain development, creativity, and physical health. It activates important brain areas, facilitating learning and adaptation, as well as promoting cellular and hormonal recovery, essential for overall well-being.

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Remarkable response to capecitabine, trastuzumab, pertuzumabe and radiotherapy in a woman with brain metastasis from breast cancer

Fernanda Lustosa Cabral Gomez[1], Roberto Sobocinski Castro[1], Ligia Henriques Coronatto[1], Natalia Silva Fernandes[1], Carolina Simão Martini[2] , Heloisa Aurora Cavalcante Soares de Melo[1], Adrialdo José Santos[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[2]Hospital Santa Paula, São Paulo SP, Brazil.

Correspondence: Fernanda Lustosa Cabral Gomez; Email: fernandalustosacg@gmail.com.

Case presentation: A 48-year-old woman was diagnosed with breast cancer following an investigation of a painful breast nodule. Anatomopathological and immunochemistry revealed an invasive carcinoma grade 3, negative for calponin, progesterone and estrogen receptors; and positive for p63 protein and HER2 expression. Systemic staging was unremarkable at the time of diagnosis. The patient underwent mastectomy and lymphadenectomy, along with adjuvant trastuzumab and paclitaxel therapy. Radiotherapy was not performed due to clinical complications at that time. One year later, systemic restaging revealed multiple metastasis (lungs, liver and brain). Brain MRI revealed multiple contrast enhanced and non-enhanced lesions suggestive of metastatic disease. Whole brain radiotherapy (WBRT) was performed (10 sessions, 3Gy each). Anti-HER2 therapy was started with trastuzumab and pertuzumabe, along with capacitabin. Radiological response two months after WBRT and one month after introduction of target therapy was remarkable. The patient is currently asymptomatic and being followed up to assess maintenance of therapeutic response. **Discussion:** Breast cancer (BC) is the second most common cancer overall and approximately 30% of BC patients will develop brain metastasis (BM). Even though the prognosis and survival are typically lower in patients with BM, specific strategies have brought the possibility of better outcomes in this scenario due to the advent of targeted therapy. Trastuzumab is a monoclonal antibody directed against the extracellular domain of HER2. Usually, it does not cross the blood-brain-barrier (BBB) but, under impaired conditions, such as radiotherapy, trastuzumab levels in cerebrospinal

fluid increase. Preclinical studies also demonstrated the role played by trastuzumab as a radiosensitizer. Pertuzumab is a humanized monoclonal antibody that inhibits the dimerization of HER2 with other HER receptors. Our patient underwent therapy with Trastuzumab, Pertuzumab along with Capacitabin, a tumor-activated fluoropyrimidine carbamate with high activity in metastatic breast cancer. As for local treatment for BM, surgical resection is the standard treatment, but in our case the high number of lesions favored the use of WBRT. Symptomatic improvement with WBRT alone has been documented in 64-83% of patients, and median overall survival also improved. **Final comments:** In our case, we believe this remarkable response was due to radiotherapy increasing levels of trastuzumab and possibly contributing to a better effect of other targeted therapies; altogether with the evidence that trastuzumab enhances the effect of radiotherapy. It is possible that the high number of metastatic lesions also played a role in augmenting drug delivery through dysfunction of BBB. We have yet to observe if this imaging response will translate into more disease-free time, however our case highlights an exciting finding for future studies on combination of WBRT and targeted therapies.

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Renal biomarkers in the evaluation of patients with hereditary amyloidosis related to ttr gene mutation

Lara Albuquerque de Brito[1], Cleonisio Leite Rodrigues[2], Hermany Capistrano Freitas[2], Gdayllon Cavalcante Meneses[3], Alexandre Braga Libório[1], Raynrich Kevin Assis Lima[1], Fernanda Martins Maia Carvalho[1]

[1]Universidade de Fortaleza, Fortaleza CE, Brazil. [2]Hospital Geral de Fortaleza, Fortaleza CE, Brazil. [3]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Lara Albuquerque de Brito; Email: laraalb.brito@hotmail.com.

Background: Hereditary amyloidosis is a disease characterized by the accumulation of abnormal proteins called amyloids in body tissues, including the kidneys. Renal biomarkers detectable in blood and urine can provide information about the presence and severity of kidney involvement in various diseases. **Objective:** To evaluate renal biomarkers, such as NGAL, MCP-1, urinary creatinine, and nephrin, and endothelial biomarkers, including VCAM, ANG-2, and syndecan-1, in patients with hereditary amyloidosis associated with the TTR gene. Additionally, to detail the profile of these biomarkers, comparing them with a healthy population. Clinical findings, especially neurological and renal manifestations, will be described to assess the degree of disease involvement. **Methods:** In this cross-sectional study, we evaluated 25 patients with pathogenic mutations in the TTR gene, investigating renal biomarkers that may provide new information about ATTR-related nephropathy. The biomarkers evaluated include NGAL, MCP-1, urinary creatinine, nephrin, VCAM, ANG-2, and syndecan-1. The biomarkers were quantified using the ELISA method. **Results:** Urinary creatinine was observed to be reduced in patients with the TTR gene compared to the control group. Regarding MCP-1, no specific pattern was observed, with similar results found between patients and the control group. Concerning syndecan-1, 48% of patients with the TTR gene had values above 155, whereas in the control group, 26% had values above 100. It was noticed that VCAM values in patients with the TTR gene were below 1300 (84%), whereas in the control group, 100% had values above 1600. **Conclusion:** The study is still ongoing, and some biomarkers have not yet been analyzed. However, with the current data, we can already observe that renal biomarkers are important tools to stage hereditary amyloidosis in association with clinical symptoms. In patients studied with the TTR gene compared to the control group, clearer abnormalities were observed, particularly in the biomarkers urinary creatinine, syndecan-1 and VCAM. We hope that by the end of the research, it will be possible to identify biomarkers associated with the progression and severity of the disease. Furthermore, it is important to emphasize the significance of studies in this area to ensure better care and management for these patients.

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Renal predictors of mortality in patients with wake-up stroke

Juliana Cerqueira Gomes[1], Aurea Maria Lago Novais[1], Marina Soledad Benitez Bitencourt[1], Renan Carvalho Castello Branco[1], Thaise Braga de Oliveira[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Juliana Cerqueira Gomes; Email: juligomesc5@gmail.com.

The literature indicates that renal function increases stroke risk and predisposes to poorer outcomes. Wake-up stroke patients often miss the thrombolysis window, potentially worsening outcomes. The association between renal function and mortality in wake-up stroke patients is not well-established. This cross-sectional cohort study aims to assess the influence of renal function on mortality in wake-up and determined ictus stroke patients. The data collected included prior renal insufficiency, dyslipidemia, diabetes, ASPECTS, atrial fibrillation, sex, obesity, alcohol use, smoking, hypertension, admission, peak, and discharge creatinine and urea levels. The outcome was death during hospitalization. Univariate analysis was performed using T-test/Mann-Whitney/chi-square tests, followed by multivariate logistic regression (p< .05).A total of 451 patients were evaluated, of which 23% were diagnosed with wake-up stroke. Among the wake-up stroke patients, the following pre-existing characteristics were observed: 52% were male, 12.5% had obesity, 19% had a history of alcoholism, 20% were smokers, 65% had hypertension, 30% were diabetic, 2% had renal insufficiency, 3% had atrial fibrillation, and 10% had dyslipidemia.Regarding renal function markers, wake-up stroke patients had a median creatinine level at admission of 1.0 mg/dL (IQR 0.85-1.2), while patients with definite ictus had a median of 1.0 mg/dL (IQR 0.83-1.2). The median peak creatinine was 1.1 mg/dL (IQR 0.9-1.4) in wake-up stroke, compared to 1.2 mg/dL (IQR 0.9-1.5) in definite ictus patients.The median discharge creatinine was 1.06 mg/dL (IQR 0.8-1.2) in wake-up stroke, compared to 1.03 mg/dL (IQR 0.9-1.1) in definite ictus patients.As for urea levels, the admission values had similar medians of 34 mg/dL (IQR 26-47) between the groups, while the peak values had medians of 39 mg/dL (IQR 33-77) for wake-up stroke and 48 mg/dL (IQR 33-72) for definite ictus patients. At discharge, the urea medians were similar between the groups, with 36 mg/dL (IQR 25-48) for wake-up stroke and 36 mg/dL (IQR 27-50) for definite ictus.In univariate analysis, there was a significant difference for previous dyslipidemia (p=0.01) and the ASPECTS score (p=0.04, median 9, IQR 7.5-10). In multivariate analysis, the ASPECTS score (p=0.06; OR=1.2, 95% CI 0.9-1.6) and the discharge urea levels were identified as independently associated with patient mortality (p=0.01; OR=1.01, 95% CI 0.98-1.05).Renal function, particularly discharge urea levels, predicted mortality in wake-up stroke patients. Despite similar median urea values between wake-up stroke and definite stroke groups, discharge urea levels independently correlated with mortality, reasons for which are not fully understood.Additionally, a higher ASPECTS score was associated with a 1.2-fold increased risk of death in wake-up stroke patients.This study underscores the significance of discharge urea levels as a critical prognostic indicator in wake-up stroke patients.

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Renal predictors of mortality in stroke patients with risk of obstructive sleep apnea: a transversal study

Juliana Cerqueira Gomes[1], Aurea Maria Lago Novais[1], Marina Soledad Benitez Bitencourt[1], Renan Carvalho Castello Branco[1], Thaise Braga de Oliveira[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Juliana Cerqueira Gomes; Email: juligomesc5@gmail.com.

Renal factors are central in disease prognosis due to their modifiability. Despite extensive research on the link between stroke and obstructive sleep apnea (OSA), the influence of renal factors on outcomes remains unclear. This cross-sectional cohort study aimed to examine how varying OSA risks affect renal factors in stroke outcomes, including hemorrhagic transformation, delirium, infection, and mortality.Patients' OSA risks were assessed using STOP-BANG and SOS scores, with data collected on prior renal insufficiency and creatinine/urea levels at admission, peak, and discharge. Statistical analyses comprised univariate tests (T-test, Mann-Whitney, chi-square) followed by multivariate logistic regression (p<0.05). We obtained data from 434 patients (69% high risk with STOP-Bang and 50% with SOS).For the STOP-BANG, the median creatinine level at admission was 0.8 mg/dL (IQR 0.71-0.94) for low-risk patients and 0.9 mg/dL (IQR 0.77-1.22) for high-risk. The median creatinine level at discharge was 0.9 mg/dL (IQR 0.6-1.01) for low-risk patients and 1 mg/dL (IQR 0.7-1.4) for high-risk. The median peak creatinine level was 0.9 mg/dL (IQR 0.8-1.1) for low-risk patients and 1.4 mg/dL (IQR 1-2) for high-risk.Regarding urea levels, the median urea level at admission was 32 mg/dL (IQR 28-53) for low-risk patients and 31 mg/dL (IQR 25-53) for high-risk. The median urea level at discharge was 33 mg/dL (IQR 24-55) for low-risk patients and 44 mg/dL (IQR 26-82) for high-risk. The median peak urea level was 55 mg/dL (IQR 31-81) for low-risk patients and 63 mg/dL(IQR 36-113) for high-risk patients.Univariate analysis showed admission, peak and discharge creatinine (p<0.01), as well as urea admission (p=0.05), discharge (p<0.01), and peak (p=0.001) were significant. For SOS score, discharge urea levels differed significantly (p=0.02).Multivariate analysis for STOP-BANG found peak urea levels were significantly associated with mortality (p=0.01;OR=0.9, 95% CI 0.90-0.94) infection (p 0.01;OR=1.0, 95% CI 0.94-0.97) and delirium (p<0.01;OR 0.9 95% CI 0.95-0.98) with admission urea also linked to infection (p=0.04;OR=1.01, 95% CI 1.0-1.03). Also, diabetes was associated with delirium (p=0.03;OR 1.8 95% CI 1-3). For the SOS score, discharge urea was associated with mortality (p=0.02;OR=0.9, 95% CI 0.97-1.05), while admission urea (p=0.04;OR=1.0, 95% CI 1-1.03), peak urea (p<0.01;OR=0.9, 95% CI 0.94-0.97) and diabetes (p=0.03;OR 1.8 95% CI 1-3) were linked to delirium.Hemorrhagic transformation was not significant.High-risk patients exhibited better urea values at admission but worse at discharge, suggesting a physiological mechanism linked to OSA, possibly influenced by hypertension and increased metabolic waste, that may contribute to worse outcomes.The analysis indicates that high urea levels, including discharge levels may act as protective factors against certain complications. Additionally, diabetes is an established risk factor for worse post-stroke outcomes.

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Repercussions of the use of oral contraceptives in women with migraine

Maria Eduarda Julião Leite[1], Artur Bruno Silva Gomes[1], Cesário Silva Souza[1], Maria Eduarda Dantas Arruda Silva[1]

[1]Centro Universitário de Maceió - Afya, Maceió AL, Brazil.

Correspondence: Maria Eduarda Julião Leite; Email: duddaleite1@outlook.com.

Background: Migraine is a disabling pain that can be associated with nausea, emesis and photo/phonophobia, lasting between 4 and 72 hours. This condition is three times more prevalent in women than in men, with a peak incidence (24%) occurring during the fourth decade of life. This is due to hormonal influences during the reproductive cycle, related to fluctuations in estrogen levels. **Objective:** To analyze the repercussions of oral contraceptive (OC) use in women with migraine. **Methods:** This is a cross-sectional observational case-control study carried out on a sample (n=25) of females aged between 18 and 60 over a 12-month period at the Doctor José Lages Filho Teaching and Care Unit (UDA) of the Centro Universitário de Maceió, Alagoas. The Migraine Disability Assessment (MIDAS) questionnaire was used to quantify the disability caused by headache over a 3-month period. The results were classified into 4 levels of severity. Level I - minimal or infrequent; level II - medium; level III - moderate and level IV - severe. Inclusion criteria: women with migraine without aura, of childbearing age and in the climacteric period, followed up at the UDA. Exclusion criteria: women using other hormonal contraceptive methods and smokers. It was approved by the Research Ethics Committee, under Certificate of Presentation for Ethical Appraisal: 70105023.2.0000.5641 and opinion: 6.163.274. **Results:** Of the 25 women interviewed, 12 were selected to be part of the sample and 13 were excluded because they had migraine with aura. Of the sample, 66.7% had used OACs at some point in their lives but did not currently use them; 25% had never used them and 8.3% used them continuously. Of the patients evaluated, 44.4% of them reported worsening of their attacks during the period in which they were using OCPs; 62.5% mentioned improvement after stopping the drug and only 25% showed worsening after stopping it. No patient reported an improvement in migraine attacks while taking the OCP. In the assessment of the disability caused by the headache, it was observed according to the MIDAS classification that 33.3% were categorized as grade I and II; 16.7% as grade III and IV, respectively. The study's limitations involved patients after menarche, as the sample only included women over 18 years of age and did not include women in the postpartum period. **Conclusion:** The group investigated showed that women who had never used OCPs or had already used them and had stopped or were post-menopausal had less disabling migraine attacks than patients who were using the drug. Those who were using OCP showed a worsening of their attacks, which indicates that OCP acts as a potentiator of migraine. It was also observed that post-menopausal women had the lowest rates of disability compared to menopausal women.

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Report of a series of patients with mosaic trisomy 18: Edwards syndrome

Maria Fernanda Peruci Felippe[1], Viviane Müller[1], Fabiana Dolovitsch de Oliveira[1], Pedro Lorenzo Neves da Silva[1], Roberta Beiser de Medeiros e Albuquerque[1], Aline Deborah de Moraes[1], Maria Eduarda Corrêa Pereira[1], Rafaela Gageiro Luchesi Soares[1], Amanda Vieira Alves[1], Thiago Menezes Cézar[1]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Maria Fernanda Peruci Felippe; Email: maria.felippe@ufcspa.edu.br.

Background: Trisomy 18 (T18) Edwards syndrome is characterized by a broad and variable clinical picture, usually associated with neurological symptoms and a poor prognosis. **Objective:** This abstract aims to describe the clinical findings of a sample of patients with mosaic T18. **Methods:** The clinical and cytogenetic findings of patients with mosaic T18 were evaluated in a Clinical Genetics Service from 1975 to 2008. All cases were diagnosed in the service's laboratory through karyotyping from peripheral blood samples. **Results:** During the study period, 50 patients were diagnosed with T18, 5 of which (10%) had mosaicism. The average number of cells analyzed was 27,8. The percentage of cells in the lineage with T18 ranged from 9 to 87%. Three of the 5 patients (60%) were male. Age at assessment ranged from 14 to 5,926 days (median 93 days). Attention was drawn to the small number of clinical findings described, both in the dysmorphological assessment and through complementary imaging exams. The main changes observed were micrognathia (n=3), low-set (n=2) and retroverted ears (n=3), clenched fists with overlapping fingers (n=2) and congenital clubfoot (n=1). Regarding internal organs, congenital heart disease was reported in 2 cases (40%), including one patient with tetralogy of Fallot. All patients had a history of delayed neuropsychomotor development. The older patient also presented cognitive deficits and seizures. **Conclusion:** The clinical presentation of our patients is consistent with that described in the literature, as they draw attention to the small number of alterations, including major malformations, such as congenital heart disease. However, neuropsychomotor delay and neurological symptoms are considered constant findings. Therefore, pediatricians and neurologists especially should be very aware of this possibility in their clinical practice.

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Report of two new cases of the extremely rare Cluster-Tic syndrome

Cassio Mota Freitas da Silva[1], Hilton Mariano da Silva Junior[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Cassio Mota Freitas da Silva; Email: cassiomotafds@gmail.com.

Case presentation: Case 1: A 72-year-old previously healthymandeveloped episodes of intense, stabbing pain in the right malar region of very short duration, without associated autonomic phenomena, triggered by touching, speaking, and chewing. He experienced pain remission after ten days of using carbamazepine. Soon after, attacks of intense pain began in the right frontotemporal and retro-orbital region, lasting 15 to 40 minutes, associated with tearing, hyaline coryza, eyelid ptosis, and ipsilateral nasal obstruction. The crises were not daily, but up to 3 episodes occurred in 24 hours. He presented significant weight loss and social isolation. A neurological examination during the interictal period and a brain MRI showed no changes. He was associated with verapamil and carbamazepine, progressing to remission of symptoms after three months. He has been followed for one year. Case 2: A 70-year-old woman started 25 years ago with episodes of intense stabbing pain in the right hemiface, lasting around an hour, associated with tearing, hyaline runny nose, and nasal obstruction, which showed a good response to the use of prednisone and verapamil. She remained asymptomatic for six years until the attacks returned and began to alternate with episodes of "shock pain" in the right malar region of strong intensity, without other associated phenomena, and lasting around 60 seconds. The radiological investigation was unremarkable. The patient had a good response to the combination of carbamazepine and verapamil. She has been followed up in our service for four years. **Discussion:** Cluster-Tic syndrome is a rare clinical condition characterized by cluster headaches associated with trigeminal neuralgia, with ipsilateral symptoms. In these cases, the maxillary division of the trigeminal nerve was affected, and there were similar symptoms of trigeminal neuralgia. It is noteworthy that the female patient had shock pain without associated factors, which is unusual in the classification of trigeminal neuralgia and for a much longer period than the male patient. Finally, is observed that the most efficient treatment of this is the combination of the drugs verapamil for cluster headache and carbamazepine for trigeminal neuralgia. **Final comments:** Rare and exceptionally debilitating, the syndrome that combines cluster headache and trigeminal neuralgia represents a profound challenge in the field of neurology. Both conditions are independently recognized as among the most painful disorders known to medicine.The first case suggests that the cluster headache appeared due to unsuccessful treatment of trigeminal neuralgia, given the short time interval between the appearance of symptoms. On the other hand, the second case indicates a lack of interrelationship between them due to the long asymptomatic period and the alternation of symptoms. Thus, our two new cases report shed light on the discussion about the legitimacy of the hypothesis of common pathophysiology in this syndrome.

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Resistance training can influence episodic memory performance in elderly people with mild cognitive impairment

Isadora Cristina Ribeiro, Camila Vieira de Ligo Teixeira[2] , Gabriel Barzon Silva[1], Marco Carlos Uchida[1], Ítalo Karmann Aventurato[1], Marjorie Cristina Rocha da Silva[1], Brenda Costa Gonçalves[1], Ana Luiza Gonçalves Rochetti[1], Paula Teixeira Fernandes[1], Marcio Luiz Figueredo Balthazar[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]West Virginia University, Rockefeller Neuroscience Institute, West Virginia, USA.

Correspondence: Isadora Cristina Ribeiro; Email: isadora.cr@hotmail.com.

Background: Mild cognitive impairment (MCI) refers to the diagnosis of individuals between preserved cognition and dementia who have a decline in one or more cognitive domains (memory, attention, executive functions, language, perception, visuospatial skills) and are at risk of developing dementia. Stimulating increased mental activity has been shown to be beneficial in altering the course of disease progression. Therefore, non-pharmacological interventions such as physical exercise have been explored as a tool to promote cognition in older adults with mild cognitive impairment. The increase in muscle strength was correlated with better cognitive performance in elderly people with MCI and decreased risk of developing dementia in elderly with preserved cognition. Resistance training's main **Objective:** To increase muscle strength. Thus, this study aimed to compare the performance of elderly people with mild cognitive impairment on different cognitive tests before and after 24 weeks of resistance training. **Methods:** This investigation includes 22 elderly people aged 65.64±6.15 years old (11 men and 11 women) and with Mild Cognitive Impairment. The diagnosis was based on NIA-AA criteria, which include complaints of cognitive changes, evidence of impairment in one or more cognitive domains, preservation of independence in functional activities, and not meeting the criteria for dementia. The participants practiced 24 weeks of resistance training consisting of 3 sets of 10 repetitions in 10 exercises for a large muscle group twice a week and with an intensity of 80% of one maximum repetition. Participants were assessed for cognitive performance before and after 24 weeks of resistance training using the following tests: RAVLT (List A6 and B1), RAVLT Recognition, Rey Complex Figure (late recall), and Memory Complaint Scale. A neuropsychologist applied the tests, and the paired T-test was used for statistical analysis in the IBM SPSS Statistics 22 software.

Results: The comparison test between the baseline and the end of the intervention showed a significant difference in the RAVLT (List A6): p=0.04 and a tendency towards significance in the RAVLT Recognition: p=0.05. No other cognitive test show significant differences: RAVLT (List B1): p=0.78, Rey Complex Figure (late recall): p=0.82, Memory Complaint Scale: p=0.06. The statistical analysis was performed using the paired T test in the IBM SPSS Statistics 22 software. **Conclusion:** Resistance training favored the learning, recall, and recognition of episodic memory in elderly people with mild cognitive impairment, suggesting that resistance exercise is beneficial for the cognition of these individuals at increased risk of developing dementia.

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Restless legs syndrome/Willis-Ekbom disease in Parkinson's disease: a cross-sectional analysis

Agatha Lopes Azedo[1], Diego Monteiro de Carvalho[2], Marcus Vinicius Della[3], Alvaro Siqueira da Silva[2], Marcello Facundo do Valle Filho[2], Raissa Costa Said[1], Emillaine Alves Noronha[1], Mariana Carvalho Ferreira[1], Yenly Gonzalez Perez[2], Carlos Mauricio Oliveira de Almeida[2]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Agatha Lopes Azedo; Email: agathalopesazedo@gmail.com.

Background: Parkinson's disease (PD) is a neurodegenerative disorder that primarily affects the substantia nigra pars compacta and basal ganglia, leading to the characteristic motor symptoms of the disease: bradykinesia, resting tremor, and muscle rigidity. Sleep disturbances are present in 90% of patients with PD, commonly including excessive daytime sleepiness (EDS) and Restless Legs Syndrome (RLS). These sleep disorders contribute to a deterioration in quality of life, complicate the treatment of PD, and can worsen its prognosis. The validated Parkinson Disease Sleep Scale (PDSS-1) includes questions about sleep complaints in PD, with total scores ranging from 0 to 150, where higher scores indicate better sleep quality. Another widely used tool is the Epworth Sleepiness Scale (ESS), which assesses the presence of EDS through 8 items measuring the propensity for sleep in various daily situations. **Objective:** To analyse the prevalence of Restless Legs Syndrome in patients with Parkinson's Disease and their quality of life using the PDSS and ESS questionnaires. **Methods:** The study was approved by the local Ethics Committee under CAAE nº 58684622.2.0000.0007. This was an observational, cross-sectional study conducted at a referral hospital in Manaus, Amazonas, over a period of 24 months. Patients with Parkinson's Disease aged > 18 years, diagnosed according to the London Brain Bank criteria, were included, while patients with diagnoses of dementia, language disorders (aphasia, dysphasia), or speech articulation disorders (dysarthria, hypophonia) that precluded the application and understanding of the questionnaires were excluded. **Results:** Of the 63 individuals selected, 43 (68.5%) were diagnosed solely with PD and 20 (31.5%) had concurrent RLS. The average PDSS score was 74.8, with a median of 71. The mean PDSS score for patients with only PD was 80.1, while for those with both PD and RLS, it was 62.3. Regarding the presence of excessive daytime sleepiness among the 63 patients, 41 (65.1%) had EDS and 22 (34.9%) did not. Among those with EDS, 13 (31.7%) also had RLS, and 28 (68.3%) had only PD. Among patients without EDS, 7 (31.9%) had both PD and RLS, while 15 (68.1%) had only PD. The sample had a p-value of 0.782. **Conclusion:** There is a high prevalence of Restless Legs Syndrome/Willis-Ekbom Disease in patients with Parkinson's Disease. Overall, there is a decline in the sleep quality of patients, which is more pronounced in those with both diagnoses, as evidenced by the nearly 20-point decrease in PDSS scores between the two populations. Additionally, patients with PD tend to be sleepier, and the coexistence of both conditions further exacerbates this, as shown by the fact that within the PD and RLS group, the number of patients with EDS according to the ESS is nearly double those without EDS.

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Rethinking hot cross bun sign: an uncommon finding on spinocerebellar ataxia type 2

Letícia Klabinske Marques Monteiro[1], Paulo Felipe Bezerra Silva dos Santos[1], Bruno Henrique Carneiro Costa Filho[1], Victor de Mores Chagas[1], Evelliny Gomes da Silva[1], Allefy Beltrão Albano[1], Ana Eunice Oliveira Rodrigues[2], Giovanna da Silva Gamba Dias[2], Miriam Carvalho Soares[2], Marcos Eugênio Ramalho Bezerra[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Leticia Klabinske Marques Monteiro; Email: leticiaklabinske@gmail.com.

The hot cross bun sign is a neuroradiological MRI finding, best visualized in T2W or T2 FLAIR sequences, mostly associated with Multiple System Atrophy Cerebellar type (MSA-C). However, over recent years, various etiologies have been associated with this radiologic finding, including neurodegenerative, genetic, autoimmune, infectious, inflammatory, neoplastic, and vascular causes. Its association with spinocerebellar ataxias (SCA) is uncommon although well documented. **Case presentation:** We present two cases of female patients, sisters, aged 29 and 32 years, with progressive postural instability and imbalance since the ages of 21 and 22, respectively. They had no relevant past medical history, except their family history of five people with a similar disorder, including their mother, maternal grandfather, two uncles, and one cousin, suggesting a genetic disease with an autosomal dominant inheritance. Neurological examination of both patients included bilateral dysmetria, scanned speech, slow saccadic eye movements, postural instability, and broad ataxic gait. No parkinsonism or dysautonomia were reported. Their Scale for the Assessment and Rating of Ataxia (SARA) scores slowly worsened over time, indicating the progressive and degenerative nature of the disease. Brain MRI of both patients revealed the hot cross bun sign, along with cerebellar atrophy. Genetic evaluation of one of the sisters disclosed a CAG trinucleotide expansion in one allele of the ATXN2 gene (41 CAG repetition, with a normal range below 31), confirming spinocerebellar ataxia type 2 (SCA2) diagnosis. **Discussion:** Cerebellar atrophy is the most common radiological finding among SCAs. Other radiological changes in SCA patients may more commonly appear after about five years of the onset. Among SCA patients, the hot cross bun sign was most frequently seen in patients with SCA2 and has already been documented in types 1, 3, 6, 7, 8, and 34. The possible pathophysiological process for this finding is due to the atrophy of the pontine neurons and transverse pontocerebellar fibers, with preservation of the pontine tegmentum and corticospinal tracts. This alteration occurs regardless of the length of the CAG repeat expansion, with age-related atrophy mainly in SCA2 and SCA7, contributing to a higher prevalence in these subtypes. **Final comments:** Despite its close association with MSA-C, the hot cross bun sign is not pathognomonic to this etiology. The association of this sign with SCA2, although uncommon, must be considered in ataxia patients, particularly in cases with a positive history, slow saccades, and absence of parkinsonism or dysautonomia.

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Retinal neurodegeneration and brain atrophy in patients with Chagas disease: a cross-sectional study

Catharina Ribeiro Lyra[1], Pedro Fernandes Abbade[1], Débora Nalígia Moraes Luna[2], Ana Beatriz Cazé Cerón[1], Bruno Felipe Santos de Oliveira[1], Graziele Carolina Araújo Soares[1], Bruna Helena Ribeiro Araújo dos Santos[1], Rebeca Souza Borges[1], Thiago Cerqueira-Silva[3], Jamary Oliveira Filho[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.
[2]Oftalmodiagnose, Salvador BA, Brazil.
[3]Fundação Oswaldo Cruz, Salvador BA, Brazil.

Correspondence: Catharina Ribeiro Lyra; Email: catharinarlyra@gmail.com.

Background: Chagas disease (CD) is associated with brain atrophy and cognitive impairment. Retinal optical coherence tomography (OCT) may be a noninvasive method to detect both outcomes but has not been previously studied. **Objective:** To investigate the association between retinal thickness and foveal vascular density with volumes of the central nervous system in patients with CD. **Methods:** Cross-sectional study performed in 2020-2024, with adult heart failure patients from a cardiology outpatient clinic in Salvador-Brazil. We excluded those with anticoagulants, metallic prostheses, cancer, chronic liver disease, dialysis, stroke, glaucoma, diabetes, or retinal diseases. CD was defined by a positive ELISA or immunofluorescence test in duplicate. Eligible participants underwent MRI to assess central nervous system (CNS) volumes using FreeSurfer software and manual counting of lacunar and territorial infarcts. Patients underwent OCT to evaluate macular, ganglion cell layer, and retinal nerve fiber layer thickness and the vascular density in the fovea at the superficial and deep plexus. OCT with quality <50% were excluded. We performed multivariate logistic regression between OCT parameters and CNS volumes, controlling for age, sex, CD, left ventricular ejection fraction (LVEF), and intracranial volume. **Results:** We included 36 participants with CD and 29 without CD. The CD group had more females (81% vs 55%) and preserved LVEF (81% vs 30%), but other clinical variables were similar. Foveal macular thickness was lower in patients with CD (230 vs 242 µm, p=0.02). Its reduction was associated with a decrease in the volume of the nucleus accumbens (β=0.62, p<0.01), putamen (β= 3.51, p<0.01), parahippocampal cortex (β=1.01, p<0.01), thalamus (β=3.53, p=0.01), cerebellum (β=17.34, p=0.01), globus pallidus (β=1.35, p<0.01), ventral diencephalon (β=2.1, p=0.01), lateral orbitofrontal cortex (β=1.32, p=0.04), and entorhinal cortex (β= 2.6, p< 0.01). The foveal vascular density in the superficial plexus was lower in patients with CD (13.4 vs 16.9%, p=0.022). Its reduction was associated with an increase in the volume of the posterior cingulate cortex (β=-4.84, p=0.03), and a decrease in the transverse temporal cortex (β=3.26, p=0.01), and the number of territorial infarctions (β=0.06, p=0.03). Foveal vascular density in the deep plexus was lower in patients with CD (26.6 vs 31.1%, p=0.031), and its reduction was related to a decrease in the volumes of the cortex in the following regions: caudal middle frontal (β=8.02, p<0.01), rostral anterior cingulate (β=4.26, p<0.01), superior temporal (β=8.85, p<0.01), lateral orbitofrontal (β=5.97, p<0.01), middle temporal (β=7.13, p=0.02), and superior parietal (β=15.27, p<0.01). **Conclusion:** Retinal thickness and foveal vascular density correlate with CNS volumes in patients with CD. This contributes to the understanding of the effects of CD in the CNS and might be used as an easy, quick and noninvasive marker of brain atrophy.

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Retrospective analysis of data from thrombectomies performed from january to march 2024 in a public hospital in João Pessoa/PB

Gilmar Leite Pessoa Filho[1], Ana Luisa Castelo Branco Gomes[1], Felipe Nunes de Araújo[1], Luciano Gonçalves do Nascimento Júnior[1], Rafael de Souza Andrade[1], Juliana Magalhães Leite[1], Matheus Gurgel Saraiva[1], Paulo Antonio Farias Lucena[1], Apolônio Peixoto de Queiroz[1], Rafael Gonçalves Duarte Cunha[1]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.

Correspondence: Gilmar Leite Pessoa Filho; Email: gilmarpessoa@gmail.com.

Background: Stroke is one of the leading causes of death and morbidity worldwide. Therapeutic advances in the treatment of acute stroke have changed outcomes. Mechanical thrombectomy has emerged as an effective treatment for the acute stroke. Although its benefits and safety have been proven for over 8 years, it was implemented in Brazil's public health system less than 1 year ago. **Objective:** Analyze epidemiological data and variables related to mechanical thrombectomies performed at a quaternary public hospital in João Pessoa/PB, Brazil. **Methods:** This is a cross- sectional, descriptive study based on the retrospective analysis of medical records of 18 patients who underwent mechanical thrombectomy between January and March 2024. The following variables were analyzed: age, sex, chemical thrombolysis, symptom-to-femoral puncture time, puncture-to-recanalization time, TICI, ASPECTS, NIHSS, pre- and 90-day mRS, symptomatic hemorrhage, occluded vessel, time of hospitalization, technique used, and admission-to-puncture time. **Results:** The mean age was 60.1 years (33-79), with a sex ratio of 1:1. Only 2 patients (11.1%) underwent both thrombolysis and thrombectomy. The average time from symptom onset to femoral puncture was 10.8 hours (2h-22.5h), with only 4 patients (22.2%) presenting less than 6 hours. The mean NIHSS was 15.8 (8-31). Only 1 patient had a prior modified Rankin Scale (mRS) score of 1 and another scored 2; all others had a previous mRS zero before admission. The mean mRS at 90 days was 4.2. Six patients (33.3%) died, and only 2 (11.1%) achieved functional independence at 90 days (mRS of 0 to 2). Symptomatic intracranial hemorrhage occurred in 2 cases (11.1%). Regarding the occluded vessel, 8 (44.4%) were in the M1 segment of the middle cerebral artery, 7 (38.8%) in the intracranial internal carotid artery, 3 (16.6%) in the basilar artery. The mean time from hospital admission to femoral puncture was 172.5 minutes (60-280). The average time from femoral puncture to vessel recanalization was 38.8 minutes (5-140). The average length of hospital stay was 21 days (3-52). Regarding procedural technique, stent retriever was used in 7 cases (38.8%), aspiration in 5 (27.7%), and a combination of both in 7 (38.8%). On the reperfusion scale, 11 (61%) achieved TICI 3, 4 (22%) TICI 2b, only 1 (5%) TICI 2a, and another TICI 1. The 90-day outcome showed that only 11% achieved functional independence (mRS 0-2). **Conclusion:** Mechanical thrombectomy has revolutionized the treatment of acute stroke, resulting in improved functional recovery. In the Brazilian Unified Health System (SUS), the availability of this treatment remains limited. Additionally, the indications for the procedure were restricted until recently. It is expected that increased access to this therapy and expanded indications will improve patient clinical outcomes.

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Rett's syndrome: a case report

Antonio Marcos Reissureição Galindo[1], Rodrigo Pernomian Cianca[1], Antonio Paulo Felice Rodrigues Leite[1], José Guilherme Pinhatti Carrasco[1], Felipe Bento Michelato[1], Maria Júlia Costa Rocha[1], Giovanna Volpato Segura[1], Emanuel Gomes Ferreira[1], Yves Henrique Ramos Mansano[1], Thiago Honório Dutra da Silva[1]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.

Correspondence: Antonio Marcos Reissureição Galindo; Email: antoniomrgalindo@gmail.com.

Case presentation: A Caucasian female patient was born at term at 40 weeks of gestation, cesarean section. The mother made adequate use of ferrous sulfate and folic acid supplementation during pregnancy. After neonatal screening, there were no significant changes in the newborn. The mother reports a perception of the delay in the child's neuropsychomotor development. At the age of seven, the patient was admitted to an Urgency and Emergency Unit complaining of muscle spasms and hypoactivity, with her oxygen saturation at 92%. The patient developed respiratory failure, bronchospasms, and recurrent epileptic seizures with clonic movements lasting more than one minute and postictal period of more than ten minutes. There was the presence of sialorrhea, muscle stiffness in the lower and upper limbs, feet in internal rotation and claw-shaped hands. Also, acquired chest deformity and scoliosis. Also, low body development and aphasia, but with a perception of understanding of what is said to the patient. The patient's diagnosis was made through the sum of the associated clinical signs and confirmed by the sequencing of the Methyl CpG Binding Protein-2 gene present on the X chromosome. **Discussion:** Rett syndrome is a rare genetic pathology related to the dominant X chromosome, resulting in the translation of proteins capable of promoting severe motor destruction. Male individuals with this syndrome die and female individuals carry severe sequelae throughout their lives. Because the evolution of the disease is characteristic, it is possible to divide four stages of this syndrome. The first has a deficiency in body development. The second consists of a rapid psychomotor regression, onset of epileptic seizures, impairment of reflexes, communication and social interaction. In addition, the patient already develops respiratory irregularities. In the third phase, there may be an improvement in communicative ability in contrast to a worsening in ambulation and balance, which is more evident in the presence of scoliosis. In stage four, present at about ten years of age, there is a visible progression of motor delay, poor ability to walk and perform manual actions. Evident signs of peripheral motor neuron injury, body rigidity, malnutrition, and gastrointestinal dysfunction. **Final comments:** Rett's disease, because it is a rare genetic syndrome and its first cases were discovered in Brazil only close to the date of 1990, still causes delays for medical professionals in making an assertive diagnosis to patients with this mutation in their genes. And, because it is a disease with no cure that occurs in multiple disabilities, starting a multidisciplinary care work early to assist in the functional capacity of the patient in question is of great importance. Therefore, popularizing the existence of this syndrome, along with its signs and symptoms, helps in the training of health professionals and reflects positively on the patient's survival.

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Reversible cerebral vasoconstriction syndrome associated with probable drug poisoning: case report

Michelle Zonkowski Ribas[1], Arthur de Oliveira Veras[2], Felipe Micelli Noleto[3], Gabriela Ferreira Paticcié[4], Júlio César Claudino dos Santos[5]

[1]Hospital Universitário Cajuru, Curitiba PR, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[3]Centro Universitário Christus, Faculdade de Medicina, Fortaleza CE, Brazil.
[4]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.
[5]Universidade Federal do Ceará, Programa de Pós-Graduação em Ciências Morfofuncionais, Fortaleza CE, Brazil.

Correspondence: Michelle Zonkowski Ribas; Email: michelle.z.ribas@gmail.com.

Case presentation: A 19-year-old woman with no relevant medical history, except for regular use of marijuana and a narcotic based on chloroform and ether, reported an onset of an acute holocranial headache a week earlier. She was found unresponsive and admitted to the hospital following a generalized tonic-clonic seizure. A non-contrast computed tomography (CT) scan of the skull revealed hyperdense foci in the splenium of the corpus callosum and the cingulate gyrus, with no other significant findings. Diagnostic cerebral arteriography showed diffuse narrowing of intracranial arteries, particularly in the P1 and P2 segments of the posterior cerebral arteries. The diagnostic hypothesis was Reversible Cerebral Vasoconstriction Syndrome (RCVS), with marijuana and the narcotic considered as precipitating factors. After 15 days, neurological improvement was observed, and a follow-up angiographic study documented the reversal of arterial narrowing. **Discussion:** RCVS is a condition characterized by arterial involvement of intracranial vessels, manifesting with vasospasm. It primarily affects individuals between 20 and 50 years of age, with a higher prevalence in females. The patient in this case fits within the typical demographic for sex but is at the lower age limit. The hallmark of RCVS is a sudden and severe headache. This can be accompanied by seizures, as seen in this patient, as well as focal neurological deficits. Laboratory tests and cerebrospinal fluid analysis are often nonspecific, typically showing no significant abnormalities, which aligns with the findings in this case. Therefore, imaging studies are crucial for the diagnosis. Non-contrast CT scans of the brain can produce variable results and are sometimes normal. However, an angiographic study is more definitive, often showing the characteristic "string of beads" appearance due to alternating segments of arterial narrowing and dilation. This patient's angiography revealed diffuse narrowing, especially in the posterior cerebral arteries, supporting the RCVS diagnosis. The etiology of RCVS is multifactorial, with precipitating factors identified in approximately 60-80% of cases. These factors include the postpartum period, preeclampsia/eclampsia, and exposure to certain medications such as antidepressants and immunosuppressants. Vasoactive substances, including illicit drugs like marijuana, cocaine, and heroin, are also well-documented triggers. In Brazil, the use of a clandestine narcotic mixture based on chloroform and ether for recreational purposes is a concern. This mixture's vasoactive properties may have contributed to the development of RCVS in this patient. **Final comments:** It is essential to recognize the variety of clinical and imaging patterns of RCVS to quickly identify suspected cases and exclude differential diagnoses. While the link between RCVS and marijuana is documented, this case suggests a potential new association with narcotics containing chloroform and ether.

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Reversible posterior encephalopathy syndrome (PRES) after endovenous iron infusion: unusual case in the literature

Carolina Flores Welker[1], Larissa Lucas Schloicka[1]

[1]Hospital de Base do Distrito Federal, Brasília DF, Brazil.

Correspondence: Carolina Flores Welker; Email: carolinafwelker@gmail.com.

M.C.N., female, 37 years old, admitted to a high-complexity hospital on 02/14/2023 after experiencing headache, vomiting, visual disturbances and reduced level of consciousness two days earlier, shortly after intravenous iron infusion. She had a previous diagnosis of uterine myomatosis and iron deficiency anemia. She had already undergone oral iron replacement, but had not yet received parenteral iron replacement. Upon admission to the emergency room, she was drowsy; inattentive, with fluctuation in level of consciousness and multidirectional rotatory nystagmus. There were

no focal neurological deficits evident. Head CT showed vasogenic edema in the left parietal lobe and ischemia in the cerebellar hemispheres, with compression of the fourth ventricle and ectasia of the supratentorial ventricular system. Hours later, the patient developed a lower level of consciousness, requiring OTI and, on the same day, a EVD was inserted . In the following days, there was significant clinical improvement, she remained without neurological deficits and there was resolution of encephalopathy. She was Extubated on 02/25/23 and the EVD was removed on 03/05/23, without complications. On 02/28/23, she was admitted to the neurology ward, alert, oriented; without any motor or sensory deficits; with preserved balance, coordination and gait. She maintained central pattern nystagmus. Skull MRI on 03/13/23 showed sequelae changes in the cerebellar hemispheres suggestive of previous vascular insult. She was discharged on 03/13/2023 with a diagnostic hypothesis of PRES. PRES is a clinical-radiological syndrome, whose clinical picture is characterized by encephalopathy, headache, visual disturbances and seizures. Imaging usually shows vasogenic edema predominantly in the occipital and parietal lobes. The most frequently described associated conditions are high blood pressure, eclampsia and use of immunosuppressants and chemotherapy. Its pathophysiology is not well understood, however, endothelial dysfunction and loss of cerebral autoregulation are the most accepted theories. The prognosis is good and, in most cases, symptoms and imaging findings are reversible. Our patient was retrospectively diagnosed with PRES and the only factor identified was intravenous iron infusion. As yet, we have not found a similar case in the literature. There are reports of PRES after infusion of packed red blood cells, which, as it is rich in iron, may have a pathophysiology similar to that of the case described.

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Reversible posterior leukoencephalopathy syndrome after intravenous iron infusion

Robercon Alves do Carmo[1], André Teixeira de Souza e Castro[1], Fernanda Sousa Lima[1], Larissa Lucas Schloicka[1], Rodrigo Torres Sena Pita[1], Carolina Flores Welker[1], André Gustavo Fonseca Ferreira[1], Wagner Afonso Teixeira[1]

[1]Hospital de Base do Distrito Federal, Brasília DF, Brazil.

Correspondence: Robercon Alvesd Carmo; Email: robercon025@gmail.com.

Case presentation: A 37-year-old female patient, previously diagnosed with uterine myomatosis, presented with significant metrorrhagia and iron deficiency anemia as a result. In February 2023, she was referred after experiencing occipital headache and uncontrollable vomiting following intravenous iron infusion (ferripolimaltose). Subsequently, the patient experienced progressive worsening of the headache associated with altered consciousness, alternating between periods of psychomotor agitation and drowsiness. Upon admission, she exhibited active movement of all four limbs but maintained a state of encephalopathy, making it difficult to cooperate with the physical examination. Initially evaluated by the neurosurgery team, a CT scan revealed extensive cerebellar ischemia leading to compression of the fourth ventricle. She underwent external ventricular drainage and was transferred to the intensive care unit, where she remained hospitalized for 10 days. During this period, she underwent conversion to a ventriculoperitoneal shunt and was subsequently referred to neurology for the etiological definition of the ischemic vascular insult that triggered her condition. Following extensive investigation during hospitalization, the patient was diagnosed with mild triglyceride elevation in the context of dyslipidemia. No other common triggering causes were found, although there are only two reported cases in the literature of a similar condition triggered by iron replacement. After identifying intravenous iron infusion as the trigger for the condition, the patient was referred to the vascular neurology clinic for follow-up, currently reporting only dizziness while being able to carry out her work activities without impairment. **Discussion:** Reversible posterior leukoencephalopathy syndrome is a rare cause of cerebral vascular insult, but should always be considered in patients presenting with symptoms suggesting a posterior fossa location. Extensive investigation is necessary for diagnostic confirmation, and whenever possible, the causative agent should be identified to prevent subsequent events. **Final comments:** In this presentation, we described a case of reversible posterior leukoencephalopathy syndrome in a patient, with the etiological agent being a medication that is poorly described in the literature.

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Rhino-cerebral mucormycosis presenting with ophthalmoplegia and intracerebral hemorrhage

Fernanda Lustosa Cabral Gomez[1], Lara Santos Machado[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Gabriela Rodrigues Tomaz[1], Leonardo Villaverde Buback Ferreira[1], Larissa Braga Mendes[1], Maria Elisabeth Matta de Rezende Ferraz[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Fernanda Lustosa Cabral Gomez; Email: fernandalustosacg@gmail.com.

Case presentation: A 55-year-old man was admitted to the emergency department with a one-day history of sudden loss of vision of the right eye. He had a previous diagnosis of uncontrolled diabetes. Upon admission, his blood glucose level was 406 mg/dL. Neurological examination revealed no perception of light, mydriasis and complete ophthalmoplegia of the right eye, as well as right facial hypoesthesia. Noncontrast brain CT showed invasive right maxillary-ethmoidal sinusitis and nasofibroscopy revealed necrosis of the lower right turbinate. Brain MRI suggested acute invasive fungal rhinosinusitis. The patient was placed on intravenous amphotericin and ethmoidal sinusectomy was performed. Two days later, he presented with reduced consciousness level and bilateral mydriasis. Emergency brain CT revealed right frontal lobe hemorrhage with midline shift and bilateral subfalcine and uncal herniation. CT Angiography was unremarkable. The patient underwent emergency decompressive surgery but two days later brain death was documented. Nasal biopsy later revealed Rhizopus spp (Mucormycosis). **Discussion:** Mucormycosis is a rare and often fatal opportunistic infection caused by the pathogenic genera Mucor, Absidia, and Rhizopus of the family Mucoraceae. Rhino-cerebral mucormycosis (RCM) is a form of cephalic mucormycosis known to occur in immunocompromised patients. The infection usually begins with symptoms of sinusitis and rhinitis and, as the fungus disseminates to the orbit, it causes involvement of the cranial nerves, such as in the case above, a classic orbital apex syndrome. Mucor fungus has a high affinity for blood vessels, particularly for the elastic membrane. Therefore, RCM may present as a stroke due to a major artery occlusion. However, case reports of intracerebral hemorrhage due to mucormycosis are extremely rare, specially with the genera Rhizopus. Although CT angiography was unremarkable in our case, we believe that it could be due to occlusion of a cortical vein or, less likely, arterial dissection or mycotic aneurysm. **Final comments:** Although rare, mucormycosis should be sought in cases of ophthalmoplegia and intracerebral hemorrhage in immunocompromised patients, such as in uncontrolled diabetics. In RCM we also highlight the importance of urgent otorhinolaryngologic evaluation. Early treatment with intravenous amphotericin B, surgical debridement and control of risk factors are imperative for survival.

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Rhino-orbital mucormycosis presenting as orbital apex syndrome: a case report

Pedro Almeida Martins Pontes[1], Filipe Marçal Junqueira[1], Henrique de Moraes Bernal[1], Ingrid Lorena da Silva Gomes[1], Aldren Thomazini Falçoni Júnior[1], Guilherme Lago Miranda[1], Eduardo Takashi Takehara[2], Mateus Paquesse Pellegrino[1], Marcele Schettini de Almeida[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Pedro Almeida Martins Pontes; Email: pedroalmeida.pontes@gmail.com.

Case presentation: An 83-year-old man was admitted with myalgia, fever and headache and was diagnosed with dengue. His medical history included diabetes mellitus (HbA1c 10.1%), heart failure with reduced ejection fraction and alcoholism. Throughout the next few days, he showed clinical improvement but had persistent thrombocytopenia. This led to a diagnosis of post-dengue Idiopathic Thrombocytopenic Purpura, and treatment with Prednisone 60 mg was started. On the seventh day of corticosteroid treatment, platelet count progressively improved. Concurrently, he developed acute ptosis, ophthalmoplegia, and reduced visual acuity in the left eye. Upon initial evaluation by Neurology, the patient showed alterations on the left side in the cranial nerve exam: ptosis, proptosis, complete paralysis of cranial nerves III, IV, and VI, absence of the corneal reflex, hypoesthesia in the V1 territory, and visual acuity with no light perception. There were no other abnormalities in the neurological physical exam. This clinical presentation is consistent with Orbital Apex Syndrome. The patient underwent etiological investigation, with main suspicions of vascular or infectious causes. Digital angiography was normal, ruling out vascular causes such as dural fistulas. CSF analysis showed no abnormalities. Brain and orbit MRI showed severe sinus disease in the left frontal sinus, along with infiltrative effects, with the presence of gas in the ipsilateral orbit. Nasofibroscopy revealed necrotic crusts throughout the left nasal cavity, with culture positive for Mucor sp., confirming the diagnosis of Rhino-Orbital Cerebral Mucormycosis. Antimicrobial treatment was started, and the patient underwent extensive surgical debridement. Fourteen days after diagnosis, the patient died from secondary bacterial complications. **Discussion:** Mucormycosis is an invasive fungal disease caused by fungi of the order Mucorales. Although rare, Mucormycosis is known to present with Orbital Apex Syndrome, especially in diabetic patients or those with prior immunosuppression. In a review conducted in India, 73.5% of patients diagnosed with Mucormycosis were diabetic, making it the main isolated risk factor for this condition. In the reported case, the patient had uncontrolled diabetes and was started on corticosteroids, which could have triggered the fungal infection. Even with early diagnosis and appropriate treatment, the mortality of Rhino-Orbital Cerebral Mucormycosis remains high (50 to 100%, depending on the reference). **Final comments:** Diabetic patients or those with general immunosuppression presenting with Orbital Apex Syndrome should be promptly investigated for Mucormycosis. It is a disease with high mortality. Neurologists need to be aware of the topic for high clinical suspicion and early therapeutic intervention to potentially improve prognosis in these high-risk populations.

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Rhomboencephalitis as a differential diagnosis of stroke: a case report

Daniele da Costa da Silva[1], Artur Gasparovic Chagas[1], Ana Carolina Grande[1], Ivo Marcos Darella

Lorenzin Fernandes Neto[2], Eduarda Basso Badalotti[2], Angelo Marcelo Wosniacki Filho[2], Gabriel Angelo Garute Zenatti[2], Renato Endler Iachinski[2], Antonio Carlos de Andrade Soares[2]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Fundação Hospitalar São Lucas, Cascavel PR, Brazil.

Correspondence: Daniele da Costa da Silva; Email: dani.costadasilva09@gmail.com.

Case presentation: A 54-year-old male was referred from primary care to the Neurology department for further investigation due to a stroke suspicion. The patient reported blurred vision and intermittent loss of strength for a few days, with spontaneous resolution. No past medical history of infections, vaccinations, and negative family history of neurological diseases. On neurological evaluation, presented with a decreased visual acuity, proportionate right hemiparesis, dysplasia, and gait ataxia relapse-remission symptoms. Lesions with hypersignal on T2/Flair sequence in the pons, left cerebellar peduncle, cerebellum, heterogeneous contrast restriction, without obeying vascular territory associated with changes in the left optic nerve suggestive of neural inflammation were described on the MR. No abnormalities on cerebrospinal fluid and no positive serology for Cytomegalovirus, Epstein-Barr, Herpes, and Cryptococci infection. Together, the Neurology and Neuroradiology team defined the condition as Rhomboencephalitis associated with idiopathic Neuritis Optica. After a 5-day methylprednisolone 1g treatment, the patient improved significantly in previous symptoms. **Discussion:** Rhomboencephalitis is defined as an inflammation of the pons, medulla oblongata, and cerebellum. Typically viral or bacterial origin and usually affects immunocompromised people, especially patients under immunosuppressants, paraneoplastic syndromes, and autoimmune conditions such as Behcet disease. The gram-positive bacterium Listeria monocytogenes, with a 9% chance of developing the disease, is the most common pathogen, normally transmitted through contaminated food. Initial clinical findings include flu-like symptoms, progressing with neurological manifestations and diagnosis is made by hematological or cerebral fluid test. **Final comments:** Rhomboencephalitis is an uncommon condition with a high morbidity and mortality rate. Due to its extensive clinical presentation, it is an important differential diagnosis in patients with sudden neurological symptoms to provide the appropriate treatment.

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Right peripheral facial paralysis in a 1-day-old neonate: a case report

Taianara Sampaio Reis[1], Amauri Pereira da Silva Filho[2], Melissa Fernandes Vilela de Freitas[3], Danielle Cortellazzi Colonna Romano[3], Gabriela Yumi Nakamura[3], Sabrina Borges Batista[3], Natália Rebeca Karpejany[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Hospital de Emergência e Trauma Dom Luiz Gonzaga Fernandes, Campina Grande PB, Brazil.
[3]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Taianara Sampaio Reis; Email: taianarasampaio21@hotmail.com.

Case presentation: A term male neonate, 1 day old (d.o.), born via cesarean section with an Apgar score of 8/8, presented for outpatient evaluation with right-sided facial mimic alteration. The mother reported inadequate prenatal care, without regular check-ups, vaccination, or serological tests. PHYSICAL EXAMINATION: The neonate was in good general condition, 1 d.o., weighing 3 kg, with a normotensive anterior fontanel open to one finger breadth, showing hemifacial alteration with deviation of the buccal rim to the left. Bell's sign was present in the right eye. After examination, the diagnostic hypothesis of right peripheral facial paralysis was raised. Subsequently, tests were requested to investigate the probable cause of the paralysis. Routine examinations of the child were normal, with no history of birth trauma, and no signs or stigmata of syndromic diseases. Therefore, cranial computed tomography (CT) and magnetic resonance imaging (MRI), lumbar puncture, and serological tests were performed to continue the investigation. The test results were: normal cerebrospinal fluid, normal cranial CT with no fractures, normal cranial MRI without clinical abnormalities, negative serology for VDLR, HIV, Hepatitis B and C, Rubella (IgM negative and IgG positive), cytomegalovirus (CMV) (IgM negative and IgG positive), Toxoplasmosis (IgM negative and IgG positive), Herpes 1 and 2 (IgM negative and IgG positive). **Discussion:** Bell's palsy is a neuropathy affecting the seventh cranial nerve, also known as the facial nerve. It can have different etiologies, such as trauma, infections, inflammation, or compressions involving the nerve. Some cases are classified as idiopathic when no probable etiologies can be found. The most common cause of Bell's palsy is the herpes simplex virus, but it can also be caused by other viruses such as Epstein Barr, HIV, and hepatitis B. The presentation of this syndrome in the pediatric population is very rare, with few reports in the literature. This makes early diagnosis and appropriate treatment crucial for better prognosis. **Final comments:** The patient was diagnosed with Bell's palsy and treated with symptomatic medications and physiotherapy. The neonate initially presented with a House-Brackman scale score of 3 and improved to a score of 2 after physiotherapy. Bell's palsy does not have a specific etiology, and it could have been due to previous infections. The patient is IgG positive for rubella, CMV, and herpes 1 and 2. Differential diagnoses considered for this patient included birth trauma, mastoid trauma, syndromic alterations, HIV, hepatitis B, brain tumor, meningitis, and meningoencephalitis. All these diagnostic hypotheses were ruled out after testing, ultimately confirming the diagnosis of Bell's palsy.

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Risdiplam treatment for spinal muscular atrophy: real life data from a Brazilian cohort

Cristina Iwabe[1], Felipe Franco da Graça[1], Marcondes Cavalcante França Jr[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Cristina Iwabe; Email: crisiwabe@hotmail.com.

Background: Risdiplam is an oral SMN2 splicing modulator used to treat spinal muscular atrophy (SMA) that became recently available in the Brazilian public health system. Real life data on Risdiplam in the Brazilian population are missing. **Objective:** To describe Ridisplam effects in a mixed SMA cohort from a referral center. **Methods:** This is an 8-month retrospective analysis of subjects taking Ridisplam, who were evaluated with motor scales before and at least 3 months after treatment onset. Motor function was assessed either using the HFMSE or the CHOP-INTEND scales depending on the clinical type of SMA. **Results:** There were 6 men and 6 women. Two patients had SMA1, 6 SMA2 and 4 SMA3. Mean age was 28.9 years (ranging from 6 to 60 years), whereas mean treatment duration was 6 months (ranging from 3 to 8 months). During follow-up, we had the same number of patients with stable (n=6) or improved (n=6) functional scores. In the stable group, there were 3 SMA2 and 3 SMA3; mean age was 36.6 years. Risdiplam use lasted on average 6.1 months. In the improved group, we had 2 SMA1, 3 SMA2 and 1 SMA3. Percentual improvement ranged from 11,10 to 33,30% in this subgroup. Mean disease duration was 21 years. None of the patients reported meaningful side effects during treatment. **Conclusion:** In this mixed Brazilian cohort, Risdiplam led to either motor functional stabilization or improvement. Further studies are needed to identify predictors for better response.

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Risk factors and prognosis of myelomeningocele associated with holoprosencephaly: a case report

Lizandra Stefani Barbosa[1], Andressa Girelli Cardoso[1], Gabrielle de Souza Bernardes[1], Jessica Helena Gomes Ferreira[1], Caroline Chaves Bittencourt[1]

[1]Universidade Nove de Julho, Guarulhos SP, Brazil.

Correspondence: Lizandra Stefani Barbosa; Email:

Case presentation: A newborn male, born on 22/03/2024, at term via a difficult cesarean section (11 minutes) due to macrocephaly and myelomeningocele (MMC), with polyhydramnios and meconium-stained amniotic fluid (2+/4+), weighing 5,190g and with a head circumference of 48 cm. He required resuscitation and intubation and was transferred to the neonatal ICU. A cranial CT scan revealed hydrocephalus (HC) and alobar holoprosencephaly (HPE), and a spinal CT scan showed lumbosacral MMC. On physical examination, he presented with macrocephaly, large and communicating fontanelles, suture dehiscence; a ruptured MMC in the sacral region, and bilateral congenital talipes equinovarus (CTEV). On 28/03/2024, he underwent a ventriculoperitoneal shunt (VPS) placement and MMC repair without complications. He was discharged from the hospital on 08/04/2024. **Discussion:** Folic acid deficiency is the main risk factor for neural tube defects, with MMC being the most common. The mortality rate reaches up to 70% within the first 6 months of life if untreated. MMC is a complex condition frequently associated with HC (in up to 80% of cases), Chiari II malformation, and orthopedic malformations (approximately 9% of cases) such as CTEV, with variability in prognosis. Macrocephaly and higher anatomical levels of the spinal defect increase the incidence of complications, which, along with HC, require multidisciplinary treatment, with shunt placement, such as VPS, being necessary in about 83.5% of cases. Shunt dysfunction affects approximately 47.7% of patients and requires frequent surgical revisions. Thus, MMC remains one of the leading causes of chronic disability, maintaining a mortality rate of 7.4% even with treatment. No risk factor has been well established for HPE, but the condition appears to be influenced by genetic, environmental, and multifactorial causes. Genetic etiologies, such as trisomy 13 and 18 and triploidy, account for most cases. Persistent hyperglycemia in pre-existing or insulin-dependent maternal diabetes, as well as salicylate consumption, are associated with a higher risk of HPE. Alobar HPE is the most severe form of this condition and has a poor prognosis, as developmental milestones, such as sitting or speaking, are not achieved even later in life. Additional complications may include dysphagia, gastroesophageal reflux, hypothalamic dysfunction, and endocrine problems. Mortality reaches 50% within the first 5 months of life, with only 30% surviving beyond the age of 1 year. **Final comments:** Despite the severity of the patient's neurological conditions–alobar HPE and MMC–with the interventions performed and the absence of critical clinical findings such as severe facial anomalies and other major dysfunctions, the patient's progress was observed. There is scarce data on the association of HPE and MMC, but it is known that these conditions separately have high mortality rates in the first year of life, and their co-occurrence worsens the prognosis.

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Risk stratification for stroke among professionals in a hospital network

Aline Fernanda Alves Ribeiro[1], Mayk Penze Cardoso[2], Andreia Insabralde de Queiroz-Cardoso[1]

[1]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.
[2]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.

Correspondence: Aline Fernanda Alves Ribeiro;
Email: aline.ribeiro@ufms.br.

Background: Stroke is one of the leading causes of mortality and disability worldwide. It is estimated that more than 15 million strokes occur each year, of which about 6 million are fatal. The increased risk for stroke is multifactorial. Additionally, epidemiological studies have shown that certain work environments and occupations are associated with a higher risk of stroke. **Objective:** To report the development of a stroke risk stratification activity among professionals in a public hospital network. **Methods:** This is an experience report regarding an action carried out in a public hospital in October 2023, aimed at professionals on "World Stroke Day". The professionals were stratified according to the "Stroke Riskometer" software questionnaire, which includes variables related to age, sex, race, anthropometric data, lifestyle habits, family history, and comorbidities. The application provided results on a scale from 0 to 100%, indicating the probability of a stroke within 5 years. Blood pressure measurements, general guidance, and risk factor control were also performed. **Results:** The activity included 35 professionals, most of whom were female (n=19). The occupations included nursing technicians (n=13), nurses (n=6), nursing assistants (n=2), radiology technicians (n=1), doctors (n=2), pharmacist (n=1), pharmacy technicians (n=4), receptionists (n=2), stretcher-bearer (n=1), pantry worker (n=1), and cleaning assistants (n=2). The stroke risk score ranged from 0.5% to 5.5% probability of developing the disease over 5 years. The highest risk scores were among males, with an average risk of 2.07%, while females had an average risk of 1.94%. Notably, in this study, the occupations with the highest indices were, respectively: radiology technicians, receptionists, nursing technicians, doctors, and pharmacy technicians. Furthermore, the main concerns of the professionals were related to risk factors and the identification of comorbidities, such as hypertension and diabetes. Recommendations included regular visits to health units to identify potential problems, regular physical activity, and maintaining a healthy diet. **Conclusion:** Awareness of modifiable stroke risk factors is essential to implement effective preventive measures and highlight the importance of occupational health in preventing this condition. Therefore, it is crucial that organizations adopt measures to promote healthy work environments, with occupational health policies aimed at reducing stroke risk factors, including health promotion programs, encouraging physical activity, and managing stress levels at work.

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Rockstar hands: a complex pattern of hand dystonia in Wilson's disease

Luis Eduardo Borges de Macedo Zubko[1], Renata Cristine Alves[1], Francisco Manoel Branco Germiniani[1], Hélio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Complexo Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Luis Eduardo Borges de Macedo Zubko; Email: luis.zubko@gmail.com.

Case presentation: A 27-year-old man presented with subacute generalized dystonia, low serum ceruloplasmin levels, and increased copper urinary excretion, which were consistent with the diagnosis of Wilson's disease (WD). Following treatment with D-penicillamine, biperiden and baclofen the generalized dystonia improved significantly, but a residual significant hand dystonia persisted. His treatment was afterwards altered to trientine and weaned-off of anticholinergic drugs. The remaining hand dystonia had a complex pattern that resembled the finger and hand positions commonly seen in electric guitar players playing rock songs with major barre chords. The fingers of the left hand exhibit a distinctive pattern, with the left 3rd and 4th fingers flexed, while the 2nd and 5th fingers are extended. The left hand showed an additional severe wrist flexion. The right wrist was flexed with an ulnar deviation, associated with 3rd finger extension, while the 2nd, 4th and 5th fingers were flexed. The hand dystonia was refractory to botulinum toxin therapy and persisted in this fixed position suggesting shortening of tendons. **Discussion:** WD is a hereditary inborn error of copper metabolism caused by a mutation of the ATP7B gene. It usually occurs between the ages of 5 and 35 years and presents with hepatic, neurological and psychiatric symptoms. The most common neurological symptoms are movement disorders, especially, dystonia, parkinsonism, tremor and ataxia. Dystonia in WD frequently presents in either a generalized distribution, or as a focal dystonia in the oromandibular region. Dystonia affecting the hands may have several etiologies and present in various forms, with cases reported as "starfish hand", claw hand, and task-specific dystonias (including musician's dystonia, writer's cramp, hairdresser's dystonia), among others. It is important to accurately identify the pattern and muscles involved for optimal treatment. Final remarks dystonia in young patients warrants prompt investigation to rule out reversible conditions such as WD and other inborn errors of metabolism. Even with disease-modifying treatment, incapacitating dystonia with complex patterns may remain. Recognition of these patterns is important to assist in muscle selection for treatment with botulinum toxin.

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Role of Cannabidiol as an adjuvant in the treatment of refractory cryptogenic epilepsy: a case report

Fernanda Valeriano Zamora, Andres Villca Zamora[2], Ana Clara F. F. Santos[3], Nicole S. Pimenta[4], João Pedro C. E. A. Salles[4], Lorhayne Kerley Capuchinho Scalioni Galvao[2], Afshan Nizami[5], Camila M. Yamada[2], Deivyd Vieira Silva Cavalcante[6], Dayana Dourado de Oliveira Costa[7]

[2]Universidade Federal de Minas Gerais, Belo Horizonte BH, Brazil.
[3]Universidade Cidade de São Paulo, São Paulo SP, Brazil.
[4]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[5]Apollo Institute of Medical Sciences and Research, Telangana, India.
[6]Universidade Federal do Maranhão, São Luís MA, Brazil.
[7]Universidade CEUMA, São Luís MA, Brazil.

Correspondence: Fernanda Valeriano Zamora; Email: dra.fernandavaleriano@gmail.com.

Case presentation: A 44-year-old female patient under neurological care due to cryptogenic epilepsy (absence type), with complex partial seizures since 1998, refractory to all previously used medications, likely due to a 15x14x13mm tumor in the right amygdala, consistent with low-grade glial neoplasia, with no surgical approach proposed. The patient has experienced progressive worsening, nearly daily seizures, significantly impacting quality of life and occupational capacity. The patient has previously tried Phenobarbital 800mg/day, Carbamazepine 800mg/day, Valproic acid 1,500mg/day, Sodium valproate 1,500mg/day, Topiramate 100mg/day, Lamotrigine 200mg/day, Oxcarbazepine, Clobazam, Phenytoin, Clonazepam, Bromazepam, and Levetiracetam, none of which reduced seizure frequency and caused various side effects such as dizziness, memory loss, and fatigue. Cannabidiol (CBD) isolate was initiated at 200mg/ml, 5mg/kg/day, with dose escalation. Seizure frequency improved from daily to approximately 1 seizure per month. Stability was achieved at 15mg/kg/day (660mg/day for a 44kg patient), with reported side effects of diarrhea and nausea. **Discussion:** Patients with drug-resistant epilepsy and no surgical option showed positive outcomes with CBD, improving seizure control and overall well-being, consistent with previous studies demonstrating significant reductions in seizure frequency and improvement in quality of life. Generally, cannabidiol is associated with minor side effects and has a favorable safety profile, offering a good risk/benefit ratio. Furthermore, there is still no consensus on the appropriate dosage for treating seizures, which leaves uncertainties among prescribing professionals and warrants extensive discussion. **Final comments:** CBD therapy presents as a promising adjunctive treatment for drug-resistant epilepsy patients and may complement surgical options. Its potential to reduce seizure frequency and improve quality of life highlights the need for further research into optimizing its use in clinical practice. Continued investigation into dosage, long-term effects, and patient-specific responses will be crucial in fully understanding CBD's role in managing refractory epilepsy.

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Role of skin biopsy in the etiological diagnosis of multiple mononeuritis

Samir Andres Ramirez Ojeda[1], Leidys Pedrozo[1], Tais Werle[1], Bruna Klein[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Samir Andres Ramirez Ojeda; Email: samir.ramirez@edu.pucrs.br.

Case presentation: Male patient, 66 years old, pre-diabetic and dyslipidemic, starts with slowly progressive symptoms over the last 2 years of fatigue and loss of strength in the lower limbs associated with muscular atrophy and later neuropathic pain. No paresthesias or autonomic changes. On examination, broad and symmetrical pulses and the presence of livedo reticularis in the lower limbs, positive Romberg, proximal hypopalesthesia and distal apalesthesia in the lower limbs. Strength grade 3 (MRC) at nadir. Severe pain on palpation of the Achilles tendons due to tendinopathy. Electroneuromyography showed a pattern compatible with mononeuritis multiplex. Systemic investigation for infectious, metabolic, neoplastic diseases and negative vitamin deficiencies. Cerebrospinal fluid unchanged. Glycated hemoglobin 6.3%. Antinuclear antibody (ANA) positive 1:640 centromeric pattern, others negative. A skin biopsy of the livedo reticularis area was performed, which demonstrated dermal lymphocytic capillaritis with extravasation of red blood cells (micropurpuric), without evidence of fibrinoid necrosis or thrombosis. He received pulse therapy with methylprednisolone 1gIV/day for 5 days followed by gradual reduction of oral corticosteroids and cyclophosphamide 1g/month with a plan of 4 to 6 doses with significant improvement and progressive functional recovery. **Discussion:** Lymphocytic capillaritis associated with livedo reticularis is a condition of unknown etiology, with possible immunological mechanisms proposed. In our patient, a skin biopsy was essential to rule out livedoid vasculopathy. Furthermore, the presence of tendinitis, reactive ANA in high titers and response to immunotherapy suggest autoimmunity as the cause of mononeuritis multiplex. **Final comments:** Consider skin biopsy as a resource for the etiological differential diagnosis of mononeuritis multiplex.

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Safety and efficacy of alternative vascular accesses in mechanical thrombectomy for large occlusion vessel strokes: a systematic review and meta-analysis

José Guilherme Maia[1], Daniel de Godoy Andreis[2], Luiza Abreu Queiroz[3], Denisarth Oliveira Nery Filho[4], André Luís e Silva Evangelista[1], Izael Pereira da Silva[1], Ítalo Barros Andrade[5], Artur Menegaz de Almeida[6], Lilianne Rodrigues Fernandes[7]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Paraná, Curitiba PR, Brazil.
[3]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.
[4]Centro Universitário Uninovafapi, Teresina PI, Brazil.
[5]Faculdade Santo Agostinho de Vitória da Conquista, Vitória da Conquista BA, Brazil.
[6]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[7]Afaya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.

Correspondence: Daniel de Godoy Andreis; Email: daniel.godoy.andreis@gmail.com.

Background: Neurological endovascular procedures have been performed via transfemoral access (TFA). This procedure has become the standard approach, given its greater ease in mechanical thrombectomy (MTB) for large occlusion stroke treatment. However, TFA can present potential complications. Thus, transradial access (TRA) and transcarotid access (ACT) have been considered viable alternatives, presenting better results and fewer complications. **Objective:** To demonstrate if using alternative vascular access (TRA or TCA) is safer and more effective than TFA in MTB for large occlusion stroke. **Methods:** MEDLINE, Cochrane, and Scopus databases were assessed to gather randomized clinical trials (RCT) and case-control studies (CCS) comparing TRA/TCA interventions versus TFA as a control group in MTB treatment for large occlusion stroke. Any complication related to any access (ACC), successful recanalization (SRC), mortality (MOR), and mean time from arrival to recanalization (ATR) were the outcomes evaluated. Data were examined using the Mentel-Haenszel method and 95% CIs. Heterogeneity was assessed using I^2 statistics. The statistical analysis was based on the R Studio, version 4.3.2. **Results:** A total of 1 RCT and 8 CCS comprising 4023 patients were included, of which 669 (16.63%) underwent MTB either through TRA or TCA and 3354 (83.37%) through TFA. The TRA/TCA group was associated with a reduction in ACC (6% vs 9%, RR 0.6, 95% CI 0.45-0.82, p=0.001, I^2=0%), compared with the TFA group. There was no significant statistical difference in SRC (90% vs 89.94%, OR 1.09; 95% CI 0.79,1.50; p=0.69; I^2=0%), MOR (12% vs 13.51%, RR 1.04; 95% CI 0.79,1.37, p=0.7964; I^2=0%) and ATR (MD -4.92; 95% CI -20.56, 10.71; p=0.5371; I^2=94%). **Conclusion:** This meta-analysis demonstrated that the use of TFA or TRA as alternative vascular accesses in MTB for large occlusion stroke reduced the risk of any type of procedure-related complication by 40% when compared to TFA. In this scenario, new studies, especially RCTs, need to be developed to clarify the effectiveness of these alternative vascular accesses in MTB for large occlusion stroke.

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Safety and efficacy of batoclimab for generalized myasthenia gravis: a systematic review and meta-analysis

Ian de Paula Bezerra[1], Giovanna de Freitas Morais[2], Helen Cristina Beckert[2], Flávia Christina Mota Kalinowski[2], Artur Menegaz de Almeida[3], Ítalo Barros Andrade[4], Lilianne Rodrigues Fernandes[5] , Michele Kreuz[6]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Paraná, Curitiba PR, Brazil.
[3]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[4]Faculdade Santo Agostinho de Vitória da Conquista, Vitória da Conquista BA, Brazil.
[5]Faculdade de Ciências Médicas da Paraíba, Abaetuba PB, Brazil.
[6]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Flávia Christina Mota Kalinowski; Email: flaviakalinowski@hotmail.com.

Background: Several neonatal Fc receptor (FcRn) inhibitors represent a possibility in the treatment of generalized Myasthenia Gravis (gMG). However, the precise therapeutic profile and efficacy of batoclimab in this context are not yet fully understood. **Objective:** To evaluate the efficacy and safety of batoclimab for the treatment of generalized Myasthenia Gravis. **Methods:** MEDLINE, Scopus, and Web of Science databases were systematically searched. Data were examined using the DerSimonian Laird method and 95% confidence intervals. Heterogeneity was assessed using I^2 statistics. R, version 4.3.2, was used for statistical analysis. **Results:** A total of 3 randomized control trials and 179 patients were included. Compared with placebo, treatment with batoclimab achieved better reduction of MG-ADL score (MD -2.07; 95% CI -2.06 - -1.49; P ≤0.000001; I^2= 0%); treatment with batoclimab had a better improvement of QMG score (MD -5.47; 95% CI -7.70 - -3.25; P=0.000001; I^2=79%). The rate of treatment-emergent adverse effects and other specific adverse effects such as diarrhea, headache, and upper respiratory tract infection did not reach a statistically significant difference between groups. **Conclusion:** Consistent results suggest that the use of batoclimab is associated with efficacy in the treatment of gMG, reducing MG-ADL and QMG scores. The drug had no significantly relevant adverse events in comparison to placebo. In conclusion, our results support the use of batoclimab for the treatment of gMG.

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Safety and efficacy of dabrafenib and trametinib in pediatric patients with glioma: a systematic review and meta-analysis

Abner Lucas Balduino de Souza[1], Edmundo Damiani Bertoli[2], Jhefferson da Silva Sousa[3], Luísa Diniz Napoleão[4], Marina Ferreira Machado[5], Fabíola Furtuoso Zarpelão[6], Larissa Irigoyen Teixeira Barbosa[7]

[1]Universidade Evangélica de Goiás, Anápolis GO, Brazil.
[2]Universidade do Sul de Santa Catarina.
[3]Afya, Faculdade de Ciências Médicas de Palmas, Palmas TO, Brazil
[4]Centro Universitário de Patos de Minas, Patos de Minas MG, Brazil.
[5]Centro Universitário FIPMoc, Montes Claros MG, Brazil.
[6]Universidade Estadual de Campinas, Campinas SP, Brazil.
[7]Faculdade Santa Marcelina, São Paulo SP, Brazil

Correspondence: Abner Lucas Balduino de Souza; Email: aabnerlucass@gmail.com.

Background: BRAF V600E mutations are prevalent in childhood cancers and are associated with a diminished response to standard chemotherapy. Recent clinical trials have demonstrated the effectiveness of dabrafenib, both as a monotherapy and in combination with trametinib, in managing refractory mutant gliomas in patients of various ages. However, safety-related outcomes and clinical improvements of this therapeutic approach in the pediatric population remain unclear. **Objective:** Evaluate the efficacy and safety of dabrafenib and trametinib in children with gliomas harboring BRAF V600E mutations. **Methods:** PubMed, Embase and Cochrane databases were searched up to April 2024, following the recommendations in the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement. Included studies were peer-reviewed randomized controlled trials (RCTs) or non-randomized studies comparing the efficacy and safety of dabrafenib in combination with trametinib among pediatric patients (<18years) with BRAF V600E mutated-gliomas. Outcomes included overall response rate (ORR), clinical benefit rate (CBR), stable disease rate, complete response rate, and incidence of adverse events. Meta-analyses were conducted with random-effects models. **Results:** Four studies met the inclusion criteria, consisting of two RCTs and two non-randomized clinical trials, encompassing a total of 326 patients (with 198 receiving dual therapy). Among these, three studies focused on low-grade gliomas (LGG), while one targeted high-grade gliomas (HGG). The pooled ORR was 44.01% (95% confidence interval [CI] 37.12–50.90), accompanied by a CBR of 81.87% (95% CI 75.9187.83), stable disease rate of 50.23% (95% CI 43.29–57.17) and complete response rate of 1.35% (95% CI 0.00–3.31). Grade 1–4 adverse events were reported in 100% of patients, with pyrexia (61.21%, 95% CI 54.46–67.96), dry skin/skin toxicity (43.46%, 95% CI 36.56–50.36), and vomiting (30.7%, 95% CI 23.58–37.82) being the most common. Adverse events necessitating dose reduction or interruption occurred in 73.66% of patients (95% CI 66.86–80.46), while treatment discontinuation due to adverse events was observed in 9.76% (95% CI 5.20–14.32). **Conclusion:** Dabrafenib and trametinib exhibit promising efficacy in pediatric glioma patients with BRAF V600E mutations, as evidenced by favorable response rates. However, the occurrence of adverse events underscores the need for further research to elucidate the long-term safety and efficacy profile of this treatment in the pediatric population.

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Safety and efficacy of duloxetine for low back pain: a meta-analysis of randomized controlled trials

Mariana Rachas Reis[1] , Kamila Ferreira de Souza[2], Frederico de Sousa Marinho Mendes Filho[3], Thais Ferreira da Silveira[4], Artur Menegaz de Almeida[5], Ítalo Barros Andrade[6], Lilianne Rodrigues Fernandes[7], Vitor Maia Arca[8]

[1]Faculdade de Ciências Médicas da Santa Casa de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal do Pará, Altamira PA, Brazil.
[3]Universidade Federal do Amazonas, Manaus AM, Brazil.
[4]Centro Universitário FAMETRO, Manaus AM, Brazil.
[5]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[6]Faculdade Santo Agostinho de Vitória da Conquista, Vitória da Conquista BA, Brazil
[7]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.
[8]Universidade Federal de Pernambuco, Hospital das Clínicas, Departamento de Neurologia, Recife PE, Brazil.

Correspondence: Mariana Rachas Reis; Email: marianarachas@gmail.com.

Background: Managing low back pain (LBP) presents a significant healthcare challenge. Duloxetine, a selective dual neuronal serotonin and norepinephrine reuptake inhibitor, is believed to have beneficial effects in LBP patients. Despite previous studies testing duloxetine in LBP patients, new trials have been conducted on the subject requiring a more comprehensive analysis to update the body of evidence. **Objective:** To assess the safety and efficacy of duloxetine for treating patients with LBP. **Methods:** MEDLINE, Embase, and Cochrane databases were systematically searched for randomized placebo-controlled trials (RCT) that tested duloxetine in LBP patients. Data were examined using the Mantel-Haenszel method and 95% confidence intervals (CIs). Heterogeneity was assessed using I^2 statistics. This meta-analysis was performed to evaluate average pain intensity (Brief Pain Inventory [BPI]), clinician1s assessment of the overall disease severity (Clinical Global Impressions of Severity [CGI-S]), and safety (Treatment Emergent Adverse Events [TEAEs] and Serious Adverse Events [SAEs]). Statistical analysis was conducted using R, version 4.3.2. **Results:** A total of 4 randomized controlled trials and 1326 patients were included, of whom 659 received duloxetine 60 mg once daily. Compared with placebo treatment, duloxetine-treated patients presented

a statistically significant reduction in all domains of BPI-Severity: worst pain (MD -0.502; 95% CI -0.737, -0.266; P<0.001; I^2=0%); average pain (MD -0.569; 95% CI 0-.779, -0.359; P<0.001; I^2=0%) and least pain (MD -0.550; 95% CI -0.750, -0.350; P<0.001; I^2=0%). There was also a decrease in BPI-Interference (MD -0.509; 95% CI -0.847, -0.170; P=0.003; I^2=63%) and CGI-S (MD -0.255; 95% CI -0.362, -0.148; P<0.001; I^2=0%). Regarding safety, the duloxetine group had higher TEAE rate compared with placebo (40.87% vs 23.15%; risk ratio [RR] 1.984; 95% CI 1.286, 3.063; P=0.002; I^2=83%) and there were no significant differences in the occurrence of SAEs between groups (2.46% vs 1.19%; risk ratio [RR] 1.657; 95% CI 0.635, 4.323; P=0.302; I^2=16%). **Conclusion:** This meta-analysis underscores the effectiveness of duloxetine in improving outcomes in patients with LBP, as evidenced by improvements in CGI-S, BPI-Interference, and all domains of BPI-Severity. In terms of safety, the increased incidence of TEAEs in the duloxetine group, alongside no significant differences in the occurrence of SAEs, emphasize the importance of vigilant monitoring and risk assessment in clinical practice. These findings support duloxetine as a valuable treatment option for LBP, with careful management ensuring its safe and effective use.

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Safety and efficacy of stellate ganglion block in patients with subarachnoid hemorrhage: a single-arm meta-analysis

Paula Schumaker[1], Lara Luisa Lopes Chrisóstomo[2], Emanuelly Maria Lima Barbosa[3], Eduardo Vinicius Gurgel Costa Rego[3], Artur Menegaz de Almeida[4], Ítalo Barros Andrade[5], Michele Kreuz[6]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Universidade Federal do Vale do São Francisco, Paulo Afonso BA, Brazil.
[3]Universidade Federal do Amazonas, Manaus AM, Brazil.
[4]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[5]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[6]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Paula Schumaker; Email: paula.schumaker@uol.com.br.

Background: Stellate Ganglion Block (SGB) is widely used to block impaired sympathetic nerve activity, affecting physiologic functions such as heart rate and blood pressure. After a subarachnoid hemorrhage, there is usually an increase in the cerebral blood flow velocity (CBFV), at which SGB may be efficient in controlling cerebral hemodynamics. **Objective:** To assess the effects of SGB on cerebral hemodynamics and functional outcomes in patients with subarachnoid hemorrhage. **Methods:** MEDLINE, Scopus, and Web of Science databases were systematically searched until May 2024. Data was examined using 95% Confidence Intervals (CIs). Heterogeneity was assessed using I2 statistics. Meta-analysis was conducted to evaluate a possible link between the Stellate Ganglion Block and the Middle Cerebral Artery Cerebral Blood Flow Velocity (MCA CBFV) in patients with subarachnoid hemorrhage. RStudio version 4.3.2 was used for statistical analysis. **Results:** Six studies and 212 patients were included. There was a statistically significant difference between groups tending for the intervention regarding Middle Cerebral Artery Cerebral Blood Flow Velocity (MCA CBFV) 24 hours before SGB (MRAW 113.30 cm/sec; 95% CI: 71.48-155.12; P<0.01; I^2=98%). After 24 hours, both the CBFV on the ipsilateral side (MRAW 115.78 cm/sec; 95% CI; 93.17-138.40; P<0.01; I^2=95%) versus CBFV contralateral to the SBG (MRAW 100.79 cm/sec; 95% CI: 85.66-115.92; P<0.01; I^2=84%) showed a statistically significant better response in the intervention group. However, in the ipsilateral method, the CBFV was higher (115.78 cm/sec). Additionally, the Glasgow Outcome Scale (GOS) between I and III after 6 months of SGB showed a proportion of (PROP 0.393; 95% CI: 0.317-0.474; P=0.46; I^2=0%). **Conclusion:** In the present study, SGB showed beneficial outcomes for subarachnoid hemorrhage patients. However, further randomized controlled trials comparing bilateral SBG and its hemodynamic effect are needed to investigate possible outcome differences between each anatomical side for SBG application.

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Safety and efficacy of tixagevimab/cilgavimab for COVID-19 prophylaxis in immunosuppresed patients with multiple sclerosis or related disorders: a meta-analysis

Paulo Roberto Moss Lopes[1], Artur Menegaz de Almeida[2], Verônica Oliveira Rodrigues[3], Juliane Lins Orrico[4], Beatriz Ramos de Godoy[5], Gabriel Nobre dos Santos Alho[6], Ítalo Barros Andrade[7], Lilianne Rodrigues Fernandes[8]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[3]Universidade Federal do Pará, Belém PA, Brazil.
[4]Universidade de Pernambuco, Recife PE, Brazil.
[5]Universidade Federal do Paraná, Curitiba PR, Brazil.
[6]Faculdade Metropolitana, Manaus AM, Brazil.
[7]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[8]AFYA Faculdade de Ciências Médicas, Abaetuba PA, Brazil.

Correspondence: Paulo Roberto Moss Lopes; Email: paulormoss3@gmail.com.

Background: Patients on certain immunosuppressive therapies often fail to develop sufficient immunity after COVID-19 vaccination, making them more susceptible to severe disease outcomes. To address this vulnerability, a monoclonal antibody combination of tixagevimab and cilgavimab has been proposed for pre-exposure prophylaxis (PrEP) against COVID-19. This strategy may be efficient in patients with multiple sclerosis (MS), neuromyelitis optica spectrum disorder (NMOSD), or myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD). However, the PrEP efficacy in reducing the incidence of COVID-19 in this population has not yet been fully elucidated. **Objective:** To assess the safety and efficacy of tixagevimab and cilgavimab pre-exposure prophylaxis against COVID-19 in patients with MS, NMOSD, or MOGAD on sphingosine 1-phosphate receptor modulators or B-cell depleting therapy. **Methods:** MEDLINE, Scopus, and Web of Science databases were systematically searched until May 13, 2024. We included trials that involved patients with MS, NMOSD or MOGAD who underwent Tixagevimab/Cilgavimab dual therapy. Data were analyzed using the Mantel-Haenszel method to calculate risk ratios (RR) and 95% confidence intervals (CI). Heterogeneity was assessed using I^2 statistics. Statistical analysis was performed using R, version 4.3.2. **Results:** We included five studies with a total of 797 participants, of whom 434 (54%) underwent Tixagevimab/Cilgavimab therapy. The primary outcome was COVID-19 infection rate between the groups. The number of COVID-19 infection events recorded in the intervention group was 69, whereas 74 events were recorded in the control group. However, no statistically significant difference was seen between groups (16% vs 20%; risk ratio [RR] 0.87; 95% CI 0.51-1.49; P=0.612661; I^2=42%). **Conclusion:** Our study found no statistically significant difference between groups concerning COVID-19 infection rates in patients treated with Tixagevimab/Cilgavimab or placebo. The potential benefit suggested by the risk ratio requires further investigation with larger studies to confirm safety and efficacy in patients with MS, NMOSD, or MOGAD on immunosuppressive therapies.

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Safety and efficacy of topiramate in migraine prophylaxis in younger patients: a meta-analysis

Natália Gabrielli Silva Alves[1], Lais Cabral Almeida[1], Ana Paula Alves Lima[1], Luana Maria da Silva Rodrigues[1], Artur Menegaz de Almeida[2], Ítalo Barros Andrade[3], Giovanna Molitor Perini[1], Michele Kreuz[4]

[1]Centro Universitário de Várzea Grande, Várzea Grande MT, Brazil.
[2]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[3]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[4]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Natalia Gabrielli Silva Alves; Email: alvesn544@gmail.com.

Background: Prophylactic treatment of migraine using the AMPA (amino-3-hydroxy-5-methyl-4-isoxazolepropionic) receptor-blocking anticonvulsant Topiramate is associated with adverse events and questionable efficacy in youngers. The combination with adjunctive vitamin D3 is discussed to be more efficient and safer in younger patients with migraine. **Objective:** Compare the safety and efficacy of Topiramate alone or in combination with vitamin D3 versus placebo in the prophylaxis of migraine in younger patients. **Methods:** PUBMED, Scopus, and Web of Science for randomized controlled trials assessing the safety and efficacy of topiramate in younger patients for migraine prevention, alone or with vitamin D. Data were analyzed using Mantel-Haenszel statistics with 95% confidence intervals. Patients older than 18 years were excluded. Statistical analysis was performed using R version 4.3.2. **Results:** We included 6 RCTs comprising 653 patients, of which 306 underwent isolated use of Topiramate 100mg and 234 received a placebo. The subgroup analysis of Topiramate (2mg/kg/day) plus adjunctive vitamin D3 (daily dose 5,000 IU daily or 500,000 IU weekly doses) encompassed 57 patients receiving the combination treatment and 56 receiving a placebo. The topiramate therapy in association with vitamin D3 demonstrated a statistically significant difference in reducing migraine frequency (MD -3.66; 95% CI -4.33 to -2.98; P< 0.000001; I^2=0), while topiramate alone did not show any difference in comparison to the placebo group (MD -0.92; 95% CI -2.39 to 0.55; P=0.22; I^2=75%). The overall frequency reduction tended to the intervention group (MD -1.99; 95% CI -3.78 to -0.19; P=0.029; I^2=93%). PEDMIDAS reduction rate was greater with topiramate alone (MD -8.51; 95% CI -16.53 to -0.49; P=0.03; I^2=59%) when compared with the combination with vitamin D (MD -6.24; 95% CI -9.84 to -2.63; P=0.00069; I^2=68%). Concerning the occurrence of adverse effects, our findings showed that patients receiving topiramate associated with vitamin D3 (3,50% vs. 5,35%; 95% CI 0.11 to 3.91; P=0.64; I^2=0%) are at no increased risk for anorexia than those receiving topiramate alone (14,38% vs. 2,91%; 95% CI 2.42 to 11.42; P=0.000027; I^2=0%). The overall risk for anorexia tended in favor of te placebo group (12,57% vs. 3,43%; 95% CI 1.86 to 7.68; P=0.000246; I^2=20%). Paresthesia was reported only in studies comparing topiramate alone versus placebo, which analysis showed an increased risk for the intervention group (35% vs 5%; RR 6.19; 95% CI 2.81-13.62; P=0.000006; I^2=18%), as well as the adverse effect of fatigue (18% vs 10%; RR 1.69; 95% CI 1.06-2.69; P=0.027752; I^2=0%). **Conclusion:** Topiramate as monotherapy showed low safety and efficacy in reducing the frequency of migraines, with side effects such as paresthesia, anorexia, and fatigue. Conversely, the combination with vitamin D3 showed better results in preventing migraines in younger patients, with a lower risk of adverse events.

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Safety of magnetic resonance-guided focused ultrasound in Parkinson's disease: a meta-analysis of randomized controlled trials

Abner Lucas Balduino de Souza[1], Thaís Pereira Mendes[2], Marco Antonnio Rocha dos Santos[3], Pierludovico Moro[4], Lauren Nirta[5], Laura de Lima Xavier[6]

[1]Universidade Evangélica de Goiás, Anápolis GO, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Universidade do Planalto Catarinense, Lages SC, Brazil.
[4]Università La Sapienza, Rome, Itay.
[5]Pusan National University Yangsan Hospital, Department of Rehabilitation Medicine, Yangsan, South Korea.
[6]University of North Carolina at Chapel Hill, USA.

Correspondence: Abner Lucas Balduino de Souza; Email: aabnerlucass@gmail.com.

Background: Magnetic Resonance-guided Focused Ultrasound (MRgFUS) is well-established for the treatment of essential tremor and is emerging as a promising non-invasive treatment for Parkinson's disease (PD). However, the safety of MRgFUS among patients with PD is not well established. **Objective:** To evaluate the incidence of adverse events among medication-refractory patients with PD treated with MRgFUS. **Methods:** PubMed, Embase, Scopus, and Cochrane databases were searched up to December 2023, following the recommendations in the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement. Included studies were peer-reviewed randomized controlled trials (RCTs) comparing MRgFUS to sham-procedure among patients with medication-refractory PD. The Cochrane risk-of-bias tool for randomized trials (RoB 2) was used for quality assessment. **Results:** Three RCTs were included, with a total of 161 patients (MRgFUS, n=115). Dizziness was the most commonly reported procedure-related adverse event (odds ratio [OR] 4.25; 95% confidence interval [CI] 1.23-14.64, p=0.02, I^2=0%). Pin-site complications (OR 0.52, 95% CI 0.23-1.20, p = 0.13, I^2 = 0%) and headache (OR 0.88, 95% CI 0.19-4.16, I^2=48%) were also notable. Given the heterogeneity related to headache, a leave-one-out sensitivity analysis was conducted, resulting in a reduction in the proportion of variance (I^2) to 0% (OR 2.14, 95% CI 0.47-9.67; p = 0.32). The most notable ablation-related adverse events included gait disturbances (OR 4.53, 95% CI 0.79-26.07, p=0.09, I^2=0%), primarily ataxia, and facial disturbances (OR 2.93, 95% CI 0.49-17.51). Most ablation-related adverse events persisted at 3 and 4-month follow-up. While no significant differences were observed between intervention and sham groups for life-threatening adverse events (OR 1.76, 95% CI 0.55-5.65; p=0.35; I^2=0%), severe adverse events included pulmonary embolism, persistent hemiparesis, cholecystitis, and falls. RoB2 analysis indicated a low risk of bias for all included studies. **Conclusion:** MRgFUS demonstrated a reasonable safety profile for the treatment of patients with medication-refractory PD. Statistically significant associations were observed for procedure-related dizziness, while gait disturbances, despite being relevant, were not statistically significant. These findings emphasize the need for further multicenter studies to clarify comprehensively the safety profiles of MRgFUS and its implementation.

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SAH: analysis of a sample from a prospective study in a Brazilian hospital

Fernanda Firmiano Casarotto[1], Hilton Mariano da Silva Júnior[1], Caroline Matielo Ricci[1], Maria Eugênia Babbini Valentino Rosa[1], Lívia Reami Braga[1], Marina Ramos Nannini[1], Lucas da Silva Oliveira[1], João Caetano Coutinho Mariuzzo[1], Isadora Miarelli Fortuna[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Fernanda Firmiano Casarotto; Email: ferfirmiano@hotmail.com.

Background: Subarachnoid hemorrhage (SAH) is a neurosurgical emergency associated with high morbidity and mortality. **Objective:** To prospectively evaluate all cases of SAH attended to by the service during 2023-24, seeking to detail their clinical presentation and the accuracy of complementary diagnostic methods. **Methods:** The sample was 127 patients (82 women - 64.6%), with an average age of 50.4 years (SD of 12.2) presenting with a clinical picture suggestive of SAH confirmed by neuroimaging. All patients were evaluated by a single examiner, who completed a questionnaire developed specifically for this study. **Results:** The most frequent presentation (64 patients - 50.4%) was a sudden headache not preceded by sentinel headache. Sentinel headache was reported by forty patients (31.5%) who sought emergency care for an unusual headache. Thirteen patients (10.2%) presented with a clinical picture suggestive of syncope as the first manifestation of bleeding. On physical examination, the average Glasgow Coma Scale score was 12.8; the average systolic blood pressure was 160 (SD: 33) and the average diastolic blood pressure was (SD: 21). Nuchal rigidity was found in 85 patients (67%), and 49 patients (38.6%) presented with focal, motor and sensory signs. Fifty-three patients (42%) had Hunt-Hess scores between 1 and 3. Cranial CT showed some SAH-related alteration in 103 cases (81%). Cerebral angiography was performed on 124 patients (3 died before the exam): 66 (53%) with aneurysm without vasospasm, 56 (45%) with aneurysm and vasospasm, and 2 (1.6%) showed no abnormalities on the first exam. **Conclusion:** The study demonstrated that a significant portion of patients presented differently from the classic descriptions in the literature of an atraumatic SAH, contributing to diagnostic errors during the initial assessment of a condition with high morbidity and mortality, even when diagnosed early and appropriately.

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Saliva whole exome sequencing utility for the molecular diagnosis of CPEO

Cristiane Araujo Martins Moreno[1], Alulin Tácio Quadros Santos Monteiro Fonseca[1], Clara Gontijo Camelo[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Cristiane Araujo Martins Moreno; Email: cristianeammoreno@gmail.com.

Background: Chronic progressive external ophthalmoplegia (CPEO) is the most common manifestation of mitochondrial diseases and is characterized by bilateral symmetrical progressive ptosis and reduced ocular motility. CPEO can be isolated or accompanied by a clinical feature of systemic involvement of mitochondrial dysfunction (CPEO plus syndrome). Both mtDNA and nDNA can be affected in mitochondrial diseases. Most common genetic variants that cause PEO are point mutations at mtDNA, large scale deletions at mtDNA or even mutation in nuclear genes interfering at mitochondrial function. The most common method used for the molecular studies in these conditions includes direct mitochondrial from muscle or urine DNA sequencing. Diagnostic rates from CPEO using saliva WES are not well defined in the literature. **Objective:** Here we present a molecular analysis of saliva WES from 28 patients presenting PEO. Patients and **Methods:** Patients had a confirmed diagnosis of mitochondrial myopathy and presented PEO at clinical exam. Whole exome sequencing was performed from saliva DNA. Large Scale deletions were also analyzed from BAM files using custom scripts. **Results:** 28 patients were included in the study. Patients were 46 ±22 years old (6 male e 21 female). 19 patients presented CPEO and nine patients presented CPEO-plus. A conclusive molecular confirmation was obtained for seven patients in the CPEO group: POLG (n=4), MYH2(n=1), DGUOK(n=1), MSTO1(n=1), PNPLA8(n=1) and mt large-scale deletion (n=1); and four patients in CPEO+ group: MT-TK (n=1), SLC24A4 (n=1), TAMM41 (=n1) and TRMT10 (n=1). **Conclusion:** CPEO is a common manifestation among mitochondrial diseases. WES was able to analyze both nuclear DNA and mtDNA and found a definite molecular diagnosis in 36% of CPEO and 44% of CPEO+, oh those, two patients from the CEPO group carried mutations in mtDNA. Large-scale deletion was detected in one single patient. In order to compare both techniques, we'll perform a WES from muscle DNA as the next step.

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Same-day discharge or postoperative day 1 in patients who underwent brain tumor resection: a systematic review and meta-analysis

Maria Lúcia Ferreira Rodrigues[1], Leticia Albuquerque Cossich[2], Jeniffer Caroline Bennes Martelet Simões[3], André Welisson Marques de Araújo[2], Artur Menegaz de Almeida[4], Ítalo Barros Andrade[5], Lilianne Rodrigues Fernandes[6], Michele Kreuz[7]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[3]Faculdade Metropolitana de Manaus, Manaus AM, Brazil.
[4]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[5]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[6]Afya Faculdade de Ciências Médicas, Abaetetuba PA, Brazil.
[7]Universidade Luterana do Brasil, Canoas RS, Brazil.

Correspondence: Maria Lúcia Ferreira Rodrigues; Email: marialuccia.rodrigues@gmail.com.

Background: The hospital length of stay (LOS) after surgical procedures, such as brain tumor resection, tends to be increasingly shorter, as it seems to be associated with lower complication rates, reduced risk of infection, and better cost-benefit. However, the safety of shorter LOS compared to longer stays in patients who underwent those kinds of procedures remains unclear. **Objective:** To assess the safety of same-day discharge compared to postoperative day 1 in patients who underwent brain tumor resection. **Methods:** We searched MEDLINE, Scopus, and Web of Science databases for studies that assessed shorter LOS among patients who underwent brain tumor resection intervention. Data were examined using the Mantel-Haenszel method and 95% confidence intervals (CIs). Heterogeneity was assessed using I^2 statistics. R, version 4.3.2., was used for statistical analysis. **Results:** Five retrospective cohort studies encompassing 1,102 patients with resectable brain tumors were included, of whom 385 (34.94%) underwent early discharge. In comparison with later discharge, same-day discharge showed no statistically significant difference in 30-day readmission rates (OR 0.54; 95% CI 0.28-1.07; p=0.078), with minimal heterogeneity (I^2=0). Similarly, no significant differences were noted in postoperative neurological deficits (RR 0.73; 95% CI 0.35-1.54; p=0.413), wound complications (RR 1.02; 95% CI 0.41-2.52; p=0.968), or mortality (RR 0.36; 95% CI 0.06-2.11; p=0.255). **Conclusion:** In this meta-analysis, no statistically significant differences were seen regarding neurological complications, wound complications, mortality, and readmission 1-30 days postoperatively, which showed SDD and POD1. Further methodologically sound randomized controlled trials are needed to produce a more accurate analysis.

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Scar dancing syndrome: a rare presentation of a unique condition

Bruno Henrique Carneiro Costa Filho[1], Vitor Maia Arca[1], Víctor de Moraes Chagas[1], João Pedro Matos de Santana[1], Ana Eunice Oliveira Rodrigues[2], Míriam Carvalho Soares[2], Marcos Eugenio Ramalho Bezerra[1]

[1]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Bruno Henrique Carneiro Costa Filho; Email: brunohccosta97@gmail.com.

Case presentation: A 40-year-old man with suffered a thoracic spinal cord trauma, at T8 level, secondary to a car accident. He was treated surgically and did not present at first any weakness or sensory sequelae related to the spinal cord injury. However, he began to complain of pain and involuntary contractions around the surgical scar, starting two months after the event. In this context, it was decided for botulinum toxin (BT) treatment, with 100 units of onabotulinum toxin A applied. It was injected in the greater and lesser rhomboid muscles bilaterally, as well as supraspinatus, latissimus dorsi, teres minor, and paravertebral muscles on the right. After 28 days of applying the toxin, the patient reported a subjective perception of 50% improvement of muscle contractions. Furthermore he reported a pain intensity reduction of 3 points in the visual analog scale (from 7 to 4). **Discussion:** Peripherally induced movement disorders (PIMD) can be defined as abnormal and involuntary movements that occur after a traumatic event involving cranial or peripheral nerves, as well as its roots. The diagnostic criteria requires a clear anatomical and temporal association with the trauma to determine causality. The phenomenology is variable and may present mainly as dystonia, tremors, and myoclonus. Among cases of PIMD, the rare dancing scar syndrome has been described, as characterized by the presence of involuntary movements around the site of a surgical incision. In a previously published systematic review, the average time for symptom onset was 21 days, with 95% of patients starting symptoms in less than a year. The presence of sensory symptoms was present in 42% of patients. The main sensory symptoms are hyperalgesia, hyperesthesia, hypoalgesia, hypoesthesia, or the emergence of a complex regional pain syndrome. In a previous case report series, in patients who received treatment with botulinum toxin, around 50% of cases showed symptomatic improvement after application. A common factor among patients who had better responses was the presence of neuropathic pain. Neuropathic pain was also present in the clinical picture of the patient described in this case report, suggesting a greater benefit of this treatment for this group of patients. **Final comments:** We present a case of scar dancing syndrome, a rare presentation of PIMDs, for which the best therapeutic option is not yet established. Even though BT injection did not cease the abnormal movements completely, it's role in controlling the patient's pain was remarkable.

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Schistosomal myelopathy mimicking longitudinally extensive myelitis in neuroimaging

Maria Weryca de Souza Belo Silva[1], Manuel Moreira Neto[1], Alberto Pereira Firmino Filho[1], Fátima de Menezes Dantas[1], Felipe Toscano Lins de Menezes[1], Felipe Olobardi Freire[1], Francisco Gil Sarmento Neto[1], Paulo Santiago de Morais Brito[1], Rodrigo Alencar e Silva[1], Nathalia Lindsay Pereira de Queiroz[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Maria Weryca de Souza Belo Silva; Email: mariaweryca@hotmail.com.

Case presentation: Male patient, 16 years old, presented with a complaint of sudden, intense pain in his thighs and calves, progressing to paresthesias in the soles of his feet and difficulty walking. He was admitted to a referral hospital, where he developed urinary and fecal retention. Neurological examination revealed flaccid paraplegia, areflexia in the lower limbs, and painful hypoesthesia up to the T12 spinal level. During the investigation, neuroimaging showed a pattern of longitudinally extensive myelitis, suggesting autoimmune/demyelinating diseases of the central nervous system that present with such findings. However, what appeared to be typical extensive myelitis was actually a spinal cord imaging alteration mimicking longitudinally extensive myelitis. After completing the investigative exams, the diagnosis of schistosomal myeloradiculitis was confirmed. **Discussion:** Schistosomiasis is a prevalent disease in tropical regions. This condition can affect the central nervous system, with schistosomal myeloradiculitis being its most common and severe form. In this manifestation, magnetic resonance imaging often reveals alterations such as spinal cord and/or cauda equina root enlargement on T1 sequences; hyperintensity in the affected region on T2 sequences; and contrast enhancement, usually heterogeneous and granular. In the reported case, these findings were visible on neuroimaging; nevertheless, the appearance of longitudinally extensive myelitis initially directed clinical reasoning towards other hypotheses. **Final comments:** The presented case illustrates a relatively common condition, especially in endemic areas. Notably, in addition to the typical neuroimaging alterations found in schistosomal myelopathy, an imaging pattern mimicking longitudinally extensive myelitis was also observed. This led to the consideration of various other hypotheses until the case was resolved through histopathological examination.

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Screening for SCA27B in a cohort of Brazilian patients with unsolved ataxias

Amanda de Jesus Araujo Dias[1], Cynthia Silveira[1], Fabricio Diniz de Lima[1], José Luiz Pedroso[2], Adriana Mendes Vinagre[1], Luciana Cardoso Bonadia[1], Luíza Alves Corazza[1], Orlando Graziani Povoas Barsottini[2], Thiago Yoshinaga Tonholo Silva[2], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Amanda de Jesus Araujo Dias; Email: a291103@dac.unicamp.br.

Spinocerebellar Ataxia 27B (SCA27B) is a recently described autosomal dominant neurodegenerative disease, prevalent in Europe and North America, but not yet properly investigated in Latin America. It is caused by a heterozygous trinucleotide repeat expansion (GAA) >300 in FGF14 gene. Since it is an expansion disorder, diagnosis requires specific molecular techniques. In this scenario, the aim of this study is to screen a large cohort of adults with unsolved ataxias for SCA27B. To date, 108 patients with ataxia were recruited, including patients negative for SCA1,2,3,6,7 and 8. DNA was extracted from peripheral blood and analyzed using different PCR techniques. Initially, long-range PCR (LR-PCR) was used to exclude patients with two alleles within the normal repeat range, 90 patients were negative for FGF14 expansion and eighteen patients showing only one allele in normal size range. Repeat-primed PCR was carried out to identify the expansion and in five out of the 18 analyzed patients the expansion pattern was detected (4.6%). The phenotype was a late-onset slowly progressive ataxia in all cases, with paroxysmal symptoms in 3/5 and pyramidal signs in 1/5. None of them had sensory manifestations. The mean age at onset was 55 years. Our results are consistent with previous publications, which showed that SCA27B ataxia presents initial symptoms at an advanced age and has slow progression. The prevalence of SCA27B we found (4.6%) was much smaller than in French Canadian (61%) and European (18%) cohorts. In conclusion, LR-PCR followed by RP-PCR is a reliable and fast approach to diagnose SCA27B. The phenotype of Brazilian patients with SCA27B is similar to previous reports, however considering that most of the patients included in this study have unsolved ataxia, often with distinct clinical manifestation, the frequency is much smaller. Further studies are needed to understand the genetic epidemiology of this new disease beyond Europe and North America.

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Seasonality of fatigue in Parkinson's disease in a high temperature location

Luma Rodrigues da Silva[1], Lara Beatriz Alves Batista[1], Pedro Henrique Ximenes Ramalho Barros[1], Isabel Maria Oliveira Macêdo Lima[1], Sabrina Ruthiele Santos de Carvalho[1], Jonatas Paulino da Cunha Monteiro Ribeiro[1], Samuel de Castro Campos[1], Gustavo Viana Castro[1], José Henrique de Melo Feitosa[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.

Correspondence: Luma Rodrigues da Silva; Email: lumarodrigues@ufpi.edu.br.

Background: Fatigue is one of the most common and disabling non-motor symptoms in Parkinson's disease (PD), and its assessment is important to measure the quality of life of these patients. Simultaneously, rising temperatures due to global warming may aggravate conditions such as fatigue. The city of Teresina has a warming scenario that will only become a reality in some regions of the globe in the next decade, making it relevant to analyze the influence of temperature on neurological diseases in this city. **Objective:** To assess the severity of fatigue and quality of life in patients with Idiopathic Parkinson's Disease (IPD) according to seasonal variations in a high temperature location. **Methods:** This is a longitudinal observational study. It included patients with IPD according to the UK Parkinson's Disease Society Brain Bank and Movement Disorders Society criteria, followed at a movement disorder center in Teresina-PI. Assessments took place in the high temperature period (September to December) and the mild temperature period (January to April), using the Unified Parkinson's Disease Rating Scale (UPDRS), Montreal Cognitive Assessment (MoCA), Parkinson's Disease Questionnaire-39 (PDQ-39), and Fatigue Severity Scale (FSS). The R software (R Core Team) was used for statistical data analysis. The normality assumption of the data was verified by the Shapiro-Wilk test. The Wilcoxon test was used to compare the UPDRS scale values in the high and mild temperature periods. The paired t-test was used to compare the levodopa equivalent dose (LED) values and the FSS and PDQ-39 scales in the high temperature period (HTP) and mild temperature period (MTP). The criterion for a statistically significant difference was p-value <0.05. **Results:** A sample of 16 patients with a mean age of 65 years, 75% male, with a mean duration of symptoms of 9.06 years and a mean diagnosis time of 8.12 years, the mean MoCA score was 18.81. The mean UPDRS was 33.18 in HTP and 31.64 in MTP (p=0.9645). The average LED in HTP was 617.31, and in MTP was 667.37 (p=0.1730). The average FSS in HTP was 4.56, while in MTP was 4.53 (p=0.9227). The average PDQ-39 was 46.33 in HTP and 45.36 in MTP (p=0.6906). There were no statistically significant differences between the high and mild temperature periods. **Conclusion:** The severity of fatigue and quality of life in patients with Idiopathic PD did not differ between seasons in a high temperature location. The restricted sample became the current findings limited.

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Secondary epileptogenesis in acquired hemispheric lesions: a case of too-little-too-late?

Carolina Heck Haubrich[1], William Alves Martins[1], Rafael Paglioli[1], Thomas More Frigeri[1], Fernanda Schuh[1], Eliseu Paglioli[1], André Palmini[1]

[1]Universidade Católica do Rio Grande do Sul, Hospital São Lucas da Pontifícia, Porto Alegre RS, Brazil.

Correspondence: Carolina Heck Haubrich; Email: carolhhaubrich@gmail.com.

Case presentation: Case 1: An 18-year-old female sustained a traumatic brain injury at age two, resulting in multiple cerebral contusions, hydrocephalus, and subsequent epilepsy with left hemiparesis. Despite normal developmental milestones, she suffered daily seizures characterized by altered consciousness, bilateral automatisms, and occasional tonic extension of both arms. MRI revealed significant atrophy, gliosis, and hyperintensities in the right hemisphere. Scalp EEG localized seizure activity to the right temporal and frontal lobes. Initial multi-lobar resection failed to achieve seizure control. Eight years later, ongoing refractory seizures prompted re-evaluation, revealing only frontoinsular connections between hemispheres and pronounced atrophy in the presumed "healthy" hemisphere. EEG indicated interictal discharges in the left frontotemporal region and ictal activity in the remaining right frontal region. Subsequently, a right hemispherectomy was performed. Initial seizure freedom lasted one month, followed by recurrence characterized by altered consciousness and automatisms. Adjustments in antiepileptic medications reduced seizure frequency to once per week. Case 2: A 15-year-old female presented with seizures from birth, featuring tonic extension of the right arm, dystonic posture of the left arm, and tonic eye and neck deviation to the left. MRI showed a congenital lesion affecting the entire right hemisphere, corroborated by ictal EEG showing hemisphere involvement. She underwent functional hemispherectomy and initially experienced three months of seizure freedom before relapse. Further investigation revealed no residual connections between the diseased hemisphere and healthy brain tissue. MRI exhibited diffuse cortical atrophy, primarily in frontal regions. Co-registered ictal SPECT with MRI (SISCOM) identified contralateral seizure activity, pinpointing it to the pre-motor regions. Despite medication adjustments, she continues to experience weekly seizures. **Discussion:** Epilepsy surgery is typically effective for patients with focal lesions, aiming for positive outcomes through meticulous resection planning, medication optimization, and surgical feasibility. However, the recurrence of seizures in the "healthy" hemisphere in both cases underscores potential challenges and complications. Hidden focal cortical dysplasia may manifest with subtle contralateral disease signs, as seen in Case 1, leading to unexpected contralateral seizure recurrence. Animal studies suggest prolonged severe epileptic activity may induce healthy neurons to synchronize into seizure activity. **Final comments:** Early surgical evaluation is critical as epilepsy surgery offers potential cure for severe cases. Prolonged epilepsy, even with acquired lesions, risks irreversible cortical damage and may predispose healthy neurons to epileptogenic changes, emphasizing the importance of timely intervention and comprehensive post-operative management.

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Secondary nummular headache: a case report

Bibiana Mayer[1], Chadi Emil Adamo[2]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.

[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Bibiana Mayer; Email: bibiana_mayer@hotmail.com.

G.V.S., male, 24 years old. One year ago, he used a psychostimulant (lisdexamfetamine 70 mg) for the first time before taking an exam, resulting in body tingling, headache, and diaphoresis, associated with tachycardia of 160 beats per minute. In the emergency room, an electrocardiogram revealed supraventricular tachycardia and a hypertensive peak. Heart rate control and analgesia were administered, leading to remission of the acute episode. Subsequently, he experienced insomnia with difficulty initiating sleep. He then developed a chronic headache localized to the high convexity, well-demarcated, with an area of approximately 3.5x2.5 cm, characterized by dysesthesias, tingling, allodynia, and pressure pain, occurring daily. The patient had previously undergone occipital nerve blocks, radiofrequency, transcranial magnetic stimulation, and prophylactic medication with topiramate, all of which showed minimal efficacy. Magnetic resonance imaging of the brain revealed a small extradural hydrocephalus adjacent to the left superior frontal gyrus and ipsilateral bone thinning. Consequently, a diagnosis of secondary nummular headache was made, secondary to the structural alteration. Beta-blocker therapy with propranolol was initiated, gradually and cautiously increased to 160 mg/day (progressing with satisfactory response), and nortriptyline was introduced, gradually increased to 50 mg/day. Upon reevaluation, the patient reported substantial improvement after three months of treatment. The pain in nummular headache is localized to a round or elliptical focus that is fixed in place, ranging in size from 1 to 6 cm and may be continuous or intermittent. In rare cases, it may be multifocal. Nummular headache is usually continuous with exacerbations, although it can be episodic. There may be focal sensory disturbances, such as allodynia or hypesthesia. The pathophysiology is unknown, although a focal neuropathic disorder has been suggested. In the authors' experience, the presentation is best treated by detailing the entire phenotype and treating the underlying primary headache disorder. Local dermatologic or bony lesions must be excluded by examination and investigation. Treatment includes tricyclics, such as amitriptyline, or anticonvulsants, such as topiramate, valproate, or gabapentin. Additionally, propranolol has been mentioned as a treatment option. In this particular case, reducing the arterial and venous gradient with propranolol was found to be an effective treatment for the patient. The condition is considered difficult to treat, although this is complex in relation to any underlying headache disorder.

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Seizure outcome in patients with malformations of cortical development other than focal cortical dysplasia in a tertiary center

Athos Paixão Silva Santos[1], Marina Koutsodontis Machado Alvim[1], Clarissa Lin Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Athos Paixão Silva Santos; Email: a231744@dac.unicamp.br.

Background: Malformations of cortical development (MCD) comprise a vast and heterogeneous group of disrupted cerebral cortex formation. The MCD may be classified according to the predominant embryological stage compromised: a) abnormal proliferation or increased apoptosis of neurons or glial cells (microcephaly, hemimegalencephaly, focal cortical dysplasia - FCD); b) abnormal neuron migration (heterotopia, lissencephaly-pachygyria); c) abnormal cortical organization (polymicrogyria, schizencephaly). MCDs are the most common cause of refractory focal epilepsy in adults after hippocampal sclerosis and the most frequent in children. **Objective:** To evaluate seizure outcomes in patients with different types of epilepsy caused by MCD other than FCD. **Methods:** The clinical data of 36 patients with MCD (excluding FCD) and epilepsy from tertiary center outpatient clinics were obtained from medical records. **Results:** The mean follow-up was 19.5 years. Two patients had hemimegalencephaly, 14 heterotopia (2 double cortex and 12 nodular heterotopia), 8 polymicrogyria, 7 schizencephaly associated with polymicrogyria, and 5 had schizencephaly associated with polymicrogyria and heterotopia. One patient underwent callosotomy and 4 patients underwent resective surgery: 2 with associated FCD (1 with hemimegalencephaly and 1 with polymicrogyria), and 2 with associated hippocampal sclerosis (1 with nodular heterotopia and 1 with polymicrogyria, schizencephaly and heterotopia combined). Of these, one (25%) was seizure-free after the procedure, one (25%) was classified as Engel IB, one (25%) as class IIA, and one (25%) as class IVA. Only 2 patients were without antiseizure medication (one after surgery and one not operated). The remaining patients were taking 1 to 5 ASM (mean 2.6). Five of the 36 (14%) patients were seizure-free for at least one year: 3 with nodular heterotopia, 2 with polymicrogyria, and one with polymicrogyria, schizencephaly, and heterotopia combined. The mean seizure frequency was 23.5 per month in patients with nodular heterotopia, 15.5 per month in those with double cortex, 38.5 per month in those with polymicrogyria, 2 per month in those with polymicrogyria with schizencephaly, 14 per month in those with polymicrogyria, schizencephaly, and heterotopia combined. Family history of epilepsy was present in 10/36 (28%): 2 patients with nodular heterotopia, one with double cortex, one with polymicrogyria, 4 with polymicrogyria and schizencephaly, and one with polymicrogyria, schizencephaly, and heterotopia combined. **Conclusion:** This tertiary center cohort study characterizes the clinical data from patients with cortical malformation other than FCD. Interestingly, a few patients (14%) were under seizure control with the use of ASM. Among the four patients who underwent surgical treatment, excluding callosotomy, 3 (75%) had significant improvement (Engel IA-IIA).

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Seizure outcomes in patients with burned-out hippocampus syndrome operated after noninvasive investigation

Leslie Ortega Hernandez[1], Renata Torres Jaimes[1], Marina Alvim[1], Enrico Ghizoni[1], Clarissa Lin Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Leslie Yadira Ortega Hernandez; Email: leslie-ortega@hotmail.com.

Background: Temporal lobe epilepsy secondary to hippocampal sclerosis (MTLE-HS) is the most prevalent subtype of refractory epilepsy among adult patients undergoing presurgical evaluation. The definition of the epileptogenic zone requires the concordance of clinical, imaging, and electrophysiological data. However, some patients with severe hippocampal sclerosis have an unusual spread of seizure discharges, leading to a significant discrepancy between MRI findings and scalp ictal EEG recordings, also known as a "burned-out hippocampus syndrome" (BHS), which makes the surgical decision due to suspicion of bilateral disease. Previous reports have shown that the intracranial EEG of these patients localizes the seizure onset in the atrophic hippocampus with a spread to the contralateral temporal neocortex before the ipsilateral side. **Objective:** To evaluate the postoperative outcome of patients with BHS operated with noninvasive investigation. **Methods:** We retrospectively reviewed the medical records of patients with temporal lobe epilepsy who

underwent surgery at the Hospital das Clínicas of Unicamp from 2011 to 2024 with the following criteria: a) unilateral hippocampal sclerosis and b) surface electroencephalogram with onset of the majority of ictal activity in the contralateral side and few or none on the side of hippocampal atrophy. **Results:** Of all patients with MTLE-HS who underwent surgery at this time, 5 met the diagnostic criteria for BHS, four women and one man, with a mean age onset of epilepsy of 21.2 years, mean epilepsy duration of 26.4 years, and mean seizure frequency prior to surgery of 8.9 seizures per month. Four patients had right, and one had left HS. Ictal activity was discordant with hippocampal atrophy in 75% of the records. The unilateral interictal activity was discordant in 28.9% of the records, and bilateral interictal EEG activity was predominantly contralateral to the affected hippocampus in 50% of the cases. FDG-PET/SPECT scans showed hypometabolism or ictal hyperperfusion lateralizing to the side of the hippocampal sclerosis in all patients. The surgical decision was based on all noninvasive investigations, including neuropsychological evaluation. At the last postoperative follow-up (mean 5,8 years) 3 (60%) were classified as Engel Ia, one (20%) as Engel Ib, and one (20%) as Engel IIb. **Conclusion:** In this series of BHS, all patients had excellent postoperative outcomes, becoming seizure-free or almost seizure-free. Some patients with MRI signs of unilateral hippocampal sclerosis have scalp ictal activity that is falsely lateralizing (BHS), which does not contraindicate surgical management when other studies are congruent. These patients may not need intracranial EEG recordings when semiology, neuropsychological investigation, and other noninvasive techniques for localizing the epileptogenic zone, such as FDG-PET and ictal SPECT, lateralize to the side of the HS.

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Seizures and myoclonus in systemic lupus erythematosus: an uncommon neurological presentation in a young woman

Carolina Oldoni[1], Alexandre Baldissera[1], Eduardo Drews Amorim[1], Raphael Machado Castilhos[1], Artur Francisco Schumacher Schuh[2], Marino Muxfeldt Bianchin[2]

[1]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Eduardo Drews Amorim; Email: eduardodrewsamorim@gmail.com.

Case presentation: A 25-year-old previously healthy woman presented to our emergency department with a one-month history of intermittent fever. Two days before arriving at the emergency department, she developed generalized myoclonus. Upon arrival, she experienced a focal seizure in her left upper limb that evolved into bilateral tonic-clonic seizures. Valproic acid and clobazam led to minimal improvement in the myoclonus. Brain MRI was unremarkable. Cerebrospinal fluid (CSF) analysis showed 9 lymphocytes/dL, normal glucose, and protein levels. Extensive testing for infectious pathogens in blood and CSF was negative. The patient had normochromic normocytic anemia, leukopenia, a positive Coombs test, decreased C3 and C4 complement levels, ANA > 1/640 (AC-1), ANA IgG > 1/320, anti-Sm = 135.4 UI/ml, and anti-RNP = 96.5 UI/ml. Anti-Ro (SS-A) and Anti-La (SS-B) were negative. Systemic lupus erythematosus (SLE) was diagnosed. **Discussion:** Neuropsychiatric SLE can initially be characterized into 19 types of neurological manifestations according to the American College of Rheumatology, though it is important to note that it is not limited to these 19 manifestations, despite them being the major subgroups related to the central and peripheral nervous systems. Among these manifestations, in terms of movement disorders, the most frequently reported is chorea. It is commonly known that there is a scarce volume of scientific publications about neuropsychiatric lupus, although there are references that cite that over the course of the disease (SLE), at least 50% of patients will present some neuropsychiatric manifestation. Among this small group of publications, cases with presentations other than chorea are even more rare, making the description of this case an important contribution to the global literature, to be remembered as a differential diagnosis in similar cases. **Final comments:** Despite being a rare clinical presentation, SLE should be considered in all patients presenting with encephalitis and myoclonus. Early identification and appropriate treatment are essential to improve the prognosis and quality of life for patients.

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Sensory aphasia as sole manifestation of focal status epilepticus

Bruno Barros de Padua Andrade[1], Rodrigo Ferreira Faria[1], Giovanna Santos Rodrigues[1], Luiz Fernando Mohallem Reynaldo[1], Victoria Lot Avezum Mestriner[1], Matheus Spilari[1], Elizardo Nogueira Junior[1], Gloria Maria Almeida Tedrus[1], João Caetano Coutinho Mariuzzo[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Bruno Barros de Padua Andrade; Email: brndepadua@gmail.com.

Case presentation: Male, 70 years old, previously epileptic due to structural cause after ischemic stroke in 2017, forwarded to tertiary hospital after two episodes of tonic clonic seizures, with no description of focal seizures preceding both events. In the past, patient had shown a pattern of impaired awareness non motor seizures with short duration and typical post ictal of confusion and drowsiness. At first assessment, patient has shown repetitive speech, descride as possible paralalia, read at first as possible new aphasia or Todd's paralysis. After realization of Magnetic Resonance Imaging (MRI), it was evidenced hyperintensity in DWI at Wernicke's área, characterizing cortical laminar necrosis. At EEG, it was reflected as frequent epileptiform paroxysms, characterized by sharp waves and spicules on frontotemporal at left cerebral hemisphere (possible dominant). Due to focal aspects of seizure, advanced age and comorbidities, it was chosen to optimize anticonvulsant treatment with later remission of language deficit, achieving full recovery during hospitalization. **Discussion:** Speech or language disturbance is frequently present during seizures, although ictal aphasia is infrequent as the sole symptom of epileptic seizures. De Pasquet et al. first reported a case of ASE in 1976. Magnetic Resonance Imaging (MRI) gives more precise information for detection of anatomical lesions associated with epilepsy and plays a key role in the management of epileptic patients when in association of EEG, that can be helpful to precise the location of the epileptiform focus and pattern. The presence of focal status must be always considerate at patients who have previously diagnosis of epilepsy, but a chance from pattern of seizure, should demands inquisition of other diagnosis, such as neurovascular diseases. **Final comments:** The RMI and EEG play a key role in the diagnosis of the structural abnormalities, but should not delay the begin of treatment in order to prevent prolonged seizure activity as cause of neuronal damage and poor outcomes. Once diagnosed, the treatment must be accurate and guided to etiological definition.

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Sensory motor axonal polyneuropathy in association with granumatosis with polyangiitis and secondary Sjogren's syndrome: a case report

Georgia Piredda Fernandes[1], Raysa Fontes Martins[1], Fernanda Mendes Carneiro[1], Ana Flávia Amitti Alvarez[1], Nicolas de Sousa Belonato Terra[1], Marcio Assis Messias Filho[1]

[1]Hospital São José do Avaí, Itaperuna RJ, Brazil.

Correspondence: Raysa Fontes Martins; Email: 190062471@aluno.unig.edu.br.

Case presentation: A 67-year-old woman presented with intense pain and distal weakness of the lower limbs, associated with edema and arthralgia with difficulty walking for 2 months. He developed disproportionate and areflexic flaccid tetraparesis, weight loss and urinary retention. On examination, there was distal muscular atrophy of the four segments, disproportionate flaccid tetraparesis with muscle strength scores as follows: distal upper limbs -3 and distal lower limbs -2, according to the Medical Research Council Scale (MRC). Deep reflexes abolished (patellar, achilleus, biccipital and triccipital). Normal cranial nerves. He reported immunization against COVID-19 (Astrazeneca) 4 days before the onset of symptoms, with post-vaccination Guillain-Barré syndrome (GBS) as the diagnostic hypothesis. The electroneurographic study revealed a primarily axonal sensorimotor polyneuropathy, affecting nerves in the upper and lower limbs, compatible with the axonal variant of GBS. Human immunoglobulin 0.4g/kg/day was administered for 5 days, there was a slight improvement in muscle strength with muscle strength scores evolving to: distal upper limbs -4 and distal lower limbs -3 according to the Medical Research Council Scale (MRC). After 6 months, it progresses again insidiously with disabling pain, progressive, asymmetric quadriparesis, worse in the left upper limb and left lower limb, in addition to xerostomia, xerophthalmia, livedo reticularis in the lower limbs, scleritis of the right eye and oral ulcers. For investigation, serology was requested, resulting in positive ANCA, positive anti-Ro, positive anti-MPO, in addition to a ground-glass interstitial lung lesion. An evaluation was carried out in conjunction with rheumatology and ophthalmology, and, based on clinical characteristics and laboratory and imaging findings, she was diagnosed with Granulomatosis with Polyangiitis (GP) and Secondary Sjögren's syndrome (SS), with sensory axonal polyneuropathy as the first symptom. engine. The patient continues to be treated with monthly cyclophosphamide, maintaining severe functional disability. **Discussion:** Early diagnosis of SS and GP remains a challenge given the complexity of clinical manifestations, in addition to the need for comprehensive immunobiological tests. In the case in question, it is worth highlighting that the neurological manifestations preceded the typical symptoms of the pathologies in question. This report allows us to discuss the clinical situation and immunological patterns of this condition and emphasizes the importance of comprehensive immunological testing for patients with GBS variants. **Final comments:** The association between GP and SS brings a high level of complexity to the clinical presentation and represents a challenge diagnosis. The specific temporal relationships and clinical presentation of this overlap syndrome can be difficult, and it is important to suspect this correlation given the early appearance of a polyneuropathy.

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Sensory-motor polyneuropathy due to the use of antiparasitic drugs: a case report

Ilana Cwajgenberg[1], Marco Antônio Orsini Neves[2], Daniel Antunes Pereira[2], Wladimir Bocca[3], Acary Bulle de Oliveira[3], Antônio Marcos da Silva Catharino[2]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Iguaçu, Itaperuna RJ, Brazil.
[3]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Ilana Cwajgenberg; Email: ilanacwajg@gmail.com.

The term polyneuropathy refers to a generalized involvement of peripheral nerves, usually involving mainly the distal nerves and, more often, presenting with sensory, motor, and autonomic symptoms and clinical findings. Drug-induced peripheral neuropathy (DIPN) is a persistent condition, most often associated with anticonvulsants, chemotherapy, cardiovascular, psychotropic, and antimicrobial drugs, such as Metronidazole. In this article, we report the case of a patient who developed polyneuropathy secondary to the use of Metronidazole. **Case presentation:** D.D.S.L., a 45-year-old female, previously healthy, presented with abdominal discomfort. Entamoeba histolytica was detected after investigation, and she started a 7-day cycle of 500 mg of Metronidazole three times a day for seven days, without clinical response. A new therapeutic approach was attempted, with three cycles of 2 grams of Secnidazole single dose and Tinidazole for four weeks. However, the patient presented dysesthesia in the distal third of the thighs, followed by allodynia in the four limbs and trunk. After new abdominal discomfort, three months after the first medication cycle, another pharmacological cycle was started. Neurological examination and electroneuromyography (ENM) examination suggested axonal sensorimotor polyneuropathy in all four limbs. **Final comments:** Polyneuropathy can be caused by many factors, including some frequently prescribed drugs such as Metronidazole and other medications in the 5-nitroimidazole group. Therefore, although the relationship between these drugs and polyneuropathy is not fully elucidated, their neurotoxicity is indisputable, even in rare cases, but with significant variability in terms of the dose-dependent potential for this disease.

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Sepsis compromises post-ischemic stroke neurological recovery and is associated with sex differences

Nathalya Camila Cruz Guerra Correa[1], Larissa Joaquim[1], Richard Simon Machado[1], Khiany Mathias[1], Solange Stork[1], Anita Dal Bó Tiscoski[1], Edgar Pereira[1], Natalia Piacentini[1], Tatiana Barichello[1,2], Fabricia Petronilho[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]University of Texas Health Science Center at Houston, Texas, USA.

Correspondence: Nathalya Camila Cruz Guerra Correa; Email: nathalyacorrea0308@gmail.com.

Infection is a complication after stroke and outcomes vary by sex. Thus, we investigated if sepsis affects brain from ischemic stroke and sex involvement. Male and female Wistar rats, were submitted to middle cerebral artery occlusion (MCAO) and after 7 days sepsis to cecal ligation and perforation (CLP). Infarct size, neuroinflammation, oxidative stress, and mitochondrial activity were quantified 24 hours after CLP in the prefrontal cortex and hippocampus. Survival and neurological score were assessed up to 15 days after MCAO or 8 days after CLP (starting at 2 hours after MCAO) and memory at the end. CLP decreased survival, increased neurological impairments in MCAO females. Early, in male sepsis following MCAO led to increased glial activation in the brain structures, and increased TNF-α and IL-1β in the hippocampus. All groups had higher IL-6 in both tissues, but the hippocampus had lower IL-10. CLP potentiated myeloperoxidase (MPO) in the prefrontal cortex of MCAO male and female. In MCAO+CLP, only male increased MPO and nitrite/nitrate in hippocampus. Males in all groups had protein oxidation in the prefrontal cortex, but only MCAO+CLP in the hippocampus. Catalase decreased in the prefrontal cortex and hippocampus of all males and females, and MCAO+CLP only increased this activity in males. Female MCAO+CLP had higher prefrontal cortex complex activity than males. In MCAO+CLP-induced long-term memory impairment only in females. The parameters evaluated for early sepsis after ischemic stroke show a worse outcome for males, while females are affected during long-term follow-up. Keywords: ischemic stroke, sepsis, sex differences, neuroinflammation, oxidative stress.

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Sepsis secondary to ischemic stroke results in sex-dependent potentiation of hypothalamic inflammation in rats

Victor Augusto Rodrigues Sulzbacher[1], Larissa Joaquim[2], Anita Dal Bó Tiscoski[1], Paulo Henrique Amboni Brunelli[1], Natália Piacentini da Silva[1], Victor Duílio Antunes Maragno[1], Naíla Maciel Andrade[1], Khiany Mathias[2], Richard Simon Machado[2], Fabricia Petronilho[1]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.

Correspondence: Victor Augusto Rodrigues Sulzbacher; Email: victor.ars@live.com.

Background: Ischemic stroke is characterized by reduced cerebral blood flow, leading to mitochondrial dysfunction, oxidative stress, and cell death, accompanied by an intense neuroinflammatory response. Inflammation is a key mechanism in the progression of ischemic stroke, affecting responses from important structures such as the hypothalamus. **Objective:** To investigate the relationship between ischemic stroke and sepsis in young rats of both sexes, analyzing the levels of cytokines in the hypothalamus. **Methods:** Male Wistar rats aged 60 days were randomized into four experimental groups, namely: sham+sham, sham+CLP, MCAO+sham, and MCAO+CLP, and subjected to the middle cerebral artery occlusion model (MCAO) for 7 days, then to the cecal ligation and perforation (CLP) sepsis model, suffering painless death after 24 hours, and the hypothalamus was removed. **Results:** All groups showed increased levels of TNF-a and IL-1B compared to sham+sham in both males and females, while only males showed increased levels of IL-6. Only the MCAO+CLP group showed a decrease in IL-10 levels in males, while females did not show significant results. **Conclusion:** We conclude that post-stroke sepsis can intensify the reduction in anti-inflammatory cytokines in the hypothalamus in male rats.

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Sequential optic neuritis secondary to aspergillosis mimicking Foster Kennedy syndrome: a case report

Luiz José Malta Gaia Ferreira Júnior[1], Larissa Braga Mendes[1], Gabriela Rodrigues Tomaz[1], Lucas Toshio Uenishi[1], Daniel Delgado Seneor[1], Lara Santos Machado[1], Maria Eduarda Slhessarenko Fraife Barreto[1], Leonardo Villaverde Buback Ferreira[1], Orlando Graziani Povoas Barsottini[1], José Luiz Pedroso[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Luiz José Malta Gaia Ferreira Júnior; Email: jr_malta@hotmail.com.

Case presentation: A 60-year-old woman underwent neurological evaluation due to a recent onset of decreased visual acuity in her left eye, which progressively worsened over two weeks before stabilizing a week ago. She also reported mild pain upon eye movement. She denied any fever, weight loss, or other systemic or local symptoms. Similar symptoms had occurred in her right eye three months prior. On neurological examination, she presented with reduced visual acuity in both eyes (20/100 bilaterally), pallor of the right optic disc, and edema with hyperemia of the left optic disc. Laboratory investigations, including tests for syphilis, HIV, toxoplasmosis, rheumatological conditions, anti-MOG, and anti-AQP4 antibodies, were unremarkable. MRI of the brain and orbits revealed hyperintensity on T2/STIR sequences in the left optic nerve, with contrast enhancement noted in the orbital segment of the nerve. Additionally, hypointense signals on both T1 and T2 images were observed, correlating with a concurrent CT scan showing a hyperdense lesion with gas pockets. This finding suggested fungal sinusitis, with apparent bone erosion at the lesion site. Furthermore, it was important to rule out compressive lesions that could be causing Foster Kennedy syndrome. Surgical resection confirmed invasive fungal rhinosinusitis upon histopathological examination, with microbiological analysis of tissue cultures indicating growth of Aspergillus fumigatus complex. Cerebrospinal fluid analysis showed no abnormalities. The patient commenced parenteral antifungal therapy, later transitioning to oral itraconazole for a three-month course. She experienced partial improvement in visual acuity, particularly in the left eye, followed by stabilization of her condition. **Discussion:** The subacute onset of visual acuity decline, coupled with pain exacerbated by eye movement, represents a significant clinical feature of optic neuritis (ON). Most cases are associated with inflammatory demyelinating conditions, particularly multiple sclerosis. However, it is crucial to emphasize that ON can also arise from other autoimmune, granulomatous, infectious, and paraneoplastic disorders. Therefore, a thorough etiological investigation is imperative to accurately select appropriate laboratory tests and complement them with imaging evaluations for precise diagnosis and treatment planning. Among the infectious causes of optic neuropathy, fungal infections are infrequent but warrant investigation, especially in diabetic and immunocompromised patients, as well as those displaying signs of sinus pathology. In our case, the presence of sinus pathology observed on facial sinus CT scans played a critical role in achieving a diagnosis. Prompt and appropriate treatment has shown promise in achieving symptomatic improvement. **Final comments:** Although rare, fungal infections should be considered as a possible cause of optic neuritis. If not treated promptly, they can lead to irreversible visual sequelae.

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Serial levels of antiseizure medications and quantitative MRI analysis in mesial temporal lobe epilepsy

Lilian Audi Goulart[1], Yasmim Nadime Jose Frigo[1], Julio Cesar dos Santos Moreira[1], Tarcisio Nunes Alvarenga[1], Maria Beatriz Bravin[1], Lucas Barbosa Napolitano de Moraes[1], Luiz Eduardo Betting[1]

[1]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Lilian Audi Goulart; Email: lilian.audi@unesp.br.

Background: In mesial temporal lobe epilepsy (mTLE), seizure control is often not achieved with antiseizure medications (ASMs). The mechanisms behind refractory epilepsy and the effects of ASMs on the brain structure of patients with mTLE are still under investigation. We hypothesized that ASM concentrations may be related to brain structural changes. **Objective:** To investigate whether areas of structural abnormalities are correlated with serial concentrations of ASMs. **Methods:** Twenty-four patients with mTLE and twenty-four control subjects were enrolled.

All subjects underwent 3T MRI scans, with T1 sequences used for anatomical analysis. ASMs were dosed on the same day as the MRI. For each patient, the relative position of the medication was calculated and standardized according to the concentration range, where a score of 0 corresponded to the lower therapeutic limit and 1 to the upper limit. For patients taking more than one medication, an overall ASM ratio was calculated by summing the individual medication scores. Negative scores were not included. Image processing was conducted using SPM12. High-resolution T1 images were automatically segmented, normalized, and smoothed using the DARTEL algorithm. Modulated gray matter maps were used for the analysis. Statistical analysis was performed using a general linear model (GLM). Comparisons between patients and controls and correlations between gray matter volume and the overall ASM ratio were performed. **Results:** Thirteen patients had left mTLE and eleven had right mTLE, with a mean age of 55±12 years. Patients were using an average of two medications, with carbamazepine (n=14) and clobazam (n=12) being the most frequently used medications. The mean serial dose ratio was 0.9±1.1 (range: -0.3 to 5). Extensive areas of gray matter atrophy were detected (135,393 mm^3, p<0.001 FWE), involving the left precuneus, gyrus rectus, thalamus, insula, superior temporal gyrus, and the right planum temporale, middle occipital gyrus, and posterior cingulate. A weak positive correlation with ASM scores was observed (3,368 mm^3, p<0.01 unc), involving the bilateral calcarine cortex, right thalamus, and hippocampus. **Conclusion:** ASM levels were mainly positively correlated with brain structures, particularly in more preserved, non-atrophic areas. This observation may indicate that preserved neural networks, which include these structures, may be responsible for a better response and tolerability to ASMs. Therefore, preserved areas may be critical for the effectiveness of ASMs in patients with mTLE.

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Serial nerve ultrasound as a predictor of therapy response in a case of chronic inflammatory demyelinating polyradiculoneuropathy

Maria Fernanda de Abreu Rodrigues[1], Liene Duarte Silva[1], Josevânia Fulgêncio de Lima Arruda[1], Rodrigo Tavares Brisson[1], Marcos Raimundo Gomes de Freitas[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Maria Fernanda de Abreu Rodrigues; Email: maferodriguesufrj@gmail.com.

Case presentation: A 60-year-old woman, diagnosed with CIDP had recurrent motor symptoms for the last 35 years and evolved with a progressive limb weakness. The neurological exam revealed tetraparesis with MRC grade 4/5 in proximal limbs and 3/5 in distal limbs, requiring unilateral support for walking. Electrophysiological studies (EMG) showed typical demyelination pattern and cerebrospinal fluid (CSF) analysis revealed high protein levels. In the past years she experienced recurrent disease, and has been receiving monthly Vig to remain asymptomatic. An ultrasonographic examination of her nerves was performed twice with high-resolution ultrasound (HRUS): once soon after an IVIg dose and once close to the next one. We found an enlargement of the ulnar (cross-sectional area [CSA] 50 mm^2) and median (CSA 68 mm^2) nerves in the humerus segment prior to the next dose, correlating with the need to restart treatment. After the IVIg course, the patient reported significant improvement in her motor function and we observed that she no longer needed crutches for walking. HRUS showed CSA of 36 mm^2 and 52 mm^2 in the ulnar and median nerves, respectively. A third measurement taken one month after IVIg, revealed a tendency for the nerves to increase in size again (CSA 48 mm^2 and 52 mm^2 in the ulnar and median nerves). **Discussion:** The diagnosis and management of inflammatory polyneuropathy are often challenging. CIDP, for example, is a potentially incapacitating disease thatrequires appropriate treatment. However, overdiagnosis and inadequate Vig prescription can compromise health systems and cause harmful effects in patients. This case shows that HRUS may contribute to the diagnosis and management of CIDP because, as previous study has already demonstrated, nerve size varies with response to treatment and disease activity over time. **Final comments:** HRUS as an interesting cost-effective tool that may aid neurological practice providing information for diagnosis, therapy decisions and follow-up especially in inflammatory polyneuropathies.

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Seronegative limbic encephalitis presenting as new-onset refractory status epilepticus (NORSE): the case for early initiation of ketamine and ketogenic diet

Caroline Schiochet Verza[1], Bruna Klein[1], Glauco Kody Nagata[1], Giuseppe Dick Bonato[1], Letícia Bassani Devens[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Caroline Schiochet Verza; Email: carolinesverza@gmail.com.

A previously healthy 32-year-old male presented with fever, cough, and headache, initially diagnosed with Influenza A and prescribed Oseltamivir with improvement of symptoms. After five days, he developed confusion, disorientation, and severe anterograde amnesia, followed by a bilateral tonic-clonic seizure prompting ICU admission. Influenza test was negative. Initial investigations including brain MRI, CSF and EEG were unremarkable. Later, he experienced temporal lobe seizures and was started on acyclovir at a tertiary center where a lumbar puncture revealed mildly elevated leukocytes. Despite treatment with phenytoin and levetiracetam, seizures worsened, progressing to focal status epilepticus necessitating mechanical ventilation and continuous EEG monitoring, which showed ongoing right and left temporal lobe seizures. Management included antiseizure drugs along with midazolam and propofol until burst-suppression was achieved. Pulsetherapy with methylprednisolone and IVIG was initiated for suspected autoimmune limbic encephalitis. Systemic screening did not reveal any tumors. Seizures persisted upon medication tapering, characterized by dysautonomia and oral automatisms, which were further treated with topiramate, clobazam, and intravenous ketamine 4mg/kg/h alongside ketogenic diet. After three weeks, seizures ceased, and intravenous medications were tapered off, although hepatic dysfunction necessitated diet and medication adjustments. Negative surface and intracellular antibody tests guided ongoing treatment with Levetiracetam, Lacosamide, Gabapentin, Clobazam, and topiramate. Rituximab was used for autoimmune encephalitis maintenance, resulting in no seizure recurrence or cognitive impairment upon discharge.NORSE (New-Onset Refractory Status Epilepticus) and limbic encephalitis are severe neurological conditions characterized by treatment-resistant seizures. NORSE manifests as prolonged seizures in previously healthy individuals without clear triggers, potentially causing significant brain damage and cognitive impairment. Its etiology involves genetic predisposition, immune dysregulation, and possible infections. Treatment includes high-dose antiepileptic drugs, immunotherapy and therapeutic coma induction when necessary to halt seizures. Diagnosis of autoimmune NORSE involves detecting specific antibodies in blood or cerebrospinal fluid, although seronegative cases are common in limbic encephalitis.Management strategies focus on seizure suppression, inflammation reduction, and addressing underlying immune dysfunction, tailored to individual patient characteristics and underlying causes. Ketamine infusion and implementation of the ketogenic diet are therapeutic approaches used in NORSE to manage refractory status epilepticus and improve seizure control. These interventions aim to modulate neuronal excitability and metabolism, potentially offering relief and enhancing the effectiveness of conventional treatments in challenging cases of NORSE.

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Severe case of cavernous sinus thrombophlebitis due to pseudomonas ecthyma

Adria Rodrigues da Silva[1], Bruna Beppler[1], Caroline Zapelini[1], Lia Maki Hatisuka Imai[1], Luis Sidônio Teixeira da Silva[1], Lúcio Baena de Melo[1], Maria Caroline Martins de Araújo[1], Jéssica Camila Fizinus[1], Manuel Victor Silva Inácio[1], Natasha Caroline Cardoso de Moraes Sanches[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Ádria Rodrigues da Silva; Email: adriarodriguess@hotmail.com.

Case presentation: A 52-year-old female patient, HIV-positive and hypertensive, on Dolutegravir, Darunavir, Ritonavir and Losartan, but with a history of poor adherence to treatment, was admitted to a tertiary hospital due to the sudden appearance two days prior of a blackened crusty papule on the left maxillary and malar region, associated with swelling, pain, fever, and difficulty chewing. Upon examination, there was gingival hypertrophy, palatal ulceration, upper lip edema, and painful cervical lymph nodes. CT scan of the face showed enlargement and infiltration of soft tissues of the left hemiface, mucosal thickening, and diffuse thick secretions in the paranasal sinuses. HIV viral load was 498,000 with a CD4 lymphocyte count of 87. Two days after admission, the patient developed acute respiratory and renal failure, requiring orotracheal intubation and hemodialysis. As the lesions progressed with necrosis, wound debridement was necessary. One month after symptom onset, the patient presented with ptosis, fixed mydriasis, conjunctival chemosis, exophthalmos, edema, and paresis of leftward gaze, indicative of cranial nerves III, IV, and VI palsy on that side. A new CT scan revealed expansive peri-orbital to submental formation in close contact with adjacent bony surfaces, alongside left cavernous sinus enlargement with poor iodinated contrast enhancement, leading to a diagnosis of septic cavernous sinus thrombophlebitis. Biopsy and culture of the lesion indicated Pseudomonas aeruginosa ecthyma, and the patient received polymyxin B, meropenem, amikacin, and moxifloxacin eye drops, remaining in the intensive care unit for 69 days. Gastrointestinal bleeding and facial wounds, along with three debridements, precluded full anticoagulation. Follow-up angiotomography after 46 days showed no evidence of sinus thrombosis. The patient was discharged with symptom improvement, underwent upper lip reconstruction, and continues with rehabilitation and multidisciplinary treatment. **Discussion:** The cavernous sinus is the most frequently infected and thrombosed due to its trabeculae and proximity to the paranasal sinuses. This complication has become rare with the advent of antibiotics, reducing mortality rates from nearly 100% to around 20-30%. Staphylococcus aureus is the primary agent involved, with sinusitis being the most prevalent primary infection. Although facial lesions due to Pseudomonas are reported in some studies, they are less common. Broad-spectrum antimicrobial therapy and surgical intervention, depending on the extent of the lesion, constitute the mainstay of treatment. However, the role of anticoagulation and corticosteroids remains controversial. **Final comments:** This case underscores the importance of early diagnosis and therapy implementation in severe cases associated with Pseudomonas aeruginosa in immunocompromised patients with bleeding and renal failure.

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Severe dysautonomia and chagas disease: a case report

Larissa Scarpini de Carvalho[1], Ana Luiza Meneses de Oliveira[1], Emily Godoi Pereira[1], Helen Maia Tavares de Andrade[1], Matheus Spilari[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Larissa Scarpini de Carvalho; Email: l.scarpinipuc@gmail.com.

Case presentation: A.B.L, a 64-year-old female, reports difficulty standing for the past 6 years, experiencing malaise and episodes of syncope during moderate physical activities and when in an orthostatic position.The condition has progressively worsened, leading to an inability to ambulate, confining her to a wheelchair, and being accompanied by constipation. She has a medical history of Systemic Lupus Erythematosus (SLE) for 26 years, hypothyroidism, and cardiac arrhythmia.The patient was on continuous use of amiodarone. On general physical examination, her seated blood pressure was 80/60 mmHg and heart rate was 50 bpm.Upon standing, she exhibited significant lip cyanosis and pre-syncope; it was necessary for her companion to support her during the orthostatic blood pressure measurement, which was 65/40 mmHg with a heart rate of 64 bpm.There were no abnormalities on neurological physical examination. Initial differential diagnoses considered included Severe Dysautonomia secondary to amiodarone-induced polyneuropathy, hypothyroidism, SLE, or an autoimmune condition. Laboratory tests showed only a positive ANA, without indications of active SLE. Cerebrospinal fluid (CSF) analysis revealed: RBC 145 (90% intact), WBC 1.25, protein 68 (reference < 45), Chloride 124, CSF glucose 48, serum glucose 70, VDRL non-reactive, CSF protein electrophoresis: Total proteins 37.5, albumin 46.6, gamma globulin 14.70% (reference >10-12%), culture negative, bacteria negative. Methylprednisolone 1g/day was administered for 5 days without improvement.Under cardiology guidance, amiodarone was discontinued. Following corticosteroid treatment, serology for Chagas disease returned positive (result: 1/30), leading to the diagnosis of dysautonomia secondary to Chagas Disease (CD) becoming our primary consideration.From the outset, therapeutic management included increased fluid and salt intake, along with the use of compression stockings. **Discussion:** Although far less common than metabolic, inflammatory, and autoimmune disorders, infectious disorders can also affect the autonomic nervous system. Chagas disease is one such infection that can cause diffuse damage to the autonomic nervous system, with proposed mechanisms being primarily denervation due to persistent parasitic infection and immune-mediated injury. Parasympathetic regulation of heart rate becomes impaired, and the tonic inhibitory mechanism exerted by the parasympathetic system fails to respond with tachycardia to rapid transient changes in blood pressure. Our patient, despite severe orthostatic hypotension, does not exhibit compensatory tachycardia. **Final comments:** Dysautonomia is a diagnosis frequently overlooked by healthcare professionals, as evidenced by our patient, who took six years to receive a diagnosis. Determining the etiology can be challenging, necessitating extensive investigation. In Brazil, given the endemic nature of Chagasd, this diagnosis should always be considered.

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Sex differences in clinical and neuropathological correlations: a large population-based autopsy study

Karen Luiza Ramos Socher[1], Alberto Fernando Oliveira Justo[2], Roberta Diehl Rodriguez[2], Renata Elaine Paraizo Leite[2], Wilson Jacob Filho[2], Carlos Alberto Pasqualucci[2], Ricardo Nitrini[1], Lea Tenenholz Grinberg[3], Sonia Maria Dozzi Brucki[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[3]University of California, San Francisco, California, USA.

Correspondence: Karen Luiza Ramos Socher; Email: klrsocher@gmail.com.

Background: i The study of dementia and its differences between the sexes is widely investigated, mainly in Alzheimer's disease. However, most studies on dementia were conducted in high-income countries, mostly in White participants. This study examined sex differences in dementia neuropathology and cognitive abilities in a diverse sample. **Methods:** Participants were classified by sex, and neuropathological lesions were assessed using immunohistochemistry according to international criteria. Cognitive abilities were evaluated using the Clinical Dementia Rating Sum of Boxes (CDR-SOB). We used ordinal logistic and linear regressions to explore the associations of sex with neuropathological and cognitive data adjusted for sociodemographic and clinical variables. **Results:** Of the 1,611 cases included, 801 were men and 810 were women, 65% were white. Women were older, had less education, and presented more Alzheimer's disease neuropathological changes, such as higher scores for Braak stages and CERAD (p> 0.001). No differences were found regarding other neurodegenerative and cerebrovascular lesions. Women presented worse cognitive abilities measured by the CDR-SOB than men in multivariate linear regression models adjusted for age and education. The association of sex, neuropathological assessments, and severity of clinical findings showed a tendency for women to present severe symptoms, regardless of the subtype of the neuropathological lesion. For Alzheimer's disease pathology, there was statistical significance with the clinical pathology and continuous staging with prevalence in women, as well as in the presence of TDP-43 pathology and hyaline arteriosclerosis in vascular pathology. **Conclusion:** Sex in the neuropathology of dementia plays an essential role in cognitive symptoms, highlighting the need to consider it in future prevention and treatment strategies.

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Sex, socioeconomic and genetic risk factors in early Alzheimer's disease type pathology

Karen Luiza Ramos Socher[1], Karen Luiza Ramos Socher[1], Roberta Diehl Rodriguez[2], Claudia Kimie Suemoto[2], Renata Elaine Paraizo Leite[2], Carlos Augusto Gonçalves Pasqualucci[2], Wilson Jacob-filho[2], Lea Tenenholz Grinberg[3], Ricardo Nitrini[2], Sonia Maria Dozzi Brucki[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[3]University of California, San Francisco, California, USA.

Correspondence: Karen Luiza Ramos Socher; Email: klrsocher@gmail.com.

Background: Alzheimer's disease (AD) is the most frequent dementia and predominantly affects women. Studies have described a possible influence of sociodemographic, biological and pathological characteristics underlying sex differences in AD. Among these, cognitive resilience, lifestyle and genetic factors, and longer life expectancy were reported. **Objective:** To analyze the burden of neurofibrillary tangles (NFT) in a large multiethnic population with AD-type pathology and determine sex differences. Additionally, investigate possible risk factors for developing cognitive impairment. **Methods:** 352 cases with clinical, neuropathological and genetic data from the Biobank for Aging Studies of the University of Sao Paulo were analyzed. Demographic, socioeconomic, and clinical data from a postmortem semi-structured interview were revised. The burden of NFT pathology was evaluated in the following areas of immunostained sections using hyperphosphorylated tau antibody (AT8): entorhinal cortex, CA1 of the hippocampus, and locus coeruleus. Additionally, neuropathological evaluation was performed according to the internationally accepted criteria. **Results:** The mean age of death was 72.1±13.4 years, 57.1% were male, the mean of education was 5.3 ± 4.4 years and 65.6% were Caucasians. Around 51% had middle socioeconomic status (SES). Cognitive impairment was observed in 22.7% (CDR>0). The most frequent neuropathological diagnosis was argyrophilic grain disease. Some degree of neurofibrillary pathology was observed in 90.4%, meanwhile, neuritic plaques were observed only in 31.4%. Only 9.7% of 360 individuals met the criteria for neuropathological AD according to NIA-AA, and only 1.6% had a vascular pathological diagnosis. APOE genotype was available only in 155 individuals (43%) and the most frequent genotype was ε3/ε3. Females were older (p<0.001), and had lower mean of education (p=0.003) and higher CDR sum of boxes (p<0.001) compared to males. Additionally, a higher burden of neurofibrillary pathology was found in women compared to men regardless of the presence of cognitive impairment. **Conclusion:** This study provides an opportunity to identify risk factors associated with sex differences that can help to understand the pathophysiological mechanisms underlying AD pathology.

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Sheehan's syndrome: a case report

Lianna da Silva Facco[1], Luidia Giacomini[2], Milena Moraes[1], Matheus Felipe Kuhn Urnau[1], Mauricio Konig Luz[1], Amanda Hedel Koerich[1], Guilherme B. Getelina[1], Júlia Fiorentin[1], Thainá Stolfi[1], Charles André Carazzo[2]

[1]Universidade de Passo Fundo, Passo Fundo RS, Brazil.
[2]Hospital São Vicente de Paulo, Passo Fundo RS, Brazil.

Correspondence: Lianna da Silva Facco; Email: lianna.facco@gmail.com.

Case presentation: Woman, 36 years old, 7th postoperative day of twin cesarean section with a history of bleeding due to placental abruption. She presented weakness, vomiting, insomnia, disorientation, and apathy. Neurology evaluation with a request for a contrasted cranial MRI. The patient developed recurrent episodes of tonic-clonic seizures. Laboratory findings reveal severe hyponatremia (Na 103 mEq/L). A hypertonic saline solution infusion of 250mL was administered. Due to a drop in saturation (80%) and vomiting, sedation and orotracheal intubation (OTI) in the ICU were indicated. Despite the administration of hypertonic saline solution, the patient's hyponatremia worsens, favoring new seizures. MRI shows an enlarged pituitary with a hyperintense lesion, suggestive of pituitary apoplexy, Sheehan's syndrome (SS). After three days of hospitalization, sodium gradually increases (Na 126 mEq/L). Hydrocortisone and levothyroxine are prescribed. Evolution to hemodynamic stability, spontaneous breathing, orientation, and communication. Presentation of diabetes insipidus was treated with desmopressin. After extubation, the patient is maintained on levetiracetam, levothyroxine, and hydrocortisone is replaced by prednisone. A new MRI shows a reduction in pituitary size. Hospital discharge 18 days after admission. **Discussion:** Sheehan's syndrome is characterized by postpartum hypopituitarism secondary to pituitary necrosis caused by shock or hypotension, resulting from massive bleeding during or after childbirth. Considered a rare syndrome, responsible for up to 6% of hypopituitary subjects in developed countries, it does not receive due attention in

textbooks. The pathology is still poorly understood, but it is believed to result from infarction of the pituitary gland, mainly in the anterior lobe, owing to decreased blood flow secondary to vasospasms. In the original clinical description of the disease, Sheehan described the inability to breastfeed, rapid involution of the breast, and menstrual cycle alterations. The symptoms are compatible with different degrees of pituitary hypofunction diagnosed days or years after childbirth. Thus, careful obstetric history is necessary for diagnosis. Management consists of the administration of glucocorticoids, fluids, and, if necessary, thyroid hormone replacement. Importantly, thyroid hormone replacement should not be started before glucocorticoid therapy unless a normal hypothalamic-pituitary-adrenal axis is evidenced in the laboratory to avoid an adrenal crisis. **Final comments:** Although rare, SS requires in-depth investigation for a better understanding of its pathophysiology. It should be considered in patients with obstetric hemorrhage and signs of hypopituitarism. Early diagnosis is essential to prevent complications and improve prognosis.

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Siblings report enforces the relationship between the polynucleotide kinase 3′-phosphatase gene (PNKP) and the clinical manifestation of Charcot-Marie-Tooth disease type 2b2

Letícia Jabur Vaz Ribeiro[1], Sarah Camargos[2]

[1]Universidade Federal de Minas Gerais, Faculdade de Medicina, Belo Horizonte MG, Brazil.
[2]Universidade Federal de Minas Gerais, Hospital das Clínicas, Ambulatório de Neurogenética, Belo Horizonte MG, Brazil.

Correspondence: Letícia Jabur Vaz Ribeiro; Email: leticiajaburvribeiro@gmail.com.

Case presentation: QS, a 19-year-old female patient, was born at term with no complications. Her parents were not related. In early childhood, she presented with global developmental delay. At 13 years old, mild imbalance and incoordination were noted by her mother. During the COVID-19 pandemic, at age of 15, she experienced a subacute worsening of her condition, leading to hospitalization for investigation and rehabilitation. Her older brother had a history of hypotonia in infancy developing impairment of gait and coordination, which also worsened rapidly in 2020. On the neurological examination, the patient presented a slurred and scanning speech, optokinetic nystagmus and absence of tendon reflexes with cold and hypotrophic extremities. Strength was reduced in distal muscles of all limbs. She could walk only with bilateral support. Magnetic resonance imaging showed cerebellar atrophy especially in the vermis. Electroneuromyography demonstrated a primarily demyelinating sensorimotor polyneuropathy. Exome sequencing revealed the presence of PNKP variants: PNKP:c.1123G> T;p.Gly375Trp and PNKP:c.1298+19_1299-37del, in trans. The variant PNKP:p.Gly375Trp variant is classified as likely pathogenic and the PNKP:c.1298+19_1299-3 7del variant is classified as uncertain significance. Segregation analysis demonstrated the presence of PNKP:c.1298+19_1299-37del variant in heterozygosity and absence of the PNKP:c.1123G> T;p.Gly375Trp variant in the mother, whereas both variants were present in the affected brother. The father's DNA was not available. Alpha fetoprotein and total cholesterol was elevated only in the affected brother. **Discussion:** The PNKP gene has three described phenotypes: "CHARCOT-MARIE-TOOTH, AXONAL, TYPE 2B2 (CMT2B2)", "ATAXIA-OCULOMOTOR APRAXIA 4" (AOA4), and "MICROCEPHALY, SEIZURES, AND DEVELOPMENTAL DELAY; MCS". The relationship with the CMT2B2 condition (MIM 605589) is provisional and still requires more data to confirm. Our patients exhibit clinical features consistent with Charcot-Marie-Tooth disease type 2B2 (CMT2B2), an axonal sensorimotor polyneuropathy with ataxia. CMT2B2 has been described in Brazil and Costa Rica. The PNKP:p.Gly375Trp variant is described in several Portuguese families with AOA4. Patients in this study show a phenotypic expansion with primarily demyelinating peripheral neuropathy with an intermittent exacerbation. **Final comments:** The relationship between the Charcot-Marie-Tooth disease type 2B2 phenotype and the PNKP gene is still uncertain. The reported siblings reinforce that this relationship may exist. It is important the report of new cases/families to further strengthen this genotype-phenotype relationship.

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Simultaneous neurocryptococcosis, neurotuberculosis and invasive aspergillosis in a patient with primary immunodeficiency

Thaís Laura Almada da Fonseca[1], Isabelly de Arruda Cardoso Slavec[1], Yasmin Coelho Patrial[1], Enã Rezende Bispo do Nascimento[1], Aline Mizuta Kozoroski Kanashiro[1]

[1]Hospital Regional de Mato Grosso do Sul, Campo Grande MS, Brazil.

Correspondence: Thaís Laura Almada da Fonseca; Email: thaisalmada123@gmail.com.

Neurosyphilis, neurotuberculosis and invasive aspergillosis are infectious diseases that usually occur in patients with severe immunosuppression, generally in the setting of AIDS or other causes of acquired immunodeficiency. **Case presentation:** P.O.S, 33 years old, from a rural area, previously healthy, working in a poultry farm, she started on 07/12/2023 with an acute headache that gradually worsened to a strong intensity, with twinge, frontal and occipital, associated with phono-photophobia, nausea and fever, being treated at the primary hospital with endovenous analgesy and released with symptomatics. On 07/17/2023, she developed mental confusion and was referred to the neurology service due to suspicion of meningitis. Upon admission, she was disoriented and showed meningeal signs. Normal head CT, cerebrospinal fluid (CSF) with opening pressure 500 mmH2O and closing pressure 320 mmH2O, cloudy, with 389 leukocytes/mm3 (lymphomonuclear 90%; polymorphonuclear 10%); glucose 31mg/dL; protein 82.2 mg/dL; direct mycological examination with presence of Cryptococcus sp; VDRL non-reactive. Serum treponemal test for Syphilis was positive, patient previously treated; positive galactomannan in serum; other non-reactive serologies (including HIV and HTLV). Amphotericin B lipid complex and flucytocin were initiated, with partial improvement of symptoms. After 3 days of CSF collection, TRM-TB was positive, and the COXIP regimen was added to the therapy. During the patient's evolution, she remained symptomatic and had increased CSF pressure levels. A new head CT was performed and communicating hydrocephalus was revealed, with a ventriculoperitoneal shunt performed. CD4 (315 mm3) and CD8 (133 mm3) counts were investigated, both decreased. After therapeutic institution, her symptoms improved completely and now she is under investigation for genetic cause of primary immunosuppression. **Discussion:** Primary immunosuppression results from qualitative and/or quantitative changes in the cellular and/or humoral immune response, which may manifest clinically at birth or in adulthood, as described above. These are genetic alterations, most often linked to the X chromosome, however they can be autosomal recessive. This hypothesis should be considered when there are multiple and recurrent infections by different microorganisms (opportunistic or not) in the absence of an acquired cause. The investigation of these conditions involves measuring T and B cells (through immunoglobulins), in addition to determining malformations, searching for cancer or dysfunctions of other systems. **Final comments:** In the case described, the patient had an infection caused by 3 opportunistic germs and did not fit into any secondary cause, therefore it was necessary to investigate the primary cause.

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Sjögren: unique surname, two men, four syndromes and one disease - a narrative review of the literature

Davi Telécio Firmino[1], Tiago César Costa da Silva[1], José Vitor Alécio Rodrigues[1], Fabio Antônio Serra de Lima Júnior[1], Daniel Pereira Maurício de Barros[1], Gustavo Leite Franklin[2], Adriana Meira Tiburtino Nepomuceno[1], Alessandra de Sousa Braz[1], Hélio A. G. Teive[3] , Alex T. Meira[1]

[1]Universidade Federal da Paraíba. João Pessoa PB, Brazil.
[2]Pontifícia Universidade Católica do Paraná, Departamento de Medicina Interna, Serviço de Neurologia, Curitiba PR, Brazil.
[3]Universidade Federal do Paraná, Serviço de Neurologia, Curitiba PR, Brazil.

Correspondence: Davi Telécio Firmino; Email: davitelecio@gmail.com.

Background: The surname Sjögren is rare outside Sweden and Finland. Two physicians with this surname became famous for describing four syndromes and one disease. The Swedish Henrik Samuel Konrad Sjögren from the Karolinska Institute and the Swedish Karl Gustaf Torsten Sjögren graduated in Medicine in Stockholm. **Objective:** To approach the five disorders to which the two physicians contributed, analyzing the historical aspects of each one and their contribution. **Methods:** This is a narrative review based on searching articles in main databases (PubMed, LILACS, Research Gate, ScienceDirect, Web of Science, Embase, Google Scholar, and SciELO) using "Sjogren", "Sjögren, "Sjogren Syndrome", "Henrik Samuel Konrad Sjögren", and "Karl Gustaf Torsten Sjögren" as keywords. **Results:** Henrik Sjögren described the sicca syndrome (Sjögren syndrome) in his doctoral thesis in 1935, describing 19 female cases with dry eyes and dry mouth and proposing treatments. Although it had already been described, it was Henrik Sjögren's contribution in correlating xerostomia and polyarthritis that coined the name of the syndrome. Sjögren reticular dystrophy is a rare dystrophy characterized by macular lesions with a reticular pattern. In 1950, Henrik Sjögren was the first to describe it in a family with 8 children. Spielmeyer-Sjögren disease(Batten disease) is a neuronal ceroid lipofuscinosis, a lysosomal storage disease that affects the retina associated with progressive brain degeneration. The first report was made in 1826, but it was only in 1931 that Torsten Sjögren conducted extensive clinical and genetic studies, demonstrating that these disorders had a monohybrid recessive inheritance pattern, which distinguished them genetically from infantile amaurotic idiocy. Marinesco-Sjögren syndrome is a rare genetic disorder that affects multiple body systems, first reported in 1904. It leads to balance problems, congenital cataracts, muscle weakness, learning and growth difficulties, leading to intellectual disability and short stature. In 1947 and 1949/1950, Torsten Sjögren advanced the understanding of the syndrome phenotype, ensuring that his name was coined in the syndrome. Sjögren-Larsson syndrome is an autosomal recessive neurocutaneous disorder caused by mutations in the ALDH3A2 gene leading to the classic triad: spastic diplegia or tetraplegia, mental retardation, and congenital ichthyosis. Torsten Sjögren was the first to describe the syndrome in 1956. Sjogren and Larsson then described 28 patients in their long monograph. **Conclusion:** Henrik and Torsten Sjögren were not the first to describe most of the disorders that bear their surnames. However, their contributions were crucial in gathering, describing, and analyzing the key elements that make up the conditions, providing the basis for more detailed studies that complemented their findings.

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Sjogren's syndrome associated with facial palsy: a systematic review

João Gabriel Pacetti Capobianco[1], Júlia Gomes[2], Hadassa Carvalho Victorino[2], Mateus Orlando Belisário[2], Beatriz Teixeira Cardoso[2], Giovanna Amorim Panissi

[1]Instituto Capobianco de Neurologia, Poços de Caldas MG, Brazil.
[i2]Pontifícia Universidade Católica de Minas Gerais, Poços de Caldas MG, Brazil.

Correspondence: João Gabriel Pacetti Capobianco; Email: joao20_03@hotmail.com.

Background: Sjögren's syndrome (SS) is considered a chronic, multisystemic condition, which is characterized by dysfunction and abnormalities of the glands and exocrine organs, one of whose characteristics is lymphocytic infiltration, leading these organs to loss of function and triggering a case of xerostomia and xerophthalmia.It affects around 2.7% of the population and the prevalence is 9 women for every man aged approximately 50 years.The association of this syndrome with facial nerve paralysis is a rare condition.**Objective:** To carry out a systematic review of the literature on the association between Sjögren's syndrome and the clinical manifestation of peripheral facial paralysis. **Methods:** A systematic review was developed based on the PRISMA recommendation.The search was carried out in the databases PubMed, Scielo, Scopus, Portal Biblioteca Virtual em Saúde (VHL) and Cochrane Library. The search strategy used was: (Sjogren's Syndrome OR Sicca Syndrome) AND (Facial Paralysis OR Facial Palsy OR Bell´s Palsy) Inclusion criteria: primary studies, published in English, Portuguese and Spanish,published in the last five years, human patients over 18 years of age.Data analysis was carried out using the qualitative metasynthesis technique. **Results:** After analysis and application of the review criteria, 7 studies were included in the final sample. One was published in 2019, three in 2020, two in 2021 and one in 2022. The studies came from Italy (n=1), France (n=1), Saudi Arabia (n=1), United Kingdom (n=1), Germany (n=1), USA (n=1) and China (n=1). Regarding the study design, (n=3) were case reports with literature review, (n=3) were case reports and (n=1) were case series. The analytical themes identified were: involvement of the central and peripheral nervous system, first manifestation of sjogren's syndrome and treatment/prognosis. **Discussion:** Despite the scarcity of data considering the rarity of this presentation, the sample analyzed followed the recommendations for diagnosis and treatment of SS with systemic characteristics. Serological tests was performed based on specific autoantibodies - anti-SSA/antiSSB and exclusion of Guillain-Barrè syndrome. The treatments carried out in the identified studies were based on the treatment of SS in the acute phase, including: intravenous human immunoglobulin, pulse therapy with methylprednisolone, cyclophosphamide and mycophenolate mofetil. The prognosis varied in the sample analyzed, ranging from clinical improvement between two weeks and two months. **Conclusion:** Peripheral facial paralysis associated with Sjogren's Syndrome is an uncommon pathological condition, rarely reported in the scientific literature. There is no specific consensus for the diagnosis and treatment of this condition, and future studies are needed to determine the prognostic relationships of this condition in relation to other systemic manifestations of Sjogren's syndrome.

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Sjögren's syndrome mimicking CIDP: the impact of correct diagnosis on therapeutic choice

Henrique de Moraes Bernal[1], Guilherme Lago Miranda[1], Aldren Falçoni Júnior[1], Pedro Almeida Martins Pontes[1], Ingrid Lorena da Silva Gomes[1], Giovanni Caniçali Simões[1], Tainá Fátima Ramos Gonçalves[1], Mateus Paquesse Pellegrino[1], Herval Ribeiro Soares Neto[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Henrique de Moraes Bernal; Email: henriquembernal@hotmail.com.

Case presentation: A 75-year-old woman with hypothyroidism and dyslipidemia was hospitalized with urinary sepsis due to a right kidney staghorn calculus. During a prolonged hospital stay, it was noticed lower limb weakness, initially attributed to immobility. Despite initial recovery with rehabilitation at home, she experienced a biphasic evolution, with progressive worsening in lower limb strength and loss of fine motor skill in her upper limbs, along with distal hypoesthesia. She sought medical care 4 months after symptoms onset. Her neurological exam revealed significant muscle weakness (proximal and distal in all limbs), areflexia, trunk instability, and predominantly distal sensory impairment. Electrophysiological studies confirmed subacute, symmetrical, sensory-motor demyelinating polyradiculoneuropathy. Systemic workup revealed elevated erythrocyte sedimentation rate (140 mm/h), positive anti-Ro (37.6 UI/mL), and antinuclear antibody (1/640). Due to the positive anti-Ro result, despite no clinical complaints of dry eyes or mouth, she underwent Schirmer's test, which confirmed dry eyes bilaterally. Salivary gland scintigraphy showed global hypofunction. Imaging ruled out neoplasms. She received immunoglobulin and methylprednisolone pulse therapy with poor response, escalated to plasmapheresis with partial recovery. Maintenance therapy with rituximab and prednisone led to significant neurological improvement on follow up. **Discussion:** Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) typically presents with symmetrical sensory-motor symptoms, areflexia, and a course longer than two months. Delaland et al. reported a 24% incidence of retrospective SS diagnosis in CIDP patients, proposing the term "NeuroSjögren" for such cases. First-line treatments for CIDP include immunoglobulin, corticosteroids, and plasmapheresis, with second-line treatments including azathioprine, mycophenolate mofetil, and cyclosporine. For refractory cases, cyclophosphamide and rituximab are options. In this case, first-line treatments provided initial response, and rituximab, an anti-CD20 monoclonal antibody, was chosen based on the consensus of the European League Against Rheumatism (EULAR), which advocates for its use in patients with NeuroSjogren who are refractory to first-line treatment. **Final comments:** The significant improvement observed with rituximab highlights the importance of recognizing NeuroSjögren as a potential diagnosis in patients with a CIDP presentation, particularly when autoimmune markers are present. Systematic screening for SS in such patients can lead to earlier diagnosis and more targeted treatments, improving patient outcomes. The successful use of rituximab in this case aligns with EULAR recommendations and highlights its role in managing severe or refractory SS. The establishment of specific diagnostic criteria for NeuroSjögren is essential in guiding clinical practice and optimizing treatment strategies.

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Skeletal muscle channelopathies: a case series and literature review

Frederico de Sousa Marinho Mendes Filho[1], Luiziane Maria da Silva Alves[1], Paulo Hermes de Lima Amaral[1], Leonardo Rodrigues Leopoldo de Menezes[1], Andreia Lira de Oliveira[1], Maria Esther Lima Rios Evangelista[1], Paulo Roberto Moss Lopes[1], Pablo Vinícius Silveira Feitoza[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Frederico de Sousa Marinho Mendes Filho; Email: frederico.smmf@gmail.com.

Background: Skeletal muscle channelopathies are rare genetic neuromuscular disorders involving sarcolemmal ion channel dysfunction, affecting muscle membrane electrochemical function. Clinically, they present as muscle weakness or myotonia and are linked to mutations in ion channels such as calcium (CACNA1S), sodium (SCN4A), chloride (CLCN1), and potassium (KCNJ2 and KCNJ18), leading to periodic paralysis, non-dystrophic myotonia, and ryanodinopathy (RYR1). In European cohorts, their prevalence is up to 2.38/100,000 in the Netherlands and 1.12/100,000 in England. **Objective:** To describe these diseases' clinical features and conduct a literature review. **Methods:** This retrospective study involves five patients treated at the neuromuscular clinic in the Araújo Lima outpatient clinic, Manaus, Amazonas, Brazil. Data were collected from medical records and analyzed by age, sex, clinical characteristics, diagnostic methods, time from symptom onset to diagnosis, and management. **Results:** Patient A: A 24-year-old female presented at 14 with acute generalized weakness, dysarthria, dysphagia, and cramps lasting 2-3 hours. She had neonatal hypotonia and feeding difficulties. Examination showed deltoid hypertrophy but no myotonia. Genetic testing revealed a heterozygous SCN4A mutation (Chr17:63,957,427 G> 4 variant). Patient B: A 45-year-old male presented at 32 with muscle weakness in the four limbs associated with myalgia and no facial involvement or triggering factors. Episodes recurred every 11-12 months, lasting 3-4 days with spontaneous relief. Physical examination showed micrognathia and short stature. Genetic testing revealed a KCNJ2 mutation (Chr17:70.175.283 C> T variant), consistent with Andersen-Tawil syndrome. Patient C: A 12-year-old male developed weakness in both legs and myalgia after a second COVID-19 vaccine shot, worsening over a week to the point of being unable to walk. Hospitalized and treated with immunoglobulin for 4 days, he improved in motor functions. Genetic testing showed a CLCN1 mutation, indicating congenital myotonia. Patient D: A 4-year-old male experienced trismus and ventilatory dysfunction after anesthesia with sevoflurane and succinylcholine for postectomy, leading to its suspension. Genetic testing identified a RyR1 mutation, indicating susceptibility to malignant hyperthermia. Patient E: A 54-year-old female reported recurrent episodes of generalized weakness lasting 3 hours, with myalgia, cramps, and fatigue. A physical exam revealed calf muscle hypertrophy. Genetic testing showed an SCN4A mutation, indicating hyperkalemic periodic paralysis. **Conclusion:** We reported five cases of rare genetic neuromuscular diseases in the Amazon region, each with distinct clinical features. These conditions are underdiagnosed, highlighting the need to explore the pathologies further.

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Skin wrinkling test in healthy adults: standardization study

Michelle Zonkowski Ribas[1], Renata Dal-Prá Ducci[2], Cláudia Suemi Kamoi Kay[2], Paulo José Lorenzoni[2], Paula Raquel do Vale Pascoal Rodrigues[2], Rosana Herminia Scola[2], Otto Jesus Hernández Fustes[2]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.
[2]Universidade Federal do Paraná, Complexo Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Michelle Zonkowski Ribas; Email: michelle.z.ribas@gmail.com.

Background: Skin wrinkling is a natural phenomenon that occurs on the palms of the hands resulting from vasoconstriction influenced by sympathetic vasomotor function. This phenomenon can be evaluated using the skin wrinkling test (SWT) after water immersion. The absence or reduction of wrinkling can be linked to conditions affecting the dense network of sympathetic nerves in these areas, such as small fiber neuropathy, particularly

in areas where biopsy is unavailable. Despite being quick, inexpensive, and readily accessible, it is not widely used because it lacks clear reference values. **Objective:** To establish SWT's characteristics and normal reference values in healthy individuals. Materials and **Methods:** A cross-sectional study was conducted involving 30 apparently healthy individuals over 18 years old. The participants underwent SWT by immersion in warm water at 40°C for 30 minutes. Wrinkling was assessed by two independent examiners using two grading scales: Clark et al. (1984) and Datema et al. (2012). In the first scale, each finger could be graded from 0 to 4 - (0) absence of wrinkling; (1) very slight, wrinkling that was just present so that the fingertip was not completely smooth; (2) slight, obvious wrinkling with 2 or fewer lines of wrinkling on the fingertip; (3) moderate, obvious wrinkling with 3 or more lines of wrinkling on the fingertip; (4) wrinkling completely distorting the pulp of the fingertip - with a maximum score of 20 for each hand. On this scale, a score of 0 to 2 was considered abnormal. The Datema scale graded wrinkling as follows: 0.0 for no wrinkling, 0.5 for minimal wrinkling, and 1.0 for clear wrinkling, based on comparison to pre-test photos. Absent wrinkling was deemed abnormal, whereas minimal and clear wrinkling was considered normal. The study received approval from the Research Ethics Committee under number: 5.546.162. **Results:** Fifteen men and 15 women, with an average age of 37 years and no history of neurological diseases or diabetes mellitus, participated in the study, with approximately one-third being students (11). The test was conducted on the right hand in all 30 individuals, and additionally on the left hand in 17 of them, with no significant differences in test scores between them. According to the Clark scale, 23 individuals exhibited normal results, while five showed slight wrinkling, and only two showed very light wrinkling. Conversely, using the Datema scale, all individuals displayed some degree of wrinkling, with 2 showing minimal and 28 showing clear wrinkling. **Conclusion:** SWT was normal in 93% of the individuals studied. The data obtained provides a reference point for future research and enables the utilization of this easily accessible and low-cost test in clinical settings. SWT is a clinical tool for assessing the integrity of sympathetic autonomic fibers during neurological examinations.

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Sleep and cognition in individuals with mild cognitive impairment and subjective cognitive decline

Elias Ramos Hurtado[1], Tania Aparecida Marchiori[1], Liara Rizzi[1], Isadora Cristina Ribeiro[1], Ítalo Karmann Aventurato[1], Marjorie Cristina Rocha da Silva[1], Brenda Costa Gonçalves[1], Emanuel Rodrigues Guimarães[1], Marcio Luiz Figueiredo Balthazar[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Elias Ramos Hurtado; Email: eliasrhurtado@outlook.com.

Background: Sleep and circadian rhythm disorders are potential markers of neurodegenerative diseases, especially for Alzheimer's disease (AD). Poor sleep is known to have a significant impact on cognitive function, and frequency and severity of sleep disturbance appear to follow the evolution of cognitive impairment. Few studies have addressed the relations of sleep quality and daytime sleepiness problems with cognitive functions in patients with milder stages of cognitive impairment. **Objective:** Investigate the possible association of sleep quality and daytime sleepiness with cognitive performance in individuals with Mild Cognitive Impairment (MCI) and Subjective Cognitive Decline (SCD), potential stages of Alzheimer's continuum. Materials and **Methods:** This is a quantitative observational cross-sectional study, part of a broader project already approved by the ethics committee. Sixty-five individuals (42 women, mean age 65.8 ±6.4) with clinical diagnosis of MCI and SCD (NIA-AA criteria) were included in this investigation. To evaluate the sleep, we used the scores in quality assessment questionnaires of sleep quality and daytime sleepiness: Pittsburgh Sleep Quality Index (PSQI) and Epworth Sleepiness Scale (ESS), respectively. We also used the score in the following neuropsychological tests: Montreal Cognitive Assessment, Clock Drawing Test, Rey Auditory-Verbal Learning Test, Rey-Osterrieth Complex Figure Test (copy, immediate recall and delayed recall), Trail Making Test (TMT-A and TMT-B), Verbal Fluency Test and Boston Naming Test. For statistical analysis, we used a partial correlation test, controlled for age and education. The significance was considered in p< 0.05. **Results:** We found a negative and weak correlation between PSQI and ROCF copy (r=-0.379; p=0.002), PSQI and ROCF immediate recall (r=-0.271; p=0.031), PSQI and ROCF delayed recall (r=-0.31; p=0.014), and a weak and positive correlation between PSQI and TMT-A (r=0.38; p=0.002), all controlled for age and education. There were no other significant correlations. **Conclusion:** These results suggest a role for sleep in cognitive performance for patients with MCI and SCD. Higher PSQI score, indicating worse sleep quality, was associated with lower ROCF score in copy, immediate recall and delayed recall, representing poor performance in constructive praxis, visuospatial abilities and visual memory. Also, it was associated with higher time in TMT-A, indication of worse sustained attention. We conclude that poor sleep quality is correlated with cognitive dysfunction, and especially executive visual episodic memory and visuospatial functions in our sample. Future research, with a longitudinal design and objective sleep metrics may better investigate this bidirectional association between sleep and AD.

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Sleep disorders in preschoolers, fundamental motor skills, and manual dexterity

Carla de Oliveira Carletti[1], Karla Carlos[1], Gilmar Fernandes do Prado[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, Departamento de Neurologia e Neurocirurgia, São Paulo SP, Brazil.

Correspondence: Karla Carlos; Email: ka.carlos1@hotmail.com.

Background: The performance of many daily tasks in childhood, school activities, and play requires mastery of fundamental motor skills such as locomotion, stabilization, and manipulation, and sleep plays an important role in acquiring these skills. **Objective:** To evaluate the association between sleep disorders, fundamental motor skills, and manual dexterity in preschool children. **Methods:** We assessed fundamental motor skills (long jump, throwing, and kicking a ball) based on fundamental movement patterns at the initial, elementary, and mature stages, and manual dexterity using the Nine Hole Peg Test (9-HPT). Poor performance was defined as children classified at the initial stage in motor skills and with an average time above 30 and 34 seconds for the dominant and non-dominant hands, respectively, on the 9-HPT. Sleep assessment was conducted using the Children's Sleep Habits Questionnaire (CSHQ), consisting of 26 items divided into 6 subscales with independent scores and the following cut-offs: Sleep-Disordered Breathing (SDB) >7, Disorders of Initiating and Maintaining Sleep (DIMS) >22, Night Wakings (NW) > 12, Sleep-Wake Transition Disorders (SWTD) >24, Excessive Daytime Sleepiness (EDS) >20, and Sleep Hyperhidrosis (SH) >8. We compared the results obtained in the group of children with and without sleep disorders. **Results:** We included 43 preschool children with an average age of 5.23 ± 0.42 years, of which 32 were boys (74.41%). The average time with the dominant and non-dominant hands to complete the entire task on the 9-HPT was 27.04s ± (4.06) and 32.68s (±5.49), respectively. For the long jump, 53.4% remained at the initial stage, 37.80% at the elementary stage, and only 9.30% at the mature stage. For overhand throwing: 51.16% at the initial stage, 34.88% at the elementary stage, and 13.95% at the mature stage. For kicking a ball: 18.60% were classified at the initial stage, 60.46% at the elementary stage, and 20.93% at the mature stage. Twelve (27.9%) children (10 boys) scored high on the SDB, SH, and DIMS subscales. There was no association between sleep disorders and fundamental motor skills or manual dexterity assessed by the 9-HPT. Discussion. SDB was one of the most prevalent issues presented in our study, with snoring and obstructive sleep apnea syndrome (OSAS) being common. SDB affects school performance, quality of life, and motor control. However, in our study, there was no association between sleep disorders and fundamental motor skills or manual dexterity. Nevertheless, for our sample of children, the expected stage for motor skills was the elementary stage, and we observed that for the skills of long jump and throwing, more than half of the children showed developmental delays. **Conclusion:** The children evaluated showed sleep impairments, with SDB, DIMS, and SH being the most prevalent. There was no association between sleep disorders, fundamental motor skills, and manual dexterity in preschool children.

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Sleep in multiple sclerosis and neuromyelitis optica spectrum disorder

Andreia Lira de Oliveira[1], Nise Alessandra de Carvalho Sousa[1], João Victor de Chagas Rossin[1], Carlos Maurício Oliveira de Almeida[1], Satiko Andrezza Ferreira Takano[1], Siane Prado Lima de Souza[1], Ronaldo Marques Pontes Rabelo[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Andreia Lira de Oliveira; Email: andreialira.med@gmail.com.

Background: Multiple sclerosis (MS) and Neuromyelitis Optica Spectrum Disorder (NMOSD) are inflammatory demyelinating diseases. They manifest with gait disturbances, spinal cord signs, motor and sensory deficits, cognitive impairment, cranial nerve dysfunction, fatigue, and sleep disturbances. In some studies, sleep and fatigue are considered disabling symptoms, occurring in > 50% of individuals affected by MS. **Objective:** Evaluate sleep disorders such as hypersomnia, fatigue, risk of sleep apnea and presence of restless legs syndrome / Willis-Ekbom disease (RLS/WED) in patients with MS and NMOSD; assess the sleep quality of these patients and correlate these findings with clinical data and imaging. **Methods:** Cross-sectional study conducted in the demyelinating diseases service of the neurology department at HUGV-UFAM, Manaus, Brazil, from January 2017 to December 2020. **Results:** The study consisted of 60 patients, 41 with MS and 19 with NMOSD. They exhibited poor sleep quality (65%) and hypersomnia (53% MS; 47% NMOSD), but low risk of sleep apnea according to the STOP-BANG score. The frequency of restless legs syndrome / Willis-Ekbom disease (RLS/WED) was 14% in MS and 5% in NMOSD. There was no correlation between sleep quality, number of relapses, and Expanded Disability Status Scale (EDSS) scores. **Conclusion:** Patients with MS and NMOSD experience poor sleep quality, hypersomnia, and low risk of obstructive sleep apnea (OSA), but the frequency of restless legs syndrome / Willis-Ekbom disease (RLS/WED) is similar to that of the general population. There does not appear to be a significant difference in sleep disorders between these demyelinating diseases.

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Sleep profile in cerebrotendinous xanthomatosis patients: a pilot study

Pedro Lucas Grangeiro de Sá Barreto Lima[1], Liandra Rayanne de Sousa Barbosa[1], Alissa Elen Formiga Moura[1], Antônio Edvan Camelo-Filho[1], Pedro Helder de Oliveira Junior[1], Mariana Michiles Santos Ramos[1], Livio Leite Barros[1], Pedro Braga-Neto[1], Manoel Alves Sobreira Neto[1], Paulo Ribeiro Nóbrega[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Pedro Lucas Grangeiro de Sá Barreto Lima; Email: pedro.lucas@alu.ufc.br.

Background: Cerebrotendinous Xanthomatosis (CTX) is a rare autosomal recessive lipid-storage disorder caused by variant in CYP27A1. The classical neurological phenotype presents with neuropsychiatric symptoms related to the brain, cerebellum, and/or brainstem with various typical systemic manifestations. There is increasing literature reporting sleep disorders in lipid-storage disorders. However, to the date of this research, there are no published studies on sleep assessment in patients with CTX, nor description of most prevalent sleep patterns reported by these patients. **Objective:** To clinically characterize sleep patterns and sleep disorders in CTX. **Methods:** This was a retrospective observational study including patients with CTX. General clinical data was collected through chart review. Scales used for sleep disorders assessment (Epworth Sleepiness Scale, Morningness-eveningness Questionnaire and screening for restless legs syndrome) and results of wrist actigraphy and polysomnography performed in their medical care were collected to characterize the sleep profile of these patients. **Results:** We included four patients with CTX from our center. All of them had diagnoses confirmed by genetic testing and high plasma cholestanol levels. The patients had clinical history compatible with classical form of CTX. Four patients related sleep disturbances. Two patients related daytime sleepiness and two complained of intense snoring. Insomnia was reported by two patients. Two patients performed polysomnography. Both had reduced latency for sleep, reduced amount of REM sleep and increased amount of N2 and/or N1 sleep for age and gender. One of these patients had evidence of moderate obstructive sleep apnea (OSA). One patient performed actigraphy, showing a circadian rhythm of 24 hours, good sleep consolidation pattern with median sleep efficiency of 91% but a median of five awakenings every night. Sleep patterns or even animal models for sleep disturbances in CTX are still to be described. Hence, there is little literature hypothesizing about how sleep disorders develop in this population. The presence of leukoencephalopathy and cerebellum impairment may be two possible reasons for the presentation of sleep disorders in CTX observed in our patients. The cerebellum is an important region in regulation of sleep, modulating function in the genesis of sleep and regulatory function on body movements during sleep. Leukodystrophies are reported to present with sleep symptoms such as hypersomnolence, insomnia and sleep-related breath disorders loke OSA. **Conclusion:** Our study shows that CTX patients may suffer from impairment in the physiological process of sleep. It highlights that the sleep assessment of these patients should be part of medical practice. Our study had several limitations due to the small sample size. More controlled studies are still needed to evaluate and intervene in the sleep of these patients.

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Sleep quality and mood disorders impact in quality of life of patients with hemifacial spasm of a public tertiary center

João Augusto de Macedo Cavalcanti de Albuquerque[1], Míriam Carvalho Soares[2], Mariana de Almeida Cruz Silva[1], Clélia Maria Ribeiro Franco[2], Pedro Augusto Sampaio Rocha Filho[1]

[1]Universidade Federal de Pernambuco, Centro de Ciências Médicas, Recife PE, Brazil.

[2]Universidade Federal de Pernambuco, Hospital das Clínicas, Serviço de Neurologia, Recife PE, Brazil.

Correspondence: Mariana de Almeida Cruz Silva; Email: mariana.almeida20021@gmail.com.

Background: Hemifacial spasm (HFS) is a condition defined by involuntary facial contractions, usually being unilateral. The literature reports that HFS is more common in women, close to 40-60 years of age. Previous studies indicate that patients with HFS are more prone to insomnia than the general population. Regarding mood disorders, although controversial, a higher frequency of depression has also been demonstrated. In recent years Botulinum Toxin (BoNT) became the most used therapy for HFS, being able to reduce the symptoms with low side effects. Even though HFS is a non-life-threatening condition, patients in this situation may experience social embarrassment, psychological illness and sleep disorders. It is essential to investigate the frequency and impact of sleep and mental disorders in HFS patients' quality of life. **Objective:** Define insomnia, depression and anxiety disorders frequency in an HFS population. Establish the correlation of the impact of insomnia and mood disorders with the quality of life of HFS patients. **Methods:** Cross-sectional observational study involving 56 HFS patients of a public tertiary hospital along 6 month. Patients with cognitive impairment, use of BoNT in the last three months and other movement disorders were excluded. Data were obtained using the following instruments: sociodemographic questionnaire, hospital anxiety and depression scale (HADS), HFS score (subjective part, for quality of life scores), insomnia severity index (ISI) and Pittsburgh sleep quality index (PSQI). The cutoff points used in this study were based on the original studies described for each scale. For HADS, a score ≥ 9 points in the anxiety and depression subscales was used to define mood disorders. For ISI, scores ≥ 14 points characterized clinically significant insomnia. A cutoff of 5 was used for PSQI defined significant sleep disturbances. Statistical analyses were conducted using the Epi Info software. **Results:** Among the 56 patients, 36 (64.2%) were female. The mean age was 66 years (SD 13.1). The average time since diagnosis was 15.4 years (SD 8.6) and the most common spasm side was left, occuring in 33 patients (58.9%). Anxiety was identified in 19 patients (33.9%) and depression in 21 individuals (37.5%). Significant sleep disturbances was identified in 39 individuals (69.6%) and clinically significant insomnia was verified in 9 patients (16%). When compared to quality of life scores (measured by subjective part of HFS score), no statistical significance was verified between anxiety (student's t-test p: 0.102), depression (student's t-test p: 0.396), PSQI (student's t-test p: 0.571) and ISI scores (student's t-test p: 0.603). **Conclusion:** This study showed a considerable frequency of mood and sleep disorders in HFS patientes of a tertiary care center. However, mood and sleep disorders did not demonstrate a significant impact on the quality of life of HFS patients. Further studies are necessary to better analyze these correlations.

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Sleep scales and motor assessment in patients with Parkinson's disease: possible associations?

Carlota Assayag Mendes[1], Diego Monteiro de Carvalho[2], Marcus Vinicius Della Coletta[3], Alvaro Siqueira da Silva[2], Marcello Facundo do Valle Filho[2], Raissa Costa Said[4], Agatha Lopes Azedo[4], Anelisa Campana Itinose[4], Yenly Gonzalez Perez[2], Carlos Mauricio Oliveira de Almeida[2]

[1]Faculdade Metropolitana, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade Federal do Paraná, Curitiba PR, Brazil.
[4]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Agatha Lopes Azedo; Email: agathalopesazedo@gmail.com.

Background: The Unified Parkinson's Disease Rating Scale (UPDRS) is globally recognised and utilised for its capability to capture various aspects of Parkinson's disease, making it a reference standard for disease assessment. The significant and ongoing development of therapies for Parkinson's disease (PD) necessitates outcome measurement strategies focused on disease progression. Hence, this project examined the disease from the perspective of the PDSS, TCSR, Friedman scales, and their comparison with the UPDRS. **Objective:** To analyse the association between the UPDRS and the PDSS (Parkinson's Disease Sleep Scale), TCSR (REM Sleep Behaviour Disorder), and Friedman scales. **Methods:** The study was approved by the local Ethics Committee under CAAE n° 58684622.2.0000.0007. This was an observational, cross-sectional study conducted over 12 months with patients diagnosed with PD. For statistical analysis, Spearman's non-parametric correlation coefficient was used. **Results:** Out of the 33 selected individuals, results indicated that non-motor UPDRS and daily life scores shared the same coefficient of variation and 95% confidence interval range, at 8% and 16.2 - 21.9%, respectively, with mean values of 17.8 and 19.1. The motor scores had a variation of 19.4%, a 95% confidence interval of 39.1 - 53%, and a mean of 46. The overall mean UPDRS score was 89.6 with a variation of 33.2% and an interval between 77.8 - 101.4%. An association analysis was performed between the sum of the UPDRS scores and the final results of the PDSS, TCSR, and Friedman scales. The results in terms of Spearman's correlation coefficient (r) and p-values were as follows: PDSS sum with UPDRS, r=-0.411, p=0.017; TCSR, r=0.76, p=0.674; Friedman, r=0.81, p = 0.654. **Conclusion:** There is no correlation between the sum and final scores of the TCSR (r=0.08, p=0.67) and a significant negative correlation between the sum and final scores of the PDSS (r=-0.41, p=0.02), indicating that as the values of one variable increase, the values of the other tend to decrease (negative correlation). Further studies with larger sample sizes are required to evaluate the associations and specific outcomes of the applied questionnaires.

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Slowly progressive als in a patient with TBK1 variant

Rodrigo Fagundes da Rosa[1], José Marcelino Aragão Fernandes[1], Francisco de Assis Aquino Gondim[1], Kauane Emilly Ribeiro Aureliano[1], Leonardo José Rodrigues de Araújo Melo[1], Francisco Duque de Paiva Giudice Junior[1], Leonardo Elias Araujo dos Santos[1], Mariana Alcântara Tavares[1], Alan Duarte Braz[1], Emanuela Freire Caetano Davi[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Rodrigo Fagundes da Rosa; Email: rodrigofdarosa@gmail.com.

Case presentation: A 67 year-old man began experiencing decreased strength in the lower limbs (LL), more pronounced on the left side, in late 2003. The condition progressed to the upper limbs (UL), right more than the left, and was associated with dysphonia. Neurological examination in 2004 revealed Hoffman and Babinski signs bilaterally, hyperreflexia, and reduced strength in both UL and LL. Also in 2004, he was started on Riluzole 50 mg BID. Brain MRI showed bilateral signal changes along the corticospinal tract, and the first EMG was reported as normal. By 2016, he exhibited spastic tetraparesis, diffuse muscle atrophy, fasciculations, dysphonia, dysphagia, emotional incontinence, and global hyperreflexia. Mild parkinsonism was also evidenced (decreased spontaneous blink and hypomimia). Motor evoked potentials in 2017 reported dysfunction of the corticospinal pathway to both upper and lower limbs. Thereafter, repeated EMG testing disclosed lower motor neuron involvement. He underwent gastrostomy in the second half of 2021 after reporting weight loss. Genetic testing in 2021 showed TBK1

c.1612C> T (p.His538Tyr) mutation in heterozygosity and LRRK2 c.3643G> A (p.Ala1215Thr) in heterozygosity. His motor impairment progressed in 2022, and since 2024 has been walking with difficulty and occasionally needs to use a wheelchair. He receives multidisciplinary care and home care. He has no significant respiratory involvement to date. Currently, he is clinically stable with a slowly progressive condition and preserved cognitive functions. **Discussion:** Recent genetic studies have identified a link between mutations in the TANK-binding kinase 1 (TBK1) gene, and Frontotemporal Dementia and ALS. TBK1, a member of the IKK-kinase family, plays a crucial role in innate immunity signaling and autophagy. Specifically, TBK1 is involved in the induction of type-1 interferons and the phosphorylation of autophagy adaptors, essential for autophagy and mitophagy processes. The impairment of autophagy due to TBK1 mutations may lead to the accumulation of protein aggregates. Our patient, carrying this variant, exhibits a slowly progressive condition that has been stable for over two decades with little clinical variation over the years. **Final comments:** Although we were not able to conduct additional genetic testing in other relatives to segregate the variant and establish causality, this study raises the possibility of the association with this variant and slowly progressive ALS without frontotemporal dementia. In addition to contributing to the relationship between TBK1 mutation and ALS, the case may contribute to a better understanding of progression patterns associated with this variant.

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Slowly progressive amyotrophic lateral sclerosis associated with SOD1 mutation

Pedro Lucas Grangeiro de Sá Barreto Lima[1], Eugênia Machado Couto[1], Pedro Vitor Ferreira Rodrigues[1], Leonardo José Rodrigues de Araújo Melo[1], Francisco Luciano Honório Barreto Cavalcante[1], Ítalo Ramon de Araújo[1], Brendow Martin Freitas Gadelha[1], Richardson Fernandes de Castro e Silva[1], Anderson Rodrigues Brandão de Paiva[1], Paulo Ribeiro Nóbrega[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Pedro Lucas Grangeiro de Sá Barreto Lima; Email: pedro.lucas@alu.ufc.br.

Case presentation: A 56-year-old woman presents with a history of slowly progressive muscular weakness, frequent falls and gait impairment. She was born from consanguineous parents and had 5 healthy siblings. She had normal motor development. By the age of 34, the patient perceived an impairment on ambulation, associated with proximal weakness in the legs. The deficit slowly progressed over the next 2 decades before the first neurological consultation in our center. At 50-year-old she started using one-arm support as walking aid. No dyspnea or dysphagia was perceived. Neurological examination revealed diffuse muscular weakness, atrophy, fasciculations and hyperreflexia. Electromyography (EMG) revealed a pattern of diffuse lower motor neuron impairment. She started use of riluzole by the age of 50 upon the hypothesis of motor neuron disease. Further evaluation with next-generation sequencing exome revealed a likely pathogenic heterozygous variant (p.Val120Leu; c.358G > C) in SOD1, reported to cause familial amyotrophic lateral sclerosis. Thirty-one years after the initial disease presentation, the patient presents independent ambulation with support, no significant dysphagia and a Forced Vital Capacity (FVC) of 56% of predicted values. **Discussion:** Amyotrophic lateral sclerosis (ALS) is the prototypical motor neuron disease in adults. Patients with ALS experience muscular weakness and fasciculations that progress with atrophy, spasticity and loss of functionality. There is no curative therapy and patients tend to have a mean life expectancy of 3-5 years after the diagnosis. Although it is not typical, ALS may manifest predominantly with features of lower motor neuron (LMN). Pathogenic variants in SOD1 gene are well-reported to cause ALS. Variants in this gene produce a wide spectrum of clinical presentations and prognostics. Interestingly, the distribution of SOD1 mutations differs markedly even among similar populations. Treatment with the antisense oligonucleotide drug, Tofersen, is a possibility for ALS-SOD1 patients and has shown to decelerate the decline of clinical function. The variant p.Val120Leu (c.358G > C) in SOD1 is reported to cause ALS with predominantly LMN signs in Brazilian families. Patients reported with this variant tend to have a better prognosis than classical ALS, even without target therapy. Our patient presentation is compatible with the current literature for ALS-SOD1. **Final comments:** We reported a case of a Brazilian woman with ALS due to SOD1 variants that experienced a slow progression of the disease, keeping independent clinical and respiratory function 22 years since the initial presentation. The present report adds to the literature by reinforcing the milder phenotype associated with the presence of this variant in a monoallelic state. Further studies are needed to assess the frequency of ALS due to this variant in northeast Brazil.

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Small vessel disease burden as a predictor for disability, mortality and cognitive impairment after mechanical thrombectomy in an acute stroke: a resilient substudy

Thiago Oscar Goulart[1] , Rui Kléber Do Vale Martins-Filho[2], Millene Rodrigues Camilo[2], Leonardo Carbonera[2], Daniel Giansante Abud[3], Fabrício Oliveira Lima[4], Francisco Mont'Alverne[4], Octávio Marques Pontes-Neto[2], Raul G. Nogueira[5], Sheila Oriques Martins[6]

[1]Harvard School of Public Health, Boston, MA, USA.
[2]Universidade de São Paulo, Ribeirão Preto SP, Brazil.
[3]Universidade de São Paulo, Hospital das Clínicas de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[4]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.
[5]University of Pittsburgh, Pittsburgh PA, USA.
[6]Hospital Moinhos de Vento, Porto Alegre RS, Brazil.

Correspondence: Thiago Oscar Goulart; Email: tgoulart@hsph.harvard.edu.

Background: Although mechanical thrombectomy (MT) has been expanding its indications over the last years, establishing a robust benefit even for the elderly, the RESILIENT trial has shown neutral effects regarding this subgroup. One possible explanation for these findings is the vulnerability of the patients treated under these trials, which means these populations were frailer and, accordingly, portended a significant burden of cerebral small vessel disease (cSVD), which may work as a surrogate marker for brain frailty and even Vascular Cognitive Impairment (VCI). **Objective:** Based on this hypothesis, we studied the influence of the cSVD markers on the baseline CT scan in the functional outcome of the patients randomized in the RESILIENT trial. **Methods:** A trained vascular neurologist blinded to clinical details and randomization allocation evaluated the CT scans of the patients in the study focusing on markers of cSVD (leukoaraiosis, old lacunes, atrophy) and its overall burden. We chose 15 points in MoCA test as the cut-off for severe VCI. Then we carried out a multivariable logistic regression to identify the variables related to disability at 90 days. Finally, the effect of MT was evaluated between subgroups defined by its cSVD imaging markers. We also evaluated the data with an artificial intelligence technique called XGBoost. **Results:** The group with disability at 90 days was older, had a higher NIHSS at admission, had more prevalence of diabetes mellitus (DM), higher percentage of smokers, higher levels of systolic blood pressure and glucose levels at admission, poorer collaterals, lower rate of MT, and higher rates of severe cSVD markers at baseline CT scan. Only NIHSS; randomization for the control group; cSVD score; and glucose levels at arrival were associated with a poor outcome. In the subgroup analyses, the allocation group (standard treatment) influenced the functional outcome just for patients with severe leukoaraiosis and old lacunes. As for VCI, glycemia at admission, systolic blood pressure , NIHSS and cSVD markers were associated with a MoCA test ≤ 15 points. **Conclusion:** Markers of cSVD on baseline CT scans are related to disability, mortality and VCI in patients with LVO and might be one of the main causes of the lack of benefit of MT in older patients observed in the RESILIENT trial.

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Smart syndrome identification and successful treatment

Thais Lins Gemir[1] , Silvana Sobreira Santos[1], Barbara Matos Almeida Queiroz[1], Alberico Carvalho Oliveira Bernardo[1], Denise Carvalho de Albuquerque[1], Natalia Saraiva Ramos e Cruz[1], Mauricea Novaes Costa Pereira[1]

[1]Rede D'Or.

Correspondence: Thaís Lins Gemir; Email: tatalg_30@hotmail.com.

Case presentation: A 62-year-old woman with hypertension, dyslipidaemia, hypothyroidism (secondary to Hashimoto thyroiditis) was admitted in 2024 to the emergency room (ER) with acute onset of global aphasia, lethargy and a left hemicranial headache. The patient had a brain tumor and was successfully treated in 2004 with chemotherapy, local radiotherapy. When the patient first arrived at the ER, she was afebrile with normal blood pressure. During the neurological examination, she was alert but exhibited impaired attention and global aphasia. She had no meningeal signs. Cerebral tomography (CT) revealed ischaemic/radiation leucoenfalopathy with no acute lesions. The cerebrospinal fluid analysis (including cytology) had 33 cels ahd 75 of protein. The following day, the patient underwent an electroencephalogram (EEG) that showed left temporoparietal cortical dysfunction, without evidence of epileptiform activity. A cerebral magnetic resonance was performed, revealing no evidence of stroke or abnormal enhancement on post-contrast sequences. However, an abnormal signal in the left parieto-occipital cortical in the T2 fluid-attenuated inversion recovery (FLAIR) sequence was observed. These revealed that the patient's antithyroid and anti-Peroxidase antibodies were positive. Seven days after admission, the patient started a 5-day regimen of methylprednisolone 1 g EV for the hypothesis of autoimmune disease and the possible diagnosis of SMART syndrome. The patient gradually improved his level of consciousness and recovered from aphasia and gait instability without new seizures or neuropsychiatric symptoms. Follow-up brain magnetic resonance imaging showed resolution of the typical findings **Discussion:** Stroke-like migraine attacks after radiation (SMART) syndrome is a delayed complication of brain irradiation characterised by recurrent neurological symptoms with episodes of complicated migraine symptoms and neurological deficits, including aphasia, hemiparesis and sensory disturbances and seizures. SMART syndrome's pathogenesis is still unknown; the vast majority of cases described in the extant literature suggest a transient syndrome, with full recovery. The clinical case discussed herein, however, is, to the best of our knowledge, unique. No treatment proved effective; nor did the patient's condition improve over time-in fact, quite the opposite occurred. These factors combined to make this case a significant diagnostic and therapeutic challenge. There is no clear consensus regarding effective treatment for SMART.9 Corticosteroids, antiplatelet agents, propranolol and verapamil have all been used without clear benefit. **Final comments:** This case displays a successful clinical evolution of a patient treated for SMART syndrome in which identification of previous radiation treatment, exclusion of other etiologies, and

prompt treatment institution were key for effectively tackling this disease.

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Smith-Lemli-Opitz syndrome in an adult patient: rare diseases are getting older

Marília de Paula Ferreira[1], Marcela Lopes de Almeida[1], Ana Paula Andrade Hamad[2], Guilherme Andrey Ariza Traslaviña[2], Maria Fernanda Ribeiro[1], Kaline Gabrielli da Cruz Benati[1], Dafinny Cela Ferreira[1], Murilo Cela Ferreira[1], Brenda Luiane Rocha de Araújo[1], Débora Marçal Novaes[1]

[1]Estacio Idomed, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, Hospital das Clínicas de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Marília de Paula Ferreira; Email: 19ferreiramarip@gmail.com.

Case presentation: A 21-year-old female patient who had been monitored since birth for various clinical signs characteristic of Smith-Lemli-Opitz Syndrome (SLO), a rare genetic disorder caused by 7-dehydrocholesterol reductase deficiency. Characteristics include retrognathism, microcephaly, protruding teeth and epilepsy of undetermined onset. Complementary tests such as electroencephalogram (EEG) and brain magnetic resonance imaging (MRI) indicated abnormal patterns compatible with diffuse encephalopathy and generalized epileptogenesis. From the beginning of his life, the patient faced health complications, including meconium aspiration syndrome and feeding difficulties. Over time, various malformations and developmental problems were identified, such as ambiguous genitalia, bilateral clinodactyly and syndactyly of the feet. At the age of 3 years and 8 months, SLO was confirmed biochemically and, at the age of 7, a gastrostomy was necessary due to persistent feeding difficulties. Therapeutic treatment included the use of Risperidone, Sodium Valproate (VPA), Clonazepam (CNZ) and a cholesterol-rich diet to control symptoms and improve quality of life. She developed aggressive behavior and absence status epilepticus at the age of 18. **Discussion:** SLO is caused by a deficiency in the enzyme 7-dehydrocholesterol reductase, caused by a recessive pathogenic variant in the DHCR7 gene, which is essential for cholesterol synthesis, resulting in a wide range of clinical manifestations such as congenital malformations and developmental delay. The patient had typical features of SLO, such as retrognathia, microcephaly and protruding teeth, along with epilepsy, which can be attributed to neurochemical dysfunctions due to cholesterol deficiency. Treatment included a combination of antiepileptic drugs (in this case VPA, CNZ and Lamotrigine) and antipsychotics (in this case Olanzapine) for symptom control. EEG and MRI brain scans revealed abnormal patterns, corroborating the neurological symptoms and helping to guide treatment. Although medical control is crucial, there is no definitive cure for SLO. The therapeutic approach's main objective is to attenuate the symptoms and optimize the patient's functionality. This case illustrates the complexity of this syndrome and the importance of its recognition and subsequent follow-up throughout adulthood. **Final comments:** Smith-Lemli-Opitz Syndrome (SLO) is a rare genetic disorder characterized by multiple anomalies and cognitive impairment due to 7- dehydrocholesterol reductase deficiency. Its management requires the collaboration of various specialties, especially Neurology, in order to control symptoms and improve the patient's quality of life.

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Snake eye myelopathy: a misdiagnosis entity

Yasmin Ramalho Mandarino[1], Natália Cardoso Darabas[2], Hannah Bang[2], Pietra Cani Linzmeier[2], Thábata Emanuelle Martins Nunes[2], Maria Eduarda Angelo Mendonça Fileti[2], Carla Heloísa Cabral Moro[2], Alexandre Luiz Longo[2]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Yasmin Ramalho Mandarino; Email: yasminramalhoo@gmail.com.

Case presentation: A 54-year-old male with history of neck trauma due to a motorcycle accident 10 years ago presented to the neurology service with complaints of strength loss in the upper limbs, worse in the left side, accompanied by myofasciculations. These symptoms started two years ago and have progressed with paresis in the lower limbs in recent months. Patient denies sphincter changes or bulbar symptoms. Physical examination revealed muscle atrophy, fasciculation in the upper limbs, grade IV strength and areflexia in left upper limb and hyporeflexia in the left lower limb. No signs of upper motor neuron involvement were found. Electroneuromyography showed chronic and asymmetric denervation with multisegmental innervation in the cervical region, most pronounced at C5-C7. An cervical spine MRI was crucial for the diagnosis, with hypersignal in the anterior horns of the C2-C5 gray matter, indicative of probable sequelae. The patient has been followed for 11 years without worsening of the condition. **Discussion:** "Snake eyes" myelopathy represents a rare form of myelopathy. Snake-eyes appearance (SEA) is a unique radiological finding characterized as bilateral hyperintense symmetric, circular or ovoid foci on T2-weighted axial magnetic resonance imaging sequences in the anterior horn cells of the spinal cord, and it's called like this because of the similarity to the face of a snake. There is no clear data on the exact prevalence and incidence, as there has been no systematic analysis of the full spectrum of clinical patterns associated with snake-eyes in larger cohorts. It's considered as an anterior horn disorder resulting from local ischemia, triggered by arterial compression from an anterior with shifting of the posterior cervical dura upon neck flexion. Trigger factors such as intensive physical exercise, trauma or carrying heavy loads are common. This "snake-eyes" appearance is not specific to any particular disease and can be seen in a variety of conditions such as spinal artery ischemia, chronic compressive myelopathy, degenerative cervical myelopathy, Hirayama disease or monomelic amyotrophy of the upper limb, amyotrophic lateral sclerosis, and spinal muscular atrophy. Therefore, clinical examination, extensive laboratory tests, nerve conduction studies, and genetic tests may be required to diagnose the underlying condition. **Final comments:** This case underscores the importance of recognizing the "snake-eyes" myelopathy as it is a disease that affects the lower motor neuron that can have a benign prognosis. This serves as a crucial differential diagnosis for amyotrophic lateral sclerosis. The "snake-eyes signal" is an often-under-recognized radiological clue, leading to frequent misdiagnosis of patients with this clinical-radiological entity. Additionally, the case highlights the significance of performing neuroimaging in the diagnostic evaluation of patients with motor neuron disease.

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Sneddon syndrome: a rare cause of stroke

Nathalye Fernanda Pedroso Dircksen[1], Marcelo Simplicio Carvalho[1], Letícia de Barros Dias[1], Lorena Fernandes Kronbauer[1], Fábio Gonçalves[1], Felipe Leonardo Semensati[1], Damacio Ramon Kaimen Maciel[1]

[1]Irmandade da Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Nathalye Fernanda Pedroso Dircksen; Email: n.athalye@hotmail.com.

Case presentation: A 41-year-old woman with mitral insufficiency treated with valve replacement 20 years ago, was admitted due to recurrent ischemic strokes. She presented paresthesia of the left upper limb and vertigo, She was confused (Glasgow scale 14), mini mental state examination loss of attention, memory and language (score of 25) and livedo reticularis was observed in the limbs and trunk. Magnetic resonance imaging (MRI) of the skull showed multiple hyperintense lesions affecting the frontal, occipital and right parietal lobes, suggesting a subacute/chronic vascular insult, in addition to diffuse volumetric reduction of the brain that was not expected for the age. Skull arterial angiography showed no changes. The echocardiogram transthoracic examination showed significant enlargement of the left atrium and mild mitral reflux - metallic valve. The VHS was 38, with no other changes in rheumatological or hematological screening. Started using warfarin with INR (International Normalized Ratio) target between 2.5-3.5 due to valve prosthesis, antihypertensives and statins. **Discussion:** Sneddon Syndrome (SS) is a rare and slowly progressive neurocutaneous disease that affects small and medium-sized arteries and is characterized by generalized livedo reticularis and recurrent cerebrovascular events. Approximately 80% of patients are women with a mean age at diagnosis of 40 years, as in the case presented. The etiology is unknown, some cases have been associated with an autosomal dominant inherited pattern. Arterio-occlusive pathogenesis is due to 2 proposed mechanisms: autoimmune/inflammatory disease and thrombotic disease. There is no specific biological marker for the diagnosis of SS, however it can be classified as antiphospholipid (APL) positive or negative, depending on the presence of any of these 3 antibodies: anticardiolipin, lupus anticoagulant and anti-beta 2-glycoprotein. Less than half are APL positive, with a higher risk of thrombocytopenia, seizures and chorea. Neurological manifestations generally occur in 3 phases. The first with prodromal symptoms (headache, dizziness and vertigo), the second with ischemic and rarely hemorrhagic strokes (rupture of collateral vessels secondary to occlusion of primary cerebral arteries) and the third with cognitive decline and early-onset dementia. The most common cardiac pathology in SS is thickening of the heart valves and/or libman-sacks endocarditis. Treatment requires control of modifiable risk factors and use of antiplatelet agents or antithrombotics (high-dose warfarin with INR > 3, preferable in APL positive patients). **Final comments:** The absence of specific exams and consensus makes the diagnosis and treatment of the disease more challenging. Currently, SS is a diagnosis of exclusion and is often forgotten. Therefore, attention should be paid to its possibility, as failure to adequately treat it can increase the morbidity and mortality of its sufferers.

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Social and occupational functioning in adult patients with epilepsy

Gloria M. A. S. Tedrus[1], Danilo Wingeter Ramalho[1]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.

Correspondence: Gloria M. A. S. Tedrus; Email: gmtedrus@uol.com.br.

Background: Adult patients with epilepsy (PWE) often face greater difficulties in employment and social activities, tending to have lower educational levels. **Objective:** To assess social and occupational functioning in PWE and examine its correlation with clinical aspects. **Methods:** Scores on the Social and Occupational Functioning Scale for Epilepsy (SOFSE) were correlated with demographic and clinical data from 65 PWE, with a significance level of $p < 0.05$. **Results:** The PWE had a mean age of 47.2 years, while their mean age at first seizure was 22.1 years, with an average epilepsy duration of 25.3 years. 40 of them were males and 33 females, 53 (72.6%) patients were employed/retired, and 20 patients were unemployed.

The total score on the SOFSE scale was 66.3±15.6. In the comparison between employed and unemployed individuals, higher scores were observed in the "employed with a salary" facet (T-test, 8.0 ± 6.7 vs 3.7 ± 5.9; p=0.011) and "instrumental activities of daily living" (19.6 ± 7.1 vs 22.8 ± 4.8, p 0.038) in employed individuals. Women and unmarried patients, as well as the occurrence of focal seizures, use of a single antiseizure medication, and epilepsy type were associated with higher scores on the SOFSE scale (facets and/or total score). The total SOFSE scores correlated with age (Pearson correlation, -0.384, p=0.001), education (0.336, p=0.004), and age at the first seizure (-0.231, p=0.049). **Conclusion:** Demographic and clinical factors are associated with impairment in functional and occupational status in epilepsy. Younger patients with higher educational levels and later onset of epilepsy present better occupational and functional adjustment.

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Sociodemographic and clinical factors affecting migraine treatment outcomes in the emergency department

Carla Bianca Alves Leite Santos[1], Artur de Oliveira Macena Lôbo[1], Claudia Cristina de Lira Santana[1], Bruno Felipe Novaes de Souza[1], Juliana Ramos de Andrade[1], Marcelo Moraes Valença[1]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil.

Correspondence: Carla Bianca Alves Leite Santos; Email: carla.biancaleite@ufpe.br.

Background: Migraine is the most prevalent neurological disease, currently considered the second leading cause of global disability, the first when it comes to women aged 50 or younger. **Objective:** To analyze the efficiency of the emergency service in treating patients with primary headache, based on the effectiveness of the treatment, taking into account the level of pain reported before and after the medical assistance, and the return rate. All correlating with the sociodemographic, economic, and clinical profile of the patients. **Methods:** An observational, cross-sectional study was designed in the emergency room of a tertiary hospital. The data was collected through interviews and surveys of all International Classification of Diseases (ICD) using the patient's electronic record. The study population comprises adult patients with ICDs for headache, migraine, and their variables, approached before and after care. For evaluation of pain, a Visual Analogue Scale was used (in which 0 indicated absence of pain and 10 represented unbearable pain). The exclusion criteria were: a) patients with a change in diagnosis during their stay in the hospital; b) patients who evaded the service before medical discharge; c) patients who progressed to hospitalization. **Results:** We evaluated a total of 102 patients, with a mean age of 36.9±12.7 years, of whom 83 (81.4%) were female. Concerning the patients' referred pain level when they arrived at the service, we found out that women reported significantly higher initial pain levels (7.48±1.95) compared to men (6.22±2.39) (p=0.009) and that the same applied to patients already under medical follow-up for migraine (8.46±1.45) compared to those without follow-up (7.08±2.11) (p=0.014). Regarding the patient's referred pain level after treatment in the service, we observed that those who had a higher frequency of migraine-related visits to the emergency service in the last year also reported higher levels of pain after treatment (p=0.017). Furthermore, it was observed that patients already under medical follow-up and those with higher purchasing power had a higher frequency of visits to the emergency service due to migraines in the last year (p<0.001 and p=0.028, in this order). No significant association was found between pain in any stage of the medical assistance or frequency of migraine-related medical appointments and the patients' workload, educational level, and use of opioids before coming to the emergency service (all p> 0,05). **Conclusion:** This study highlights potential factors that affect migraine treatment outcomes in the emergency room. While women and those under medical follow-up for migraine reported higher initial pain, those with frequent emergency room visits had higher post-treatment pain levels. These findings suggest the need for a more individualized approach to migraine care in the emergency department, potentially considering alternative treatment pathways for specific patient profiles.

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Socioeconomic burden of multiple sclerosis in Brazil: an overview of the last 10 years

João Lucas Lima Diogo[1], Ana Clara Costa de Siqueira Campos[1], Gabriela Morais de Medeiros Dias Melo[1], Joanna Sousa da Fonsêca Santana[1], Lara Millena Miranda Frota[1], Luísa Lettiery Andrade Santos[1], Maria Antônia Vianna Santos[1], Maria Eduarda Kobayashi Teixeira[1], Ruben Costa Santos Neto[1], Felipe Oliveira Costa[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: João Lucas Lima Diogo; Email: joaodiogo20.2@bahiana.edu.br.

Background: Multiple sclerosis (MS) is an autoimmune and chronic neurological disease, resulting from the demyelination of neurons in the white and gray matter of the central nervous system, with a prevalence of 8.69 cases per 100,000 inhabitants in Brazil. This condition is the second most prevalent cause of permanent neurological disability before the age of 50 in the country. Therefore, the scarcity of socioeconomic analyzes on this disease highlights the importance of this study for health policies and resource allocation, ensuring an equitable treatment for all patients. **Objective:** To analyze the socioeconomic burden and demographic distribution of MS in-hospital environment in the Brazilian Unified Health System (SUS). **Methods:** Descriptive ecological study involving the number of hospitalizations for MS in Brazil, between the years 2014 and 2023. All hospitalizations were listed based on access to data from the SUS Hospital Information System (SIH/SUS) using the TABNET tabulator. **Results:** When analyzing data for costs of hospitalizations due to MS in the SUS from 2014 to 2023, we found that the total national amount spent in this period was R$24.797.196,40. The region with the highest average cost of hospitalization was the North Region (R$1.448,50). The region with the highest annual growth in investments was the Central-West Region (18%) from 2015 to 2016. The Central-West Region had the greatest reduction in absolute and relative values, with a decrease of R$214.605,81 (74,62%) between 2018 and 2019. Finally, the highest annual average cost of hospitalization was observed in the Southeast Region (R$1.530.645,38). Regarding the number of hospitalizations, the region with the highest number of hospitalizations during those years was the Southeast Region (31.006 hospitalizations) and the region with the lowest was the North Region (730 hospitalizations). The Southeast Region had an increase of 1.340 hospitalizations from 2021 to 2022 and the Central-West Region showed a reduction of 748 hospitalizations from 2018 to 2019. **Conclusion:** The study reveals a significant regional divergence between costs of hospitalization for MS. The Southeast Region stands out both in terms of expenditure and the number of hospitalizations, denoting a possible greater access to specialized health services. The North Region, despite having the lowest total number of hospitalizations, had the highest average value per hospitalization, suggesting that the cases treated may have greater complexity or higher individual costs. The results suggest the need for a more equitable distribution of health resources and strategic planning to improve access and quality of MS treatment. The limitations of this study include its dependence on SIH data and the lack of data on the quality of care provided. Future studies shall investigate explanations for resource allocation and the inclusion of qualitative data on the impact of treatments.

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Somatosensory evoked potentials in neuromyelitis optica spectrum disorders

Otto Jesus Hernandez Fustes[1], Cláudia Suemi Kamoi Kay[1], Paulo José Lorenzoni[1], Renata Dal-Prá Ducci[1], Paula Raquel do Vale Pascoal Rodrigues[1], Rosana Herminia Scola[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Otto Jesus Hernandez Fustes; Email: otto.fustes@hc.ufpr.br.

Background: Neuromyelitis optica spectrum disorders (NMOSD) are antibody-mediated disorders of the central nervous system with a predilection for the spinal cord and optic nerves. NMOSD was previously thought to be a variant of multiple sclerosis (MS), but the identification of aquaporin 4–immunoglobulin G (AQP4-IgG), has led to an appreciation of these diseases as distinct entities. The clinical utility of evoked potential recordings (EPs) has already been established for MS, in which EPs can detect clinically silent lesions in visual, auditory, sensory, and motor pathways. In addition, several studies have suggested a prognostic value for EPs in MS and an association between EP abnormality and the level of clinical disability. On the other hand, there have been few reports on EPs in patients with NMO. At the NMO, recent studies are beginning to reveal the role of EPs and their possible value as a diagnostic tool. **Objective:** Determine the characteristics of somatosensory evoked potentials (SSEP) in patients with NMOSD. **Methods:** The SSEP in the upper and lower extremities was prospectively performed in patients with NMOSD according to the International Panel for NMO Diagnosis criteria, recruited from the outpatient clinic of a university neurology center. Recorded as recommended by the IFCN, using the Nihon-Kohden Neuropack software. The latency was measured at the peak amplitude, and the amplitude was measured between the peak and the end of the potential. The interpretation was based on our laboratory standards using the same technique and apparatus. For each latency variable (N20, P40), patients were categorized as "Normal" or "Abnormal". Then the variables of age, gender, AQP4-Ab, and latencies were compared between the two groups (Abnormal vs. Normal). The study was approved by the Local Ethics Committee for Human Research with a number 4.340.521. All studies were conducted following ethical principles after obtaining patient informed consent. **Results:** We evaluated 12 females with a mean age of 42 years old and one man of 25 years old. When categorized concerning the variable SSEP (Normal vs. Abnormal), there was a statistical difference in N20, N21, and P40 latencies. 76.9% of our patients had AQP4-IgG positive. No significant differences in age at onset were observed between anti-AQP4 antibody-positive and --negative patients. Normal patients had an average N20 latency of their limbs of 18.69 ± 1.6 and Abnormal patients had an average of 25.21±10.09 (p=0.016 two-tailed hypothesis). **Conclusion:** SSEP showed a significant increase in N20 and P40 latencies. SSEP is a non-invasive, painless, fast, and low-cost exam that provides neurophysiological data for diagnosis of NMOSD.

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Spastic paraplegia type 3: case report

Laura Altomare Fonseca Campos[1], Jéssica Cristina Silveira Damasceno[1], Marcelo Sobrinho Mendonça[1], Thiago Cardoso Vale[1]

[1]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.

Correspondence: Laura Altomare Fonseca Campos; Email: lauraaltomare8@gmail.com.

Case presentation: A 66-year-old hypertensive male patient is evaluated with a history of childhood onset of slowly progressive gait impairment. Family history included a father, a paternal uncle and a brother with a similar clinical condition. On physical examination, there was a slight reduction in the distal strength of the lower limbs, associated with significant spasticity, worse in the right lower limb, with scissoring gait. Imaging and laboratory tests were unrevealing, what led us to request a genetic panel, that revealed a mutation in the ATL1 gene. Spastic paraplegia Type 3 (OMIM 182600, variant c.1483 C > T p.(Arg 495Trp) was diagnosed. **Discussion:** Type 3 spastic paraplegia is part of the Hereditary Spastic Paraplegias (HSP) group and is, therefore, a disease with a genetic inheritance pattern, which can be autosomal (recessive or dominant) or X-linked. Despite the genetically diverse forms, it is known that the disease evolves with progressive degeneration of the axons of the corticospinal tract, which typically manifests with spasticity and weakness predominantly in the lower limbs, in addition to other findings that may be systemic or neurological. The phenotype of HSP varies according to the gene involved and can present with the typical clinical features mentioned above, as well as other neurological alterations such as seizures, dementia, extrapyramidal disorders, peripheral neuropathy, amyotrophy, ataxia and hydrocephalus. Some forms may also present with cognitive alterations, which can be observed from the onset of symptoms. Type 3 spastic paraplegia occurs in around 4 to 6% of autosomal dominant HSP, and is usually a pure form, without other neurological involvement, and with onset in childhood, as in the patient reported. **Final comments:** Correct diagnosis is important because the differentials include motor neuron diseases, structural impairments of the spinal cord, leukodystrophies, dystonia, metabolic or infectious disorders, and hereditary ataxias. Although there is no disease-modifying treatment, it is possible to carry out genetic counseling and to provide symptoms relieve through medication, physiotherapy, rehabilitation, and botulinum toxin.

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Spatial distribution of incidence and mortality by malignant tumors of the central nervous system in the Southern region of Brazil

Pamella Fracisquini Gardin[1], Rodrigo Penomian Cianca[1], Yves Henrique Ramos Mansano[1], João Marcelo de Souza Baptista[1], Matheus Henrique Arruda Beltrame[1], Miyoko Massago[1], Luciano de Andrade[1]

[1]Universidade Estadual de Maringá, Maringá PR, Brazil.

Correspondence: Pamella Francisquini Gardin; Email: pamfgardin@gmail.com.

Background: Malignant neoplasms of the central nervous system (CNS) have increased in recent years around the world, including Brazil, where this group ranks 11th among the most frequent neoplasms, showing higher incidence and mortality in the Southern region of the country. **Objective:** To analyze geographical disparities in incidence and mortality by CNS neoplasms in the Southern region of Brazil, between the years of 2017 and 2021 **Methods:** This is a cross-sectional ecological study using secondary data obtained from the Oncology Panel, DATASUS Mortality Information System and Brazilian Institute of Geography and Statistics. Data on incidences and deaths from malignant CNS tumor (C70, C71, C72), in individuals aged 20 to 64 years, were filtered. Crude incidence and mortality rates were calculated by the ratio between the number of cases and the resident population adjusted by age. These rates were smoothed by the empirical Bayesian estimator method, being named smoothed incidence and mortality rates. Subsequently, the Global Moran's Index and the Local Indicator of Spatial Association (LISA) were applied to detect spatial autocorrelation at global and local levels of incidence and mortality rates. **Results:** During the study period, there were 4,666 cases and 3,042 deaths by malignant CNS tumor, with 1,922 cases in Paraná, 1,107 in Santa Catarina and 1,637 in Rio Grande do Sul, and a mortality of 100, 998, and 1,944 deaths in the respective states. Analyzing only incidence rates yielded a Global Moran's Index of 0.546 ($p<0.001$). There were 151 municipalities identified with high-high clusters (incidence rate from 5.3 to 17.6 cases per 100,000 inhabitants), that is, municipalities with high incidence rates surrounded by municipalities with high rates, in the northeast, southwest, central and southeast regions of Paraná, west-central Santa Catarina and north Rio Grande do Sul. Regarding mortality, a Global Moran's Index of 0.593 ($p<0.001$) was obtained. Mortality rates varied from 0 to 16.6 deaths per 100,000 inhabitants, with 150 municipalities showing high-high clusters predominantly distributed in the states of Santa Catarina and Rio Grande do Sul, especially on their border, as well as clusters in the extreme south of Rio Grande do Sul, western Santa Catarina and scattered municipalities in Paraná. **Conclusion:** The highest mortality rates were concentrated in Rio Grande do Sul and Santa Catarina, and although the state of Paraná showed high incidence rates of malignant CNS tumors, it corresponds to the lowest mortality rates when compared to the others. Such divergence may be due to variables such as accessibility and quality of reference services. However, more in-depth studies are necessary to elucidate this hypothesis.

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Spectrum of ataxic neuropathy in a young woman: case study of POLG gene mutation

Iago César Lopes Tenório de Cerqueira[1], Caio Roberto Alexandre Cavalcante dos Santos[1], Isaías Henrique Martins de Vasconcelos Norte Bezerra[1], Maria Eduarda Vieira de Oliveira[1], Déborah Emmily de Carvalho[1], Lívia Barbosa Cavalcanti[1], Leticia Gomes Fonseca[1]

[1]Universidade de Pernambuco, Recife PE, Brazil.

Correspondence: Iago César Lopes Tenório de Cerqueira; Email: iagocesartenorio@gmail.com.

Case presentation: A 29-year-old woman with no significant prior clinical history developed gait ataxia 16 years ago, beginning with frequent falls and ankle sprains, progressing to independent walking difficulty and clumsy speech. Examination revealed cerebellar ataxia with Romberg sign, wide-based gait, global areflexia, mild proximal tetraparesis, auditory hypersensitivity, spastic dysarthria, and limited upward gaze. General laboratory tests were unremarkable. ENG indicated purely sensory distal peripheral axonal polyneuropathy in the lower limbs. Brain MRI showed cerebellar atrophy and T2 hyperintense lesions in bilateral cerebellar deep white matter, thalami, and inferior olivary nucleus. Genetic analysis confirmed a homozygous pathogenic variant, c.2243G> C, diagnosing a POLG-related disorder with SANDO phenotype. **Discussion:** The Ataxia Neuropathy Spectrum (ANS) is a rare genetic condition, one of six major syndromes associated with POLG gene mutations, affecting a small fraction of the population. These mutations are the most prevalent cause of mitochondrial disease across all ages, accounting for 10% of adult cases. The POLG gene encodes the catalytic subunit of DNA polymerase gamma, essential for mitochondrial DNA replication and repair. This mitochondrial disease primarily affects the nervous system, causing neurological and systemic manifestations, such as auditory neuropathy indicated by auditory hypersensitivity and unilateral hearing loss. Additionally, this dysfunction may present with chronic headaches and osteopenia due to altered bone metabolism. The patient exhibited progressive cerebellar ataxia, dysarthria, and sensory ataxic polyneuropathy with ophthalmoplegia, common characteristics of POLG mutations, with the latter being significant. MRI findings, such as cerebellar atrophy and T2/FLAIR hyperintensities in white matter and deep brain structures, are typical of mitochondrial disorders, supporting the ANS diagnosis. Electroneuromyography corroborates the patient's peripheral neuropathy, also associated with ANS. **Final comments:** The findings described corroborate the complexity of the syndromes caused by mutations in the POLG gene. Due to the rarity of the condition highlighted in this case, it tends to be underdiagnosed, resulting in a poor quality of life for affected patients. To enable early diagnosis and effective management, further studies on ANS are needed, aiming to provide better outcomes for affected individuals.

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SPG related to the spast gene (SPG4): case report of a novel variant

João Felipe Pissolito[1], Wilson Falco Neto[1], Gustave Rocha Rodrigues[1], Eliana Meire Melhado[1], Natália Prando[1], Renan Cenize Guardia[1], Laura Alonso Matheus Montouro[1,2]

[1]Centro Universitário Padre Albino, Faculdade de Medicina de Catanduva, Catanduva SP, Brazil. [2]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: João Felipe Pissolito; Email: felipe.pissolito@gmail.com.

Case presentation: Female patient, 65 years old, non-consanguineous parents, weakness and stiffness in the lower limbs 25 years ago in a progressive evolution. She denies abnormalities in sensitivity, dysphagia or cognitive complaints. Father, two brothers, a sister and four nieces with a similar condition. On physical examination: preserved cognition, significant spasticity tone in the lower limbs (Grade IV - Ashworth Scale), positive Hoffmann and Babinski signs bilaterally, grade 5 strength in the upper limbs and grade 3 strength in the lower limbs (MRC Scale), preserved coordination and exteroceptive/proprioceptive sensitivity. She underwent brain MRI, which showed signs of corpus callosum atrophy and mild diffuse brain volumetric reduction. Commercial genetic panel for hereditary spastic paraparesis revealed a heterozygous mutation: c.886_887del:p.Asn296*, in exon 6 of the SPAST gene. Baclofen 10 mg/day prescription, indicated motor physiotherapy and genetic counseling. **Discussion:** Our proband presents the pure phenotype of hereditary spastic paraparesis (HSP) as progressive weakness of the lower extremities and spasticity without other signs. This variant is a frameshift deletion mutation that results in the deletion of two nucleotides from position c.886 to c.887, resulting in an earlier stop codon compared to the wild-type protein. This leads to the truncation of the synthesis of the coding protein, thus losing its normal function. This new variant was predicted as probably pathogenic by the American College of Medical Genetics and Genomics, absent from gnomAD and ClinVar and scientific literature. Corpus callosum thinning and mild diffuse brain volumetric reduction, it's unusual but can occur in SPG4 and other HSP, although it is most frequently associated with SPG11. Therefore, given the compatible clinical condition, neuroimage, autosomal dominant inheritance pattern and probable pathogenic variant, in heterozygosity, in the SPAST gene, the diagnosis of SPG4 was confirmed by a new genetic variant. **Final comments:** Epidemiological studies and further case reports of HSP in Brazil are necessary to characterize genotype and phenotype of the disease. Performing genetic study to diagnose and classify the disorder is essential in the control and targeted treatment of the disease and genetic counseling.

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Sphenoidal sinusitis with neurological complications: a case report

Ricardo Kendy Nishiuchi de Paiva[1], Breno Gonçalves Medeiros[1], Matheus Hisamitsu Facine[1], Lucas Do Rego Barros Correa Martins[1], Humberto Ramos Crispim[1], Maria Izabella Grigoli Passarello Afonso[1], Maria Antunes Villa[1], Leticia Akemi Rama Yamamoto[1], Gabriela Rodrigues Sousa[1], Nasser Faiçal Abou Zeenni Nfa Zeenni[1]

[1]Hospital de Base de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Ricardo Kendy Nishiuchi de Paiva; Email: ricaknpaiva@gmail.com.

Sphenoidal sinusitis is the rarest type and can cause severe damage, mainly ophthalmological. Involvement of the II, III or IV cranial nerves can lead to permanent deficits or, in severe cases, fatalities. Analysis of the clinical evolution of a patient with optical impairment due to sinusitis. **Case presentation:** A 78- year old female presented with a 45-day history of pain in the left cheek and a left frontal parietal headache, which initially improved with over-the-counter analgesics. Two weeks prior to seeking medical attention, the headache returned and she experienced a sudden decrease in visual acuity in the left eye, accompanied by proptosis and ophthalmoparesis with difficulty on left eye adduction. Brain MRI revealed presence of a T2-hyperintense signal suggesting secretion accumulation in the right maxillary sinus, sphenoid sinus and left ethmoidal cells. Comparison with a previous MRI showed a bone dehiscence on the left lateral wall of the sphenoid sinus and the left posterior wall of the ethmoid, resulting in a communication with the left orbital fissure. This extension of the affected area with secretion has impacted the left orbit and, mainly, the III and IV cranial pairs. The patient underwent sinus surgery. Fluid-cultures revealedPseudomonas aeruginosas growth. Empiric treatment with piperacillin-tazobactam was followed by antibiotic de-escalation to levofloxacin, prescribed for two weeks. No improvement of the cranial nerves affected was observed on follow-up. **Final comments::** Although optical impairment is a rare complication of sinusitis, the patient had severe sequelae due to the delay in diagnosing the infection, which resulted in permanent deficits.

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Sphenopalatine ganglion block in refractory cluster headache: a case report

Luis Armando Sawada[1], Annelise Akemi Higa Lee[1], Ana Carolina Montouro Storarri[1], Beatriz Marques dos Santos[1], Bruno Pissolati Mattos Gonzaga[1], Lorrane Campidelli Arthuzo[1], Renan Barros Domingues[1]

[1]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Annelise Akemi Higa Lee; Email: a.higalee@gmail.com.

Case presentation: A 58-year-old woman presented with intense, right-sided orbital pain, with 60-minute duration attacks occurring 1-2 times a day, mostly at night. These were associated with right-sided ptosis, conjunctival hyperemia, lacrimation, and rhinorrhea. The patient had previously tried prophylactic treatment with verapamil 320 mg/day, achieving only partial reduction in attacks. Valproic acid and lithium were added at different times to optimize treatment but were discontinued due to adverse reactions. Initially, the cluster headache responded to prednisone 1 mg/kg; however, due to long-term effects, the medication was tapered off, leading to a return of attacks shortly after. Acute treatment with sumatriptan (oral and subcutaneous) resulted in poor responses, providing only mild relief. In most cases, complete relief was achieved only with oxygen therapy in the emergency department. Due to the refractory nature of the condition, bilateral occipital nerve block with lidocaine and dexamethasone was performed while maintaining verapamil at 320 mg/day. However, the patient still experienced severe headaches, responsive only to oxygen therapy. Finally, an alternative approach using a sphenopalatine ganglion block was performed. The patient showed significant improvement on the same day of the procedure, with complete resolution of attacks and no need for any medications. This improvement was maintained in the subsequent days, allowing her to return to normal daily activities. **Discussion:** Cluster headache is a severe and debilitating trigeminal-autonomic headache. It is often underdiagnosed, has few therapeutic options, and in 10-20% of cases, patients can be refractory to acute and prophylactic medications. This case highlights the complexity of cluster headaches management and underscores the importance of an individualized approach in treatment. Although occipital nerve block is evidenced to be effective for many patients, therapeutic response can vary. In this case, the sphenopalatine ganglion block proved to be a safe and effective alternative, providing complete symptom relief. **Final comments:** Thus, it is crucial to consider individual characteristics when choosing treatment, recognizing that the sphenopalatine ganglion block can be a viable and highly effective option for patients with refractory cluster headaches.

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Spinal cord compression by cystic IgG4-related pachymeningitis mimicking neurocysticercosis: a case report

Cássia Caroline Aguiar da Ponte[1], David Augusto Batista Sá Araújo[1], Pedro Lucas Grangeiro de Sá Barreto Lima[1], Dânton Campos de Queiroz[1], Milena Sales Pitombeira[2], Bernardo Martins[3], Pablo Picasso de Araújo Coimbra[2], Pedro Braga Neto[4], Guilherme Diogo Silva[5], Paulo Ribeiro Nóbrega[6]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Hospital Geral de Fortaleza, Fortaleza CE, Brazil.
[3]Rede SARAH de Hospitais de Reabilitação, Brasília DF, Brazil.
[4]Universidade Federal do Ceará, Departamento de Medicina Clínica, Fortaleza CE, Brazil.
[5]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[6]Universidade Federal do Ceará, Departamento de Medicina Clínica, Fortaleza CE, Brazil.

Correspondence: Cássia Caroline Aguiar da Ponte; Email: cassiacaroline0311@gmail.com.

Case presentation: A 40-year-old woman presented in 2016 with imbalance and shuffling gait due to hydrocephalus, necessitating a ventriculoperitoneal shunt that moderately improved her symptoms. However, four months later, she developed progressive weakness in her left arm and leg, followed by dysphagia and inability to walk. Spinal cord magnetic resonance imaging (MRI) revealed extensive pachymeningitis with associated cysts causing spinal cord compression. A meningeal biopsy showed nonspecific inflammatory changes. Despite normal laboratory tests, serum and cerebrospinal fluid ELISA for Cysticercus cellulosae were positive, leading to a diagnosis of pachymeningitis associated with cysticercosis. Treatment with albendazole, praziquantel and intravenous methylprednisolone resulted in partial improvement. In September 2017, she was admitted to a neurology ward with dysphagia, dysphonia and tetraparesis. Subsequently, in October 2017, she underwent posterior fossa decompression and cervical laminectomy with dural excision. Histopathological examination of the dural samples indicated significant IgG4-positive plasma cells and storiform fibrosis, showing no signs of cysticercosis and leading to a diagnosis of IgG4-associated pachymeningitis with no evidence of other organ involvement. Treatment included IV methylprednisolone followed by cyclophosphamide and rituximab. Following treatment, the patient experienced improved motor functions and was able to walk with unilateral assistance. However, in 2019, she experienced recurrent dysphagia and dysarthria. In April 2022, she was diagnosed with advanced invasive ductal carcinoma of the breast and she died of sepsis and multiple organ failure. **Discussion:** IgG4 related disease (IgG4-RD) is a multisystemic disease that is characterized by storiform fibrosis associated with fibrogenic cytokines and IgG4 tissue deposition. The condition usually presents with tumor-mimicking lesions, more often affecting lacrimal glands, orbits, pancreas and pachymeninges. Previous studies reported that up to 35% of patients with IgG4-RD present with pachymeningitis, a rare inflammatory process affecting the dura mater that requires a comprehensive diagnostic approach due to its association with various underlying conditions, such as cysticercosis. In our case, the atypical presentation of cystic pachymeningitis and the positivity of serum and cerebrospinal fluid ELISA for Cysticercus cellulosae resulted in a misdiagnosis of neurocysticercosis. **Final comments:** We described a case of cystic IgG4-RD spinal pachymeningitis causing cervical spinal cord compression initially misdiagnosed as neurocysticercosis, delaying immunotherapy. Diagnosis of IgG4-RD remains challenging, as clinical, laboratory, and imaging findings can mimic other conditions. Early identification is crucial since spinal IgG4-RD pachymeningitis responds well to corticosteroids and anti-CD20 agents, thus averting further neurological disability.

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Spinal cord damage progression in hereditary spastic paraplegia SPG4

Carelis Gonzalez-Salazar[1], Thiago Junqueira R. Rezende[1], Luciana Ramalho[1], Marcondes Cavalcante França Jr[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Marcondes Cavalcante França Jr; Email: mcfrancajr@uol.com.br.

Background: Hereditary Spastic Paraplegia is a neurodegenerative disease characterized by length-dependent degeneration of the spinal corticospinal tracts. SPG4 is the most frequent subtype worldwide. Spinal cord (SC) atrophy is the neuroimaging hallmark of SPG4, but so far little is known about its progression over time. **Objective:** To characterize SC structural damage progression in symptomatic patients with SPG4 and to identify potential genotypic-phenotypic correlates. **Methods:** We performed a cross-sectional as well as longitudinal study of upper SC (C1-C4) cross-sectional area (CSA) and eccentricity in subjects with SPG4 and matched controls. Images were acquired on a 3T scanner Paired clinical evaluations were also obtained - the severity of the disease was quantified with the Spastic Paraplegia Rating Scale (SPRS-BR). **Results:** There were 42 patients (% male) and 44 controls (% male) in the first evaluation; 31 patients (% male)/ 32 controls (% male) for the second evaluation and 16 patients and 14 controls for the third evaluation, with 3.6- and 3.1-years interval between base-line and second evaluation for patients and controls respectively and 5.7- and 5.9- years interval between base-line and third evaluation for patients and control group respectively. Patients with SPG4 had significantly reduced CSA at all spinal cord level at the 3 cross-sectional analyses (base line, second and third scan). At C2 and C3, progressive atrophy was noticed in the interval between first and second evaluation ($p<0.01$) whereas at C3, progression was noticed between 2nd and 3rd visits. We did not find significant differences in the eccentricity (cross-sectional or longitudinal). **Conclusion:** Quantitative SC MRI is able to capture disease progression in SPG4 and should be explored in further studies.

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Spinal cord infarction and subarachnoid hemorrhage secondary to aortic dissection: a case report

Pedro Almeida Martins Pontes[1], Giovanni Caniçali Simões[1], Fernanda Maria Gonçalves de Sousa Moura[1], Henrique de Moraes Bernal[1], Ingrid Lorena da Silva Gomes[1], Tainá Fátima Ramos Gonçalves[1], Eduardo Takashi Takehara[2] , Mateus Paquesse Pellegrino[1], Marcele Schettini de Almeida[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Pedro Almeida Martins Pontes; Email: pedroalmeida.pontes@gmail.com.

Case presentation: A 73-year-old man presented with sudden chest pain radiating to the back. Initial exams included a positive troponin curve and a normal electrocardiogram. Therefore, a non-ST-segment elevation myocardial infarction was suspected. A coronary angiography showed a 90% proximal stenosis of the circumflex artery and a 70% middle stenosis of the left anterior descending artery. Suddenly after the procedure, he developed chest pain radiating to the back, followed by loss of pain and temperature sensation and weakness in the lower limbs, as well as distal weakness in the upper limbs. On neurological examination, there was paraplegia and distal paraparesis of the upper limbs. Also, there was loss of sensation at the sensory level of the seventh thoracic spinal segment and absent deep tendon reflexes in the lower limbs. Urgent brain computed tomography (CT) and angio CT of the head, neck, and thoracic aorta were performed. Brain CT showed a hyperdense area in the subarachnoid space of the upper cervical spinal cord and medulla, suggestive of hemorrhage. Angio CT was notable for ulceration, a 14 cm fusiform aneurysm and a mural thrombus in the descending portion of the thoracic aorta. Spinal cord MRI showed a hyperintense signal in the spinal cord at the C4 level, "snake eyes" sign indicating spinal ischemia at the T8-T9 level and intramedullary blood in cervical, thoracic, and lumbosacral regions. Thus, the diagnosis of spinal cord infarction and spinal subarachnoid hemorrhage secondary to thoracic aortic dissection was made. During the next few days, the patient showed substantial improvement of neurological symptoms, fully recovering strength and sensation in the upper and lower limbs. Due to this improvement we opted for expectant management without surgical treatment. He was discharged one month following the event, with plans to reassess the possibility of thoracic aortic surgery later. **Discussion:** Aortic dissection is a potentially fatal condition that presents with neurological symptoms in up to one-third of cases. Spinal cord infarction is one of the less common presentations, being even rarer when associated with subarachnoid hemorrhage. The proposed mechanism for this condition is extension of arterial dissection from the aorta to the vessels supplying the spinal cord, leading to bleeding into the subarachnoid space and secondary spinal cord ischemia. Treatment options include conventional or endovascular surgery to correct the aortic dissection, along with cerebrospinal fluid drainage for cases associated with spinal cord infarction. However, up to half of the patients treated conservatively show significant improvement, as it happened in this case. **Final comments:** Aortic dissection can rarely present with subarachnoid hemorrhage and spinal cord infarction, and early diagnosis and intervention may be related to better clinical outcomes.

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Spinal cord ischemia after endovascular aneurysm repair of abdominal aortic aneurysms: a case study

Ana Francisca de Barros Coelho Brandalise[1], Alexandre Luiz Siano[1], Matheus Zago[1], Ivo Marcos Darella Lorenzin Fernandes Neto[2], Eduarda Basso Badalotti[2], Angelo Marcelo Wosniacki Filho[2], Gabriel Angelo Garute Zenatti[2], Renato Endler Iachinski[2], Antonio Carlos de Andrade Soares[2]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Fundação Hospitalar São Lucas, Cascavel PR, Brazil.

Correspondence: Ana Francisca de Barros Coelho Brandalise; Email: anafbrandalise@gmail.com.

Case presentation: Female, 57 years, underwent repair of a pararenal aneurysm of the abdominal aorta, using an endoprosthesis. She was admitted to the ICU for the post-operative period and, 2 hours after the procedure, she developed sudden, symmetrical paraplegia in her lower limbs, associated with tactile and painful hypoesthesia in both lower limbs. In the neurological physical examination, he presented motor strength grade 0 in the lower limbs and grade 5 in the upper limbs, tactile and painful hypoesthesia referred to in the legs, and digital rectal examination with absence of sphincter control / absent bulbo-cavernous reflex. A computed tomography of the skull was requested, which showed no acute changes. During hospitalization, laboratory tests were performed (which did not show significant changes) and MRI of the thoracolumbar region, which demonstrated acute myelopathy, probably of a vascular nature, and a fracture due to probable insufficiency of the L1 vertebral body with slight retreat of the posterior wall. During the period, the patient showed improvement in sensitivity on physical examination, with tactile sensitivity with asymmetry between the hemibodies (right - T9 and left - T10), but maintained paraplegia in the lower limbs, rectal examination with lack of sphincter control and bulbo- cavernous absent. He remained clinically stable, with the possibility of being discharged for outpatient follow-up. **Discussion:** The diagnosis of spinal cord infarction is an infrequent entity that represents around 1% of all ischemia. Iatrogenic spinal cord ischemia is a rare and debilitating complication of some open or endovascular surgical procedures, including the repair of thoraco-abdominal aortic aneurysms, resulting from the interruption of blood flow in the aortic branches that supply the spinal cord. In these cases, treatment is still controversial and there is no evidence necessary for an optimal protocol to adequately treat iatrogenic ischemic injury. Some studies support the use of lumbar drainage in patients with spinal infarction after an aortic surgical procedure. In the remaining patients, it is recommended to control vascular risk factors and initiate antiplatelet or anticoagulant medications when clinically appropriate, a strategy used in our case due to the already established ischemic injury. **Final comments:** Spinal cord ischemia is an uncommon pathology, therefore the number of clinical cases reported is low. Therefore, cases like this need to be discussed so that situations like this can be prevented and managed. There is still no literary consensus on what would be the best method adopted to treat this adversity once it has occurred, but it is known that it is necessary to increasingly carry out prevention measures, especially in surgical procedures in which there is a risk of spinal cord ischemia in order to avoid further harm to patients.

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Spinal muscular atrophy and home care: the relevance of an accurate follow-up in clinical care

Fabrício Loffredo D'ottaviano[1], Luis Fernando Grossklauss[1], Acary Souza Bulle Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Fabricio Loffredo D'Ottaviano; Email: fabriciodotta@gmail.com.

Background: Among the neuromuscular disorders, the hereditary proximal 5q spinal muscular atrophy (SMA) is a severe disease that causes progressive muscle weakness, mainly with infantile and childhood onset. SMA is caused by the lack of the survivor motor neuron (SMN), due to the deletion of the SMN1 gene. A less active gene, the SMN2, acts as a severity modifier. The number of SMN2 copies is inversely related to the clinical severity. **Methods:** Our study aims to describe the clinical features and the scope of home care in treating children with SMA. This is a retrospective review of records of pediatric neurology patients with SMA 1 or 2, over a 10-year period. **Results:** 12 SMA1 and 5 SMA2 patients (N=17) were discharged from the hospital and attended in home health care, provided by a multidisciplinary team. All SMA1 patients had nursing supervision 24 hours a day, whereas SMA2 had at most 12 hours daily. Concerning the use of ventilatory support, 5 patients used invasive ventilation and 12 used non-invasive ventilation. All SMA2 children used non-invasive mechanical ventilation only during night sleep, but most of SMA1 were ventilated for more than 20 hours a day (N=10, or 58% relative to N=17). In terms of monitoring colonization and guiding the rational use of antibiotics, 5 children had + Pseudomonas aeruginosa in quantitative cultures of endotracheal aspiration (all SMA1). Every patient underwent gene therapy such as Nusinersen (N=11), Risdiplam (N=2), Onasemnogene Abeparvovec plus Nusinersen (N=2), or Onasemnogene with Risdiplam (N=1). Six patients with Type 1 underwent a tracheostomy, 5 of them before their first birthday. Among these patients, one SMA1 infant had a positive therapeutic response; he was tracheostomized when 3 months old, started Nusinersen when 5 months old, took Onasemnogene at 7 months, and achieved decannulation (removal of tracheostomy tube) when he was 1 year and 9 months old. Unfortunately, one patient passed away when her tracheostomy tube was pulled out accidentally, while she was being transferred from wheelchair to bed. **Conclusion:** Considering SMA as a disease of the motor neuron associated with weakness and progressive motor impairment, gene therapy has shown to be an effective treatment. No functional loss was demonstrated in our patients, and substantial improvement in motor milestones has been observed in some of the children under treatment. In the overall follow-up, developing home care plans and preparing professionals to work with neuromuscular disorders (NMD) with continuous care, optimized by the clinical aspects of each patient, seems fundamental in the therapeutic process. This accurate follow-up in clinical care contributes to early diagnosis and assertive interventions and has led to great results in the patients' daily lives, including the possibility of attending formal school education for some of them.

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Spinal muscular atrophy diagnosis in Latin American: the LATAM RegistrAme clinical registry

Elice Carneiro Batista[1], Edmar Zanoteli[2], Marcela Câmara Machado Costa[3], Jaqueline Almeida Pereira[4], Marcondes Cavalcante França Junior[5], Juliana Gurgel Giannetti[5], Rejane de Souza Macedo[1], Otávio Berwanger[6], Gisele Sampaio Silva[1], Luiz Vicente Rizzo[1]

[1]Hospital Israelita Albert Einstein, São Paulo, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.
[3]Escola Bahiana de Medicina e Saúde Pública, Fundação Para o Desenvolvimento das Ciências, Salvador BA, Brazil.
[4]Universidade Federal do Rio de Janeiro, Instituto de Puericultura e Pediatria Martagão Gesteira, Rio de Janeiro RJ, Brazil.
[5]Universidade Estadual de Campinas, Hospital de Clínicas, Campinas SP, Brazil.
[6]The George Institute for Global Health and Imperial College of London, London, UK.

Correspondence: Elice Carneiro Batista; Email: elice.batista@einstein.br.

Spinal muscular atrophy (SMA) is a rare genetic disease characterized by progressive loss of motor neurons located in the anterior horn of the spinal cord and brainstem, leading to progressive muscle atrophy and weakness. The most common form of SMA (5q-SMA) is classified into subtypes according to the age of onset and the maximum motor function achieved. This study is an international multicenter clinical Registry for SMA diseases (LATAM RegistrAme study NCT05475691) that includes patients with 5q-SMA types 1, 2, 3, and 4 of all ages and both sexes, treated at reference centers in six Latin American countries (Argentina, Brazil, Chile, Colombia, Mexico, and Uruguay). This study aims to describe the clinical characteristics of the participants, as well as the time to start disease-modifying treatment in patients who are using some disease-modifying therapy. Currently, the study already has 361 patients included (Argentina: 63; Brazil: 180; Chile: 35; Colombia: 54; Mexico: 20, and Uruguay: 7). The results show that subtype 2 (158 participants; 43.8%) constitutes most of the participants in this study, followed by subtypes 3 (114 participants; 31.5%) and 1 (86 participants; 23.8%). The average age of the included participants is 12.8 years old (SD 10.9 years), regarding sex at birth, 177 female (49.0%), 184 male (51.0%), regarding declared race, 214 (59.4%) white, 15 (4.2%) black, 3 (0.8%) indigenous and 116 participants (32.2 %) declare themselves or present the breed classification registered in their medical records, as others, and 12 (3.3%) as unknown. Symptom onset period 90.6% postnatal (2 weeks of age or older), at birth (up to 2 weeks) 4.2%, asymptomatic 2.8% and unknown 2,5%. Homozygous deletion of exon 7 of SMN1 is present in 323 included participants, heterozygous (deletion of exon 7 of SMN1) in 18 participants, heterozygous for two substitutions in SMN1 (point mutation) in 1 participant, and 19 participants present genetic reports where only the description "mutation compatible with 5q-SMA". Regarding the need for ventilatory support, 315 (87.3%) have never used invasive ventilation, 25 (6.9%) currently use invasive ventilation, and 114 (31.6%) currently use non-invasive ventilation. To date, 245 (67.9%) use or have had access to disease-modifying therapy, and 116 (32.1%) have never had access to therapies. Knowing the reality of patients' journeys is relevant for greater assertiveness in the clinical management of the disease and management of access to treatments that modify the course of the disease.

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Spinocerebellar ataxia type 19: an early-stage onset disease

Isabela de Carvalho Florêncio[1], Thiago Dias Fernandes[1], Victória Almeida Villamil[2], Artur Marques Ferreira[2], Guilherme Ribeiro Correa Garcia[2], Pedro Henrique Andreotti Arakaki[2], Mariana dos Santos Zanelato[2], Vinícius de Lima Montello Jardim[2], Nathan Cândido Teixeira[2], Michelle Fonseca Acosta[2]

[1]Universidade Federal de Mato Grosso do Sul, Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.
[2]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.

Correspondence: Vinícius de Lima Montello Jardim; Email: vmontello1@gmail.com.

Case presentation: Female, 28 years old, daughter of non-consanguineous parents, with no family history of neurological or psychiatric diseases, with a history of delayed neuropsychomotor development and gait ataxia even after starting to walk. The patient's motor skills worsened at the age of 18, with a loss of autonomy for the activities she was capable of performing, as well as behavioral changes (hallucinations and aggression). On examination, she presented with cognitive deficit, ataxic gait, slurred speech, head tremor, difficulty fixing her gaze, dystonic hand posture, bradykinesia and symmetrical appendicular ataxia. Magnetic resonance imaging of the skull showed marked cerebellar atrophy and the exome showed a point mutation (alanine for valine at codon 336) in the KCND3 gene compatible with autosomal dominant spinocerebellar ataxia 19 (SCA19). The patient was referred for multidisciplinary follow-up, with partial improvement in her psychiatric symptoms and, to date, slow progression of her neurological condition. **Discussion:** Spinocerebellar ataxia type 19 is a rare autosomal dominant neurodegenerative disease secondary to a pathogenic mutation in the KCND3 gene, which codes for the Kv4.3 potassium channel proteins present in neurons in the cerebellum and other areas of the central nervous system. The mutation leads to neuronal dysfunction, which is expressed in a slowly progressive cerebellar syndrome. It manifests as cerebellar syndrome, bradykinesia, dystonia, myoclonus, frontal lobe executive dysfunction and epileptic seizures. The age of onset of symptoms is variable and can occur from 2 years to young adulthood. There is no specific treatment. **Final comments:** Although spinocerebellar ataxias mostly manifest after the second decade of life, neurologists should be on the lookout for cases that start early, even in childhood.

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Spinocerebellar ataxia type 23 (SCA23): a case report of a rare cause of ataxia

Bruna Andrade de Oliveira[1], Natália da Costa Chiote Pinheiro[1], Carolina Rocha da Silveira Brugger Cardoso[1], Lucas Leroux de Ricchezza[1], Luiz Felipe Vasconcellos[1]

[1]Universidade Federal do Rio de Janeiro, Instituto de Neurologia Deolindo Couto, Rio de Janeiro RJ, Brazil.

Correspondence: Bruna Andrade de Oliveira; Email: brunaado97@gmail.com.

Case presentation: A 19-year-old woman presented a progressive dysarthria associated with bilateral resting and kinetic tremor that worsened when she was anxious. At the time she was submitted to a frenectomy without improvement. When she was 21, she began to present difficulty walking with frequent falls. At the age of 23, she started to present dysphagia, worse for liquids. All the symptoms continue to worsen over the years. Physical examination at the time she was 25 revealed gait ataxia, upbeat nystagmus, dysarthria, dysdiadochokinesia worse in the right upper limb, dysmetria worse in the right upper and lower limbs and no pyramidal signs. The patient had no relevant family history, has always been in good health and did not take any regular medications. At five years of disease progression she was submitted to a brain MRI that showed cerebellar atrophy, with atrophy of the middle and superior cerebellar peduncles with faint hypersignal on T2/Flair, which extends to the cerebellar cortex, apparent atrophy of the pons and the Hot Cross Bun sign. She was submitted to extensive investigation and the genetic testing identified a pathogenic variant in PDYN c.414G> T (p.Arg138Ser), so she was diagnosed with Spinocerebellar Ataxia type 23 (SCA23). She was referred to phonoaudiology, physiotherapy and occupational therapy and received genetic counseling. **Discussion:** SCA 23 is an autosomal dominant spinocerebellar ataxia that is characterized by mutant prodynorphin (PDYN) gene. The prevalence of SCA 23 is estimated to be less than 1:1.000.000 people. The clinical features, brain magnetic resonance imaging (MRI), and neuropathological findings are indistinguishable from other SCA subtypes, so the diagnosis is only made by genetic testing. The inheritance is autosomal dominant and the symptoms begin in adulthood. The treatment consists in genetic counseling and complementary therapies. Disease progression can be slow within wheelchair bound ocurring in more than 20 years after symptomatic disease onset. **Final comments:** SCA 23 is a very rare cause of adult-onset hereditary ataxia. It's important that physicians are aware of this condition because not all genetic panel for hereditary ataxia are able to identify the mutation that causes SCA 23. The diagnosis is important for genetic counseling and to bring awareness to this condition.

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Spinocerebellar ataxia type 8: case report and literature review

Bruno Wendorff Lucci[1], João Carlos Cervelin[1], Fernanda Coan Antunes[1], Pricila Bernardi[1], André Dias de Oliveira[1], Gisele Espíndola[1]

[1]Universidade Federal de Santa Catarina, Florianópolis SC, Brazil.

Correspondence: Bruno Wendorff Lucci; Email: brunowlucci@hotmail.com.

Case presentation: A 22-year-old male with non-consanguineous parents and two healthy siblings presented with no relevant family history. Motor and cognitive development were normal in early childhood. At age 9, he began experiencing uncoordinated movements, progressing to gait instability and falls. Symptoms included muscle spasms, orofacial and cervical dystonia, moderate dysarthria, mild dysphagia, malnutrition, and low weight. Medical history: Sleep apnea syndrome; recurrent febrile seizures in childhood; focal motor dysperceptive seizures (ceased at age 16); sensorineural hearing loss; ADHD and major depressive disorder. Neurological Exam: Bilateral winged scapula; pectus carinatum; orofacial and cervical dystonia; scandent speech; preserved eye movement with mild nystagmus; global hypotonia with diffuse muscle atrophy; upper limb hyporeflexia; bilateral patellar hyperreflexia with pendular reflexes; bilateral Achilles hyperreflexia with clonus; hypermetria and dysdiadochokinesia; bilateral intention tremor; broad-based gait with difficulty in tandem walking. Initial tests, including electroneuromyography, ataxia and epilepsy panels, array-CGH, Friedreich's ataxia, and exome sequencing, were normal or inconclusive. Whole genome sequencing showed one allele with 183 CTG*CTA repeats in the 3'UTR region of the ATXN8OS gene and one allele within the normal range (22 repeats). Cranial MRI showed bilateral cerebellar volume reduction. A trial with L-dopa and regular botulinum toxin applications to the neck showed a good response. **Discussion:** Spinocerebellar Ataxia Type 8 (SCA8) is a rare autosomal dominant disease characterized by CAG and CTA/CTG repeat expansions in the ATXN8 and ATXN8OS genes. Symptoms include scandent speech, dysarthria, swallowing disturbances, slowly progressive cerebellar ataxia, oculomotor disturbances, pyramidal syndrome, cognitive and motor disturbances, and sensory neuropathy. Atypical cases can present with epilepsy. The ATXN8 gene encodes a polyglutamine expansion protein, while the ATXN8OS gene expresses a noncoding RNA. SCA8 expansions involve toxic mechanisms at both protein and RNA levels. Polyglutamine toxicity correlates with repeat sizes, showing an inverse correlation between age at onset and repeat size. Typical SCA8 repeat sizes range from 91 to 200. Patients with ≥200 repeats may exhibit reduced toxicity. Pure CTG expansions are more pathogenic and correlate with earlier onset. This bidirectional expression parallels other triplet expansion disorders. **Final comments:** SCA8 mutations have been identified in isolated cases in Brazil, with higher prevalence in Nordic countries. The frequency of SCA8 in Brazilian ataxia patients highlights the importance of comprehensive genetic testing, including whole genome sequencing, for accurate diagnosis. The exclusion of SCA8 from standard ataxia panels may lead to underdiagnosis. Further research is necessary to understand the pathomechanisms and genetic variations of SCA8.

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Split-hand-like syndrome in X-linked form of Charcot-Marie-Tooth disease (CMTX) caused by GJB1 mutation: a comparative study of electrophysiological features

Ricardo Vergara Gianvechio[1], Wilson Marques Junior[1], Pedro José Tomaselli[1], Brenda Cristina Navas[2], Gabriela Rodrigues Ramos[2], Lucas Gondim Briand Vieira[1]

[1]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Hospital das Clínicas, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Ricardo Vergara Gianvechio; Email: ricardo.gianvechio@alumni.usp.br.

Background: The X-linked form of Charcot-Marie-Tooth disease (CMT1X) is a common form of inherited neuropathy. It's caused by mutations in the GJB1 gene. Some reports in the literature suggest that a "split hand syndrome-like" condition can be found in patients with CMT1X, characterized by more prominent atrophy in the abductor pollicis brevis (APB) than in the abductor digiti minimi (ADM). Nonetheless, we found no studies numerically describing this possible predilection by means of neurophysiologic parameters. **Objective:** This retrospective study was conducted to objectively evaluate the possible predilection of involvement of the APB (in detriment of ADM) in CMT1X, comparing them to CMT1A patients and to control patients without evidence of peripheral neuropathy and with normal nerve conduction studies (NCS). **Methods:** We analyzed the data from electroneuromyography of 59 patients (21 patients with GJB1 mutations, 26 patients with PMP22 duplication - CMT1A group, and 12 patients without peripheral neuropathy and with normal values of NCS). The epidemiological and clinical baseline characteristics were also collected for comparison. The mean age of the patients undergoing electroneuromyography was 36.1 years in the CMT1X group and 35.5 years in the CMT1A group. The ratio APB/ADM (the ratio between the CMAPs in the APB and in the ADM, respectively) was calculated in each patient. In patients where it was possible to calculate the ratio from both arms, we conventionally used the smaller ratio. We then compared the results from the three groups using a two-tailed Student's t-test. **Results:** The mean APB/ADM in the CMT1X group was 0.646 with a standard deviation of 0.444, while in the CMT1A group, the mean APB/ADM was 1.286 with a standard deviation of 0.578. The mean APB/ADM ratio was significantly lower in patients in the CMT1X group compared to patients in the CMT1A group (p=0.0001) and to patients in the control group (p=0.04). There was no statistical difference between the CMT1A group and the control group (p=0.06). **Conclusion:** Although this is a preliminary study, the results suggest a significant predilection for more pronounced atrophy in the APB compared to the abductor digiti minimi ADM in patients with CMT1X. This finding was quantitatively supported by the significantly lower APB/ADM ratios in the CMT1X group compared to both the CMT1A and control groups. The absence of a significant difference between the CMT1A and control groups suggests that this phenomenon is specific to CMT1X. This could be useful to generate hypotheses surrounding the pathophysiology of the disease. Further prospective and/or longitudinal studies should be conducted to evaluate the reproducibility of these results in larger samples and to determine the predictive values of the clinical and neurophysiological findings of "split hand-like syndrome," and the APB/ADM ratio in patients with suspected inherited neuropathies.

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Spontaneous intracranial hypotension with atypical presentation: a case report

Rodrigo Fernandes Nitsch[1], Julyana Leoni do Prado[1], Mateus Coltro Dornelles[1], Patrícia Áurea Andreucci Martins Bonilha[1]

[1]Pontifícia Universidade Católica do Paraná, Hospital Universitário Cajuru, Curitiba PR, Brazil.

Correspondence: Mateus Coltro Dornelles; Email: mateuscdornelles@gmail.com.

Case presentation: C.A.P, female, 70 years old with a history of depressive and anxiety disorders and obstructive sleep apnea and hypopnea presented to a Neurology outpatient clinic with a 1 year history of cognitive decline and postural instability, that began with sudden, severe holocranial headache, that resolved spontaneously after one week. There was no history of head and spinal trauma, nor previous surgeries Later, she developed an anterograde episodic amnestic symptoms associated with brief, self-limited confusional states, and, in one year, she has lost independence in instrumental and some basic activities of daily living. As time passed, her cognitive symptoms worsened while new ones manifested: liquid dysphasia, dysarthria, and ataxic gait disorder with recurrent falls. Initial laboratory evaluation was normal and polysomnography evidenced enhanced initial and REM sleep latency and reduced sleep efficacy; her brain magnetic resonance (MRI) revealed meningeal enhancement, Obliteration of perimesencephalic cisterns, straightening of the distance between mamillary bodies and pons, Inferior herniation of cerebellar compatible with Saggy brain Syndrome and intracranial hypotension diffuse volumetric reduction and numerous signs of Intracranial Hypotension, while her spinal MRI enlarged epidural space. Treatment with caffeine 200mg daily was initiated. After initial improvement, the Caffeine dosage was adjusted to 300mg a day with an almost complete clinical resolution. **Discussion:** Spontaneous Intracranial Hypotension is a clinical-radiological syndrome usually caused by dural tear, leaking meningeal diverticulum or venous-CSF fistula with unestablished national epidemiology. It is known to be a rare cause (but treatable and important differential diagnosis) of new-onset headaches with an approximate incidence of 1-3.8/100.000 habitants per year, being an orthostatic headache its most common (approximately 98%) symptom. It is known to mimic a myriad of neurological syndromes, such as motor, cranial nerve, ataxic and cognitive decline. Its diagnosis is made with clinical and imaging investigation, and etiologic diagnosis is important for invasive treatment (such as targeted blood patches or surgical interventions) which, coupled with intensive hydration and caffeine supplementation, help prevent catastrophic complications and potentially reverse severe debilitating symptoms. **Final comments:** We present a case of a patient with atypical presentation, facing limiting symptoms, impairing daily life activities, due to delayed diagnosis. This is a rare, though treatable central nervous system disease with satisfying response to clinical and surgical treatments, highlighting the importance of including Spontaneous Intracranial Hypotension as a differential diagnosis of rapidly progressive dementia on clinical practice.

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Sporadic Creutzfeldt-Jakob disease and MRI use: a case report

Áurea Maria Salomão Simão[1], Murilo Caldas Bellotto[1], Beatriz Hog Jorge Silva[1], Davi Legora[1], Diogo Haddad Santos[1]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.

Correspondence: Beatriz Hog Jorge Silva; Email: beatrizhog@gmail.com.

Case presentation: A 68-year-old woman, with a high school degree, came to Santa Casa de São Paulo's neurology clinic in July 2022 due to memory loss. Her symptoms started in December 2021 and worsened ever since. In those seven months, she also started presenting personality and behavioral changes and an unstable gait. Her family history is positive for Alzheimer's disease in two first-degree relatives. During this first visit she was still able to perform Basic Activities of Daily Living (BADL), and her laboratory and neurological exams were unremarkable, except for a Mini Mental State Examination (MMSE) score of 19/30. In August 2022, she reported sleep pattern alterations and repetitive speech, while maintaining her previous symptoms. The neurological exam showed bradykinesia and a worsening MMSE score of 14/30. She was then submitted to a Magnetic Resonance Imaging (MRI), which showed both the cortical ribbon and the hockey stick signs, the classic imaging presentation of Creutzfeldt-Jakob Disease (CJD). The cortical ribbon sign refers to a curvilinear hyperintense signal involving the cerebral cortex of parietal and occipital lobes on DWI images. The hockey stick sign is described as a hyperintense signal involving the pulvinar and dorsomedial thalamic nuclei bilaterally on the FLAIR view. Once the hypothesis was made, we communicated it to the family, who refused to follow up with the investigation, denying the electroencephalogram (EEG) and cerebrospinal fluid (CSF) exams. One month later, she developed urinary and fecal incontinence and had lost the ability to feed herself. Being the MRI the only exam available in this patient's case, we would like to bring light to the importance of recognizing the typical imaging signs of Creutzfeldt-Jakob disease (CJD). **Discussion:** CJD is a rapidly progressive dementia associated with neurodegeneration and the most common prion disease in humans. Its diagnosis still depends on neuropathological confirmation through biopsy, but the use of other tools such as imaging (MRI), along with the clinical presentation, EEG and CSF analysis for the 14-3-3 protein have shown to be enough for managing the patient. Before assuming it is a CJD case, we must first exclude reversible causes of dementia through imaging and blood tests. We should also first consider more prevalent etiologies such as Alzheimer's disease. In this context, the MRI is considered by the literature as a fundamental tool to detect early signs of neurodegeneration, with a high sensitivity (91%) and a high specificity (97%). Approximately 80% of the patients with CJD had alterations in the MRI, and 60% of them exhibited the classical signs. **Final comments:** In conclusion, CJD is an irreversible dementia with a 100% mortality rate. The fact that 85% of its presentation is idiopathic does assure us the importance of laboratory tests and imaging, especially the MRI in indicating the diagnosis and, therefore, granting the best care for the patient.

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Sporadic Creutzfeldt-Jakob disease as a prototype of rapidly progressive dementia: a case report

Caique Alberto Dosualdo[1,2], Lucas Do Rego Barros Correa Martins[1,2], Matheus Hisamitsu Facine[1,2], Humberto Ramos Crispim[1,2]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.
[2]Fundação Faculdade de Medicina de Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Caique Alberto Dosualdo; Email: caiquedosualdo100@gmail.com.

Case presentation: From the review of medical records, we found the case of a patient who evolved for six months with progressive gait ataxia and motor incoordination, associated with postural instability and symptom of depression and apathy. Patient presented several hospitalizations due to confusional episodes, associated with progressive cognitive dysfunction, mainly short-term memory, spacial disorientation and executive dysfunction. After this period, the patient became bedridden and developed generalized myoclonus, associated with akinetic mutism. Diagnostic approach was performed with cerebrospinal fluid, without alterations and with negative 14-3-3 protein, but with evidence of an hypersignal basal

ganglia bilaterally and parashagitais region of frontal lobes in brain resonance. Electroencephalogram showed frequent generalized epileptiform discharges in periodic pattern. Patient evolved with death in the same hospitalization after infectious complications. **Discussion:** Sporadic Creutzfeldt-Jakob disease is one of the most dramatic causes of rapidly progressive dementia, and it is characterized by neuropsychiatric symptoms such as depression, apathy, and cognitive changes, especially in memory, social cognition, and executive functions within a period of less than one year. It is often accompanied by complaints of cerebellar ataxia, signs of pyramidal syndrome, and myoclonias and other movement disorders. This pathology represents more than 90% of the sporadic diseases caused by prions. **Final comments:** Rapidly progressive dementias have been considered medical emergencies due to the need of quick diagnosis, since some of the possible etiologies can be reversible.

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Ss-amyloid deposition, impairment cognitive and glymphatic system dysfunction after pneumococcal meningitis experimental

Beatriz Brandão Lima[1], Samuel Brasil Stonoga[1], João Victor Vicente Costa[1], Cristiano Julio Faller[2], Amanda Christine da Silva Kursancew[2], Eloa Melpomene Piva[1], Bruno Marcon[1], Natalli Studnicka[1], Diogo Dominguini[2], Jaqueline S. Generoso[2]

[1]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.
[2]Universidade do Extremo Sul Catarinense, Programa de Graduação em Ciências da Saúde, Laboratório de Neurologia Experimental, Criciúma SC, Brazil.

Correspondence: Beatriz Brandão Lima; Email: beatrizbrandao183@gmail.com.

Bacterial meningitis is a severe infection caused mainly by Streptococcus pneumoniae in adults, inducing brain dysfunction with microglial activation and release of inflammatory mediators. This pathology is a medical emergency, and the neuroinflammation triggered can lead to a generalized lesion of the meninges and brain tissue. The central nervous system relies on the glymphatic system to remove debris and agents in an infection. This system is highly dependent on aquaporin-4 (AQP4) water channels, and its dysfunction may contribute to the worsening of neuroinflammation caused by meningitis. This study aimed to evaluate the glymphatic system activity, β-amyloid (βA) peptide deposition, and behavioral parameters in adult Wistar rats 30 days after induction of pneumococcal meningitis. At 60 days of age, rats Wistar were subjected to anesthesia with intraperitoneal administration of ketamine and xylazine. Then, the animals received, via the cisterna magna, 10 μL of S. pneumoniae suspension (group meningitis) or with artificial cerebrospinal fluid (sham group). The animals were treated with ceftriaxone (100 mg/kg) twice daily for seven days. Thirty days after meningitis induction, animals were subjected to behavioral tests. Immediately after the behavioral tests, 25 μL of albumin and Evans blue (EBA) were injected into the cisterna magna to evaluate the activity of the glymphatic system. Other animals were used for cerebral evaluation of the βA peptide. The animals in the group with meningitis presented impairment of habituation memory and a reduced ability to recognize memory. Furthermore, this study demonstrated an increase in positive cells for βA expression in the prefrontal cortex and hippocampus of animals in the meningitis group when compared to the sham group and lower concentrations of EBA in the serum of the meningitis group about the sham group, indicating impairment of glymphatic system activity due to retention of the EBA in the brain. Our results showed that during pneumococcal meningitis, there were losses in the drainage of βA due to the malfunction of the glymphatic system, leading to cognitive impairment. Loss of function of the glymphatic system may, therefore, be one of the causes of sequelae neurological disorders after pneumococcal meningitis.

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Stability of control of epileptic drop attacks with selective posterior callosotomy: do good results persist over the years?

André Palmini[1], Eliseu Paglioli[1], Thomas Frigeri[1], Natalie Donida[1], Bibiana Thome[1], William Martins[1], Fernanda Schuh Martins[1], Rafael Paglioli[1], André Gus[1], Rhana Santos[2]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Universidade do Vale do Rio dos Sinos, Porto Alegre RS, Brazil.

Correspondence: Andre Luis Fernandes Palmini; Email: alpalmini@gmail.com.

Epileptic drop attacks are perhaps the most malignant type of seizures, often leading to recurrent trauma and demanding constant supervision while patients are standing and walking. The underlying etiology is usually associated with severe diffuse brain dysfunction and these seizures are virtually always refractory to medications. Corpus callosotomy has been the cornerstone of the surgical treatment, based upon the premise that bilateral hypersynchronization of epileptiform activity is the starting point of a very fast multisynaptic cascade leading to falls. Moreover, several studies suggested that complete callosal sections were the more effective strategy, compared to partial disconnections. However, because new data showed that fibers originating from epileptic premotor and motor cortex - ie, the most relevant fibers for tonic and atonic seizures - cross through the posterior body and isthmus of the callosum, we proposed 8 years ago a novel surgical strategy to control drop attacks, in which the posterior half of the corpus callosum is sectioned, preserving prefrontal interhemispheric connectivity. Results were very good and comparable to those reported with complete sections. Here we report on the stability of those results between and x 8 years later in 41 patients. We hypothesized that should selective posterior callosotomy truly section the relevant fibers for drop attacks, results would remain stable, despite preservation fo the anterior callosum. Forty-one patients operated until 2008, and who had a minimum follow up of 8 years when results were reported in 2016, were re-evaluated in 2024 through outpatient consultations or structured telephone interview. Frequency of drop attacks in the last 2 years was classified as daily, weekly, monthly, yearly or absent (ie, complete control). An estimate of the absolute number of falls per month was also obtained. Seizure frequency at 3 time points - before surgery (t0), at least 8 years later (t1), and 8 years following th previous follow-up (ie, at least 16 years after surgery; t2) were compared according to the Jonckheere-Terpstra test. Mean median frequency of drop attacks at the 3 timepoints was 180 ([Q1 90 - Q3 540]), 0 ([Q1 0 - Q3 25.5]) and 0 ([Q1 0 - Q3 30]) at t0, t1 e t2, respectively (p=0.000). These results suggest that the degree of control of drop attacks with selective posterior callosotomy is stable over large periods of time, giving credence to the hypothesis that the most relevant fibers leading to these seizures are sectioned with this procedure, and that anterior callosal fibers do not need to be sectioned to obtain very good seizure control.

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Stability of functionality for activities of daily living following section of the posterior half of the corpus callosum to control epileptic drop attacks

André Palmini[1], Eliseu Paglioli[1], Frederico Orlando Friedrich[1], Bibiana Thome[1], Natalie Donida[1], William Martins[1], Fernanda Schuh Martins[1], Rafael Paglioli[1], Rhana Santos[2], André Gus[3]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Universidade do Vale do Rio dos Sinos, Porto Alegre RS, Brazil.
[3]UPF, Passo Fundo/RS;

Correspondence: Andre Luis Fernandes Palmini; Email: alpalmini@gmail.com.

Epileptic drop attacks are severe and disabling, usually medically refractory, seizures often presented by children and adolescents with diffuse encephalopathies. Most have no or limited schooling and variable degrees of dependence for activities of daily living. We have shown that selective section of the posterior half of the corpus callosum (selective posterior callosotomy) brings forth dramatic control of the falls, comparable to that seen with complete callosotomy, with preservation of the prefrontal interconnectivity. Because traditional neurology regards preservation of the posterior callosum as relevant to avoid what has been known as posterior disconnection syndrome (ie, symptomatology related to lack of functional integration between the posterior areas of the hemispheres in real time), selective posterior callosotomy has been seen with some suspicion as bound to provoke clinically relevant disconnection syndrome. We hypothesize, however, that because the anterior callosal fibers are preserved, section of the posterior callosum would not lead to interference with tasks related to activities of daily living. M: 41 patients operated until 2008, and who had a minimum follow up of 8 years when results were reported in 2016, were re-evaluated in 2024 through outpatient consultations or structured telephone interview. Relatives were interviewed to assess level of dependence in activities of daily living (ADL) - feeding, dressing, toilet hygiene and sphincter control. Neurological examination focused on axial tonus, gait and word articulation. Gait could be independent or needing assistance and word articulation could be fluent, comprehensible or incomprehensible. Autonomy to eat, dress and go to the toilet, normal axial and jaw tonus, sphincter control, praxis to button and use a spoon, and ability to read scored 1 point each. Fluent speech and independent gait scored 2 each (barely comprehensible, dysarthric speech and walking with assistance scored 1 each, while inability to speak or walk scored 0). From these neurological and functional data we generated a composite functional score ranging from 0 to 13. Functionality scores were then obtained at 3 time points medians and interquartile ranges - before surgery (t0), at least 8 years later (t1), and 8 years following the previous follow-up (ie, at least 16 years after surgery (t2) using the Jonckheere-Terpstra test. RC: The trend analyses of the functional scoring showed significant variation between pre- and both post-operative time points. Medians were 7 ([Q1 4 - Q3 9]) at t0, 8 ([Q1 6 - Q3 10]) at t1 e 9 ([Q1 6 - Q3 10]) em t2 (p=0.041), suggesting a tendency of functional improvement over the years with then stabilization between t1 and t2. These results confirm the hypothesis that disconnection in patients udergoing selective posterior callosotomy does not worsen, but rather improve functionality in patients with severe epilepsies and intellectual deficiency.

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Statin-induced myopathy: a case report

Bruno Barreto de Oliveira[1] , Eustaquio Claret dos Santos[1]

[1]Hospital Socor , Belo Horizonte MG, Brazil.

Correspondence: Bruno Barreto de Oliveira; Email: brunobarreto2014@yahoo.com.br.

Case presentation: Female patient, 71 years old, with comorbidities (chronic CAD, dialysis CKD due to polycytic kidneys, previous TIA in January/24), was using AAS 100 mg/day, Ticagrelor 90 mg/day, Concor 5 mg/day, and Rosuvastatin / Ezetimibe - 20mg/10mg - began use 2 weeks before admission. Admitted due to UTI - treated with Ceftriaxone. It started suddenly with proximal paresis of the MSD and lower limbs, progressing rapidly. MSD paresis attributed to AVF. A brain MRI was performed which showed chronic ischemic microvascular changes and volumetric reduction of the brain parenchyma. Cervical MRI showed degenerative changes and reduced amplitude of the spinal canal, without signs of compressive myelopathy. Initial CK dosage on 03/12/24: 6278 U/L . The decision was made to suspend the statin, intensify physiotherapy and perform ENMG. It evolved with a progressive reduction in CK (4150 / 3020 / 231). ENMG of the lower limbs performed on 03/18/24 showed changes suggestive of a myopathic process with the presence of pseudomyotonic discharges - polyphasic of low amplitude and short duration. Pulse therapy was performed with methylprednisolone 1g/day for 5 days. The patient developed a slow and progressive improvement in proximal strength in the lower limbs. He was discharged with instructions to maintain Prednisone 60mg/day in addition to home physiotherapy. **Discussion:** Statin-associated muscle symptoms (SAMS) range in severity from mild-to-moderate muscle pain, weakness, or fatigue to potentially life-threatening rhabdomyolysis. The mechanisms causing SAMS are not defined, but probably result from decreased production of noncholesterol endpoints of the mevalonate pathway. Muscle symptoms, with or without elevation of creatin kinase are one of the main adverse effects of statin therapy, a fact that sometimes limits their use . Statin-related myotoxicity is a common, probably underdiagnosed problem in clinical practice. **Final comments:** Periodic monitoring of patients using statins, knowledge of the pathophysiology of statin-induced myopathy, in addition to the correct management of adverse events, tends to minimize situations of discontinuity in the use of the drug, maintaining the benefits that statins provide to patients at risk. high cardiovascular.

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Statins as potential modifiers of Parkinson's disease: a systematic review with meta-analysis

Jonatas Paulino da Cunha Monteiro Ribeiro[1], Ocílio de Deus da Rocha Ribeiro Gonçalves[1], Arquimedes Barros Nascimento[1], Victor Gonçalves Soares[2], Gielson de Sousa Silva[1], Laura Martins Soares Cortez[1], Lara Beatriz Alves Batista[1], Kayron Rodrigues Lima[1], Társis Vinícius Cronemberger de Carvalho Moura Mendes[1], Kelson James Almeida[1]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Universidade Federal dos Vales do Jequitinhonha e Mucuri, Diamantina MG, Brazil.

Correspondence: Jonatas Paulino da Cunha Monteiro Ribeiro; Email: jonatas.ribeiro@ufpi.edu.br.

Background: Recent evidence indicates that statins have anti-inflammatory and antioxidant effects, leading to several investigations of whether statins have a neuroprotective effect and may benefit patients with Parkinson's disease (PD). However, the potential mechanisms of this class of medications as modifiers of the course of PD in already diagnosed patients is still unclear. **Objective:** To assess the effectiveness of statins as modifiers of established PD. **Methods:** Two investigators systematically searched for randomized clinical trials and observational studies comparing PD patients treated with statins versus those treated without statins. The search was conducted in PubMed, Embase, Cochrane, and Web of Science. The outcome of interest was the change in the MDS-UPDRS III (Movement Disorder society- Unified Parkinson's Disease Rating Scale Part III) score from baseline. Statistical analysis was performed using R Studio 4.3.2. **Results:** Four studies, totaling 472 patients, were included. The mean difference for the MDS-UPDRS Part III scale between statin and non-statin groups was-0.98 points (95% CI-3.40 to 1.45; p=0.09; I^2=54%). Meta-regression results showed that follow-up time accounted for 77.37% of heterogeneity among studies and 69% of the mean difference. The test for residual heterogeneity was not significant (p=0.283). The regression graph intercept was-6.5714 points, and the coefficient for follow-up time was 0.2913 points, although it did not result in a significant increase in the MDS-UPDRS Part III score over time (p=0.094). **Conclusion:** There was no statistical significance for the potential clinical benefit statins as modifiers of the disease course in PD patients.

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Statistical analysis of the use of disease-modifying therapies in multiple sclerosis in a neuroimmunology outpatient clinic in Cuiabá-MT

Helen Luiza Silvestrini[1], Leticia Lage Silva[1], Maryam Yehya Ghalfi[1], Sophia Oliva Schommer[2], Beatriz Fulador Maciel[3], Rafaella Resplande Xavier[3], Bianca Barbosa Araldi Freitas[3], Klesia Adaynny Rodrigues[3], Victor Hugo de Souza Silva Gomes[3], Heloise Helena Siqueira Borges[3]

[1]Universidade de Cuiabá, Cuiabá MT, Brazil.
[2]Universidade Federal do Mato Grosso, Cuiabá MT, Brazil.
[3]Hospital Geral de Cuiabá, Cuiabá MT, Brazil.

Correspondence: Helen Luiza Silvestrini; Email: helensilvest@gmail.com.

Background: Multiple sclerosis (MS) is an autoimmune disease of the central nervous system that affects about 15 out of every 100,000 Brazilians. It is characterized by an inflammatory response against oligodendrocytes and neuronal dysfunction. Its clinical manifestation is extremely variable and complex due to the various sites of inflammation. Treatment is divided into acute phase therapies and disease-modifying therapies (DMTs), which aim to minimize relapses and the progression of disability by suppressing inflammation. These therapies include low/medium efficacy options such as dimethyl fumarate and fingolimod, and high efficacy options such as monoclonal antibodies and cladribine. **Objective:** To analyze the different disease-modifying therapies for Multiple Sclerosis in a neuroimmunology outpatient clinic and the profile of patients undergoing treatment, as well as to evaluate the use of high-efficacy therapies implemented. **Methods:** This is a cross-sectional study observing DMTs in 31 patients diagnosed with MS. Data were obtained from medical records, covering variables such as sex, age, race/color, Expanded Disability Status Scale (EDSS), clinical form, and current medication. Informed consent was obtained from participants and the study was submitted to the hospital's Ethics Committee. **Results:** The sample consisted of 31 patients diagnosed with MS, with a female to male ratio of 2.4:1. The average age was 37 years, ranging from 16 to 65 years. Regarding ethnic distribution, 55% (n=17) of patients were mixed-race. The average EDSS score was 3 among the studied forms. The most prevalent clinical form in this hospital service was relapsing-remitting MS, identified in 27 patients. It was observed that 61% (n=19) of patients use high-efficacy medications, such as natalizumab (16) and ocrelizumab (3), while 39% (n=12) use low-efficacy DMTs like beta interferon (3), dimethyl fumarate (4), and teriflunomide (5). **Conclusion:** The analysis of different disease-modifying therapies revealed a preference for high-efficacy agents like natalizumab and ocrelizumab over less effective immunomodulators. This study highlights the importance of continuous personalized therapeutic adaptation to minimize relapses and disability progression caused by MS, aiming to preserve the quality of life of affected patients. As consolidated in various global references, this service opts to employ a high-efficacy strategy across different types of MS over a stepwise therapy approach to minimize disease progression.

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Stem-cell transplant for refractory myasthenia gravis: first Brazilian case report and literature review

Michelle Abdo Paiva[1], Beatriz Carneiro Gondim Silva[1], José Pedro Soares Baima[1], Luiz Gomes de Sá-Neto[1], Mariana Hiromi Manoel Oku[1], Carlos Wilson de Alencar Cano[2], Mariana Nassif Kerbauy[2], Nelson Hamerschlak[2], Edmar Zanoteli[1], André Macedo Serafim Silva[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.
[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Michelle Abdo Paiva; Email: michelle.abdop@gmail.com.

Background: Myasthenia gravis (MG) is an autoimmune disorder that can significantly impact functionality and quality of life. Approximately 10% of patients remain refractory to treatment. Stem cell transplant (HSCT) has been reported as an option, although no clinical trials are available yet. To our knowledge, HSCT for MG was not previously reported in Brazil. **Objective:** To report the first brazilian case of autologous HSCT (aHSCT) for MG and to review reported cases in literature, presenting all results compiled. **Methods:** We prospectively followed medical records and assessed response to treatment from our patient who underwent aHSCT. We also systematically searched in Pubmed and Google Scholar "stem cell transplant" and "myasthenia gravis". We identified 10 articles and one book chapter reporting MG patients that underwent HSCT. **Results:** Female patient diagnosed with MG anti-acetylcholine receptor antibody (ACh-R) positive at the age of 46 after presenting global weakness and dyspnea. Over a period of more than 3 years, she failed to respond to various therapies including pyridostigmine, ambenonium, prednisone, azathioprine, tacrolimus, cyclosporine, rituximab, eculizumab, ravulizumab, intravenous cyclophosphamide, thymectomy and plasma exchange. Due to the severity of her symptoms and recurrent symptoms, she received intravenous immunoglobulin (IVIG) every 15 days during the last three years and monthly cyclophosphamide over the last year, which significantly affected her quality of life. After conjoint decision, she underwent aHSCT. Before HSCT the patient scored 15 points on the MG-ADL and 34 points on the MGC. Early post-transplant complications included bloodstream infection and pseudomembranous colitis. Neutrophil engraftment occurred on D+10.The patient experienced improvement of symptoms on D+12. Her MG-ADL and MGC scored 0 points at discharge which was maintained until the last follow-up at 3 months. In total, 26 cases were identified in the literature (27, after including ours). Of the total, 17 patients were female, 5 males and 5 had no report on sex. Mean age at the time of HSCT was 44 years-old. Twenty cases were positive for ACh-R antibody and five for anti-muscle specific kinase. MGFA status previous to HSCT ranged from III-V. All patients had already tried multiple treatments, ranging from four to 11 different drugs and 59% had undergone thymectomy. Allogeneic HSCT was reported in two cases and the remaining were aHSCT. Most complications during HSCT were transplant-expected, such as febrile neutropenia and herpes family virus reactivation. MGFA status post-HSCT showed remission or improvement for 25 patients (92%). One death was reported during HSCT and one had no outcome reported. **Conclusion:** HSCT has been described as an option to treat many auto-immune disorders, such as multiple sclerosis. For MG, according to our data, HSCT appears to be relatively safe and effective and may be an option to treat refractory MG patients.

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Stereotactic radiosurgery versus whole brain radiotherapy in brain metastasis: a systematic review and meta-analysis of randomized controlled trials

Luiza Cersosimo Costa[1], Riesly de Oliveira Vasconcelos[2], Caio Paulain Cavalcante[3], Vitor Lopes[2], Artur Menegaz de Almeida[4], Ítalo Barros Andrade[5], Lilianne Rodrigues Fernandes[6]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.
[3]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[4]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[5]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[6]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.

Correspondence: Caio Paulain Cavalcante; Email: caiocavalcante754@gmail.com.

Background: The treatment landscape for brain metastases has significantly evolved in recent decades, driven by advances in radiotherapy techniques. Two primary approaches have been extensively studied: stereotactic radiosurgery (SRS) and whole brain radiotherapy (WBRT). SRS offers millimeter-precision in radiation delivery, limiting exposure to surrounding healthy tissues, whereas WBRT traditionally aims to treat diffuse brain micrometastases more broadly. Choosing between these modalities remains a subject of ongoing debate in clinical practice, focusing on maximizing local disease control, preserving cognitive function, and improving patients' quality of life. In this meta-analysis, we explore the outcomes of randomized controlled trials directly comparing SRS and WBRT in patients with brain metastases. **Objective:** Comparison of stereotactic radiosurgery versus whole brain radiotherapy, analyzing control of intracranial tumor (ITC), overall survival (OS) and quality of life (QOL). **Methods:** We performed a systematic review and meta-analysis assessing the effect of stereotactic radiosurgery in comparison with whole-brain radiotherapy in patients with brain metastatic cancer. We searched PubMed, MEDLINE, Scopus, and Web of Science for eligible trials. A random-effects model was used to calculate the odds ratio with 95% confidence intervals (CIs). Statistical analyses were performed using RStudio version 4.3.2. **Results:** Fifteen trials were included in this study, with all patients with a history of brain tumors. The choice foi stereotactic radiosurgery did not have significant differences over whole brain radiotherapy in the control of intracranial tumor (OR 2.05; 95% CI 0.58-7.19; p=0.61). There were also no significant differences between groups for overall survival (OR 1.14; 95% CI 0.83-1.58; p=0.61) and quality of life (OR 2.02; 95% CI 0.75-5.42; p=0.62). **Conclusion:** The results did not show a benefit of stereotactic radiosurgery over whole brain radiotherapy in all three parameters analyzed, revealing no difference between them about performance for control of intracranial tumors, providing a better quality of life, and overall survival. Therefore, we need more studies for better avaliation of these therapies.

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Stiff person syndrome (sps) presenting as lumbar subacute pain: a case report

Isabella Nascimento Guedes[1], Karoline da Silva Guimarães[1], Gilvan Pires de Castro Junior[1], Matheus Pinto Munoz[1], Lilian Eduarda Bastos da Silva[1], William Corá[1], Darah Fontes da Silva Assunção, Marina Buldrini Filogonio Seraidarian[2]

[1]Universidade Estácio de Sá, Rio de Janeiro RJ, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Isabella Nascimento Guedes; Email: isabellaguedes21@icloud.com.

Case presentation: A 48-year-old female reported intense pain in the lumbar region and hips for 4 months. The case was initially managed as a low back pain, but the diagnosis was later on switched to radiculopathy. The patient progressed with worsening of pain, gait impairment and lower limb tremors, which led her to falls. Clinical examination revealed painful spasms, increase in muscle tonus and presence of rigidity when extending lower limbs. It was also observed resistance in passive and active flexion, particularly on the left lower extremity. Imaging exams performed were: brain CT with no significant findings and spine MRI with lumbar discopathy but no myeloradicular conflict. Laboratories findings were normal. EMG revealed mild axonal distal neuropathy of left limb deep fibular nerve. Anti-GAD assay: results greater than, 2000U/mL suggesting SPS. In this case, the patient was treated with baclofen and clonazepam. Although the patient had a good overall response, she sustained a tonic posture on the left lower limb. However, there were no new episodes of painful spasms. The increased muscle tonus was managed with drug dosage adjustment. **Discussion:** Stiff Person Syndrome (SPS) is a rare neurological autoimmune disease affecting 1 in 1 million people. It is more predominant between 20 and 50 years, and it has a higher incidence in women population. It is characterized by painful spasms and progressive rigidity of axial and proximal limb muscles, mostly. Muscle stiffness can be triggered by noise, tactile stimuli, variations of temperature or psychological stress. Moreover, the disease can progress to gait impairment and recurrent falls. The pathophysiology resides in the production of autoantibodies against glutamic acid decarboxylase (GAD), which leads to a decreased GABA production in the presynaptic neuron and also intense muscle contractions. Treatment consists in using drugs that increase GABAergic tone such as benzodiazepines, and immunotherapy to be further used to reduce neurological 2 / 2 symptoms. Finally, the diagnosis is challenging and often reached by the exclusion of other diseases. SPS's prognosis is reserved, and its progressive course can potentially lead to disability and fatality. **Final comments:** SPS is a rare disease and the literature regarding this pathology is very limited. Thus, cases are often underdiagnosed. This report aims to contribute to the medical literature and improve diagnostic accuracy, therefore reducing morbimortality.

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Stiff person syndrome with anti-glycine receptor antibody: a case report

Pedro Maia Nobre Rocha Saffi[1], Leonardo Jadyr Silva Rodrigues Alves[1], Andresa do Rêgo Barros Vieira Santos[1], Talita Novak Thomezyk[1], Lucas Ferreira Alves[1], Ana Carolina Araujo[1], Maria Cecília Mendes Resende[1], Felipe Von Glehn Silva[1], Márcia Silva Santos Neiva[1], Pedro Renato de Paula Brandão[1]

[1]Hospital Universitário de Brasília, Brasília DF, Brazil.

Correspondence: Pedro Maia Nobre Rocha Saffi; Email: pedromaiabsb@gmail.com.

Case presentation: A 33-year-old female patient attended her first consultation in March 2024, reporting hip pain associated with the femoral head rotation, which began in June 2023. Over time, she developed pain extending from the thoracic to the sacral region and rigidity predominantly in the right lower limb (RLL), with difficulty walking due to inversion and plantar flexion of the right foot. She stated that her left foot was starting to show similar symptoms. She denied having any complaints regarding her upper limbs. During the consultation, she also reported urinary urge incontinence, bowel urgency, and a burning sensation in both feet. Physical examination revealed global hyperreflexia, 3+/4+ in the upper limbs (UL) and 4+/4+ in the lower limbs (LL), with an increased reflexogenic area. Muscle strength was grade III in the RLL for right foot dorsiflexion and right thigh extension and grade IV for leg flexion and extension, thigh flexion, and right foot plantar flexion, with hypertonia in the RLL (2+/4+). Muscle strength was grade V in the left lower limb (LLL). Bilateral extensor plantar reflex and positive bilateral Hoffmann's sign were observed. Anti-GAD65 was requested and came back negative. At the following consultation, she reported worsening pain in the LL, arthralgia in the knees and ankles bilaterally, and progression of rigidity in the abdomen and LL. She experienced two episodes of falls during this period. A lumbar puncture was performed, revealing oligoclonal bands restricted to the cerebrospinal fluid. Other tests, such as mammography and contrasted CT scans of the abdomen, pelvis, and chest, showed no abnormalities. During the same consultation, she presented weak positive results for anti-glycine receptor antibodies, and hospitalization for immunoglobulin therapy was requested. **Discussion:** Stiff Person Syndrome (SPS) is characterized by dysfunction in the GABAergic inhibitory pathway, presenting symptoms such as rigidity and painful muscle spasms, primarily in axial and proximal limb muscles. It is associated with high levels of anti-glutamic acid decarboxylase (anti-GAD) antibodies, with other antibodies such as anti-glycine receptor antibodies (anti-GlyR) also described. The classic form of the disease has an estimated prevalence of 1 to 2 cases per million, affecting more women than men. Diagnosis is based on recognizing the symptoms and is corroborated by serologies, response to diazepam, and electromyography (EMG) studies results, which show continuous motor unit activity. Symptomatic treatment involves the use of GABA-stimulating medications, such as benzodiazepines and baclofen, to reduce rigidity and painful muscle spasms. Immunotherapy aims to reduce or remove the antibodies, usually through intravenous immunoglobulin. **Final comments:** SPS me is progressive and generally ensues varying levels of disability. Early diagnosis and treatment initiation significantly impact the patient's prognosis.

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Stiff person syndrome: case report of an atypical presentation of the classic form of the disease

Pâmela Ritzmann de Lima[1], Laís Cristina Marques Moura[1], Renan Augusto Campos Manoera[1], Fabio de Paula Conforto de Oliveira[1], Brenda Jahn Henning[1], Patrícia de Morais Della Justina[1], Isabella Flach Antonioli[2], Renan Pires Carneiro da Cunha[1], Chelin Auswaldt Steclan[3], Iuri Pereira dos Santos[1]

[1]Universidade do Contestado, Mafra SC, Brazil.
[2]Universidade do Vale do Taquari, Lajeado RS, Brazil.
[3]Universidade Federal de Santa Catarina, Florianópolis SC, Brazil.

Correspondence: Pâmela Ritzmann de Lima; Email: pamelaritzmann@gmail.com.

Case presentation: Female, white patient, started with loss of muscle strength in the lower limbs and recurrent falls at 48 years of age. Over time, the symptoms progressively worsened, with difficulty walking, imbalance, paresthesias and painful muscle spasms in the lower limbs. At the age of 55, he sought medical help for the first time, due to a marked impairment in his quality of life. He consulted with several specialists, having normal brain imaging and electroneuromyography exams, and underwent countless symptomatic treatments with analgesics and anti-inflammatories and even with anxiolytics and

antidepressants, for associated anxiety symptoms, without getting a response. The diagnosis was only confirmed at the age of 56, in the face of an intense symptomatic crisis, after referral to a neurology service and extensive investigation, with normal imaging tests and positive anti-glutamic acid decarboxylase (anti-GAD) serum (> 2,000 IU/ml). Partial improvement was achieved with symptomatological treatment, using neuromodulators (carbamazepine, bacoflene, pregabalin and diazepam) and immunotherapy (IV immunoglobulin), although they were ineffective in controlling clinical worsening and preventing crises. Therefore, at the age of 63, the patient also started to present paresthesia in the thoracolumbar spine and upper limbs. **Discussion:** Stiff-person syndrome (SPS) is a rare autoimmune disease that affects one to two people in every million, causing muscle stiffness, spasms and chronic pain. The pathophysiology is not yet defined, but it is known that it is related to the production of anti-GAD against the synthesis of gamma-aminobutyric acid (GABA), resulting in disturbance of the inhibitory pathways. This case is classified as classic SPS, in which there is positive anti-GAD in the serum. However, this is a classic atypical presentation, as the tendency is for symptoms to begin in the trunk and, later, spread to proximal and distal limbs, and the opposite occurred here. In addition to this and the fact that SPS is a rare disease, constituting a diagnosis of exclusion, the patient also had associated psychiatric symptoms that served as confusing factors, resulting in a late diagnosis. As for treatment, immunotherapies were not enough to reverse the symptomatology. The clinical worsening continued, culminating in the patient's neurocognitive impairment and immobility. **Final comments:** This case discusses an atypical evolution of SPS, which made diagnosis even more difficult, and the ineffectiveness of the therapies adopted, reinforcing that early diagnosis is one of the biggest challenges of this condition, as well as controlling crises and disease progression through available treatments.

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Stroke and hemogram: warning signs in blood tests

Santhiago Calvelo Graça[1], Tainá Mosca[1], Vivian Dias Baptista Gagliardi[2], Wilma Carvalho Neves Forte[1], Rubens José Gagliardi[2]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Santhiago Calvelo Graça; Email: santhiagocalvelo@hotmail.com.

Background: Stroke is a significant cause of death and disability worldwide, with its prognosis closely tied to the severity of this acute condition. This condition is linked to an inflammatory process in the central nervous system, involving various types of cells such as monocytes, lymphocytes, and neutrophils. The inflammatory process can be reflected in peripheral blood through the hemogram, test frequently used in urgent situations such as this vascular disease. Research indicates that relationships derived from the peripheral blood cell count can serve as valuable markers of various inflammatory processes. **Objective:** To investigate the relationship between the severity of stroke and inflammatory markers derived from the peripheral blood cell count. **Methods:** It is a retrospective study based on medical records of patients aged over 18 years, who had ischemic stroke between 2017 and 2022. The clinical data includes the severity of the ischemic stroke (according to the National Institutes of Health Stroke Scale (NIHSS)) and the peripheral blood cells values from the patients' medical records. Based on these last values, the following relationships were established: systemic immune inflammation (SII) index (neutrophils multiplied by platelets and divided by lymphocytes), neutrophil-lymphocyte ratio (NLR), monocyte-lymphocyte ratio (MLR) and platelets-lymphocyte ratio (PLR). NIHSS scores were categorized into the following ranges: ≤ 5, 6-10, 11-15, and > 15. **Results:** Data from 194 stroke patients were collected. It was observed that patients with more severe conditions, as measured by the NIHSS scale, exhibited higher SII, NLR, MLR, and PLR values within the first 72 hours post-stroke ($p \leq 0,05$). There was a statistically significant difference between all groups across all ratios (≤5, 6-10, 11-15 and > 15), apart from the comparison between the 11-15 and > 15 groups. **Conclusion:** Stroke patients manifest an inflammatory process within the CNS, which can be evidenced through peripheral blood markers such as SII, NLR, MLR, and PLR. These relationships are correlated with the severity of the condition according to the NIHSS. Given the complexity of the SII ratio and the fact that all ratios convey similar information, it is advisable to utilize only one of the other three ratios, preferably the one that is the easiest to calculate for the patient in question, thereby obviating the need to determine all others.

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Stroke due to neurosarcoidosis: a case report

Roberto Pereira Santos[1], Cícero Cunha Lima[1], Filipi Brito[1], Ingrid Pereira Marques[1], Matheus Jorand Cecatto[1], Celmir Vilaça[1], Carolina Rouanet[1], Gustavo Honório[1], Marcos Martins da Silva[1], Marco Antonio Sales Dantas de Lima[1]

[1]Universidade Federal do Rio de Janeiro, Faculdade de Medicina, Hospital Universitário Clementino Fraga Filho, Serviço de Neurologia, Rio de Janeiro RJ, Brazil.

Correspondence: Roberto Pereira Santos; Email: r.santos.ufrj@gmail.com.

Neurosarcoidosis represents a rare systemic inflammatory condition characterized by the formation of non-caseating granulomas in various organs, including the central nervous system (CNS). Its clinical presentation spans a spectrum of neurological disturbances, posing diagnostic challenges due to its heterogeneous manifestations. Among its phenotypic variants, vascular neurosarcoidosis stands as an uncommon form, fraught with the potential for severe ischemic complications, notably stroke. Herein, we report a case of vascular neurosarcoidosis complicating with ischemic stroke, underscoring the diagnostic conundrums and the imperative of early recognition for the judicious management of these patients. **Case presentation:** A 72-year-old woman with a history of dyslipidemia and hypertension presented to the emergency department of Clementino Fraga Filho University Hospital with a 2-week history of left upper limb paresis and sudden dysarthria onset 2 days prior. Clinical and imaging evaluations unveiled suggestive signs of vascular neurosarcoidosis, encompassing pre-nuclear facial paresis, horizontal gaze nystagmus, positive Romberg sign, and magnetic resonance imaging (MRI) findings consistent with multifocal cerebral ischemia. The patient also bore antecedents of prior acute neurological episodes, aligning with the recurrent course of the disease. Further investigation confirmed perivascular contrast enhancement on dark blood imaging, corroborating the diagnosis of inflammatory vasculopathy concomitant with neurosarcoidosis. **Discussion:** This case epitomizes the diagnostic intricacies inherent to vascular neurosarcoidosis, particularly when manifesting as acute ischemic events. The presence of clinically suggestive signs such as pre-nuclear facial paresis and nystagmus, coupled with typical imaging findings, proved pivotal in the differential diagnosis between inflammatory and atheromatous etiologies of stroke. Perivascular contrast enhancement on dark blood imaging furnished additional evidence, fortifying the hypothesis of inflammatory vasculopathy. This case underscores the imperative of multidisciplinary assessment and consideration of uncommon systemic maladies in patients presenting with atypical neurological phenotypes. **Final comments:** Vascular neurosarcoidosis represents a rare yet potentially grave complication culminating in acute cerebral ischemic events. This case report accentuates the significance of early clinical suspicion and meticulous differential diagnosis to ensure optimal management of such patients. The identification of distinctive clinical and radiological features, coupled with validation through advanced imaging modalities like perivascular contrast enhancement on dark blood imaging, is indispensable in directing therapy and optimizing clinical outcomes.

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Stroke epidemiology trends in Brazil: a decade analysis

Wilson Falco Neto[1], Christian Ken Fukunaga[2], Raiza Brito Cipriano[3], Júlia Gon Perez Nardoque[1], Laura Alonso Matheus Montouro[1]

[1]Faculdade de Medicina de Catanduva, Catanduva SP, Brazil.
[2]Centro Universitário Faculdade de Medicina do ABC, Santo André SP, Brazil.
[3]Escola Superior de Ciências da Santa Casa de Misericórdia, Vitória ES, Brazil.

Correspondence: Wilson Falco Neto; Email: wilson.academic.mail@gmail.com.

Background: Despite advances in the treatment and prevention of strokes, the condition still has a high morbidity and mortality rate and is a major public health problem. Understanding the incidence and mortality of this condition is fundamental for public health. **Objective:** Our study therefore assessed trends in the distribution, demographics and evolution of strokes in Brazil. **Methods:** This is an ecological study. The data on hospitalizations was extracted from the DATASUS platform, with the period selected being 2010-2019 and the origin of the patients accounted for according to their place of origin. Only hospitalizations related to the ICD codes "Transient Stroke" and "Stroke of unidentified cause". Population data was obtained according to the IBGE census. Since the census does not take place every year, a projection based on the 2010 census was used. Geospatial analysis was carried out using geoda software in order to obtain the spatial autocorrelation coefficient and inferential statistics were carried out using R studio software. The age groups were transformed into numbers (1-16) and a simple linear regression was carried out, with confidence intervals for the coefficient obtained using bootstrap. Furthermore, simple linear regressions were also performed to observe the annual evolution of hospitalizations, deaths and mortalities. **Results:** A total of 1,644,590 hospitalizations occurred during the period, with an average hospitalization time of 7.4 days, a total cost of R$1,977,622,972.08, an average cost of 1,202.50 per hospitalization and a total of 250,196 deaths, giving a mortality rate of 15.21. Regarding the epidemiological profile, there were more men (51.81%) than women (48.18%). The main age group was 80+ years (20.87%) and the main race was white (48.52%). A simple linear regression analysis was carried out to investigate the extent to which the age group explained the number of hospitalizations. A statistically significant influence was found ($F(1.16)=88.43$, $p<0.001$; adjusted $R2=0.83$). The regression coefficient B (B=18102.73 , 95% [CI=14165.69 - 24222.12]) indicated that, on average, there was an increase of 18102 hospitalizations for each age group. Spatial autocorrelation was observed when analyzing the incidence of deaths ($I^2=0.439$; $p<0.01$) and hospitalizations ($I^2=0.363$; $p<0.01$). The evolution of hospitalizations showed a linear pattern (($F(1.8)=105.5$, $p<0.001$; adjusted $R2=0.92$), where there was an increase of 3457.921 each year (B=3457.921, 95% [CI=271.903 - 4089.415]). **Conclusion:** Our study observed a growing trend in the number of deaths and hospitalizations, which is in line with that observed in other countries, but differs in terms of epidemiological profile. Investments in structure and training for strokes care are essential for public health.

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Stroke in a young transgender patient using estrogen therapy: a case report

Andressa Camilo Oliveira[1], Lucas Blanco Insaurralde da Luz Silva[1], Amanda Boutrik[1], Anny Gabriela Marçal de Carvalho[1], Isabela de Carvalho Florêncio[2], Renata Gratão Rezende[2], Rodolfo Antonio Corona[2], Lucas Luiz Fitipaldi Ferreira[3], Maria Luiza Ferri Cury[2], Gabriel Pereira Braga[2]

[1]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.
[2]Hospital Universitário Maria Aparecida Pedrossian, Campo Grande MS, Brazil.
[3]Santa Casa de Misericórdia de Campo Grande, Campo Grande MS, Brazil.

Correspondence: Lucas Blanco Insaurralde; Email: lucasblancoms@gmail.com.

Case presentation: A 30-year-old transgender woman, using estrogen hormone therapy without medical guidance since the age of 17 (cyproterone 2mg + ethinylestradiol 0.035mg 1/day), HIV positive (viral load on December 4, 2023, of 413, and CD4 count on October 3, 2023, of 413) with irregular treatment, and previously treated for syphilis. One day before hospitalization, patient presented with a moderate intensity holocranial headache. In the next day, she woke up with sudden-onset right-sided hemiparesis and associated homonymous hemianopsia. In the admission, her vital signs were blood pressure 130/80 mmHg, heart rate 104 BPM, and oxygen saturation 99%. She underwent an infectious screening and cerebrospinal fluid analysis, which excluded opportunistic neuroinfections. The initial computed tomography scan showed no abnormalities, but magnetic resonance imaging revealed signs of ischemia in the left posterior cerebral artery territory. An etiological investigation for stroke was conducted with transthoracic and transesophageal echocardiograms, carotid and vertebral Doppler ultrasound, electrocardiogram, Holter monitoring, arteriography, rheumatological tests, and thrombophilia workup, all of which were unremarkable. Therefore, the final etiological diagnosis was concluded as TOAST classification other causes due to improper estrogen therapy. After diagnosis, she was discharged on antiplatelet therapy with aspirin 100mg daily, simvastatin 20mg daily, and fluoxetine 20mg daily, with hormone supplementation suspended, and referred to a transgender care clinic and vascular neurology outpatient services. **Discussion:** Ischemic stroke in young adults comprises approximately 10% to 12% of all stroke cases. Among young adults, etiologies such as infections, drug abuse, heart diseases, genetic conditions, and autoimmune diseases must be considered. In the present case, the patient had been on irregular estrogen therapy without medical supervision, and other possible etiologies were excluded. The use of estrogen therapy has been discussed in the scientific community, with evidence indicating increased cardiovascular risk at various levels. This is manifested by increased cases of venous thromboembolism, elevated blood pressure, and acute myocardial infarction. Therefore, the potential for hormone therapy to also cause cerebrovascular diseases is noted, especially after prolonged exposure, which likely led to the pathology presented in this case. The magnitude of this effect in transgender individuals undergoing hormone supplementation remains uncertain. **Final comments:** This case highlights the increased risk of cerebrovascular events in the transgender population with inadequate hormone therapy. Therefore, with the diffusion of services aimed at the transgender population and access to hormone therapy, it has become essential to expand studies on the risk and prevention of cerebrovascular diseases associated with hormone therapy use.

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Stroke in young adults in Brazil: analysis of hospital morbidity in the SUS in the pre and post COVID-19 pandemic periods

Henrique Chiarini Batistella[1], Heloísa Chiarini[2], Natalia Alini Haubenthal[2], Gisandra de Fátima Stangherlin[2], Luiza de Gregori Dutra[2], Clândio Timm Marques[2], Lucas Henrique Picur Tavares da Silva[2], Natalia Lançanova da Silveira[2], Kelly de Oliveira Harada[2]

[1]Pontifícia Universidade Católica de Campinas, Campinas SP, Brazil.
[2]Universidade Franciscana, Santa Maria RS, Brazil.

Correspondence: Henrique Chiarini Batistella; Email: hcbatistella@gmail.com.

Background: Stroke stands as the second leading cause of death and the biggest cause of permanent disability in the world and can affect individuals at any age, being uncommon before the age of 40. Studies have shown an increase in the incidence of stroke following the COVID-19 pandemic. **Objective:** To analyze selected aspects of hospital morbidity due to stroke from January 2014 to December 2023. **Methods:** retrospective, cross-sectional ecological study. Data was obtained from the Information Technology Department of the Unified Health System. The population consisted of patients aged between 20 and 49, hospitalized with a diagnosis of stroke (ICD-10 I61, I63, I64, I67 and G45), from January 2014 to December 2023. We considered the period 2014-2019 as pre-pandemic COVID-19 and the period 2020-2023 as post-pandemic. The variables of interest: age group, gender, hospitalization period, ICD-10 and number of deaths. The normality of the variables to be studied was verified using the Shapiro-Wilk test and the t-test for independent data. Differences were significant when $p<0.05$. IBM SPSS Version 25 software was used for statistical analysis. **Results:** The study revealed a total of 350,161 hospitalizations for stroke among individuals aged 20 to 49 years from 2014 to 2023, with 21.8% (76,303) classified as CID-10 I61, 8.3% (29,078) as I63, 53.5% (187,152) as I64, 9.1% (32,018) as I67, and 7.3% (25,610) as G45. Regarding age groups, 11.7% (40,864) were aged 20-29 years, 26% (91,081) were aged 30-39 years, and 62.3% (218,216) were aged 40-49 years. When analyzing the total number of stroke hospitalizations in the pre- and post-pandemic periods, a significant difference ($p=0.023$) was observed among young adults (20 to 49 years) comparing before (33569 ± 1234.3) and after (36452.5 ± 2060.2) the pandemic; the 40-49 years age group showed a significant increase ($p=0.012$) comparing 2014-2019 (20761.3 ± 824.4) to 2020-2023 (22948 ± 1332.6). Concerning sex, a significant increase in the number of stroke hospitalizations was noted in men ($p=0.012$) post-pandemic, particularly in the 30-39 years ($p=0.024$) and 40-49 years ($p=0.007$) age groups comparing pre (4140.2 ± 78.5 and 9873.7 ± 453.4, respectively) and post (4451.7 ± 264.2 and 11125.2 ± 728.2, respectively) pandemic periods. In women, a significant increase ($p=0.025$) was observed only in the 40-49 years age group comparing pre (10888.2 ± 458.6) and post (11822.5 ± 619.9) pandemic periods. Furthermore, a significant difference was found in the number of deaths in stroke hospitalizations in the 40-49 years age group ($p=0.019$). **Conclusion:** Despite the limitations of this study, such as the inability to correlate with risk factors, define the type of cerebrovascular disease and SARS-CoV-2 infection, the results demonstrate an increase in hospital morbidity due to stroke among young adults in the post-pandemic period, particularly in males and the 40 to 49 years age group.

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Stroke knowledge, clinical-epidemiological aspects, and odds of undergoing intravenous thrombolysis in patients with acute ischemic stroke

Bárbara Nascimento Moutinho[1], Mário Luciano de Mélo Silva Júnior[1], João Pedro Wanderley Soares[1], Mariana Leandro Silva[2], Karyne de Souza Nobre[3], Renata Maria de Melo Moraes[3], Teresa Virgínia Macedo de Aquino[3], Yasmin Gabrielle Pereira Pacheco Cavalcanti de Albuquerque[3], Ilo de Vasconcelos Ximenes Júnior[1], Bárbara Francisca do Nascimento[3]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil.
[2]Faculdade de Medicina de Olinda, Olinda PE, Brazil.
[3]Centro Universitário Maurício de Nassau, Aracaju SE, Brazil.

Correspondence: Bárbara Nascimento Moutinho; Email: barbara.moutinho@ufpe.br.

Background: Ischemic stroke is among the leading causes of death and disability around the globe. Early therapeutic intervention for reperfusion is the keystone to enhancing prognosis, but it is time-sensitive. Although guidelines have expanded the time window for intravenous thrombolysis, data suggest that the number of patients with acute ischemic stroke who undergo intravenous thrombolysis remains low, especially in underdeveloped regions. Then, improving comprehension of the factors contributing to undergoing intravenous thrombolysis is crucial. **Objective:** To evaluate the relation between clinical-epidemiological aspects of patients with acute ischemic stroke, prior knowledge about this disease, and receiving intravenous thrombolysis. **Methods:** Cross-sectional study enrolling patients with acute ischemic stroke admitted in a tertiary care center. Data collection was performed using an electronic form developed by our team and administered directly to the patients. The form consisted of objective questions regarding sociodemographic data, personal history, comorbidities, presguenting symptoms, time of symptom onset, time taken to reach the hospital, awareness of stroke warning signs and risk factors, and knowledge about the Mobile Emergency Service (SAMU) telephone number. We also obtained the status of intravenous thrombolysis. **Results:** The sample consisted of 245 patients with acute ischemic stroke, with a median age of 66 (IQR 57-74) years, with 53% being women. Of the total, 48 (19.5%) patients underwent intravenous thrombolysis. Regarding the time taken to reach the referral service, in minutes, the median in the thrombolysis group was 177.5 (IQR 120-210) vs. 240 (IQR 160-507) minutes among controls ($p<0.001$). Regarding the NIHSS, the median was 9 (IQR 6-12) in the intravenous thrombolysis group vs. 3 (IQR 1-8) points in patients who did not undergo it ($p=0.012$). As for patient characteristics, we have for the intravenous thrombolysis group and the control group, respectively: systemic arterial hypertension 71.4% vs. 84.9% ($p=0.069$); diabetes 25.6% vs. 44.7% ($p=0.031$); dyslipidemia 20.6% vs. 38.9% ($p=0.050$); coronary artery disease 2.8% vs. 15.1% ($p=0.053$); health insurance coverage 8.5% vs. 3.6% ($p=0.232$); symptom onset at work 11.4% vs. 3.5% ($p=0.070$); wake-up stroke 13.3% vs. 26% ($p=0.081$). Finally, the variables years of formal education, knowing the SAMU number, knowing at least one stroke risk factor, knowing the affected organ in stroke, and knowing at least one stroke sign or symptom did not differ between the groups. **Conclusion:** The absence of diabetes and dyslipidemia, initial NIHSS score, and a shorter time between symptom onset and hospital arrival were the factors associated with undergoing intravenous thrombolysis. General knowledge about stroke and other variables, however, did not show statistical significance.

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Stroke mimics profile in a tertiary hospital in Salvador-BA

Filipe Nolasco de Souza e Silva[1], Rayanderson Nunes da Gama[1], João Gabriel Teixeira Mota[1], Rubson Soares Rocha[1], Marcel Leal Ribeiro[1], Daniel Lordelo San Martin[1], Renan Carvalho Castello Branco[1], Karla Oliveira Couto[1], Pedro José da Silva Júnior[1]

[1]Hospital Santa Izabel, Salvador BA, Brazil.

Correspondence: Filipe Nolasco de Souza e Silva; Email: filipe.nolasco95@gmail.com.

Background: Stroke mimics are non-vascular disorders that clinically resemble acute stroke. Despite numerous studies in the literature about this topic, there remains limited information about this condition in Brazil. **Objective:** To describe the clinical and epidemiological profile of patients diagnosed with stroke mimics. Secondary objectives were to identify the most prevalent etiologies, management practices recommended by the neurology team, and to determine the outcomes of these patients. **Methods:** This was an observational, descriptive, retrospective study consisting of a case series of patients classified as stroke mimics at a tertiary hospital from August 2020 to August 2023. **Results:** A total of 441 patients were admitted, with 152 (34%) classified as stroke mimics. Among these, 99 were women with a mean age of 61.7 years and a median National Institute of Health Stroke Scale of 4 on admission. Of the patients, 110 (72.4%) had at least one risk factor, including hypertension in 100 patients, diabetes mellitus in 57, prior stroke in 24, coronary artery disease in 11, and atrial fibrillation in 9. The most prevalent etiology for stroke mimics was epileptic events, accounting for 29.6%. None of the stroke mimic patients underwent thrombolysis or thrombectomy, but 25 received single antiplatelet therapy and 30 received dual antiplatelet therapy upon admission. The median modified Rankin Scale at discharge was 0, with a mortality rate of 7.2%. **Conclusion:** Stroke mimics constitute a significant proportion of patients admitted with suspected stroke, with a prevalence higher than the average reported in the literature. Epileptic events were identified as the most prevalent etiology.

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Stroke mimics: assessing the most common differential diagnoses of stroke in the emergency department

Luiz Carlos Porcello Marrone, Victor Garcia Ortiz[1], João Pedro Farina Brunelli, William Alves Martins, Luciano Passamani Diogo, Laura Fuchs Bahlis, Leonardo Pisani, Matheus Bom Fraga, Antônio Carlos Huf Marrone

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Victor Alfonso Garcia Ortiz; Email: victorgarcia.or@gmail.com.

Background: Sudden onset of focal neurological deficits in a vascular distribution, such as hemiparesis, facial weakness, and aphasia, typically suggests a stroke. However, other disorders can mimic these clinical manifestations. “Stroke mimics” describe non-vascular disorders that present similarly to stroke, such as multiple sclerosis and facial palsy. Previous studies have found varying frequencies of stroke mimics among patients suspected of acute ischemic stroke, with younger patients and those with fewer stroke risk factors being more likely to have a stroke mimic diagnosis. **Objective:** To evaluate the prevalence and characteristics of stroke mimics in a neurologic emergency department at Hospital São Lucas in Porto Alegre. **Methods:** Over two years and six months, data were collected from 330 patients diagnosed with stroke mimics after being admitted to the emergency room. These patients were initially diagnosed with stroke but later excluded by a neurologist who confirmed other diagnoses. Our study evaluated sex, age, symptom onset, and final diagnosis. Two main groups were analyzed: those arriving within 4.5 hours of symptom onset and those arriving after 4.5 hours. Stroke mimics were categorized into 14 subtypes, including functional disorders, delirium, seizures, migraine, among others. **Results:** Of the 330 patients included, 50.3% were male, with a mean age of 67.1 years. Functional disorders (14.2%) and hydroelectrolytic imbalances (14.2%) were most found, followed by delirium (12.7%). Among patients admitted within 4.5 hours of symptom onset, 73.6% were older than 60, compared to 62.3% of those who arrived later. In the early presentation group, hydroelectrolytic imbalances and seizures were most common, while in the late presentation group, delirium and psychiatric disturbances were more frequent. **Conclusion:** Our study highlighted a variety of disorders presenting similarly to stroke, posing a risk of thrombolysis overtreatment by unprepared professionals. Although symptomatic intracranial hemorrhage in stroke mimics is rare, the correct use of thrombolysis is economically significant due to the high cost of these drugs. Misdiagnosis can also delay appropriate treatment for conditions like seizures and metabolic disorders. Our findings underscore the importance of recognizing stroke mimics in emergency settings. The most common stroke mimics differ between patients eligible for thrombolysis and those who are not, emphasizing the need for careful triage. This knowledge can guide clinicians in accurately diagnosing and treating stroke mimics, ultimately improving patient outcomes. In summary, awareness of the most common stroke mimics is crucial for emergency department clinicians, with functional disorders and hydroelectrolytic imbalances being the most frequent in this study.

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Stroke palliative care protocol implementation in a reference hospital for stroke treatment in Joinville, Santa Catarina

Maria Eduarda Angelo de Mendonça Fileti[1], Patrícia Vera[1], Lila Jerusa Nascimento Pereira Abreu[1], Marta Gabriele Pires da Rosa Schmen[1], Camilla Ramos Mohr[1], Jane Rossi[1], Brenner Henrique de Oliveira Marques da Silva[1], Joselma Siqueira de Aquino[1], Carla Heloísa Cabral Moro[1]

[1]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Maria Eduarda Angelo de Mendonça Fileti; Email: neuro.mef@gmail.com.

The concept of palliative care has awakened a new vision about the integrality of a person, with its physical, psychological, social and spiritual dimensions. This approach consists of assistance provided by a multidisciplinary team that aims to provide greater comfort to patients and their families and reduce expenses with measures that are disproportionate to the patient’s condition. Studies show difficulties in symptom management, communication, support family and decision-making in patients who present with acute stroke in hospital care. They also identified high case fatality after severe stroke, numerous long-term physical problems, substantial emotional distress, prognostic uncertainty, and a sense of abandonment after hospital discharge. Our experience in a reference center for stroke treatment has shown the need of Post-Stroke Palliative Care Protocol establishment. In 2019 a Protocol for Post-stroke Palliative Care was written, based on NIHSS, Hemphill, Fischer, Palliative Performance, Modified Rankin, Barthel and NECPAL scales. Patients who match the following criteria are eligible for the Protocol: NIHSS > or = 18 one week after the event; Palliative Performance Scale ≤ 30%; mRankin ≥ 4; and NECPAL CCOMS-ICO indicating Palliative Care. Because of the COVID-19 pandemics in 2020, we had difficulties in setting down the Protocol. So in 2023, we were finally able to put it into practice. We created a multidisciplinary committee composed by a neurologist, nurses, psychologist, social worker, occupational therapist, physiotherapist, nutritionist and speech therapist. Bedside visits are taken twice a week, for control and management of symptoms and treatment and multidimensional care planning. Family conferences for effective communication between the patient, health professionals, family members and caregivers are routine, so are conducting lectures for caregivers and family members about care. Promotion of continuous education to update the team’s knowledge is also a concern of ours. From June 2023 to June 2024, 81 patients were followed by our team. Concomitantly with the implementation of the palliative care protocol, a reduction in the need for prolonged hospitalizations was noticed, resulting in fewer emergency visits and readmissions, due to the information offered to family members and the patient himself about the real health condition and its perspectives; in addition to greater awareness among professionals involved in care and a complete demystification related to Palliative Care - avoiding aggressive interventions and unnecessary invasive procedures.

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Stroke presenting as bilateral hearing loss: a case report

Guilherme Bitencourt Barbosa[1], Renato Morais Bueno[1], Victor Ezaki Alves Silva[1], Rayanne Pereira Mendes[1], Yasmin Pagano Monteiro[1], Evelyn de Paula Pacheco[1], Miriam Eva Koch[1], Ruda Alessi[1], Eduardo Macedo de Souza Tieppo[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Guilherme Bitencourt Barbosa; Email: guibtencourt@gmail.com.

A 63-years-old male patient, with hypertension, admitted to the Emergency Room due to sudden bilateral hearing loss associated with occipital headache for one day. The patient was referred to an external hospital for evaluation with an otolaryngologist and imaging investigation. Despite brain magnetic resonance imaging (MRI) having no alterations (DWI was not included), MR Angiography of the brain revealed a basilar artery subocclusion in the middle segment. The patient refused hospital admission, but hours later sought a Neurology Emergency Department due to persistent hearing loss for 3 days. His physical examination was remarkable for right facial hypoesthesia, absent right vestibulo-ocular reflex (VOR), hypoacusis, broad-based gait, unable to tolerate Tandem, and a positive Romberg sign with a tendency to fall to the right. A brain CT on admission showed no abnormalities. A new brain MRI was requested due to posterior fossa stroke hypothesis, but his level of consciousness deteriorated rapidly, showing signs of decerebration and Cheyne-Stokes pattern hyperventilation. On the new physical examination, the patient showed no response in both VOR, absent bilateral corneal reflex, and absence of horizontal and vertical gaze. Due to instability to perform MRI, a new brain CT was done instead, showing hypodensity in the cerebellum and right cerebellar peduncle, territory of the anterior inferior cerebellar artery. The patient died due to stroke complications. Commonly, vertebrobasilar territory stroke manifests with prodromal symptoms, with the most characteristic being vertigo and headache. There are less common symptoms including hearing loss, as in the case. These symptoms can start days to weeks before the vascular event and pose a significant challenge for stroke diagnosis, as they are non-characteristic stroke symptoms and are present in other pathologies. Finally, with obstruction of the basilar artery, various signs and symptoms of basilar syndromes can occur, ranging from cortical blindness to decreased level of consciousness and altered breathing patterns. It is a serious and potentially fatal condition, as observed in the reported case. Although rare, hearing loss can be a symptom of a stroke, and should be considered, especially when there are simultaneous focal deficits and a sudden onset. Posterior fossa ischemia is hardly visualized on a brain CT - which is the first and sometimes only available exam in several Brazilian centers. Symptoms of posterior circulation involvement tend to be overlooked, either due to low specificity, absence of typical stroke manifestations, slower progressions and lower CT sensitivity. A detailed neurologic examination by a skilled professional can differentiate the early basilar artery symptoms from a peripheric disease and initiate a precocious treatment to prevent a poor prognosis.

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Stroke prevention by combining medical education in the context of primary health care and social media: an extension project

Lorenzo Marinho Morais[1], Chiara Gübel Portugal[1], Gabriel Macedo Cavalcante[1], Júlia Lima Vasconcelos[1], Matheus Lima de Oliveira[1], Gustavo Paes de Andrade Saraiva[1]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Lorenzo Marinho Morais; Email: lorenzo.morais@aluno.uece.br.

Background: Stroke is the second leading cause of death worldwide and a major cause of disabilities. Today, it is known that five risk factors account for more than 80% of the global risk for developing a stroke, whether ischemic or intracerebral hemorrhage: hypertension, smoking, obesity, sedentary lifestyle, and diet. Despite this, multicenter studies have shown that most people cannot define what a stroke is, nor can they recognize and respond to a probable episode. In light of this, the Neurology and Neurosurgery League of the State University of Ceará (Neuruece) created an extension project aimed at educating the population about strokes by visiting a primary health care unit to provide valuable information on the subject and also using social media. **Objective:** To educate the general population about the various characteristics of stroke, from understanding the risk factors to recognizing a probable episode. **Methods:** Educational activities were carried out by six medical students at a Primary Health Care Unit in Fortaleza. The unit was chosen due to the high number of users with stroke risk factors. Three actions were conducted throughout the year, using informative materials and a small questionnaire to assess people's level of knowledge on the subject. Regarding social media, several educational posts were made throughout the year on the Neuruece Instagram account. **Results:** The actions reached hundreds of people at the health unit throughout the year. The main topics taught to the population were: definition and types, differentiating ischemic and hemorrhagic stroke; risk factors; main symptoms; and the Brazilian mnemonic SAMU ("Sorriso", "Abraço", "Música" and "Urgência"), a quick and easy method to identify a probable stroke and how to act. Regarding social media, twelve posts were made throughout the year, covering topics from basic to more technical specifics, expanding the project's reach to medical students seeking relevant and evidence-based information. A total of 4669 accounts were reached, averaging 389.1 accounts reached per post. **Conclusion:** The project's relevance was evident, as it proved the lack of knowledge among most users involved regarding basic stroke topics. By disseminating this information, new stroke cases can be prevented, as the vast majority are triggered by modifiable risk factors. This type of project highlights to health authorities the positive impact that medical education can bring to the population, influencing the creation of public policies.

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Stroke profile of patients with high risk of obstructive sleep apnea: a transversal study

Aurea Maria Lago Novais Castello Branco[1], Juliana Cerqueira Gomes[1], Marina Soledad Benitez Bitencourt[1], Thaise Braga de Oliveira[1], Renan Carvalho Castello Branco[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Hospital Municipal de Salvador, Salvador BA, Brazil.

Correspondence: Renan Carvalho Castello Branco; Email: castellorenan@yahoo.com.br.

Obstructive sleep apnea (OSA) is a disease of upper airways, diagnosed with polisomnography, that is well associated with stroke throught diverse mechanisms. Despite that, it still lacks studies about stroke profile in patients with high risk of OSA. This study aims to describe the diferences between stroke in patients with high and low risk of OSA. We performed a transversal study, part of a cohort of stroke patients. Patients were evaluated of risk of OSA with STOP-Bang and SOS scores; also, data of wake-up stroke, stroke type and etiology, transient ischemic attack (TIA), thrombolysis, circulation evolved, ASPECTS (Alberta stroke program early computed tomography score), PC-ASPECTS (posterior circulation - acute stroke prognosis early computed tomography score), hemorragic transformation and its type (ECASS), ABCD2, National Institute of Health Stroke Scale (NIHSS) and death during internment were colectted. We obtained 430 patients, 50.5% female. 69% were classified as high risk with STOP-Bang, median 3 (interquartile range 2-4) and 50% with SOS score, median 10 (interquartile range 7-16). Considering STOP-Bang, there were significant diferences with previous stroke - most prevalent in high risk - (p=0.01) and admitional dyastolic pressure (p<0.001). Considering SOS, we obtained admitional capillary blood glucose (p=0.005), admitional dyastolic pressure (p=0.03). ASPECTS > 7 (p=0.02). In both scores, high risk patients evolved more to death during internment (p=0.04 and p=0.01, respectively). In baseline characteristics of STOP-Bang, there was a higher prevalence in high risk group of male sex (p<0.001), alcoholism (p=0.01), obesity (p=0.01), hypertension (p<0.001) and diabetes (p=0.002); of SOS score, female sex (p=0.007), obesity (p<0.001), hypertension (p=0.001) and diabetes (p=0.001). The stroke profile of patients with high risk of obstructive sleep apnea appears to follow the pattern of stroke in general: a higher prevalence of ischemic type and similar rates of hemorragic transformation. Otherwise, high risk of OSA patients have more recurrence of cerebrovascular event and tend to be admitted with higher measures of capillary blood glucose, dyastolic pressure and worst images, which is concordant with the information of higher prevalence of hypertension and diabetes. This phenomenon also can be explained due to metabolic pattern of this diasease, in addition to vasoreactivity impairment, which is demonstrated most with dyastolic measures.

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Stroke rehabilitation after hospital discharge is a gap that needs to be addressed in Brazil: highlights from a multicenter area study

Roberta de Oliveira Cacho[1,2], Carla Heloisa Cabral Moro[3], Suzete Nascimento Farias da Guarda[4], Elen Beatriz Pinto[5], Gustavo José Luvizutto[6], Suellen Mary Marinho dos Santos Andrade[7], Kelson James Almeida[8,9], Rodrigo Bazan[10], Gabriel Pereira Braga[11], Adriana Bastos Conforto[2]

[1]Universidade Federal do Rio Grande do Norte, Santa Cruz RN, Brazil.
[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.
[3]Hospital Municipal São José, Joinville SC, Brazil.
[4]Universidade Federal da Bahia, Salvador BA, Brazil.
[5]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[6]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.
[7]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[8]Universidade Federal do Piauí, Teresina PI, Brazil.
[9]Centro Universitário Facid Wyden, Teresina PI, Brazil.
[10]Universidade Estadual Paulista, Botucatu SP, Brazil.
[11]Universidade Federal de Mato Grosso do Sul, Campo Grande MS, Brazil.

Correspondence: Roberta de Oliveira Cacho; Email: ro_fisio1@hotmail.com.

There is limited information about stroke rehabilitation in low- and middle-income countries. This multicenter study investigated the access to rehabilitation for post-stroke patients in the first 6 months after discharge from public hospitals in Brazil and compared these outcomes between regions stratified by Human Development Indexes (HDI). Sixteen centers in 11 Brazilian states were grouped in Group HDIHigher (>0.750, n=225, South and Southeastern regions) and Group HDILower (≤0.750, n=248, Western and Northeastern regions). A survey regarding referrals to, assessment and treatment by rehabilitation professionals (physiotherapist-PT, speech therapist-ST, occupational therapist-OT, physiatrist) was answered by patients/caregivers. The primary outcome was the percentage of patients who were assessed at least once by a PT within the first six months post-discharge from acute care. Comparisons between groups were performed with Student´s t, Mann-Whitney, Chi-square or Fisher´s exact tests. Multivariate stepwise logistic regression models were planned, with assessment at least once by a PT as the dependent variable. Participants were recruited from January 2020, until October 2023. Participants in Group HDILower were significantly younger (p=0.018), had more hemorrhagic events (p<0.001), and higher NIH Stroke Scale (NIHSS) scores (p<0.001) than patients in Group HDIHigher. Referrals to at least one professional upon discharge from the acute stroke care facility were restricted to 54.5% patients. Referrals for PT were significantly more frequent in Group HDILower (p=0.001) than in Group HDIHigher. The participants referred to PT (p<0.001), ST (p<0.001), OT (p<0.001), were more likely to be assessed at least once, than patients not referred. Only 54.1% of the patients were assessed at least once by a PT, and less than 25%, by other rehabilitation professionals, with no significant between-group differences. The overall rate of PT treatment was higher (42.5%) than rates of treatments by other professionals (range, 1.5% to 17%), with no significant between-group difference. Only 5.1% were assessed by PT, ST, and OT, with no significant between-group difference. Age, gender and timing of data collection were not significantly associated with PT assessment and were removed from the regression model. Education ≤4 years was independently associated (p=0.001 CI 1.29-2.90), while higher NIHSS scores were inversely associated (p<0.001; CI 0.82-0.90) with lack of PT assessment. The AReA study provides objective, novel data about the gaps in stroke rehabilitation in Brazil, to guide effective public policies. AReA study group: Cesar Minelli, Luciane Souza, Luana Miranda, Luana Amaral, Gisela Tinone, Beatriz Neves, Renata Jucá, Maria Elisa Piemonte, Tatiana Ribeiro, Ana Paula Fontana, Lenise Valler, Octávio Pontes-Neto, Millene Camilo, Marco Tulio Pedatella, Isabel Lima.

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Stroke secondary to thyrotoxic crisis: case report

Rachel Leirner Argelazi[1], Maria Clara Motta Barbosa Valente[2], Santhiago Calvelo Graça[1], Filipe Brito Ferraz da Silveira[2], Camila Lopes Figueiredo[2], Rafael Chaves Claudino de Queiroga[2], Letícia Machado Dumont[2], Cícero Rodrigues Veloso[2], Vivian Dias Baptista Gagliardi[2], Rubens Jose Gagliardi[2]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil./

Correspondence: Rachel Leirner Argelazi; Email: racheleirner@gmail.com.

Case presentation: A 44-year-old female patient presented to the emergency department with a history of left hemiparesis and hypoesthesia for the past 10 days. She denied any comorbidities and upon admission was disoriented in time and space, fluctuating between agitation and lethargy. She was tachycardic, hypertensive, and hyperthermic, with exophthalmos. Admission cranial computed tomography (CT) revealed hypodensity in the right occipital and frontal regions. Further investigations

showed free T4 >12 (reference range) and TSH < 0.0008 (reference range). Electrocardiogram showed evidence of sinus tachycardia, and the Burch Wartofsky index was 50 points, defining the diagnosis of thyrotoxic crisis. Despite clinical measures for thyrotoxic crisis, the patient deteriorated with decreased level of consciousness and required orotracheal intubation. A repeat CT scan revealed multiple acute/subacute hypodense areas, as well as central venous thrombosis of the right transverse and sigmoid sinuses. Atrial fibrillation was incidentally detected during the course of intensive care unit (ICU) admission. Despite this, imaging findings suggested venous infarction secondary to thrombosis, and anticoagulation was continued in this context. Despite optimized medical therapy, the patient died on the eleventh day of hospitalization. **Discussion:** Several mechanisms are related to vascular events in the context of hyperthyroidism, including both cardioembolic events precipitated by atrial fibrillation and an increased risk of thrombotic events due to alterations in coagulation factors, such as increased factor VIII and the risk of antiphospholipid antibody syndrome. The patient in the reported case experienced venous infarction secondary to sagittal sinus thrombosis, which was the cause of the stroke. **Final comments:** Systemic conditions should be considered as risk factors for cerebrovascular events; early recognition and rapid intervention significantly influence outcomes. Furthermore, there is a need for better elucidation of thromboembolic and cardioembolic aspects related to hyperthyroidism, as well as the use of anticoagulation as secondary prophylaxis for stroke in this patient group.

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Structural integrity analysis of temporal lobe components and the thalamus in juvenile myoclonic epilepsy through T2 relaxometry

Carlos Dibbern[1], Ingrid Carolina da Silva Cardoso[1], Brunno Machado de Campos[1], Marina K. Alvim[1], Clarissa L. Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Campinas SP, Brazil.

Correspondence: Carlos Dibbern; Email: c222428@dac.unicamp.br.

Juvenile myoclonic epilepsy is the most common type of generalized epilepsy. A few studies have investigated functional and structural abnormalities of the brain in this form of epilepsy, indicating a possible role of temporal lobe structures and the thalamus on the onset and activity of juvenile myoclonic epilepsy; however, their structural integrity remains unclear. For that reason, here we assessed the structural integrity of the hippocampus, amygdala, temporal lobe pole, and thalamus in patients with juvenile myoclonic epilepsy through T2 relaxometry, a technique capable of detecting T2 signal irregularities (that correlate to tissue abnormalities) undetectable by conventional visual analysis based on MRI. We included 38 healthy controls and 40 patients diagnosed with juvenile myoclonic epilepsy without other neurological comorbidities, followed by the Epilepsy Service of Universidade Estadual de Campinas. All had a multi-echo T2 weighted magnetic resonance imaging with 3mm thickness acquired in a 3 Tesla Philips Achieva® MRI Scanner. We measured the T2 signal of the four structures in both groups through the software "Relaxo LNI" by manually drawing the regions of interest over the T2 relaxometry map in multiple slices to fully represent the components. We performed the statistical analysis using a bicaudate Student's t-test. At first, we compared the T2 signal of the structures between both brain hemispheres: the only structure that presented a significant asymmetry was the thalamus in the group of patients ($p<0.001$; 95% CI [-1.53; -0.53]). The control group and the other structures in the group of patients presented no T2 signal significant difference between hemispheres. In a second step, we compared the T2 signals between groups, which demonstrated an increased T2 signal of the right hippocampus ($p<0.001$; 95% CI [1.84; 4.35]), the left hippocampus ($p=0.009$; 95% CI [0.38; 2.63]) and the right amygdala ($p=0.001$; 95% CI [1.08; 4.57]) in the group of patients. We found no T2 signal differences in the other structures. We conclude that there is a microstructural integrity alteration detectable by T2 relaxometry in the hippocampus, amygdala, and thalamus in patients with juvenile myoclonic epilepsy, which may be relevant for the pathophysiology of this form of epilepsy and reinforces previous studies. However, the cause-effect relationships of these alterations with seizures or medications are unknown and require further investigation.

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Study of chronic meningitis at Clementino Fraga Filho University Hospital - UFRJ

Amanda Carvalho de Oliveira[1], Marco Antonio Sales Dantas de Lima[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Amanda Carvalho de Oliveira; Email: amanda-c16@hotmail.com.

Background: Chronic meningitis is defined as one where the duration of symptoms exceeds four weeks. Typically, patients present with symptoms such as headache, vomiting, and slowly progressive fever. Due to its extensive differential diagnoses and the need for careful analysis of complementary tests, the diagnostic and therapeutic management of chronic meningitis remains poorly explored, with limited studies in the literature. A better understanding of the epidemiology of chronic meningitis can aid in early diagnosis, optimizing treatments, and thereby reducing morbidity and mortality. **Objective:** Through an epidemiological analysis, we aim to create a database providing insights into the most prevalent etiologies, associated conditions, responses to different proposed treatments, and outcomes of cases. Our goal is to contribute to the literature, aiming to enhance protocols for diagnostic investigation and treatment. **Methods:** This is a retrospective observational study of chronic meningitis cases over the past decades at the Clementino Fraga Filho University Hospital, covering the period from 2001 to 2023. Data collection is guided by a questionnaire established by researchers, converted into a table format to accommodate information in a numerical list. Upon completion of data collection, statistical analysis will be performed, presenting descriptive data in numbers, percentages, means, and standard deviations. **Results:** We obtained a total of 1581 medical records, of which 524 have been evaluated, and 92 patients met the inclusion criteria for the study. Among the 92 evaluated patients, 64% of cases encompass the most frequent etiologies: Cryptococcosis and Tuberculosis, with approximately 76% associated with immunosuppressive conditions. Additionally, neoplastic conditions significantly contributed to the number of cases, representing about a quarter of the total. Meningeal syndrome was the initial manifestation in 70% of cases, and hydrocephalus and/or meningeal enhancement were the most common findings on imaging. Interestingly, the diagnosis of chronic meningitis was confirmed even in the absence of such data. Hospitalization duration varied with diverse outcomes. Using the modified Rankin scale at discharge, approximately 70% of patients left the hospital with sequelae, and around 20% of cases resulted in death. **Conclusion:** The study confirms the diverse etiology and clinical/radiological manifestations of chronic meningitis. Despite over two decades, treatment protocols have seen minimal changes. With the establishment of a comprehensive and meticulous database, we aim to identify crucial areas for modification in chronic meningitis protocols, ultimately improving outcomes for future patients.

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Study of neuropsychiatric symptoms as post-acute sequelae of COVID-19 during pregnancy

Leonardo José Rodrigues de Araújo Melo[1], Danielle Mesquita Torres[1], Luíza Madureira Silva[1], Marylane da Silva Viana[1], Danielle Macêdo Gaspar[1], Manoel Alves Sobreira Neto[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Leonardo José Rodrigues de Araújo Melo; Email: lleonardomello98@alu.ufc.br.

Background: The SARS-CoV-2 pandemic has had severe impacts on global mental health. In the acute phase of COVID-19, 36% of patients exhibit Neuropsychiatric Symptoms(NS) such as mood changes due to factors like systemic toxemia and metabolic alterations. NS are also common in the Post-Acute Sequelae of SARS-CoV-2 infection (PASC), such cognitive impairment, fatigue, and depression lasting more than six months, regardless of the infection severity. Studies show a higher prevalence of PASC in women, including pregnant women, who already experience cognitive and immunological impairment during pregnancy, known as "pregnancy brain fog", which is exacerbated by viral infections and psychological stress. **Objective:** To evaluate NS due PASC in women exposed to the SARS-CoV-2 virus during pregnancy. **Methods:** The research was conducted through a 12-month follow-up of women at the Maternity School Assis Chateaubriand (MEAC), in Fortaleza/CE, who contracted COVID-19 during pregnancy with long COVID symptoms (PASC-G). COVID-19 is confirmed by serological testing for the detection of anti-SARS-CoV-2 antibodies and/or a nasopharyngeal swab test. Participants underwent neuropsychological and sleep evaluation. Neurophysiological assessments of sleep was made by polysomnography. Neuropsychological evaluation included Mini-Mental State Examination (MMSE), Addenbrooke's Cognitive Examination (ACE), Beck Anxiety and Depression Inventories, Pittsburgh Sleep Quality Index and Epworth Sleepiness Scale. **Results:** Data were collected from 8 PASC-G patients, with an average age of 29.9 years (±5.54). Among these patients, only 2 had neuropsychiatric symptoms (insomnia) before COVID-19. Only one patient experienced severe COVID-19 symptoms, requiring hospitalization. 75% of the patients reported some symptom associated with anxiety disorder. MMSE the average score was 27.3 (±1.91), with only one patient scoring below the expected level for her education. Regarding the ACE, the average score was 81.9 (±8.64), with 4 patients scoring below 84 points, indicating cognitive disability. The average Pittsburgh Sleep Quality Index score was 15 (±4.28), with all participants showing poor sleep quality. The average Epworth Sleepiness Scale score was 6.75 (±3.01), with only one patient showing excessive sleepiness. The average scores for the Beck Anxiety Inventory and Beck Depression Inventory were 15.6 (±4.37) and 24.9 (±12.1), respectively, with significant anxiety symptoms in 4 patients.Two polysomnographies were performed, showing reduced sleep latency and changes in N2, N3, and REM sleep stages. **Conclusion:** It was found that neuropsychiatric symptoms are common in PASC-G patients. Therefore, especialized evaluation of these patients should be considered. The study is still ongoing and, due to the small number of patients, more research is needed to validate the hypothesis.

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Study of sympathetic small fibers using the skin wrinkle test in patients with amyotrophic lateral sclerosis

Thalita Aparecida dos Santos[1], Webert Alex dos Santos Benetti[1], Felipe Jorge Abdo[1], Paulo José Lorenzoni[1], Cláudia Suemi Kamoi Kay[1],

Renata Dal-Prá Ducci[1], Paula Raquel do Vale Pascoal Rodrigues[1], Rosana Herminia Scola[1], Otto Jesus Hernandez Fustes[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Otto Jesus Hernandez Fustes; Email: otto.fustes@hc.ufpr.br.

Background: Amyotrophic Lateral Sclerosis (ALS) is the most common form of Motor Neuron Disease. However, although the motor component changes in ALS are widely known, new research also indicates sensory impairment in this disease. As Weis et al. noted in 2011, 79% of patients with ALS diagnosed in their study had a reduction in the number of fine sensory fibers. In this sense, as a disease with sensory impairment, there are methods that can investigate this characteristic of ALS, of which the Skin Wrinkling Test (SWT) stands out. This can provide foundations to understand more deeply how the sensory component is related to ALS and allow substrate for new clinical correlations. **Objective:** To evaluate small sympathetic fibers using the SWT in patients with ALS. **Methods:** We performed a cross-sectional study on 22 individuals over 18 years old, diagnosed with ALS being monitored in a specialized outpatient clinic, who underwent SWT by immersion in warm water at 40°C (104°F) for 30 minutes. Currently, the parameters for evaluating wrinkling are based on the location and quantification of wrinkles produced by the test, which are evaluated on all fingers except the thumb, due to the difficulty of photographing it, so there are 3 possible classifications using the DATEMA scale: 0.0 = not wrinkled, 0.5 = minimally wrinkled and 1.0 = obviously wrinkled. Scores of 0.5 and 1.0 are considered as positive wrinkling, while 0.0 is considered negative. **Results:** 13 men and 9 women were evaluated, with a mean age of 58 and 63 years, respectively. 19 with sporadic ALS and 3 with the P56S mutation linked to ALS of the VAPB genetic type. The scale used for evaluation was DATEMA. Compared to a group of 30 patients without comorbidities, in which 6.4% of the sample tested negative (no visible wrinkles), 31.8% of the population with ALS tested negative, which revealed a statistically significant association between the negative test and the presence of a diagnosis of ALS by Fisher's exact test (p=0.027). **Conclusion:** small fibers controls skin wrinkling, which is present in the majority of the population. The absence of wrinkling may demonstrate abnormalities in the fine fibers. The fact that SWT has significantly more negative results in the population diagnosed with ALS demonstrates that this disease, historically known for its motor impairment, may also have sensory impairment involved in its pathophysiology. Further studies are needed to confirm these findings and to investigate the potential clinical implications of skin wrinkling assessment in ALS.

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Study of the clinical-demographic profile of patients with amyotrophic lateral sclerosis treated at a university hospital in the city of Rio de Janeiro

Rômulo Cerqueira Heckert dos Santos[1], Larissa Bittencourt de Carvalho[1], Lígia Rocha Andrade[1], Clarissa Neves Spitz[1], Salim Lázaro Balassiano[1], Robson Teixeira Vital[1], Luiz Claudio Santos Thuler[2], Márcia Maria Jardim Rodrigues[1]

[1]Universidade do Estado do Rio de Janeiro, Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Marcia Maria Jardim Rodrigues; Email: jardim.mm@gmail.com.

Background: Amyotrophic lateral sclerosis (ALS) is characterized by the progressive and inexorable neurodegeneration of upper and lower motoneurons. The study of ALS cases in a reference institution, such as a university hospital in the city of Rio de Janeiro, can enable the identification of epidemiological patterns in this population and support improvements in care. **Objective:** To characterize the clinical-epidemiological profile of patients with ALS treated at a university hospital in the city of Rio de Janeiro. **Methods:** The study was designed as an ambispective cohort. In the retrospective segment, data was collected from patients diagnosed with ALS from 12/13/2002 to 06/28/2023. In the prospective segment, new cases were incorporated into the final series between 06/29/2023 and 04/08/2024. The data comprised demographic, clinical and neurophysiological factors, which were statistically analyzed and compared to data published in other studies, nationally and internationally. **Results:** 84 cases of ALS were gathered, of which male patients predominated (57.1%), with an average age of first symptoms in the sixth decade of life (54.3 years), with the main initial motor symptom being impairment of one or both lower limbs (58.4%), with idiopathic etiology (91.7%), clinical phenotype of classic ALS (71.4%), systemic arterial hypertension as the main comorbidity (28.6%) and category El Escorial of defined ALS (48.8%). The time required to establish the diagnosis from the first symptom had a median of 19 months. Specific treatment with riluzole was performed in 59.5% of cases. Forty-five percent of patients had died by the end of the study, among whom an average interval between the first symptom of the disease and death was observed to be 35 months. **Conclusion:** All this compiled data is in line with data published in national and international bibliographic reviews on the disease to date.

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Study of the permeability of the blood-brain barrier and the intra-thecal inflammatory profile in patients with cerebral manifestations due to SARS-CoV-2

Rhuann Pontes dos Santos Silva[1], Lucas Coêlho Bernardo-Menezes[1], Elisa de Almeida Neves Azevedo[1], Marton Kaique de Andrade Cavalcante[2], Flávia Falcão Albuquerque[3], Maria Lúcia Brito Ferreira[3], Marta Rodrigues de Carvalho[4], Jurandy Junior Ferraz de Magalhães[2], Cristiane Campello Bresani-Salvi[1], Clarice Neuenschwander Lins de Morais[1]

[1]Fundação Oswaldo Cruz, Instituto Aggeu Magalhães, Laboratório de Virologia e Terapia Experimental, Recife PE, Brazil.
[2]Fundação Oswaldo Cruz, Instituto Aggeu Magalhães, Recife PE, Brazil.
[3]Hospital da Restauração, Recife PE, Brazil.
[4]Hospital de Base, Brasília DF, Brazil.

Correspondence: Marta Rodrigues de Carvalho; Email: martinhabhs@gmail.com.

Background: The severe acute respiratory syndrome coronavirus type 2 (SARS-CoV-2) has had effects on the central nervous system in cases of acute neurological manifestations (ANM). Although cytokines, chemokines and immunoglobulins were investigated in patients with acute brain syndromes and SARS-CoV-2 infection, data on the integrity of the blood-brain barrier (BBB) and intrathecal production of these biomarkers in these patients are still scarce. **Objective:** To investigate whether patients with acute brain syndromes after exposure to SARS-CoV-2 have inflammatory BBB dysfunction. **Methods:** Experimental study, part of the NeuroCOVID study, funded by Fiocruz (nº VPPCB-005-FIO-20-2-22). Samples from 39 patients admitted in the neurology service of Hospital da Restauração, in Recife, were analyzed. The case series was selected according to three comparison groups: Exposed cases (acute central neurological syndrome and exposure to SARS-CoV-2), exposed controls (peripheral neurological syndrome and with exposure to SARS-CoV-2) and unexposed controls (without neurological syndrome and without exposure). Exposure was defined as positivity in the molecular test for SARS-CoV-2 in swab or serology (IgG or IgA anti-SARS-CoV-2) in serum or CSF. The neurological syndromes comprised acute brain manifestations and were confirmed by clinical-radiological evaluation by neurologists at the service. Inflammatory cytokines and chemokines were measured using CBA in a flow cytometer. The evaluation of the BBB function was evaluated through the protein profile by gel electrophoresis, with calculation of the CSF:serum coefficients and the reibergrams. Clinical and laboratory data were processed by GraphPad, R and CSF Software Tool software. **Results:** Of the 39 patients, 16 exposed cases (mean 59 years; 33-82), 17 exposed controls (mean 49 years; 26-68), and 7 unexposed controls (mean 37 years; 28-53) were included. When compared with controls exposed and not exposed to SARS-CoV-2, patients with acute neurological syndromes exposed to the virus had significantly ($p<0.05$) higher values of IL-6, CCL2, IL-8 and CXCL9 in both serum and CSF; also showed values of CXCL10 only in serum and CXCL5 in CSF. In the BBB integrity study, there were no significant differences in the values of total proteins and fractions (albumin and IgG) of exposed cases when compared with the other groups. The "reibergrams" indicated the absence of total intrathecal IgG production in the three groups. **Conclusion:** Patients with ANM associated with subclinical SARS-CoV-2 infections did not show signs of BBB breakdown or intrathecal production of immunoglobulins. Despite not presenting systemic manifestations of COVID-19, these patients presented increased values in both serum and CSF of some cytokines involved in the systemic inflammatory response. In turn, the highest CXCL5 values only in the CSF suggest that it may have a role in the neurological manifestations in our sample.

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Subacute combined degeneration associated with severe hematological disorder: a case report

Raphaell de Souza Barbosa[1,2], Edson Junior da Silva Cordova[1,2], Mariana Martins Simoes[1,2], Guilherme Michelon[1,2], Thais Magalhães da Silva[3], Ana Luísa dos Santos Carregosa[3], Carlos Eduardo Marisco Ceretta[3], Eduarda Leitzke[3], Daniel Marchi Kieling[2], Daniel Lima Varela[4]

[1]Hospital de Clínicas de Passo Fundo, Passo Fundo RS, Brazil.
[2]Universidade Federal da Fronteira Sul, Passo Fundo RS, Brazil.
[3]Universidade de Passo Fundo, Passo Fundo RS, Brazil.
[4]Hospital de Clínicas de Passo Fundo, Serviço de Neurologia, Passo Fundo RS, Brazil.

Correspondence: Ana Luísa dos Santos Carregosa; Email: carregosaluisa@gmail.com.

Case presentation: A 42-year-old male, with a history of alcohol abuse and bedridden status, was admitted to the emergency department due to a two-month history of temporospatial disorientation with disorganized speech, which had worsened in the past 48 hours. The patient also exhibited anorexia and paraparesis, which had begun a year prior and progressively worsened over the past four months, eventually leading to an inability to walk. Physical examination revealed pale mucous membranes, afebrile status, spastic paraparesis, tactile and pain hypoesthesia in a "glove" distribution, and reduced vibratory sensation in the lower limbs. Initial supportive measures were provided, and laboratory tests showed pancytopenia associated with vitamin B12 deficiency (104.7 pg/mL). Cranial magnetic resonance imaging (MRI) revealed T2/FLAIR hyperintensity in the corticospinal tracts, while thoracic spinal MRI showed T2/FLAIR hyperintensity in the posterior columns of the spinal cord (inverted "V" sign). No abnormal gadolinium enhancement was observed.

Discussion: Subacute combined degeneration of the spinal cord is a neurological complication that affects neuronal myelin integrity due to vitamin B12 (cobalamin) deficiency, either from nutritional deficiency or inefficient absorption. While this condition is more prevalent in the elderly and often associated with pernicious anemia due to cobalamin malabsorption, it's crucial to consider it in alcoholic patients. Clinical manifestations include sensory deficits, paresthesia, weakness, and gait disturbances, which can progress to paraplegia. Laboratory analysis may reveal anemia, macrocytosis, leukopenia, and thrombocytopenia. Additionally, methylmalonic acidemia and homocysteine levels should be measured when characteristic findings are present but vitamin B12 levels are normal or borderline. Imaging studies should also be conducted to rule out other neurological causes; however, in patients with this condition, thoracic spinal MRI may reveal bilateral paired regions of hyperintensity in the dorsal portions of the spinal cord, forming the inverted "V" sign, and in critical cases, there's an increased likelihood of involvement of the lateral columns. Treatment involves cobalamin supplementation via intramuscular injections followed by an oral maintenance protocol. **Final comments:** Subacute combined degeneration is a rare condition typically associated with macrocytic anemia. However, it can also present with pancytopenia, an even rarer association, making it imperative to consider vitamin B12 deficiency in patients with hematologic abnormalities. Prognostic factors indicating a more favorable outcome include age under 50 years, a brief disease course, and the absence of sensory deficits. Moreover, prompt initiation of treatment can arrest disease progression and significantly reduce the risk of permanent neurological damage.

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Subacute paraneoplastic cerebellar ataxia syndrome in Hodgkin's lymphoma: a case report

Larissa Canela Ruiz Evangelista[1], Ana Beatriz Esteves Pires[1], Sarah de Souza Chinelato[1], Luiz Henrique Soares Santos Stefano[1]

[1]Faculdade de Medicina de Marília, Marília SP, Brazil.

Correspondence: Larissa Canela Ruiz Evangelista; Email: larissacanelaruiz30@gmail.com.

Case presentation: We report the case of a 47-year-old male patient diagnosed with Hodgkin lymphoma during evaluation and treatment of subacute cerebellar ataxia syndrome. In July 2021, the patient presented with vertigo, frequent headaches, bilateral visual blurring, slurred speech, dysphagia, and difficulty walking, rapidly evolved to oromandibular dystonia. Six months after that, he sought neurological care, with appendicular ataxia, inability to walk, dysdiadochokinesia, diplopia, photophobia, visual scotomas, and scanning speech. Laboratory and neuroimaging exams revealed signs of gliosis and cerebellar atrophy, suggesting a previous ischemic event. Cerebrospinal fluid analysis showed no significant alterations. The patient received human immunoglobulin, exhibiting partial symptom improvement, yet maintained significant ataxia associated with orthostatic incapacity. Three months after hospitalization, he developed painful cervical nodules, nocturnal fever and weight loss, which led to a diagnosis of advanced-stage classical Hodgkin's lymphoma, with enlarged cervical and axillary lymph nodes and lytic bone lesions in the spine. The palliative treatment was recommended due to the unfavorable prognosis and the impossibility of an emergency chemotherapy. **Discussion:** Paraneoplastic syndromes are rare complications of neoplasms outside the central nervous system, frequently preceding tumor detection. Subacute paraneoplastic cerebellar ataxia syndrome (SPCA) is characterized by cerebellar dysfunction, such as appendicular ataxia, vertigo, visual abnormalities, nystagmus, dysarthria, and scanning speech, with subacute onset and rapid progression to severe ataxia within less than a month. This condition is immunologically mediated, resulting in partial destruction of Purkinje cells, astrogliosis and cerebellar demyelination and the neoplasms associated with SPCA include ovarian, breast, lung carcinomas, and lymphomas. The diagnosis of SPCA involves excluding metastases and other neoplasms through laboratory exams, neuroimaging and identification of onconeural antibodies. The magnetic resonance imaging provides superior visualization of the cerebellum, and the laboratory tests include complete blood count, glucose, liver function tests, and cerebrospinal fluid analysis to rule out metabolic disorders and infections. Neurological complications in neoplasms may occur through direct or indirect immune-mediated mechanisms. Therefore, early identification and treatment of the primary neoplasm can reduce neuroimmunological symptoms; furthermore, aggressive immunotherapy, intravenous immunoglobulin, plasmapheresis, corticosteroids, and immunosuppressants are considered therapeutic options with variable response. **Final comments:** In this reported case, immunotherapy resulted in partial improvement of symptoms and, due to the advanced stage of Hodgkin's lymphoma, a palliative approach was adopted, preceding any investigation of onconeural antibody panels.

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Subacute spinal cord degeneration due to vitamin B12 deficiency in an alcoholic patient

Marcelo Simplicio Carvalho[1], Nathalye Fernanda Pedroso Dircksen[1], Felipe Leonardo Semensati[1], Fábio Gonçalves[1], Letícia de Barros Dias[1], Lorena Fernandes Kronbauer[1], Mônica Meneguette Silveira[1], Barbara Okabaiasse Luizeti[1], Damacio Ramon Kaimen Maciel[1], Mônica Marcos de Souza[1]

[1]Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Marcelo Simplicio Carvalho; Email: marcelo.neurolog@gmail.com.

Case presentation: Male patient, 42 year old, heavy drinker, was admitted to the emergency room, complaining of difficulty walking started 10 days ago, followed by paresis of the flexor muscles of the hands and paresthesia on the back of the feet, on neurological examination: areflexia of deep reflexes, sensitive ataxia, diffuse hypopalestesia. The patient denied fever, allergies and infectious prodromes. On admission, skull tomography, chest radiography and electrocardiogram tests showed no changes. Performed analysis of cerebrospinal fluid (CSF) revealed leukocytes 1, glucose 49, lactate 2.2, proteins 25.7, and absent fungi. Serological and laboratory tests showed no significant reactivity, except for herpes simplex IgG reagent and Epstein-Barr IgG positive. CBC and liver function were normal, with blood glucose and lipid profile within the expected limits. Vitamin B12 was in 213 pg/ML elevated homocysteine (33.90 µmol/L). MRI scans of the dorsal and cervical spine revealed diffuse prominence in the pachimeninge region and venous vascular structures of the cervical spinal canal, changes in the posterior aspect of the spinal cord compatible with subacute combined degeneration due to vitamin B12 deficiency. Patient was treated with Vitamin B12 replacement, evolving with neurological gains. **Discussion:** The patient presents a picture of subacute combined degeneration of the spinal cord due to vitamin B12 deficiency, associated with toxic-metabolic polyneuropathy, exacerbated by excessive alcohol consumption. Clinical and radiological findings are compatible with a subacute myelopathy, often related to vitamin B12 deficiency, which can lead to degeneration of the dorsal and lateral columns of the spinal cord, resulting in severe neurological symptoms such as paresthesia, paresis and loss of proprioception. Vitamin B12 deficiency is commonly associated with pernicious anemia and can be precipitated or exacerbated by chronic alcohol consumption, which interferes with the absorption of essential nutrients. Low vitamin B12 (213 pg/ML) and elevated homocysteine (33.90 µmol/L) reinforce the diagnosis. The absence of significant changes in other laboratory and imaging tests suggests that the main etiology of the patient's symptoms is related to vitamin B12 deficit. **Final comments:** The case highlights the importance of considering nutritional and toxic-metabolic causes in patients with progressive neurological symptoms and a history of substance abuse. Early identification and proper treatment of vitamin B12 deficiency is crucial to prevent permanent neurological damage. In this patient, vitamin B12 supplementation and cessation of alcohol consumption were essential for neurological recovery.

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Subarachnoid hemorrhage and socioeconomic indices in Brazil

Eliane Batista Silva[1], Samuel Oliveira de Amorim[1], Matheus da Silva Ferreira[1], Gabrielly da Silva Costa[1], Kelson Vinicius Dallabrida Padilha[1], Camila de Almeida Silva[1], Felipe Henrique Lima Pereira[1], Átila Barros Magalhães[1], Marcos Daniel Borges Melo[1], Marcos Manoel Honorato[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Eliane Batista Silva; Email: elianebs2712@gmail.com.

Background: Subarachnoid hemorrhage (SAH) is bleeding into the subarachnoid space of the brain, frequently caused by ruptured aneurysms, and may present with severe headache, neck stiffness, and loss of consciousness. SAH is a medical emergency that reflects social and economic inequalities, affecting different groups in disparate ways. Effective treatment depends on rapid diagnosis and a comprehensive understanding of the social determinants of health. **Objective:** To investigate the correlation between SAH mortality and socioeconomic indicators (SI) in Brazil from 2016 to 2021. **Methods:** This is a retrospective study with a quantitative approach, utilizing data from the Mortality Information System (SIM) of DATASUS, according to the International Classification of Diseases (ICD-10), and the Human Development Atlas (HDA) in Brazil, for the years 2016 and 2021. The average SAH mortality rate for each federative unit was calculated from the annual mortality rates (deaths due to SAH / resident population * 100,000). The human development index (HDI) and its sub-indices (education, longevity, and income) were analyzed. Through simple linear regression with a 95% confidence interval, the Pearson correlation coefficient (r) was obtained for the correlation between average mortality and socioeconomic indicators. **Results:** Statistically significant ($p<0.05$) and positive correlations ($r>0$) were observed between SAH mortality and the overall HDI (r=0.56), HDI-education (r=0.53), and HDI-longevity (r=0.42). However, the HDI-income index did not demonstrate a significant correlation. The state of São Paulo reported the highest average SAH mortality rate (3.42), whereas Acre exhibited the lowest average (1.19). These regional disparities highlight notable differences in SAH mortality rates across the states. **Conclusion:** As the level of human development increases, measured by access to healthcare, education, and income distribution, so does the impact of SAH on total deaths. This relationship is attributed to factors associated with regions with high human development, such as a large geriatric population, availability of diagnostic equipment, early screening, access to primary care, strict reporting practices, and conditions inherent to industrialization, as well as occupational exposure to carcinogenic agents, which may contribute to these findings.

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Subarachnoid hemorrhage with anterior wall ST segment depression (1-V4): a rare electrocardiographic change in acute brain disease

Amauri Pereira da Silva Filho[1], Danielle Cortellazzi Colonna Romano[2], Gabriela Yumi Nakamura[2], Melissa Fernandes Vilela de Freitas[2], Natália Rebeca Karpejany[3], Sabrina Borges Batista[2], Taianara Sampaio Reis[3]

[1]Hospital de Emergência e Trauma Dom Luiz Gonzaga Fernandes, Campina Grande PB, Brazil.
[2]Universidade Nove de Julho, São Paulo, São Paulo SP, Brazil.
[3]Universidade Federal do Piauí

Correspondence: Amauri Pereira da Silva Filho; Email: amauri.filho@yahoo.com.br.

Case presentation: A 45-year-old patient arrived at the emergency department with a sudden, severe headache, described as the worst of their life, accompanied by nausea, vomiting, and significant chest discomfort. Vital signs showed hypertension with a blood pressure of 190/120 mmHg and a heart rate of 130 bpm; the respiratory examination was normal. Neurological examination yielded a Glasgow Coma Scale of 15, isocoric and reactive pupils, no appendicular deficits, and mild nuchal rigidity (+1/+5). The patient underwent an immediate non-contrast CT scan which returned normal results. An ECG indicated ST-segment depression from V1 to V4, while routine laboratory tests were normal. Given the suspicion of subarachnoid hemorrhage and acute coronary syndrome, further investigations included a lumbar puncture and ultra-sensitive troponin measurement. The cerebrospinal fluid appeared xanthochromic with numerous red blood cells, and troponin levels were at the upper limit of normal. Following clinical stabilization, cerebral arteriography identified a 3mm aneurysm at the right posterior communicating artery, and cardiac catheterization showed no ischemic heart disease. The ECG changes were attributed to acute pain and a significant rise in blood pressure, mimicking acute coronary syndrome. **Discussion:** Subarachnoid hemorrhage (SAH) with ST-segment depression in anterior leads (V1-V4) is a rare but significant electrocardiographic finding in acute cerebral disorders. SAH, often caused by aneurysm rupture or trauma, can lead to neurological and cardiac manifestations due to the brain-heart interaction. ST-segment depression, typically associated with cardiac ischemia, often indicates neurogenic stunned myocardium rather than coronary artery disease in this context. This results from a surge in catecholamines due to acute brain injury, causing transient myocardial dysfunction. Recognizing these patterns as indicative of SAH's secondary cardiac effects is crucial for appropriate management and improved outcomes. **Final comments:** This case highlights the complexity of the correlation between cardiac manifestations and acute neurological events like SAH. Identifying electrocardiographic changes, specifically ST-segment depression, underscores the importance of integrated management of neurological and cardiac conditions. These findings emphasize the need to consider neurogenic stunned myocardium in SAH patients, recognizing that ECG changes may be neurally mediated rather than due to primary coronary disease. Ongoing investigation and treatment algorithms addressing both neurological and cardiac complications are essential for improving outcomes and patient quality of life.

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Subdural empiema due to contiguity of sinusopathy

Lorena Fernandes Kronbauer[1], Letícia de Barros Dias[1], Fábio Gonçalves[1], Felipe Leonardo Semensati[1], Nathalye Fernanda Pedroso Dircksen[1], Marcelo Simplicio Carvalho[1], Bárbara Okabaiasse Luizeti[1], Mônica Meneguette Silveira[1], Damacio Ramon Kaimen Maciel[1], Monica Matos de Souza[1]

[1]Irmandade Santa Casa de Londrina, Londrina PR, Brazil.

Correspondence: Lorena Fernandes Kronbauer; Email: lorenafkronbauer@gmail.com.

Case presentation: DMS, 65 years old, holocranial headache with frontal predominance for 15 days, mental confusion, edema in the left orbital and periorbital region, and presented congestion of the left maxillary sinus and hypodensity in the left frontal. Lumbar puncture: leukocytes 213 - predominance of lymphocytes, glucose 68, protein 186, with negative fungal analysis or culture.. She underwent therapy with ceftriaxone and vancomycin. After 5 days, course with right hemiparesis and clear appearance of left frontal subdural empyema adjacent to the frontal hypodensity; There was a decrease in the level of consciousness, with worsening hypodensity e now with left subdural empyema. Subjected to drainage, without clinical response. Metronidazole, amphotericin and polymyxin added. The exams did not demonstrate cavernous sinus thrombosis or any other finding that could explain the ocular edema. Cerebrospinal fluid culture, culture of secretion drained from the empyema and brain biopsy were performed, with negative results. Patient died. **Discussion:** The most affected are young male adults. There is difficulty in relating cause and effect between meningitis and sinusitis because, un like abscesses, it mostly originates from chronic sinusitis. The clinical picture includes fever, headache that increases in intensity, neck stiffness, irritability and delirium. There are two routes of infection from the paranasal sinuses to the intracranial cavity: Retrograde thrombophlebitis through diploic veins of the skull and ethmoid bone or communicating veins (Breschet's diploic veins) - by continuity; and by direct extension of sinus infection through congenital or traumatic dehiscences, erosion of the sinus wall, that is osteomyelitis, and existing foramina (e.g. olfactory nerves) - contiguity. Complications: meningitis, epidural abscess, subdural empyema, venous sinus thrombophlebitis and brain abscess. The frontal sinus is the most commonly associated with intracranial infection. In subdural empyema, only large collections cause a mass effect, leading to the appearance of an evident clinical picture. Anaerobic germs represent 29% of endocranial complications. Non-betahemolytic streptococcus are the most commonly found. Staphylococcus is isolated in subdural empyemas and brain abscesses, as well as in cavernous sinus thrombophlebitis and osteomyelitis. Pneumococcus and Haemophilus influenzae are more commonly found in meningitis of upper respiratory tract etiology. Anaerobes may occur in up to 50% of non-traumatic brain abscesses. Negative cultures occur in 21% of cases. **Final comments:** A CNS infection resulting from infectious sinusopathy is serious, includes disabling sequelae and is difficult to diagnose due to the chronicity of the symptoms and the exclusive attribution of the symptoms to sinusopathy, as in the case of our patient. CSF and empyema cultures have high rates of false negatives.

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Subsyndromal delirium as a predictor of death, loss of independence and functionality in post-stroke patients in the semi-intensive care unit

Thaís Barbosa de Oliveira[1], Gabriel Souza Medrado Nunes[1], João Pedro Fernandes Gonçalves[1], Alex de Cerqueira Silveira Figueiredo[1], Antonio Lázaro Santos Soares Junior[1], Felipe Asaf Borges Millard[1], Júlia Xavier Oliveira[1], Pedro Antônio Pereira de Jesus[1]

[1]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Thaís Barbosa de Oliveira; Email: thaisbo@ufba.br.

Background: Delirium is an acute, fluctuating confusional state defined by attention disturbances, associated with altered consciousness and/or disorganized thinking. It is common in semi-ICU settings and often underdiagnosed, leading to clinical deterioration, increased mortality, and functional decline. Previous studies also suggest that intermediate stages between normal cognition and delirium can hinder recovery. Patients with acute cognitive dysfunction, not meeting full delirium criteria–subsyndromal delirium (SSD)–may benefit from interventions due to their poorer prognosis compared to those without cognitive symptoms. This is especially relevant for post-stroke patients, as stroke-related brain injury lowers the threshold for cognitive impairment. **Objective:** This study investigates the association between SSD and functional loss in post-stroke patients, and whether SSD differs significantly from asymptomatic conditions. **Methods:** This prospective cohort study was conducted in a semi-ICU for stroke at a hospital in Bahia, Brazil, from 2023 to 2024. Eligible patients had a confirmed stroke diagnosis within 72 hours of symptom onset, excluding those under age of 18, unresponsive at admission, or diagnosed with blindness, complete hearing loss, severe aphasia, and/or dysarthria. Patients were screened for delirium using the Confusion Assessment Method for the ICU (CAM-ICU) every 2 days for up to 30 days. They were classified into 3 groups: non-delirium (4 negative criteria in all assessments), SSD (at least one altered criterion without fulfilling delirium diagnosis), and delirium (positive CAM-ICU test in at least one assessment). The main outcome was independence, functionality, or death, assessed by the Modified Rankin Scale (mRS) ninety days after stroke onset. **Results:** Two hundred sixty-two patients were enrolled, mostly women [138 (52.7%)], brown and hispanic individuals [131 (50%)], with a mean age of 65.34 (±12.4) years-old. The most common highest educational level was primary education [125 (47.7%)]. The mean for Short IQ-Code score was 3.13 (±0.82), 108 (41.2%) participants had a Charlson Comorbidity Index > 5 and most were independent and fully functional before the stroke (mRS < 2) [220 (84.0%)]. The median NIHSS at admission was 7.0 [IQR 4-13], with most cases being ischemic stroke [245 (93.5%)], and 109 (41.6%) were thrombolyzed. Univariate analyses showed a significant difference between non-delirium and SSD groups for mRS-90 < 2 [OR=2.60, 95% CI 1.32-5.09], though smaller than between non-delirium and delirium groups [OR = 10.8, 95% CI 3.67-31.80]. Multivariate analysis confirmed SSD as an independent predictor for mRS-90 <2 (p<0.001), compared to non-delirium. NIHSS at admission (p=0.011) and hospitalization duration (p<0.001) were also independent predictors for mRS-90 <2. **Conclusion:** Subsyndromal delirium predicts loss of functionality, dependency, or death within 90 days after stroke onset in a semi-ICU setting.

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Successful use of immunoglobulin as a treatment for post-infectious cerebellitis related to dengue fever refractory to steroids

João Vitor Assumpção Silva[1], Henrique de Moraes Bernal[1], Pedro Almeida Martins Pontes[1], Ingrid Lorena da Silva Gomes[1], Herval Ribeiro Soares Neto[1], Sônia Maria César Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: João Vitor Assumpção Silva; Email: joaosilva7890@hotmail.com.

Case presentation: A 17-year-old boy presented with difficulty standing and walking, along with a sense of imbalance, slurred speech, severe drowsiness, and vertigo. Additionally, he experienced uncontrollable nausea and vomiting refractory to symptomatic medications. The patient's medical history included only atopic dermatitis. Dengue, confirmed by NS1 test, was diagnosed 7 days prior to the onset of neurological symptoms. On examination, the patient was drowsy but responsive to commands, with slurred and scanning speech. Notable findings included bilateral ptosis, opsoclonus, hypermetric saccades,

saccadic pursuit, and significant axial and appendicular ataxia. He was unable to stand or walk without assistance, displaying a wide-based gait. When administering the MoCA test, the patient scored 20 points. Cerebrospinal fluid (CSF) analysis and brain MRI revealed no abnormalities. Serum tests for viral infections, including EBV, Varicella, HIV, Hepatitis, and Herpes Virus, were negative. Treatment with intravenous methylprednisolone (1g/day for 5 days) was initiated, followed by intravenous human immunoglobulin (IVIG) (0.4mg/kg/day for 5 days). After treatment, the patient achieved independent ambulation, and cerebellar symptoms markedly improved. At the conclusion of the treatment, the patient scored 24 points at MoCA. **Discussion:** Acute cerebellar ataxia is characterized by the acute onset of appendicular and axial ataxia associated with symptoms such as nystagmus, hypotonia, dysarthria, and gait disturbances. Causes are divided according to the nosology. This is the tenth reported case of post-infectious cerebellitis related to dengue worldwide. Initially reported in 2012, this condition was first associated with EBV before being observed in isolated cases of dengue infection. Currently, the disease mechanism is believed to involve both viral invasion of the central nervous system and an immune-mediated response. The prevailing hypothesis posits that dengue triggers a 'cytokine storm,' leading to endothelial injury, blood-brain barrier disruption, fluid leakage, and subsequent brain damage. Cerebellar involvement may occur during the acute phase but typically manifests 1 to 3 weeks after the onset of dengue symptoms. Neuroimaging abnormalities are rare, and cases currently described in the literature haven't demonstrated abnormal CSF analysis. Some cases are self-limiting and may not require therapy. When treatment is necessary, corticosteroids are the initial option. In this case, the decision to administer IVIG was based on the lack of response to high-dose corticosteroid therapy and supported by literature for its use in refractory post-viral cerebellitis. **Final comments:** Post-infectious acute cerebellar ataxia must be promptly recognized and investigated as it is a treatable condition with a generally favorable outcome. IVIG should be considered in patients who do not respond adequately to steroids.

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Sudden unexpected death in epilepsy (SUDEP): a retrospective longitudinal study of the Brazilian population over the last 10 years

Elvis Lucas Rabelo Shintay Chan[1]

[1]Universidade Federal de Ouro Preto, Ouro Preto MG, Brazil.

Correspondence: Elvis Lucas Rabelo Shintay Chan; Email: elvislucaschan2011@hotmail.com.

Background: Sudden Unexpected Death in Epilepsy (SUDEP) still has an etiology that is not yet fully known, and it is suggested that death occurs in the context of a seizure, which usually occurs at night, during sleep. Cardiogenic, pulmonary and neurogenic causes have been suggested. However, this event is not always witnessed and the diagnosis is not always possible. Although relatively uncommon, SUDEP represents between 2 and 18% of all deaths in patients with epilepsy, thus having a significant impact on the survival. **Objective:** This study sought to evaluate the epidemiology of SUDEP in relation to the Brazilian population, with a view to identifying risk factors or confirming what has already been observed in the population of other countries. METHOD: Epidemiological data on SUDEP between 2013 and 2022 were extracted from the Department of Informatics of the Unified Health System (DATASUS), from the Brazilian Ministry of Health. The outcomes analyzed were incidence by year, incidence by sex, incidence by skin color and mortality by age group, considering 10-year-old intervals. Descriptive and inferential statistical analysis was used to compare variables. **Results:** There were 31959 cases, with a statistically significant increase from 2020 onwards (p=0.004905401). There were more male deaths, in a ratio of 1.85:1 and with a significant difference between the groups (p=0.000210717). The highest incidences occurred in white people and pardo people, but without significant difference (p=0.4307446). There was a significant difference between the incidence between white and black individuals (p=0.00001572). Asian and indigenous populations could not be compared due to the small sample size. Regarding the age group, those who accounted for the majority of cases were between 40 and 49 years old (16,5%), 50 and 59 years old (16,68%), 60 and 69 years old (12,9%), 70 to 79 years old (10,15%) and 80 years old or more (10,3%). CONCLUSIONS: The increase in the incidence of SUDEP raises a warning about the need for careful monitoring of these patients, since one of the main risk factors is the lack of supervision. This implies less medical monitoring and risk of new crises. In fact, the increase observed occurred in 2020, when many individuals lost close contact with family members and health services during the Covid-19 pandemic. The greater occurrence of events in male patients follows the international literature. The differences observed in the incidence by skin color can be explained by genetic factors, since an association with genes related to sodium channelopathies, congenital heart diseases and some syndromes has already been identified. The prevalence of these mutations in the Brazilian population is, however, unknown. Finally, this work showed a higher proportion of cases occurring in people over 40 years of age, which contradicts other authors, who despite this already mention in their works the possibility of underreporting in this age group.

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SUNCT: a challenging diagnosis

Márcio Alves da Cruz Júnior[1], Iasmim Portela Maifrede[1], Carlos Eduardo Reis Silva[1], Maria Paula Cezar de Castro[1], Luiz Paulo Bastos Vasconcelos[1]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Márcio Alves da Cruz Júnior; Email: marcio.jr14@yahoo.com.br.

Case presentation: Case 1: JARL, a 54 years-old male patient, diagnosed with bipolar affective disorder, reported a new headache in 2016, characterized by shock pain in the right temporal/retro-orbital region, of severe intensity, lasting 60 seconds and associated with ipsilateral tearing and ocular hyperemia. The pain was triggered by chewing and tactile stimulation in the supra-orbital region of the right eye and had a frequency of 15 to 20 episodes per day. Initially, he was diagnosed with trigeminal neuralgia (TN). He used carbamazepine and gabapentin in optimized doses, without success. After confirming the diagnosis of SUNCT by ICHD-3 criteria, lamotrigine was started. After 6 months of follow-up, the patient had complete pain resolution at a dosage of 200mg/day. Case 2: FCR, a 71 years-old male patient, previously healthy, started experiencing headaches in 2016, characterized by excruciating shock/stabbing pain in the left orbital and parietal regions, with dozens of daily episodes, associated with tearing and ipsilateral conjunctival hyperemia . His condition worsened in 2018, after starting to use a dental prosthesis in the upper arch. Initially, he was diagnosed with TN and later with cluster headache (CH). He used carbamazepine and gabapentin for neuralgia, without success. The use of verapamil 240mg/day for CH resulted in little relief in the intensity of the attacks. The diagnosis of SUNCT by ICHD-3 criteria was performed and lamotrigine was prescribed. After 1 year of follow-up, the patient had complete control of the condition at a dosage of 300 mg/day. **Discussion:** The short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) are included in the trigemino-autonomic headaches (TCAs) group, listed in ICHD-3. In both patients presented, the initial diagnosis was attributed to other types of facial pain, such as CH and TN, given their semiological similarities. SUNCT and CH are listed in the TCAs group and share clinical characteristics such as the same pain territory and the presence of autonomic symptoms. The differential diagnosis between them is based mainly on the frequency and duration of pain attacks. Regarding NT, the similarities with SUNCT include pain in the trigeminal territory, the presence of pain trigger zones upon touch and stabbing/shock pain. The differential diagnosis is mostly based on the manifestation of autonomic symptoms in SUNCT and on the preferential involvement of the V2 territory of the trigeminal nerve in TN. **Final comments:** SUNCT is rare and may lead to severe functional disability. Both clinical cases presented here illustrate how its diagnosis is challenging. The objective of this report is to reinforce the importance of a detailed cephaliatric assessment to differentiate the types of CTAs. Furthermore, it is important to highlight that facial shock pain is not defining of TN, as it also occurs in SUNCT and other facial neuralgias.

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SUNHA case report: exceptional response to lacosamide treatment

Verena Miranda de Souza[1], Felipe Franco da Graça[1], Hiago Antunes Silva[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Verena Miranda de Souza; Email: vemirandasouza@gmail.com.

Case presentation: A 90-year-old individual presented with a new-onset headache described as a severe stabbing pain in the right supraorbital region, lasting for seconds to minutes, associated with photophobia, ipsilateral rhinorrhea and bilateral lacrimation. The pain was triggered by any local tactile stimulus, followed by a period of pain relief, without suspending it. Neurological examination did not reveal any noteworthy abnormalities. Therapies with carbamazepine and gabapentin were initiated with improvement in the frequency and intensity of pain. However, the patient continued to require rescue analgesic therapies. After two weeks, carbamazepine was discontinued due to major-like drug-induced erythema multiforme. The following month, a headache exacerbation led to another hospitalization. Therapeutic attempts were made but failed to significantly released the pain: levetiracetam, valproic acid, chlorpromazine, dexamethasone and anesthetic blocks. Subsequently, lacosamide was initiated at a dose of 100 mg intravenously every 12 hours leading to significant improvement in headache, allowing hospital discharge with maintenance of oral medications and suspension of corticosteroids and chlorpromazine. Due to effective pain management, the dosages of valproic acid and gabapentin were gradually reduced on an outpatient basis without pain recurrence. The patient is currently on lacosamide 100mg/day and gabapentin 600mg/day, maintaining complete headache control. **Discussion:** Short-lasting unilateral neuralgiform headache attacks (SUNHA) are similar to V1 trigeminal neuralgia, with autonomic features and no refractory period to cutaneous triggering. Classified in two subtypes: Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT) and Short lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA). Since attacks are short, medical therapy is primarily preventive. Lamotrigine is the treatment of choice for SUNCT and SUNA, with a clear dose-response relationship. As a second line there are an assortment of therapies used for trigeminal neuralgia and other syndromes: oxcarbazepine, duloxetine, topiramate, carbamazepine, gabapentin and pregabalin. In a prospective, open and non-randomized, single-center study of the six patients who tried lacosamide two reported improvement in headache (33%). Both of these patients were considered responders but required a dose higher than the recommended 300 mg daily to achieve a beneficial effect. This report describes a case of SUNHA

refractory to several medications but showing an excellent response to low-dose lacosamide, offering another treatment option for this condition. **Final comments:** So far, there is no consistent evidence for the use of lacosamide in the treatment of SUNHA. Further studies with larger sample sizes are needed to evaluate the true efficacy of lacosamide in this condition.

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Sunken eyes as a peculiar finding in neuronal ceroid lipofuscionoses

Raphael Pinheiro Camurugy da Hora[1], Victor Rebelo Procaci[1], Thiago Yoshinaga Tonholo Silva[1], Flávio Moura de Rezende Filho[1], José Luiz Pedroso[1], Orlando Graziani Povoas Barsottini[1], Manuela Magalhães Dardenne Tenório[1], João Victor Cabral Correia Férrer[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Raphael Pinheiro Camurugy da Hora; Email: raphaeldahora@hotmail.com.

Background: Neuronal ceroid lipofuscinoses (NCLs) are lysosomal storage disorders characterized by progressive neurodegeneration and the accumulation of autofluorescent lysosomal storage material called lipofuscin. These disorders result from genetic mutations causing enzymatic dysfunction, with certain types associated with lysosomal enzyme deficiencies and others with mutations in genes encoding transmembrane proteins, ATPase, and potassium channels. Sunken eyes are characterized by deeply recessed eyes within the orbits, resulting in a hollowed appearance, often associated with prominent, bushy brows, narrow palpebral apertures, hooded eyelids, and prominent noses. This facial abnormality is hardly ever described in patients with NCLs, such as in a Turkish series, and could be more frequent than previously though **Objective:** This study aims to report a unique clinical finding of sunken eyes in a Brazilian family with NCL type 2, suggesting it may be a previously under-recognized feature of the disorder. **Methods:** A case series approach was used to document the clinical presentation, genetic testing, and neurological assessments of three siblings with NCL type 2. **Results:** Case 1: A 41-year-old man presented with progressive ataxia, generalized seizures, severe imbalance, dysarthria, dysphagia, and was restricted to a wheelchair. Neurological examination revealed sunken eyes, and brain MRI showed olivopontocerebellar atrophy. Genetic testing identified a pathogenic variant in the TPP1 gene. Case 2: A 42-year-old woman exhibited unsteady gait, motor incoordination, emotional lability, and aggressive behavior, leading to wheelchair dependency by age 14. Similar neurological features and olivopontocerebellar atrophy were observed, with sunken eyes noted upon examination. Genetic testing supported a diagnosis of NCL type 2. Case 3: A 37-year-old woman experienced seizures from age 3, progressing to gait imbalance and slurred speech. Her examination also showed sunken eyes and other neurological symptoms, with brain MRI revealing olivopontocerebellar atrophy. A presumptive diagnosis of NCL type 2 was made based on familial genetic results. **Conclusion:** This case series highlights sunken eyes as a notable clinical feature in patients with NCL type 2. This finding, potentially linked to malnutrition and disease severity, suggests that sunken eyes could be an indicator of the progression of NCLs. Further research is warranted to explore this association and its implications for clinical practice.

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Superficial siderosis of the central nervous system: cause of sensorineural deafness and cerebellar ataxia - case report

Mário Natalício de Gasperi[1], Claudio Henrique do Amaral[1], Paulo Roberto Wille[1]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.

Correspondence: Mário Natalício de Gasperi; Email: mario.natalicio2@gmail.com.

Superficial siderosis of the central nervous system is a clinical entity discovered in 1908 by Hamill with the classic symptoms of progressive bilateral sensorineural deafness, cerebellar ataxia, imbalance and sometimes myelopathy. Its clinical findings are very rare in the population, with around 300 cases reported in the literature around the world. **Case presentation:** M.B.R, 73 years old, male, started 3 years ago with progressive bilateral hearing loss and imbalance when walking. He denied dizziness. History: severe head trauma 15 years before the first symptoms, systemic arterial hypertension, atrial fibrillation, benign prostatic hyperplasia and no history of surgery. Family history, nothing noteworthy. Former smoker and reports social alcohol consumption until the age of 62. Currently using Fluoxetine, doxazosin, simvastatin, losartan and apixaban. In the neurological examination, he is conscious, oriented, with preserved speech, severe bilateral hearing loss, other cranial nerves preserved, motor strength and sensitivity preserved, symmetrical reflexes, mild bilateral dysmetria, without didiadokinesia, Romberg test with mild instability, Tanden without conditions to perform and ataxic gait. Skull MRI in ferromagnetic susceptibility sequences reveals areas of signal void characterizing hemosiderin deposits involving the cortical and sylvian sulci bilaterally in a marked way, the cisterns on the cerebellar folia and perimesencephalic cisterns as well as ambiens and pentagonal and the ependymal cerebrospinal fluid space characterizing superficial hemosiderosis. Fazekas grade 3 leukoaraiosis, signs of volumetric involution of the cerebral and cerebellar hemispheres. **Discussion:** Superficial siderosis of the central nervous system is a rare clinical entity whose main cause of pathophysiology is the leakage of blood into the cerebrospinal fluid, which degrades into free iron, ferritin and hemosiderin, which are deposited in the superficial layers of the central nervous system (CNS) and lead to cellular changes such as reactive gliosis, demyelization, neuronal loss. Among the possible causes of hemorrhage in the subarachnoid space is a previous clinical history of surgical procedures in the posterior fossa, trauma, intradural surgery, spinal tumors or vascular malformations of the CNS; Such bleeding is low in volume, chronic and recurrent and may take decades until the first symptoms of superficial siderosis appear. The classic symptoms are associated with areas of susceptibility of the central nervous system to hemosiderin. **Final comments:** The symptom of bilateral progressive sensorineural deafness and ataxia makes a differential diagnosis with neurosyphilis, multiple sclerosis and other autoimmune causes. The rarity of the incidence of superficial siderosis of the central nervous system makes imaging findings on magnetic resonance imaging an ally in the diagnosis in the face of clinical suspicion.

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Surgical outcome in patients with mesial temporal lobe epilepsy and unilateral hippocampal sclerosis associated with calcified neurocysticercosis

Karen Karol Vásquez Quicaño[1], Marina K. Alvim[1], Fabio Rogerio[1], Enrico Ghizoni[1], Clarissa Lin Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas Campinas, São Paulo SP, Brazil.

Correspondence: Karen Karol Vásquez Quicaño; Email: karenvasquez1807@gmail.com.

Background: Calcified neurocysticercosis (cNCC) is considered a frequent etiology of epilepsy in developing countries. In Brazil, cysticercosis is an endemic disease, and seizures are the most frequent symptoms, occurring in up to 92% of patients. **Objective:** This study aimed to analyze postsurgical outcomes of patients with mesial temporal lobe epilepsy with unilateral hippocampal sclerosis (MTLE-HS), with and without cNCC, who underwent surgical treatment. **Methods:** A retrospective cross-sectional study was conducted on 82 patients with MTLE-HS who underwent anterior temporal resection plus amygdalohippocampectomy between January 1997 and December 2020 at the epilepsy center of the University of Campinas Universidade Estadual de Campinas. All patients had ictal semiology indicating MTLE and EEG findings concordant with the side of HS diagnosed with high-resolution 3T MRI epilepsy protocol. The presence of cNCC was determined with CT scans or SWI MRIs. We collected clinical data and neuroimaging information. The postoperative results were analyzed using bivariate analysis according to the Engel classification. Differences in proportions were analyzed using Fisher's exact test. **Results:** The median age was 42.15 years, and 54.87% were women. Thirty-three (40.2%) had multiple cNCC lesions in addition to HS, and 49 (59.8%) did not. Fewer patients with cNCC reported a history of febrile seizures compared to the group without cNCC (2 [6.1%] vs. 7 [14.3%]), but without statistical significance (p=0.3). The mean postoperative follow-up was 12.5 years for those with cNCC and 5.5 years for those without. Overall, 58/82 (70.72%) achieved Engel IA. At the last follow-up, 27/33 patients (81.8%) with cNCC were categorized as Engel IA compared to 31/49 (63.3%) in those without cNCC; however, this difference was not significant (p=0.086). **Conclusion:** Our results confirm previous studies showing that in patients with refractory unilateral MTLE-HS with concordant ictal semiology and EEG findings, cNCC does not change the probability of excellent postoperative seizure control and does not contraindicate surgery. These findings also indicate that the real cause of ongoing seizures in these patients who became seizure-free after surgery was not the cNCC but the HS. Therefore, the finding of cNCC on CT or MRI does not necessarily indicate the etiology of epilepsy, and perhaps neurocysticercosis acts as a precipitating insult in some patients with MTLE-HS.

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Surgical outcome in patients with severe epilepsies, intellectual disability and bilateral structural lesions: comparison of the Engel's and the pasis (Porto Alegre seizure impact scale)

André Palmini[1], Eliseu Paglioli[1], Rafael Paglioli[1], Francine Hehn[1], William Martins[1], Vicenzo Zarpellon[1], Thomas Frigeri[1], André Gus[1], Natalie Donida[1], Bibiana Thome[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Andre Luis Fernandes Palmini; Email: alpalmini@gmail.com.

IO: The link between surgery and seizure freedom narrows the spectrum of epilepsy surgery candidacy to people with well localized epileptogenic zones, often with visible lesions. However, many children and adolescents with severe epilepsies, do not "fit" the link, ie, may have surgically remediable epilepsies but without a reasonable expectation of seizure freedom. In many Centers, patients with severe epilepsies, intellectual disability and bilateral structural lesions are not considered suitable for surgery. A recurrent argument is that these patients fall in the unfavorable classes (III, IV) of the Engel's outcome scale. However, moving from a dramatic to a manageable situation honors the ultimate goal of epilepsy surgery, ie, to significantly improve the life of patients and families. Many have a potential surgical target, and although

seizure freedom is not likely, the prospect of shorter and milder seizures, eliminating the need to frequent visits to the emergency room and preventing developmental regression is a much desirable outcome. Such post-operative gains beyond complete seizure freedom are seldom captured by the Engel outcome scale. We designed the Porto Alegre Seizure Impact Scale (PASIS), quantifying and integrating objective and subjective data following epilepsy surgery, considering changes in frequency, severity, duration and circadian distribution of seizures and post-ictal abnormalities. Here, we present a comparison between the Engel and the PASIS outcome scales in patients with severe epilepsies, intellectual disability and bilateral structural lesions. M: 31 patients with daily focal onset and generalized seizures and bilateral lesions underwent presurgical evaluation followed by resective surgery. All had a history of delayed psychomotor development associated with bilateral structural lesions. Surgical outcome was classified according to both scales. R: Eleven patients had tuberous sclerosis and the other 20 included: 6 with diffuse atrophy due to perinatal hypoxia , 4 with bilateral MCDs, 4 with bilateral cortico-subcortical encephalomalacia due to severe prematurity, 3 with bilateral cortico-subcortical abnormalities due to meningitis/encephalitis; and one each with diffuse MRI atrophy due to chromosomal translocation or CDKL5 mutation. Mean pre-operative PASIS score was 11 (range: 9 - 12) and 4.3 (- 4 to + 16) after operation. 24/31 (77%) patients were on PASIS 1, 5/31 (16%) on PASIS 2 and the other two on (7%). In comparison, only 7/31 (23%) were in Engel class I, and the other 24 (77%) in classes III and IV. C: Pragmatic assessment of the impact of surgery in the daily life of patients with severe epilepsies and their families demand an outcome scale that integrates objective and subjective observations. Within this framework, many patients with diffuse brain dysfunction and bilateral structural lesions will be candidates for presurgical evaluation and attain a very significant positive impact with surgery.

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Survival analysis in patients with myasthenia gravis in a city in the countryside of the Brazilian Amazon

Marcos Manoel Honorato[1], Abraão Luiz Colares Gualberto[1], Jean Augusto de Sousa Tavares[1], Cleizimara Cavalcante Nunes[1], Ilga Milla Chaves Silva[1], Samuel Oliveira de Amorim[1], Ana Flávia Ribeiro Nascimento[2], Matheus da Silva Ferreira[1], Kelson Vinicius Dallabrida Padilha[1], Átila Barros Magalhães[1]

[1]Universidade do Estado do Pará, Santarém PA, Brazil.
[2]Santa Casa de Misericórdia de Barretos, Barretos SP, Brazil.

Correspondence: Marcos Manoel Honorato; Email: marcos.honorato.dr@gmail.com.

Background: Myasthenia gravis is an autoimmune disease characterized by weakness and rapid fatigue of voluntary muscles. This occurs due to a disruption in communication between nerves and muscles, caused by antibodies that block or destroy acetylcholine receptors at the neuromuscular junction. The survival of patients with myasthenia gravis has significantly improved due to advances in treatments and clinical management of the disease. However, variability in access to treatments and socioeconomic conditions can influence these outcomes, especially in specific regions such as the interior of the Brazilian Amazon. **Objective:** To evaluate the profile and survival of patients diagnosed with Myasthenia Gravis in a city in the interior of the Brazilian Amazon between 2005 and 2020. **Methods:** Historical cohort, observational, and retrospective study. Survival analysis methods were used to identify prognostic factors for disease remission during the 36-month observation period. The correlation between variables and the outcome of death was performed using the chi-square test. **Results:** A total of 36 participants were included in the study. The majority of the patients were female (66.6%) and had the generalized form of the disease (86.1%). The most prevalent symptoms were: ophthalmoparesis (97.2%), fatigue (75%), and dysphagia (72.2%). Among the complications, 19.4% experienced a myasthenic crisis. The dosage of anti-acetylcholine receptor antibody (AChR) was positive in 58.3% of the patients, and 69.4% underwent electroneuromyography, with 72% of these showing an electro-decrement. Most patients responded to the standard stepped treatment and achieved remission (83.3%), while 16.6% died. As some patients did not have complete control, it was necessary to introduce adjuvant therapy with corticosteroids (69.4%), in addition to azathioprine (33.3%) or cyclosporine (25%) in patients resistant to corticosteroid therapy. A portion required thymectomy (19.5%) and 33.5% required hospitalization due to disease exacerbation. Survival analysis using Kaplan-Meier curves and Log-rank test showed that the variables related to poor control were male gender (p=0.01), thymic disease (p=0.01), and the use of cyclosporine (p=0.02). **Conclusion:** The study showed that the progression of individuals with Myasthenia Gravis and our patients had a similar progression to those described in the literature.

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Survival analysis of risk factors for the West syndrome's refractoriness in Brazilian Amazon's children

Marcos Manoel Honorato[1], Jonas Wesley Pinto de Lima[1], Kariny Facioni Scalabrin[2] , Jaqueline da Rocha Calvo[1], Marissol Rabelo de Almeida[1], Arthur Severiano dos Santos Pontes[1], Clara Sofia da Silva Oliveira[1], Norma Vitória da Silva Fialho[1], Kelson Vinicius Dallabrida Padilha[1], Samuel Oliveira de Amorim[1]

[1]Universidade do Estado do Pará, Santarém PA, Brazil.
[2]Hospital Adventista de Manaus, Manaus AM, Brazil.

Correspondence: Marcos Manoel Honorato; Email: marcos.honorato.dr@gmail.com.

Background: West syndrome is a severe epileptic encephalopathy characterized by infantile spasms, electroencephalographic pattern with hypsarrhythmia and delay in neuropsychomotor development. It presents a poor prognosis and high morbidity, with direct implications for quality of life. **Objective:** To perform a survival analysis related to the control of seizures in patients with West syndrome, as well as estimate the response to treatment, evaluate the clinical and electroencephalographic evolution and identify the main outcomes and prognostic factors. **Methods:** A retrospective observational study was carried out, analyzing the medical records of 34 patients diagnosed with West syndrome treated between 2005 and 2020 in two specialized services in a city in the interior of the Brazilian Amazon. The research was approved by the Ethics Committee of the Baixo Amazonas Regional Hospital, following international bioethical standards. We calculated the proportions of categorical variables, and then performed a survival analysis with an observation time of 6 months from the first consultation, obtaining Kaplan-Meier survival curves. The Log Rank statistical test was used and the risk rate was calculated. **Results:** The average age at first crisis was 4.9 months. Complications during pregnancy and childbirth were observed in 50% and 47%, respectively, perinatal hypoxia was also reported in 47%. The majority of patients (70%) presented changes in the neuroimaging examination. All patients presented changes in the initial neurological examination, predominantly spasticity (64%) and motor deficit. Among the medications, vigabatrin had the highest frequency of use (51%). Only 41.2% of patients achieved complete control of spasms. The median time to control was 4.5 months. The majority progressed to non-evolutionary chronic encephalopathy or intellectual disability. There was converseion to Lennox-Gastaut Syndrome, progressing to focal epilepsy or other epilepsies, in 58.8%. Only 19% progressed to normal neurodevelopment. The most significant factors for poor seizure control over time were non-use of vigabatrin (p= 0.049, relative risk= 2.73), low birth weight (p=0.05, relative risk= 2.64) and spasticity (p=0.05, relative risk=1.99). **Conclusion:** West syndrome has a variable evolution, but the patients in the study had worse control and outcome percentage, compared to those reported in the most recent literature, which increased the risk of cognitive and motor sequelae. This was possibly due to poor access to specialized medical care in the interior of the Amazon and the low availability of high-cost medications in the initial years of the study. Furthermore, among the therapies most used in treatment, vigabatrin was the only one that proved to be relevant for controlling spasms.

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Susac syndrome accompanied by Wernicke: a rare presentation

Márcio Alves da Cruz Júnior[1], Luiz Felipe Mota Freitas[1], Leopoldo Antonio Pires[1], Ana Laura Maciel Almeida[1], Daniel Sabino de Oliveira[1], Thiago Cardoso Vale[1]

[1]Universidade Federal de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Márcio Alves da Cruz Júnior; Email: marcio.jr14@yahoo.com.br.

Case presentation: A 22-year-old woman, with 9 years of schooling, with a history of chronic alcoholism, use of electronic cigarettes and illicit drugs (LSD, MDMA and cocaine), began experiencing apathy, hyporexia and headache that progressed over a period of one week with weakness of lower limbs and difficulty walking and after two weeks with dizziness, mental confusion and imbalance. On neurological examination, she presented spastic asymmetric tetraparesis with hyperreflexia, anarthria, ataxia, fluctuating consciousness and right hearing loss. Brain MRI demonstrated numerous hyperintense foci on T2/FLAIR throughout the supra and infratentorial white matter, with numerous small foci of diffuse enhancement with a non-perivascular miliary pattern, in addition to signs of supra and infratentorial leptomeningeal enhancement. The presence of high signal foci on T2/FLAIR in the corpus callosum accompanied by headache with hearing loss raised a strong suspicion for Susac syndrome. Cerebrospinal fluid (CSF) identified proteinocytological dissociation, increased CSF IgG and albumin, and absence of oligoclonal bands. Serology was negative for rheumatological analysis and infectious diseases. Given the history of long-term alcoholism combined with mental confusion and ataxia, with the presence of hyperintense lesions in the supra and infratentorial white matter, Wernicke's syndrome hypothesis was made, and the patient underwent vitamin B1 replacement, achieving substantial motor and confusional improvement. However, the patient developed marked cognitive impairment (Montreal Cognitive Assessment - MoCA 15 points out of 30), and upon reassessment in an imaging exam 9 months later, the lesions in the corpus callosum persisted, what led us to suspect of Susac syndrome. **Discussion:** Susac syndrome is a rare disease characterized by the triad of encephalopathy, retinal artery occlusions and hearing loss, with an undefined pathogenesis. We report here a case of leukoencephalopathy with a radiological pattern typical of Susac syndrome, with rapid progression of cognitive decline. The syndrome is marked by presumably autoimmune retinocochleocerebral microangiopathy. The toxic-metabolic and infectious relationships are not well established, however the literature has already shown an association between cocaine use and leukoencephalopathy. CSF examination shows mild lymphocytic pleocytosis and hyperproteinorrhachia, and MRI typically shows lesions involving midline areas of the corpus callosum. Wernicke syndrome can typically present with edematous lesions that can be reversed with the use of

thiamine. **Final comments:** This case shows an association of Wernicke's syndrome with significant clinical and radiological improvement with the use of thiamine, and Susac syndrome with cognitive impairment and hearing loss associated with typical lesions in the corpus callosum.

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Susac syndrome (retino-cochlear-cerebral vasculitis)

Luis Fernando Guimarães[1], Luiz Guilherme Gerhardt Fernandes[1], Osvaldo Pantoja de Oliveira[1], Bárbara Luisa Coraspe Gonçalves[1], Maria Luiza Levindo Coelho Martinis[1], Eduardo Peliky Fontes[2]

[1]Hospital Federal dos Servidores do Estado, Rio de Janeiro RJ, Brazil.
[2]Universidade Estadual de Montes Claros, Montes Claros MG, Brazil.

Correspondence: Luis Fernando Guimarães; Email: luisguimaraesneuro@gmail.com.

Case presentation: A 64-year-old man has progressively developed ataxic gait for approximately 10 years. He was referred to the demyelinating diseases clinic due to lesions in the corpus callosum and other white matter, although with a vascular appearance. Approximately 5 years ago, acute bilateral hearing loss of central etiology (sensorineural hearing loss) began through audiometry (Figure 1). In the last 3 years, he has presented dementia symptoms, behavioral changes, loss of anterograde amnesia, aggressiveness, executive dysfunction and prosopognosia. The magnetic resonance imaging of the brain shows lesions in the corpus callosum and white matter of a vascular nature, volumetric reduction of the hippocampal gyri and cortical atrophy (Figure 2 e 3). His optical coherence tomography shows a reduction in the thickness of nerve fibers in the temporal sector of the right eye and a reduction in the thickness of fires in the foveal region of the left eye (Figure 4). His ocular angiography showed prolonged circulatory time and an increase in the foveal avascular zone in both eyes. In analysis of the clinical findings and complementary exams, the patient was diagnosed with Susac Syndrome and started on immunosuppressants. **Discussion:** Susac syndrome is a rare occlusive microangiopathy (vasculitis) of unknown etiology and mechanism involving retinal arteries, cochlea and brain. Ophthalmological signs and symptoms are the first to be noticed in 50% of cases, encephalopathy affects up to two thirds of patients and is expressed by cognitive changes, headache, personality or behavioral changes. Magnetic resonance imaging shows involvement of the corpus callosum with typical multifocal snowball-shaped lesions on T2. Hearing loss is usually acute and bilateral. The typical age of onset of the disease is between 20 and 40 years of age, but the age range is from 2.5 to 72. It predominantly affects women with an estimated male/female ratio of about 1:3. Treatment generally involves corticosteroids and other immunosuppressants such as mycophenolate mofetil, azathioprine, rituximab and infliximab or cyclosporine. Plasmapheresis and the use of immunoglobulins also constitute treatment for the disease. **Conclusion:** The reported case is diagnosed with Susac Syndrome. The patient presents the classic clinical triad of the disease, manifested by visual disturbances, due to occlusion of a branch of the retinal artery, hearing loss and encephalopathy. The exact prevalence is unknown, but to date, just over 300 cases have been published worldwide.

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Susac's syndrome: a case report

Yago Marcos Pessoa-Gonçalves[1], Victor Hugo Palhares Flávio Reis[1] , Chamberttan Souza Desidério[1], Marlos Aureliano Dias-sousa[2], Weslley Guimarães Bovi[3] , Gabriela Terra Silva[1], Alex Eduardo da Silva[2], Marcos Vinicius da Silva[1], Carlo José Freire Oliveira[1]

[1]Universidade Federal do Triângulo Mineiro, Instituto de Ciências Biológicas e Naturais, Laboratório de Imunologia e Bioinformática, Uberaba MG, Brazil.
[2]Universidade Federal do Triângulo Mineiro, Hospital de Clínicas, Departamento de Neurologia, Uberaba MG, Brazil.
[3]Yale University School of Medicine, Department of Comparative Medicine, New Haven, CT, USA.

Correspondence: Yago Marcos Pessoa Gonçalves; Email: yagompg98@gmail.com.

Case presentation: A 23-year-old male office assistant with no significant medical history presented in April 2023 with hand paresthesia and tinnitus, which resolved spontaneously. In May 2023, he experienced a self-limited episode of headache. In June 2023, he developed diplopia and scotoma, seeking care at the emergency room of the Clinics Hospital at the Federal University of Triângulo Mineiro. Upon admission, there was a reduction in vision in the lower field of the left eye; fluorescein angiography revealed branch retinal artery occlusion (BRAO). Laboratory-serological investigation, electrocardiogram, carotid and vertebral artery Doppler ultrasound, and echocardiogram showed no abnormalities. In July 2023, brain MRI revealed numerous hyperintense lesions distributed in the corpus callosum in a "snowball" pattern and throughout the cortex and white matter of both cerebral hemispheres. Suspecting Susac's Syndrome (SuS), audiometry and lumbar puncture were performed, both without abnormalities. In August 2023, there was a catastrophic worsening, with increased visual loss, right hemi-hypoesthesia, and hearing loss. A new audiometry revealed sensorineural hearing loss in the left ear, confirming the diagnosis of SuS according to the European Susac Consortium. The patient was hospitalized and treated with human immunoglobulin and methylprednisolone for five days, with cyclophosphamide on the fifth day. **Discussion:** SuS is characterized by the triad of visual deficit (BRAO), hearing loss, and encephalopathy. Studies show that CD8+ T cells anchor to the endothelium, release granzymes and perforins, cause microvascular bleeding, activate the coagulation cascade, and promote vascular occlusion. It is a rare condition, with fewer than 700 cases reported in the literature, potentially progressing to blindness and deafness without adequate treatment. **Conclusion:** SuS should be considered as a differential diagnosis in diseases with components of its triad for appropriate investigation and treatment.

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Syndrome of delayed neuropsychomotor development secondary to a rare genetic alteration due to a deletion in chromosome 8q21.13

Lais Cabral Almeida[1], Izabela Passadore Hauagge dos Santos[1], Ana Maria Sversut Briante[1], Leonardo Cittadella Alves[1], Ana Paula Galvão Paim[1], Giovanna Molitor Perini[1], Natália Gabrielli Silva Alves[1], Gustavo Baranhuk Oliveira[1], Wanclis Pinheiro Poussan[1], Luana Maria da Silva Rodrigues[1]

[1]Centro Universitário de Várzea Grande, Várzea Grande MT, Brazil.

Correspondence: Laís Cabral Almeida; Email: laiscabralalmeida1@gmail.com.

Case presentation: E.F.V.M, 7 years and 10 months old, woman, with no complications during pregnancy or childbirth. The mother reports using unknown medication for arthrosis in the first trimester of pregnancy and a family history of mood and neurodevelopmental disorders. The patient has a history of congenital heart disease, atopic dermatitis, asthma, obesity and astigmatism. She was referred to the pediatric neurology clinic due to global neuropsychomotor developmental delay. Physical examination revealed prominent ears with low implantation, appendicular hypotonia and reduced muscle strength. CGH array, a genetic test, showed a rare deletion of a 6.5Mb interstitial segment of the long arm of chromosome 8 (8q21.13). During clinical evaluation, the patient presented a pattern of global hypotonia, speech articulation delay, dependence in activities of daily living, delay in personal-social and school development. She is currently being treated with risperidone 0.2 mg, melatonin 9 mg and asthma medication, without any follow-up from the multidisciplinary support team. However, she was referred for multidisciplinary therapeutic follow-up to ensure a better prognosis and quality of life. **Discussion:** Chromosome 8 band deletion is considered rare, with only 18 reported cases in literature. The diagnosis is complex, observed during the postnatal period in individuals with multiple anomalies and developmental delay that do not characterize known genetic syndromes. In this patient, a deletion of the 6.5 Mb interstitial segment of the long arm of chromosome 8 (8q21.13) was identified by chromosomal microarray analysis. The deleted segment of the genome is responsible for approximately 48 genes, 15 of which encode proteins and 4 of which are associated with comorbidities, such as heart disease. The genetic investigation was carried out due to the characteristic phenotype of chromosomal alterations, such as facial dysmorphism, global hypotonia, gait instability and delayed neuropsychomotor development, which could be explained by the syndrome. Given her pathological history, she had atrial septal defect at birth, which was surgically resolved at the age of 4. This comorbidity may be related to the chromosomal alteration presented by the patient. However, the detection of genetic alterations allows for multidisciplinary monitoring, prognosis assessment and genetic counseling. **Final comments:** This shows that genetic tests are fundamental for differentiating cases of global developmental delay, a frequent issue in practice, but with various possible clinical manifestations. Genetic research in this case provided prognostic information regarding clinical evolution and treatment. Therefore, multidisciplinary therapy is also crucial for the development of impaired skills, despite the challenges of access in the Unified Health System and high costs in the health private network.

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Syndrome of nine: case report of an infarct in the dorsal pontine tegmentum

Loyslene Maria de Paula Bonifácio[1], Julia Lima Vilela[2], Jessica Cristina Silveira Damasceno[2], Thiago Cardoso Vale[2]

[1]SUPREMA, Juiz de Fora MG, Brazil.
[2]Hospital e Maternidade Therezinha de Jesus, Juiz de Fora MG, Brazil.

Correspondence: Loyslene Maria de Paula Bonifácio; Email: loyslene.med@gmail.com.

Case presentation: A 49-year-old right-handed male truck driver, with a history of hypertension, diabetes, dilated cardiomyopathy, and a previous ischemic stroke (with residual left central facial paralysis), presented with a history of fronto-ocular trauma due to his truck's load had fallen over his face two days prior to admission. Since the event, he reported localized pain, malaise, subjective vertigo, moderate left-side headache, and elevated blood pressure. Neurological examination revealed dysarthric speech, right peripheral facial paralysis, horizontal diplopia with conjugate gaze paralysis when attempting to look at right, with left eye adduction paralysis, right eye adduction and abduction paralysis. Only his left eye abduction was preserved. Vertical and convergence eye movements were intact. Additionally, there was decreased tactile, thermal, and pain sensation on his left side of the

body. Cranial computed tomography (CT scan) showed no significant acute abnormalities. Due to the trauma, vertebral artery dissection was hypothesized, but cervical and cranial CT angiography were also normal. However, brain magnetic resonance imaging (MRI) revealed focal diffusion restriction in the mid-dorsal tegmentum of the pons extending dorsally to the right medulla, consistent with an acute vascular insult, confirming a medial pontine infarct as the underlying cause of symptoms. Based on his presentation and neuroimaging, a diagnosis of syndrome of nine (one-and-a-half syndrome - horizontal conjugate gaze palsy with internuclear ophthalmoplegia - plus ipsilateral fascicular seventh nerve palsy and contralateral hemianesthesia due to medial lemniscus involvement in the pons) was made. The patient was admitted to the ischemic stroke unit and treated with antithrombotic therapy, including enoxaparin and clopidogrel, along with supportive measures such as blood pressure control and motor rehabilitation with a speech therapy team. **Discussion:** Syndrome of nine is a rare neurological condition that combines one-and-a-half syndrome with ipsilateral facial paralysis and contralateral hemianesthesia. It is characterized by the presence of internuclear ophthalmoplegia and horizontal conjugate gaze paralysis, along with ipsilateral facial nerve palsy and contralateral hemianesthesia or hemiparesis due to involvement of the medial lemniscus or corticospinal tract, respectively. The syndrome of nine is associated with pontine lesions, predominantly of vascular origin (infarct or hemorrhage). This case report aligns with the literature, where most described cases involve more medial pontine lesions. **Final comments:** This case of syndrome of nine underscores the need for thorough neurological examination, rapid diagnosis, and appropriate treatment to minimize neurological sequelae. Early identification and targeted therapy are crucial for improving the prognosis of patients with this rare syndrome.

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Syringomyelia and vocal cord paralysis

Moacir Pereira Leite Neto[1], Ricardo Queiroz Gurgel[2], Kelly Rejane Leite Figueiredo de Oliveira[3], Pedro Garcia Dias de Barros[1], Isabel Monique Leite Romualdo[4]

[1]Universidade Federal do Cariri, Barbalha CE, Brazil.
[2]Universidade Federal de Sergipe, São Cristóvão SE, Brazil.
[3]Afya Educação Médica, São Paulo SP, Brazil.
[4]Escola Estadual de Educação Profissional Presidente Médici, Presidente Médici MA, Brazil.

Correspondence: Moacir Pereira Leite Neto; Email: moacir.leite@ufca.edu.br.

Chiari malformation (CM) is an anomaly that can cause various neurological deficits. We describe a patient with MC type I and cervical syringomyelia (CS) accompanied by bilateral vocal cord paralysis. **Case presentation:** Patient F.D.S., male, 59 years old, living in Crato-Ce, 14 years ago began progressive and rapid onset of hoarseness, progressing to complete aphonia in three days, associated with dysphagia, followed by due to dyspnea, having been admitted to the ICU on the same day due to acute respiratory failure, and an emergency tracheostomy was performed. Exams carried out during hospitalization showed a normal chest X-ray, serum tests (blood count, serum electrolytes, CPK, lactic acid, creatinine and blood glucose), cervical ultrasound, electroneuromyography of the four limbs and skull tomography were normal. Laryngoscopy revealed bilateral paralysis of the vocal folds, with minimal opening of the glottis during inspiration. MRI of the skull and cervical spine showed tapered cerebellar tonsils, migrated 1.5 cm below the foramen magnum, compatible with MC type 1, and SC. The patient received a permanent tracheostomy after one month, and over the next 3 months he gradually developed paresis of the right upper limb. The patient was evaluated by a neurologist in March 2021, finding paresis and muscular atrophy in the right upper limb, predominantly distal, and paresis for elevation of the palate, with the presence of nausea reflex, dysphagia and using a metal tracheal cannula, and the patient underwent a surgical approach to treat CS associated with MC. **Discussion:** CM consists of the caudal displacement of the cerebellum and brainstem through the foramen magnum, which may be accompanied by compression of the brainstem and spinal cord, and syringobulbomyelia. Initial symptoms include recurrent headaches, neck pain, progressive ataxia and dysphagia. CS commonly occurs in patients with CM, and consists of a cystic cavity in the spinal cord, which can cause sensory loss and brachial amyotrophy. Its association with bilateral vocal cord paralysis or dyspnea is uncommon but well recognized, but is rarely seen in adults. When the corticobulbar fibers are affected, which culminate in the caudal portion of the nucleus ambiguus, laryngeal nerve cells can be affected, and a lesion below the decussation of the pyramids favors vocal cord paralysis. CS treatment is based on reducing cavity pressure and improving CSF flow, with shunt implantation (SI), lysis of adhesions or cyst fenestration. We describe a patient with type I CM and CS accompanied by acute bilateral paralysis of the vocal folds, treated by SI, and who evolved with no functional recovery, and it should be noted that the presence of long-lasting gliosis limits the surgical benefit. **Final comments:** We highlight the importance of early diagnostic recognition of MC and CS, including through bulbar muscle paresis, as well as rapid treatment, in order to obtain a better prognosis for symptomatic patients.

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Syringomyelia associated with xanthomatous plaques in dermis: a case report

Jonnathan Camara da Silva Vianna[1], Eduardo Martiniano de Lima[1], Moisés Vasco de Freitas[1], Gabriel Belo Cavalcante Linhares[1], Yasmin Luíse Lucena e Silva[1], Emanuela Maria de Aguiar Azevedo[1], Samila Marissa Pinheiro Gomes[1], Aglagilson Fernandes das Chagas[1]

[1]Universidade Federal Rural do Semi-Árido, Mossoró RN, Brazil.

Correspondence: Jonnathan Camara da Silva Vianna; Email: jonnathanvianna@gmail.com.

Case report: A 37-year-old woman, the only child of non-consanguineous parents, with two siblings who died as infants due to malformations. She has had scoliosis since childhood. She was referred to the neurology clinic due to a neurological condition of proximal tetraparesis. In 2024, magnetic resonance imaging (MRI) revealed extensive syringomyelia, extending from the cervical region to the T11-T12 level, along with moderate diffuse reduction in medullary caliber. On examination, she exhibited grade IV strength in bilateral deltoids and iliopsoas, impaired deep artresthesia sensitivity in the lower limbs, and abolished vibratory sensation up to the iliac crest. Additionally, she presented Hoffmann's sign in the left hand and Romberg's sign with tendon dancing. Dermatological examinations revealed thick-textured skin with dark spots and increased elasticity. In 2018, histopathological study of the lesions indicated fragmented elastic fibers in the reticular dermis region, with calcium deposition, suggesting pseudoxanthoma elasticum (PXE). A genetic test was requested for diagnostic confirmation. **Discussion:** Syringomyelia is characterized by the formation of a cavity within the spinal canal, commonly associated with central nervous system malformation. Additionally, the MRI showed signs of vertebrobasilar invagination and platybasia, configuring communicating syringomyelia type I. Symptoms vary according to the level and extent of involvement, and in this patient, muscle weakness, progressive scoliosis, ataxia, and reduced superficial abdominal sensitivity were observed. Furthermore, PXE is a rare genetic disorder, inherited in an autosomal dominant or recessive manner, caused by mutations in the ABCC6 gene, leading to abnormal calcium deposition in elastic fibers of the skin, eyes, and blood vessels, resulting in leather-like skin lesions and increased cardiovascular risk. For diagnostic confirmation, ocular changes such as angioid streaks and confirmation of ABCC6 gene mutations by genetic testing are necessary. PXE is progressive and requires serial evaluations by an ophthalmologist and a cardiologist due to the risk of atherosclerosis, reduced walking capacity, and serious complications such as thromboembolic events and ophthalmological changes like drusen and pattern dystrophy. Moreover, even rarer cases of PXE harbor pathogenic variants in the ENPP1 gene, causing generalized arterial calcification of infancy (GACI), inherited in an autosomal recessive manner, and resulting in infant death by the first year of life, which may explain the cause of death of the patient's siblings as infants. **Final comments:** This report illustrates the rare association between PXE and extensive symptomatic syringomyelia. It highlights the importance of careful differential diagnosis and interdisciplinary approach. Early detection and treatment of complications from both conditions can significantly improve quality of life.

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Systematic review and meta-analysis of mortality in neuromyelitis optica spectrum disorder (NMOSD)

Leonardo José Rodrigues de Araújo Melo[1], Francisco Luciano Honório Barreto Cavalcante[1], Leonardo Elias Araujo dos Santos[1], Emanuela Freire Caetano Davi[1], Rodrigo Fagundes da Rosa[1], Yasmim Alves de Maria Nunes[2], João Pedro de Oliveira Gouveia Marcotti[1], Alan Duarte Braz[1], Wagner Patrik Sousa Silva[1], João Marcelo Albuquerque Beserra de Sousa[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Leonardo José Rodrigues de Araújo Melo; Email: lleonardomello98@alu.ufc.br.

Background: Neuromyelitis Optica Spectrum Disorder (NMOSD) is an autoimmune disease of the central nervous system, characterized by the production of autoantibodies against astrocytic aquaporin-4(AQP4), leading to demyelination and neuronal damage, which causes serious clinical complications such as optic neuritis and longitudinally extensive transverse myelitis. As a result, NMO is a severely disabling disease with insufficiently known mortality rates. **Objective:** To estimate the mortality rate (MR) in NMOSD and identify clinical factors associated with it. **Methods:** A systematic literature search was conducted on PubMed and Web of Science platforms, using DeCS/MeSH descriptors "Neuromyelitis Optica Spectrum Disorder" or "NMOSD" and "Mortality" or "Survival rate". The inclusion criteria were cohorts or observational studies published in English that cites MR. Reviews, case reports or clinical trials were excluded. Articles were analyzed by two blind observers using the Rayyan AI platform, with divergences analyzed by a third independent individual. Data and statistical analysis were processed using jamovi software. Initially, 67 papers on the subject were obtained. **Results:** Thirteen articles met the inclusion criteria, including 12 cohort studies and 1 observational study, from 10 different countries, totaling 29.534 patients of which more than 80% were women, with an average follow-up of 11.8 years (ranging from 2 to 27 years). The total number of deaths among NMOSD patients was 748, with an average age of death of 58.5 years (ranging from 54 to 66 years). The MR was 7.35% (95% CI: 5-10%, $p< 0.01$) and the mean Expanded Disability Status Scale (EDSS) of the deceased patients was 6.35, estimated by 7 articles that presented EDSS data.Standardized mortality ratio (SMR)variables from 2,54% to 3,52%, higher than the general population.The mean EDSS is high in group deceased patients compared to survivors group at

the follow-up time.A later onset of symptoms, especially after the age of 50, was associated with a higher risk of death. Delayed treatment and the number of attacks per year were other conditions associated with high mortality in NMOSD patients. The main causes of death related to NMOSD were infections due immunotherapy, mainly of the respiratory tract, and respiratory failure due to extensive cervical myelitis. **Conclusion:** Patients with NMOSD have a high mortality rate and a reduced life expectancy compared to the general population. High mortality in NMOSD was associated with high EDSS values and a later onset of symptoms. Study limitations include the lack of EDSS data in some papers and the lack of articles investigating risk factors associated with mortality and comparing them with control groups. Further studies with more standardized methodologies are needed to more accurately evaluate MR in NMOSD and risk factors associated with it.

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Systemic lupus erythematosus associated with posterior reversible encephalopathy syndrome: a case report

Mariana Costa Borges[1], Jessica Vargas Lopes[2], Caroline Dresch Sabadin[3], Maria Júlia Todero[3], Carolina Aparecida Donofrio Santos Costa[1], André Luiz Pereira Martins[1], Diego Antonio Fagundes[4], Olga Cassol Silva[1], Maria Eduarda Andreatta[5]

[1]Universidade do Sul de Santa Catarina, Palhoça SC, Brazil.
[2]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[3]Universidade Estadual do Oeste do Paraná, Cascavel PR, Brazil
[4]Universidade do Extremo Sul de Santa Catarina, Criciúma SC, Brazil.
[5]Universidade Federal de Santa Catarina, Florianópolis SC, Brazil.

Correspondence: Mariana Costa Borges; Email: maricostaborges@hotmail.com.

Case presentation: A 16-year-old male patient initially presented with bacterial tonsillitis, progressing to lower limb swelling, dyspnea and headache. He sought hospital care which revealed hematuria and pleural effusion, and was treated for pneumonia with levofloxacin. After 10 days, he developed two episodes of seizures lasting 10 minutes each, with tonic-clonic tremors, and came to our service. On admission, he had hypertension 160/90mmHg, a positive Brudzinski sign and new seizure episodes. Pulse therapy of 1mg/kg methylprednisolone was carried out in the suspicion of autoimmune encephalitis and the patient showed clinical improvement. The electroencephalogram showed moderately disorganized baseline activity with an excess of slow rhythms in the delta band superimposed by faster rhythms in the alpha band, as well as intermittent rhythmic delta activity in the fronto-temporal regions and a slow sharp wave epileptiform discharge with greater electronegativity in C3P3. Laboratory tests revealed: C3 consumed (< 11), CH 50 consumed, C4 normal, FAN positive (AC-4) (AC-8) 1:320, ASLO positive (349.9), proteinuria of 3.8g. Partial urine test with leukocyturia and hematuria. Magnetic Resonance Imaging (MRI) of the skull showed hypointense signals in T1, hyperintense in T2/ FLAIR, without diffusion restriction and gadolinium impregnation, indicating PRES. Treatment for SLE was started in conjunction with rheumatology. Control MRI of the skull identified a significant reduction in the lesions. The patient is currently taking cyclophosphamide and is stable.The condition, characterized by acute diffuse glomerulonephritis, serositis and laboratory tests, indicates a case of reversible posterior encephalopathy triggered by systemic lupus erythematosus. **Discussion:** Posterior reversible encephalopathy syndrome (PRES) is a rare but serious neurological condition often associated with systemic lupus erythematosus (SLE). The patient's initial presentation of headache followed by seizures and MRI alterations indicates PRES. Characterized by rapid onset symptoms such as seizures, headache, and visual disturbances, PRES involves reversible brain edema primarily in the posterior regions. Treatment with corticosteroids and cyclophosphamide,targeting SLE activity and hypertension control, resulted in clinical and imaging improvement. **Final comments:** Investigating the underlying causes of PRES and initiating early treatment are critical to prevent sequelae associated with delayed management of autoimmune diseases, particularly those involving the central nervous system. Early diagnosis and proper management can prevent irreversible neurological damage and improve patient prognosis.

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Task-specific stroke program: developing an extension program to increase exercise intensity

Maria Eduarda Salum Aveiro Henrique[1], Clara Rezende Rocha[1], Danilo Santana Gonsalves[1], Marco Túlio Almeida Lenza[1], Jayne Siqueira Cagliari[1], Pedro Henrique Sousa de Andrade[1], Caroline Rodrigues Osawa[1], Luciane Aparecida Pascucci Sande de Souza[1], Gustavo José Luvizutto[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Maria Eduarda Salum Aveiro Henrique; Email: aveiromariaeduarda@gmail.com.

Background: Stroke is a leading cause of death and disability among adults worldwide. Following a stroke, individuals may experience motor, sensory, and/or cognitive impairments that diminish independence and hinder the performance of core tasks such as sitting, standing, walking, stepping up/down, and reaching/grasping/manipulating objects. Despite numerous studies on post-stroke rehabilitation, the literature lacks an exercise protocol specifically based on the essential biomechanical components of these core tasks. **Objective:** To develop an exercise protocol tailored to tasks relevant to post-stroke rehabilitation within an extension university project. **Methods:** The protocol was designed with exercises focused on essential activities involved in executing core tasks, emphasizing gains in muscle strength and range of motion specific to each task phase. The biomechanical components of each task were defined based on existing literature. The protocol's efficacy was assessed using scales measuring improvements in patient structure and function, activity, and participation. **Results:** Exercises for each task were based on Task-Oriented Training (TOT), known for effectively enhancing upper limb function, mobility, and walking post-stroke. Each core task was segmented by its essential biomechanical components, resulting in a protocol comprising 50 exercises - one for each component with two ways to increase difficulty and two to simplify, facilitating protocol customization based on patient needs. Exercises incorporated external focus techniques to optimize muscular recruitment during execution, using readily available materials to ensure reproducibility across different environments. Dual-task exercises were integrated for cases requiring cognitive rehabilitation alongside motor training. Exercises were documented with photographs and organized into a flowchart indicating activities adaptable to patient-specific restrictions. **Conclusion:** Post-stroke individuals often face challenges that impact independence in essential daily activities. This study developed a neurofunctional rehabilitation protocol based on TOT, aiming to restore autonomy in performing core tasks. Exercises consist of straightforward activities using common objects as external focal points, enabling replication in diverse settings. Efficacy will be evaluated based on improvements in function and structure, activity, and participation using validated stroke assessment scales and questionnaires.

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Teaching tool in neoplasms of the central nervous system: seeking integration between theory and practice

Ana Caroline Siquara de Sousa[1], Bruna dos Santos Serra de Alencar[1], Rafael de Oliveira Telles[1], Stella Coelho Rodrigues[1]

[1]Instituto de Educação Médica, Rio de Janeiro RJ, Brazil.

Correspondence: Ana Caroline Siquara de Sousa; Email: anasiquara@gmail.com.

Background: Central nervous system (CNS) neoplasms are one of the most challenging diseases in various fields of medicine. The national incidence is around 11 thousand cases annually for the three years from 2023 to 2025. This number makes this group of neoplasms the 11th most common in the country, excluding cases of metastases. The study of these neoplasms during graduation is an opportunity to reinforce general pathology concepts and learn about such a specific diagnosis and its classification. Our research group then proposed to create a tool for the histopathological study of CNS tumors, to help undergraduate students become familiar with these diseases. **Objective:** The objectives of the tool are: to promote integration between theoretical concepts of pathology and the practice of the pathologist, with the observation of microscopy images; and to encourage the use of clinical cases in the diagnostic approach of these neoplasms. **Results:** In the first stage, a histopathological review was carried out on the main types of CNS neoplasms, studying clinical and epidemiological aspects and providing microscopy images of these tumors. Subsequently, a brochure was created with other microscopy images and clinical cases, and a board game (trial type) with 40 questions about these neoplasms. The board game must have the participation of at least two players. The theoretical material, then, was used as guidance for the student to prepare for performance in the match. In the next stage of the research, the game will be promoted among Medicine students by members of the research group themselves. This is a topic that students generally have little contact with during the course, despite its importance mainly for understanding the early signs and symptoms of most CNS tumors. Among the reasons that explain the importance of knowledge about these diseases, the following stand out: their high morbidity and mortality; incidence in all age groups, being common in children; and complexity of histopathological and biomolecular aspects. The study of these neoplasms during graduation was an opportunity to reinforce general pathology concepts learn about such a specific diagnosis and its classification, and stimulate reasoning through the study using clinical cases. Furthermore, there was integration with other subjects in the curriculum, such as Human Anatomy, Oncology, and Neurology. **Conclusion:** There are several ways to diversify teaching-learning formats, such as creating conceptual maps, anatomic-clinical case seminars, creating games such as "escape rooms", etc. These strategies may represent new ways of approaching case studies or problem-based learning, bringing new discussion tools to the classroom.

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Tegumentary assessment as a diagnostic clue in demyelinating neuropathies: don't underestimate the tendons!

Támara Larissa Silva Barbosa[1], Isis Maria Lima Cruz[1], André Luis Dias de Figueiredo[1], Vinícius Araújo do Vale[1], Pedro Victor Pinheiro Bezerra Melo[1], Gabryel Felipe Alves de Sousa[1], Rafael Fontes de Carvalho Amorim[1], Ademir Aragão Moura[2]

[1]Universidade Federal do Piauí, Teresina PI, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Isis Maria Lima Cruz; Email: isismarialc@gmail.com.

Case presentation: A 47-year-old male patient sought medical care presenting with episodes of psychomotor agitation, hallucinations, and incoherent speech for the past three months. Family members reported intellectual disability and dysphagia since childhood. Treatment with risperidone 3mg/day and bromazepam 6mg/day was initiated by the psychiatry team. The patient then developed dysarthria, sialorrhea, quadriparesis, weight loss, vomiting, drowsiness, and worsening dysphagia, leading to the discontinuation of medications and neurological evaluation. Upon examination, the patient presented with dysarthria, bilateral Achilles tendon thickening, moderate generalized hypotrophy, bilateral pes cavus, hypoalgesia in the upper and lower limbs, and abolished patellar and Achilles reflexes bilaterally. Subsequently, the following tests were requested: Electroneuromyography (ENMG) of the four limbs, which evidenced mild to moderate, symmetrical, mixed sensorimotor polyneuropathy; cranial magnetic resonance imaging (MRI), which showed hyperintense foci in the periventricular white matter; MRI of the lower limbs, which showed thickening and diffuse signal alteration of the bilateral Achilles tendons with intratendinous deposits suggestive of xanthomas. Genetic testing confirmed the diagnosis by identifying a pathogenic heterozygous variant in the CYP27A gene. **Discussion:** Cerebrotendinous xanthomatosis is an autosomal recessive disease caused by mutations in the CYP27A1 gene, resulting in lipid accumulation in tissues. The reported patient did not present early non-neurological symptoms typical of the disease, such as juvenile cataracts and premature osteoporosis. In line with the literature, psychiatric symptoms usually appear in childhood or adolescence, manifesting as behavioral disorders and learning difficulties, while in the advanced stage of the disease, it progresses with dementia or complex neuropsychiatric syndromes. Neurological examination findings, such as quadriparesis, Achilles tendon thickening, pes cavus, and abolished reflexes, were commonly described in previous studies. As for the test results, the ENMG showed the expected result, as polyneuropathy is a characteristic finding of this disease, which progresses with demyelination; the literature describes characteristic MRI with bilateral lesions in the dentate nucleus, cerebellum, and mild white matter lesions; MRI of the lower limbs usually shows fusiform thickening and heterogeneous signals of the Achilles tendons. In Brazil, there is no drug therapy for the disease. The primary therapeutic approach to delay disease progression is multidisciplinary follow-up. **Final comments:** Progressive and irreversible neurological involvement is the primary manifestation of the disease. To delay disease progression, the main therapeutic option is multidisciplinary follow-up. Early diagnosis is essential for the patient's quality of life.

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Teleconsultations specialized in difficult diagnoses: Whipple's disease

Lizandra Lavinia Santos de Albuquerque Menezes[1], Carlos Eduardo Aliatti Mantese[2,3], Rhaná Carolina Santos[3], Aline Oliveira dos Santos Meneses[3]

[1]Serviço Médico Odontológico, Capim Grosso BA, Brazil.
[2]Hospital Sírio Libanês
[3]Universidade do Vale do Rio dos Sinos, São Leopoldo RS, Brazil.

Correspondence: Lizandra Lavinia Santos de Albuquerque Menezes; Email: lizandramenezes18.2@bahiana.edu.br.

Case presentation: A 48-year-old man, of mixed race, residing in the countryside of Bahia, without comorbidities, attended a consultation at the basic health unit in June 2023 with cervical dystonia, dysphonia, slowed gait, and unquantified, progressive weight loss over the past 5 months. The dystonia presented as involuntary contraction of the bilateral platysma muscle, more pronounced on the left, associated with jaw clenching, worsening during stress. He reported depressed mood, insomnia, and, on physical examination, absence of horizontal eye movement in abduction, no pupillary changes, and difficulty keeping eyes closed. He had additive polyarthralgia and poorly characterized diarrhea, which improved spontaneously 10 months earlier, investigated in tertiary care but lost follow- up. Laboratory tests revealed hypochromic, microcytic anemia with plateletosis, albumin/ globulin inversion, and slightly elevated transaminases. A cranial MRI showed gliosis and compensatory ventricular system ectasia. Among the previous tests, which he lost follow-up on, included a chest and abdomen CT scan from August 2022 showing mediastinal, mesenteric, and retroperitoneal lymph node conglomerates suggestive of lymphoproliferative disease. There was also a biopsy of a sentinel lymph node showing reactive inflammatory histiocytosis. **Discussion:** Due to multisystemic alterations, dystonia was considered a guiding symptom, and teleconsultations with neurology have been conducted since September 2023, raising the hypotheses of Wilson's disease, Whipple's disease, and paraneoplastic syndrome. Wilson's disease was an important differential diagnosis, as changes in pigment were observed in the descemet membranes during the slit-lamp examination, consistent with corneal pigment deposits, suggestive of Kayser-Fleischer ring (K-F). Among the differentials for the ring, there is the pseudo-K-F ring, present in hepatobiliary diseases, cholestasis, and exposure to metals. The patient in question has a work history involving aluminum, likely the source of the pigment deposit. By correlating the case with the reactive histiocytosis in lymph node clusters, the absence of alteration in serum/urinary copper and ceruloplasmin, despite the ophthalmological finding, led to the hypothesis of Wilson's disease being ruled out, with Whipple's disease being a possibility to consider. As the condition progressed, the patient was referred to Salvador via the state central regulation system, where gastric and duodenal biopsies in October 2023 were positive for Tropheryma whipplei. Antibiotic therapy was initiated with favorable evolution. **Final comments:** Whipple's disease, although rare, should be remembered in differential diagnoses as it can be a lethal infection if not treated properly. Currently, the patient continues to be under basic and tertiary care, showing improvement in symptoms, with persistent cervical dystonia and resolution of laboratory abnormalities.

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Temporal analysis of stroke morbidity in Brazil. What has changed in two decades?

Sávio Roberto Silva Costa[1], Mayla de Carvalho Zavarise[1], Chan David Ribeiro[1], Danusa Neves Somensi[1], Mariângela Moreno Domingues[1], Emanuel de Jesus Soares de Sousa[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Sávio Roberto Silva Costa; Email: saviorsc20@gmail.com.

Background: Stroke is a neurological condition resulting from the interruption of blood flow to the brain, which causes cell damage and brain dysfunction. This condition is one of the leading causes of morbidity and mortality in Brazil and worldwide, resulting in motor, sensory and intellectual deficits that significantly affect patients' quality of life and generate substantial economic costs for healthcare systems. **Objective:** To compare variables related to stroke and analyze their possible changes in two time series. **Methods:** This is a cross-sectional, observational study with a quantitative approach whose data were collected from the Department of Information and Informatics of the Unified Health System (DATASUS), in the hospital morbidity section (SIH/SUS). Variables related to region, age, sex and race were analyzed in two different time series (TS), TS1 (2004-2013) and TS2 (2014-2023). Bioestat 5.3 software was used to statistically analyze and validate the data. A p-value of less than 0.05 was considered statistically significant for this study. **Results:** It was observed that morbidity due to stroke in these two STs showed an increase of 33.62%. As for the regional aspects, all regions showed a significant increase, with the Southeast region having the highest absolute number of morbidities in TS2 with n = 676,247, but showing the lowest increase between the two TSs (21.48%). In addition, the Northeast region showed the largest increase between the two series (54.99%). In terms of age group, all categories showed an increase, especially the under 9 age group, with an increase of 72.19%. In addition, the elderly group (60 years old or more) showed an increase of 35.59% in relation to TS1, with the 60 to 69 age group standing out, which recorded a significant increase of 42.39%. Analyzing gender among the TS, there was an increase in both sexes, 35.69% in males and 31.42% in females, and the predominance of males increased from 6.16% in TS1 to 9.61% in TS2. Finally, with regard to race, in ST2 there was a predominance of brown people with 602,680 cases, followed by white people with 530,523 cases. There was no data on race in ST1. **Conclusion:** Overall, the data indicates a significant increase in stroke morbidity in Brazil between the time series studied, with an increase in variables related to region, age group and gender. This highlights the need to develop more effective public health strategies to identify and mitigate risk factors in the populations most vulnerable to stroke.

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Temporal aspects of hospitalizations due to tuberculous meningitis in Brazil

Annayza de Oliveira Bessa[1], Deborah Araujo Silva[1], Rynele Almeida de Fonseca[1], Larissa Bianca Borsone[1], Amanda Oliva Spaziani[1], Francieli Gomes Lima[2], Gustavo Henrique da Silva[3], Luis Carlos Spaziani[4], João Carlos Bizinotto Leal de Lima[1], Vanessa Maria Gonçalves de Souza[1]

[1]Universidade Brasil, Fernandópolis SP, Brazil.
[2]AME Fernandópolis, Fernandópolis SP, Brazil.
[3]UPA Santa Fé do Sul,Santa Fé do Sul SP, Brazil.
[4]Fundação Educacional de Fernandópolis, Fernandópolis SP, Brazil.

Correspondence: Annayza de Oliveira Bessa; Email: annabessa01@gmail.com.

Background: Tuberculosis, caused by the bacterium Mycobacterium tuberculosis, is a disease that can affect the lungs and other sites in the body and is the second leading cause of death globally. It is characterized by a subacute or chronic inflammation of the meninges due to the dissemination of *M. tuberculosis* into the subarachnoid space. This form of tuberculosis typically results from a complication of pulmonary tuberculosis and usually manifests within the first six months after an individual is infected. The diagnosis of tuberculous meningitis is often delayed due to the limited efficacy or availability of laboratory techniques, and the treatment is based on protocols for pulmonary tuberculosis. **Objective:** To evaluate the profile of hospitalizations due to tuberculous meningitis in Brazil between the years 2019 and 2023. **Methods:** Data collection was performed using a retrospective longitudinal study design with a quantitative descriptive approach. Data were grouped by Brazilian regions, type of care, sex, age group, and race/ethnicity. Statistical inferences were conducted using BioEstat 5.3 software, employing the paired T-test for related samples to compare groups. Descriptive statistics, measures of central tendency, and dispersion were used to present the data. **Results:** Between 2019 and 2023, there were 1,629 hospitalizations for tuberculous meningitis in Brazil. A majority, 65.56% (n=1,068), were male (p<0.0017), and 46.35% (n=755) were of mixed race (p=0.0371). The age group most affected was 30-39 years, accounting for 20.38% (n=332; p<0.0001) of the cases. The Southeast region had

the highest concentration of cases (n=770), corresponding to 47.27% (p=0.0011), with an average annual treatment cost of R$ 721,076.02. Mortality was also highest in the Southeast, with 53.89% (n=104) of deaths (p=0.0024) and a mortality rate of 24.50 (p=0.1933). The number of hospitalizations peaked in 2023, accounting for 24% (n=391) of the total, compared to only 17.37% (n=283) in 2021. Most hospitalizations, 87.78% (n=1,430; p<0.0001), were classified as urgent. **Conclusion:** Hospitalizations due to tuberculous meningitis are predominantly concentrated in the Southeast region of Brazil, with higher mortality rates and associated treatment costs. Most cases occur in males aged 30-39 years of mixed race, with most hospitalizations requiring urgent care.

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Temporal trend of CNS neoplasms mortality rate in Brazil between 2001 and 2022

Victor Santos Vilares[1], Érica Otoni Pereira Miranda[1], Guilherme Rocha Cardoso[1], Lukas Santos Freire[1], Maria Fernanda Aragão Andrade[1], Rebeca Gabrielle Almeida Maciel[2], Priscila Santos Oliveira[1], Emmanuel Santos Trindade[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.
[2]Universidade Federal da Bahia, Salvador BA, Brazil.

Correspondence: Priscila Santos Oliveira; Email: priscilaoliveira35001@gmail.com.

Background: Central Nervous System (CNS) tumors are a heterogeneous group of neoplasms. Although some types have a high mortality rate, prognosis and evolution depend on their cellular origin, time of diagnosis and location. The most prevalent are gliomas, meningiomas and neurinomas. **Objective:** To analyze the temporal trend of CNS neoplasms mortality rate in Brazil between 2001 and 2022. **Methods:** Death data was collected from the Mortality Information System (SIM) through DATASUS, including those from malignant neoplasm of meninges (C70), brain (C71), spinal cord, cranial nerves and other parts of central nervous system (C72), stratified by sex, ethnicity and age group. A joinpoint regression model was used for the temporal analysis. **Results:** From 2001 to 2022, the average annual mortality rate (MR) from CNS neoplasms was 3.59/100,000 inhabitants in the general population, with higher rates in males (3.82/100,000 inhabitants), in line with the literature regarding the higher prevalence and worse prognosis in males. The white ethnicity was the one with the highest MR (4.78/100,000 inhabitants), in compliance with studies showing a higher prevalence of glioblastoma, one of the most aggressive CNS neoplasms, among whites. In regard to age groups, those aged 60 or over had the highest MR (14.95/100,000 inhabitants), which reinforces the literature, since glioblastoma is more incident from the 5th decade of life onwards. In the general population there was an increasing trend in MR between 2001 and 2022 (AAPC: 1.80 [1.50; 2.10]; p<0.001), with a more pronounced increasing trend from 2001 to 2013 (APC: 2.58 [2,21; 2.95]; p<0.001) than from 2013 to 2022 (APC: 0.78 [0.22; 1.34]; p:0.009). A similar trend was identified in both sex. In addition, whites and browns had the highest MR in 2022, with 5.72 and 2.99, respectively. Between 2001 and 2022, whites (AAPC: 2.19 [1.71; 2.66]; p<0.001) and browns (AAPC: 4.17 [3.70; 4.64]; p<0.001) showed an increasing trend in MR; blacks, however, had a stationary trend (AAPC: 1.78 [-1.32; 4.98]; p:0.26). Those aged 60 or over presented an increasing trend in MR (AAPC: 0.92 [0.39; 1.45; p<0.001) from 2001 to 2022, with a initial increasing trend from 2001 to 2015 (APC: 2.06 [1.56; 2.56]; p<0.001) followed by a stationary trend (APC: -1.32 [-2.68; 0.06]; p:0.06). Those aged between 50 and 59 years showed a stationary trend (AAPC: 0.19 [-0.20; 0.57]; p: 0.32). The other age groups presented decreasing trends in MR. **Conclusion:** Two epidemiological patterns in CNS neoplasms mortality rate were identified from 2001 to 2022: an increasing trend between 2001 and 2013, followed by a stationary trend, which could be partly explained by the improvement in the notification system and diagnosis. Besides, it was noticed a decreasing trend in mortality rate in younger individuals, which could be related to better diagnosis and treatment in those patients.

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Tenecteplase versus alteplase in thrombolysis for ischemic stroke

Luiz Mateus Alves de Góis[1], Edla de Andrade Alves da Silva[1], Marcela Carvalho do Nascimento[1], Felipe Hermes Amorim da Silva[1], Thayna Costa Tenório Ribeiro Neves[1], Vinicius Rodrigues Ramos[1], Giovana Gonçalves de Souza[1], Ana Beatriz Benassi de Oliveira[1], Tainá Torres Pedro[1], Analuiza Silva Tenório Luna Sarmento[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.

Correspondence: Luiz Mateus Alves de Góis; Email: luiz.gois@famed.ufal.br.

Background: Stroke is the second leading cause of global mortality, with 84.4% being of ischemic origin (IS). Currently, alteplase is considered the first-line treatment for patients within the therapeutic window for thrombolysis. However, recent studies have raised the possibility of using a modified variant of alteplase (rtPA), tenecteplase (TNK), citing advantages such as a rapid onset of action. **Objective:** To analyze the efficacy and safety profiles of tenecteplase compared to alteplase in thrombolysis for patients with IS. **Methods:** For this systematic review, the descriptors «tenecteplase (OR) tissue plasminogen activator» combined (AND) with «alteplase» and (AND) «ischemic stroke» were used in the PUBMED and BVS databases. The filters «last 10 years» and «clinical trial" were used. After removing duplicates and thoroughly reading 27 randomized clinical trials, 13 were selected for this article. Exclusion criteria were: addressed individuals under 18 years old; included patients outside the 4.5-hour window for thrombolysis; were not freely available. **Results:** Data from 16,699 individuals were evaluated, among whom 65.57% were treated with rtPA and 34.43% with TNK. The minimum dose of TNK evaluated was 0.1 mg/kg, and the maximum was 0.4 mg/kg, the 0.25 mg/kg bolus dose was evaluated in all the analyzed studies. The rtPA maintained the standard dose of 0.9 mg/kg. 12 of the 13 studies involved individuals over 18 years old, and one study focused on individuals over 80 years old. The main outcomes were a score of 0-1 on the Modified Rankin Scale (mRs) within 90 days or a reduction of 4 or more points on the NIHSS within 24 to 72 hours, in addition to safety outcomes such as symptomatic intracranial hemorrhage (ICH) and 90-day mortality. The neurological improvement observed with TNK use slightly surpassed that obtained with rtPA in all studies that used the 0.25 mg/kg dose. Symptomatic ICH within 48 hours followed this trend, being lower with TNK than with rtPA in 44.4% of the cases, as well as 90-day mortality. **Conclusion:** The efficacy of TNK was similar to that of rtPA regarding the primary outcomes, with a statistically insignificant difference in acute IS within the 4.5-hour window from symptom onset. There were no statistically significant differences in the proportions of ICH within 90 days after symptom onset or in mortality rates within the 3 months following therapy. Thus, the results support the implementation of TNK 0.25 mg/kg as an alternative thrombolytic agent to the standard treatment with rtPA 0.9 mg/kg.

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Ten-year neurological IgG4-related disorder: a case report presenting with mild seizures evolving with late inflammatory brain tumor

Polyana Favero Ferreira Caetano[1], João Henrique Fregadolli Ferreira[1], Lívia Almeida Dutra[1]

[1]Instituto do Cérebro, São Paulo SP, Brazil.

Correspondence: Polyana Favero Ferreira Caetano; Email: polyanafaveroc@hotmail.com.

Case presentation: A 43-year-old woman had focal motor seizures for the past 10 years, characterized by tonic movements in the right arm, progressing to bilateral tonic-clonic seizures. Initial brain MRI and EEG were normal. Despite treatment with valproate, she continued to have occasional mild seizures. In 2022, she started with focal seizures involving the right leg. A new MRI showed a Gd+ mass lesion in the left precentral gyrus and brain biopsy showed perivascular lymphoplasmacytic infiltrate with storiform fibrosis. Immunohistochemistry was negative for CD1a, and positive for S100 with IgG4+ plasmocytes < 40 and IgG4/IgG > 40%. Whole body CT scans revealed pulmonary involvement with ground glass infiltrates. After 4 months, she developed amenorrhea and reduced FSH and LH levels, normal hypophysis on MRI. She was treated with azathioprine, steroids, levetiracetam, topiramate and clobazam. After 12 months, she developed abnormal movements in the right foot, and was admitted for further investigation. Physical examination revealed mild weakness and sensory loss in the right inferior foot with hyperreflexia. Brain MRI showed enlargement of the initial lesion, indicating a recurrence of IgG4-related disease. VEEG was normal and ENMG showed upper motor neuron discharges. She received IV methylprednisolone for 5 days and was started on rituximab. **Discussion:** IgG4-related disease (IgG4-RD) is a systemic immune-mediated fibroinflammatory disease with variable clinical manifestations, depending on the affected organ. Most common neurological manifestations are hypertrophic pachymeningitis and hypophysitis. In this case neurological manifestations had a chronic course of ten years, with slowly progressive symptoms and an initial normal brain imaging, resembling epilepsy with unprovoked seizures. Although brain MRI is subject to specificities of equipment and imaging acquisition, this case indicate that some IgG4-RD patients may present functional neurological impairment rather than structural abnormalities. The clinical pituitary involvement without sella turcica abnormalities also exemplifies this hypothesis. Moreover, this report indicates that the disease may remain silent for a long period and present with a late severe flare. Another important goal learning of this case is the interpretation of brain biopsy results. The findings of perivascular lymphoplasmacytic infiltrate with fibrosis should prompt a focused immunohistochemical study for immune cells as well as IgG4. **Final comments:** This case illustrates the potential challenges of IgG4-related disease neurological manifestations. Patients may present with initial normal brain MRI, the slow progression of the disease may delay the diagnosis and the disease may remain silent for a long period of time. Neurologists should be able to correct interpret brain biopsy results and to ask for complimentary investigation when needed.

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Thalamic deep brain stimulation for treatment of refractory epilepsy: a systematic review and meta analysis

Emanuela Freire Caetano Davi[1], Leonardo José Rodrigues de Araújo Melo[1], João Pedro de Oliveira Gouveia Marcotti[1], Alan Duarte Braz[1], Wagner Patrik Sousa Silva[1], Rodrigo Fagundes da Rosa[1], Caio Levi Mendes Costa[1], Leonardo Elias Araujo dos Santos[1], Francisco Duque de Paiva Giudice Junior[1], Mariana Borges Falcão Costa[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Emanuela Freire Caetano Davi; Email: emanueladavi@alu.ufc.br.

Background: Among the target areas for treatment of refractory epilepsy with Deep Brain Stimulation (DBS), the thalamus was shown to have great importance in propagation of seizure activity in experimental evidence, raising the necessity to review its impacts in clinical context. **Objective:** To evaluate the efficacy of DBS in the thalamus of patients with refractory epilepsy on the decrease of seizure frequency (SF) in the existing literature. **Methods:** This systematic review was executed according to the PRISMA 4 checklist. The search on the databases PUBMED, Scielo and EMBASE with "Deep Brain Stimulation" AND "epilepsy" AND "Thalamus", with 41 results. Inclusion criteria were controlled trials in English and Portuguese that evaluated the efficacy of thalamic DBS on the decrease of SF on human patients. The RoB2 guidelines were used to assess risk of bias, and the GRADE tool to assess certainty of evidence. The effect measures analyzed for outcome were the median percentage of SF decrease from baseline, responder's rate and value p, evaluated in meta analysis with Jamovi. The software Rayyan was used to automate exclusion and 12 studies were selected for assessment and 3 were discarded due to data duplicity or high risk of bias. **Results:** Across the 9 studies reviewed, data of 356 patients, mean age 32.4, was analyzed in follow-up periods varying between 3-84 months. The change of monthly SF from baseline while on active stimulation was evaluated using seizure diaries registered by patients or caretakers. FIAS was the most common type of seizure in this population, and the temporal lobe was the most common location of seizure origin. In studies that compared location subgroups, temporal lobe seizures had the highest decrease on SF of 70%. The decrease of SF was achieved and statistically significant by the population in follow-up studies, with the median percentage of decrease varying between 33% to 75% and overall mean of 53.9% of decrease (CI, 63, 44.7), with reported rates of 69% (CI, 43, 96.5) reduction in 5 years. 48% of the total population reached the responder rate of ≥ 50% SF reduction in comparison to baseline. Patients with intellectual disabilities (42 patients, 11.7%) present less efficacy in decrease of SF, ranging across 25% on studies with this population, though this group presented higher baseline SF. Notably, 4 trials reported subjects with improvement of SF after surgery but previous to stimulation. **Conclusion:** This systematic review indicates that the stimulation of the thalamus for the treatment of refractory epilepsy is an effective treatment in support of antiepileptic medication with objective decrease of SF in most populations. On study limitations, it's noted the use of Antiepileptic drugs are a possible confounding, divergence in inclusion criteria for participants, increasing standard deviation, and the sample was limited in the current literature. Due to this, the GRADE level of certainty was assessed as III.

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The 'rosetta' sign: a peculiar gyral conformation of type II focal cortical dysplasia

Rafael Paglioli[1], Eliseu Paglioli[1], Francine Hehn[1], William Martins[1], Ricardo Paganin[1], Victoria Bizzi Schvartzman[1], André Gus[1], Natalie Donida[1], Bibiana Thome[1], André Palmini[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Victoria Bizzi Schvartzman; Email:Bizzivictoria@gmail.com

Focal cortical dysplasia (FCD) is a frequent pathology underlying an epileptic focus. The lesions can be subtle and despite much better imaging, clinical studies with 3T magnetic resonance imaging (MRI) still report a significant number of patients with histologically proven FCD that are 'MRI-negative'. Here, we report a series of patients with histology-proven type II FCD with a distinct, peculiar, 3D pattern of cortical gyration resembling what in botany is called a rosetta. M: Were evaluated 20 patients operated for refractory seizures due neocortical epilepsies over two years, in whom pathology revealed type II FCD. All had daily or weekly socially disabling attacks and presurgical evaluation, including clinical epileptological investigation, video-EEG scalp recordings, 3T MRI and neuropsychological testing. Following visual analysis of the 2-D MRI data by two neuroradiologists, a Visual Manipulation Toolkit for NeuroImages (VMTN) software was used to provide 3-D images of the sulci and gyri, at sequential sections, starting at the outermost cortical surface. After integration of presurgical data and delineation of the epileptogenic zone, patients underwent stepwise resective surgery, fine-tuned by electrocorticographic recordings with 16 individual electrodes that allowed acute probing of electrical activity from the surface, lateral banks and depths of gyri in sequential fashion during resection. Resected tissue was stained with hematoxylin-eosin and immunostained with neuronal nuclear antigen (NeuN), vimentin, GFAP and Olig2. FCD II was diagnosed by dyslamination on NeuN and the presence of large, dysmorphic neurons, accompanied or not by balloon cells. R: In 10 patients (50%), reformatted 3D images using VMTN software showed that gyri had a roughly circular arrangement radiating from a deep sulcus - a depressed, void center (the '3D image of the dimple') - the whole set akin to a rosetta of leaves. In all, dysplastic cortex was found in the gyral banks surrounding the depressed center and highly epileptogenic electrocorticographic discharge patterns were recorded acutely from variable portions of the gyri emanating from the depressed center. In 5 of these patients, conventional 2D-images showed discrete dysplastic lesions, variably characterized by thickened cortex, increased signal and blurring of the cortical-subcortical transition. In the other 5, images were initially read as normal and finding the rosetta pattern was crucial to confirm localization, supplemented by VEEG and semiological data. The 10 patients in whom the rosetta pattern was not seen had visible, large dysplastic lesions. C: The rosetta sign showed to be a reliable indicator of dysplastic epileptogenic cortex often not otherwise identified in many patients. Using the software and paying attention to the rosetta pattern are important assets to surgical decision in many patients with refractory neocortical seizures, obviating the need for costly and invasive studies.

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The advances in monoclonal antibody research and its influence on Alzheimer's early prognosis

Nathan Chinelato Afonso[1], Victoria Lima Marques[1], Carlos Eduardo Fernandes Poppi[1], Júlia Carolina Fernandes Poppi[1], Otávio Antigo Orlandini[1], Amanda Piveta Nicodemo[1]

[1]Centro Universitário Ingá, Maringá PR, Brazil.

Correspondence: Nathan Chinelato Afonso; Email: nathan.afonsoch@gmail.com.

Background: Alzheimer's disease (AD) is characterized as chronic neurodegenerative complex and multifactorial pathology; being know for it's progressive loss of memory, cognitive functions, as well as synaptic dysfunction. Statistical data predicts an increase in the number of AD patients in America, surpassing 13.8 million cases by 2050. In addition, the observed pathological characteristics are, high levels of amyloid-β (Aβ) proteins composing plaques and hyperphosphorylated tau (p-tau), aggregated intracellularly. Recently findings review positive results with therapies with monoclonal antibodies, which act on the reduction of beta-amyloid Plaques. **Objective:** To demonstrate the efficiency of specific antibodies in the reduction of cerebral amyloid plaques accompanied by a reduction in clinical decline, taking into account that it brings promising expectations in the delay of the disease. **Methods:** This is an integrated review by means of a literature review in the PubMed and SciELO databases. The established descriptors were "lecanemab", "alzheimer" and "monoclonal antibody" connected by the Boolean operator and, published in English, during the last 5 years (2019-2024). A total of 140 articles were found, excluding those that did not involve the proposed theme, repeated in the databases, or addressed only one aspect or specific population, and 14 were initially chosen to be part of the research. **Results:** The studies show that patients affected by Alzheimer's disease present cognitive impairment and dementia at an early stage, due to mainly the neurodegeneration created by the amyloidogenic pathway, being a product of the successive cleavage of the amyloid precursor protein, which is extracellularly deposited in the parenchyma and cerebral vasculature. Monoclonal antibodies (mAbs), such as lecanemab, represent an approach based on high-affinity ligands in Aß plaques, such action promotes removal of aggregates by phagocytosis, mediated by receptors, altering the course of the disease. Concomitantly, randomized controlled trials using mAbs have shown that reduction of β-amyloid protein can be achieved in 3 months of treatment, with clinical efficacy in 6 months. According to the tests carried out, one of the most common side effects presented by this therapy occur in ARIA-E and ARIA-H, edema and hemorrhage, respectively, affecting only 12.6% of patients. At the same time, research has shown relevant improvements in biomarkers of the Tau protein (a potentially fundamental molecule in the pathogenesis of neurodegenerative diseases). **Conclusion:** In summary, the data obtained by clinical tests represent an effective way to improve the manifestations of symptoms, delay or even interrupt the progress of AD. Essentially, the therapeutic approach of monoclonal antibodies in pathology should be further explored.

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The association between statin use and dementia risk: an update meta-analysis of observational studies

Fernando Luiz Westphal Filho[1], Artur Menegaz de Almeida[2], Paulo Roberto Moss Lopes[1], Vitor Kendi Tsuchiya Sano[3], Fernanda Moraes Tamashiro[4], Francinny Alves Kelly[5], Pablo Vinícius Silveira Feitoza[1]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[3]Universidade Federal do Acre, Rio Branco AC, Brazil.
[4]Universidad de Buenos Aires, Buenos Aires, Argentina.
[5]Instituto de Cardiologia Dante Pazzanese, São Paulo SP, Brazil.

Correspondence: Fernando Luiz Westphal Filho; Email: fernandowestphal1@gmail.com.

Background: The potential impact of statin therapy on cognitive function and the risk of developing dementia has been a topic of considerable debate. Primarily prescribed for their lipid-lowering effects and cardiovascular benefits, statins have been hypothesized to exert neuroprotective effects due to their anti-inflammatory and antioxidant properties, as well as their ability to modulate cholesterol metabolism in the brain. Observational studies have often reported a lower incidence of dementia among statin users compared to non-users. However, the results from randomized clinical trials have been less consistent, with some studies failing to demonstrate a significant benefit of statins on cognitive outcomes. **Objective:** To provide a current and thorough review of the association between statin use and the incidence of dementia. **Methods:** MEDLINE, Embase and Cochrane databases were systematically searched until May 20, 2024. Observational studies that evaluated the association of statins and the development of dementia were included. We computed the Hazard Ratios (HR) of developing dementia in the statin group, while the reference was the non-statin group.

Additionally, subgroups' HRs were collected for further analysis. Data were analyzed using the Mantel-Haenszel method and 95% confidence intervals (CI). Heterogeneity was assessed using I^2 statistics. Standard Error and logHR were calculated through Review Manager 5.4.1 and statistical analysis was performed using R, version 4.3.2. **Results:** This meta-analysis included 34 observational studies evaluating the effect of statin use on the risk of developing dementia. The overall analysis demonstrated a statistically significant reduction in dementia risk among statin users compared to non-users (HR 0.87; 95% CI 0.83 to 0.90; I^2=86%; P<0.001). Among the analysed subgroups, statistically significant reduction was identified in the Lipophilic (HR 0.84; 95% CI 0.78 to 0.91; I^2=83%; P<0.001), but not in the Hydrophilic group. Atorvastatin and Rosuvastatin were the only specific statins that showed statiscally relevant reduction in dementia risk (HR 0.88; CI 0.83 to 0.94; I^2=86%; P<0.001 and HR 0.71; CI 0.61 to 0.83; I^2=91%; P<0.001, respectively). Furthermore, statins reduced the risk among Diabetes Mellitus patients (HR 0.87; CI 0.85 to 0.89; I^2=38%; P<0.001). **Conclusion:** In this comprehensive systematic review and meta-analysis, statin use is associated with a significantly reduced risk of developing dementia. Subgroup analysis revealed that rosuvastatin is the most effective in reducing this outcome. These findings support the potential neuroprotective role of statins, highlighting their importance in dementia prevention strategies.

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The association between the incidence of encephalitis and dengue fever in Brazil: an epidemiologic analysis of the notifications between 2013 and 2023

Ana Luiza Rossetto[1], Ana Paula Stofel Fernandes[2], Gabriela Maria Vicente de Melo[3], Lucas Aires Alencar Ferreira[4], Millena Ribeiro Bessa[5], Paulo Henrique Oliveira Santos[6], Renato do Nascimento de Martine[2], Tarcila Fontes Lucena[7]

[1]Universidade de Marília, Marília SP, Brazil.
[2]Universidade Federal do Espírito Santo, Vitória ES, Brazil.
[3]Universidade Federal do Rio Grande, Rio Grande RS, Brazil.
[4]Universidade Estadual de Roraima, Boa Vista RR, Brazil.
[5]Faculdade de Petrolina, Petrolina PE, Brazil.
[6]Universidade Nove de Julho, Mauá SP, Brazil.
[7]Universidade Federal do Rio Grande do Norte, Natal RS, Brazil.

Correspondence: Ana Luiza Rossetto; Email: rossettoanaluiza23@gmail.com.

Background: The dengue fever is an arbovirosis with the major number of infections in Brazil, defining a public health problem in the country. We can estimate around 390 million infections occurring annually. Of those, between 1-5% show some compromise with the nervous system like encephalitis, an inflammatory condition of the brain generated by an infection of the central nervous system (CNS). **Objective:** Analyze the quantitative relation between the outbreaks of dengue fever with the increase the amount of cases of viral encephalitis in Brazil. **Methods:** Studies with ecological design, data extracted from the Departamento de Informática do Sistema Único de Saúde (DATASUS), in which were analyzed the number of hospitalizations by encephalitis in the months with the major incidence of hospitalization by dengue fever in Brazil in the year of 2023. The data were analyzed by descriptive statistics. **Results:** In Brazil, there were 432,423 hospitalizations by dengue fever between the years 2013 and 2023, being around 239,632 (55.4%) in the months of march, april, may and june. In this same period, 21,643 hospitalizations by encephalitis were notified, being 6,680 (30.8%) in the months of major incidence of hospitalizations by dengue fever. There was an increase of 30% in the cases of encephalitis in comparison with the months of low incidence of the disease. Furthermore, the age range that was affected the most during the outbreaks of dengue fever was between 20 to 40 years old, representing 45% of the cases in this period. **Conclusion:** Lastly, the results of this study suggest a significant correlation between the outbreaks of dengue fever and an notable increase in the cases of encephalitis, especially between young adults. This highlights the necessity of executing preventive measures and strengthening the epidemiological vigilance for a better understanding and control of these diseases in a more efficient way in the Brazilian territory.

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The benefits of physical training on physical and cognitive performance, quality of life and activities of daily living in patients with Alzheimer's disease: a systematic review and meta-analysis

Bernardo de Faria Moraes[1], Cândido Celso Coimbra[1], Cauã Viana Fernandes de Sá Leitão[1], Amanda Gonçalves Duarte[1], Renata Campos Correa dos Santos[1], Gabriel Moraes de Oliveira[1], Gabriel André Pedral Diniz Leite[1], Jair Antônio Bessa da Silva[1], Marcos Vinícius Gonçalves da Silva[1], Fernando Augusto Barcelos Andrade[1]

[1]Universidade Federal de Minas Gerais, Belo Horizonte MG, Brazil.

Correspondence: Bernardo de Faria Moraes; Email: bernardofmoraes@outlook.com.

Background: Alzheimer's disease is the most common type of dementia in the elderly, being characterized by slow and gradual impairment of mental function. It is a progressive neurodegenerative disorder that manifests itself as damage to cognition, memory, judgment, thinking and motor skills. Being aware of the already known benefits of physical activity in the elderly, this practice needs to be considered as a possible strategy for improvement of prognosis in this disease. Considering the increase in the number of cases due to population aging, the potential benefits of combined physical training are of great importance and need to be further elucidated. **Objective:** Therefore, this study aims to identify the possible benefits of physical training on cognitive performance, physical performance, quality of life and activities of daily living in patients with Alzheimer's. **Methods:** The bibliographic research was conducted according to the guidelines outlined in PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analysis). The following electronic databases were used: PubMed, Web of Science and Embase. The systematic search was performed without date and language restrictions. The research used the following descriptors as a search strategy: Alzheimer's disease, exercise, aerobic training, strength training, multimodal training, cognitive function, functional independence and neuropsychiatric symptoms. After the selection process, 113 studies were included in the systematic review and 26 studies (82 trials and 2231 individuals) were included in the meta-analysis. The studies were grouped according to the impacted Alzheimer's axis and the duration of combined physical training: cognitive performance, physical performance, quality of life and activities of daily living up to 3 months and up to 6 months of training. **Results:** The practice of combined physical training for up to 3 months had a moderate effect on cognitive performance, by Mini Mental State Examination (SMD: 0.34; 95% CI: 0.07-0.61; p=0.016), physical performance (SMD: 0.75; 95% CI: 0.43-1.06; p=0.000) and quality of life (SMD: 0.40; 95% CI: 0.17-0.63 ; p=0.567) of the patient. However, activities of daily living remained stable (SMD: -0.10; 95% CI: -0.31-0.12; p=0.621). The combined physical training lasting from 4 to 6 months did not alter cognitive performance (SMD: 0.01; 95% CI: -0.22, 0.24; p=0.397) or activities of daily living (SMD: 0.00; 95% CI: - 0.12, 0.12; p=0.444), but there was a moderate effect on physical performance (SMD: 0.51; 95% CI: 0.23-0.79; p=0.000). **Conclusion:** The practice of physical exercise for up to 3 months generates positive impacts on the cognitive performance, physical performance and quality of life of individuals with Alzheimer's, with longstanding improvements in physical capacity.

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The challenger thalamic anatomy and clinical spectrum of posterior circulation stroke: a anteromedial bitalamic ischemic presentation by artery of percheron infarct

Renato Dumbá Monteiro de Castro[1], Taís Luise Denicol[1], Fernanda Nicoli Broch[1], Alexandre Laisson Prado Taschetto[1], Juan Sebastian Sanchez Léon[1], Amanda Cipriano Torquato[2], Carolina Matté Dagostini[1], Julian Gomez Gonzalez[1], Diogenes Guimaraes Zan[1], Marlise de Castro Ribeiro[1]

[1]Santa Casa de Porto Alegre, Porto Alegre RS, Brazil.
[2]Universidade Vale do Rio Doce, Governador Valadares MG, Brazil.

Correspondence: Renato Dumbá Monteiro de Castro; Email: renato.castro@ufcspa.edu.br.

Case presentation: A 68-year-old man, metabolic syndrome (hypertension, dyslipidemia, obesity) unsatisfactory medication control, admitted to the emergency department after 5 hours of sudden behavioral change and partial depression of consciousness, impaired verbal response, confusing but understandable words, temporal and spatial disorientation followed by hypersomnia, doesn't remain alert with eyes open for more than 10 seconds even with vigorous stimulation. There was response to commands with difficulty, overcoming gravity and supporting the four limbs with preserved strength, despite impairment of executive functions and attention, scoring 10 according to the Glasgow Coma Scale (GCS), National Institutes of Health Stroke Scale (NIHSS) = 2 and Head Impulse Test (HINTS) non suggestive for central nervous system damage. Other focal neurological deficits were not identified and cranial nerve examination was normal. Laboratory tests and liquor analysis ruled out infections and systemic metabolic changes. The initial cranial and neck CT angiography showed microangiopathies, diffuse atheromatous stenosis minor 50% with parietal irregularities, worse in the right vertebral artery and 0.8 cm subdural hematoma(SDH) on left cerebellar tentorium. Furthermore, complementary magnetic resonance showed a well-defined area suggestive of recent bitalamic anteromedial ischemia. Conservative therapy was followed for SHD and ischemic stroke. Etiological screening identified heart failure with a slightly reduced fraction, 46% with diffuse hypokinesia, serological tests and prothrombotic factors with negative results. Initially classified as an embolic stroke of undetermined source (ESUS). Control neuroimaging after 10 days showed SDH resolution. According to clinical stability, he was discharged home on multidisciplinary support in partial improvement, dysexecutive rehabilitation and secondary prophylaxis. **Discussion:** Although less prevalent, posterior circulation stroke has atypical presentation that can mimic other neurological conditions. Thalamus disorders are true semiological challenges that can involve from purely language syndromes to motor, cerebellar, memory and behavioral associations. The medial and intralaminar thalamic nuclei are the most affected by vascular lesions, prefrontal cortex connection territories and the center of wakefulness through ascending activating fibers (SARA), respectively. Bilateral thalamic involvement suggests vascular supply through a single trunk of the paramedian arteries, the proximal branch of the posterior cerebral artery. This anatomical variation occurs in 4-12% of the population known to have Percheron's artery. **Final comments:** Recognizing the presentation of stroke is crucial for early diagnosis and therapeutic planning according to the time window for indications. Therefore, it is important to describe and publicize less prevalent cases among health professionals and the scientific community.

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The challenges of diagnosing chronic inflammatory demyelinating polyneuropathy. The experience of Hospital Universitário Pedro Ernesto

André Rodrigues Poton[1], Rafael Oliveira Vidon[2], Salim Lázaro Balassiano[1], Eduardo Rodrigues Davidovich[1], Robson Teixeira Vital[1], Clarissa Neves Spitz[1], Larissa Bittencourt de Carvalho Chaffin[1], Marcia Rodrigues Jardim[1]

[1]Hospital Universitário Pedro Ernesto, Rio de Janeiro RJ, Brazil.
[2]Hospital Universitário Gaffrée e Guinle, Rio de Janeiro RJ, Brazil.

Correspondence: André Rodrigues Poton; Email: andrepoton@gmail.com.

Background: Chronic inflammatory demyelinating polyneuropathy (CIDP) affects both proximal and distal nerve segments, characterized primarily by muscle weakness and various types of sensory impairment. It presents in multiple forms and is an immune-mediated disease with a pathophysiology that is not fully understood. Despite being treatable, CIDP is often misdiagnosed, especially its variants, prompting the development and revision of diagnostic criteria. The 2021 revised clinical and electrophysiological criteria by the European Academy of Neurology/Peripheral Nerve Society (EFNS/PNS) are the most widely accepted. **Objective:** To analyze CIDP and its variants among patients in our hospital's Neurology service, focusing on the clinical and electrophysiological 2021 EFNS/PNS Consensus criteria. **Methods:** Retrospective study analyzing data from patients admitted with the initial diagnostic hypothesis of CIDP, seen at our Peripheral Neuropathies outpatient clinic between 2004 and 2024. Patients diagnosed with CIDP were included if sufficient medical records for evaluating 2021 EFNS/PNS Consensus clinical and corresponding electromyography data were found. Those with alternative diagnosis, insufficient medical data, or unavailable electromyography were excluded. **Results:** Among 1681 clinic patients, 68 (4%) were admitted with suspicion of CIDP. Among these, 21 (30.9%) were selected for analysis of the EFNS/PNS criteria. Five (7.3%) were excluded due to alternative diagnoses, and 42 (61.8%) due to inadequate data or unavailable electromyography. Of the 21 analyzed, 7 (33.4%) met typical CIDP criteria, while 13 (61.9%) met variant criteria: 10 (47.7%) Distal, 2 (9.5%) Motor, and 1 (4.7%) Sensory; 1 (4.7%) did not meet clinical criteria. Sixteen (76.2%) out of 21 met EFNS/PNS electrodiagnostic criteria. There were 12 (57.2%) men and 9 (42.8%) women, with an average age at symptom onset of 53.3 years (17-74), and an average time from symptom onset to electromyography of 27.9 months (1-180). **Conclusion:** Diagnosing CIDP is a challenge even in large neuromuscular disease centers. The diagnosis is primarily based on neurophysiological findings. There are various diagnostic criteria proposed across different centers, with the EFNS/PNS criteria currently being widely accepted. However, due to the diverse clinical presentations of CIDP, diagnostic complexities persist. It is important to discuss the criteria used for diagnosing CIDP to reduce both false positive and, especially, false negative diagnoses, as these prevent the treatment of a potentially disabling disease.

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The correlation between long-term sleep deprivation and the development of Alzheimer's disease: a systematic literature review

Flávia Brady de Carvalho Gauziski Figuerêdo[1], Carolina Louise Costa de Faria[1], Fernanda Pais Mainiere[1], Fernando Tarcsay Marques Neto[1], Giovanna Prudente Buccino[1], Larissa Enes Cota[1], Leonardo Figueira Magno[1], Letícia de Castro Carvalho Oliveira de Faria[1], Letícia Jabor Veiga[1], Matheus Rocha Clemente[1]

[1]Faculdades Souza Marques, Rio de Janeiro RJ, Brazil.

Correspondence: Flávia Brady de Carvalho Gauziski Figueredo; Email: flaviabrady@outlook.com.

Background: Sleep deprivation refers to a reduction in sleeping time below individual's basal needs. Inefficient sleep is a risk for human development of acute and chronic pathologies. Long-term sleep deprivation is responsible for beta-amyloid protein deposition in the brain, forming neuroinflammatory plaques and hyperphosphorylation of Tau resulting in neurofibrillary tangles, damaging the functions of neurons and synapses. These represent risk factors for the pathogenesis and progression of AD, a neurodegenerative disorder which consists in progressive cognitive decline, causing behavioral, personality and functional capacity changes in the patient. Therefore, investigating the effects of sleep deprivation on Alzheimer's disease is relevant for a scientific foundation on prevention and treatment of this pathology. **Objective:** Systematic review of articles that expose the correlation existent between long-term sleep deprivation and the progression of Alzheimer's in patients in different approaches so that the knowledge of this topic is unified and critically analyzed. **Methods:** Systematic review of articles from 2012 to 2023 selected from Ministério da Saúde, PubMed and Scielo database with the following keywords: "Alzheimer's Disease", "sleep deprivation" and "memory". Each database was screened independently by all the authors of this review. **Results:** All the 6 studies analyzed showed a degree of relation between sleep and the increase in beta-amyloid levels in the brain. A bidirectional relation was demonstrated, in which sleep alterations present themselves both as a symptom of AD and as an aggravating factor, since they favor the early onset and faster progression of the disease. There is a specific relationship between interruptions in slow-wave sleep (SWS) and Alzheimer's pathophysiology, highlighting that SWS is essential for memory-related functions and exhibits a causal and bidirectional relationship with beta-amyloid accumulation. It was observed that the preferred locations for beta-amyloid accumulation are cortical regions, due to higher expression of the HOMER1 gene in these areas, contrasting with other studies that demonstrated a higher affinity of beta-amyloid for hippocampal regions. During NREM slow-wave sleep, neuronal hyperpolarization and reduced neuronal firing decrease beta-amyloid production. Self-reported sleep problems in cognitively normal individuals correlate with AD biomarkers, such as lower Aß42 levels and higher Tau protein levels in cerebral-spinal fluid. Sleep deprivation and circadian rhythm dysfunction can lead to dysfunctional tau protein metabolism, contributing to dementia even without Aß alterations. **Conclusion:** In brief, the intricate relationship that exists between sleep deprivation and AD can either be a symptom or an aggravating factor for its quickest advance. Furthermore, this insight is crucial for prevention and treatment of this disease.

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The correlation between manual dexterity, executive function, and grip strength in individuals with Parkinson's disease

Julia Vitória Bento[1], Maria Eduarda Chaves da Silva[1], Hayslenne Andressa Gonçalves de Oliveira Araújo[1], Renan Alexandre Mregalote[1], Sara Garcia Amaro dos Reis[1], Francis Camila de Oliveira[1], Larissa Laskovski[1], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Maria Eduarda Chaves da Silva; Email: maria.eduarda.chaves@uel.br.

Background: The clinical spectrum of Parkinson's disease (PD) is broad and is characterized by motor and non-motor symptoms, as well as impairments in manual dexterity and grip strength. Equally important, the prevalence of non-motor symptoms such as depression and cognitive decline greatly impact the functionality and quality of life of individuals who suffer from it. These changes, combined with motor disturbances, impair the executive functions responsible for efficient motor planning. Since these outcomes play an important role in the independence of individuals with PD, their evaluation needs to be routine, detailed, and individualized, as well as their interrelationships understood. **Objective:** To analyze the correlation between manual dexterity, executive function, and grip strength in individuals with PD. **Methods:** This cross-sectional study involved individuals diagnosed with PD who were receiving care at a specialized neurofunctional physiotherapy outpatient clinic at the State University of Londrina, encompassing both sexes. All individuals were evaluated during the "on" phase of their medication. Initially, the Mini-Mental State Examination (MMSE), modified Hoehn & Yahr scale (HY), and Movement Disorder Society Unified Parkinson's Disease Rating Scale (MDS-UPDRS) were all used to characterize the sample. For the evaluation of manual dexterity, executive function, and grip strength, the Squares Test, Trail Making Test - parts A and B (TMT A and B), and manual dynamometry were all used respectively. Clinical and demographic data was described in median and interquartile range, and the correlation between manual dexterity, executive function, and dynamometry data was analyzed using Spearman's correlation test, adopting a significance level of < 0.05. The study was approved by the Research Ethics Committee under opinion number (CAAE: 23272819.8.0000.5231). **Results:** 27 individuals were analyzed, with a median age of 68 [56.0-74.0], between mild to moderate stages of PD, with a diagnosis time of 6 [4.0-10.0] years. In the analysis between manual dexterity and the executive function tests, moderate correlations were observed between the Squares Test on the right and left with TMT A (r = -0.69 and r = -0.68, respectively) and TMT B (r = -0.53 and r = -0.52, respectively). However, there was no significant correlation between manual dexterity and executive function tests with grip strength. **Conclusion:** In this study, manual dexterity correlated with better performance in executive function tests, which did not occur with grip strength. This result aids therapeutic planning in clinical practice, indicating a greater interconnection between executive function activities and manual dexterity compared to grip strength. Training these skills is important for activities of daily living and can improve the autonomy of people with PD.

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The critical role of accurate diagnosis in trigeminal autonomic cephalalgias: a case study

Rhaná Carolina Santos[1], Aline Oliveira dos Santos Meneses[1], Giovanna Polo[1,2], Carlos Eduardo Mantese[1]

[1]Universidade do Vale do Rio dos Sinos, São Leopoldo RS, Brazil.
[2]Hospital Mãe de Deus, Porto Alegre RS, Brazil.

Correspondence: Rhaná Carolina Santos; Email: santoscrhana@gmail.com.

Case presentation: A 31-year-old female patient, previously healthy, has been diagnosed with cluster headache for four years. She started treatment with verapamil 480mg/day and occipital block with lidocaine. Despite treatment, the patient reported intense attacks of right periorbital headache, with pain intensity of 10/10 on the visual analog scale. Episodes were associated with trigemino-autonomic phenomena such as ptosis, hyperemia and rhinorrhea, occurring up to six times a day and lasting 15-20 minutes. A good response to high-flow

oxygen was observed. The episodes occurred in cycles of 1-2 months, each cycle lasting 40 days. Treatment with topiramate was initiated but no response. Subsequently, the use of indomethacin began, with a positive response at a full dose of 100mg/day. Considering the response to indomethacin, the diagnosis of paroxysmal hemicrania was confirmed. **Discussion:** This case illustrates overlapping characteristics between cluster headache and paroxysmal hemicrania, including a good response to oxygen therapy, an important confounding factor in this example. The patient demonstrated an excellent response to "Indotest", which is the diagnostic test for detecting headaches responsive to oral indomethacin. Although paroxysmal hemicrania is less common, the clinical overlap and excellent response to indomethacin highlight the importance of precise differential diagnosis. Paroxysmal hemicrania (PH) is characterized by severe, strictly unilateral head pain attacks, localized to orbital, supraorbital, or temporal areas, accompanied by one or more ipsilateral cranial autonomic features. For a diagnosis of PH according to The International Classification of Headache Disorders (ICHD-3), a minimum of 20 attacks is required, with the frequency exceeding five per day for more than half of the time. The duration of attacks typically ranges between 2 to 30 minutes, although duration can vary widely. Usually, PH attacks are spontaneous, but they can be triggered by various stimuli. The primary treatment for preventing paroxysmal hemicrania attacks is indomethacin. An absolute response to indomethacin is one of the diagnostic criteria; however, achieving this response may require very high doses. Paroxysmal hemicrania can be very similar to cluster headache, but PH usually presents with more frequent and shorter attacks and less predilection for nocturnal occurrence. The key factor distinguishing between these two conditions for diagnosis lies in the response to indomethacin. **Final comments:** Cluster headache management is complex, often taking years to properly diagnose. The need to consider Indotest in cases of diagnosis or refractoriness to conventional treatment is reinforced. Differential diagnosis is essential, since trigeminal-autonomic headaches significantly impact the quality of life of those affected. Accurate identification and appropriate management can prevent years of avoidable suffering.

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The effect of GLP-1 analogs on the motor function of patients with Parkinson's disease: a systematic review and meta-analysis

Luís Fernando Ferreira Cavalcante[1], Giovanna Costa[1], Sophia Massafelli Battistuta[1], Pedro Faria Makabe[1], Isadora Silva Fanucci Bueno[1], Bruno Yuamoto[1], Felipe Endrigo Gonçalves Vilela[1], Diogo Haddad Santos[1]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.

Correspondence: Luís Fernando Ferreira Cavalcante; Email: luiscavalcante.med@gmail.com.

Background: Parkinson's disease (PD) stands as the second most prevalent neurodegenerative disorder. Presently, symptomatic management through medication is standard practice, yet effective disease-modifying therapies remain elusive in routine clinical care. The imperative now lies in advancing treatments to preempt and mitigate disabilities such as balance issues, cognitive decline, speech impairments, and swallowing difficulties among PD patients. Recent literature underscores the pivotal role of microglia in perpetuating chronic neuroinflammation characteristic of this condition. Against this backdrop, emerging research is spotlighting glucagon-like peptide-1 (GLP-1) analogs for their potential to restore dopamine levels, curtail dopaminergic loss, mitigate neuronal degeneration, and alleviate both motor and non-motor symptoms of PD. Originally developed for diabetes management, GLP-1 analogs have since revealed additional anti-inflammatory and neuroprotective properties, notably in suppressing microglial activation. To elucidate the possible effect on the motor function of patients with PD, we present a comprehensive meta-analysis and literature review encompassing four randomized controlled trials evaluating GLP-1 analogs versus placebo. **Objective:** To assess the effect of GLP-1 analogs on the motor function of patients with PD. **Methods:** We performed a systematic review and random effects meta-analysis of randomized control trials comparing GLP-1 analogs to placebo in patients with PD through the change from baseline of Part III of the Movement Disorder Society - Unified Parkinson's Disease Rating Scale (MDS-UPDRS). **Results:** The standard mean difference (SMD) of the change in MDS-UPDRS Part III from baseline of patients with PD was not statistically significant (SMD -0.41; 95% CI -0.93 - 0.10; I2=89%; p =0.11). **Conclusion:** Although there is a trend toward improvement, the role of GLP-1 analogs in the improvement of the motor function of patients with PD remains unclear.

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The effectiveness of dual task training when combined with exercise on cognition in individuals with Parkinson's disease: a randomized clinical trial

Rogério José de Souza[1], Alessandra Cattaneo Estrada Melanda[1], Guilherme Lopes Barbosa da Silva[1], Andressa Leticia Miri[1], Patrícia Gonçalves Broto[1], Victor Hugo Kenzo Ishii[1], Ana Raquel Lindquist[2], Adriana Costa-Ribeiro[3], Suellen Mary Marinho dos Santos Andrade[3], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.
[2]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.
[3]Universidade Federal da Paraíba, João Pessoa PB, Brazil.

Correspondence: Guilherme Lopes Barbosa da Silva; Email: guilhermelps_@hotmail.com.

Background: Cognitive decline is among the most frequent non-motor symptoms of Parkinson's disease (PD). This decline affects daily activities, gait, postural control, and the ability to perform dual tasks (DT). Managing PD, especially its non-motor symptoms, is challenging, leading to a growing interest in studies investigating the effectiveness of various treatment protocols, whether isolated or combined. **Objective:** To verify the effectiveness of dual task training when combined with walking on a treadmill as well as anodal transcranial direct current stimulation (tDCS) on cognition in individuals with PD. **Methods:** This study is a randomized, controlled, blinded clinical trial involving 36 individuals with mild to moderate PD without cognitive decline. Participants were divided into two groups: 1) the Experimental Group (EG): underwent dual task training + walking on a treadmill + anodal tDCS and 2) the Control Group (CG): underwent walking on a treadmill + anodal tDCS. There were 12 intervention sessions, that were conducted three times a week. The dual task training protocol consisted of three levels, with six tasks per level, progressively increasing in complexity, and was applied for 18 minutes while walking on a treadmill. The current was applied to the left dorsolateral prefrontal cortex at an intensity level of 2mA, while walking on a treadmill. A cognitive assessment was conducted using the Montreal Cognitive Assessment (MoCA), the Trail Making Test, and the Stroop Test. Evaluations were performed three times at different points: pre-intervention, post-intervention, and after a 4-week follow-up period. Repeated measures ANOVA was used to compare the groups based on intervention, time, and group-time interaction. The study was approved by the Ethics Committee (CAAE: 30668420.7.0000.5188) and registered at Clinical Trials (NCT04581590). **Results:** There was a significant improvement in cognition in the EG (time effect) between pre- and post-intervention, and between pre-intervention and follow-up in the delayed recall domain and the total MoCA score. Additionally, there was improvement from pre-intervention to follow-up in the Stroop Test performance. For the CG, there was improvement between pre- and post-intervention in the Stroop Test performance, and between pre-intervention and follow-up in the delayed recall domain of the MoCA. No differences were observed between groups or in the time vs. group interaction. **Conclusion:** The addition of dual task training to a combined walking on a treadmill and anodal tDCS protocol was not effective in improving cognition in individuals with PD. Cognition is a complex construct, and more studies are needed to advance this investigation and seek an optimal protocol based on the type, number of sessions, frequency, duration, and intensity of the intervention to aid clinical decision-making.

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The effects of general anesthesia on pediatric neurodevelopment: a systematic review

Ana Beatriz Landim Ribeiro[1], Lara Barbosa Vale[1], Estela Rodrigues Cardoso Chiappetta[1], Giovana Rodrigues França[1], Thiago Antonino Gonçalves[1], Gabriela de Sousa Carvalho[1], Gilberto Sadala Mendes[1]

[1]Universidade Nove de Julho, São Paulo SP, Brazil.

Correspondence: Ana Beatriz Landim Ribeiro; Email: ablr2016@icloud.com.

Background: General anesthesia is crucial for safe and painless pediatric procedures, but inadequate sedation can result in adverse outcomes. Animal studies suggest that anesthetics like isoflurane, sevoflurane, and ketamine may cause persistent cognitive dysfunctions. Retrospective studies indicate potential neurodevelopmental risks in children exposed to anesthesia, such as anxiety and learning disorders, particularly with repeated or prolonged exposure. **Objective:** To analyze the neurodevelopmental impairments caused by pediatric exposure to general anesthesia. **Methods:** This systematic review used the PICOT strategy to investigate the effects of general anesthesia (GA) on pediatric neurodevelopment. Searches in PubMed and Scielo up to February 2024 yielded 55 studies. Following PRISMA, articles on GA in children up to 4 years old were included, excluding reviews, meta-analyses, and unrelated articles. The quality of evidence was assessed using GRADE, resulting in 14 articles analyzed in this review. **Results:** According to the National Anesthesia Clinical Outcomes Registry, pediatric anesthesia poses greater neurodevelopmental risks with longer exposures. The majority of the preclinical studies showed that anesthetics like ketamine and sevoflurane cause persistent cognitive dysfunctions, especially with longer exposure. These findings were similar to the results of the clinical trials analyzed, which indicated negative neurodevelopmental effects with more frequent and prolonged exposures, especially in the first year of life. Similar results were seen in a study in Singapore with 100 children undergoing surgeries with anesthetic sedation before 1 year, that found a higher risk of learning disorders at 12 years old. A cohort study with 97 infants undergoing prolonged anesthesia was associated with reduced outcomes on the Bayley-III test at 12 months. The Bayley-III test assesses domains such as cognitive, language, motor, social-emotional, and adaptive behavior in children. In more extensive research, a cohort investigating 100,144 births revealed a higher risk of developmental delays with three or more exposures to GA. There were also contradictory results, such as a study with 33 infants who underwent general anesthesia and were assessed using the Griffiths Mental Development Scales (GMDS), showing no developmental impairment. However, these trials have limitations like small sample

sizes and exposure. **Conclusion:** This review indicates that clinical studies show prolonged exposure to GA in humans is associated with neurodevelopmental delays, varying according to age, clinical condition, and duration of drug exposure. In contrast, single and brief exposures do not appear to impact infant neurodevelopment. It is essential to continue investigating with larger and more specific clinical studies to confirm these findings and better understand the risks associated with anesthetics, particularly sevoflurane and ketamine.

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The epidemiological profile of patients with meningitis treated on an urgent basis in the state of são paulo from 2019 to 2023

Leticia Dardes Barbosa[1], Jamile Rodrigues Cosme de Holanda[2], Lara Schiavinato Merloto[3], Maiana Lourenço Benvindo da Rocha Martins[4], Ana Percya Sais da Rocha[5]

[1]Centro Universitário Barão de Mauá, Ribeirão Preto SP, Brazil.
[2]Universidade Federal de Sergipe, São Cristovão SE, Brazil.
[3]Universidade Anhembi Morumbi, São Paulo SP, Brazil.
[4]Centro Universitário Novafapi, Teresina PI.
[5]Universidad Central del Paraguay, Ciudad del Este, Paraguai.

Correspondence: Letícia Dardes Barbosa; Email: leticiadardes@hotmail.com.

Background: Meningitis is a disease that affects the membranes lining the brain and spinal cord. Caused by viruses, bacteria, or other infectious agents, it can lead to serious complications if not promptly treated, and it can be prevented through vaccination. **Objective:** To conduct an epidemiological analysis of patients with meningitis treated as emergency cases in the state of São Paulo between the years 2019 and 2023. **Methods:** This is an ecological study that obtained data collection through the Hospital Information System (SIH/SUS). The studied population consisted of individuals from the state of São Paulo with meningitis who were treated as emergency cases during the period from 2019 to 2023. The analyzed variables included the number of hospitalizations, age group, gender, and race/ethnicity. Data were collected using descriptive statistical analysis techniques. **Results:** In this five-year period studied, 7,514 cases of meningitis were reported, with the year 2019 recording the highest number of cases, at 2,041 (27.29%), and the year 2021 recording only 947 cases (12.60%), representing the lowest result in these periods. The most affected age group is infants under 1 year old, with 1,559 cases reported (20.74%), and the second most affected age group is children aged 1 to 4 years, with 1,267 cases (16.86%). Elderly individuals over 80 years old had only 64 hospitalizations (0.85%) over the last 5 years. The predominance in terms of race/ethnicity occurs in the white population, with 3,837 cases (51.06%), notably in the year 2019, when 1,013 episodes (13.48%) were recorded. The second most affected population is the mixed-race (parda) population, with 2,311 occurrences (30.75%), with the year 2023 having the highest incidence, with 652 cases (8.67%). The male population predominated over the female, with 4,300 (57.22%) notifications, notably in the year 2019, when 1,182 (15.73%) cases were recorded, while females accounted for 3,214 (42.77%), also with emphasis in 2019, with 859 hospitalizations (11.43%). According to the analysis of the profile of affected patients seeking emergency care, a significant increase in cases was observed in 2019, especially among children under one year old, white individuals, and males, which is consistent with previous studies. It was found that the elderly population was less affected, accounting for only 0.85% of emergency care over the last 5 years. **Conclusion:** Meningitis poses a major challenge to public health. According to these data, there is a predominant distribution in the pediatric population, thus necessitating the implementation of sanitary, educational, and adequate medical care measures to reduce and control meningococcal disease in the study area. Additionally, it underscores the importance of public actions regarding prophylaxis against such pathology, such as extensive vaccination campaigns against influenza and pneumococcus.

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The first experience with the use of deutetrabenazine in a young patient diagnosed with tardive dyskinesia in the Brazilian Amazon: a case report

Maria Esther Lima Rios Evangelista[1], Emanuelle Campos Amaral[2], Fernanda Almeida Carvalho[2], Danilo Lemes Reis[2], Manuela Brandão Ferreira Teles[2], David França Ferreira Cruz[2], Victor Hugo Soares dos Santos[2], Jonathas Taumaturgo Soares[2], Marcus Vinicius Della Coletta[2]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Maria Esther Lima Rios Evangelista; Email: mariaesther.rios@hotmail.com.

Case presentation: Male patient, 24 years old, diagnosed with tardive dyskinesia in 2019 due to the indiscriminate use of neuroleptics following psychiatric hospitalization for a psychotic episode with acute symptoms. After four years of diagnosis and a significant worsening of his quality of life, he began using deutetrabenazine to treat the severe chorea he presented, under medical supervision and recommendation. **Discussion:** Deutetrabenazine (DTBZ) is a highly selective reversible inhibitor of VMAT2, whose mechanism of action involves the depletion of monoamines in the presynaptic terminals of the CNS. It is metabolized via the hepatic route, producing alpha-dihydrodeutetrabenazine and beta-dihydrodeutetrabenazine as metabolites. In 2023, six years afts is approval by FDA, in the USA, and two years after its approval by ANVISA, in Brazil, the first patient in the state of Amazonas began treatment with this medication, obtained through legal means due to its high cost. Therapy began in September 2023 with one 6 mg pill of DTBZ per day for one week, followed by weekly dosage adjustments until reaching a stable dosage of two pills, 6 mg and 9 mg per day. One month after starting treatment, there was a reduction in chorea in the upper limb extremities and the previously present buccolingual stereotypies. After three months of treatment, a significant change in the patient's routine was noted due to the reduction of motor symptoms, allowing a return to activities that were previously not possible, for example playing soccer and feed himself, and reducing the risks of bronchoaspiration due to food. Currently, he exhibits a progressive improvement in symptoms. **Final comments:** As well as demonstrated in randomized studies conducted abroad, deutetrabenazine showed significant efficacy in the symptomatic treatment of tardive dyskinesia in a young patient in Amazonas, leading to a substantial improvement in his quality of life through the modulation of neurotransmitters by the medication.

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The great mimicker strikes again: neurosyphilis mimicking viral and autoimmune limbic encephalitides - a case report

Gabriel Abrahão Stoliar[1], Carolina Mibielli[1], Hélio Afonso Ghizoni Teive[1], Caio César Diniz Disserol[1,2,3]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.
[2]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.
[3]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Gabriel Abrahão Stoliar; Email: gabrielstoliar@gmail.com.

Case presentation: A 47 years old man with a history of smoking and drug abuse (crack) presented with 2 months of behavioral changes, aggressiveness, disorientation and focal evolving to to bilateral tonic-clonic seizures. Upon admission he was febrile and there was no respiratory, gastrointestinal or urinary symptoms. Neurological examination revealed spatial disorientation and psychomotor agitation, without changes in cranial nerves, strength, sensitivity, reflexes, balance, coordination or meningoradicular signs. Brain MRI showed T2/FLAIR hyperintensities affecting the right medial temporal lobes. CSF revealed pleocytosis (20 cells; 98% lymphocytes) and elevated protein levels (98.7); glucose was normal. Due to the high suspicion of herpetic encephalitis, IV acyclovir was started. However, two viral panels were negative for HSV-1, HSV-2, VZV and CMV, and CSF VDRL was positive (1:8). Tissue and cell-based assays for superficial and intracellular neuronal antibodies was negative. The treatment was switched to Crystalline Penicillin 4,000,000 IU every 4 hours for 14 days, and the patient had partial clinical improvement, with residual memory deficit and behaviour changes, but without new seizures or neurological deficits. **Discussion:** infection of the central nervous system by Treponema pallidum is known as neurosyphilis. This is a clinically challenging disease because it affects any part of the CNS, explaining its multifaceted presentations and why is a great mimicker of many disorders. Syphilitic limbic encephalitis mimicking viral or autoimmune encephalitis is rare. Some cases of rapidly progressive neuropsychiatric symptoms with or without seizures and limbic encephalitis on MRI affecting one or both medial temporal lobes have been described. Limbic encephalitis secondary to neurosyphilis still has an uncertain cause. It is possible that intense meningovascular inflammation may cause vasogenic, cytotoxic, and interstitial edema. Furthermore, ischemic changes in small vessels induced by infection can cause gliosis. **Final comments:** This case represents an atypical manifestation of neurosyphilis with limbic system involvement, typical topography of viral and autoimmune etiologies, with partial recovery after antibiotic therapy. Physicians should be aware of this very uncommon presentation of neurosyphilis, which mainly mimics viral and autoimmune encephalitides.

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The impact of a stroke unit at Northeastern Brazil on the national institute of health stroke scale and the average rankin: an original study

Maria Luiza Vasconcelos Montenegro[1], Maria Eduarda Vieira de Oliveira[1], Lívia Barbosa Cavalcanti[1], Isaías Henrique Martins de Vasconcelos Norte Bezerra[1], Noelle Ventura Jordão[2], Maria Eduarda Moreira Cardoso[2], Ana Dolores Firmino Santos do Nascimento[2]

[1]Universidade de Pernambuco, Recife PE, Brazil.
[2]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Maria Luiza Vasconcelos Montenegro; Email: marialuizavasc534@gmail.com.

Background: Stroke is a leading cause of death worldwide, accounting for approximately 100,000 deaths annually in Brazil alone. Survivors often experience functional deficits; approximately three-quarters suffer moderate impairments, while 15 to 30% endure severe deficits.

Various scales play crucial roles in stroke assessment, notably the NIH Stroke Scale (NIHSS) for evaluating clinical severity and progression, and the Rankin Scale for assessing patient disability levels. In Brazil, the average cost per stroke patient is $11,300, significantly lower compared to developed countries. Specialized stroke units are pivotal in delivering prompt and effective treatment, thereby enhancing recovery prospects and minimizing long-term sequelae. **Objective:** To evaluate the neurological deficits and functionality of patients admitted to a Stroke Care Unit (SCU), using the NIHSS and RANKIN scales, respectively. **Methods:** This study is a retrospective cohort carried out in a SCU at a reference center, in Pernambuco, Brazil, between January of 2023 and May of 2024, evaluating the NIHSS and the RANKIN scales. The inclusion criteria were patients with stroke of ischemic etiology with age and sex registered, and individuals with at least one of the scales assessed at the time of entry and exit from the hospital. **Results:** 1,224 patients admitted to the SCU were identified, however, 701 were excluded as they did not meet the inclusion criteria, leaving 523 individuals. The study consisted of 276 (52.8%) male, with the average age of the patients being 65.7 years old. Among them, 256 (49%) underwent intravenous administration of thrombolytic agents. The average NIHSS on admission was 12.2. At the time of hospital discharge, the average NIHSS was 9.7 points. About RANKIN, the average value at admission was 0.52, while at hospital discharge there was an increase to 2.6. **Conclusion:** A higher prevalence of ischemic stroke was observed as people aged and in men, with the majority of individuals being treated conservatively. The average NIHSS value was reduced by 20.4% when comparing the moments of hospital admission and discharge, which reveals the importance of stroke units in transforming the patient's prognosis. Despite the increase in RANKIN, the average found in this study was below that reported in another Brazilian study.

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The impact of dysphagia on quality life in Parkinson's disease

Vivian Kaori Segawa[1], Arthur Jun Deguti Hata[1], Ana Luiza Fabris Moraes[1], Margarete Jesus Carvalho[1], Alice Estevo Dias[1], Emerson Gisoldi[1]

[1]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Vivian Kaori Segawa; Email: viviankaorisegawa@gmail.com.

Background: Idiopathic Parkinson's disease (IPD) is a neurodegenerative, chronic and slowly evolving pathology that mainly affects the population over the age of 65. Dysphagia is one of the disabling clinical manifestations and affects approximately 80% of patients with IPD. **Objective:** To assess the presence and degree of severity of dysphagia symptoms in patients with ILD. **Methods:** A study was carried out using the Eating Assessment Tool (EAT-10), made up of 10 questions graded from 0 to 4, with 0 corresponding to "no problems" and 4 to "severe problems". The instrument's cut-off point is 3 points, a score equal to or greater than this value indicates a risk of dysphagia. The patients were assessed using the Hoehn & Yahr (H&Y) scale and the Unified Parkinson's Disease Rating Scale (UPDRS) parts I, II, III, IV and V. Pearson's correlation test was used. **Results:** 25 patients with IPD aged between 40 and 80 years were assessed, 14 (56%) of whom were female and 11 (44%) male. The H&Y scale showed that 21 patients (84%) had mild to moderate disability, with answers between 1 and 3, and 4 patients (16%) had severe disability. The UPDRS ranged from 9 to 96. The EAT-10 showed that 4 patients (16%) complained of pain when swallowing; 6 patients (24%) were stressed by swallowing difficulties and had to make efforts to swallow medicines; 7 patients (28%) lost weight due to swallowing problems, reported straining to swallow solids and liquids and reported a reduction in the pleasure of eating; 8 patients (32%) reported that food stuck in their throats; 9 patients (36%) coughed while eating and 12 patients (48%) said they stopped eating out because of swallowing difficulties. There were 12 (48%) scores below 3, representing a low risk of dysphagia, and 13 (52%) scores equal to or greater than 3, with the highest score being 32. There was a weak correlation between H&Y and EAT-10, while the correlation between UPDRS and EAT-10 was moderate. **Conclusion:** This study showed that the EAT-10 was effective in identifying patients' swallowing difficulties and how they affected their quality of life, and could be used during neurological assessment to make a diagnosis. High UPDRS scores denoted severe dysphagia and prompted immediate referral for speech therapy analysis and rehabilitation.

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The impact of external attentional focus on prefrontal cortex activity: a preliminary stud

Maria Eduarda Salum Aveiro Henrique[1], Clara Rezende Rocha[1], Pedro Henrique Sousa de Andrade[1], Caroline Rodrigues Osawa[1], Jayne Siqueira Cagliari[1], Marco Túlio Almeida Lenza[1], Luciane Aparecida Pascucci Sande de Souza[1], Gustavo José Luvizutto[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Maria Eduarda Salum Aveiro Henrique; Email: aveiromariaeduarda@gmail.com.

Background: Motor control depend on the interaction of the individual with the task and the environment. To develop successful daily tasks, the individual must control movement synchronicity and coordination, which requires prefrontal lobe activity. The frontal lobe is the part of the cortex responsible for executive functions such as planning, organization, and attention targeting. It is possible to optimize coordination during a task to target attention to an external focus. Despite literature proving that an external focus may improve muscular activity during movement, no studies assess the influence of a focus of attention on brain activity. **Objective:** To measure the effect of different foci of attention on prefrontal lobe activity in healthy people during a lower limb task. **Methods:** This is a cross-sectional study with ten neurologically healthy individuals. The prefrontal activity was measured by hemoencephalography (HEG Neurobots®) with sensors allocated at the Fp1 and Fp2 areas during three different activities. Brain activation was assessed by the increase of oxyhemoglobin (oHb) during the realization of each task according to the optical density (OD) presented by the device. Higher levels of oHb increase the OD due to lower absorption of light sent by the device. In the first task, the volunteer had to touch a step 20.3 cm high with the right (RS) and left (LS) legs; the second task demanded touching a plastic cup 8 cm high and 7.5 cm in superior diameter with both legs (RCA/LCA); in the third task, the individual had to perform the same movement as task two but with a plastic cup 4 cm high and 5.3 cm in superior diameter (RCB/LCB). The cups were placed on a platform to measure their height with the step. Each task was performed three times, and the third attempt was analyzed. The OD comparison between tasks and moments of the tasks was analyzed by the ANOVA test of repeated measures and the Tukey test of multiple comparisons. The data was considered statistically significant if $p<0.05$. **Results:** Five women and four men with a mean age of 27.1 years (±5.5) were evaluated. In most tasks, the OD mean registered on HEG was higher at the beginning of the activity than in the other moments (execution and end). There was a statistical difference only in the moments of the RCB ($F=4.0$; $p=0.046$). In the multiple comparison analysis, differences were observed between the beginning and the task ending (MD: 4.3; $p=0.032$). **Conclusion:** Prefrontal lobe activity is highest at the beginning of the task compared to the end when it comes to the RCB activity. This finding suggests that the executive functions related to planning during a task are more significant in its initial phase than in execution and end, especially in tasks requiring more precision.

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The impact of motor, non-motor, and social aspects on the sexual health of women living with Parkinson's disease

Kátia Cirilo Costa Nóbrega[1], Isaíra Almeida Pereira da Silva[1], Raissa Amorim Gonçalves[2], Thalyta Silva Martins[1], Geovanna Ferreira Santos[1], Bruno Rafael Antunes de Souza[3], André Helene Frazão[3], Antônio Carlos Roque[4], Maria Elisa Pimentel Piemonte[1]

[1]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.
[2]Universidade Federal do Amapá, Macapá AP, Brazil.
[3]Universidade de São Paulo, São Paulo SP, Brazil.
[4]Universidade de São Paulo, Ribeirão Preto SP, Brazil.

Correspondence: Kátia Cirilo Costa Nóbrega; Email: katiacirilo@usp.com.

Background: Sexual dysfunction (SD) is a common non-motor symptom (NMS) in people with Parkinson's disease (PwPD). Sexual health (SH) depends on several biological, mental, and social factors that PD may affect. Despite its prevalence and relevance for quality of life, SD in women with Parkinson's disease (WwPD) is poorly understood, and research in this area is scarce. **Objective:** To investigate the impact of motor, non-motor, and social aspects on the SH of WwPD. **Methods:** We conducted a cross-sectional study of 100 women with confirmed diagnosis of idiopathic PD, (mean age 54.45±8.31), in stages 1-3 of Hoehn and Yahr classification, who reported having an active sex life in the last six months. The following data were collected by remote interview for each participant: (1) Demographic and clinical features; (2) global cognitive capacity; (3) Non-Motor Aspects of Experiences of Daily Living; (4) Motor Aspects of Experiences of Daily Living; (5) Fatigue; (6) Self-esteem; (7) Sleep disorder; (8) Couple relationship quality (CRQ); (9) Depressive signals; (10) Short-term sexual health by Female Sexual Function Index; and (11) Long-term sexual health by Sexual Quotient-Female. **Results:** We found that several motor, non-motor, and social factors were correlated with SH. However, only CRQ and sleep quality were able to predict short and long-term SH in WwPD. We did not find any significant correlations between SH and age, disease onset, post-menopausal period or medication daily dosage. **Conclusion:** Our findings highlight the importance of a multi-professional team approach to provide comprehensive care to improve SH in WwPD.

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The impact of PLCG2 gene variants on the pathophysiology of Alzheimer's disease

Savicevic Ortega Silva de Melo[1], Rebeka Mayara Santana Carneiro[1]

[1]Faculdade Pernambucana de Saúde, Recife PE, Brazil.

Correspondence: Savicevic Ortega Silva de Melo; Email: ortegasavicevic@gmail.com.

Background: Alzheimer's disease (AD) is the leading cause of dementia worldwide, characterized by memory loss and progressive cognitive decline. Genome-wide association studies (GWAS) have highlighted the role of microglia in AD pathology, with phospholipase C gamma

2 (PLCγ2) being a gene of particular interest. PLCγ2, expressed in microglia, is involved in immune signaling and cerebral inflammatory response. The P522R variant of the PLCG2 gene has been associated with a reduced risk of AD and other neurodegenerative diseases. This systematic review investigates the impact of PLCG2 gene variants on the pathophysiology of AD, exploring their effects on microglial activity, neuroinflammation, and disease progression, aiming to contribute to the development of new therapies and diagnostic strategies. **Objective:** This study aims to investigate the impact of PLCG2 gene variants on the pathophysiology of AD. We seek to identify specific PLCG2 variants associated with the disease, evaluate their effect on microglial activity, and explore the underlying molecular mechanisms. **Methods:** This is a systematic review based on PRISMA methodological recommendations. The databases used were Lilacs and PubMed. The search descriptors were: "Alzheimer" and "Phospholipase C Gamma". Inclusion criteria were: articles published between 1998 and 2024, in English and Portuguese, specifically investigating the relationship of the PLCG2 gene with AD. We excluded studies in rodents and unavailable texts. **Results:** Of the 44 initially selected articles, 15 were reviewed, providing a detailed view of the central role of PLCγ2 in neurodegenerative diseases, focusing on AD. These studies showed that specific genetic variants, such as P522R, are associated with a reduced risk of AD. The P522R variant was consistently related to the promotion of beta-amyloid clearance and the enhancement of a protective microglial function, characterized by modulated inflammatory responses and increased phagocytic capacity. Conversely, hypermorphic variants such as S707Y were identified as causing exacerbated dysfunctions in microglial cells. These hyperactive PLCγ2 variants demonstrated significant adverse effects, impairing microglial function, potentiating inflammatory processes, and contributing to pathological progression in neurodegenerative disease models. **Conclusion:** The reviewed studies highlighted PLCγ2's crucial role in regulating immune cell processes, including signaling, immune response, and phagocytosis. Optimized fluorogenic substrates enabled advanced methods to assess PLCγ2 activity, facilitating the identification of potential therapeutic activators or inhibitors. These findings enhance our understanding of AD and other neurodegenerative conditions, offering promising avenues for targeted therapies that improve microglial function and reduce the pathological burden in the central nervous system.

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The impact of population aging on hospitalizations for Alzheimer's disease: a temporal analysis

Sávio Roberto Roberto Silva[1], Marcos Lude da Silva Ferreira[1], Bruno Kauê Rodrigues Vilhena[1], Danusa Neves Somensi[1], Mariangela Moreno Domingues[1], Emanuel de Jesus Soares de Sousa[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Sávio Roberto Silva Costa; Email: saviorsc20@gmail.com.

Background: From 2010 to 2022, the elderly population in Brazil grew rapidly, increasing by 57.4% to 22.2 million, which represents 10.9% of the Brazilian population. Since aging is considered to be the most significant risk factor for developing Alzheimer's disease (AD), the demographic transition implies an enormous public health challenge. In this context, epidemiological studies on AD in the country are important, in order to provide data to help plan health policies focused on the population studied. **Objective:** To analyze how the aging of the population over two decades has affected hospitalizations for AD in Brazil. **Methods:** This is an ecological, cross-sectional, observational study with a quantitative approach. Secondary data was collected from the Department of Information and Informatics of the Unified Health System (DATASUS), in the hospital morbidity section (SIH/SUS). The data collected refers to the years 2009 to 2023. Variables related to the number of hospitalizations, type of hospitalization, gender, ethnicity, age group, average cost of hospitalization, average length of stay and mortality rate were analyzed. A p-value of less than 0.05 was considered statistically significant for this study; Bioestat 5.3 software was used for the statistical analysis. **Results:** In 10 years, 19,598 hospitalizations for ad were recorded in Brazil. The population aged 65 and over accounted for 92.8% of hospitalizations, while patients with early-onset Alzheimer's (under 65) accounted for 7.2% of the records. In this context, the population aged 80 and over accounted for the majority of hospitalizations, with 57.2% of the total, followed by the 75-79 age group, which accounted for 18% of cases. The age group with the highest relative growth in hospitalizations in the period was 80 years or older (350%), followed by the 60 to 64 age group (334%). In terms of gender, 35% of inpatients were male and 65% female. In addition, 49.7% of patients were identified as white, 20% as brown and only 4% as black. The mortality rate for the period was 4.46, with an average length of stay of 5.4 days. The average cost of hospitalization during the period was R$1,233.92. **Conclusion:** The study revealed that the population most affected is over 65 years old, which highlights the importance of aging. In addition, there was an increase in hospitalizations of patients at an early age for the disease, suggesting the existence of risk factors to be better investigated. In addition, there was a clear predominance of females and whites in hospitalizations, with fewer hospitalizations of the black population. This study can help define public health policies so that they can be more efficient and based on the national reality.

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The impact of sex differences on the systemic consequences of diabetes associated with ischemic stroke: a preclinical study

Khiany Mathias[1], Richard Simon Machado[1], Nathalya Camila Cruz Guerra Correa[2], Edgar Pereira[2], Solange Stork[1], Maiara de Aguiar da Costa[2], Victoria Linden de Rezende[2], David dos Santos[2], Gislaine Tezza Rezin[1], Fabricia Petronilho[2]

[1]Universidade do Sul de Santa Catarina, Tubarão SC, Brazil.
[2]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Khiany Damaris Mathias Maurilio; Email: khianymathiasft@gmail.com.

Background: Diabetes mellitus (DM) is a disease characterized by elevated serum glucose levels, a condition known as hyperglycemia. Hyperglycemia plays a crucial role in the occurrence of complications related to DM, such as ischemic stroke (IS), which results in the interruption or reduction of cerebral blood flow in a focal region of the brain, compromising neurological function. **Objective:** Considering the hypothesis that DM and its hyperglycemic state may not only predispose to IS but also enhance post-ischemic immune and glycemic pathways, the present study aimed to understand the pathophysiological mechanisms of DM in the occurrence and outcome of IS, examining their association with worse outcomes in affected individuals. **Methods:** We randomly assigned male Wistar rats, aged 60 days, into four experimental groups: (1) sham + sham, (2) sham + middle cerebral artery occlusion (MCAO), (3) DM + sham, and (4) DM + MCAO. Animals were experimentally induced into a DM model by a single administration of alloxan 150 mg/kg intraperitoneally, and after 2 days, they underwent the MCAO model to induce IS. Twenty-four hours after painless euthanasia, blood was collected for biochemical analyses, weight evaluation, temperature, neurological score, glycemic, and immunological parameter assessment. **Results:** There was an increase in glucose levels in both sexes on day 2 after alloxan induction compared to day 0, confirming the hyperglycemia model. In both male and female rats, diabetic animals subjected to MCAO showed a significant increase in glucose levels in urine and blood compared to the control group without diabetes. In females, there was a significant decrease in segmented cells in the DM + MCAO group compared to the DM + sham group and a significant increase in lymphocytes in the DM + MCAO and sham + MCAO groups compared to their respective controls, with a decrease in total leukocytes in DM + MCAO females. In males, there was a significant decrease in size in the DM + MCAO group compared to the sham + sham group, possibly explained by the recruitment of immune cells from the spleen. In terms of weight, there was a decrease in spleen weight in DM+MCAO females and males. In females, there was a decrease in the spleen weight/body weight ratio in both the sham + MCAO and DM + MCAO groups. **Conclusion:** The hyperglycemic state, in addition to promoting the occurrence of the disease, is capable of altering the immune response after cerebral ischemia and worsening the outcome of the disease caused by the animal's sex.

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The impact of statin therapy on cognitive function: a systematic review and meta-analysis of observational studies and clinical trials

Fernando Luiz Westphal Filho[1], Artur Menegaz de Almeida[2], Luís Otávio Souza Nogueira[3], Luis Henrique Soares Silva[4], Luana Mendes dos Santos[5], Luiz Eduardo Oliveira Bezerra[6], Lilianne Rodrigues Fernandes[7], Ítalo Barros Andrade[8], Michele Kreuz[9], Vitor Kendi Tsuchiya Sano[10]

[1]Universidade Federal do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[3]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[4]Universidade de Uberaba, Uberaba MG, Brazil.
[5]Universidade de São Paulo, São Paulo SP, Brazil.
[6]Universidade Estadual do Piauí, Teresina PI, Brazil.
[7]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.
[8]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[9]Universidade Luterana do Brasil, Canoas RS, Brazil.
[10]Universidade Federal do Acre, Rio Branco AC, Brazil.

Correspondence: Fernando Luiz Westphal Filho; Email: fernandowestphal1@gmail.com.

Background: Statins have attracted attention for their possible effects on brain function and cognition in addition to their cardiovascular benefits. Observational studies suggest that statins may lower the risk of Alzheimer's disease and other types of dementia, but evidence from clinical trials is mixed. Furthermore, the body of evidence lacks thorough evaluations of whether statins are linked to slower cognitive decline. **Objective:** To provide a current and thorough review of the link between statin use and cognitive decline while also pointing out the limitations of the existing evidence. **Methods:** MEDLINE, EMBASE, and Web of Science databases were systematically searched until May 14, 2024. We included observational studies and trials that evaluated the impact of statins in the deterioration of cognitive function in patients with and without dementia through Mini-Mental State Examination (MMSE) score assessment. We computed the MMSE scores at baseline and at the end of follow-up to calculate the mean difference. Data were analyzed using the Mantel-Haenszel method and 95% confidence intervals (CI). Heterogeneity was assessed using I^2 statistics. Statistical analysis was performed using R, version 4.3.2. **Results:** A total of 4 studies were included in this meta-analysis, comprising 3 observational studies and 1 randomized clinical trial, with a combined total of

2,345 patients. Among these, 847 patients were under statin therapy. When compared with the control group, there was no statistically significant difference regarding improvement in the deterioration of cognitive function (MMSE MD 0.70; 95% CI - 0.25 to 1.66; P=0.149; I^2=96%). **Conclusion:** In this comprehensive systematic review and meta-analysis, no statistically significant association was found between statin use and the improvement of cognitive function deterioration, as measured by the MMSE scores. The results highlight a critical gap in the literature: the current evidence, primarily derived from a limited number of observational studies and a single randomized clinical trial, is insufficient to draw definitive conclusions. Therefore, our study underscores the urgent need for more robust and well-designed randomized clinical trials assessing the impact of statin therapy on cognitive function.

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The impacts of non-pharmacological therapies on the quality of life of patients with Alzheimer's disease: a systematic review

Priscila Santos Oliveira[1], Emmanuel Santos Trindade[1], Victor Santos Vilares[1], Luídi Neves Nascimento[1], Naila Maria Magalhães da Silva[1], Philip George Glass Andrade[1], Davi Tanajura Costa[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Emmanuel Santos Trindade; Email: emmanuelstrindade@gmail.com.

Background: Alzheimer's disease (AD) is a neurodegenerative condition that challenges the physical and mental health of individuals but also affects their Quality of Life (QoL). The concept of QoL transcends mere absence of disease, encompassing multidimensional aspects reflecting physical, psychological, social, and emotional well-being. Addressing the various dimensions of AD, in which the insidious progression of the disease often deprives patients of their autonomy and identity, preservation and promotion of quality of life emerge as essential objectives in caring for these individuals. **Objective:** To conduct a systematic review of the impacts of non-pharmacological therapies on the Quality of Life of patients with Alzheimer's Disease described in the literature in the last 10 years. **Methods:** A systematic literature review was conducted in PubMed and BVS databases on March 18, 2024. The descriptors "Alzheimer's disease", "non-pharmacological treatment", and "quality of life" were used, combined with the boolean operator AND. Systematic reviews and meta-analyses published in the last 10 years in English, Portuguese, and Spanish were included. 48 articles were found, of which 22 were selected after initial reading of the abstracts. After full-text reading, 9 were excluded and 13 were included in this research. **Results:** Among non-pharmacological therapies, music therapy, whether singing or listening to music, appears as one of the main alternative therapies for AD, with improvements in some cognitive domains such as attention and language, and in the patient's QoL. Additionally, dance can be a useful tool in the promotion of wellbeing in patients with AD and Mild Cognitive Impairment well-being, assisting in balance and, primarily, in promoting mental health, which is helpful in depression management. Reminiscence therapy presented positive results in the global cognitive and neuropsychiatric aspects of AD patients, with improvement in depressive symptoms. It is a low-cost technique that improved the QoL of patients after its application. Structured physical activity is also a tool that promotes QoL, assisting in cognitive improvement and sleep. Cognitive stimulation was another approach that proved relevant improvement in QoL and overall cognition, providing benefits even in ADLs. These activities, especially if applied in a multimodal form, can help promote the comfort of these patients. **Conclusion:** It is observed that there is a definitive responsiveness of patients with Alzheimer's Disease (AD) to non-pharmacological treatments. In this sense, despite their apparent simplicity, these activities directly contribute to the positive QoL of individuals with AD and indirectly affect neurocognitive aspects that sometimes reflect consequences of disease progression. Therefore, such non-pharmacological therapies are treatment modalities capable of addressing not only AD but also the associated negative impacts.

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The importance of differential diagnosis in acute vestibular syndrome

Daniela Aparecida Lima Viana[1], Amanda Soares Pimenta[1], João Henrique da Matta Clementino[1], Ana Luisa Rufino de Sousa[1], Elisângela Ferraz Pazinat[1], Giovani Zago Borges[1], Alex Eduardo Silva[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Daniela Aparecida Lima Viana; Email: danialviana@hotmail.com.

Case presentation: Man, 70 years old, started experiencing dizzying symptoms 15 days ago associated with nausea and vomiting that improved in less than 24 hours and began treatment for labyrinthopathy. He was admitted to the emergency room with vertigo, peripheral facial paralysis on the left, tongue and lip rhyme with deviation to the right, tactile and painful hypoesthesia on the left hemiface, absence of strength deficits and nystagmus. Previously hypertensive and denied smoking. The patient was admitted to the neurological unit in a window for thrombolysis, but with a head tomography showing acute infarction of the left middle cerebellar pendunculus. The investigation was complemented with transthoracic echocardiogram and electrocardiogram without changes that would justify the ischemic event. Arterial CT angiography of cervical and intracranial arteries, small amorphous filling defects in the V1 and V2 (proximal) segments of the left vertebral artery and slight irregularity in the contours of the P2 segment of the left posterior cerebral artery. **Discussion:** Posterior fossa stroke can cause dizzying symptoms, changes in motor and sensory coordination, facial and swallowing impairment. **Final comments:** it is extremely important to carry out the differential diagnosis between peripheral and central vertigo, since the treatment is directed to the cause of the vertigo with the possibility of definitive treatments when possible and rehabilitation.

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The importance of multidisciplinary management in Miller Fisher syndrome associated with bickerstaff encephalitis: a case report

Guilherme Antonio Niemeyer Di Mango[1], Bruna de Andrade Hees[2]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.
[2]Estacio de Sá, Rio de Janeiro RJ, Brazil.

Correspondence: Guilherme Antônio Niemeyer Di Mango; Email: guilherme.dimango@gmail.com.

Background: Miller Fisher syndrome, a benign variant of Guillain-Barré syndrome, characterized by extrinsic ophthalmoplegia, cerebellar ataxia and profound areflexia, may be associated with Bickerstaff encephalitis, constitutes a pathology that requires intensive and multidisciplinary care. **Objective:** To report the clinical and therapeutic evolution of a patient with Miller-Fisher syndrome, Bickerstaff encephalitis and other complications, to discuss its particularities in intensive care, based on the analysis of a case in 2015. **Methods:** This work consists of a case report, with narrative review of articles, whose information and a data contained were obtained through careful reading and review of the medical record, interview with the patient and photographic record of complementary exams, with proper authorizations and signatures on the consent form. **Results:** It is a case report from the year 2015 of an adult female patient who developed Miller Fisher syndrome after Zika virus infection, also presenting Bickerstaff encephalitis. She was treated with venous immunoglobulin in the context of an intensive care unit, supported by a multidisciplinary team trained to manage the case, showing a good response to the therapeutic proposal. **Conclusion:** Treatment for Miller-Fisher syndrome, when indicated, should not be delayed due to the high potential for complications. For an adequate approach to the patient, it is important that there is a multidisciplinary support for the management of the case, in addition to pharmacological or non-pharmacological disease-modifying treatment.

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The influence of ischemic stroke on cognitive performance impairment

Jocsã David Gomes Santos[1], João Gabriel Araújo Coelho[1], Lucas Falcão Ferreira[1], José Felipe Modesto Cordeiro[1], Luís Felipe Caputo Oliveira Servulo[1], Leiz Maria Costa Véras[2], Ítalo Vecchi Figueiredo[1], João Lucas Gomes Carvalho[1], Ricardson Rolim Ricarte Neto[1]

[1]Instituto de Educação Superior do Vale do Parnaíba, Parnaíba PI, Brazil.
[2]Centro Universitário Maurício Nassau, Parnaíba PI, Brazil.

Correspondence: Jocsã David Gomes Santos; Email: jocsadavid.jd@gmail.com.

Background: Stroke is defined as the rapid onset of an episode with symptoms characterized by focal or generalized deficiencies in brain function, vascular motivation, and manifestations lasting at least 24 hours. These symptoms cause changes in the cognitive and motor planning of the affected individual, depending on the location and extent of the lesion. **Objective:** Investigate, through a literature review, the main aspects related to ischemic stroke and possible relationships with changes in cognitive performance. **Methods:** The databases Up To Date, Scielo, Science Direct, and Pubmed were searched with the descriptors "ischemic stroke," "cognitive performance," and "symptoms." Studies of the last 10 years were considered, in addition to English, Portuguese, and Spanish. **Results:** The literature states that approximately 50% of individuals affected by an ischemic stroke manifest cognitive deficits six months after the episode. In addition, stroke has a high incidence, prevalence, and mortality rate, especially in economically less developed countries, and affected elderly individuals are more likely to have impaired cognitive performance after stroke compared to young people. **Conclusion:** Therefore, stroke often leaves sequelae in surviving patients; among these sequelae is the alteration of cognitive performance. To understand the severity, it is necessary to consider the affected area, the extent of the damage, and the structures associated with this brain region.

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The influence of the gut microbiome on stroke incidence and recovery: mechanisms and therapeutic interventions

Jocsã David Gomes Santos[1], Maria Clara de Sousa Morais[1], Raquel de Sousa Lima[1], Renata Amélia Léda Moura[1], Luan Kelves Miranda e Souza[1], Lucas Mainardo Rodrigues Bezerra[1], João Felipe Fernandes Brito[1], Maria Tereza Viana Lima[1], Ana Júlia Araújo Fortes[1]

[1]Instituto de Educação Superior do Vale do Parnaíba, Parnaíba PI, Brazil.

Correspondence: Jocsã David Gomes Santos; Email: jocsadavid.jd@gmail.com.

Background: The interaction between the gut microbiome and the cerebrovascular system has been shown to be crucial in determining neurological health. Recent studies have shown that the composition of the gut microbiome can influence both stroke incidence and recovery. This emerging field offers a new perspective on the pathophysiological mechanisms of stroke, as well as suggesting possible therapeutic interventions to improve clinical outcomes. Investigating the gut microbiome opens avenues to understand how alterations in gut flora may affect stroke risk and subsequent recovery. **Objective:** To explore the influence of the gut microbiome on stroke incidence and recovery, elucidating the biological mechanisms involved and evaluating therapeutic interventions based on microbiome modulation. **Methods:** This is an integrative literature review, using descriptors such as "Gut Microbiome" and "Stroke" combined by the Boolean operator AND. The databases consulted were PubMed and Scielo, covering publications in Portuguese, English and Spanish between 2019 and 2024. Original studies, systematic reviews and meta-analyses were included, excluding letters to the editor, editorials and opinion pieces. The analysis focused on studies investigating the composition of the microbiome in stroke patients, mechanisms of interaction between the microbiome and the central nervous system, and therapeutic interventions such as the use of probiotics and prebiotics. **Results:** The review revealed that stroke patients often present with dysbiosis, characterized by a reduction in microbial diversity and a predominance of pro-inflammatory bacteria. In this sense, suggested mechanisms include bacterial translocation and modulation of the gut-brain axis, influencing systemic inflammatory responses and neuroinflammation. Interventions with probiotics, especially with strains of Lactobacillus and Bifidobacterium, have shown potential in restoring the healthy composition of the microbiome, reducing inflammatory markers and improving functional recovery in pre-clinical and clinical models. In addition, diets rich in fiber and prebiotics have shown benefits in modulating the intestinal microbiota and protecting against cerebrovascular lesions. **Conclusion:** Modulation of the gut microbiome is emerging as a promising strategy for stroke prevention and treatment. Understanding the mechanisms by which the microbiome influences cerebrovascular health is key to the development of innovative therapies. Personalized interventions based on the individual microbial profile could optimize clinical outcomes, offering a new approach to post-stroke rehabilitation. This interdisciplinary field has the potential to transform clinical practice and significantly improve the quality of life of patients with cerebrovascular diseases.

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The joint increase in deaths and hospitalizations of patients affected by malignant neoplasm of the brain in Northeastern Brazil: analysis of a 12-year epidemiological overview

Marconny Alexandre Oliveira de Medeiros Cavalcante[1], Thiago Luís Marques Lopes[1], Mateus Dutra Balsells[1], Hildel Freire Leite Filho[1], Matheus Lima de Oliveira[1], Oliver Reiks Miyajima[1], Lorenzo Marinho Morais[1], Késia Sindy Alves Ferreira Pereira[1], Chiara Gübel Portugal[1], Daniel Pereira dos Santos[1]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Marconny Alexandre Oliveira de Medeiros Cavalcante; Email: alexandremarc1912@gmail.com.

Background: Malignant brain neoplasms (MBN), also known as brain cancers, represent the eighth most prevalent type of tumor in adults over forty years of age, and metastases are closely related to other very prevalent neoplasms, such as breast, skin and lung neoplasms. Due to the limitation of standard therapies, MBN are often endowed with a poor prognosis and the quality of life of patients is greatly affected. Despite this, this is a topic that, in the Brazilian northeast, needs further investigation. Thus, based on the need for a better understanding of this scenario, the justification of this study is given by the intention to better understand this panorama through epidemiology. **Objective:** To analyze the relationship between deaths and hospitalizations caused by MBN in the northeastern region of Brazil, using a twelve-year epidemiological overview. **Methods:** As this is an epidemiological study of secondary data, of the observational, descriptive and analytical type, the "TabNet" platform was accessed, where the axis "Epidemiological and Morbidity" and the topic "Hospital Morbidity of the SUS (SIH/SUS)" were chosen. Next, we selected "General, by place of hospitalization - from 2008 onwards" and "Brazil by Region and Federation Unit". Subsequently, in order to obtain the data for this study, the line "Age group 1", the column "Year of processing" the period "Jan/2012- Dec/2023" and the contents "deaths" and "hospitalizations" were adopted. Finally, from the available selections, "2 Northeast Region" and "Malignant neoplasm of the brain" were chosen in the ICD-10 Region and Morb List, respectively. **Results:** Considering the period from January 2012 to December 2023, there were 4,635 deaths due to MBN in northeastern Brazil, of which the majority, 2,553 (55.1%), were recorded in people aged 50 to 79 years. Dividing this period into four three-year intervals, it is possible to perceive, comparing the 2012-2014 interval (1,037 deaths) with the 2021-2023 interval (1,324 deaths), an increase of 27.7% in the number of deaths. In the case of hospitalizations, during the 12-year period mentioned above, 35,646 occurrences were recorded, of which a considerable portion, 16,659 (46.7%), were associated with people aged 40 to 69 years. Comparing the last three years (10,506 hospitalizations) with the oldest three-year period under analysis (8,165 hospitalizations), an increase of 28.7% is seen. **Conclusion:** Therefore, it is inferred that, together with a higher number of hospitalizations due to MBN treatment, there was a relevant increase in the number of deaths related to this scenario. In addition, the older portion of the adult population and the younger portion of the elderly population were found to be the most affected by the incidence of hospitalizations and deaths associated with this disease. Thus, due to this progressive relevance, more and more studies are needed to improve the management of this scenario of increasing aggravation.

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The MCT8 deficiency (Allan-Herndon-Dudley syndrome) from the child neurodevelopment point of view

Marina Ottmann Boff[1], Gabriella Sityá Moojen da Silveira[1], Ana Carolina Baldi Pasqualini[1], Alessandro Batista Soares[1], Eduarda Gandolfi Horst[1], Vanessa Pellenz Soares[1], Ana Lima[1], Luísa Castro Gomes[1], Frederico Orlando Friedrich[1], Magda Lahorgue Nunes[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Marina Ottmann Boff; Email: mottmannboff@gmail.com.

Background: Thyroid hormones (TH) are crucial for the regulation of the metabolism and development, including that of the brain. A mutation in SLC16A2 causes the deficiency of MCT8, a TH transport protein in the brain. It manifests as the Allan-Herndon-Dudley Syndrome (AHDS), which is characterized by severe intellectual and motor deficits resultant from the brain hypothyroidism and by multisystemic sequels secondary to the peripheral hyperthyroidism. A study aiming to identify which outcomes were most valued by the patients' parents showed that the improvement of motor and language abilities occupied 1st place, meaning that the AHDS's neurodevelopmental aspect is responsible for most of the burden in the patients' and their relatives' lives. **Objective:** To gather recent scientific evidence regarding the neurodevelopment in AHDS. **Methods:** This systematic review has been conducted in the Scopus, PubMed and Embase databases with the following Medical Subject Headings (MeSH) terms and boolean operators: "Neurodevelopmental disorder" AND "Monocarboxylate transporter 8 deficiency"; as recommended by the Preferred Reporting Items for Systematic reviews and Meta-Analysis (PRISMA). The inclusion criteria were original articles, open access, published from 2014 to 2024, in english or portuguese, and the exclusion one was non-correspondent text subject. The initial search resulted in 10 distinct articles from which 9 were selected. **Results:** In the nervous system SAHD causes hypomyelination, impairment of the differentiation of GABAergic interneurons and delay in the development of the cerebellum. Global hypotonia, hypertonia due to dystonia and spasticity and persistence of primitive reflexes are present in 90% of the cases and their identification is essential for surveillance and early differential diagnosis. Treatments are based on the administration of THs or their analogues. Triiodothyroacetic acid, an MCT8-independent THs metabolite, inhibits THs production and has low peripheral thyromimetic activity, but it has only been evaluated for neurodevelopment preclinically and exploratorily, presenting to be effective on the neurodevelopmental aspect only if administered early. Sob-AM2, a prodrug of an HT analogue capable of crossing the placental and blood-brain barrier and being specifically metabolized in the brain, is a strong candidate for the prenatal treatment of SAHD, since neurological improvement depends on early treatment. The effectiveness of levodopa, carbidopa and botulinum toxin is quite variable between patients, and may improve spontaneous movements temporarily or not. There was no improvement in the development with diiodothyropropionic acid (recommended treatment), T3 and T4. **Conclusion:** Given the rarity of SAHD, little scientific evidence is produced about it. Even case reports are extremely important for its better characterization. Ongoing trials should better elucidate its treatment from a neurodevelopmental perspective.

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The mitigation of "neurophobia" among medical students through the use of active learning methodologies in neuroanatomy

Samuel Natal Rodrigues[1], Eduarda Kei-Lan Tavares Pierre[1], Cristiane Aschidamini[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.

Correspondence: Samuel Natal Rodrigues; Email: snr.med20@uea.edu.br.

Background: Neurology is a broad medical field that requires great intensity of training and studying to master it. However, there is only a small percentage of professionals who have neurology as their specialty of choice, in contrast with the growing necessity of qualified physicians in the area. The construction of knowledge begins in neuroanatomy, the anatomic field focused on the nervous system, which is an integral portion of the basic sciences curriculum at medical schools. Nonetheless, despite the appreciable value of the subject, neuroanatomy is one of

the most pronounced obstacles for medical students, especially due to difficulties in exposing the material clearly, new technical language for students, and challenges in understanding anatomical elements and linking them to clinical knowledge. These problems lead to a negative perception among scholars regarding neuroanatomy and jeopardize a possible future in neurology. Thus, since 1994, the term "neurophobia" has existed to characterize this immense fear of neurological apprenticeship that a large portion of students can relate to, which is intensified by the first contacts with neuroanatomy. **Objective:** To analyze the efficacy of active learning methods in the teaching of neuroanatomy in order to reduce neurophobia, according to the perspective of first-year students enrolled in a Human Anatomy course at a public medical school in Amazonas, Brazil. **Methods:** A longitudinal, exploratory, descriptive study was conducted with a qualitative-quantitative approach through administration of surveys in a virtual environment. The goal was to evaluate study participants in 3 distinct phases: 1) pre-learning process assessment; 2) process of learning neuroanatomy with active learning methodologies; and 3) post-process assessment of learning. **Results:** Over 50% of study participants, out of 80 in total, had been aware of a "negative reputation" of neuroanatomy, and this knowledge influenced the participant's own perception in 88% of cases; the main factors hindering the study of neuroanatomy were the neuroanatomical terminology and the complexity of the subjects covered by the subject; 75% of participants evaluated the study of neuroanatomy as an important factor in deciding whether or not to delve deeper into future studies in neurology; 95% of participants considered the active methodology in learning neuroanatomy as an important factor in mitigating the symptoms of neurophobia. **Conclusion:** Therefore, it is evident that this study is aligned with the current literature on the topic which states that active learning methodologies in teaching neuroanatomy represent a vital importance in reducing the effects of neurophobia and encourage interest in the future study of neurology among students.

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The moral history of seizures: reported causes of seizures in the 19th century

Márcio Pinheiro Lima[1], Adriel Rêgo Barbosa[1], André Felipe Candêas Amorim[1], Gabriel Pinheiro Martins de Almeida e Souza[1], João Victor Cabral Correia Férrer[1], Aline Agatha Lima de Oliveira[1], Rafael Tuzino Leite Neves Maffei[1], Ruan Gambardella Rosalina de Azevedo[1], Victor Cardoso de Faria[1], Fernanda Lustosa Cabral Gomez[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Márcio Pinheiro Lima; Email: mplima1997@gmail.com.

Background: The first description of epilepsy dates from approximately 2000 BC, from an Akkadian tablet describing a person with "his neck turning left, hands and feet are tense, and his eyes wide open, and from his mouth froth is flowing without him having any consciousness". Nowadays epilepsy is recognized conceptually as "a disorder of the brain characterized by an enduring predisposition to generate epileptic seizures". Seizures were attributed to the different causes throughout history. In the 19th century neurology started organizing as a medical specialty, with many of its great names (such as Charcot, Duchenne and Romberg) acting in this time period. However, in the scientific communications of the time it's often possible to see "unwanted" or "antisocial" daily habits and other characteristics mentioned as causes of seizures, many times with judgmental undertones. **Objective:** Report some of the causes for seizures seen in case reports and statistical tables from 1800-1900 and their relationship to moral concepts at the time. **Methods:** A literature review was made on the PubMed and SciELO databases for works containing the words "epilepsy", "seizure", "epilepsia", "convulsão", convulsión", and "crise epiléptica" from 1800 to 1900. 666 papers were found, all in the English language. Those papers were analyzed for descriptions of the causes of epilepsy, either describing current medical theories or personal opinions of the professionals. **Results:** In the timeframe analyzed it was possible to find various references to "abnormal" behaviors causing epilepsy. Multiple papers ascribed the development of epilepsy in children to mistakes made by the mother during feeding, such as "inadequate attention". The idea that different kinds of personality would be imprinted on the children was also seen, with several papers describing that children of "nervous" or "hysterical" mothers would be predisposed to epilepsy. There were also descriptions about the possibility of cure if the parents raise the children correctly. Sexual behavior was also thought to be correlated with epilepsy, with multiple sources correlating masturbation with seizures - with one source explicitly describing masturbation as evil, and several papers describing the excitation of the genital organs as the origin of reflex epilepsy. The behavior of women was scrutinized, with a report attributing seizures to the use of heavy dresses with the consequent pressing down of the ovaries. Women were also described as more emotional, and therefore more prone to epilepsy, especially after puberty. **Conclusion:** Even though the 19th century was a time of scientific experimentation for neurology - then a budding specialty – it's possible to notice multiple examples of how the moral thought of the European/North American society of the time influenced the thought about epilepsy. This serves as a reminder of the influence of the notions of the wider society in medicine.

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The negative impact of anxiety-depression symptons on cognition after mild COVID-19: analyses of 151 individuals without past history

Beatriz Amorim da Costa[1], Leila Camila Santos Silva[1], Mateus Nogueira[1], Marjorie Cornejo Pontelli[1], Ana Paula Beppler Lazaro Lino[1], Lucas Scárdua Silva[1], Italo Karmann Aventurato[1], Fernando Cendes[1], Clarissa Lin Yasuda[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Beatriz Amorim da Costa; Email: b165550@dac.unicamp.br.

Background: Although cognitive impairment has been observed in patients with LONG-COVID, little is known about the negative impact of mood dysfunction on cognition after mild infection. **Objective:** To investigate the impact of symptoms of fatigue, daytime somnolence, anxiety and depression on the neuropsychological performance of patients (without psychiatric history) after mild SARS-CoV-2 infection. **Methods:** We studied 151 post-COVID-19 patients with a median of 85 days after diagnosis (range 18-420). Subjects had a median of 15 years of education and no history of psychiatric events. Individuals responded the BDI (Beck Depression Inventory) and BAI (Beck anxiety inventory) for quantification of symptoms; we then divided the subjects into ASYMPT-Group (no symptoms of depression or anxiety) (69 subjects), DOUBLE-group (combined symptoms of anxiety and depression) (43 subjects), ANX-group (exclusively anxiety symptoms) (30 subjects) and DEPRESS-group (exclusively depressive symptoms) (9 subjects). Daytime somnolence was assessed with the Epworth Sleepiness Scale (ESS) (median 10 points, range 0-21), and fatigue was evaluated with the Chalder Fatigue Scale (CFQ-11) (median 18, range 0-33). We included different cognitive test domains, including language (Verbal Categorical Fluency Test and Phonemic Verbal Fluency Test), verbal episodic memory (Logical Memory), visuoconstruction and visuospatial episodic memory (the Rey Complex Figure Test), manual dexterity (9-hole Peg Test), selective attention (FDT-Choice), inhibitory control (FDT-Inhibition), alternate attention and cognitive flexibility (FDT-Alternation and Flexibility), and sustained attention, perceptual tracking and motor skills (the Color Trail Test [CTT]-parts A and B). Subjects were balanced for diagnosis interval (p=0.24), sex (p=0.90) and age (p=0.86). We performed Mann-Whitney tests to compare continuous variables among groups and Spearman tests for correlations. **Results:** The DOUBLE-group presented higher scores of somnolence (ESS) and fatigue (CFQ) compared to the ASYMPT-group (p<0.005) and ANX-group. Overall, the cognitive performance was worse than expected for most of the patients, according to their age and education. Performance of language, verbal memory, mental flexibility/inhibition and FDT were more affected in the DOUBLE-group compared to the ASYMPT-group (p<0.05). Manual dexterity z-scores (bilaterally) were similarly low for all groups. **Conclusion:** Our results suggest that any individual can present some degree of cognitive dysfunction after a mild infection. The impairment seems worse in individuals with symptoms of anxiety and depression (who also showed more symptoms of fatigue and somnolence). Neuropsychiatric and cognitive dysfunction in long-covid are part of a complex phenomenon and require multidisciplinary attention for effective resolution.

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The nobel prize awarded to Torsten Wiesel and David H. Hubel: progress in visual neuroscience and its connection to neuroesthetics

Maria Clara Jackson de Carvalho Sousa[1], Luiz Henrique Mattos da Silva Santos[1], João Ignacio Ferrara Neto[1], Maria Isabella Osório Cavalcanti de Jardim Sayão[1], Roberto Pereira Santos[1], Marleide da Mota Gomes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Maria Clara Jackson de Carvalho Sousa; Email: mariaclarasousa93@gmail.com.

Background: Torsten Wiesel, the Swedish neurophysiologist, worked in visual neuroscience, in collaboration with David H. Hubel (1926-2013), which has significantly advanced our comprehension of how the brain processes visual stimuli. As we honor the birth anniversary of Wiesel, it is imperative to recognize his contributions to the field of neurophysiology. Wiesel and Hubel were co-recipients of the Nobel Prize in Physiology or Medicine in 1981 for their research on the complexities of visual information processing. Their partnership marked an advance in visual neuroscience, illuminating the brain's interpretation and response to visual stimuli and laying the groundwork for the neuroesthetics. **Objective:** This paper aims to delve into the contributions of Torsten Wiesel and David H. Hubel to the field of visual neuroscience. Additionally, the paper seeks to explore the concept of neuroesthetics and its role in uncovering the neural mechanisms underlying aesthetic experiences. **Methods:** To accomplish the objective, a comprehensive review of the seminal research conducted by Wiesel and Hubel on the visual system will be conducted. By analyzing their experiments and findings, the key advancements in our understanding of how the brain processes visual information will be elucidated. Additionally, the emergence of neuroesthetics as a discipline will be explored. **Results:** The research conducted sheds light on the achievements of Torsten Wiesel and David H. Hubel in visual neuroscience, emphasizing their significant findings on orientation selectivity in visual cortex neurons and the columnar organization of the visual cortex. Additionally, the exploration of neuroesthetics demonstrates the discipline's potential to bridge art and science, offering fresh perspectives on aesthetic experiences. **Conclusion:** The contributions of Torsten Wiesel and David H. Hubel in visual neuroscience have provided

new insights into brain functioning. The emergence of neuroesthetics presents exciting possibilities for delving into the neural basis of aesthetic experiences and their implications for various fields. By combining the findings of Wiesel and Hubel with the principles of neuroesthetics, new avenues for research and collaboration at the intersection of art and science can be unlocked.

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The pain points of Alzheimer's disease pain management

Gabriella Chinaglia Zanin[1], Caio Hussid de Góes[1], Diego Olympio Peixoto Lopes[2], Maria Tereza Martinez[3], Fernanda Herculano[4], Diogo Haddad Santos[4]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Universidade Santo Amaro, São Paulo SP, Brazil.
[3]CP Quali, São Paulo SP, Brazil.
[4]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Gabriella Chinaglia Zanin; Email: gabriella.zanin@aluno.fcmsantacasasp.edu.br.

Pain is commonly reported by patients with Alzheimer's disease, and daily pain rates could be as high as 70% of nursing home patients. Consequently, analgesic medications are some of the most prescribed drugs for patients with Alzheimer's disease, and yet current literature shows that undertreatment of pain is common in context of dementia, as it is a referred symptom and dementia patients progressively loose communication ability, concurrent with a progressive increase in nociception disturbances. Thus, the present study's aim was to analyze the use of analgesic medications prescribed to patients with Alzheimer in the last 5 years in reference neurology center in São Paulo, while comparing it to the general Brazilian usage of analgesic in Alzheimer patients. An electronic medical record aggregator called TriNetX was used to anonymously obtain the patient data used in the study, using exclusively hospital related records. The Brazilian cohort was built using a database that coalesces patient records of several institutions throughout national territory, via selecting patient with Alzheimer diagnosis who were at least 50 years old when they received the diagnosis. The reference center cohort was built using the same criteria as the Brazilian but pulled data only from the database of a reference neurology center in São Paulo. Analgesic use event was defined as the first prescription of the drug, regardless of time of use or subsequent administrations. Test results revealed a Brazilian cohort of N=5,944, of whom 42% had been prescribed any type of analgesic at some point. Opioids where the most common at 34%, distributed between Tramadol at 22%, Morphine at 18%, Fentanyl at 9% and Remifentanil at >1%. Non-opioids usage totaled 24% of patients, with acetaminophen at 12% and metamizole at 11%. The reference center cohort had N=250 patients, whom 48% had been prescribed any type of analgesic. Opioids where the most common at 40%, distributed between Tramadol at 28%, Morphine at 28%, Fentanyl and 16% and Remifentanil at 12%. Non-opioids usage totaled 24% of patients, with acetaminophen at 16% and metamizole at 16%. While prevalence of chronic pain in community dwelling patients with Alzheimer is close to 40%, in nursing homes the prevalence increases to between 60 to 70%. Based on the obtained results, this may point to both an undertreatment, as even in the reference center only 48% of patients were prescribed any analgesic, possibly pointing to a difficulty of identifying pain in these patients, as well as an overtreatment, due to the preference for opioids, an option with unclear consequences as some studies suggest no negative impact of use, while others associated it with increased mortality. Thus, it can be concluded that pain management remains an important, complex and easily overlooked aspect of Alzheimer's disease treatment.

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The path of a young patient with a lower motor neuron syndrome: a case report

Filipe Tupinambá Di Pace[1], Frederico Mennucci de Haidar Jorge[1], Nicolas Ruan dos Santos Cavalcante[1], André Macedo Serafim Silva[1], Rodrigo de Holanda Mendonça[1], Mariana Hiromi Manoel Oku[1], Gabriella Corrêa Dousseua[1], Gabriel Saboia de Araújo Torres[1], Dagoberto Callegaro[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Filipe Tupinambá Di Pace; Email: filipedipace@gmail.com.

Lower motor neuron syndromes encompass diseases affecting the anterior horn cell, the motor axon, and/or its surrounding myelin, sometimes clinically indistinguishable from neuromuscular junction disorders. Here, we present a case of a young man with progressive motor neuron degeneration. **Case presentation:** A 15-year-old right-handed male with no significant medical history started complaining about weakness in the abduction of the fifth digit of his right hand. Over several months, this weakness extended to his entire hand, accompanied by significant muscle wasting, leading to writing impairment. Eighteen months later, similar symptoms manifested in his left hand. After multiple investigations, including neurophysiological studies, the diagnosis was classified as motor neuron disease localized to the cervical spine with no clear etiology. Initial treatment included corticosteroids and physical rehabilitation. The patient's condition continued to progress slowly, with increased weakness and muscle wasting in both hands. Four years after the onset of the symptoms, he was referred to our service, now with right forearm weakness and wasting. The electromyography showed signs of active denervation (fibrillations) at rest and a chronic reinnervation pattern in voluntary muscle activation in C7-T1 myotomes bilaterally (predominance on the right). A dynamic cervical MRI revealed a "snake eye" sign and mild spinal atrophy from C5 to C6, along with posterior epidural enlargement during neck flexion. These findings confirmed the diagnosis of Hirayama disease. Following spinal surgery, the patient's disease progression halted, and he experienced some improvement in both hands, now able to write with adaptations. **Discussion:** Hirayama disease is a rare motor neuron disorder predominantly affecting unilateral or asymmetrical bilateral upper extremities, more common in juvenile Asian males. It is typically benign and self-limiting course but can occasionally lead to significant disability and progressive symptoms over time. The optimal treatment strategy remains debated, including long-term neck brace use or surgical intervention. Our decision to proceed with surgery was influenced by the patient's rapid initial progression, the four-year duration of unstable disease, and the patient's informed preference after discussing the available options. **Final comments:** This case exemplifies a classical presentation of Hirayama disease, which, despite extensive investigation, took four years to be diagnosed, resulting in irreversible damage. Awareness of rare but classic diseases, coupled with an integrated healthcare system facilitating specialist referrals, is crucial to prevent permanent disability in similar patients.

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The pattern of neuropsychological performance according to presence and side of hippocampal sclerosis in mesial temporal lobe epilepsy

Carolina Ferreira Tessler[1], Luciana Ramalho Pimentel-Silva[1], Bruna Augusto Martins[1], Mateus Henrique Nogueira[1], Clarissa Lin Yasuda[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Carolina Ferreira Tessler; Email: carolinatessler@gmail.com.

Neuropsychological assessment has emerged as an indispensable instrument in the comprehensive care of individuals with mesial temporal lobe epilepsy (MTLE). This approach plays a fundamental role in enhancing the accuracy of diagnosis, tailoring effective treatment strategies, and refining prognostic considerations for MTLE patients. The significance of neuropsychological evaluation becomes particularly evident when distinguishing performance patterns among MTLE patients with or without hippocampal sclerosis (HS) and side of the lesion. Therefore, we aimed to differentiate the cognitive performances associated with MTLE. We selected 167 individuals with MTLE, divided according to the presence of HS on the MRI: 23 MRI-negative, 54 had right-HS, 80 had left-HS, and 10 had bilateral-HS. The neuropsychological assessment utilized the following tests: Wechsler Memory Scale - Revised; Wechsler Adult Intelligence Scale - Revised; Phonemic and Semantic Verbal Fluency Tests; and the Boston Naming Test. To test differences in epilepsy duration and cognition, we employed univariate ANOVA or ANCOVA and partial correlation controlling for education. The duration of epilepsy was similar among groups, except between MRI-negative and left-HS ($p=0.022$), with longer duration in the second group. We found differences in the performance of the cognitive domains of verbal memory, general memory, delayed memory, and semantic verbal fluency between MRI-negative and right-HS (all $p\leq0.009$), MRI-negative and left-HS (all $p\leq0.01$), and MRI-negative and bilateral-HS (all $p\leq0.004$), with all the HS groups showing poorer performance when compared to MRI-negative. Therefore, MRI-negative MTLE patients had better performance when compared to patients with right, left, and bilateral HS in the cognitive domains related to verbal, general, and delayed memory, as well as language, suggesting that the presence of HS, regardless of side, influence the cognitive impairment in MTLE. In this series, we found no difference in verbal and visual memory between patients with right and left HS.

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The potential of cannabidiol as a novel adjuvant therapy for myotonic dystrophy type 1 (Steinert's disease): a case report

Fernanda Valeriano Zamora[1], Andres Villca Zamora[2], Ana Clara F. F. Santos[3], Lorhayne Kerley Capuchinho Scalioni Galvao[2], Nicole S. Pimenta[1], João Pedro C.E. A. Salles[1], Afshan Nizami[4], Camila M. Yamada[2], Deivyd Vieira Silva Cavalcante[5], Dayana Dourado de Oliveira Costa[6]

[1]Universidade Estadual do Rio de Janeiro
[2]Universidade Federal de Minas Gerais
[3]Universidade Cidade de São Paulo, São Paulo SP, Brazil.
[4]Apollo Institute of Medical Sciences and Research, Telangana, India.
[5]Universidade Federal do Maranhão, São Luís MA, Brazil.
[6]Universidade Ceuma, São Luís MA, Brazil.

Correspondence: Fernanda Valeriano Zamora; Email: dra.fernandavaleriano@gmail.com.

Case presentation: Male patient, 17 years old, diagnosed with Steinert's disease at age 14, presents with significant language delay, impairments in communication and social interaction, as well as restricted and repetitive patterns of behavior, interests, or activities. He also experiences progressive muscle weakness, myotonia, muscle fatigue, and sleep-related respiratory difficulties, along with painful muscle spasms. The patient has undergone

various prior treatments, including Carbamazepine, sodium valproate, baclofen, and topiramate, as well as sessions of occupational therapy, speech therapy, psychology, and neuropsychopedagogy. While there is no specific treatment to reverse the effects of Myotonic Dystrophy Type 1, treatments can help improve quality of life and manage associated complications. It is crucial that the treatment plan be personalized for each patient, considering the severity of symptoms and the stage of the disease. However, the patient has experienced significant impairment in quality of life despite symptomatic treatment and multidisciplinary care. Given the patient's family history of schizophrenia in his father, it was decided to initiate therapy with Broad Spectrum Cannabidiol (CBD) at a dosage of 200mg/ml, 5 mg/kg/day, starting at low doses and gradually increasing. The patient showed improvement in overall condition, behavior, social interaction, and patterns of interest. He also reported reduced muscle fatigue and muscle spasms. There were no observed side effects from the medication. Liver transaminases showed a slight increase during treatment but stabilized after 3 months of follow-up. **Discussion:** CBD is already used to treat spasticity and muscle pain in chronic conditions such as multiple sclerosis, cerebral palsy, among others. Its potential use in Steinert's disease is attributed to its action on the endocannabinoid receptors CB1 and CB2, which promote muscle relaxation, neuroprotection, and anti-inflammatory effects. Its application has led to an improvement in these symptoms in this patient. It is also known that the presence of various cannabinoids in Broad Spectrum CBD allows for the use of lower doses to achieve its therapeutic potential, thanks to the entourage effect. This justifies the reported improvement of the patient with a low dosage of CBD. **Final comments:** CBD demonstrated potential in improving overall condition, behavior, social interaction, and alleviating muscle symptoms without adverse effects, suggesting it could be a beneficial adjunctive treatment for managing symptoms of Steinert's disease. Further research is warranted to confirm these findings and investigate CBD's long-term efficacy and safety in similar patient populations.

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The potential peripheral effects of long COVID: critical neuropathy after COVID-19 infection: a case report

Antonia Cardoso Silva[1], Olagide Wagner Castro[1], Ênio José Bassi[1], Amanda Pacheco[1], Abelardo Silva Júnior[1], Fernanda Souza[1], Alanna Michella Oliveira de Albuquerque da Silva[1], Gabriel de Oliveira Souza[1], Sarah Antunes Figueiredo[1], Mariana Reis Prado[1]

[1]Universidade Federal de Alagoas, Maceió AL, Brazil.

Correspondence: Antonia Cardoso Silva; Email: antonia.silva@famed.ufal.br.

Case presentation: Patient, male, 32 years old, with grade 1 obesity, diagnosed with COVID-19 in November 2020. He progressed to acute respiratory failure, requiring orotracheal intubation (OTI) for 35 days, followed by a tracheostomy. He developed pneumonia, laryngotracheal injury, and laryngotracheobronchospasm, leading to the decision for endoscopic gastrostomy and removal of the nasoenteric tube. He developed peritonitis, necessitating an exploratory laparotomy, which revealed gastric injury. He presented with wound dehiscence, incisional hernia, and femoral head osteonecrosis. After 87 days of hospitalization, he had reduced mobility, requiring a wheelchair and rehabilitation sessions. In August 2023, with grade 2 obesity and after a mild case of COVID-19 reinfection, he developed long COVID (LC) symptoms. Clinical evaluation revealed current symptoms of fatigue, arrhythmia, dyslipidemia, anxiety, memory impairment, mild to moderate depression, insomnia, constipation, and hip pain. He also presented with paresis in all four limbs, more pronounced in the lower limbs, right foot drop and steppage gait, painful, thermal, and tactile hypoesthesia in the right lower limb with a similar pattern in the upper limbs. Electroneuromyography indicated peripheral sensorimotor axonal polyneuropathy with mild secondary demyelination, predominantly distal. There was significant involvement of the right fibular nerve, with an axonal pattern throughout its extent, and moderate involvement of the left tibial nerve, with an axonal pattern in the middle and lower third of the left leg. **Discussion:** Long COVID (CL) arises following acute infection with SARS-CoV-2 and is generally associated with severe cases of the infection, occurring across all degrees of severity. The syndrome encompasses physical, mental, and cognitive symptoms, with muscular and neuromuscular effects being among the most prevalent and persistent. CL is associated with muscular conditions characterized by the presence of peripheral and/or central demyelination. Additionally, neuromuscular alterations in CL may be linked to axonal neuropathies in peripheral and even cranial nerves. COVID-19 has been strongly linked to the condition of critical illness polyneuropathy (CIP), predominantly observed in severely ill patients subjected to prolonged intensive care. It is characterized by sensory and motor axonal neuropathic alterations. Causes include sepsis, systemic inflammatory response syndrome, microcirculation damage, and increased cytokine secretion, conditions that are possible in COVID-19 and CL. The mechanisms linking COVID-19 to axonal pathologies and the relationship between CL and CIP are not clearly elucidated. This reported case presents electroneuromyographic characteristics consistent with axonal sensory-motor peripheral neuropathy. **Final comments:** This case highlights the severe impacts of COVID-19 and CL, emphasizing the complexity of recovery and the importance of comprehensive care.

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The prevalence of delirium in hospitalized patients of a tertiary hospital in Curitiba.

Mariana de Oliveira Trintinalha[1], Giulia Schaidt[1], Juan Carlos Pacheco[1], Bruna Segaspini Felber[1], Letícia Graniska Salanti[1], Carlos Alexandre Twardowschy[1]

[1]Pontifícia Universidade Católica do Paraná, Hospital Universitário Cajuru, Curitiba PR, Brazil.

Correspondence: Mariana de Oliveira Trintinalha; Email: marianaatrintinalha@hotmail.com.

Background: Delirium is a confusional state characterized by fluctuating symptoms, primarily affecting the elderly, which can lead to a series of detrimental events to the patient. The development of such a condition depends on multiple factors–modifiable or not–and the clinical presentation is often variable and fluctuant, making the diagnosis mostly based on clinical evaluation. Therefore, delirium is generally highly underdiagnosed. **Objective:** This study aimed to assess the prevalence of delirium in patients admitted to a tertiary hospital. **Methods:** A cross-sectional observational study was conducted between October 2022 and March 2023 on hospitalized patients over 18 years. Epidemiological data (age, current and previous health status, presence of metabolic, neurological, and infectious factors) were collected and the diagnosis of delirium was using the Confusion Assessment Method (CAM). The results obtained were analyzed, according to the type of variable considered, using the Chi-square test or Fisher's exact test, where applicable; the non-parametric Mann-Whitney test; and the Wald test. The confidence interval used was 95%, with p-values < 0.05 indicating statistical significance. **Results:** Among the patients included (n=100), 76% did not have a neurological cause for hospitalization and 83% did not require mechanical restraint. The analysis showed that 20% of the sample experienced delirium during the interview, with metabolic factors being the most common in the population. The risk factor most associated with the occurrence of the phenomenon was the use of mechanical restraint during hospitalization. **Conclusion:** Delirium has a high prevalence even in patients without neurological causes or comorbidities, thus healthcare professionals need to broaden their knowledge on the topic to identify and prevent it, reducing its occurrence during hospitalizations. The routine use of mechanical restraint should be avoided, as it represents the main precipitating factor of delirium. Clinical observation of hospitalized patients with risk factors is essential for diagnosis.

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The prevalence of migraine in Brazil from 2013 to 2023

Rebeca Alessi Tedeschi Pachega[1], Daniele Pelegrini Coral[1], Guilherme Henrique Dalaqua Grande[1], Leandra Ernst Kerche[1]

[1]Universidade do Oeste Paulista, Presidente Prudente SP, Brazil.

Correspondence: Rebeca Alessi Tedeschi Pachega; Email: rebeca.alessi@hotmail.com.

Migraine is a prevalent global health issue and the most common neurological disorder worldwide, significantly impacting public health. It predominantly affects females more than males, with a ratio of 3:1 respectively, and ranks as a leading cause of years lived with disability among adult women. The condition affects approximately 15% of the global population, exceeding one billion individuals. Prevalence is highest in the working-age population, particularly between 25 and 55 years, resulting in an annual economic burden of 27 million dollars due to debilitating migraine episodes. In Brazil, the annual prevalence is 15.8%, affecting 22% of women and 9% of men aged between 30 and 50 years old. This ecological research is crucial for optimizing public policies aimed at the migraine-affected population. To analyze migraine hospitalization rates in Brazil from 2013 to 2023, focusing on gender, age groups, and years of disability (YLDs). This ecological research utilized databases from the Hospital Information System of Brazil (HIS/SUS) and the Burden of Diseases database for 2021. Prevalence rates per 100,000 inhabitants were analyzed for hospital admissions related to migraines and other headache syndromes (ICD-10-G43) from 2013 to 2023 in Brazil. The analysis included age groups, gender distribution, public healthcare costs, and disability duration (YLDs) caused by migraine. The study investigated migraine-related hospitalizations in Brazil from 2013 to 2023, revealing a higher prevalence among females than males, with an approximate ratio of 2:1. A significant peak occurred in 2019, with prevalence rates of 4.21:7.26 (men:women), resulting in 7,796 female hospitalizations. The main affected age group was 20-59 years old, accounting for 69.4% of total hospitalizations. In these cases, 69% were women, more than twice the number of men admitted. During this period, hospital services incurred total costs of 26,712,306.73 Brazilian reais funded by SUS. According to the Burden of Diseases in 2021, women accumulated nearly 27 million years of disability due to migraine (YLDs), which is 30% higher than men. In Brazil, in the same year, over one million days were lost due to migraine, double the number observed in men.Migraine represents a significant public health concern, profoundly affecting quality of life and national economies due to productivity losses and healthcare expenditures. The analysis of migraine hospitalizations in Brazil in this article emphasizes a consistent trend of higher prevalence among economically active women. Furthermore, there is a need for additional research to refine treatment strategies and enhance understanding across diverse demographics. These initiatives aim to reduce hospital admissions, lower healthcare expenditures, and improve the quality of life for individuals living with chronic migraines.

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The profile of hospitalizations for multiple sclerosis in Brazil

Annayza de Oliveira Bessa[1], Deborah Araujo Silva[1], Rynele Almeida de Fonseca[1], Larissa Bianca Borsone[1], Amanda Oliva Spaziani[1], Vanessa Maria Gonçalves de Souza[1], João Carlos Bizinotto Leal de Lima[1], Luis Carlos Spaziani[2], Gustavo Henrique da Silva[3], Francieli Gomes Lima[4]

[1]Universidade Brasil, Fernandópolis SP, Brazil.
[2]Fundação Educacional de Fernandópolis, Fernandópolis SP, Brazil.
[3]UPA Santa Fé do Sul,Santa Fé do Sul SP, Brazil.
[4]AME Fernandópolis, Fernandópolis SP, Brazil.

Correspondence: Annayza de Oliveira Bessa; Email: annabessa01@gmail.com.

Background: Multiple sclerosis (MS) is a chronic autoimmune disease of the central nervous system characterized by the development of autoantibodies against the myelin sheath of neuronal axons. This results in inflammatory sites and oxidative stress, leading to neurodegeneration and demyelination of affected neurons. MS is a global disease with incidence varying by geography, age, ethnicity, and sex. It is one of the leading causes of neurological symptoms in young adults, with no known cure, primarily affecting individuals between 20 and 40 years old. The main symptoms include fatigue, paresis, spasticity, weakness, mood alterations, apathy, euphoria, anxiety, depression, and issues with physical and cognitive functions. **Objective:** To evaluate the profile of hospitalizations for Multiple Sclerosis in Brazil from 2019 to 2023. **Methods:** Data collection was performed using publicly available information from the Hospital Information System of the Brazilian Unified Health System (SIH/SUS) on Tabnet/DATASUS between January 15 and May 15, 2024. This retrospective longitudinal study employed a quantitative descriptive design. Data were grouped by Brazilian macro-regions, service classification, sex, age group, and race/ethnicity. Statistical inference was conducted using BioEstat 5.3 software, with paired t-tests for related samples to compare groups and data presentation using descriptive statistics, central tendency, and dispersion measures. **Results:** Between 2019 and 2023, there were 27,586 hospitalizations for Multiple Sclerosis in Brazil. The majority (69.10%, n=19,062) were among females (p=0.0023), and 56.59% (n=15,612) were among white individuals (p=0.0071). The age group most affected was 30 to 39 years, accounting for 28.05% (n=7,738; p<0.0001) of the hospitalizations. The Southeast region had the highest concentration of cases (n=20,790), representing 75.36% (p=0.0010) of the total, with an average annual cost of R$3,183,762.06 for treatments. Deaths were more frequent in the Southeast, with 43.53% (n=74) of the total (p=0.0056) and an average mortality rate of 6.65 (p=0.2311). The highest number of hospitalizations occurred in 2022, comprising 26.46% (n=7,299) of the total, compared to only 14.40% (n=3,972) in 2020. Most hospitalizations were for urgent care, accounting for 52.33% (n=14,436; p=0.2964) of the cases. **Conclusion:** Hospitalizations for Multiple Sclerosis are predominantly concentrated in the Southeast region of Brazil, with the highest proportionality of deaths and healthcare costs. Many cases occur among white females aged 30 to 39, with urgent care being the most common type of service required.

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The relationship between deaths and hospitalizations by age group caused by epilepsy in Brazil: analysis of a 12-year epidemiological overview

Chiara Gübel Portugal[1], Lorenzo Marinho Morais[1], Thiago Luís Marques Lopes[1]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Chiara Gübel Portugal; Email: chiaraportugal@gmail.com.

Background: Epilepsy is defined as a brain disorder characterized by a chronic predisposition to generate epileptic seizures. These seizures are transient occurrences of signs and/or symptoms due to abnormal, excessive, or synchronous neuronal activity in the brain, alongside the neurobiological, cognitive, psychological, and social consequences of this condition. Epilepsy has a high prevalence, affecting approximately 50 million people worldwide and 2% of the Brazilian population. It is also one of the most common neurological disorders in childhood, challenging in terms of diagnosis and treatment, which vary depending on classification, type, and age group. **Objective:** Given the need for a better understanding of this scenario, this study is justified to analyze the epidemiological profile of patients and their respective therapeutic responses. **Methods:** This is an epidemiological study using secondary data, of observational, descriptive, and analytical nature. Data were accessed through the "TabNet" platform, focusing on the "Epidemiological and Morbidities" axis and selecting the topic "Hospital Morbidity of SUS (SIH/SUS)". The subtopic "General by Place of Hospitalization from 2008" was chosen. The study period covered "2012-2023", examining "Hospitalizations per Year processed by Age Group 1", "Deaths per Year processed by Age Group 1", and "Morbidity List CID-10: Epilepsy". **Results:** From 2012 to 2023, there were a total of 639,924 hospitalizations and 15,539 deaths due to epilepsy. Of all hospitalizations, 47,433 (7.4%) occurred in children under one year old and 106,832 (16.7%) in the age group of 1 to 4 years, which had the highest number of hospitalizations, followed by a 54% decrease in hospitalizations in the 5 to 9 years age group. In terms of deaths, 461 (3%) occurred in infants under 1 year old and 502 (3.2%) in children between 1 and 4 years old, followed by a 50% decrease in the 5 to 9 years age group. Based on these data, the mortality rate among hospitalized patients was 0.97% for children under 1 year old and 0.47% for those aged 1 to 4 years. **Conclusion:** Therefore, there is a controversy in the age group of children under 1 year old, as despite having fewer hospitalizations, they proportionally have twice the number of deaths compared to children aged 1 to 4 years. This suggests lower therapeutic efficacy and a higher likelihood of lethal outcomes in infants under 1 year old. The decrease in hospitalizations and deaths among children aged 1 to 4 years indicates that appropriate diagnosis and treatment of epileptic syndromes reduce the recurrence of seizures, emphasizing the importance of studies that continually improve the management of this condition.

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The relationship between response to antiseizure medication and temporo-limbic atrophy in patients with mesial temporal lobe epilepsy and hippocampal sclerosis

Pamela Regina Henning[1], Aline de Fátima Dias[1], William Souza Martins Ferreira[1], Lucas Scárdua Silva[1], Luciana Ramalho Pimentel da Silva[1], Marina Koutsodontis Machado Alvim[1], Clarissa Lin Yasuda[1], Fernandos Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Pamela Regina Henning; Email: pamrhenning@gmail.com.

Background: Mesial temporal lobe epilepsy (mTLE) associated with hippocampal sclerosis (HS) is the most common type of focal refractory epilepsy in adults. Quantitative MRI studies have shown evidence for progressive atrophy in these patients; however, the relationship between patterns of response to antiseizure medication (ASM) and the degree and distribution of gray matter atrophy is not yet clear. **Objective:** To correlate response to ASM with cortical surface area (CSA), cortical thickness and subcortical volumes (SV) in patients with mesial temporal lobe epilepsy with hippocampal sclerosis (mTLE-HS). **Methods:** 224 patients with mTLE-HS who did not undergo surgery because they refused it, had contraindications, or had good seizure control with ASM (mean follow-up of 17 years) were categorized in three groups: pharmacoresponsive (seizure-free in the last two years or more), fluctuating-response (seizure-freedom periods of one year or longer) and pharmacoresistant (never had a seizure-free period of more than one year). T1-weighted MRIs (1x1x1mm) were processed with Freesurfer. We analyzed the following regions of interest for CSA: inferior, middle and superior temporal gyrus, parahippocampal gyrus and insula. Volumes of thalamus, hippocampus and amygdala, along with the CSA from each ASM response-groups were compared with 223 controls. No violations of models' assumptions were found. **Results:** The 224 patients were balanced for sex, and side of HS. 58 were, pharmacoresponsive, 61 had a fluctuating-response to ASM and 105 were pharmacoresistant. Age of onset was earlier ($p<0.05$) in the pharmacoresistant group than the others. Compared to controls, there was significant volume reduction for ipsilateral hipoccampus and amygdala for the three groups and bilateral thalamus also in the three groups (all $p<0.002$). Significant reduced cortical thicknesses were found in ipsilateral limbic regions in all three groups (all $p<0.02$) and only contralateral parahippocampal region was reduced in pharmacoresponsive and pharmacoresistant groups ($p<0.005$). Regarding CSA, contralateral areas of limbic regions were significantly reduced ($p<0.04$) but not the ipsilateral CSA. Interestingly, for these ipsilateral regions, we found that age significantly adjusted the model, with weak to moderate effect sizes. Meaning that an effect of aging might be more sensitive to these changes than the clinical outcome. **Conclusion:** Contrary to our initial hypothesis, the findings of reduced CSA, cortical thickness and volume of limbic structures and subcortical regions did not differ among pharmacological responses in these patients. Factors other than the degree and distribution of gray matter damage should explain the differences in response to ASM in mTLE-HS.

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The relationship between the chronic use of benzodiazepines and the risk of developing Alzheimer's disease

Raquel Fakhouri Cardoso[1], Evelyn Victória Braselino[1], Marina Maiolino de Carvalho[1], Isabelle Campos Campi[1], Sarah Mística Simplício Silva[1], Mariana Lima de Moura[1], Jennifer Carvalho Garcez Obeid[1], Emanueli Vicente da Silva Honorato[1], Pedro Henrique Cabral Vilhena[1], Matheus Alves da Silva[1]

[1]Universidade Anhembi Morumbi, São Paulo SP, Brazil.

Correspondence: Raquel Fakhouri Cardoso; Email: raquelfakhouri@gmail.com.

Background: Alzheimer's disease (AD) is a chronic neurodegenerative disorder that manifests itself through cognition deterioration and compromise of daily life activities. AD occurs due to deposits of abnormal proteins in the central nervous system (CNS), creating neurotoxic protein fragments that induce a progressive neural loss in areas of the brain such as the hippocampus. In this context, recent evidence suggests that the chronic use of benzodiazepines (BZDs) is one of the risk factors for the development of AD. BZDs are psychotropic drugs whose mechanism of action consists of the depression of the CNS and can treat psychiatric diseases. However, its chronic use can lead to complications, as well as aggravation of pre-existing pathologies. **Objective:** To explore the relation between the effects of the chronic use of BZDs and

the development of AD. **Methods:** A PubMed Central database search was made in April 2024. The DeCS/MeSH descriptors "Alzheimer's Disease", "Benzodiazepines" and "Risk Factors" were used and filtered for publications from the last 5 years written in English. This resulted in 18 articles, 6 of which met the selection criteria of being free of charge and correlating with the topic studied. **Results:** BZDs are potentially inappropriate medications (PIMs) for the elderly, but are commonly used. Studies suggest that the prolonged use of BZDs promotes cognitive disorders in older persons and can predispose to the genesis and progression of AD, mild cognitive impairment, and dementia. Therefore, many hypothetical mechanisms aim to elucidate this relationship, such as the role of BZDs in the reduction in the number of chloride channels in the GABA A receptors; the alterations in synaptic plasticity and memory; as well as acting as intensifiers in the aggregation of hyperphosphorylated tau, enhancing the cytotoxicity and neurodegeneration in AD. Moreover, there are ponderations as to the neurotoxicity induced by the chronic use of BZDs being drug and dose-dependent, and occur mainly in females, however, while some studies highlight short and intermediate-acting BZDs as the major inducers of dementia, others delegate this importance to long-acting BZDs. Nonetheless, the association of the binomial BZD-AD is controversial, considering some studies have reported that BZDs do not compromise cognition or the progression of AD long-term, and even if it does happen, there are no significant disparities between genders. **Conclusion:** There is a questionable association between the chronic use of BZDs and AD, because not all studies highlight the potential impact of these drugs on the progression of cognitive deterioration in elderly, through the drug and dose-dependent neurotoxicity. Since there are disparities between studies on the association of chronic BZD use and DA, more investigations on this subject are needed to define these implications.

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The relationship between the increase in deaths from Alzheimer's disease and the heterogeneity of macro regions in Brazil: analysis of a 12-year epidemiological overview

Chiara Gübel Portugal[1], Lorenzo Marinho Morais[1], Thiago Luis Marques Lopes[1]

[1]Universidade Estadual do Ceará, Fortaleza CE, Brazil.

Correspondence: Chiara Gübel Portugal; Email: chiaraportugal@gmail.com.

Background: Alzheimer's disease (AD) is currently the leading type of dementia. This condition is neurodegenerative, leading to cognitive and functional decline, making it a major source of morbidity and mortality among the elderly population. The estimated prevalence of AD is 50 million cases worldwide and 1.6 million in Brazil, with projections suggesting a tripling of cases by 2050. Despite many advances in research, the disease's pathophysiology is not fully understood. Therefore, this study aims to better analyze the progression trend and evaluate the patient population profiles, focusing on mortality data. **Objective:** To analyze the epidemiological profile of deaths by region in Brazil over a 12-year period. **Methods:** This is an epidemiological study using secondary data, of an observational, descriptive, and analytical type. Data were accessed through the "TabNet" platform, selecting the "Epidemiological and Morbidities" axis and the topic "Hospital Morbidity of SUS (SIH/SUS)". The subtopics "General by Place of Hospitalization from 2008" and "Brazil by Region and State" were then selected. The period analyzed was "2012-2023", focusing on "Alzheimer's Disease", "Deaths by Age Group", and "Deaths by Region". **Results:** Between 2012 and 2023, there were 3,524 recorded deaths from AD, 98.6% of which were among the elderly (3,474). Dividing this period into three four-year intervals (2012-2015, 2016-2019, and 2020-2023), there were 765, 1,240, and 1,519 deaths, respectively. In percentage terms, there was a 62% increase between the first two intervals and a 22.5% increase between the last two, totaling an overall increase of approximately 99%, which aligns with the global trend. However, when breaking down the deaths by region, significant regional discrepancies are evident. The Southeast region had the highest number of deaths (2,167), with a 72% increase, while the North, Northeast, South, and Central-West regions showed increases of 136%, 172%, 87%, and 721%, respectively. **Conclusion:** The increase in deaths from AD was significant, but this increase did not follow a uniform pattern across Brazil's five macroregions, a higher number was expected in the Southeast, as it is the most populous region, but it obtained a lower percentage increase. This reinforces the idea that, in addition to the aging population, other modifiable risk factors that vary by region–such as education, hypertension, smoking, depression, and air pollution–also significantly influence the development of the disease, contributing to these interregional disparities in death rates.

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The role of cutaneous silent period in sensory ganglionopathies avaliaton: a case series and review

Hendrick Henrique Fernandes Gramasco[1], Maria Clara Foloni[1], Marcos Antônio de Lemos Júnior[1], Cristina Saade Jaques[1], Márcio Luiz Escórcio Bezerra[1], Karlo Faria Nunes[1]

[1]Universidade Federal de São Paulo, Escola Paulista de Medicina, São Paulo SP, Brazil.

Correspondence: Hendrick Henrique Fernandes Gramasco; Email: gramasco.hendrick@gmail.com.

Background: Sensory ganglionopathies consist of conditions characterized by involvement of the dorsal root ganglion, leading to a pure sensory deficit syndrome, not length-dependent, with involvement of the face, trunk and upper limbs before involvement of the lower limbs, in general. Most ganglionopathies affect primarily the large fibers, leading to symptoms such as sensory ataxia. Less commonly, ganglionopathy can affect only the small sensory fibers, leading to burning pain, tingling, shock pain and dysautonomia. In general, routine neurophysiological assessment does not evaluate the small sensory fibers, requiring specific tests, such as the cutaneous silent period, to access them. **Objective:** We present a series of cases of sensory ganglionopathy at different stages of evolution, from initial to chronic conditions, followed in a referenced center, in which we perform a cutaneous silent period test protocol. Our objectives were to use this information to differentiate cases of ganglionopathy from cases of asymmetric peripheral polyneuropathy, evaluate the involvement of small sensory fibers and the evolution of this involvement according to the course of the ganglionopathy. **Methods:** We selected cases already monitored as ganglionopathy or included patients diagnosed by our group based on standard electroneuromyography to carry out a protocol to evaluate the cutaneous silent period and the autonomic nervous system using Ewing tests, in addition to a targeted questionnaire and clinical examination. **Results:** Until the moment this work was sent, it was possible to observe that there is preservation of the cutaneous silent period and Ewing tests in cases recently diagnosed as ganglionopathy, while in later cases, we observed changes in the tests. It is possible that there is an association between the underlying disease and the involvement of fine fibers and the autonomic nervous system. **Conclusion:** This pilot study allows us to establish a low-cost standardized evaluation protocol to evaluate the involvement of small sensory fibers in cases of ganglionopathy, to assist in the differential diagnosis and in an attempt to establish a prognostic assessment and relationship between the underlying cause and the type of compromised fiber, thus assisting in the etiological investigation.

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The role of etiologic subtypes in predicting functional impairment after stroke: a longitudinal study

Maria Vitória do Nascimento Silva[1], Maria Laura Lima da Silva[1], Jéssica dos Santos Coelho[1], Joan Lins Serafim[1], Thiago Bezerra dos Anjos[1], Ana Luiza Sampaio Lopes[1], Américo Danúzio Pereira de Oliveira[1], Rita Di Cassia Oliveira Angelo[2]

[1]Universidade de Pernambuco, Serra Talhada PE, Brazil.
[2]Universidade de Pernambuco, Garanhuns PE, Brazil.

Correspondence: Maria Vitória do Nascimento Silva; Email: vitoria.nsilva@upe.br.

Background: Stroke is the second leading cause of mortality and disability worldwide, arising from various causes and resulting in differing levels of neurological impairment and disability. Identifying the etiological subtypes of ischemic stroke (IS) is crucial for personalized patient management, guiding subtype-specific treatments, and delineating distinct disease patterns across populations. **Objective:** To investigate potential associations between etiological subtypes of IS and levels of functional impairment among patients treated at a public hospital in Serra Talhada, Pernambuco, Brazil. **Methods:** A longitudinal study was conducted involving IS patients treated at a public healthcare in Pernambuco between 2021 and 2023. The analysis employed the Trial of Org 10.172 in Acute Stroke Treatment (TOAST) classification and the Modified Barthel Index (MBI), administered at hospital discharge. **Results:** The sample included 157 patients, predominantly male (n=89; 56.7%), self-identified as Black (n=115; 73.3%), aged 61-80 years (n=86; 54.8%), and diagnosed with arterial hypertension (n=127; 80.9%). Etiological classification revealed a higher prevalence of stroke of undetermined cause (SUC) (n=94; 59.9%), followed by large artery atherosclerosis (LAA) (n=29; 18.5%) and small vessel occlusion (SVO) (n=20; 12.7%). The median MBI score at discharge was 24 [14-38], indicating severe functional dependence. Thus, the majority of patients exhibited some degree of functional dependence in their post-stroke activities of daily living (n=147; 93.6%). After excluding patients with undetermined etiology (i.e., SUC), a statistically significant association was observed between SVO and lower functional dependence (p=0.015). **Conclusion:** The findings of this study suggest that individuals over 60, males, Black individuals, and those diagnosed with arterial hypertension are at higher risk of experiencing a stroke. The high prevalence of SUC may be due to challenges in determining IS etiology within healthcare facilities. Patients in the SVO subgroup appeared to exhibit lower functional dependence compared to those with other etiologies. Identifying the underlying cause of IS seems to significantly influence functional dependence, highlighting the importance of etiological identification for prognosis and treatment. Furthermore, the prevalence of severe functional dependence underscores the need for improved access to recanalization therapies and post-stroke rehabilitation programs.

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The role of tonic motor activation (TOMAC) for restless legs syndrome: a systematic review and single-arm meta-analysis

Daniela Lopes Vieira, Ana Beatriz da Cruz Pessoa[1], Stephanie Senna Cotta Queiroz de Oliveira[1], Wanessa Almeida Luna[1], Daniele Fernandes Holanda[2]

[1]Universidade Federal de Minas Gerais
[2]Universidade Federal do Amazonas

Correspondence: Daniela Lopes Vieira; Email: daniela.medufmg@gmail.com.

Background: Restless legs syndrome (RLS) is a neurological condition characterized by a distressing need to move the legs, especially during periods of immobility, resulting in chronic conditions that worsen the patient's health and quality of life. Conventional pharmacological treatment for RLS has limitations, including the worsening of symptoms and adverse effects, with many patients developing refractory RLS as a common scenario. In light of this situation, Tonic Motor Activation Therapy (TOMAC) emerges as a non-invasive intervention alternative, consisting of peripheral nerve stimulation of the fibular nerve using a wearable device bilaterally on the lower legs. **Methods:** PubMed, Embase and Cochrane databases were searched to compose our systematic review and single-arm meta-analysis of randomized clinical trials (RCTs) to evaluate TOMAC's efficiency in treating RLS. We pooled outcomes for treatment responder rate and for change in the severity of symptoms. Subgroup analyses were performed for (1) medication-resistant and (2) medication-naïve participants. Mean differences (MD) were pooled with 95% confidence intervals (95%CIs) for continuous outcomes using a random-effects model. We used OpenMeta-Analyst for statistical analyses. **Results:** 5 RCTs and 270 patients were included in the meta-analysis. The mean time of follow-up was 5,2 weeks. Statistically, there was no significant improvement on psychopathology (3 RCTs; 55.8%; n=106; 95%CI 0.379 to 0.737; p=0.06; I2=64.49%), assessed with the Clinical Global Impressions-Improvement scale (CGI-I). TOMAC resulted no reduction for severity of RLS symptoms (5 RCTs; n=167; MD -7.034; 95%CI -8.110 to -5.957; p=0.610; I2=0%), evaluated with the International RLS Rating Scale. Subgroup analysis indicated that medication-resistant (3 RCTs; n=102; MD -7.351; 95%CI -8.613 to -6.088; p=0.880 I2=0%) and medication-naïve participants (3 RCTs; n=49; MD -7.177; 95%CI -10.136 to -4.217; p=0.284 I2=20.5%) both exhibited similarly responses. **Conclusion:** This is a single-arm meta-analysis due to the scarce scientific production in the field. Despite no statistically observed benefit for RLS psychopathology, TOMAC potential for medication-refractory RLS treatment should be measured by further studies with larger numbers of participants, better study design and longer follow-ups.

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The spectrum of IL6 signaling-related genetic variants in a cohort of AQP4+ and AQP4- NMOSD: towards for personalized anti-IL6R therapy in NMOSD

Elielson Veloso da Silva[1], Renan Amphilophio Fernandes[1,2], Andreza Lemos Salvio[1], Jessica Raposo Vedovi[1], Larissa Araujo Duarte[1,2], Helena França Alcaraz Ferreira[1,2], Anna Beatriz Scherer Vanzan[1], Elisa Gouvea Gutman[1,2], Valéria Coelho Santa Rita Pereira[2], Soniza Vieira Alves-Leon[1,2]

[1]Universidade Federal do Estado do Rio de Janeiro, Instituto Biomédico, Laboratório de Neurociências Translacional, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Rio de Janeiro, Hospital Universitário Clementino Fraga Filho, Centro de Referência em Doenças Inflamatórias Desmielinizantes, Rio de Janeiro RJ, Brazil.

Correspondence: Elielson Veloso da Silva; Email: elielsonveloso@hotmail.com.

Background: Interleukin-6 (IL-6) plays a pivotal role in the pathogenesis of NMOSD, prompting the development of anti-IL-6R biologics such as tocilizumab and satralizumab as treatment options. However, treatment response variability between AQP4+ and AQP4- patients underscores the necessity of investigating genetic factors influencing therapeutic efficacy. Currently, the mechanisms behind treatment response differences in these serostatus groups are still poorly understood. Exome sequencing in polygenic and multifactorial disorders like NMOSD offers a powerful tool to identify biological pathways contributing not only to our understanding of disease mechanisms but also to the identification of genetic variants associated with treatment failure or suboptimal response. **Objective:** Our aim was to identify genetic variants in IL-6 signaling-related genes in a cohort of 41 NMOSD patients, stratified by AQP4 serostatus, and to uncover a novel spectrum of genetic variants that warrant further investigation for predicting treatment response in clinical trials of anti-IL6R biologics for NMOSD. **Methods:** For that, we analyzed exome sequencing data from 41 NMOSD patients (27 AQP4+ and 14 AQP4-) to identify genetic variants within genes associated with IL-6 signaling (IL6R, FCGRT, ADAM10, and ADAM17). Fisher's exact test assessed the significance of differences in variant frequency between the two groups. Expression Quantitative Trait Loci (eQTL) data for the most significant variants were examined using the FIVEx and GTEx databases and relevant scientific literature. **Results:** The variants rs3810194 and rs2878342 (FCGRT), rs7179724 (ADAM10), and rs2228145 (IL6R) showed the most significant differences in frequency between the groups. Notably, variants rs3810194 and rs2878342 within the FCGRT gene, encoding the IgG transport receptor FcRn, demonstrated statistically significant differences (p=0.0303) with higher frequency in the AQP4- group. Furthermore, eQTL databases suggest these variants may decrease FCGRT gene expression. Since FcRn plays a crucial role in satralizumab recycling, reduced expression of FCGRT could potentially impact its efficacy in AQP4- NMOSD. **Conclusion:** This study identified genetic variants in IL-6 signaling-related genes that may contribute to the variability in treatment response among AQP4+ and AQP4- NMOSD patients. Notably, variants rs3810194 and rs2878342, potentially associated with reduced FCGRT gene expression and observed at higher frequencies in AQP4- patients, suggest a potential mechanism influencing the efficacy of satralizumab. It's important to note that variants rs2228145 in IL6R and rs7179724 in ADAM10, although not statistically significant in our cohort, warrant further investigation in cohorts treated with anti-IL6R biologics or larger NMOSD cohorts. These findings emphasize the need for genetic profiling in clinical trials to optimize the use of anti-IL6R therapies, paving the way for personalized medicine in NMOSD management.

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The storm within: unmasking thunderclap headache and reversible cerebral vasoconstriction (RCVS) in pheochromocytoma: a case report

Ingrid Lorena da Silva Gomes[1], Tainá Fátima Ramos Gonçalves[1], Giovanni Caniçali Simões[1], Guilherme Lago Miranda[1], Pedro Almeida Martins Pontes[1], Henrique de Moraes Bernal[1], Eduardo Takashi Takehara[1], Roberta Arb Saba Rodrigues Pinto[1], Marcele Schettini de Almeida[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, Hospital do Servidor Público Estadual, São Paulo SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Ingrid Lorena da Silva Gomes; Email: ilsgomes@yahoo.com.

Case presentation: A 43-year-old female teacher presented with a history of thunderclap headaches (TCH) starting 15 days prior to admission. She reported recurrent episodes occurring more than three times daily, accompanied by cardiac palpitations, high blood pressure episodes, and a sensation of impending head explosion. Symptoms typically resolved spontaneously within 5 minutes, yet residual headache often persisted. Eleven days after the initial TCH, the patient developed sudden blurred vision and complete disproportionate left hemiparesis, prompting emergency admission to a secondary unit under suspicion of stroke. After a normal brain CT scan, she was discharged within 24 hours due to the complete resolution of symptoms. Due to recurrent headache episodes, she visited several emergency units before presenting to our service on the 16th day from the initial symptom onset. She denied using any vasoconstrictors or illicit drugs. Upon thorough evaluation, we found that the patient had been experiencing recurrent acute symptoms of sweating, high blood pressure and fainting lasting less than one minute, typically occurring after large meals several times daily for at least 3 years. Despite initiating antihypertensive medication prescribed by a general practitioner, her symptoms did not improve. The neurological examination was unremarkable, prompting further investigation. Imaging included CT angiography of the brain and cervical arteries, followed by MRI of the brain and cerebral angiography, revealing multiple segmental stenoses in the bilateral anterior cerebral arteries and branches of the middle cerebral arteries. After hypothesizing TCH secondary to RCVS with no identifiable trigger, we explored the possibility of an adrenergic crisis due to pheochromocytoma based on the patient's prior symptoms. Abdominal MRI revealed two exophytic nodular lesions in the left adrenal gland and kidney, with elevated urinary catecholamine levels. Symptomatic treatment for RCVS and preoperative alpha-blocker therapy to prevent an adrenergic crisis were initiated. After resolution of headache symptoms, the patient was discharged. Three months later, scheduled left adrenalectomy and nephrectomy confirmed histological and immunohistochemical findings consistent with pheochromocytoma. **Discussion:** Various factors contribute to the development of RCVS, and its management depends on addressing the underlying cause. Although rare, catecholamine-secreting tumors are significant contributors to RCVS. Accurate history-taking and appropriate diagnostic tools are crucial for reaching a definitive diagnosis. **Final comments:** Pheochromocytoma represents a significant yet frequently overlooked cause of RCVS. A multidisciplinary approach is crucial for effective patient management. Clinicians must maintain a high index of suspicion for this diagnosis, particularly in patients experiencing recurrent episodes of thunderclap headache without an identifiable trigger.

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The use of anti-Tau therapy in patients with Alzheimer: a systematic review

Raquel de Sousa Lima[1], Maria Clara de Sousa Moraes[1], Ingred Lauane Balbino Alves[2], Myrella Evelyn Nunes Turbano[1]

[1]Instituto de Educação do Vale da Parnaíba, Parnaíba PI, Brazil.
[2]Universidade do Estado do Rio Grande do Norte, Mossoró RN, Brazil.

Correspondence: Raquel de Sousa Lima; Email: rl1918042020@gmail.com.

Background: Alzheimer's disease (AD) is a neurodegenerative condition. The accumulation of the Tau protein in this disease accompanies cognitive decline and plays a subsequent role in its progression. Therefore, therapies such as anti-Tau monoclonal antibodies aimed at slowing or stopping this accumulation are of utmost importance. However, there are persistent controversies regarding the efficacy of these drugs in treating the pathology. **Objective:** To analyze the efficacy of antibody therapy targeting the Tau protein AD. **Methods:** This scientific production is a systematic review based on randomized studies of an adult and elderly population with AD and high Tau load, who used anti-Tau monoclonal therapy and showed improvement. Furthermore, the databases used were: PubMed, Cochrane Library, and Scopus. The research was conducted based on the PRISMA strategy

(Preferred Reporting Items for Systematic Reviews and Meta-Analyses). The inclusion criteria for the analyzed studies were: randomized studies, without temporal restriction, focused on the topic. **Results:** Based on the strategies and criteria, 5 articles were selected for data collection. The use of anti-Tau monoclonal antibodies, such as: SEMORINEMAB, GOSURANEMAB, TILAVONEMAB, and ABBV-8E12, proved to be safe and well-tolerated by patients who used them, capable of reducing the ADAS-Cog11 score (11-item version of the Alzheimer's Disease Assessment Scale - Cognitive Subscale) compared to the placebo group. Additionally, it was noted that the Tau protein load is reduced in cerebrospinal fluid when compared to baseline value when exposed to antibodies at low doses. **Conclusion:** Therefore, it was evidenced that anti-Tau therapy with monoclonal antibodies in patients with Alzheimer's associated with high Tau protein load caused significant reductions in non-bound N-terminal Tau levels in cerebrospinal fluid compared to placebo. This encourages the continuation of studies to ensure greater treatment effectiveness.

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The use of decompressive craniectomy in young patients with ischemic stroke: a case report

Yuri Victor Barbosa Silva[1], Carolina Diniz Furtado[1], José Nilo Ribeiro Neto[1], Márcio Luís Soares Ferreira[1], Roseliny de Moraes Martins Batista[1], Sanzylly Soares França Ferreira[2], João Guilherme de Brito Rodrigues[1], Lael Mendonça Moraes[1]

[1]Centro Universitário UNDB, São Luís MA, Brazil.
[2]Universidade Estadual do Maranhão, São Luís MA, Brazil.

Correspondence: Yuri Victor Barbosa Silva; Email: yurifisiocenter@hotmail.com.

Case presentation: Patient, 16 years old, female, was admitted to the regional hospital in the city of Bacabal-MA, presenting with syncope 6 hours after admission. Patient denies Sys-temic Arterial Hypertension (SAH), Diabetes Mellitus (DM) and other pathologies. Laboratory tests, electrocardiogram and head tomography were requested. Skull to-mography with signs of hypodensity in the territory of the middle cerebral artery on the left with slight deviation of the midline structures. Laboratory tests and electrocardio-gram were unremarkable. The patient subsequently underwent decompressive craniectomy with bone flap stor-age in the abdomen. After surgery, the pupils were isochoric and photoreactive, and the patient re-mained sedated in an intensive care unit bed. Sedation was weaned after 72 hours. The cranioplasty was scheduled within six months, with no complications oc-curring during the procedure. **Discussion:** Ischemic stroke (IS) is defined as partial or total occlusion of a cerebral blood vessel by an atherosclerotic plaque or clot (David et al, 2024). IS has a high mortality rate and, above all, functional disability, causing a major impact on the lives of indi-viduals and their families. The prevalence rate is higher than that of hemorrhagic stroke, averaging around 80% to 85% of cases (Feitosa et al, 2022). Ischemic brain injury results from a cascade of events, from energy depletion to cell death, some intermediate factors include excess of extracellular excitatory amino acids, formation of free radicals and inflammatory processes (Leite, 2020). Rios et al (2021) mentions that manifestations of ischemia are caused by biochemical sequenc-es that lead to the destruction of the brain parenchyma and follow three main path-ways: in the neuron, in the glia and in the microcirculation, called ischemic cascades.Several treatment approaches are carried out for patients with ischemic stroke, both conservative and surgical, based on the level of consciousness and imaging tests performed every 48 hours for monitoring in cases of clinical signs of neurological disease (Neufeuld et al, 2020). According to Wang et al (2021), decompressive craniectomy is the removal of a segment of the skull large enough to minimize refractory intracranial pressure and maintain brain compliance with the aim of preventing injury to other structures and neurological deterioration. According to Leite (2020), the purpose of the surgery is to promote decompression above the swollen cerebellum to open space. **Final comments:** As the updated studies state, it is notable that the causes of stroke must be in-vestigated mainly and especially in cases of young patients. Decompressive craniec-tomy is an appropriate surgical intervention in cases of extensive mass effect, since conservative treatment is not very effective or when the subject presents considerable changes in consciousness levels, thus obtaining a greater probability of survival.

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The use of fibroblasts culture to evaluate treatment for mitochondrial diseases

Celia Harumi Tengan[1], Paloma Ramos de Macedo[1], Aryane Coutinho[1], Vinicius Gomes da Silva[1], Jomênica de Bortoli Livramento[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Celia Harumi Tengan; Email: chtengan@unifesp.br.

Background: Mitochondrial diseases are characterized by oxidative phosphorylation (OXPHOS) deficiencies and are caused by mutations in the nuclear or mitochondrial DNA. Despite significant advances in discovering the genetic causes, no proven effective treatments are available for patients with these diseases. The large number of genes associated with mitochondrial diseases, the rarity and the highly variable phenotypes hinder the study of potential therapies in clinical trials. **Objective:** To evaluate the effect of potential therapies for mitochondrial diseases using fibroblast culture. **Methods:** Cultured fibroblasts were obtained from skin biopsies of patients (n=4) with mitochondrial diseases. Fibroblasts with no abnormalities in OXPHOS were used as controls (n=2). We evaluated the effects of two antioxidants (N-acetyl cysteine, NAC, and mitoquinone, MitoQ) and the precursor of nitric oxide synthesis, L-Arginine (L-ARG). Treatments were performed for 12 days with the following concentrations: 1 mM and 5 mM NAC; 10 nM and 100 nM MitoQ; and 1 mM L-ARG. Fibroblasts were cultured in two different media: (1) with high concentrations of glucose and (2) in a restrictive condition where glucose was replaced by galactose, not favoring the growth of mitochondrial-deficient cells. The effect of treatment was evaluated by measuring cell growth (or cell proliferation) in both conditions and at different time points. We expected that improving mitochondrial function would result in better cell proliferation in the restrictive medium. **Results:** Controls exhibited an exponential growth in cell number in both glucose and restrictive media, whereas patient cultures showed a good exponential growth only in the glucose medium, except one culture (P-1), which also had good cell proliferation in the restrictive medium. Treatment with MitoQ, NAC and L-ARG did not affect control samples. MitoQ (10 nM) led to a higher number of cells in P-2 fibroblasts at day 12 (4188±277 cells vs 6125±554 cells, p=0.02), with this tendency starting at day 8. Treatment with 1 mM L-ARG improved cellular growth in the restrictive medium in P-4, with a statistically significant difference in cell number starting from day 8. The increases in cell number were 151% (day 8), 307% (day 10) and 790% (day 12) compared to untreated cells. This increment reached 10625±946 cells with L-ARG versus 1344±157 cells with no treatment at day 12 (p=0.03). The other treatments with 100 nM MitoQ and NAC (1 mM and 5 mM) did not modify cellular growth in a restrictive medium. **Conclusion:** These preliminary results demonstrate that this assay can detect improvements in cell growth with MitoQ and L-ARG treatment. Variable results may indicate individual variability, highlighting the importance of personalized study on drug responses. Further studies are still necessary to confirm the beneficial effects of these treatments.

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The use of neural networks to predict stroke diagnosis: a systematic review

Vanessa de Oliveira e Silva[1], Luciana Cartaxo[1], João Victor Loiola[1], Kael Costa Santana[1], João Wilson de Luna Freire Neto[1], Matheus Oliveira Lira[1], Marcela Vasconcelos Montenegro[2], Maria Luiza Vasconcelos Montenegro[2], Maria Clara Formiga de Mello[1], Maria das Graças Loureiro das Chagas Campêlo[1]

[1]Universidade Federal de Campina Grande, Campina Grande PB, Brazil.
[2]Universidade de Pernambuco, Recife PE, Brazil.

Correspondence: Luciana Cartaxo; Email: lucianaestudos1000@gmail.com.

Background: Stroke is a syndrome of acute neurological deficit attributed to vascular damage to the Nervous System (NS). The Artificial Intelligence (AI) techniques in Medicine – such as Artificial Neural Networks algorithms (ANNs) – have helped in making clinical decisions for this condition. **Objective:** To evaluate how Artificial neural networks are being used to predict the diagnosis of stroke. **Methods:** This is a systematic review of articles indexed in databases PubMed, BVS, SciELO, Cochrane and SpringerLink, between January and February of 2022. The inclusion criteria and filters for this study were: articles related to the topic, randomized studies, cohort and clinical trials, studies with humans, carried out in the last 5 years, only in Portuguese, English and Spanish and with the full text available free of charge. The parameters of exclusion were: duplicated articles, off-topic studies, review articles and papers that did not meet all inclusion criteria. The Kappa coefficient was calculated and it was obtained a value equal to 0.795 (substantial agreement). Furthermore, the review has a protocol registered on the International Prospective Register of Systematic Reviews (PROSPERO) website with registration number CRD42021290522. **Results:** ANNs are being used mainly to evaluate areas of ischemic and hemorrhagic injury by segmentation methods and the most used exams for modeling the programs have been Magnetic Resonance Imaging (MRI) and Computed Tomography (CT). In addition to CT and MRI, magnetic resonance angiography and CT angiography are also being used for algorithm modeling and they are useful because they have greater sensitivity for detecting infarcts. **Conclusion:** Segmentation and classification algorithms applied to ANNs are part of personalized medicine and serve as a basis for doctors in clinical practice.

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The use of physical exercise as a treatment for Alzheimer's disease: a systematic review

Júlia de Castro Couto Santos[1], Helena Oliveira Farage[1], Letícia Dalton Moreira dos Santos[1], Lucas Vieira Nogueira[1], Raquel Toledo Martins de Almeida[1], Roberta de Oliveira Lima Duarte[1], Sofia Oliveira Duarte[1]

[1]Faculdade de Ciências Médicas e da Saúde de Juiz de Fora, Juiz de Fora MG, Brazil.

Correspondence: Júlia de Castro Couto Santos; Email: juliacastroccs@yahoo.com.

Background: Alzheimer's dementia (AD) is a hypocholinergic disease, in which beta amyloid deposition and tau protein hyperphosphorylation occur, resulting in the loss of neurons and, consequently, synapses in brain regions, compromising cognitive function and the consolidation of new memories. **Objective:** To analyze the influence of physical exercise in the treatment of patients diagnosed with AD. **Methods:** Controlled and randomized clinical trials originally published in English, from the last ten

years, on humans were analyzed, using the National Library of Medicine (MedLine) database as a reference. The search for descriptors and terms used was carried out by consulting the Medical Subject Headings (MeSH), through the U.S. National Library of Medicine (NLM) portal and the descriptors used were: Alzheimer Disease, Physical Exercises, Treatment. Studies with a sample of individuals over 50 years of age and a clinical diagnosis of probable mild to moderate AD were included. Articles that evaluated individuals without evidence of cognitive impairment and studies lasting less than 16 weeks were excluded. The PRISMA scale was used to systematize the reporting of this review. **Results:** Initially, 7 studies were found and, after selecting inclusion and exclusion criteria, only 4 articles were part of the scope and final analysis. When analyzing cerebral blood flow, no benefits were observed over 16 weeks of moderate to high intensity aerobic exercise, and a limitation noted in this study was the duration of the intervention. However, in a study lasting 6 months, the angiogenic process, in response to the increased energy demand promoted by physical exercise, during this period of time contributed to significant functional changes and improvements in cerebral blood flow, which are factors affected by AD. Furthermore, it is known that, through biomarker analysis, physical exercise contributes to increased levels of neuroprotective proteins and can positively affect cognition in individuals with AD. Furthermore, training in activities of daily living is a viable and effective intervention in improving other issues affected by the disease, such as physical resistance and depressed mood in nursing home residents. **Conclusion:** Physical exercise proved to be significantly effective, especially in periods longer than 16 weeks in the treatment of patients with AD.

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The use of Wobble Chair to assess trunk postural control in individuals with Parkinson's disease: a reliability study

Hayslenne Andressa Gonçalves de Oliveira Araújo[1], Patrícia Gonçalves Broto[1], Luzia Aparecida Trinca[2], Alessandra Cattaneo Estrada Melanda[1], Marcelle Brandão Terra[1], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.
[2]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Patrícia Gonçalves Broto da Silva; Email: pbroto@hotmail.com.

Background: Postural instability is a relevant symptom that can occur from the early stages of Parkinson's disease (PD), resulting in loss of functionality and independence. Trunk control represents a large portion of body mass and plays an essential role in maintaining postural control. However, there is still a lack of specific instruments to assess trunk control in PD. **Methods:** A cross-sectional study with 44 individuals with mild to moderate idiopathic PD, according to UK Brain Bank criteria, recruited from an outpatient of specialized physiotherapy (GPFIN), with no cognitive impairment, and walking independently. Trunk control was assessed using the Wobble Chair, (which is an instrument based on force platform and composed by a pivot and springs structure, aiming to provide instability during seated evaluation). Participants were assessed under three conditions: 1) eyes open (EO); 2) eyes closed (EC); and 3) no feet support (NS). The outcome variables included: center of pressure (COP) area (cm^2), AP/ML amplitude (cm), AP/ML mean frequency (Hz) and AP/ML velocity (cm/s). The test and retest were performed within a 7-day interval and compared using a paired t-test. The instrument reliability was quantified using an intraclass correlation coefficient (ICC), 95% confidence intervals (CI), and Bland Altman plot. Statistical Package for the Social Sciences 29 (SPSS 29) and GraphPad Prism 10.0 were used for the analysis, and we considered a statistical significance of $p< 0.05$. CAAE: 31775420.8.0000.0108. **Results:** The ICC ranged from 0.51 to 0.86 in the different analyzed tasks, and overall, the EO, EC, and NS conditions showed similar results, since the CI measurements overlapped. The EO condition showed excellent reliability for AP and ML velocity (ICC=0.82, for both) and ML mean frequency (ICC=0.75), as well as good reliability for the other analyzed parameters (area, amplitudes, and AP mean frequency). The EC condition showed excellent reliability for AP and ML velocities, and ML mean frequency (ICC=0.75, ICC=0.82, and ICC=0.75, respectively) and good reliability for COP area, amplitudes, and AP mean frequency. Finally, the NS condition showed excellent reliability for the COP area (ICC=0.75), AP velocity (ICC=0.82), AP and ML mean frequencies (ICC=0.75 and ICC=0.79, respectively), as well as good reliability for amplitudes and ML velocity. Considering the variables analyzed, velocity showed the most consistent reliability, with ICCs ranging from 0.74 to 0.86. **Conclusion:** The Wobble Chair is a reliable and accurate instrument to assess trunk postural control in individuals with PD, especially considering the velocity and EO situations (with and without feet support). Further studies are suggested to explore different conditions, populations, and assessment protocols.

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The utilization of telemedicine and computed tomography in the management of stroke in Brazil: a focus on emergency referral in 14 secondary care hospitals

Guilherme Nobre Nogueira[1], Rafaela Fernandes Gonçalves[2], Adrielle Holler Pykocz[2], Guilherme Batista do Nascimento[3], Gustavo Rassier Isolan[2]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Faculdade Evangélica Mackenzie do Paraná, Curitiba PR, Brazil.
[3]Centro Universitário de Adamantina, Adamantina SP, Brazil.

Correspondence: Guilherme Nobre Nogueira; Email: guiermenobre@gmail.com.

Background: The recognition of stroke (AVC) and its symptoms is a topic that significantly impacts the treatment time for suspected strokes, mainly due to delays in reaching medical care. In this context, telemedicine provides specialized support promptly for neurological emergencies. **Objective:** To demonstrate the experience of using telemedicine in managing cases of suspected stroke, investigating the average time of care and the type of stroke. **Methods:** This study is a descriptive, cross-sectional, and retrospective study involving 587 patients from 14 hospitals in the states of Rio Grande do Sul and São Paulo, from April 2019 to October 2022. All patients signed a consent form for the research, respecting the general data protection law. **Results:**A total of 587 patients were evaluated. The age ranged from 0 to 98 years, with an average of 63.1 (standard deviation - SD ±18.7) and a median of 65. There was a slight majority of men, accounting for 51.2%. The distribution of patients by age group showed that 10.56% of individuals were between 1 and 40 years old, 28.28% were in the age group of 41 to 60 years, 44.12% were between 61 and 80 years, and 17.04% were between 81 and 100 years old. Regarding medical history, headache was reported by 46.68% of patients, progressive neurological symptoms were mentioned by 23.68%, and sudden neurological symptoms were recorded in 29.64% of the analyzed individuals. Out of the 587 patients, 40.37% had hemorrhagic stroke, 23.68% had old ischemic stroke, 29.64% had new ischemic stroke, and 6.30% had Aneurysmal Subarachnoid Hemorrhage. In terms of response time, it was observed that all patients had telemedicine initiated within 30 minutes. Considering the start of care, CT evaluation, treatment recommendation, and referral, the average time was 93.4 minutes (SD = 120.4). There was a significant association between tomographic changes and emergency referral. The chi-square test results showed a value of 130.05 with 3 degrees of freedom and a P-value < 0.001. These results suggest a statistically significant association between the variables under study. The effect size (φ) was 0.47 between the variables, indicating that the emergency referral variable has a strong and significant influence on the tomographic change variable. **Conclusion:** Since emergency referral was recommended after telemedicine consultation, we can infer that it influenced the management of these conditions. Telemedicine is relevant in caring for patients with suspected stroke, particularly due to its prompt service in a country with continental dimensions like Brazil. These results provide valuable information about the clinical and tomographic profile of patients, emphasizing the importance of effective strategies for stroke through real-time specialist opinions.

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The weight of genotype on clinical presentation of COQ7 related hereditary motor neuropathy: case series and literature review

Laura Alonso Matheus Montouro[1], Amanda Selvátici dos Santos Dias[1], Maria da Penha Ananias Morita[1], Érica Nogueira Coelho[1], João Aris Kouyoumdjian[1], Carla Renata Graça[1], Fábio Nazaré Oliveira[1], Charles Lourenço Marques[1], Eduardo de Paula Estephan[1,2]

[1]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, São Paulo SP, Brazil.

Correspondence: Laura Alonso Matheus Montouro; Email: lauramontouro@hotmail.com.

Background: The mitochondrial enzyme 5-demethoxyubiquinone hydroxylase, encoded by COQ7, is involved in CoQ10 biosynthesis. Mutations in the COQ7 gene result in primary CoQ10 deficiency-8. First case was published in 2015, presenting as a multisystemic form (MF). Subsequently, other phenotypes described: spastic paraparesis (SP), polyneuropathy, and motor neuron disease and distal hereditary motor neuropathy (dHMN). Since 2022, dHMN related to p.1met? variant, mostly in Brazil. A founder effect evidence of this variant in Brazil was published recently. **Objective:** To report five Brazilian patients from three unrelated families with p.1met? variant and dHMN. Also perform narrative review of COQ7 disorders and analyze possible correlations between the variants and phenotypes/clinical sign and also between p.1met? and other dHMN from Brazil. Reinforce the notion of founder effect and the phenotype probably related to this mutation. **Methods:** Descriptive study of our cases. Our case series clinical features were compared with all COQ7-patients reported and Brazilian dHMN cohorts in literature so far. Categorical variables were expressed as absolute frequencies and compared using Fisher's test (two-tailed) and Odds Ratio (OR), with confidence interval 95% (CI). **Results:** Patient 1 presented dHMN plus with cerebellar (CS) and pyramidal syndrome (PS). Patient 2 had pure dHMN. Patients 3 and 4, siblings, both had a dHMN plus (PS) and patient 5 had dHMN plus with cognitive impairment. Our search identified 20 studies with 47 patients with COQ7 related disorders, with origin from 10 countries. Comparing the phenotypes among the presence of variants, the p.1met? was significantly more frequent in patients with dHMN (21 out 30, $p<0.001$). Brazilian origin was also significantly related to p.1met? variant compared to other regions ($p<0.001$, OR: 10.5, CI: 2.6-40.8). Brazilian origin is also related to the dHMN phenotype ($p=0.009$, OR: 8.6, CI: 1.6-45.8). All patients with CS were from Brazil, and this relation was also statistically significant ($p=0.0063$). Compared to 170 cases of dHMN from Brazil, PS ($p<0.001$; OR: 7,2273, CI: 2.42-21.5) and CS ($p<0.001$) associated with dHMN had a strong relation with p.1met?. SPG-COQ7 is related

to p.Leu111Pro with statistical significance (p=0.00181, OR:34, CI:3.3-427.9). This variant had a strong association with Iran (p<0.001). The p.Arg54Gln was statistically associated with SPG (p:0.029, OR:11.1 95%, CI: 1.4-83.5). **Conclusion:** Specific variants in COQ7 can be associated with specific phenotypes. The variant p.1met? has a strong association with dHMN, Brazil and cerebellar syndrome. Among Brazilian patients with dHMN with CS and/or PS suggests a higher chance of COQ7 disease. The finding of CS only in Brazilian patients raises the hypothesis of environmental influence in disease expression.

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Therapeutic effects of human dental pulp stem cell derived exosomes in transgenic mice SOD1G93A

João Pedro Nunes Gonçalves[1], Rodrigo Pinheiro Araldi[2], Cristiane Valverde Wenceslau[3], Irina Kerkis[4], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]BioDecision Analytics Ltda., São Paulo SP, Brazil.
[3] Cellavita Pesquisas Científicas Ltda., Valinhos SP, Brazil.
[4]Instituto Butantã, São Paulo SP, Brazil.

Correspondence: João Pedro Nunes Gonçalves; Email: j227991@dac.unicamp.br.

Background: Amyotrophic lateral sclerosis (ALS) is a devastating condition for which treatment still represents an unmet need. Exosomes derived from human dental pulp stem cell (hDPSCs) secrete neurotrophic factors and have shown positive therapeutic results in some neurodegenerative disease models, but not yet properly explored in ALS. **Objective:** To evaluate the potential therapeutic effect of hDPSC-derived exosomes in the ALS SOD1G93A transgenic mice model. **Methods:** Seventy-day-old transgenic mice were treated intravenously either with exosomes (n=5) or saline (n=5) for 5 weeks and were sacrificed at the age of 15 weeks. A 3rd third group of wild type mice was also used for comparison. We then proceeded quantified motor neurons in the ventral horns of lumbar spinal cord using Nissl staining. Between-group comparisons were accomplished with ANOVA followed by post-hoc Tukey test. **Results:** At 15 weeks, neuronal survival was significantly different across experimental groups. Neuronal loss was significantly higher in non-treated SOD1 G93A mice compared to the other two groups (p<0.0001, CI:95% for both groups). In contrast, neuronal survivability was similar in treated and wild type animals (p=0.025, CI:95%). **Conclusion:** While still in the early stages, our findings show that spinal cord motor neuron survival in this ALS mouse model is preserved by repeated IV injections of hDPSC-derived exosomes, though they still vary from wild-type animals. Further studies are needed to understand the mechanisms underlying such therapeutic effects.

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Therapeutic plasma exchange in steroid-refractory attacks of myelin oligodendrocyte glycoprotein antibody-associated disease: a systematic review and meta-analysis

Roger Santana de Araujo[1], Thiago Trajano da Silva[1], Guilherme Diogo Silva[1], Samira Luisa dos Apóstolos Pereira[1], Dagoberto Callegaro[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Roger Santana de Araujo; Email: rogersantana.araujo@gmail.com.

Background: Therapeutic plasma exchange (TPE) has been extensively studied for its role in disability recovery during attacks of demyelinating diseases like Multiple Sclerosis and Neuromyelitis Optica Spectrum Disorder. However, its efficacy in Myelin Oligodendrocyte Glycoprotein Antibody-Associated Disease (MOGAD) remains uncertain. While MOGAD attacks typically respond well to glucocorticoids, the effectiveness of TPE in steroid-refractory cases is still debated and under investigation. **Objective:** To evaluate the response to TPE in steroid-refractory MOGAD attacks. **Methods:** We conducted a systematic review and meta-analysis, searching PubMed/MEDLINE, Embase, and Scopus for manuscripts published up to March 21, 2024, using synonyms for "plasma exchange" and "MOGAD." We included cohorts, case reports, and case series of patients with MOGAD attacks who underwent TPE. Two evaluators selected studies and extracted data, resolving disagreements by consensus. We collected information on age, sex, and core clinical demyelinating events. The primary outcome was the clinical response to TPE in MOGAD attacks at the last follow-up, categorized as complete response, partial response, or no response. We performed a meta-proportion analysis using R software with a random-effects model. **Results:** We included 40 studies, comprising 36 case reports and 4 cohorts, totaling 278 attacks (mean age 34 years, 46% male). Core clinical demyelinating events included optic neuritis (49%), transverse myelitis (28%), acute demyelinating encephalomyelitis (3%), cerebral with focal deficits (9%), brainstem or cerebellar deficits (7%), and cortical encephalitis (4%). Complete recovery was observed in 36% (95% CI 26%; 48%) of cases, while 13% (95% CI 6%; 25%) showed no response. **Conclusion:** TPE may improve neurological recovery in steroid-refractory MOGAD attacks. This study is ongoing.

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Therapeutic window for thrombolysis at a stroke center in Northeast Brazil: current overview and future perspectives

Raphael Pinheiro Camurugy da Hora[1], Ana Dolores Firmino Santos do Nascimento[1], Manuela Magalhães Dardenne Tenório[1]

[1]Secretaria de Saúde de Pernambuco, Hospital da Restauração, Recife PE, Brazil.

Correspondence: Raphael Pinheiro Camurugy da Hora; Email: raphaeldahora@hotmail.com.

Background: Stroke is a complex, multifactorial disease with a high social impact. Its most prevalent subtype, ischemic stroke, has undergone a radical therapeutic revolution in the last thirty years with the advent of intravenous thrombolysis and thrombectomy. Even so, the outcome of patients with stroke in Brazil is still suboptimal, with a small percentage of patients receiving thrombolytic treatment. **Objective:** To evaluate the conversion rate to thrombolysis and the reasons why patients admitted to Hospital da Restauração (HR) in the therapeutic window for intravenous thrombolysis had the procedure contraindicated. **Methods:** This study is classified as observational, cross-sectional, retrospective and analytical. Data were collected through review of medical records, tabulated and analyzed in IBM SPSS Statistics®. **Results:** 3,140 medical records were reviewed over a period of nine months in the neurological emergency department of HR. Of these, 14.1% (445) were flagged as possible stroke within a therapeutic window of up to 270 minutes. The conversion rate to intravenous thrombolysis was 42.6% (190) and considering only patients diagnosed with ischemic stroke (352), this percentage was 53.98% (190). When analyzing the prevalence of contraindications to the use of thrombolytics, we observed an ASPECTS scale <7 (13.5%), AVCH (11.9%), AIT (6.3%), more than one reason (5.8 %), minimal deficit or NIHSS <5 (4%), gastrointestinal or genitourinary hemorrhage <21 days (2.2%), use of anticoagulant drugs (1.3%) and others with less statistical significance. Secondly, 57 ASPECTS contraindications were reviewed by the neuroradiology team, revealing a 35.7% agreement rate. **Conclusion:** Analysis of the results revealed a very satisfactory therapeutic index within our reality. Furthermore, the implementation of vessel studies and extended protocols across all shifts could increase this rate, as well as the active presence of radiology in the acute ischemic stroke protocol.

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Theta burst stimulation and spasticity in multiple sclerosis: a meta-analysis of randomized controlled trials

Mariana Rachas Reis[1], Isabella Christina Amaral de Lara[2], Amanda Gabriela Freitas Rodrigues[3], Matheus Ricardo Malveira Camacho[4], Bruno Lopes Santos Lobato[3]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Universidade Cesumar, Maringá PR, Brazil.
[3]Universidade Federal do Pará, Belém PA, Brazil.
[4]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Mariana Rachas Reis; Email: marianarachas@gmail.com.

Background: Multiple sclerosis (MS) is an autoimmune disorder that affects the central nervous system, often showing suboptimal responses to traditional treatments. This has prompted significant interest in alternative symptom management approaches. Non-invasive brain stimulation (NIBS), particularly theta burst stimulation (TBS), has emerged as a promising method to mitigate the disease's morbidity. TBS delivers rapid impulses to enhance neural activity, fostering beneficial neuroplasticity. We conducted a meta-analysis comparing outcomes in patients receiving theta-burst stimulation versus placebo, aiming to evaluate its efficacy in managing MS symptoms. **Objective:** To assess the efficacy of theta-burst stimulation for the treatment of patients with Multiple Sclerosis spasticity. **Methods:** MEDLINE, Embase, and Cochrane databases were systematically searched for randomized placebo-controlled trials (RCT) that tested theta-burst stimulation in multiple sclerosis patients. Data were examined using the Mantel-Haenszel method and 95% confidence intervals (CIs). Heterogeneity was assessed using I^2 statistics. This meta-analysis was performed to evaluate measures of spasticity through the Modified Ashworth Scale (MAS). Statistical analysis was conducted using R, version 4.4.0. **Results:** The initial search identified 619 results. After removing duplicate records and screening the studies based on title and abstract, 412 studies were eligible for full-text review according to predefined criteria. Among these, three randomized placebo-controlled trials (RCT) were included, with a total number of 59 patients, of whom 32 (54.2%) received TBS and 27 (45.7%) received sham stimulation. Mean age ranged from 38.4 to 49.8 years, with a majority of female patients. Our pooled analysis showed that, compared with the placebo group, TBS was associated with a significant reduction in MAS score (Mean Difference -0.82; 95% CI -1.11-0.53; p<0.000001; I^2=0%), indicating an improvement in muscle spasticity. **Conclusion:** This systematic review and meta-analysis highlights the effectiveness of theta-burst stimulation in improving spasticity among patients with MS, as evidenced by significant improvement on the Modified Ashworth Scale.

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Three-territory sign in acute ischemic stroke as a manifestation of ovarian tumor: a case presentation

Ana Luísa Castelo Branco Gomes[1], Davi Telécio Firmino[2], Gilmar Leite Pessoa Filho[1], Luciano Gonçalves do Nascimento Júnior[1], Felipe Nunes de Araújo[1], Diego Almeida de Souza[1], Rafael de Souza Andrade[1], Francisco Anderson de Sá Carvalho[1], Paulo Antonio Farias Lucena[1], Juliana Magalhães Leite[1]

[1]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[2]Universidade Federal da Paraíba, João Pessoa PB, Brazil.

Correspondence: Ana Luísa Castelo Branco Gomes; Email: analuisa_gomes@hotmail.com.

Case presentation: I.F., 75 years old, female, smoker, hypertensive, history of acute vertigo for 2 months, associated with headache and disorientation. Three days after admission, she presented a new acute event of drowsiness, emetic episodes and worsening vertigo. Neurological examination revealed dysarthria, right facial palsy and right hemiparesis. During the investigation, brain MRI showed an area of diffusion restriction in the parietal lobes, right cerebellar hemisphere and right posterolateral region of the medulla oblongata, as well as late subacute intraparenchymal hematomas in the left temporal lobe. An etiological investigation was carried out with EKG, TTE, TEE and cervical and intracranial arterial AngioMRI without abnormalities. Laboratory investigation for systemic lupus erythematosus and antiphospholipid syndrome were negative. Once the possibility of stroke due to hypercoagulability was raised, neoplastic investigation was initiated. Pelvic MRI showed a nodule in the right ovary with hyposignal on T1 and T2 without gadolinium enhancement. The patient was referred for neoplastic investigation and anticoagulation was started. **Discussion:** Trousseau syndrome(TS) is a hypercoagulative state associated with cancer that leads to venous or arterial thromboembolic events. The three-territory sign is a radiographic marker of TS, which is described as an ischemic stroke involving three vascular territories, including involvement of the bilateral anterior and posterior circulations. The primary approach to treating TS is to eliminate, if possible, the causative tumor, as well as anticoagulation, with heparin being the treatment of choice. **Final comments:** In the present case, the lesions in three different vascular territories raise the possibility of a hypercoagulable state, after excluding possible causes of stroke, mainly cardioembolic causes. Therefore, this case report is relevant in the etiological study of patients with stroke, leading to early diagnosis of neoplastic diseases based on cerebrovascular conditions.

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Thrombectomy in SUS: a new perspective to the AIS treatments

Vítor de Oliveira Cei[1], Jéssica Helena Gomes Ferreira[1], Gabrielly Gomes de Oliveira[1], Leticia Fernandes de Freitas[1], Priscila Maria de Oliveira[1], João Gabriel Alves Coimbra Chaves[1], Caroline Chaves Bittencourt[1]

[1]Universidade Nove de Julho, Guarulhos SP, Brazil.

Correspondence: Leticia Fernandes de Freitas; Email: lecaff1984@gmail.com.

Background: Recently, the Brazilian Ministry of Health instituted Ordinance: GM/MS No. 1996 of 11/24/2023, regarding the implementation of Mechanical Thrombectomy (MT) in the SUS, guaranteeing this treatment option in applicable cases of Acute Ischemic Stroke (AIS) with a symptomatic signals between 8 and 24 hours. MT consists of removing the obstructive content that impedes blood flow to the brain, resulting in acute ischemia and neurological deficit. According to DATASUS, between 2022-2023, AIS caused 55 thousand deaths. Therefore, with this possibility, there are expectations of reducing the number of deaths and alleviating serious functional disabilities. **Objective:** To evaluate the effectiveness of treating AIS with mechanical thrombectomy and outline prospective parameters for Brazil with the implementation of GM/MS regulation nº1996 of 11/24/2023. **Methods:** Records were collected on the PubMed platform with the descriptors: Thrombectomy, Acute Ischemic Stroke. Randomized clinical trial studies were selected that highlight the advantages and disadvantages of thrombectomy. **Results:** Studies have shown a positive prognosis with MT. Sarraj (2023), in a randomized and controlled clinical trial on AIS covering the United States, Canada, Europe, Australia and New Zealand, compared 178 patients who underwent MT in 24 hours with 174 who received only medical care with local protocols, the median ischemic volumes 74ml vs 77ml, both with a median of 4 on the ASPECTS (Alberta Stroke Early Score) and a median of 19 on the NIHSS (National Institutes of Health Stroke Scale) score. In the MT group, 142 (79.8%) had successful reperfusion, with 20.3% showing functional independence at 90 days (Modified Rankin Scale [mRS] from 0 to 2) and 37.9% with independent ambulation at 90 days (mRS from 0 to 3). Furthermore, 11.5% showed early neurological improvement (decreasing 8 points in the NIHSS), however 24.7% showed worsening of the neurological condition related to a larger ischemic area when compared to the group without neurological worsening. The hospital death rate was 23.6%. In the medical care group at 90 days, the median MRS was 5; functional independence at 90 days 7% (mRS 0-2); independent ambulation at 90 days 18.7% (mRS 0-3); deaths at 90 days 41.5% and neurological worsening 15.5%. Comparatively, MT obtained better results compared to the group that received only clinical care. **Conclusion:** The regulation of GM/MS No. 1996/24/11/2023 to accomplish thrombectomy in the SUS indicate a promising future, given the high effectiveness of the procedure and a better prognosis in reperfusion and the patient's clinical condition 90 days after management, especially the decrease in the death rate. Therefore, the 12 institutions accredited for the procedure in Brazil can minimize national morbidity and mortality, with the need for further Brazilian comparative studies to assess the status before and after its implementation.

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Thrombolysis in primary brain tumor: anaplastic meningioma simulating recurrent ischemic stroke

Caroline Schiochet Verza[1], Glauco Kody Nagata[1], Victor Alfonso Garcia Ortiz[1], Carolina Heck Haubrich[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Caroline Schiochet Verza; Email: carolinesverza@gmail.com.

Case presentation: A 55-year-old woman presented to the emergency room with acute aphasia that began one hour earlier. Her medical history was notable only for mild, treated hypertension. Upon arrival, her National Institute of Health Stroke Score (NIHSS) was 4. A CT scan showed no signs of bleeding or early stroke, and there were no contraindications to administering tissue plasminogen activator (tPA). Thirty minutes into the tPA infusion, she regained her language function but began to experience a severe headache. The tPA infusion was stopped, and an emergency CT scan revealed a small hemorrhage in the left parietal lobe. She remained asymptomatic during her hospital stay, and an MRI showed an area of diffusion restriction along with hypointense images on gradient echo, attributed to the tPA-induced hemorrhage. Subsequent stroke investigations revealed no specific etiology, and she was discharged 20 days later on aspirin and simvastatin. Three months later, she returned due to acute right-side weakness. This lasted about one hour and was not accompanied by changes in consciousness. A new CT scan showed a significant area of brain edema in the left parietal lobe. She was admitted and started on dexamethasone. MRI revealed a 4 cm mass in the temporo-parietal region with heterogeneous gadolinium enhancement. Extensive surgery was performed, successfully resecting the entire lesion. Histopathological examination confirmed an anaplastic meningioma (WHO grade III). The patient is now undergoing radiochemotherapy and remains free of permanent neurological deficits. **Discussion:** Thrombolysis in patients with brain tumors is rarely reported in the literature, resulting in the systematic exclusion of these patients from such therapy. Differentiating small, undiagnosed brain tumors from stroke can be challenging. A small percentage of patients treated with thrombolysis are later found to have stroke mimics rather than true strokes. In a systematic review, seizures were identified as the most common cause of stroke mimics. Our patient had a reversible deficit following a seizure caused by an anaplastic meningioma, a rare, malignant, and rapidly growing tumor. Since patients with brain tumors were excluded from thrombolysis trials, the risk of intracranial bleeding can only be inferred from case reports. Etgen and colleagues reviewed the use of thrombolysis in patients with brain tumors, concluding that it is potentially safe in cases of extra-axial lesions without significant mass effect. Some studies highlight the difficulty in differentiating ischemic stroke from seizures, suggesting that CT perfusion may assist. Thrombolysis minimizes morbidity and represents the current state of the art in treating acute ischemic stroke. Understanding conditions that mimic stroke and the associated risks of brain hemorrhage with thrombolytic therapy is crucial for expanding inclusion criteria and translating clinical trial results to real-life situations.

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Thrombolytic treatment to isolated brachiocephalic dissection related stroke: case report

Rodrigo Targa Martins[1], Cristiane de Oliveira[1], Kelin Cristiane Martin Hackenhaar[1], Ana Carolina Rodrigues Farias[1]

[1]Grupo Hospitalar Conceição, Porto Alegre RS, Brazil.

Correspondence: Cristiane de Oliveira; Email: c.deoliveirasm@gmail.com.

Ischemic stroke (IS) sums 62% of all strokes in the world, its cause includes frequently large vessel atheromatosis, cardioembolic event, small vessel occlusion and less often arterial dissections. In young arterial dissection plays a major role in IS etiology with carotid and vertebral most frequently involved. Dissections involving extracranial portions of these arteries are routinely treated with intravenous thrombolysis (IVT) in the acute phase. In contrast, aorta dissection represents a contraindication to IVT and demands surgical to early brain hypoperfusion correction. The occurrence of IS related to isolated brachiocephalic trunk dissection (IBD) is rare with even fewer known results of IVT treatment to this condition (n=1 to our knowledge). **Case presentation:** We describe a 63 y/o woman hypertensive with acute onset of left hemiplegia including face, neglect, dysarthria, right ocular deviation admitted 90 minutes after symptoms onset and stroke severity scale (NIHSS) 17. She denied recent trauma, arm hyperextension or chest pain - but had light pain in the back of the neck. She had a symmetrical radial pulse and admission blood pressure was 110/90mmHg. Brain CT without contrast didn't show hyperdense artery or early hypodensity. She underwent IVT with 131 minutes of onset with improvement of symptoms finishing infusion with NIHSS 4. Because of worsening of neck pain now radiating to the right shoulder and appearing difference in BP between arms right 90x62mmHg, left 103x71mmHg she underwent and cervical CT-angio that showed BC dissection with 20 mm of maximal diameter, extending to right carotid artery and sparing aortic arch. She maintained improvement of neurologic symptoms attaining

NIHSS 0 in the first 24 hours, was urgently evaluated by cardiovascular surgery that defined conservative management based on resolution of neurologic symptoms and no critical arm hypoperfusion. Was maintained with anticoagulation with false lumen reduction and no new neurologic symptoms. **Discussion:** Our report involves uncommon stroke syndrome IS attributed to arterial dissection involving isolated IBD treated with IVT and improvement of neurologic deficits without worsening of arm perfusion or aortic involvement. The only case treated with IVT suggest succession of stroke mechanisms to explain early response and latter deficits reappearing. Similarly, as the cases described by Sansoni, ours didn't involve the base of BCT and in our cases atherosclerosis was notably absent in the arterial tree. **Final comments:** We conclude only with reliable estimates of main complications of IVT in this condition it will be possible to decide when to proceed to thrombolytic treatment vs surgical/endovascular approaches or if one would be generally advisable in neurologic presenting cases. So, this case does not ensure safety by absence of enough data but intends to contribute to this peculiar acute stroke etiology.

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Thunderclap headache in a puerperal patient

Luma Santos de Oliveira[1], Emanuelly Victoria Andrade Fonseca[1], Igor Flavio Gonçalves do Vale[1], Daniel Mariani Favalessa[1], Bruno Batitucci Castrillo[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Luma Santos de Oliveira; Email: luma.oliveira@edu.ufes.br.

Case presentation: A 35-year-old woman presented with Thunderclap headache during the immediate puerperium briefly after administration of metaraminol hemitartrate due to convulsive syncope. The headache was in the frontal region, of strong intensity, pulsatile, improving after 30 minutes. She had other 4 episodes of thunderclap headache over the next 10 days, interrupting her sleep. She denied visual symptoms, paresthesias and focal deficits. She had a history of vasovagal syncope with manifestation of convulsive syncope for years. She also reported a diagnosis of primary headache since the age of 18. She was taking sertraline and denied use of ilycit drugs. On examination, she had only pain on palpation of the occipital nerves and cervical muscles. Magnetic resonance angiography (MRA) was normal 3 days after the initial symptom. Patient preferred not to use any medication because she was afraid of lactating meanwhile using any pills. She had more episodes of thunderclap headache during the next days and a new MRA showed vascular narrowings and confirmed reversible cerebral vasoconstriction syndrome (RCVS). Verapamil was started and patient had an excellent response. **Discussion:** RCVS is understood as a thunderclap headache, with sudden onset and reversible multifocal narrowing of the cerebral arteries, lasting around three months with or without focal neurological symptoms. It comes on rapidly, peaking in less than a minute, and can be unilateral or bilateral, followed by diffuse, short-lasting pain. Its pathophysiology is partially understood, involving multiple mechanisms, such as dysregulation of cerebral vascular tone and genetic predisposition. In the case mentioned, the combined use of sympathomimetics after vasovagal syncope and serotonergic agents in a puerperal woman with a history of primary headache contributed to the development of CRVCS. It should also be noted that SSRIs can play a vasoconstrictive role in larger cerebral arteries, due to the increase in serotonin in the CNS, and that the puerperium also contributes to an increase in blood pressure and loss of cerebral autoregulation. Diagnosis is based on tests that prove diffuse cerebral vasoconstriction. Angiography can reveal alternating smooth and elongated narrowings and dilations, which can resemble a string of pearls. It should be borne in mind that the initial CT scan or MRI may be normal, and will have to be repeated a few days later if the suspicion is high. Treatment is carried out by removing possible causal agents, using calcium channel blockers as a prophylactic action, but there is still little scientific evidence to prove that the course of the disease has changed. **Final comments:** This highlights the intricate relationship between CRVCS, the use of medication and the postpartum period in women with a history of headache. It is of the utmost importance to better understand CRVCS, including its pathophysiological mechanisms and treatment strategies.

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Thyrotoxic hypokalemic periodic paralysis in a Brazilian male: a case report

Victor Hugo Soares dos Santos[1], Eduarda Kei-Lan Tavares Pierre[1], Emanuelle Campos Amaral[1], Fernanda Almeida Carvalho[1], Fernando Luiz Westphal Filho[2], Jonathas Taumaturgo Soares[1], Manuela Brandão Ferreira Teles[1], Vanise Campos Gomes Amaral[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Victor Hugo Soares dos Santos; Email: vhsds.med21@uea.edu.br.

Case presentation: A 49-year-old Brazilian male was referred for neurological evaluation due to the worsening of paresthesias in the calves and feet, muscle weakness, myalgia, and tremors in the lower limbs with periods of fluctuation over the past three months. Upon admission, the patient reported a recent flu-like illness with fever, nasal congestion, dry cough, and headache. Additionally, he had been diagnosed with hyperthyroidism two months prior and, despite treatment with methimazole, continued to experience insomnia, anxiety, fatigue, retro-orbital pain, exophthalmos, resting tremor in the upper limbs, hair loss, night sweats, cervical swelling, and a weight loss of 15 kg over five months. The patient was admitted to the medical ward, where he continued to experience episodes of flaccid paralysis in the lower limbs despite therapeutic intervention. On physical examination at hospital admission, the patient was in good general condition, alert, and oriented. Vital signs included a heart rate of 115 bpm. He exhibited asymmetric resting tremors in the upper limbs, cervical swelling, bilateral and asymmetric resting tremors in the upper limbs, and proximal paraparesis in the lower limbs graded as 2/5, and distal paraparesis graded as 3/5 bilaterally. Laboratory tests showed normal magnesium and phosphorus levels, with a serum potassium level of 3.0 mEq/L. **Discussion:** Thyrotoxic hypokalemic periodic paralysis (THPP) is frequently underdiagnosed due to its nonspecific clinical presentation and symptoms common to various pathologies. Differential diagnoses for ascending muscle weakness include Guillain-Barré syndrome; however, in this patient, cerebrospinal fluid analysis did not show cytoalbuminologic dissociation. The hypothesis of THPP is supported by the clinical presentation and low potassium levels, with the condition being more prevalent in males at a ratio of 20:1. **Final comments:** THPP should be considered in any patient presenting with muscle weakness associated with low serum potassium levels. Potassium replacement must be done cautiously due to its intracellular presence.

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Ticagrelor versus clopidogrel dual antiplatelet therapy for unruptured intracranial aneurysms: a systematic review and meta-analysis

Clara Rocha Dantas[1], Hideki Zimermann Kamitani[2], Williames Matheus Malaquias da Silva[2], Artur Menegaz de Almeida[3], Ítalo Barros Andrade[4], Lilianne Rodrigues Fernandes[5], Michele Kreuz[6], Isabela Junger Meirelles Aguiar[7], Vanessa Lopes Senssulini[7]

[1]Universidad de Buenos Aires, Buenos Aires, Argentina.
[2]Universidade Federal do Vale do São Francisco, Paulo Afonso BA, Brazil.
[3]Universidade Federal do Mato Grosso, Sinop MT, Brazil.
[4]Faculdade Santo Agostinho, Vitória da Conquista BA, Brazil.
[5]Afya Faculdade de Ciências Médicas, Abaetuba PA, Brazil.
[6]Universidade Luterana do Brasil, Canoas RS, Brazil.
[7]Universidade Anhembi Morumbi, Piracicaba SP, Brazil.

Correspondence: Vanessa Lopes Senssulini; Email: vsenssulini@gmail.com.

Background: Dual antiplatelet therapy (DAPT) with aspirin plus clopidogrel is the first-line antithrombotic regimen for unruptured intracranial aneurysms. However, in recent years, ticagrelor has been increasingly investigated to replace clopidogrel for this purpose since it has a faster effect and a more intense platelet inhibition. **Objective:** To perform a systematic review and meta-analysis comparing ticagrelor versus clopidogrel dual antiplatelet therapy in patients with unruptured intracranial aneurysms. **Methods:** PubMed, Scopus and Web of Science databases were searched for randomized controlled trials that compared ticagrelor to clopidogrel dual antiplatelet therapy for unruptured intracranial aneurysms and reported the outcomes of (1) Thromboembolic complications; (2) Mortality, and (3) Major Bleeding. Mean differences (MDs) with 95% confidence intervals (CIs) were calculated using a random-effects model. Heterogeneity was examined with I^2 statistics. Pvalues of ≤0.05 were considered statistically significant. Statistical analyses were performed using RStudio, version 4.2.3. **Results:** Three studies and 534 patients were included, of whom 252 (47%) received ticagrelor. The follow-up ranged from 3 to 12 months and the mean age was 57,15. Incidence of thrombo-embolic complications [7.49% ticagrelor vs. 8.60% clopidogrel RR 1.14 95%CI 0.56, 2.32 p=0.71; I^2=28%], major bleeding [8.88%ticagrelor vs. 6.52% clopidogrel RR 0.69 95%CI 0.38, 1.25 p=0.22; I^2=0%] and mortality [0.96% ticagrelor vs. 1.34% clopidogrel; RR 1.20 95%CI 0.26, 5.55 p=0.80; I^2=0%] did not reach a statistically significant difference between groups. **Conclusion:** Although most of the recent data indicate that ticagrelor appears to be safe and effective in replacing clopidogrel in DAPT settings for unruptured aneurysms, our results show no significant difference between the clopidogrel and the ticagrelor dual antiplatelet therapy arm.

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Time series analysis in pediatric stroke: investigating the differences in stroke trends and patterns according to age groups

Alisson Rafael de Oliveira Pereira[1], Lucas Abreu Dias[1], Pedro Henrique de Vilhena[1], Rodrigo Ferreira de Moura[1]

[1]Universidade Federal de Lavras, Lavras MG, Brazil.

Correspondence: Alisson Rafael de Oliveira Pereira; Email: alissonropereira@gmail.com.

Background: Stroke, the leading cause of death in Brazil since 2022, does not affect age groups uniformly, with a predominance in adults and the elderly, affecting less the pediatric group. The biggest difference between these groups was related to etiology. In adults, the main related factors are hypertension, smoking, and arrhythmias. In

the pediatric population, the deficit is caused by arteriovenous malformations (AVMs), coagulation dysfunction, and other congenital alterations. Although the possibility of recovery from neurological deficits in children is greater than that in adults due to neuroplasticity, it is important to understand the trends in the occurrence of the syndrome to anticipate interventions generating better prognosis. **Objective:** To evaluate trends in hospitalizations and deaths due to stroke in the pediatric age group as well as their types in the same age group. **Methods:** A retrospective study was conducted by tabulating the data recorded in SIH by DATASUS. Data on hospitalizations and deaths from ischemic, hemorrhagic, and unspecified strokes were collected. The data was grouped according to the WHO definitions of age group as childhood, adolescence and "pediatric" (sum of the groups). The data was analyzed using regression models. **Results:** During the study period, there was a global trend of increase in the rate of hospitalization for all types of stroke in the pediatric age group (AAPC= 1.5; 95%CI: 0.6 to 2.0; p<0.01), maintaining a stationary trend most of the time, with a joinpoint followed by a significant increase in 2021 (APC: 10.3, p<0.001). However, the overall trend of death decreased in the total period (AAPC= -2.3; 95%CI: -3.2 to -1.6; p<0.01). The analysis of the subgroups according to the type of stroke revealed that the trend of hospitalization for ischemic stroke did not change, maintaining a significant increase throughout the study period (APC= 2.4; p=0.01), while the hemorrhagic stroke group had a reducing effect (AAPC= -1.2, p<0.01). The unspecified stroke group showed a significant increase (AAPC= 5.4, p<0.01). The correlation between the three subgroups was evaluated according to covariances in an ARIMA model (0,1,0), seeking to identify whether the decrease in hospitalization due to hemorrhagic stroke would be compensated by the increase in unspecified cases; however, no significance was identified in the estimated effect (B= -0.17; SE= 0.3; t= -0.5; p=0.6). **Conclusion:** In the pediatric age group the trend of increase in the rate of hospitalizations due to stroke occurred at the same time that there was a decrease in deaths from the same disease. As for the types of stroke, cerebral infarctions have maintained their initial upward trend, and there is insufficient evidence to suggest that the decrease in hospitalization for intracerebral hemorrhage is related to the increase in unspecified stroke cases.

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Time to diagnosis of dmd patients with or without development delay

Marco Antônio Veloso de Albuquerque[1], Karlla Danielle Ferreira Lima[1], Fernando Kok[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Karlla Danielle Ferreira Lima; Email: karlla.dfl@gmail.com.

Background: Duchenne muscular dystrophy (DMD), the most common inherited childhood neuromuscular disease, typically starts around 3-5 years with progressive weakness, but a group of boys presents symptoms in early childhood with delayed development milestone, including walking and language delay. Despite the availability of genetic tests, the age of diagnosis has remained unchanged in the last decade. **Objective:** To analyze the age of diagnosis in a cohort of 127 DMD boys, comparing the time to diagnosis between patients with normal development milestones (group 1) and those with a delay of walking and / or speech acquisition (group 2). **Results:** The mean age of diagnosis in the cohort was 6.9 years (range: 2-16 years), with the meantime interval until genetic diagnosis of 3.6 years (range: 1-13 years). From the total, 89/127 (70%) showed an initial motor symptom with normal development. In this group, the mean age at presentation was 3.8 years (range: 1 to 7 years), and the age of genetic diagnosis was 7.5 years, (range: 2-16 years). On the other hand, 20% of patients (29/127) showed some development delay (group 2). In this group, the initial symptom was noted in a mean age of 1.4 years (ranged 0.5 - 2 years), with a delay of 3.4 years until diagnosis; 2.7 years earlier than group 1. **Conclusion:** Although patients with DMD and developmental delay presented earlier manifestations and diagnosis, the time to diagnosis did not differ much from those without delay in motor development. The importance of considering the diagnosis of DMD in patients with delayed motor development and elevated serum CK levels is highlighted.

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Time to high-efficacy treatment for multiple sclerosis in Brazil

Caio Hussid de Góes[1], Gabriella Chinaglia Zanin[1], Diego Olympio Peixoto Lopes[2], Maria Tereza Martinez[3], Fernanda Herculano[4], Diogo Haddad Santos[4]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Universidade Santo Amaro, São Paulo SP, Brazil.
[3]CP Quali, São Paulo SP, Brazil.
[4]Hospital Alemão Oswaldo Cruz, São Paulo SP, Brazil.

Correspondence: Caio Hussid de Góes; Email: caio.goes@aluno.fcmsantacasasp.edu.br.

Multiple sclerosis is a central nervous system affecting, chronic demyelinating disease, mediated via inflammatory mechanisms. It can result in progressive and permanent disabilities in both the physical and cognitive spectrum, the degree of impairment associated with time to initial treatment. Recent data challenges the previous status quo of initiating treatment with low-efficacy treatments and escalating to High-Efficacy disease modifying drugs based on relapse, due to reduced annualized relapse rate and reduced disability progression associated with early High-Efficacy treatment. Thus the study's objective is to evaluate the time to introduction of High-Efficacy drugs within 5 years of newly diagnosed Multiple Sclerosis in a Brazilian population. An electronic medical record aggregator called TriNetX was used to anonymously obtain the patient data used in the study, using exclusively hospital related records. The cohort was built using a Brazilian-only database via selecting patients who received the Multiple Sclerosis diagnosis in the last 10 years time frame, had never received an MCI or Multiple Sclerosis diagnosis before, and were at least 18 years at the time of the diagnosis. The Multiple Sclerosis diagnosis was considered the index event, while the outcome was the introduction of the high-efficacy medications available within 5 years of the index event. The high-efficacy drugs investigated in the study were natalizumab, fingolimod, alemtuzumab, cladribine, rituximab, ofatumumab and ocrelizumab. The resulting cohort had N=3,181 patients, with 2,747 patients who did not used High-Efficacy therapies as first line treatment. The remaining 434 patients had a median follow-up of 490 days. In this group, quickest therapy to introduce was Fingolimod, used by 25 patients, with a median time to treatment of 32 days, followed by Natalizumab, used by 228 patients, with a median time to treatment of 39 days. Rituximab was next, used by 34 patients, and with a median time to treatment of 76 days, followed by Ocrelizumab, used by 76 patients, with a median time to treatment of 79 days. The longest time to treatment was found with Alemtuzumab, used by 6 patients with a median time to treatment of 591 days. No patients used ofatumumab or cladribine as first line therapy. Established literature demonstrates benefit associated with introduction of High-Efficacy therapy within the first 6 months (approximately 180 days) of diagnosis. Based on the gathered results, apart from Alemtuzumab, the investigated therapies have been administered in adequate time. An unforeseen result obtained was the 86% of the initial cohort with Multiple Sclerosis who were not under High-Efficacy therapies, likely pointing to a lack of accessibility to these therapies. This is possibly a reflection of the high price associated these therapies or insurance policies restricted by specific clinical presentations of the disease.

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Very early-onset Alzheimer's disease in a 29-year-old patient from the interior of Bahia

Sheila Ferreira Pessoa[1], Sonia Maria Dozzi Brucki[1], Cristiane Araújo Martins Moreno[1], João Luiz de Carvalho Santos[2], Vitor Guimarães Corrêa[1], Raphael Ribeiro Spera[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Sheila Ferreira Pessoa; Email: sheilaferreirapessoa@hotmail.com.

Case presentation: A 29-year-old woman with no history of previous comorbidities began experiencing progressive amnestic impairment and executive function decline at the age of 25. Over four years, this progressed to rigid-akinetic parkinsonism, myoclonus, and severe language impairment. Her paternal family history was unknown, and her maternal family history did not reveal any similar conditions. She tested negative for potentially reversible causes. CSF analysis was within normal limits but showed a biological signature for Alzheimer's disease (AD) with elevated P-tau: 176 pg/mL (normal range < 61); beta-amyloid 1598.00 ng/L (normal range < 550 pg/mL), and an AB42/AB40 ratio of 0.100 (normal range ≤ 0.10). MRI revealed global volumetric reduction. Given the clinical and biological findings, and the complexity of the case, including the early onset of symptoms, molecular analysis via DNA sequencing was performed. This identified a pathogenic mutation in the PSEN1 gene in heterozygosity (c.617G> T, p.Gly206Val) and an APOE genotype of E3/E4 in heterozygosity. **Discussion:** AD is the most common progressive neurodegenerative condition and is associated with aging. However, it is also the leading cause of pre-senile dementia. Very early-onset forms can present with atypical initial symptoms and are more strongly related to genetic factors. About 10% of cases are caused by mutations in three genes: presenilin 1 (PSEN1), presenilin 2 (PSEN2), and amyloid precursor protein (APP). Mutations in the PSEN1 gene present phenotypic heterogeneity, including amnestic presentations, executive function impairment, attention deficits, and also epileptic seizures, myoclonus, spastic paraparesis, and parkinsonism. Several mutations in the PSEN1 gene are associated with very early-onset AD (< 35 years of age). The patient's described variant (p.Gly206Val) has been previously reported to be associated with symptom onset before the age of 30, an extremely rare condition. Additionally, our patient has the E4 allele in heterozygosity, which may have accelerated the appearance of symptoms. **Final comments:** Given its severity and the particularities in terms of diagnosis and treatment, early-onset AD represents a crucial field of research. Identifying genetic biomarkers and elucidating underlying molecular mechanisms are essential steps for advancing early diagnosis, monitoring disease progression, and developing more effective therapies. Better understanding the genetic bases and pathological processes of early-onset Alzheimer's will not only help manage this specific condition but may also offer valuable insights into the more common late-onset form of the disease.

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TLR4 (rs10759931 and rs2737190) and FCGR2A (rs1801274) polymorphisms as potential pharmacogenetic predictors of natalizumab therapeutic failure in multiple sclerosis

Matheus Dumans de Matos[1], Rafaella de Carvalho Cardoso[1], Bárbara Dahmer Guimenes[1], Larissa de Araujo Duarte[2], Vinícius Batista Domingues[1], Felipe da Rocha Schmidt[1], João Gabriel Dib Farinhas[1], Guilherme Carneiro Montes[1], Soniza Vieira Alves-Leon[2], Fabrícia Lima Fontes-Dantas[1]

[1]Universidade do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Matheus Dumans de Matos; Email: matheusdmatos@gmail.com.

Multiple sclerosis (MS) is an autoimmune, degenerative and demyelinating disease that affects the central nervous system (CNS) and represents the leading cause of neurological disability in young adults. Natalizumab (NTZ) is recognized as a safe and effective monoclonal antibody (mAb) in the treatment of MS as it suppresses the migration of immune cells to the CNS mitigating inflammation. Genetic factors can significantly influence the therapeutic outcomes of MS treatment, however, there are few pharmacogenetic predictors of therapeutic outcomes in the treatment of MS patient. Therefore, this study aims to analyze the feasibility of TLR4 (rs10759931 and rs2737190) and FCGR2A (rs1801274) polymorphisms as a potential pharmacogenetic predictor of therapeutic failure.Were collected 104 blood samples from patients admitted to two university hospitals in Rio de Janeiro (CAAE 64171122.7.0000.5259 / 69411517.2.0000.5258). A detailed analysis of each patient was conducted, including an extensive clinical history (demographics, age of onset, disease duration, Expanded Disability Status Scale (EDSS) score and treatment history) and brain magnetic resonance images. The sample was categorized into two groups according to their response to NTZ treatment (responders and non-responders). Genomic DNA was extracted from leukocytes for subsequent genotyping by allelic discrimination using the TaqMan® PCR assay. TLR4 rs10759931 functional assay was conducted on peripheral blood mononuclear cells (PBMCs) of healthy volunteers. This was followed by stimulation with 1μg of LPS for 24h and subsequent assessment of mRNA expression. Among the patients, 9 confirmed therapeutic failure to NTZ treatment. The genotypic distributions were in Hardy-Weinberg equilibrium, there was no linkage disequilibrium (LD) between the two TLR4 SNPs (D >0.9). Statistical analysis identified significant association between the GG genotype of FCGR2A (p=0.0004, OR=0.3530) and therapeutic failure. Furthermore, a higher frequency of the AG genotype was observed in patients who did not experience treatment failure (p=0.0002, OR=3.144). Individuals carrying the TLR4 (rs10759931) AA genotype were observed in the good response group (p> 0.0002) associated with decreased expression of TLR4 mRNA. Our prior results for the TLR4 (rs2737190) also support the AA genotype as a good response group (p>0.0001), corroborating a possible protective role. Given the genetic diversity of the Brazilian population due to admixture, our preliminary findings suggest the need to investigate the role of FCGR2A rs1801274, TLR4 rs10759931 and rs2737190 as a potential pharmacogenetic predictor for MS patients' response to NTZ.

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Tocilizumab as second line treatment option to refractory anti-NMDAR encephalitis with severe clinical complications

Renato Pierre Lima[1], João Henrique Fregadolli Ferreira[1], Marcos Vinícius Tadao Fujino[1], Ana Luíza Vieira de Araújo[1], Lívia Almeida Dutra[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Renato Pierre Lima; Email: rpierrelima@gmail.com.

Case presentation: A prior healthy 30-year-old female patient presented nausea and vomiting for 4 days. She evolved with aphasia and two episodes of tonic-clonic seizures. Brain MRI and CSF had normal results. After a week she developed psychosis and catatonia and was admitted to a psychiatric ward. During the next 14 days, she seizures frequency increased, she developed orofacial dyskinesias, and was admitted to the ICU. Antineuronal antibodies panel in CSF and serum confirmed anti-NMDAR encephalitis (anti-NMDARE). She was started on IVIG and steroids 35 days after firs symptoms with no improvement, and evolved with refractory status epilepticus, acute renal injury requiring hemodialysis, segmental pulmonary thromboembolism, and sepsis due to Pseudomonas with gluteus abscess. Upon admission to the ICU, the patient was under sedation, on mechanical ventilation, with RASS -3, receiving propofol and fentanyl. Pupils were isofotoreactive. The patient exhibited orofacial-limb dyskinesias and appendicular rigidity. Clinical evaluation at this point revealed mRs 5 and CASE of 22. Laboratory exams showed Hb of 6.4g/dL, platelets 46.000/ml, Cr 2.52mg/dL, normal serum immunoglobulins, negative TB Gold, and anti-HBs and IgG anti-HBc positive. EEG showed extreme delta brush. Due to the multiple clinical complications and severe infection, along with the risk of hepatitis B virus reactivation, she was started on tocilizumab 8mg/kg and tenofovir 300mg/day. She evolved with slowly clinical and neurological improvement in the following 30 days, did not have additional clinical complications, was weaned from mechanical ventilation and became seizure-free. Rituximab was started after a washout period of 30 days from the last tocilizumab dose. **Discussion:** We report a patient that received tocilizumab as second line option for anti-NMDARE, due to many clinical complications and formal contraindication to rituximab. Tocilizumab effects start 5 days after infusion and lasts for 30 days, which is suitable for unstable patients with severe complications in the ICU, as opposed to rituximab, which may take 15-30 days for initial effect. Although no controlled trial evaluated the efficacy of tocilizumab versus rituximab in anti-NMDARE, this case illustrates how patients in the ICU may benefit from other therapies, as time to treatment is a variable with prognostic value. **Final comments:** Tocilizumab may be considered as second-line treatment to patients with severe complications in ICU.

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Tocilizumab for cryptogenic fires with hemofagocytic syndrome: a case report

Anna Maria Gomes[1], João Henrique Fregadolli Ferreira[1], Rejane de Souza Macedo Campos[1], Carol Ladeia-Frota[1], Victor Rebelo Procaci[1], Livia Almeida Dutra[1]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.

Correspondence: Anna Maria Gomes; Email: g.annamgomes@gmail.com.

Case presentation: A previously healthy 11-year-old boy presented with fever, rash and vomiting. Five days later, he developed bilateral tonic-clonic seizures, and status epilepticus. He had experienced childhood infections associated with disproportionate febrile reactions and migratory arthritis, but no previous autoimmune diseases diagnosis. Family history was unremarkable. Lab results showed mild leukocytosis 14120 and anemia 10.3g/dl. Brain MRI was normal. CSF analysis demonstrated 1 cell, 24 mg/dL protein, 13.5mg/dl lactate, 62 mg/dL glucose, OBC were absent, antineuronal and antiglial antibodies were negative. He evolved with super-refractory status epilepticus, despite the use of eight different antiseizure medications. He was diagnosed with Febrile Infection-Related Epilepsy Syndrome (FIRES) and was started on methylprednisolone and plasmapheresis (4 cycles) two weeks after the initial symptoms followed by anakinra. After twelve days, anakinra was discontinued due to posterior reversible encephalopathy syndrome. CSF cytokine profiling showed significantly increased IL-6 (31.2 pg/mL) and IL-1 (<6.5 pg/ml). Forty-four days after the onset of seizures, tocilizumab was administered and repeated biweekly. Seizures improved, nonetheless the patient developed persistent fever, increased ferritin, bicytopenia, hypertriglyceridemia and elevated transaminases, indicating hemophagocytic lymphohistiocytosis (HLH), for which intrathecal dexamethasone and etoposide were introduced (5 cycles). One dose of cyclophosphamide was administered the day after dexamethasone, resulting in improvement of the EEG and systemic inflammation. The patient neurological condition gradually improved, and he was discharged home, ambulatory with mild cognitive impairment, with tocilizumab and antiseizure medication. Exoma was normal. **Discussion:** FIRES is rare type of New-Onset Refractory Status Epilepticus (NORSE), and it is characterized by the sudden onset of a febrile infection between 24 hours and 2 weeks before the onset of refractory status epilepticus, with or without fever at the onset of RSE. This case illustrates that cytokine profile can help in switching medications, and also shows that brain inflammation can trigger systemic inflammation leading to HLH. Moreover it shows how FIRES requires intensive immunossupressive treatment and association of depleting agents as well as cytokine blockage. **Final comments:** Cryptogenic FIRES is a challenging disease, which can evolve with multiple complications, including HLH.

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Todd's paralysis as a differential diagnosis for stroke

Emanuelly Valentim Oliveira[1], Maria Luiza Rodrigues Defante[1], Diogo de Souza Vargas[1], Henrique Rosa Henriques[2]

[1]UniRedentor, Itaperuna RJ, Brazil.
[1]Universidade Iguaçu, Itaperuna RJ, Brazil.

Correspondence: Emanuelly Valentim Oliveira; Email: valentimanu@outlook.com.

Case presentation: A 42-year-old male patient with no previous comorbidities presented for blood donation. During the procedure, he experienced a tonic-clonic seizure and was taken to the local emergency department, where he had another seizure episode. Around 2 PM that day, he exhibited sudden onset focal neurological deficit, presenting with sensory aphasia and complete brachiocrural hemiplegia disproportionately affecting the right side with brachial predominance. Initial suspicions included Todd's Paralysis (TP) and acute ischemic stroke (AIS). A cranial CT scan showed no bleeding, leading to a conservative approach with acetylsalicylic acid, lamotrigine, clopidogrel, and simvastatin. The patient was admitted for further investigation and transferred to the Intensive Care Unit. Electroencephalography ruled out epileptogenic activity. A follow-up CT scan after 24 hours revealed ischemic lesions, confirming the diagnosis of AIS. **Discussion:** AIS, the most common cerebrovascular disease, refers to ischemia resulting from obstruction, vasospasm, or narrowing of cerebral arteries. Clinical manifestations depend on the occluded artery's location. The middle cerebral artery is most commonly involved, presenting with hemiparesis, aphasia, hypoesthesia, central facial paralysis, and hemineglect. TP, on the other hand, is a postictal state following tonic-clonic and absence seizures, manifesting as hemiparesis, aphasia, hemineglect, and visual disturbances, lasting from minutes to up to 36 hours. Differentiating TP from AIS can be challenging, particularly due to the focal impairments in the postictal state. Diagnosing TP requires investigating the patient's medical history to identify possible AIS risk factors and previous seizure occurrences, as well as knowing the medications being used. A CT scan, showing ischemic lesions 24 hours post-event, and an EEG, which ruled out epileptogenic activity, are crucial in this differentiation. **Final comments:** Differentiating between

TP and AIS is vital. While TP does not require medical intervention, AIS management depends on blood pressure and vascular control, utilizing medications that affect blood viscosity along with fibrinolytic administration. Thus, understanding the differential diagnosis is essential to guide appropriate clinical management, avoiding potential iatrogenesis in TP cases and/or delays in AIS treatment.

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Tongue tremor as a presentation of focal seizure: a rare case report

Ana Letícia Amorim de Albuquerque[1], Cândida Driemeyer[2], Carolina Oldoni[3], Rhaná Carolina Santos[1], Rafaela Jucá Lindner[1], Amanda da Silva Araújo[1], Natália Toebe Giudice da Costa[1]

[1]Universidade do Vale do Rio dos Sinos, São Leopoldo RS, Brazil.
[2]Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre RS, Brazil.
[3]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Cândida Driemeyer; Email: candidaplena@gmail.com.

Case presentation: A 43-year-old female patient, with unknown comorbidities, was brought to the emergency room with involuntary episodes of tongue tremors, associated with sialorrhea and dysphagia, occurring over the past 24 hours. The episodes last about 2 minutes, are involuntary, and are associated with twitching of the second and third fingers of the right hand and dysarthria during the episodes. In the preceding week, she reported a mild headache without alarm signs, which was relieved by dipyrone. There were no focal deficits on physical examination and no laboratory abnormalities. A cranial MRI with contrast revealed a recent intraparenchymal hemorrhage in the left inferior frontal gyrus measuring 1.8 cm, associated with a vascular structure extending towards the subependymal region of the left lateral ventricle, suggestive of a developmental venous anomaly. A contrast-enhanced CT angiography confirmed the presence of an arteriovenous malformation (AVM). Given the semiology of the movements and the imaging findings, the possibility of focal seizures was considered. Administration of diazepam during an episode resulted in the cessation of movements, supporting this impression. Consequently, the patient was treated with phenytoin without further recurrence of seizures. **Discussion:** AVMs are congenital anomalies of intracranial vessels, characterized by a cluster of abnormal arteries and veins, resulting in abnormal and high cerebral blood flow. AVMs tend to present clinically around the age of 30, with intracranial hemorrhage, progressive neurological deficit, and/or seizures. Focal epilepsy presenting as isolated tongue tremor is a rare condition, highlighting the diversity in the presentation of epileptic seizures. This condition is particularly relevant in the context of an AVM, as the vascular anomaly can create an environment suitable to epileptic activity due to altered blood flow and brain structure. Thus, seizures may manifest in less common cerebral areas, such as the tongue. **Final comments:** Although previously reported in other studies, focal epilepsy of the tongue area is a rare manifestation with diverse causes. We present a case where the epileptic focus is related to a structural origin of abnormal cerebral blood vessels. Medical follow-up is essential for monitoring the condition to prevent further seizures and, in the case of AVMs, new hemorrhages. Therefore, we highlight the importance of considering AVMs in the differential diagnosis of similar cases, especially in atypical presentations in young patients. The diagnostic and therapeutic approach should be comprehensive and multidisciplinary, aiming at both seizure control and the correction of the underlying vascular anomaly when deemed possible.

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Tongue ultrasound assessment in patients with nemaline myopathy

Ana Lucila Moreira[1], Cristiane Araujo Martins Moreno[2], Clara Gontijo Camelo[2], Edmar Zanoteli[2]

[1]CENEC Neurofisiologia Clínica, Campinas SP, Brazil.
[2]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Ana Lucila Moreira; Email: ana.lucila.moreira@gmail.com.

Background: Dysphagia in Nemaline Myopathy (NM) is commonly associated with altered orofacial structure, but tongue function may be a key feature in the evaluation of these patients. **Objective:** To describe the changes found on ultrasound of the tongue muscles in patients with NM. **Methods:** Fifteen patients with NM underwent ultrasound using muscle-specific setup with fixed gain (GE Logic E) with a 4 to 12 MHz transducer, evaluating the muscles of the tongue in coronal section with B-mode and M-mode, the latter being used with a plane on the digastric muscle to evaluate the number of movements performed during saliva swallowing. The muscles were graded using the modified Heckmatt scale (Grade 1 - normal muscle; Grade 2 - probably abnormal; Grade 3 - abnormal). **Results:** Nine patients were male, aged between 1 and 54 years (median 16 years). Five patients had severe dysphagia with exclusive use of gastrostomy, six had mild dysphagia and four had normal swallowing. All patients showed involvement of the genioglossus muscle on ultrasound. The geniohyoid was the least commonly affected muscle on ultrasound, being altered in only seven patients, five of which had tongue atrophy. The number of movements performed for swallowing was evaluated in eleven patients and was greater in patients with involvement of more than one muscle evaluated. **Conclusion:** Ultrasound of tongue muscles is capable of detecting changes in patients with NM even before the development of atrophy or dysphagia. Early recognition of these changes is important for therapeutic guidance.

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Toxoplasmic encephalitis in a patient with multiple myeloma: a case report

Josenice Gomes Trindade Seidel[1], Ahmad Kassem El Zein[1], Joana Laura Aihara Barbosa[1], Matheus Rodrigues Nogueira[1], Lucas Henrique de Moura Rogerio Garcia[1], Romulo Loss Mattedi[1], Julia Martins Brunelli[1], Lúcio Huebra Pimentel Filho[1], Bruno Fukelmann Guedes[1], Tarso Adoni[1]

[1]Hospital Sírio Libanês, São Paulo SP, Brazil.

Correspondence: Josenice Gomes Trindade Seidel; Email: josegomes15@hotmail.com.

Case presentation: An 83-year-old woman previously diagnosed with IgG Kappa multiple myeloma (MM) currently in remission was admitted to the emergency department due to a 10-hour history of mental confusion and impaired speech. The previous oncological approach was induction therapy with lenalidomide and dexamethasone followed by maintenance treatment with lenalidomide, and the patient was considered ineligible for an autologous hematopoietic stem-cell transplantation (AHSCT). On neurological evaluation, the patient presented global aphasia, psychomotor agitation, and right-hand paresis. Brain magnetic resonance imaging (MRI) identified a ring-enhancing oval cortical and subcortical lesion in the left temporoparietal region. An 18F-FDG PET/CT scan showed increased metabolic expression in the same region. IgM antibodies for toxoplasmosis were negative, but IgG antibodies were positive. Serology tests for HIV, hepatitis B and C, syphilis and tuberculosis were negative. Cerebrospinal fluid analysis revealed increased protein with lymphocytic pleocytosis and normal glucose levels, negative bacterioscopy and cultures, as well as negative for syphilis and toxoplasmosis. Proton MRS and perfusion MRI showed eccentric target sign and scattered nodular foci of enhancement in both cerebral hemispheres. Increased blood volume in the left hemisphere and reduced neuronal viability were noted. The possibility of brain metastases was considered. Given the neurological worsening with seizures and diagnostic uncertainty, a brain biopsy was performed and demonstrated inflammatory infiltrate with extensive coagulative necrosis and the presence of grouped parasites in the form of pseudocysts, as well as free in the parenchyma, suggestive of Toxoplasma gondii bradyzoites and tachyzoites. Both polymerase chain reaction and immunohistochemistry were positive for toxoplasma in the brain tissue. Treatment with trimethoprim/sulfamethoxazole was promptly initiated, resulting in a significant improvement in the patient's clinical condition. **Discussion:** Toxoplasmosis is the most common opportunistic infection of the central nervous system in patients with HIV, but patients with MM also have an increased risk of infections due to dysfunctional adaptive immunity. Furthermore, the use of chemotherapy and immunomodulatory therapy has increased typical and atypical neurological infections. Rare cases of cerebral toxoplasmosis in HIV seronegative patients with MM, were described in the literature, most of them in patients undergoing AHSCT or using multiple chemotherapy drugs. We report a rare case of toxoplasmosis reactivation, manifested as acute encephalitis, in a patient with MM in remission, considered ineligible for an AHSCT, HIV seronegative, and using lenalidomide only. **Final comments:** Toxoplasmic encephalitis should be considered as a differential diagnosis in patients with MM, regardless of disease status and therapy.

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Tracing the legacy of arnold pick on his death centennial: insight into frontotemporal dementia

João Ignacio Ferrara Neto[1]

[1]Universidade Federal do Rio de Janeiro, Faculdade de Medicina, Rio de Janeiro RJ, Brazil.

Correspondence: João Ignacio Ferrara Neto; Email: joaoignacioferrara@gmail.com.

Background: Arnold Pick (1851-1924), born in Velké Mezi í í, Moravia, graduated from Vienna's medical school in 1875. He completed an elective at Vienna's Landesirrenanstalt and served as a student assistant to Theodor Meynert from 1872 to 1874, focusing on neuroanatomy. Pick, alongside Carl Wernicke, distinguished himself as a Czech psychiatrist and neurologist, advancing behavioral neurology. In 1892, Pick described what is now known as Pick's disease or frontotemporal dementia (FTD). He outlined distinct clinical symptoms and identified two types of agrammatism: temporal, characterized by defective syntax, rapid speech, and incorrect inflections; and frontal, marked by substantive word juxtaposition without syntactical elements. Pick's findings on brain atrophy in the frontal and temporal lobes significantly advanced the understanding of presenile dementia, solidifying his legacy in neurological research. **Objective:** This historical report aims to investigate Arnold Pick's pioneering work during the late 19th and early 20th centuries, focusing on his discovery and characterization of frontotemporal dementia (FTD), later termed Pick's disease. **Methods:** This historical analysis of Arnold Pick's work and legacy entails an extensive literature review of primary and secondary sources, encompassing medical journals and biographies. Key documents and case studies authored by Pick will be scrutinized to elucidate his contributions to neurology. Furthermore, a comparative analysis with contemporaries such as Alois Alzheimer will be conducted. **Results:** Arnold Pick's early articles lacked

histological descriptions of FTD until Alois Alzheimer's 1911 discovery of Pick Bodies–silver-staining globular inclusions. Today, Pick's disease is recognized by pathology featuring Pick bodies, Pick cells, and circumscribed brain atrophy. FTD is classified into three categories: behavioral variant, semantic variant primary progressive aphasia, and non-fluent variant primary progressive aphasia, all based on symptoms identified by Pick. Thus, Pick pioneered research on these topics and neuroanatomy and left a lasting legacy in the field. **Conclusion:** In conclusion, Arnold Pick's groundbreaking contributions to the understanding of frontotemporal dementia have profoundly influenced the field of neurology. His meticulous observations and pioneering descriptions continue to shape our understanding of neurodegenerative diseases, underscoring the enduring legacy of his work in modern medicine.

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Tract-specific spinal damage in SCA2, SCA3 and SCA6

Fabrício Catro de Borba[1], Joyce Macedo Sanches Fernandes[1], Thiago Junqueira Ribeiro de Menezes[1], Carelis González-Salazar[1], Lucas de Melo Teixeira Branco[1], Paulo Schneider Wolmer[1], José Luiz Pedroso[2], Orlando Graziani Povoas Barsottini[2], Marcondes Cavalcante França Junior[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Fabrício Castro de Borba; Email: fabriciocborba@gmail.com.

Background: Spinocerebellar ataxias (SCAs) are a group of neurodegenerative disorders characterized by progressive ataxia. **Objective:** To characterize spinal cord abnormalities in patients with SCA2, SCA3, and SCA6 and identify their phenotypic correlates. **Methods:** A multimodal spinal neuroimaging study was conducted on 26 SCA3, 16 SCA2, and 14 SCA6 patients, along with matched healthy controls. MRI scanning was performed using a 3 Tesla device, and the Spinal Cord Toolbox (SCT) was employed for morphometric and diffusivity analyses of the cervical spinal cord. **Results:** Significant spinal cord atrophy and altered white matter microstructural metrics were observed in SCA3 and SCA2 patients compared to controls, with no abnormalities in SCA6. A strong negative correlation was found between cross-sectional cord area and disease duration in SCA2, suggesting its potential as a biomarker for disease progression. **Conclusion:** This study highlights the importance of spinal cord imaging in understanding the pathophysiology of SCAs and demonstrates the utility of MRI-based metrics in identifying structural deviations and their clinical correlates. Further longitudinal studies are needed to validate these findings and explore their implications for clinical trials and therapeutic interventions.

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Transcranial direct current stimulation in patients with refractory epilepsy: a meta-analysis study

Marcos Vinícius do Nascimento da Silva[1], Katharine Aguiar de Souza[1], André Welisson Marques de Araújo[1], Lucas da Silva Pereira[1], Letícia Albuquerque Cossich[1], Artur Menegaz de Almeida[2]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.
[2]Universidade Federal do Mato Grosso, Sinop MT, Brazil.

Correspondence: Marcos Vinícius do Nascimento da Silva; Email: ufrj.marcosnascimento@gmail.com.

Background: Epilepsy is a neurological disorder characterized by recurrent, unprovoked seizures. A significant part of people affected by this disease has a condition called refractory epilepsy, which does not respond adequately to anti-seizure drugs. Therefore, transcranial direct-current stimulation (TDCS) is a neuromodulation technique and a promising treatment for those with this condition, given its ease of use and non-invasive nature. However, its benefits for this purpose remain unclear. **Objective:** To clarify the safety and efficacy of TDCS in reducing seizure frequency in patients with refractory epilepsy. **Methods:** We searched MEDLINE, Scopus, and Web of Science databases for randomized clinical trials (RCT) and observational studies that assessed transcranial direct current stimulation in refractory epilepsy patients. A random-effects model was used to calculate the mean differences (MDs) with 95% confidence intervals (CIs). Heterogeneity was assessed using I^2 statistics. R, version 4.3.2, was used for statistical analysis. **Results:** A total of four studies comprising 88 patients with epilepsy were included. For seizure frequency, TDCS demonstrated a statistically significant reduction compared with sham treatment, with a mean difference (MD) of -4.19 (95% CI -6.54 to -1.83, p=0.0005; I^2=62%).In contrast, for interictal epileptiform discharge (IED) frequency, TDCS showed no statistically significant difference compared with sham treatment (MD -3.69; 95% CI -8.20 to 0.81; p=0.11; I^2=64%). **Conclusion:** In this meta-analysis, consistent results suggest that while TDCS may significantly reduce seizure frequency, its effect on IED frequency remains inconclusive. Therefore, to evaluate the benefits of TDCS in IED, further studies are needed.

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Transcranial doppler assessment as an ancillary tool in the diagnosis of convulsive syncope with disrupted cerebral autoregulation in a pregnant woman

Vinícius Canhestro Roedel[1], Rodrigo Santiago Gomez[1], Mirian Fabiola Studart Gurgel Mendes[1], Aretha Miranda Silva[1], Bárbara Cristina Silveira de Oliveira[1], Rachel Laguardia Rego[1], Larissa Cruvinel Leite[1], Samuel Inácio Oliveira Araújo[1], Verônica Moreira Belo[1]

[1]Hospital Madre Teresa, Belo Horizonte MG, Brazil.

Correspondence: Vinícius Canhestro Roedel; Email: roedelvinicius@gmail.com.

Case presentation: A 22-year-old woman with depression and attention deficit hyperactivity disorder experienced an unplanned pregnancy and began having recurrent episodes of loss of consciousness. These episodes started around the twenty-sixth week of pregnancy, gradually increasing in frequency to several times a day, significantly impairing her daily life. The loss of consciousness was preceded by subjective sensations of "everything turning white" and "pressure in the ears," lasted a few seconds, and was notably triggered by standing up. During these episodes, the patient exhibited eye deviation and flexion posturing of hands and fingers, raising concerns about an epileptic origin of her symptoms. Investigations included magnetic resonance imaging, which was normal, and a video electroencephalogram (EEG), during which four events were recorded. These events were characterized by a drop in blood pressure and severe, generalized slowing of EEG waves. The patient was then evaluated using transcranial Doppler (TCD) for 35 minutes in a sitting position. Paroxysmal periods of tachycardia were observed, and during one of these periods, another episode of loss of consciousness occurred. This episode was marked by interrupted diastolic flow in the left middle cerebral artery, reduced systolic flow velocity, and a significant increase in the pulsatility index. **Discussion:** This case underscores the importance of a well-established differential diagnosis between epileptic seizures and convulsive syncopes. It also highlights the utility of TCD monitoring during an event to demonstrate a hemodynamic disorder as the cause. The patient and her family were oriented, avoiding unnecessary use of anticonvulsant drugs, which could be problematic during pregnancy. Furthermore, the TCD findings indicated increased cerebrovascular resistance due to vasoconstriction in the middle cerebral artery, consistent with findings from other studies using TCD during tilt-tests. This cerebral vasoconstriction in response to systemic hypotension is paradoxical, as the expected response would be vasodilation, suggesting a link to the pathophysiology of syncope. However, this case does not fit the classic neurocardiogenic syncope profile, as there was a tachycardia response instead of bradycardia, nor does it align with postural orthostatic tachycardia syndrome, since systemic hypotension was recorded. The pathophysiology might be related to the decreased venous return typical of pregnancy, resembling a hypovolemic shock and triggering an exaggerated adrenergic response that causes tachycardia and cerebral vasoconstriction. **Final comments:** TCD use in assessing episodes of loss of consciousness is not routine, but it can aid in diagnosis and provide new insights into cerebral circulation autoregulation. To our knowledge, this is the first documented case of using TCD assessment combined with EEG for episodes of loss of consciousness in a pregnant woman.

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Transcranial sonography in diseases with hippocampal atrophy

Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Rita de Cassia Leite Fernandes; Email: rilefe2016@gmail.com.

Background: Neurological sonography is a cost-effective and safe neuroimaging method. In many scenarios it may help to quickly establish a diagnosis, sometimes at the bedside, in patients with limited or delayed access to routine neuroimaging. The indications of Neurosonology are rapidly expanding outside the field of cerebrovascular diseases. Transcranial sonography (TCS) may assist in the differential diagnosis of movement disorders and a good correlation to MRI imaging in Alzheimer's dementia (AD) was found. Our **Objective:** To check the ability of TCS to visualize medial temporal lobe structures (MTLs). **Methods:** Preliminary proof-of-the-concept study to evaluate MTLs in patients of a universitary hospital. TCS was performed through the temporal bone window bilaterally with HD11XE (Philips) equipment using a sectorial 1-5MHz transducer. Axial and coronal insonation planes were used. A group with patients diagnosed with ILAE criteria for MTL epilepsy and MRI showing unilateral hippocampal sclerosis (MTL-HS) had the following structures analyzed and compared by both methods: choroidal fissure height (CF), temporal pole ventricle width (TP) and hippocampal height. The MTL atrophy index was calculated for both methods (MTLA=CF+TP). A second group was divided in AD patients with a > 2y diagnosis of a probable AD and a healthy control no dementia group. Hippocampi were evaluated and compared; the following measures were obtained: (A) lateral temporal pole, (B) MTL height, (C) MTL width, (D) ambiens cistern width and (E) choroidal fissure height. **Results:** Visualization of MTL structures by TCS was easy and feasible in all 23 participants. Only 1 patient had no bone window on one side. MTLE-HS group (n=4) had 3 men and ages 24-50y. Comparison between measures obtained by TCS and MRI showed a good correlation, specially the MTLA index, with a 74% agreement. The worst measures were the TP, with only 43% agreement. The laterality definition of HS by TCS

and MRI was 100%. In the other group we examined 4 DA patients and 15 controls, similar age. There was significant difference in MTL width between the groups (1.54+0.11 cm vs 1,91+0,33 cm; p=0.0027). The ratio B/E was the best to identify the dementia patients (2.23+0.22 vs 3.17+0.47; p=0.0001). **Conclusion:** TCS depicts MTL structures easily for an experienced operator. We compared measures obtained by TCS with MRI in a small group of HS patients and found them quite similar with identical laterality identification. Also, AD patients showed a ratio between MTL height and choroidal fissure height (B/E) significantly different from controls. Although the number of patients is small, this work points to a possible usefulness of TCS in evaluating MTL structures and, therefore, in diagnosing diseases with hippocampal atrophy. Studies with larger participant groups are highly desirable to prove TCS accuracy in clinical neurological practice as a neuroimaging modality in these diseases.

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Transient ischemic attack in a young patient with rete mirabilis circulation pattern in the middle cerebral artery: a case report

Hélio Aquaroni Farão Gomes[1], Itamar Meireles Andrade Santos[1], Dominick Esthephanny Monteiro Silva[1], Ana Beatriz Marangoni Baston[1], Pedro Machry Pozzobon[1], Caroline de Barros Viegas[1], Eduardo Abrão Spinola Rezk[1], Gabriela Rossini Dantas[1], José Eduardo Tentim Longo[1], Gabriel Pinheiro Modolo[1]

[1]Universidade Estadual Paulista, Botucatu SP, Brazil.

Correspondence: Hélio Aquaroni Farão Gomes; Email: heliofarao@hotmail.com.

Case presentation: GFO, 31 years old, in February 2024 presented with sudden onset of complete and proportionate right hemiparesis and dysarthria lasting around 25 minutes, denies head trauma, denies headache, in evaluation patient admitted asymptomatic, kept in neurological observation in stroke unit for 48 hours without clinical recurrence of deficits. He was investigated with a CT scan of the brain with an ASPECTS of 10 and no acute ischemic lesion or even indirect signs of ischemia. On CT angiography, he appeared to having a stop in M1 to the left, with heterogeneous and posterior hyperdensity in M2 which may correspond to an atypical circulation pattern; laboratory investigation has not found alterations in rheumatological or coagulation tests, or metabolic tests. MRI angiography and arteriography were then requested to complete the diagnosis and the study of the two exams revealed a network of exuberant insular perforating branches and slowed distal filling of the left middle cerebral territory, disconnecting the distal M1 portion with M2, corresponding to the rete mirabilis/Twin like pattern. Once the investigation was concluded, the patient was followed up as an outpatient with secondary prophylaxis with double antiaggregation for 21 days followed by monoaggregation and there has been no recurrence of the condition. **Discussion:** The rete mirabilis pattern is more common in other animal species, its formation does not occur during the fetal period and is usually formed by a network of physiological collaterals after a situation of vascular agenesis, whether of branches of the carotid or vertebral portion, shown on imaging exams with a hypoplastic vessel and circulatory compensation has a network shape maintaining circulation. Patients with a rede mirabilis circulation pattern are usually discovered in the event of an ischemic stroke, subarachnoid hemorrhage, intraparenchymal hemorrhage and carotid-cavernous fistula, showing the association with various vascular abnormalities, as was also the case with the patient presented in this case, with vascular hypoplasia noted in the middle cerebral artery. Suspicion of the condition is consistent when vascular hypoplasia/agenesis is seen in association with a network of small vessels in close proximity to the preserved subsequent circulation, with arteriography being the definitive diagnostic method for the condition. The best prophylaxis for the condition has not yet been defined in the bibliography, so the choice of prophylaxis was based on the prophylaxis normally used in transient ischemic attacks in this case. **Final comments:** with the development of new imaging methods, circulation variants are increasingly recognized and may be important causes of cerebrovascular alterations. Their recognition and increased presentation will motivate further research for better monitoring, prophylaxis and surgical approaches to avoid consequences for patients.

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Transverse myelitis following arbovirus infection (dengue): a case report

Rodrigo Batista Irikura[1], Isabella Brasileiro Quintella Migueis[1], Anouar Sacca Colognesi[1], Guilherme Leonel Gomes da Cunha[1], Talles Camin Rajab[1], Sarah de Souza Chinelato[1], Bruna Santos Silva Guzzardi[1], Gustavo Carvalho Viveiros[1]

[1]Faculdade de Medicina de Marília, Marília SP, Brazil.

Correspondence: Isabella Brasileiro Quintella Migueis; Email: isabellabrm2@gmail.com.

Case presentation: A 33-year-old male patient presented to the Emergency Department with a history suggestive of dengue fever starting around May 27, 2024, characterized by fever (not measured), myalgia, and petechiae throughout the body. On June 5, 2024, around 10 PM, he experienced sudden onset of lower limb weakness resulting in a fall, and the following day (June 6, 2024), he woke up with paraparesis and paresthesia, prompting him to seek emergency evaluation. The patient developed urinary retention, necessitating placement of an indwelling urinary catheter. He denied lower back pain, upper limb weakness or paresthesia, dysphagia, visual changes, or other neurological complaints. He was admitted under the Neurology team for diagnostic investigation, including Brain MRI and cervical and thoracic spine MRI, as well as Dengue serology. The diagnosis of Transverse Myelitis following Arbovirus (Dengue) infection was established. **Discussion:** Transverse myelitis is a neuroimmune disorder affecting the spinal cord, characterized by rapid onset of muscle weakness, bladder dysfunction, and sensory disturbances. It typically follows an infectious illness but can also be associated with neuroinflammatory disorders such as multiple sclerosis. **Final comments:** Transverse myelitis is a rare condition, with an annual incidence of one to nine cases per million people. Therefore, enhanced understanding of the disease is crucial for suspicion and diagnostic investigation, as early treatment initiation may help prevent neurological damage.

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Trauma car accident and Miller-Fisher syndrome: a case report

Bárbara Borges de Oliveira[1], Brenda Letícia Lopes Batista[1], Victor Souza Torres de Vasconcelos[1], Yanne Almeida Aguiar[1], Monique Evelyn Mendonça do Nascimento[1], Mariana Holanda Gameleira[1], Ana Carolina Paiva Farias[1], Rosevaldo Alves Junior[1], Maria Lúcia Brito Ferreira[1], Ana Rosa Melo Correa Lima[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Bárbara Borges de Oliveira; Email: barbaraborgesneurologia@gmail.com.

Case presentation: A 25-year-old male patient presented with symptoms of binocular diplopia and imbalance 3 days after being admitted to the hospital. He had been involved in a car accident 4 days prior to the onset of symptoms but did not experience any head trauma or other complications at the time. The patient had no recent history of infection or vaccinations. The neurological examination revealed bilateral mydriasis, semiptosis, ophthalmoparesis, global arreflexia, dysmetria, and Romberg's sign. CSF analysis showed protein-cytological dissociation, and viral serologies were negative. MRI of the brain showed no abnormalities, and ophthalmologic evaluation revealed no issues. The patient was diagnosed with Miller-Fisher syndrome (MFS) possibly related to the trauma and treated with intravenous human immunoglobulin for 5 days. Although there was partial improvement in oculomotricity, the patient still had some symptoms upon discharge, which eventually resolved over the following months. **Discussion:** MFS is considered a rare variant of Guillain-Barré syndrome, an acute inflammatory and immune-mediated demyelinating polyradiculopathy, with an estimated incidence of 1 to 5% of cases in Western countries. It is characterized by the triad of ataxia, arreflexia and ophthalmoplegia. In addition to protein-cytological dissociation in the CSF, the presence of anti-GQ1b antibodies reinforces the diagnosis, as they are positive in more than 90% of MFS cases, although this is not pathognomonic. Although the syndrome is usually triggered by respiratory or gastrointestinal infections, it is known that other factors, although infrequent, can act as precipitants, such as vaccination and even cases of trauma. Regarding trauma, the pathogenesis is thought to involve various biochemical cascade reactions, including the release of catecholamines and cytokines, increased permeability of the blood-brain barrier. This allows macrophages to secrete neurotoxic mediators such as tumor necrosis factor alpha and nitric oxide and thus allowing the inflammatory process with damage to the myelin sheath and axon. **Final comments:** In MFS, it is important to carry out a more thorough clinical examination and ask about the patient's pathological history, especially a history of trauma. The patient had no other clinical event besides the trauma that could be related to the condition, and similar cases have already been described in the literature. Despite the favorable prognosis for MFS, with the possibility of self-limiting remission, treatment with IVIG is typically chosen to expedite recovery and minimize complications.

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Traumatic brain injuries in military police: a case series of 11 officers wounded by high-caliber firearms

Raquel Maria Ayres Monteiro[1], Raphaela Coelho Veloso Gomes[1], Uri Adrian Prync Flato[2], Carlos Gilberto Carlotti Júnior[3], Wellingson Silva Paiva[3], Mário Vicente Campos Guimarães[3]

[1]Faculdade Israelita de Ciências da Saúde Albert Einstein, São Paulo SP, Brazil.
[2]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.
[3]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Raquel Maria Ayres Monteiro; Email: raquelmamonteiro@gmail.com.

Background: Traumatic brain injury (TBI) is a major global health issue, with combat TBI, often resulting from armed conflicts, accounting for 2% of cases. Advances in technology and military tactics have altered injury patterns, increasing both fatality and morbidity. Although generally mild, up to 25% of combat TBIs lead to chronic impairments like cognitive, behavioral, and sensory changes, known as post-concussion syndrome. Firearm-related TBIs tend to cause more severe sequelae than blunt traumas. In Brazil military police officers face a higher risk of firearm fatalities than civilians, predominantly outside work hours. These incidents mainly affect men and Black

individuals (97% and 67%, respectively), highlighting the professional risks and the physical, psychological, and social consequences of their duties. **Objective:** This case series analyzes the profile of 11 military police officers from São Paulo State, aiming to compare findings with global and Brazilian statistics. **Methods:** This study included 11 military police officers from the State of São Paulo, who survived gunshot wounds to the skull and were treated between 2014 and 2017. Variables analyzed included sex, age, number of projectiles, entry wound, need for decompressive craniectomy, and cranioplasty, with a descriptive analysis conducted using JASP software. **Results:** Patients in this case series, aged 23 to 35 years with an average age of 26.9, included 10 men and one woman among 11 police officers. The most common injury was a frontotemporal entry wound (54.54%), followed by parietal wounds (18.18%), with occipito-mastoid, frontotemporoparietal, and temporoparietal entry wounds each accounting for 9.09%. Decompressive craniectomy was performed in 9 cases (81.8%), and cranioplasty in 8 (72.7%). The injuries were primarily caused by two types of projectiles, 5.56 and 7.62, with the latter responsible for 90% of the injuries; notably, one patient was struck by projectiles of two different calibers. Neurologically, 5 patients (45.4%) exhibited left hemiparesis, 4 (36.3%) had right hemiparesis–with half of these also presenting with aphasia. Additionally, one case involved diparesis accompanied by aphasia, and another presented with vertigo and right-sided anacusis. **Conclusion:** This study of traumatic brain injuries in São Paulo's military police underscores the severe implications of high-caliber firearm wounds. Extensive surgical interventions were necessary due to the injury severity. The officers experienced significant chronic impairments, affecting their quality of life and necessitating a multidisciplinary medical approach. The findings call for more research and improved safety measures for law enforcement professionals.

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Traumatic brain injury and correlations with psychiatric disorders: a systematic review

Rafaela Coelho Rachid[1], Lizen Clare André Moreira[2], Gustavo Joji Yoshida[2], Fernanda Beatriz Venturi Araújo[1], Augusto Neuberger[1], Anita Ferreira Das Neves[1]

[1]Universidade Federal do Rio Grande, Rio Grande RS, Brazil.
[2]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Rafaela Coelho Rachid; Email: rafacoelhorachid@hotmail.com.

Background: Traumatic brain injury (TBI) is a neural injury that affects millions of people each year. While physical and cognitive impairments are extensively studied, psychological effects are relatively understudied. Recent studies indicate a significant relationship between TBI and post-traumatic mental disorders, emphasizing the importance of understanding this connection to guide treatments. **Objective:** To identify the main mental disorders following TBI in patients with no psychiatric history. **Methods:** A systematic review was conducted following the PRISMA protocol across three databases: Scopus, Web of Science, and PubMed. Initially, 1232 articles were found based on the PICO criteria, but 1082 were excluded after title and abstract screening. Of the remaining 150 articles, 97 were included after full-text review, with 17 excluded due to lack of access. **Results:** Symptoms analyzed in TBI patients included anxiety, depression, memory and sleep disturbances, irritability, obsession, suicidal ideation, mood disorders, post-traumatic stress disorder (PTSD), and personality disorders. Assessment tools used included the Structured Clinical Interview for DSM-IV, Pittsburgh Sleep Quality Index, and Mini Mental Health State Examination. Udala M found a negative correlation between anxiety and working memory in TBI patients. Katzin identified mood and personality changes in inmates with TBI (66.9% with personality changes). Maguen S noted a higher tendency for mental disorders, PTSD, and depression in blast-related TBIs. Collen J found specific sleep disorders like obstructive sleep apnea related to blunt traumas, with insomnia prevalent in blast-related traumas. Lange RT reported irritability in 30-40% of mild TBIs and 66% in severe cases. Choi Y identified depression as the most common disorder, with an incidence of 34.6 per 1000 persons per year compared to 21.42 in the control group. Yeh TC found a doubled risk of developing disorders in TBI patients compared to non-TBI individuals. Sivolap YP highlighted increased substance abuse, and Talaslahti T noted a 1.9 to 2.5 times higher suicidal tendency in TBI patients. Some studies suggest symptoms worsen over time and with TBI severity, indicating a risk for cognitive decline but not necessarily neurodegenerative diseases. **Conclusion:** TBI is associated with psychiatric disorders. It is evident that a more accurate assessment of individuals' neuropsychiatric state occurs when there is a correlation between neuropsychological domains, neurobehavioral questionnaires, and self-perception. The trauma mechanism was a determining factor in the development of specific neuropsychiatric disorders. Depression, anxiety, sleep disturbances, and suicidal behavior were the most frequent disorders. However, more studies with general populations are needed, as many of those included only inmates or military personnel.

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Treatment for 5q spinal muscular atrophy with risdiplam: safety and preliminary efficacy in a Brazilian cohort

Raquel Diógenes Alencar Sindeaux[1], Rodrigo de Holanda Mendonça[1], Graziela Jorge Polido[1], Eduardo Vital de Carvalho[1], Leandro Augusto de Almeida[1], Edmar Zanoteli[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Departamento de Neurologia, São Paulo SP, Brazil.

Correspondence: Eduardo Vital de Carvalho; Email: duvital1983@gmail.com.

Background: Risdiplam is approved for the treatment of all types 5q spinal muscular atrophy (5q-SMA) from 16 days of age onwards. Although clinical trials have shown improvements or stabilization in motor and respiratory function, real-world data is still scarce. **Objective:** To describe the experience of a neuromuscular specialized center in Brazil in treating 5q-SMA patients with risdiplam and to analyze clinical aspects during a follow-up period up to 32 months (range 3-32; mean 10,95 months). **Methods:** Eighteen 5q-SMA patients have been followed by a multidisciplinary team since the beginning of treatment with risdiplam from November 2021 to May 2024. These patients have been monitored in a specialized public outpatient clinic and a private clinic in São Paulo-Brazil. At each follow-up visit, subjective global impression of the patient or caregiver and adverse treatment events have been collected. Motor scales and respiratory assessments have also been performed when feasible. **Results:** All patients were treatment-naïve, and they started risdiplam between 17 days and 49 years old (mean 18,62 years). Three patients began treatment in a pre-symptomatic phase. One patient was SMA type 1, eleven were SMA type 2 and three were SMA type 3. Fifteen patients (83.3%) reported improvement or stability with risdiplam. Motor scales were regularly applied only to pre-symptomatic or type 1 SMA children, who demonstrated score gains like those described in clinical trials. All pre-symptomatic patients had 2 copies of SMN2 and achieved maximum score on Children's Hospital of Philadelphia Infant Test of Neuromuscular Disorders (CHOP INTEND) at 4 months old. CHOP INTEND score of type 1 SMA patient increased from 29 to 44 within 3 months of treatment. Among the 14 patients with type 2 or type 3 SMA: nine reported improvement, two stability and three worsening in motor function; one also reported improvement in cough strength and another in vocal power. Three patients with type 2 SMA underwent spirometry before and after 6 months of treatment: two showed stabilities in forced vital capacity and one showed an increase of 6%. Six patients reported adverse effects: one had diarrhea, one constipation, one abdominal pain, one anemia, one headache and one hair loss. **Conclusion:** Risdiplam was well tolerated, and no patient changed or discontinued treatment. Furthermore, most patients experienced clinical improvement or stabilization. This real-world data from risdiplam showed results like those from clinical trials.

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Treatment of NEDAMSS syndrome by deep brain stimulation: report of the first successful treatment case in the world

Sophia Keiko Inuzuka[1], Gabriel de Castro Micheli[1], Marcello Reis da Silva[1], Gustavo da Fontoura Galvao[1], Marlos Melo Martins[1], Hanid Fontes Gomes[1], Roberto Santos Pereira[1], Rita de Cássia Leite Fernandes[1]

[1]Universidade Federal do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Sophia Keiko Inuzuka; Email: sophia.keiko@gmail.com.

Case presentation: A 9-y-old girl had a confirmed diagnosis of NEDAMSS ("Neurodevelopmental Disorder with Regression, Abnormal Movements, Loss of Speech, and Seizures"), through genetic testing that revealed the pathogenic variant p.(Val76Cysfs*50). The recently described ultra-rare syndrome is a progressive condition caused by mutations of the intronless gene IRF2BPL ("interferon regulatory factor 2 binding protein like"), mapped to 14q24.3. Its relationship with neural development has not yet been fully established, but its mutation is manifested by neurological regression beginning around 3.5 y of age. There is loss of gait and speech accompanied by epilepsy, dysphagia, dysarthria and dystonia. To date, there is no specific treatment. Our patient was born at 32 w with 1.9 kg. She had continued monitoring due to asthma, low weight and slow cognitive development. At the age of 5, she began to show impairment in gait. The diagnosis of the syndrome was not established at the time. Over the next two years, there was a progressive worsening with mother reports of "involuntary muscle contractions" and pain that led to the use of morphine and cannabidiol in high doses, without adequate relief. At that time, genetic testing diagnosed the mutation responsible for NEDAMSS. Treatment with gabapentin, trihexyphenidyl and clonazepam was unsuccessful and generalized dystonia developed with 80 points on the Fahn-Marsden Scale (BFMDRS), associated with loss of speech and gait. Cranial MRI was unremarkable DBS implantation in the Globus Pallidus Internus (GPi) was done using the Vercise Genus P16 implantable pulse generator. Four weeks after bilateral symmetrical stimulation began, guided by three-dimensional reconstruction and set to 1.5 mA pulse, width of 60 µs and frequency of 130 Hz. After a week of stimulation, significant improvement in clinical picture was observed. She was able to sit up without assistance, cooperated with physiotherapy and medication doses reduced by a third. The BFMDRS score decreased to 24, indicating a positive response to DBS treatment. **Discussion:** The effectiveness of DBS in the treatment of generalized dystonias is widely recognized, evidenced by significant reductions in post-procedure BFMDRS scores. Although the only previous report indicated refractoriness of dystonia in a male patient with NEDAMSS undergoing pallidal DBS, our patient showed a remarkable response to DBS. We remain optimistic that the benefits will sustain and improve with subsequent device schedules. **Final comments:** This case represents a significant milestone by demonstrating, for the first time, a positive response to DBS treatment in the GPi in patients with dystonia.

This suggests a therapeutic potential for patients with NEDAMSS refractory to conventional treatment. Further studies are needed to validate these findings and better understand the mechanisms underlying the response to DBS in this ultra-rare syndrome.

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Treatment of neurodegenerative diseases: comparative study of the spatio-temporal distribution between Brazilian regions in the years 2019 to 2023

Luan Nascimento Pereira de Amorim[1], Dangilla Ribeiro dos Santos[1], Bruno Kenji Hosoda Mineshita[1], Fernando Lourenzo Rocha Vianna Oliveira[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Luan Nascimento Pereira de Amorim; Email: luanamorim010@gmail.com.

Background: Neurodegenerative diseases are incurable and debilitating conditions, such as Alzheimer's disease and Parkinson's disease. They result from the progressive degeneration of neurons in structures in various anatomical regions of the nervous system and reflect gradual functional alterations, leading to movement and cognitive deficits. Specialized healthcare and expensive treatments are associated with high treatment costs and a high demand for hospitalizations. **Objective:** To characterize the spatio-temporal distribution of treatment for neurodegenerative diseases in Brazil between 2019 and 2023. **Methods:** This is a quantitative, time-series ecological study based on data from the Hospital Information System of the Unified Health System on the treatment of neurodegenerative diseases in Brazil between January 2019 and December 2023. The following variables were used: number of hospitalizations, cost of treatment, average length of stay, character of care, mortality rate and deaths. **Results:** In the period from 2019 to 2023, 25,446 hospitalizations were recorded with an annual average of 5.08±0.97 procedures, with the most in the Southeast region, with 8,849 hospitalizations (34.77%), followed by the South with 8,643 (33.96%), and then the Northeast with 5,536 (21.75%). With regard to the type of care, 88% (N=22,393) were urgent and 12% (N=3,051) elective. During hospitalization, the average length of stay was 13.1 days between the regions, in which the North and Midwest were the only ones to reduce the average from 2019 to 2023, with the Northeast showing an increase of 40.43%, from 9.4 to 13.2 days. Regarding the cost of treatment, the average value per hospitalization was R$1,408.64, the lowest in 2019 (R$1,190.46) and the highest in 2022 (R$1,500.40), an increase of 26.04% between these years and a drop of 4.34% in 2023. The North region had the highest value among the regions (R$2,552.90). Regarding the evolution of the condition, 2,484 deaths were recorded, denoting 9.76% of all hospitalizations, with an annual average of 496.8±121.7, with the Southeast once again in first place in this regard (N=967/38.93%). The Northeast had the highest mortality rate (18.23%), above the national average of 9.76%. **Conclusion:** There has been an increase in the treatment of neurodegenerative diseases, especially emergencies, in the Southeast and South. Expenditure increased over 5 years, especially in the North, possibly due to the impact of the pandemic on labor and input costs. In the Midwest, the diagnosis of death has improved, reflected in the higher annual variation in the mortality rate. This highlights the importance of effective management and preventive strategies to reduce cases of these pathologies.

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Treatment of Parkinson's disease in Brazil's public heath system: is there still room for dopaminergic agonists?

André Borges Ferreira Gomes[1], Raynrich Kevin Assis Lima[2], Isabela Cacau Sousa Santos[2], Carolina Helen Soares de Macêdo[2], Flavia de Paiva Santos Rolim[2], Antonia Rosivalda Teixeira Marinho[1], Vivia Linhares Mesquita[1], Manoel Albano Amora Neto[1], Amanda Araujo Braga[1], Fernanda Martins Maia Carvalho[2]

[1]Hospital Geral de Fortaleza, Fortaleza CE, Brazil. [2]Universidade de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Andre Borges Ferreira Gomes; Email: andrebfg@gmail.com.

Background: Parkinson's disease (PD) is a heterogeneous condition in which no agents have been shown to have unequivocal evidence of disease-modifying effects so far. Treatment for motor symptoms involves pharmacologic approaches (typically with levodopa preparations), particularly in regions where access to other therapies are restricted or not available. **Objective:** To determine the clinical and epidemiological profile of patients with PD in a movement disorder outpatient clinic of a public hospital in the state of Ceará. **Methods:** A descriptive, observational, cross-sectional study, with a quantitative approach, was carried out in 2023. A total of 636 patients were screened and were evaluated using specific questionnaire and review of medical records. In this first stage, patients meeting the Movement Disorder Society criteria for the diagnosis of PD were included. A descriptive statistical analysis of epidemiological, clinical and therapeutic data of the patients was obtained. **Results:** A total of 226 PD patients were included. Most patients were male (62%), presenting a mean age was 65 years. The vast majority (92%) were using some form of levodopa, with a mean dosage of 710mg per day. The second most commonly used medication among patients was pramipexole (61.9%), followed by rasagiline (50.8%), amantadine (41.1%) and entacapone (31.4%). The mean duration of the disease was 13 years and the mean result of the Hoehn and Yahr was 3. Dyskinesia and motor fluctuations were referred by 50.4% and 79.6% of the patients, respectively. When any psychiatric symptom was investigated, it was evidenced in 54.8% of the patients. **Conclusion:** Although levodopa therapy is the most effective and recommended therapy for PD, pramipexol is maintained in most patients in this tertiary public health care clinic in the Northeast of Brazil, in a higher proportion than rasagiline and entacapone. Even though dopaminergic agents are progressively becoming less prescribed worldwide, the limited access to surgical and infusion therapies for advanced PD may be a reason for maintaining such therapies. Despite the side effects described in the literature and restriction of use in some regions of the world, more studies are needed to understand the role of dopamine agonists in the treatment of PD in low-income regions.

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Treatment of Parkinson's disease in Brazil from 2019 to 2023: a comparative analysis between regions

Luan Nascimento Pereira de Amorim[1], Fernando Lourenzo Rocha Vianna Oliveira[1], Dangilla Ribeiro dos Santos[1], Bruno Kenji Hosoda Mineshita[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.

Correspondence: Luan Nascimento Pereira de Amorim; Email: luanamorim010@gmail.com.

Background: Parkinson's disease is a neurodegenerative syndrome characterized by the involvement mainly of the neurons of the substantia nigra pars compacta, which can lead to bradykinesia, resting tremors and rigidity. The WHO estimates that around 1% of the world's population over the age of 65 has the disease, which means that as the population ages, the number of cases is expected to rise. Therefore, continued treatment is essential for the patient's survival, quality of life and well-being. **Objective:** To analyze the distribution of Parkinson's disease treatment in Brazil from 2019 to 2023. **Methods:** This is a descriptive, cross-sectional and quantitative study, based on data from the Hospital Information System tabulated by the Department of Informatics of the Unified Health System (SIH/DATASUS), recorded from 2019 to 2023, in Brazil. Data was collected for ICD-10: Parkinson's Disease. The variables used for analysis were the average value per hospital admission authorization, approved hospitalizations, average length of stay and mortality rate, which, due to the lack of a population estimate for the year 2023, absolute data were used. Microsoft Excel 2021 software was used for the calculations and analysis. **Results:** During the period analyzed from 2019 to 2023, a total of 2640 hospital care authorizations (AIH) were found, with an average of 528 AIH per year, with the Region with the highest number being the Southeast with 1016 (38.4%), followed by the South, 870 (32.9%) the North appears in last place with 168 (6.3%), As for the average value per AIH, The regions with the highest values were the Midwest (R$1017.9) and the Northeast (R$804.61). When analyzing the average length of stay variable, it can be seen that the two regions mentioned have the longest with 9.9 and 10.1 days respectively, while the value per AIH and the average time in the North are among the lowest (R$796.93 and 8 days). With regard to the annual mortality rate, the North (10.12) leads this variable and exceeds the national rate (7.8), while the Midwest (6.04) has the lowest rate. **Conclusion:** It can therefore be seen that the lack of greater investment in the North Region in the treatment of Parkinson's Disease, with fewer AIHs approved, the lower average length of stay and average values per AIH contribute to it having the highest mortality rate in the country, but the opposite is observed in the Midwest Region, which has a lower rate than the country. Therefore, attention to the dichotomies between regions can help to understand the dynamics of negative or positive outcomes and the relationship with the promotion of treatment for the disease.

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Treatment of Parkinson's disease with botulinum toxin: a systematic literature review

Flávia Brady de Carvalho Gauziski Figuerêdo[1], Bruna Meschesi Dantas[1], Enzo Pelajo Mosca[1], Fabrícia Elysio Linhares dos Santos[1], Gabrielle Brandão Vasconcelos[1], Giovana Mattos Delgado[1], Maria Carolina Luiz Rodrigues Meireles[1], Pedro Vieira Ahouagi Cunha[1], Reynaldo Yuji Souza Tanaka[1], Natalia Conti Lessa Pereira[1]

[1]Faculdades Souza Marques, Rio de Janeiro RJ, Brazil.

Correspondence: Flávia Brady de Carvalho Gauziski Figueredo; Email: flaviabrady@outlook.com.

Background: Botulinum toxin (BoNT) is considered a promising alternative for treating various symptoms of Parkinson's disease (PD). This neurodegenerative condition, in addition to its characteristic motor symptoms such as tremor, bradykinesia, and rigidity, also includes non-motor symptoms that significantly affect patients' life quality. Among these, sialorrhea often occurs, causing difficulties such as infections, swallowing problems, and social impact. The treatment of sialorrhea with BoNT injections into the salivary glands, often guided by ultrasonography, has been studied and suggested as an effective and safe approach. Additionally, BoNT has shown utility in managing spasticity, which is an increase in muscle tone due to central nervous system impairment, offering a therapeutic alternative for various disabling aspects of PD. **Objective:** Systematic review of methodological practices in the treatment of PD through the use of botulinum toxin in the literature, taking into account recent work and the analysis of changes in the practice of this treatment. **Methods:** Systematic review of articles from 2012 to 2023 selected from Ministério da Saúde, PubMed and Scielo database with the following keywords: "Parkinson's Disease", "botulinum toxin" and "neurodegenerative".

Each database was screened independently by all the authors of this review. **Results:** Five articles were included in the final review, all of which identified BoNT injections as a promising treatment for symptoms linked to PD, including sialorrhea, dystonia and tremor. Two of the articles reinforced the positive effects on the reduction of sialorrhea, mainly in volume of salivation, in PD patients regarding the weekly use of local toxin injections for up to 96 weeks. However, three articles emphasized the need of additional studies to fully elucidate BoNT's role in managing various motor disorders correlated to PD, such as at rest tremors and dystonias. Furthermore, four articles reported adverse effects associated with the BoNT injections, such as muscle weakness, fatigue, dry mouth, neck pain, dysphagia, diarrhea, worsened gait, and dysarthria. These adverse effects were more prevalent in the use of BoNT for motor disorders, which were described as having a more challenging approach in the administration of the drug due to the number of muscles and articulations in arms and legs of the patients that needed to be properly medicated. **Conclusion:** BoNT injections are a promising therapeutic alternative for the symptomatic treatment of PD, due to the subjective and objective improvement in the condition and life quality of these patients, especially those with sialorrhea, despite the possible side effects. However, further studies are still needed to prove the safety and efficacy of this potential therapeutic intervention.

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Treatment of perioral dystonia with botulinum toxin in case of Meige's syndrome

Thais Lins Gemir[1], Silvana Sobreira Santos[1], Bárbara Matos Almeida Queiroz[1], Alberico Albanês Oliveira Bernardo[1], Natalia Saraiva Ramos e Cruz[1], Denise Carvalho de Albuquerque[1], Mauricea Novaes Costa Pereira[1]

[1]Rede D'Or.

Correspondence: Thaís Lins Gemir; Email: tatalg_30@hotmail.com.

Case presentation: A 37-year-old woman with a history of recurrent facial paralysis for 2 years and perioral dystonia . She also had difficulty opening her eye several times a day. The patient was hospitalized for investigation and electroneuromyography compatible with bifacial dystonia and blepharospasm component in the left eye. Normal skull MRI. She started epileptic seizures. She didn't show dystonia and can be associated with complex movement of lower fany improvement with diazepam, haloperidol, levodopa or baclofen, and the application of botulinum toxin was indicated with excellent results. **Discussion:** "Meige's syndrome" is a type of cranial dystonia characterized by blepharospasm and oromandibularacial muscles, mouth, jaw, tongue, pharyngeal and cervical muscles. Frequently, blepharospasm is the earliest clinical manifestation, which spreads over a period of time to involve other cranial and extra-cranial muscles. Common characteristics of this syndrome are well known, but their variety is wide. In the majority of the patients Meige's syndrome is primary or idiopathic, where the cause of spasm is not known, however secondary cases can occur following prolonged use of neuroleptics or secondary to underlying brain disorders. Most of the patients are successfully treated with injection of botulinum toxin, however deep brain stimulation has emerged as a good therapeutic option in intractable patients. The result of the intramuscular botulinum toxin A injections was characterized by the patients as "much improved"; correspondingly, dystonia was significantly reduced in visual analog scale scores, on examiner-based assessments, and in recordings of EMG. **Final comments:** Meige's syndrome is a type of cranial dystonia characterized by a combination of blepharospasm and orofacial-cervical dystonia. Most of the cases are primary or idiopathic, however secondary cases also occur usually due to chronic neuroleptics exposure or focal brain lesions. Neurophysiological studies have demonstrated abnormal excitability of the sensorimotor cortex and brainstem inter-neuronal pathways. Incapacitating perioral dystonia in Meige's syndrome may be safely controlled by recurrent EMG-guided botulinum toxin A injections in the orbicularis oris muscle, in combination with general medication

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Treatment of trigeminal autonomic cephalalgia: a case report

Gabriella Mara Arcie[1], Eduarda Menin da Silva[2], Valentina Lima Cartaxo da Silva[2], Victor Frandoloso[3]

[1]Faculdade Evangélica Mackenzie do Paraná, Curitiba PR, Brazil.
[2]Universidade Positivo, Curitiba PR, Brazil.
[3]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.

Correspondence: Eduarda Menin da Silva; Email: eduardamenindasilva@gmail.com.

Case presentation: A 69-year-old male patient was diagnosed with trigeminal autonomic cephalalgia, SUNCT syndrome, in 2021. The patient presented with intense, intermittent pain in the left periorbital region, lasting for seconds, associated with conjunctival hyperemia, ptosis, and ipsilateral tearing. He reported that the pain was triggered by daily activities such as talking, drinking water, and brushing his teeth. His medical history included hypertension and dyslipidemia. He was taking Carbamazepine 200 mg; Pregabalin 75 mg; Micardis; and Rosuvastatin 10 mg. Neurological examination was unremarkable. In 2021, the patient underwent conventional and pulsed radiofrequency procedures of the left sphenopalatine ganglion. **Discussion:** SUNCT syndrome is a trigeminal autonomic cephalalgia characterized by short episodes of unilateral pain associated with autonomic symptoms such as conjunctival hyperemia and tearing. The patient in this case presents with typical symptoms of the syndrome, including intense paroxysmal pain in the left periorbital region and ipsilateral autonomic symptoms. SUNCT is a challenging condition to treat, often requiring a multifaceted approach. Initial pharmacological treatment with anticonvulsants aims to reduce the frequency and intensity of pain attacks, although efficacy may be limited. Conventional radiofrequency (CRF) creates thermal lesions on target nerves, interrupting pain signal transmission and providing long-lasting pain relief. Pulsed radiofrequency (PRF), on the other hand, uses intermittent energy pulses, modulating neural function without causing significant thermal lesions. In this patient's case, the combination of CRF and PRF performed in 2021 on the sphenopalatine ganglion proved to be an appropriate approach. PRF, in particular, has a favorable safety profile and can be repeated if necessary. **Final comments:** Managing the syndrome can be challenging due to the intensity and frequency of pain episodes. Therefore, SUNCT treatment must be personalized, taking into account individual response to treatments and safety profile. Combining pharmacological treatments with radiofrequency interventions, such as CRF and PRF, can offer significant relief, improving patients' quality of life.

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Treatment of uncontrolled epileptic seizures in Brazil: an epidemiological analysis over the last 5 years

Matheus Prado dos Santos[1], Rafael Hendrick Freitas de Paula[1], Luan Nascimento Pereira de Amorim[1], Fernando Silva Galvão[2], Daniel Lucas Araujo Ramos[2], Paola Carolina Santos Foinkinos[1]

[1]Universidade do Estado do Pará, Belém PA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Matheus Prado dos Santos; Email: imatheusprado@gmail.com.

Background: An epileptic seizure is caused by an abnormal electrical discharge that occurs in the gray matter of the brain, disrupting brain function. Typically, an epileptic seizure results in an altered level of consciousness, abnormal sensations, focal involuntary movements, or motor crises. Understanding the efficacy of treatment and its specific action concerning the epidemiological variations among the regions of Brazil is crucial for formulating strategies to control epileptic seizures in our country. **Objective:** To analyze the epidemiological profile of uncontrolled epileptic seizures in Brazil from 2019 to 2023. **Methods:** This is an ecological, cross-sectional, descriptive, and analytical study with a quantitative approach, conducted by collecting secondary data from the Department of Informatics of the Unified Health System (DATASUS) from 2019 to 2023. The following variables were analyzed: number of hospital admissions, average cost per admission, average length of stay, mortality rate, and the nature of the care provided for approved admissions. **Results:** Between 2019 and 2023, there were 228,181 hospitalizations for uncontrolled epileptic seizures in Brazil, with an approximate annual average of 45,636 hospitalizations. According to region, during this period there were 14,730 hospitalizations in the North (6.4%), 61,208 in the Northeast (26.8%), 94,777 in the Southeast (41.5%), 37,195 in the South (16.3%), and 20,271 in the Midwest (8.8%). As for the number of hospitalizations per year, there were 44,012 in 2019 (19.2%), 40,073 in 2020 (17.5%), 44,156 in 2021 (19.3%), 51,700 in 2022 (22.6%), and 48,240 in 2023 (21.1%). Regarding the cost per hospitalization, the average was 565.75 reais in the North, 784.71 reais in the Northeast, 988.05 reais in the Southeast, 903.4 reais in the South, and 806.6 reais in the Midwest. The mortality rate was 3.02 in the North, 3.24 in the Northeast, 3.52 in the Southeast, 2.09 in the South, and 2.19 in the Midwest. Concerning the type of care, 8,600 hospitalizations were elective (3.7%) and 219,576 were emergencies (96.3%). Additionally, the average length of stay per year was 5.8 days in the period studied: 5.3 days in the North, 6.1 days in the Northeast, 5.9 days in the Southeast, 5.3 days in the South, and 5.1 days in the Midwest. **Conclusion:** The Southeast region had the highest number of hospitalizations, the highest average cost per hospitalization, the highest mortality rate, and the highest absolute number of deaths in the period studied. Therefore, there is a need to improve health services in the Southeast of Brazil, which, despite having the highest cost of hospitalization, has high mortality rates. Finally, there is a need for further studies to understand the factors influencing the current scenario in the Southeast and Northeast regions of the country.

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Trends in door-to-needle and door-to-puncture times in ischemic stroke: a quality improvement study

Caio Pinheiro Santos Souza[1], Murilo Santos de Souza[2], Luiz Eduardo Fonteles Ritt[2], Eduardo Sahade Darzé[2], Alexandre Drayton[2], Gabriel Xavier[2], Humberto Castro-Lima[2], Matheus do Rosário[2], Barbara Costa[2]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.
[2]Instituto D'or de Pesquisa e Ensino, Hospital Cardio Pulmonar, Salvador BA, Brazil.

Correspondence: Caio Pinheiro Santos Souza; Email: caiopsouza21.2@bahiana.edu.br.

Background: Stroke will affect one in four adults over their lifetime, causing the destruction of billions of synapses every minute. Furthermore, delays from the onset

of symptoms to reperfusion therapy progressively reduce its effectiveness. The protocol implemented in our institution stipulated: 1) Early identification of patients with signs of stroke and direct referral to CT by nursing staff; 2) Early assessment by a neurologist, with an expected interval of <20 minutes between contact and the arrival of the on-call neurologist; 3) Stroke training and simulations; 4) Real-time feedback to the emergency room team. **Objective:** Therefore, our single-center observational study aimed to evaluate the effect of a treatment protocol designed to reduce delays in treatment. **Methods:** This retrospective cohort study included all patients eligible for Chemical Thrombolysis or Mechanical Thrombectomy from 2011 to 2023. The impacts of the protocol were evaluated by calculating the absolute frequencies and the annual median of door-to-needle time (DNT) and door-to-puncture time (DPT), with interquartile range. Given that the data did not follow a normal distribution, the Kruskal-Wallis test was used to analyze differences between times. Temporal trends were assessed using the Mann-Kendall test. **Results:** Between January 2011 and December 2023, 1014 patients were admitted under the stroke protocol. The median age was 74 years (63-83) and the NIHSS was 3 (1-9). Reperfusion therapy was offered to 174 patients; in this group, the median age was 74 years (61-84), and the NIHSS was 8.5 (4-18). We observed a significant increase in the thrombolysis rate from 7.9% to 15% (P<0.001) and in the thrombectomy rate from 1.1% to 7.6% (P<0.001) when comparing the periods 2012-2014 and 2015-2023. The DNT during the studied years showed a statistically significant difference (P=0.008), with a reduction in this time from 89 minutes (84-116) in 2012 to 31 minutes (18-51) in 2015, and later to 28 minutes (17-43) in 2019. When analyzing the pre-pandemic period, a moderate to strong statistically significant decreasing temporal trend was identified in median values over time (tau = -0.64, P =0.02). Despite the trend disruption due to the COVID-19 pandemic, the DNT did not exceed 45 minutes during the period, ultimately reaching 34 minutes (18-48) in 2023. There was no difference in mortality (P=0.89) or the rate of hemorrhagic transformation (P=0.33) between the years in patients undergoing reperfusion therapy. **Conclusion:** During the protocol implementation period, which included validated strategies, there was a considerable reduction in treatment delays and a significant downward trend in times. An increase in reperfusion rates was also observed, without an increase in mortality or complications. Thus, the study demonstrates that the implementation of robust protocols in healthcare institutions can positively impact stroke care.

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Trigeminal neuralgia secondary to basilar invagination responsive to botulinum toxin type a: a case report

João Vitor Andrade Fernandes[1], Tiago César Costa da Silva[1], Camilla Vanessa Araújo Soares[1], Gilmar Leite Pessoa Filho[2], Ana Luisa Castelo Branco Gomes[2], Davi Telécio Firmino[1], Raissa N. L. F. Leite[3], Adriana M. T. Nepomuceno[4], Maurus M. de A. Holanda[5], Alex T. Meira[5]

[1]Universidade Federal da Paraíba, João Pessoa PB, Brazil.
[2]Hospital Metropolitano Dom José Maria Pires, João Pessoa PB, Brazil.
[3]Nova Diagnóstica por Imagem, João Pessoa PB, Brazil.
[4]Universidade Federal da Paraíba, Programa de Mestrado Profissional em Gerontologia, João Pessoa PB, Brazil.
[5]Universidade Federal da Paraíba, Departamento de Medicina Interna, Serviço de Neurologia, João Pessoa PB, Brazil.

Correspondence: Tiago César Costa da Silva; Email: tiagocesarcs@hotmail.com.

Case presentation: A 34-year-old male patient presented with paroxysmal shock-like pain in the right V2 and V3 branches, persisting for approximately 12 years. Neuroimaging revealed basilar invagination and neurovascular conflict of the trigeminal nerve. The patient was refractory to treatments with duloxetine (120 mg/day), carbamazepine (1,200 mg/day), valproate (1,500 mg/day), lamotrigine (200 mg/day), and cannabidiol. Surgical intervention was deemed inappropriate. Physical examination revealed a short neck, allodynia, hyperalgesia in the right V2 and V3 branches, and signs of poor self-care. Treatment was initiated with type A botulinum toxin (Botox®) (BoNT/A), 30 units distributed subcutaneously in the V2 and V3 territories, with 2 cm between application points. An 80% improvement in pain (visual scale from 10/10 to 2/10) and frequency (from constant to few attacks per day) was observed after the first dose. Quality of life increased from 0 to 10 (assessed by the Quality Of Life Scale from the American Chronic Pain Association), sleep and mood improved (according to the Beck Depression Inventory from 36 to 2), remitting his suicidal ideation and resumption of his employment. Results were observed from the 3rd day, without adverse effects, and remained consistent over successive applications. **Discussion:** Resistance or adverse effects to conventional therapies often necessitate alternatives when treating patients with trigeminal neuralgia (TN). In this case, optimized pharmacotherapy did not control the pain, highlighting the refractory nature of some TN cases. BoNT/A demonstrated excellent efficacy, substantially alleviating pain and improving the patient's well-being, quality of life, and mood, emphasizing the importance of non-conventional approaches when standard treatments fail. Recent studies indicate that BoNT/A reduces pain in more than 50% of patients with TN at 1- and 3-months follow-up. Surgical interventions can lead to severe and intractable complications. The mechanisms of pain modulation by BoNT/A include inhibition of neurotransmitters (substance P, glutamate, calcitonin gene-related peptide), interference with synaptic vesicle fusion, modulation of neurons in the dorsal root ganglia and spinal cord, and indirect effects on the central nervous system. Interaction with glial cells suggests an additional analgesic mechanism. **Final comments:** This case underscores the efficacy of BoNT/A in managing complex and refractory pain syndromes associated with trigeminal neuralgia secondary to structural anomalies, offering a promising therapeutic avenue for patients resistant to conventional treatments.

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Trisomy 14 and it's manifestations: a case report

Gabrielle Miranda Magalhães Pinto[1], Lourdes Maria Silva de Assis[1], Isabelle Diniz Melo[1], Maria de Fátima Miranda de Oliveira[2], José Lucivan Miranda[3], Fabiane Elpídio de Sá Pinheiro[1], João Joaquim Freitas do Amaral[1], Norma Martins de Menezes Morais[3]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Universidade de Fortaleza, Fortaleza CE, Brazil.
[3]Núcleo de Tratamento e Estimulação Precoce, Fortaleza CE, Brazil.

Correspondence: Gabrielle Miranda Magalhães Pinto; Email: gabriellemmp1@hotmail.com.

Case presentation: A 2 year old female patient, born at full term via vaginal delivery (APGAR 9/10). During the gestation, the mother, besides taking fluoxetine and diazepam, presented with urinary tract infection during the first trimester and hypertension during the third. At 1 month of age, the patient suffered a traumatic brain injury (TBI), undergoing a cranial tomography (CCT) that revealed an occipital aligned fracture on the left and subgaleal hematoma. She had a positive clinical recovery. At 5 months, it was detected in a new CCT corporal assimetry with hemihypertrophy on the right side and cervical inclination on the left. At 10 months, besides having a hypoplasic left kidney detected by ultrasound, the patient was diagnosed with thyroglossal duct cyst and Waldeyer's ring hypertrophy after b3ing submitted to a craniocervical magnetic ressonance. The Karyotype revealed trysomy on gene 14 with low grade mosaicism (2% of cells). At 16 months, she exhibited hypocromic linear macular lesions throughout the whole body, apendicular hypotonia, difficulty with chewing, shuddering attacks, aggressive behavior and motor stereotypes. At 22 months, she showed dysfunctional play, hearing hypersensitivity and difficulty to communicate. At 2 years, she began to walk without support, while continuing with significant delay in speech, hyperfixations, food selectivity, agitated sleep, difficulty to comprehend simple commands and to sustain attention, irritated behavior. Currently, the patient persists with developmental delay (DD), but has been improving non-verbal communication, attention and social interactions. **Discussion:** This case describes manifestations related to Trisomy 14 in mosaic, a rare chromossomic disorder caused by anomalous cell division on a certain amount of cells, being incompatible with life in most cases. This genetic condition predisposed the presence of body assimetry, genitourinary abnormalities, pigmented skin lesions, intelectual disability, hypotonia and DD, symptoms that were detected on the presented patient. It is also relevant that the DD secondary to this condition is also supported by the TBI suffered by the patient and the gestational complications, that may have altered intrauterine environment and, as a result, this child's neurodevelopment. **Final comments:** In front of the complications that carriers of this Trisomy face, it is necessary for this chromossomal disorder to be more studied by the scientific community and detected early, once this alteration substancially affects the neurodevelopment. Furthermore, it is important for the individuals that carry this Trisomy to have access to a multidisciplinary care, since it is a genetic condition that affects multiple systems, with the goal of improving the patient's and their family's quality of life.

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TTN gene and proximal myopathy: a case report

Ivo Gabriel Ribeiro Carvalho[1], Guilherme Marques Nunes Mamede[1], Fabio Victor Vieira Rocha[1], Renann Nunes Pirola[1], Daniel José Siqueira Tosi[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Ivo Gabriel Ribeiro Carvalho; Email: ivo.carvalho@edu.ufes.br.

Case presentation: A 44-year-old female with tremors during intense exercise and difficulty walking since childhood. She has been using a wheelchair since the age of 40 following hospitalization for dengue fever. The patient reports inability to fully extend her arms, but without distal motor deficits. Neurological examination revealed symmetric proximal tetraparesis with absent reflexes, elbow contractures limiting extension, preserved facial mimicry, and speech. Electroneuromyography showed neurophysiological findings suggestive of chronic myopathy affecting all four limbs. Muscle biopsy displayed dystrophic pattern and nuclear centralization. Genetic testing conducted in 2023 revealed a homozygous mutation in the titin gene, indicating suspicion of TTN-related myopathy. **Discussion:** Titin, encoded by the TTN gene, is a giant sarcomeric protein that maintains myofibrillar structure, positioning filaments within the sarcomere and providing elasticity and stability during muscle contraction. Mutations in the TTN gene can cause cardiac and muscular disorders, including limb-girdle muscular dystrophy type 2J (LGMD2J). This autosomal recessive disorder typically begins in adulthood or childhood with progressive weakness and proximal muscle atrophy,

predominantly affecting the shoulder and pelvic girdles. Cardiac involvement is common and can significantly impact life expectancy. Physical examination in this case revealed proximal tetraparesis, areflexia, distal muscle atrophy in the lower limbs, and preservation of the extensor digitorum brevis (EDB). In titinopathy, patients may exhibit atrophy of the anterior compartment of the leg (tibialis anterior) while preserving EDB function, distinguishing it from neuropathic patterns. Genetic testing identified a homozygous mutation in the TTN gene (2q31.2), resulting in truncated titin isoforms in skeletal and cardiac muscles. Electroneuromyography demonstrated a myopathic pattern with loss of functional fibers in all limbs. Muscle biopsy findings showed dystrophic pattern with nuclear centralization. Regular cardiac monitoring is crucial to prevent morbidity and sudden death due to the high frequency of cardiomyopathy and arrhythmias associated with TTN mutations. **Final comments:** Unlike neuropathies, titinopathy is characterized by distal muscle hypotrophy with preserved EDB function, underscoring the importance of evaluating the pattern of distal muscle involvement. The disease exhibits a wide age range and phenotypic diversity, posing diagnostic challenges. This case highlights the importance of detailed clinical evaluation, muscle biopsy, and genetic analysis for accurate diagnosis.

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Tumefactive demyelination: atypical presentation of multiple sclerosis?

Bruno Paulo Teles Chaves[1], Vanessa Cristina Fragoso Cassiano Alencar[1], Marcilio Oliveira Filho[1], Nathalia Saraiva Ramos e Cruz[1], Mauricéa Novaes Costa Pereira[1], Lidiane Alves de Macedo Souza[1], Maria Iris Moraes Machado[1], Maria Lucia Brito Ferreira[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Bruno Paulo Teles Chaves; Email: brunopaulo.c@gmail.com.

Background: Atypical demyelinations exhibit pathological characteristics similar to those observed in multiple sclerosis (MS), although their clinical manifestations and radiological patterns differ from those of classic MS. The relationship between MS and its demyelination variants is still unclear, making it difficult to identify patients at risk of developing MS. Tumefactive Demyelination associated with MS is a rare and understudied variant due to the lack of large patient cohorts. The estimated incidence is 1-2 per 1000 MS cases, although other authors suggest a prevalence of 1.4 to 8%. The most common clinical manifestations include hemiparesis or hemiplegia, and headache, aphasia, cognitive, visual, and sensory disturbances, as well as epilepsy, may also be identified. Several important aspects stand out in this aggressive form of demyelination, characterized by tumor-like lesions that are equal to or greater than 2 cm in size. These lesions can occur as single or multiple, and a biopsy may be indicated if the imaging is inconclusive. These lesions may be present in patients with diagnosed MS in 30% of cases or may present as the first episode. **Objective:** To analyze the cases attended with tumefactive demyelination, following up on these cases and subsequently defining the diagnosis of MS. **Methods:** Patients included in the database of the referral service for MS patients care between 1997 and 2019 at the Hospital da Restauração (Recife, Pernambuco) were analyzed. The inclusion criterion was the presence of clinical and imaging findings compatible with tumefactive lesions and the possibility of a final MS diagnosis. **Results:** Seven cases were analyzed, including six female and one male patient, with ages ranging from 11 to 42 years. The predominant signs and symptoms were headache, low visual acuity, motor deficit, seizures, and altered sensorium. MRI revealed multiple pseudotumoral lesions. Three patients underwent biopsy due to suspected primary brain tumor. All patients received pulse therapy with methylprednisolone (MTP), one underwent plasmapheresis (PLEX), three used beta-interferon, one used glatiramer acetate (GA), and two used natalizumab as a second option. The EDSS ranged from 0 to 4 at the onset and from 0 to 7 at the last evaluation, with one death recorded. **Conclusion:** Variants of MS have demonstrated tumefactive lesions and distinct immunological signatures. Tumefactive demyelinating lesions (TDLs) can also occur in patients with other atypical demyelinating syndromes, such as ADEM, NMOSD seropositive for AQ4 IgG or seronegative, as well as MOGAD and other neuroinflammatory diseases (neurosarcoidosis and Behçet's disease), posing a constant challenge, especially in these cases.

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Tumor severity of diffuse gliomas and immunohistochemical markers (S100, GFAP, ATRX, IDH1, synaptophysin, P53): a retrospective study from a tertiary center

Guilherme Nobre Nogueira[1], Pedro Yago Lima de Mesquita[1], Osvaldo Mariano Viana Neto[1], Cauan Miranda Mateus[1], Iasmin Maria Rodrigues Saldanha[1], Vivian Helen Alves Bezerra[1], Lourdes Maria Silva de Assis[1], Maria Jânia Teixeira[1], Gunter Gerson[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Guilherme Nobre Nogueira; Email: guiermenobre@gmail.com.

Background: The prognosis of gliomas is directly related to the tumor grade. The molecular profile allows specific characterization of gliomas, enabling the customization of therapeutic plans to improve prognosis and treatment efficacy. In this context, immunohistochemical analysis stands out as a good alternative for the early identification of tumor severity and consequently the management to improve patient prognosis. **Objective:** To associate the main immunohistochemical alterations with the tumor grade in patients undergoing glioma resection at a tertiary referral hospital. **Methods:** A descriptive, cross-sectional, and retrospective study was conducted with patients who underwent brain tumor resection at a tertiary referral hospital in the state of Ceará between 2013 and 2023. A total of 158 patients were analyzed, with the appropriate authorization from the ethics committee. **Results:** Regarding the characterization of the included patients, the mean age was 48 years, ranging from 17 to 85 years, with a median age of 50 years. Of these patients, 56% were male, and 44% were female. Concerning the diagnosis of brain tumor severity, no patients with grade 1 gliomas were identified. Seventy-four patients were diagnosed with grade 2 gliomas, of which, in terms of immunohistochemistry, 23 (31%) were positive for synaptophysin, 37 (50%) showed P53 alteration, 47 (63%) had IDH1 mutation, 38 (51%) had preserved ATRX, 50 (65%) were positive for GFAP, and 32 (43%) for S100. Eighteen patients were diagnosed with grade 3 gliomas, with 5 (28%) positive for synaptophysin, 9 (50%) showing P53 alteration, 12 (67%) had IDH1 mutation, 8 (44%) had preserved ATRX, 14 (78%) were positive for GFAP, and 6 (33%) for S100. Sixty-six patients were diagnosed with grade 4 gliomas, with 11 (17%) positive for synaptophysin, 40 (60%) showing P53 alteration, 46 (70%) had IDH1 mutation, 36 (54%) had preserved ATRX, 54 (81%) were positive for GFAP, and 39 (59%) for S100. **Conclusion:** The high positivity of GFAP in all three types of gliomas suggests it could be considered a diagnostic marker for brain tumors. Prognostically, ATRX was primarily identified in low-grade gliomas, while S100 was more present in glioblastomas. The IDH1 mutation is associated with worse prognoses in patients with gliomas, as observed in this study, consistent with the literature. Thus, the importance of immunohistochemical analysis for the diagnosis and prognosis of brain tumors is evident, highlighting the need for further studies to demonstrate the correlation between severity and altered markers.

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Two-year longitudinal evaluation of brain structural changes and ataxia severity in RFC1-related disorder

Camila Caroso Lobo[1], Thiago Junqueira Ribeiro Rezende[1], Gustavo Moreira Jarola[1], Fabrício Diniz Lima[1], Paula Camila Alves de Assis Pereira Matos[2], Alberto Rolim Muro Martinez[1], José Luiz Pedroso[2], Orlando Graziani Povoas Barsottini[2], Wilson Marques Jr[3], Marcondes Cavalcante França Jr[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.
[2]Universidade Federal de São Paulo, São Paulo SP, Brazil.
[3]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Ribeirão Preto SP, Brazil.

Correspondence: Camila Caroso Lobo; Email: camilaloboc@gmail.com.

Background: The structural characteristics of the brain in RFC1-related ataxia are well-documented; however, the progression of brain damage over time remains poorly understood. **Objective:** To delineate the evolutionary patterns of brain changes in RFC1-related ataxia through longitudinal neuroimaging assessments and to compare these findings with clinical evaluations of ataxia severity using the SARA scale. **Methods:** A cohort comprising ten patients diagnosed with RFC1-related ataxia and ten age- and sex-matched healthy controls underwent 3T MRI scans at two-year intervals. Gray matter volumes were analyzed using FreeSurfer, cerebellar structures were assessed with CerebNet, white matter integrity was evaluated via DTI multiatlas analysis, and spinal cord morphology was examined using the Spinal Cord Toolbox. Longitudinal changes between the groups were investigated using covariance analysis, with effect sizes (ES) calculated for significant findings. **Results:** Compared to healthy controls, patients with RFC1-related ataxia exhibited more pronounced progressive atrophy in the brainstem (ES=2.18), left hippocampus (ES=1.21), and right thalamus (ES=1.24). Additionally, there was a reduction in cross-sectional area at C1 of the spinal cord (ES=1.09) and an increase in axial diffusivity of the middle cerebellar peduncle (ES=1.44) over the study period. Concurrently, mean SARA scores increased from 15.2±5.1 to 19.4±7.9 (ES=0.62). **Conclusion:** Quantitative neuroimaging methods effectively capture the progressive nature of RFC1-related ataxia, demonstrating greater sensitivity compared to clinical evaluation based on SARA scores. These findings underscore the utility of longitudinal MRI assessments in understanding the evolving brain changes associated with RFC1-related ataxia and their potential as a sensitive biomarker.

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Ultra-high-field brain imaging at 7 Tesla magnetic resonance in LAMA2-RD patients

Clara Gontijo Camelo[1], Maria da Graça Morais Martin[1], Khallil Taverna Chaim[1], Leandro Tavares Lucato[1], Suely Fazio Ferraciolli[1], Claudia da Costa Leite[1], Umbertina Conti Reed[1], Paulo Ribeiro Nóbrega[2], Edmar Zanoteli[1]

[1]Universidade de São Paulo, São Paulo SP, Brazil.
[2]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Clara Gontijo Camelo; Email: claragc@gmail.com.

Background: LAMA2-related muscular dystrophy (LAMA2-RD) is an autosomal recessive disorder that causes muscle weakness in a broad spectrum of phenotypic severity. The disease affects muscle strength and causes brain abnormalities, such as high signal intensity in the white matter on T2 and FLAIR, and in rarer cases, cortical malformations. The correlation between motor severity and CNS abnormalities is not well understood, and few studies have detailed the brain MRI alterations in these patients. Ultra-high-field MRI [≥7 Tesla (7T)] has improved the capability to depict and characterize brain structures efficiently, with better signal-to-noise ratio and spatial resolution. **Objective:** To compare the brain imaging findings at 7T magnetic resonance with the images obtained with 3 Tesla magnetic resonance techniques in patients with LAMA2-RD. **Methods:** Patients with LAMA2-RD who had previously 3T brain MRI were selected, and were submitted to a 7T brain MRI protocol. The patients included both the congenital muscular dystrophy (CMD) form of the disease (unable to walk) and the limb-girdle muscular dystrophy form (LGMD). Patients under 16 years old who could not tolerate the exam without anesthesia were excluded, as well as those who had previously undergone spine surgery. **Results:** Four patients were submitted to the 7T brain MRI by now, aged between 26 and 40 years. Two patients had the phenotype of CMD, and two had LGMD. Three patients had epilepsy. The images revealed generalized white matter alterations with both field strengths, although the 7T better depicted a tigroid pattern, that could represent some perivascular preserved fibers. The subcortical area was preserved in certain regions among patients with the LGMD phenotype. All three patients with epilepsy showed cortical malformations, specifically polymicrogyria / cobblestone lissencephaly. These malformations were predominantly located in the occipital region and were not widespread across the cortex. Although the 7T MRI provided better resolution, it did not reveal additional cortical malformations compared to the 3T MRI. Also, due to 7T technical limitations, the occipital cortex was of limited identification, mainly in patient with accentuated kyphosis. **Conclusion:** Both patients with CMD and LGMD related to the LAMA2 gene exhibit CNS abnormalities. The 7T MRI provides better resolution, but did not identify more cortical malformations than previously identified with 3T.

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Ultrasonographic characteristics in prevalent myopathies: a systematic review

Lucas Frigo[1], Matheus Henrique Brustolim Marcucci[1], Ana Luiza Souza Santos[1], Leslie Mateus de Sousa Nunes[1], Leonardo Elias Araujo[1], Letícia da Costa Rios[1], Letícia Gadelha Caldas[1], Júlia Viana de Castro[1], Leonardo José Rodrigues de Araújo Melo[1], David Augusto Batista Sá Araújo[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Lucas Frigo; Email: lucas.frigo2@gmail.com.

Background: Ultrasonography has emerged as a valuable tool in the assessment of prevalent myopathies, offering real-time, detailed visualization of skeletal muscle tissue. Advanced ultrasound modalities including shear wave elastography have expanded diagnostic capabilities, providing insights into muscle morphology, edema, atrophy, and lipid infiltration. **Objective:** To summarize and analyze the ultrasonographic characteristics of prevalent myopathies, aiming to enhance diagnostic precision and therapeutic strategies. **Methods:** A systematic search of literature was conducted on PubMed, following PRISMA protocol, using DeCS/MeSH descriptors ("idiopathic inflammatory myopathies" OR "inflammatory myopathies" OR "myositis") AND ("muscle ultrasound" OR "neuromuscular ultrasound" OR "ultrasound").The inclusion criteria were cohorts or clinical trials published in English from 2019-2024. Reviews, case reports or animal studies were excluded. Initially, 140 articles were found, of which 9 studies met the inclusion criteria. **Results:** The systematic review of ultrasonographic findings in prevalent myopathies highlighted significant insights. In healthy controls aged 50 years and older, shear wave speed (SWS) in the deltoid muscle during passive stretch was notably lower compared to younger controls (2.80 vs. 2.47m/s). Echogenicity and power Doppler values varied widely across muscles examined, ranging from 1 - 3 and 0 - 4, respectively. Age, sex, and BMI significantly influenced muscle metrics. Age inversely correlated with SWS in the deltoid (Rs:-0.40) and positively with echogenicity in the flexor digitorum profundus (Rs:0.38). Higher BMI was associated with lower SWS (Rs:-0.45, p=0.018) and higher echogenicity (Rs:0.69) in the deltoid. Comparative analyses between healthy controls and patients with inclusion body myositis (IBM) and necrotising autoimmune myopathy (NAM) revealed specific differences. In IBM, the flexor digitorum profundus exhibited increased echogenicity and reduced muscle thickness compared to controls and patients with polymyositis/dermatomyositis. These findings underscore ultrasound's utility in differentiating myopathies, providing valuable quantitative data for clinical assessment and monitoring of muscle pathologies. **Conclusion:** This review examined ultrasonographic characteristics in prevalent myopathies, revealing significant insights into diagnostic and monitoring capabilities. The findings underscored age, sex, and BMI as critical factors influencing muscle metrics such as shear wave speed and echogenicity. Moreover, comparisons between healthy controls and patients with specific myopathies like inclusion body myositis highlighted distinctive ultrasound markers such as increased echogenicity and reduced muscle thickness. These quantitative findings enhance our understanding of muscle pathology and emphasize ultrasound's potential as a non-invasive tool for precise diagnosis and therapeutic monitoring in clinical practice.

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Ultrasound in the diagnosis of hereditary sensory neuronopathies: report of two cases

Vitor Santos de Souza[1], Rodrigo Santiago Gomez[1]

[1]Universidade Federal de Minas Gerais, Hospital das Clínicas, Belo Horizonte MG, Brazil.

Correspondence: Vitor Santos de Souza; Email: vitorsantosdesouza@outlook.com.

Case presentation: Patient 1, 67 years old, parents were cousins. Progressive course started 10 years ago, with main complaints of imbalance and chronic dry cough. Clinical examination revealed bilateral and symmetrical foot paresis, mild paresis of thigh adductors, myotatic areflexia in upper and lower limbs. Bilateral extensor plantar cutaneous reflex. Reduced superficial and deep sensation up to the iliac spine. Ocular motricity showed difficulty with pursuit eye movements and absence of vestibulo-ocular reflex. Mild gait ataxia. Patient 2, 65 years old, parents were cousins. Progressive course started 4 years ago, with main complaint of gait disturbance and chronic dry cough. Clinical examination showed reduced superficial and deep sensation in lower limbs, myopathic areflexia, absence of oculocephalic reflex, and Romberg test with non-specific side preference. Ultrasonographic evaluation of both patients revealed significant reduction in cross-sectional area of various peripheral nerves in upper and lower limbs. Genetic testing for cerebellar ataxia, neuropathy, and vestibular areflexia syndrome (CANVAS) was requested, showing biallelic expansion compatible with the diagnosis. **Discussion:** CANVAS affects middle-aged individuals. Diagnosis is made by identifying biallelic expansions of AAGGG in the replication factor C complex 1 (RFC1). Despite primarily sensory features, the phenotype of RFC1-related disease is expanding, including reports of motor neuron involvement, leading to a broad and sometimes challenging differential diagnosis. In the two cases described above, the presence of reduced cross-sectional area of peripheral nerves was crucial in suspecting CANVAS. Most sensory neuronopathies exhibit this reduction. Recently, ultrasound has gained prominence for its utility in recognizing and screening tests for predominantly sensory neuronopathies or late-onset ataxias of unknown cause. This is feasible due to ultrasound characteristics that can predict a hereditary or acquired origin. Recent studies show that acquired sensory neuronopathies have normal or enlarged nerves, whereas hereditary ones have abnormally small nerves. In the aforementioned case reports, besides clinical phenomenology, finding reduced average nerve diameters was fundamental for diagnostic reasoning and for recommending intronic biallelic expansion study in the RFC1 gene. **Final comments:** The use of ultrasound played a pivotal role in diagnosing the above-mentioned cases, due to its ability to aid in the identification of hereditary sensory neuronopathies. This underscores the advantages of using this tool in neuromuscular studies due to its practicality, low cost, detailed imaging capabilities, and minimal potential for harm.

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Underreporting of meningitis cases in the Northeast region of Brazil during the COVID-19 pandemic: an epidemiological overview

Guilherme Carvalho de Souza[1], Analuiza Silva Tenório Luna Sarmento[1], Alícia Eduarda Rios Soares[1], André de Oliveira Paiva[1], Antonia Cardoso Silva[1], Cecilia Maria Monteiro Barbosa[1], Gabriela Barbosa de Sá Rocha[1], João Victor Vasconcelos Tavares Maximiliano[1], Julio César Soares Barros[1], Larissa da Silva Almeida[1]

[1]Universidade Federal de Alagoas, Faculdade de Medicina, Maceió AL, Brazil.

Correspondence: Guilherme Carvalho de Souza; Email: guicadesouza@gmail.com.

Background: Meningitis is the inflammatory process of the membranes surrounding the brain and spinal cord, which can be of infectious or non-infectious origin. Due to its clinical and epidemiological significance, it is a compulsory notification condition. However, the onset of the COVID-19 pandemic in 2020 significantly impacted healthcare systems, affecting the epidemiological scenario and the reporting of conditions such as meningitis. **Objective:** To evaluate the underreporting of meningitis cases in the Northeast region of Brazil compared to the rest of the country during the COVID-19 pandemic. **Methods:** This is a quantitative, cross-sectional analytical study of meningitis notifications in the Northeast region of Brazil compared to the rest of the country from 2015 to 2021, using data from the Notifiable Diseases Information System (SINAN/DATASUS) for the variables of year of diagnosis and region of notification. **Results:** Between 2020 and 2021, the average number of meningitis notifications in the Northeast region of the country was 1139.5 cases, compared to an average of 2352.6 cases between 2015 and 2019, representing a decline of approximately 51.5%. This pattern was also observed in the national epidemiological analysis, which showed a reduction of approximately 56.9% in notifications during the same period. **Conclusion:** During the pandemic period of 2020 and 2021, there was a significant decrease in meningitis notifications both in the Northeast region of Brazil and nationally, which may be correlated with the coronavirus pandemic. Possible contributing factors to this epidemiological scenario include reduced access to healthcare services for individuals with meningitis, as well as difficulties in patient monitoring and updating health surveillance information due to the overload associated with the high rate of COVID-19 hospitalizations. Additionally, the adoption of social distancing, self protection and hygiene measures during this period may have contributed to the decline in the incidence of infectious meningitis.

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Understanding the impact of neuropsychiatric symptoms of dementia on caregiver burden: a systematic review

Ana Maria Porto Cortines Laxe Martins da Silva[1], Leonardo Sena de Souza[1], Luisa Lara Calazans[1], Marianna Leite[1], Carolina Braga Moura[1], Raquel Quimas Molina da Costa[1]

[1]Universidade Federal Fluminense, Niterói RJ, Brazil.

Correspondence: Ana Maria Porto Cortines Laxe Martins da Silva; Email: analaxe@id.uff.br.

Background: Behavioral and Psychological Symptoms in Dementia (BSPD) can significantly contribute to caregiver burden, leading to physical, psychological, social, and financial strains on caregivers. The impact is particularly observed on family members, often women, who experience higher burden levels than men. These symptoms can also contribute to institutionalization, escalating care costs. Recognizing and managing these symptoms is crucial for enhancing the patient's and caregiver's quality of life. **Objective:** To explore the relationship between BPSD in people living with dementia, as assessed by the Neuropsychiatric Inventory (NPI), and caregiver burden scales. **Methods:** A systematic review was conducted, searching PubMed, LILACS, Scielo and Embase databases for articles in English, Spanish, and Portuguese without time restrictions. Articles focusing on dementia and utilizing the NPI for symptom measurement were included. Screening and data extraction were performed independently by three reviewers, with disagreements resolved through discussion. **Results:** Of the 368 articles identified, 16 met the inclusion criteria. Various types of dementia were included. Cognitive decline was assessed mainly by the Mini Mental State Examination, Barthel Index, and Global Deterioration Scale, while the caregiver's burden was measured by Zarit Burden Interview(ZBI) and Neuropsychiatric Inventory caregiver Distress (NPI-D) Neuropsychiatric symptoms such as Apathy/indifference and Agitation/aggression were significantly associated with caregiver burden, being the former the most prevalent. **Conclusion:** Specific BPSD are linked to caregiver burden, urging early identification and management to prevent caregiver overload and preserve the quality of life for both patients and caregivers. Efforts should be made to address these symptoms effectively to improve the overall care experience for all involved parties.

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Unexpected presentation: case report of inflammatory cerebral amyloid angiopathy in a young adult with Down syndrome

Mateus Gustavo Favaro[1], Thales Pardini Fagundes[1], Gustavo Maximiano Alves[1], Joyce Yuri Silvestre Yamamoto[1], Jorge Alberto Martins Pentiado Junior[1]

[1]Hospital São Lucas, Ribeirão Preto SP, Brazil.

Correspondence: Mateus Gustavo Favaro; Email: mateusgustavofavaro@gmail.com.

Case presentation: A 32-year-old female patient with Down syndrome was admitted to our hospital with a multisystemic disease, characterized by fever, vomiting, diarrhea, and dyspnea. Basic laboratory tests presented elevated serum inflammatory markers (C-reactive protein of 30mg/L and a erythrocyte sedimentation rate of 135 mm/h), and thoracic imaging revealed pleural and pericardial effusions. After one week of hospital admission, the patient developed neurological symptoms, characterized by dysarthria, right hemiparesis, and an altered mental status with seizures. On the brain MRI it was found an asymmetric leptomeningitis leptomeningitis, predominantly in the left hemisphere, along with T2/FLAIR cortical/subcortical hyperintensities on the precentral gyrus, and multiple cortical microbleeds on SWI sequence. The cerebrospinal fluid (CSF) analysis showed high inflammatory activity (CSF protein of 363mg/dL and 365 white blood cells/mm^3, with a lymphocytes predominance). No microorganismswere isolated on CSF cultures. The combination of these findings leads to the diagnosis of Inflammatory Cerebral Amyloid Angiopathy (CAA-RI). Initially, the patient received an intravenous pulse of methylprednisolone (1g/day for 5 days) followed by oral prednisone for maintenance, which resulted in partial improvement. However, a few months later, she was readmitted with exertional dyspnea and moderate pericardial effusion. A follow-up brain MRI showed improvement but persistent inflammation. CSF analysis and the absence of clinical response suggested refractoriness to corticosteroids. At this point additional immunosuppression was required. Rituximab was chosen for its efficacy and favorable safety profile. **Discussion:** The patient exhibited a spectrum of symptoms including cognitive decline, seizures, and focal neurological deficits, consistent with CAA-RI. The pathophysiology of CAA involves beta-amyloid deposition in leptomeningeal arteries, sharing mechanisms with Alzheimer's disease. In Down syndrome, triplication of chromosome 21, which includes the APP gene, leads to augmented APP expression and beta-amyloid accumulation, contributing to early Alzheimer's neuropathology. This may explain the presentation at such a young age. Initial treatment with methylprednisolone pulse therapy followed by prednisone maintenance showed partial improvement, but relapse necessitated additional immunosuppression.. Rituximab was chosen for its efficacy and lower risk of adverse effects. **Final comments:** Immunomodulatory treatment can significantly improve clinical and radiographic outcomes in CAA-RI patients, reducing recurrence probability. However, studies on CAA-RI lack robust data due to limited cases and lack of large-scale randomized trials. More rigorous research is needed to elucidate underlying mechanisms, optimize treatment strategies, and improve clinical outcomes.

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Unilateral protruding ear as a novel clue for Fragile-X-associated tremor ataxia (FXTAS)

Luis Eduardo Borges de Macedo Zubko[1], Renata Cristine Alves[1], Francisco Manoel Branco Germiniani[1], Hélio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Complexo Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Luis Eduardo Borges de Macedo Zubko; Email: luis.zubko@gmail.com.

Case presentation: A 51-year-old woman presented with a history of a troubled gait associated with imbalance and falls. Although initially, she reported having no relevant past medical history, during follow-up her family mentioned that the patient had global developmental delay and significant learning difficulties during childhood. Family history was also noteworthy, with 2/7 brothers, having formerly been diagnosed with cerebral palsy, with no previous history of perinatal complications. Additionally, 1 brother and 5 cousins had similar abnormal gait and possible intellectual complaints. Her maternal grandparents were first-degree cousins. Her gait deteriorated until she was wheelchair-bound within 2 years since symptom's onset. During that time, she also had a significant cognitive and functional decline, leading her to become completely dependent for self-care activities. Physical examination was unremarkable, except for an asymmetrical protruding right ear. Neurological exam showed severe executive dysfunction with frontal release signs, severe ataxic and hypophonic dysarthria, moderate bilateral dysmetria and dysdiadochokinesia, mild bilateral bradykinesia, bilateral hypertonia due to rigidity and paratonia, and wide-based stance with severe postural instability requiring either constant bilateral support or wheelchair. Brain MRI disclosed moderate global cortical atrophy and severe cerebellar atrophy with a vermis predominance. Laboratory work-up showed only signs of ovarian insufficiency and no other cause of ataxia. Final diagnosis was achieved with specific sequencing of the FMR1 gene with 114 repeats in one allele and 30 repeats in the other allele (Premutation 55-200), thus confirming the diagnosis of Fragile-X-Associated Tremor Ataxia (FXTAS). She has been treated with levodopa, buspirone and trehalose with mild improvement in her overall quality of life. **Discussion:** Fragile-X-associated tremor ataxia (FXTAS) is a hereditary X-linked ataxia characterized by increased CGG repeats in the FMR1 gene in the premutation range between 55-200. It presents clinically between 50-70 years of age with progressive gait ataxia, intention tremor, and mild parkinsonism. Constitutional abnormalities may include protruding ears in 20% of cases, similar to the complete phenotype seen in fragile X syndrome. Asymmetry in constitutional findings is rare in FXTAS, with only one previously reported case with unilateral macroorchidism. Cognitive decline with compromise of memory and executive function, mood lability, and/or anxiety varies amongst cases. MRI findings may include global brain atrophy and a specific focal increased signal in the middle cerebellar peduncles on T2-weighted images. **Final comments:** FXTAS is the most common hereditary ataxia with a X-link pattern of inheritance. Clinical clues for its diagnosis include protruding ears, personal, or family history of intellectual disability, or early ovarian failure.

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Unmasking hidden culprits: the role of neurodiagnostic imaging in ulnar neuropathy investigation

Carolina Rodrigues Dal Bo[1] , Mary Angela O'Neal[2], Miriam E. Defilipp[2], Jim S. Wu[2], Aditi V. Varma-Doyle[2]

[1]Hospital Israelita Albert Einstein, São Paulo SP, Brazil.
[2]Harvard Medical School, Brigham and Women's Hospital, Boston, MA, USA.

Correspondence: Carolina Rodrigues Dal Bo; Email: carolinardalbo@gmail.com.

Case presentation: A 33-year-old man presented with left elbow pain radiating into the forearm and the left fifth digit, occurring 1-2 times weekly, often triggered by specific positions. Physical exam revealed tenderness at posterior medial epicondyle of humerus (Tinel's sign), positive elbow flexion test, sensory deficits in fingers 4 and 5, consistent with McGowan Grade 1 ulnar neuropathy. Electromyography (EMG) and nerve conduction studies (NCS) were normal. Neuromuscular ultrasound and MRI identified an accessory anconeus epitrochlearis muscle overlying the ulnar nerve, which was compressing the ulnar nerve. **Discussion:** The anconeus epitrochlearis is a rare anatomical variant muscle that extends from the medial epicondyle of the humerus to the olecranon of the ulna. Although often asymptomatic, it can sometimes compress the ulnar nerve, leading to an ulnar neuropathy, mimicking a cubital tunnel syndrome. Traditional diagnostic tools such as EMG and NCS can be normal. Neurodiagnostic imaging, specifically neuromuscular ultrasound and MRI, played a pivotal role in identifying the accessory anconeus epitrochlearis muscle as the underlying cause. This underscores the importance of incorporating advanced imaging techniques when initial diagnostic tests are inconclusive but clinical suspicion remains high. **Final comments:** In cases of normal EMG/NCS, neurodiagnostic imaging (Neuromuscular ultrasound +/- MRI) is crucial for investigating ulnar neuropathy, especially in younger patients with activity-related symptoms even in absence of palpable mass. Excision of this muscle and anterior ulnar nerve transposition can be effective surgical treatments.

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Unmasking the great masquerader: neuroimaging of general paresis in neurosyphilis

Samir Andres Ramirez Ojeda[1], William Alves Martins[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Samir Andres Ramirez Ojeda; Email: samir.ramirez@edu.pucrs.br.

Case presentation: A 38-year-old female presented with a 3-month history of behavioral changes, initially attributed to a psychiatric disorder. Neurological examination showed cognitive impairment, sensory ataxia, pyramidal tetraparesis with exaggerated deep tendon reflexes. Serum VDRL was found to be reactive (1/64). Both reactive VDRL test (1/32) and positive FTA-Abs on cerebrospinal fluid confirmed the diagnosis of neurosyphilis. HIV testing was negative. Brain MRI demonstrated diffuse atrophy associated to frontotemporal hyperintensities. Serological and CSF studies confirmed syphilitic meningoencephalitis. She was treated with intravenous crystalline penicillin and presented negative follow up VDRL on CSF and serum. Her symptoms stabilized and she presented slight improvement in motor function. She remained severely compromised with disinhibition and a frontal lobe syndrome. **Discussion:** Syphilis is a multisystem chronic infection caused by treponema pallidum support. Although there is a decrease in the number of cases of syphilis with the introduction of penicillin into use, it is still an important cause of the sexually transmitted diseases in developing countries because of the increase of incidence of AIDS and HIV seropositivity in the world. Syphilis progresses into four stages if untreated as primary, secondary, latent and tertiary stages. Tertiary syphilis is seen 1-30 years after primary infection. This inflammatory disease progresses slowly as neurosyphilis or gummatous syphilis. Neurosyphilis is classified as early and late syphilis. Cerebrospinal fluid (CSF), meninges and vascular structures are involved in the early stages of neurosyphilis, while in the late stage; cerebral tissue and spinal cord parenchyma are affected. Therefore, neurosyphilis can manifest with many different symptoms. In this article, we present a case of neurosyphilis with progressive cognitive changes and intractable behavioral and psychiatric problems whose primary and secondary phases were not detected. **Final comments:** Neurosyphilis affects the brain, meninges and/or spinal cord, varying from asymptomatic to several specific syndromes. Initial misdiagnosis of a psychiatric disorder is not an uncommon presentation of general paresis. Although MRI is variable, severe atrophy associated to patchy hyperintensities in frontotemporal regions should raise suspicion, helping to unmask this 'great masquerader'. Neurosyphilis has been a rarely seen clinical entity within the last decade. It can be encountered in almost all psychiatric disorders including dementia, personality changes, mania, depression, psychosis and delirium.

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Unprecedented ZIC2 variant linked to holoprosencephaly 5: case report and literature review

Arthur Caribé Sena[1], Hugo Nunes Pustilnik[2], Emerson Raí Araújo Azevedo[3], Elvis Oliveira Fonseca[2], Juliana Coelho Xavier[1], Gustavo Felipe Dourado Abreu[1]

[1]Faculdade Zarns, Salvador BA, Brazil.
[2]Universidade Salvador, Salvador BA, Brazil
[3]Universidade do Estado da Bahia, Salvador BA, Brazil.

Correspondence: Arthur Caribé Sena; Email: arthurcaribe.17@hotmail.com.

Case presentation: Patient, 9-month-old, only child of non-consanguineous couple, followed up due to motor delay associated with malformation of the central nervous system and renal agenesis. Physical examination revealed axial hypotonia, spasticity in the extremities, bitemporal narrowing, ocular hypotelorism, upward slanted palpebral fissures, prematurely closed fontanel, prominent ears, deep philtrum and PC of 41.5cm (p3). Cranial MRI revealed agenesis of the corpus callosum, monoventricle, interventricular fusion in the midline - alterations in the formation of the supratentorial brain suggestive of semilobar HPE, in addition to delay in the formation of cerebral sulci and cisterns, and incomplete myelination of the white matter. Whole Exome Sequencing detected a probably pathogenic heterozygous variant in the ZIC2 gene, associated with AD holoprosencephaly (p.Val197Trpfs*21). 46, XY karyotype. Unaltered CGH-array. BERA revealed moderate/severe bilateral hearing loss. Urinary tract USG revealed renal agenesis on the right and mild hydronephrosis on the left. Thus, the diagnostic hypothesis of HPE5 was strengthened. **Discussion:** Holoprosencephaly occurs due to incomplete cleavage of the prosencephalon between the 18th and 28th day of gestation, affecting both the forebrain and the face and resulting in neurological manifestations and facial anomalies of variable severity. Mutations in the ZIC2 gene are the second most common heterozygous variants detected in patients with HPE, accounting for 4.8%-5.2% of patients with semilobar holoprosencephaly. The variant (p.Val197Trpfs*21) in ZIC2 described in this case is novel in the literature. Patients with ZIC2 mutations often have neurologic impairment, and renal/urinary anomalies may be more common than those with pathogenic variants in other genes. **Final comments:** The reported case represents the first description of the p.Val197Trpfs*21 variant in ZIC2 associated with HPE5. Neurological involvement is frequent in patients with HPE, and renal/urinary anomalies may be more common than those with pathogenic variants in genes other than ZIC2. This is a genetic condition, therefore chronic and without cure, and treatment should focus on symptoms and follow-ups. Early diagnosis and management, with the support of a multidisciplinary team, are important for treatment and prevention of complications related to this condition. The risk of recurrence for the couple's offspring is low because it is a non-inherited condition.

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Unraveling acquired hepatocerebral degeneration: a case study

Rhaná Carolina Santos[1], Aline Oliveira dos Santos Meneses[1], Laura Porto Nunes[1], Ana Letícia Amorim de Albuquerque[1], Carolina Oldoni[2]

[1]Universidade do Vale do Rio dos Sinos, São Leopoldo RS, Brazil.
[2]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Rhaná Carolina Santos; Email: santoscrhana@gmail.com.

Case presentation: 74-year-old female with type 2 diabetes mellitus, systemic arterial hypertension, hypothyroidism, previous hepatitis A, and hepatic steatosis as comorbidities. Using the following medications: metformin, metoprolol, rosuvastatin, ezetimibe, levothyroxine, duloxetine, mirtazapine, primidone. She reported the onset of tremors in the upper limbs (UL) 10 years ago, without specifying whether they occurred at rest or during movement, with symptom progression. She currently reports bradykinesia and constipation but denies hyposmia or sleep disturbances. Physical examination showed facial hypomimia, right eyelid ptosis, jaw tremor, Moderate bradykinesia, Resting tremor more evident in the right upper limb, postural tremor in both upper limbs, cogwheel rigidity worse on the left side, gait characterized by small and shuffling steps. Laboratory investigation was carried out with blood count, renal function and vitamins without changes, and serologies (HIV, hepatitis B and C and syphilis) were non-reactive. Investigation for Wilson's disease was negative. Other results indicated altered liver function: TGO 78, TGP 61, GGT 451, FA 138, INR 1.54. Abdominal MRI showed chronic liver disease. Brain TRODAT scintigraphy revealed symmetrical uptake and preserved neurotransmitter activity, suggesting integrity of the dopaminergic system. Brain MRI showed microangiopathy and T1 hyperintense areas in the basal ganglia, suggesting Acquired Hepatocerebral Degeneration (AHD). **Discussion:** AHD is characterized by extrapyramidal and neuropsychiatric symptoms, including lethargy, somnolence, emotional lability, hallucinations, and movement disorders such as parkinsonism, ataxia, and myelopathy. It is a chronic and rare condition that manifests in patients with advanced hepatobiliary diseases, such as cirrhosis. Although the pathogenesis of AHD is not fully elucidated, it is known to be associated with manganese (Mn) accumulation in the basal ganglia. This accumulation can be detected by MRI, evidenced by T1 hyperintensities. Due to the morbidity associated with AHD and its limited response to pharmacological treatment, early diagnosis is crucial. Liver transplantation is a therapeutic option, with improvement in the progression of AHD observed post-procedure. **Final comments:** In the presented case, the integrity of the dopaminergic system and the metabolic alterations secondary to chronic liver disease observed in MRI support the diagnosis. This example underscores the importance of multidisciplinary assessment and continuous monitoring in patients with chronic liver diseases, given their prevalence, for the early identification and proper management of neurological complications, which, although rare, ad significant morbidity. Therapeutic intervention should focus on optimizing the control of the underlying liver disease and managing the imposed neurological symptoms to improve patient's quality of life.

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Unraveling the genetic complexity of hereditary spastic paraplegias in a large Brazilian multicenter cohort

Cynthia Silveira[1], Carelis Del Valle González Salazar[2], Fabrício Diniz de Lima[2], Karina Carvalho Donis[3,4], Diana Maria Cubillos-Arcila[5], Pedro José Tomaselli[6], Orlando Graziani Povoas Barsottini[7], Júlian Letícia de Freitas[7], Roberta Paiva Magalhães Ortega[8], Sérgio Rosemberg[8], Fernando Freua[9], Sarah Teixeira Camargos[10], Daniel Sabino de Oliveira[11], Marcelo Maroco Cruzeiro[11], Helio Afonso Ghizoni Teive[12], Bruna Nadiely Victor da Silva[13], Thiago Cardoso Vale[11], Fernando Kok[9,14], José Luiz Pedroso[7], Wilson Marques Junior[6], Jonas Alex Morales Saute[3,4,5], Marcondes Cavalcante França Junior[2]

[1]Universidade Estadual de Campinas, Faculdade de Ciências Médicas, Departamento de Neurologia,Campinas SP, Brazil.
[2]Universidade Estadual de Campinas, Departamento de Neurologia,Campinas SP, Brazil.
[3]Universidade Federal do Rio Grande do Sul, Departamento de Neurogenética, Porto Alegre RS, Brazil.
[4]Hospital de Clínicas de Porto Alegre, Porto Alegre RS, Brazil.
[5]Universidade Federal do Rio Grande do Sul, Faculdade de Ciências Médicas, Programa de Pós-Graduação em Medicina, Porto Alegre RS, Brazil.
[6]Universidade de São Paulo, Faculdade de Medicina de Ribeirão Preto, Divisão de Neurologia, Divisão de Neurociências, Ribeirão Preto SP, Brazil.
[7]Universidade Federal de São Paulo, Departamento de Neurologia, São Paulo SP, Brazil.
[8]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
[9]Universidade Federal de São Paulo, Hospital das Clínicas, Departamento de Neurogenética, São Paulo SP, Brazil.

[10]Universidade Federal de Minas Gerais, Faculdade de Medicina, Departamento de Medicina Interna, Belo Horizonte MG, Brazil.
[11]Universidade Federal de Juiz de Fora, Hospital Universitário, Serviço de Neurologia, Juiz de Fora MG, Brazil.
[12]Universidade Federal do Paraná, Hospital de Clínicas, Departamento de Medicina Interna, Serviço de Neurologia, Unidade de Distúrbios do Movimento, Curitiba PR, Brazil.
[13]Universidade Federal da Paraíba, Programa de Neurociência Cognitiva e Comportamento, João Pessoa PB, Brazil.
[14]Mendelics Análise Genômica, Departamento Médico, São Paulo SP, Brazil.

Correspondence: Cynthia Silveira; Email: silveiracynthia89@gmail.com.

Background: Genetic heterogeneity is a hallmark of hereditary spastic paraplegias (HSPs) with more than 90 associated genes reported so far. Despite that, few studies attempted to characterize the genetic epidemiology of HSPs and none of them in the highly admixed Latin American population. **Objective:** To describe the genetic profile of HSP in a large multicentric Brazilian HSP cohort recruited through an ongoing collaborative effort involving ten groups from five states (São Paulo, Rio Grande do Sul, Minas Gerais, Paraná and Paraíba) and to evaluate the diagnostic yield of next-generation sequencing (NGS) for suspected HSP. **Results:** The present cohort is composed of 668 patients (358 men and 310 women) from 490 families. Declared consanguinity was reported in 102/490 families, not present in 316/490 and unknown/not available in 72/490. Considering all families, 236 were inconclusive or not genetically tested yet. From those index-cases who underwent NGS, 192/490 (39,18%) had non-diagnostic results whereas 298/490 (60,82%) had diagnostic results. The top five most frequent genotypes were: SPG4 [SPAST] 61/490 (12,45%), SPG11 50/490 (10,20%), SPG7 22/490 (4,49%), SPG76 [CAPN1] 19/490 (3,88%) and SPG5A [CYP7B1] 15/490 (3,06%). Apart from the 'known' HSP genes, we also uncovered newly recognized genes associated with the HSP phenotype: COQ7 gene variants were present in 6 patients from 5 families - the p.Met1? variant appearing in 4 patients either in homozygous state or combined with p.Arg54Gln or Arg107Trp, and in 2 patients with p.Arg54Gln in homozygous state; RNASEH2B p.[Ala177Thr];[Ala177Thr] is present in two HSP sisters, SNAPC4 p.[Trp404Ter];[Ala415Val] present in two siblings and SPTAN1 (SPG91) last gene related with HSP appearing in 2 relatives with the described variant p.[Arg19Trp]. **Conclusion:** The study emphasizes the heterogeneity of HSP and the importance of NGS methods for genetic testing also the need for further genetic analyses for the inconclusive cases.

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Unraveling the genetic landscape of Brazilian patients with undiagnosed cerebellar ataxia

Luiz Eduardo Novis[1], Shahryar Alavi[2], David Pellerin[3], Marcus Vinicius Della Coleta[4], Salmo Raskin[5], Mariana Spitz[6], Andrea Cortese[7], Henry Houlden[2], Helio Teive[1]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Programa de Pós-Graduação em Medicina Interna e Ciências da Saúde, Curitiba PR, Brazil.
[2]UCL Queen Square Institute of Neurology and the National Hospital for Neurology and Neurosurgery, Department of Neuromuscular Diseases, London, UK.
[3]McGill University, Montreal Neurological Hospital and Institute, Departments of Neurology and Neurosurgery, Montreal, Canada.
[4]Universidade do Estado do Amazonas, Departamento de Neurologia, Manaus AM, Brazil.
[5]Laboratório Genetika, Curitiba PR, Brazil.
[6]Universidade Estadual do Rio de Janeiro, Serviço de Neurologia, Departamento de Especialidades Médicas, Rio de Janeiro RJ, Brazil.
[7]University of Pavia, Department of Brain and Behavioral Sciences, Pavia, Italy.

Correspondence: Luiz Eduardo Novis de Faria; Email:luizeduardonovis@hotmail.com

Background: Hereditary ataxias (HAs) encompass a diverse and genetically intricate group of rare neurodegenerative disorders, presenting diagnostic challenges. Whole-exome sequencing (WES) has significantly improved diagnostic success. **Objective:** To elucidate genetic causes of cerebellar ataxia within a diverse Brazilian cohort. **Methods:** Biological samples were collected from individuals with sporadic or familial cerebellar ataxia, spanning various ages and phenotypes, excluding common SCAs and Friedreich ataxia. RFC1 biallelic AAGGG repeat expansion was screened in all patients. For AAGGG-negative cases, WES targeting 441 ataxia-related genes was performed, followed by ExpansionHunter analysis for repeat expansions, including the recently described GGCZFHX3. Variant classification adhered to ClinGen guidelines, yielding definitive or probable diagnoses. **Results:** The study involved 76 diverse Brazilian families. 16 % received definitive diagnoses, and another 16 % received probable ones. RFC1-related ataxia was predominant, with two definitive cases, followed by KIF1A (one definitive and one probable) and SYNE-1 (two probable). Early-onset cases exhibited higher diagnostic rates. ExpansionHunter improved diagnosis by 4 %.We did not detected GGC-ZFHX3 repeat expansion in this cohort. **Conclusion:** This study highlights diagnostic complexities in cerebellar ataxia, even with advanced genetic methods. RFC1, KIF1A, and SYNE1 emerged as prevalent mutations. ZFHX3 repeat expansion seem to be rare in Brazilian population. Early-onset cases showed higher diagnostic success. WES coupled with ExpansionHunter holds promise as a primary diagnostic tool, emphasizing the need for broader NGS accessibility in Brazil.

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Unraveling toxic-metabolic encephalopathy: the decisive role of EEG in hyperemesis-related seizures

Luís Guilherme Pacheco de Almeida[1], Luiza Gonçalves Fraga[1], Patrícia Coral[1], Marlon Wycliff Caeira[1], Carlos Eduardo Soares Silvado[1], Luciano de Paola[1], Mayara de Rezende Machado[1]

[1]Universidade Federal do Paraná, Complexo Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Luiza Gonçalves Fraga; Email: luizaag.fraga@gmail.com.

Case presentation: A 24-year-old pregnant woman at 15 weeks of gestation, with no significant medical history, presented with her first epileptic event in the context of severe hyperemesis gravidarum. The episode was followed by persistent altered consciousness. An electroencephalogram (EEG) revealed diffuse cerebral dysfunction with occasional multifocal epileptiform activity, predominantly bifrontal. The diffuse cerebral dysfunction observed on EEG, in association with the patient's altered level of consciousness, suggested an encephalopathic state overlaid by an acute symptomatic seizure. Given the clinical context, Wernicke's encephalopathy was suspected and later confirmed by MRI findings. Following thiamine administration, the patient showed progressive electroclinical improvement, with resolution of epileptiform activity and recovery of consciousness, and was discharged without further antiseizure medication. **Discussion:** Wernicke's encephalopathy (WE) is caused by thiamine deficiency, commonly associated with alcohol abuse, malabsorption, malnutrition, increased metabolism, or hemodialysis. A brief literature review was conducted to examine the relationship between thiamine deficiency and epileptic manifestations. In series of patients with WE or Korsakoff's psychosis, epileptic seizures are frequently mentioned, often attributed to alcohol withdrawal. In a group of 50 adult patients, 16 with neurological manifestations demonstrated epileptiform activity on their EEGs. Thiamine deficiency interferes with cerebral electrogenesis through several mechanisms. It impacts the biosynthesis of neurotransmitters such as acetylcholine and gamma-aminobutyric acid affects membrane stability and conduction along the neurilemma, provokes osmotic imbalance with perineural edema and neuronal swelling, and causes periventricular lesions due to high metabolic rates dependent on vitamin B1. These factors seem to contribute to transient epileptogenicity, which can be reversed with treatment before severe damage permanently affects neuronal circuitry. **Final comments:** This case illustrates the important role of EEG in diagnosing and managing encephalopathy in hyperemesis gravidarum. Although EEG findings are not specific, they offer significant diagnostic insights when combined with clinical history, aiding in the rapid identification of WE. Early thiamine treatment led to marked improvement, preventing irreversible neurological damage. This highlights the necessity of early recognition and intervention, particularly in pregnant women with severe hyperemesis gravidarum, to prevent long-term neurological sequelae.

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Unsteadiness of gait in terminal renal failure: Miller Fisher syndrome triggered by haemodialysis

Natália Contini[1], William Alves Martins[1], Lucas Porcello Schilling[1]

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Porto Alegre RS, Brazil.

Correspondence: Natália Contini; Email: nataliacontini4@gmail.com.

Case presentation: A 79-year-old man presented acutely with dizziness, unsteadiness of gait and falls during a session of hemodialysis, which had started two months earlier due to hypertensive nephropathy. Brain MRI ruled out structural or acute ischemiclesions. Neurological examination showed downbeat nystagmus, right facial paresis and sixth nerve palsies bilaterally. Muscle strength remained normal, but deep tendon reflexes were absent diffusely. He showed an unstable and wide-based gait, with a tendency to fall in all directions. Cerebrospinal fluid analysis showed high protein (79mg/dl) with a normal cell count (<5cell/mm3). Nerve conduction studies disclosed absent sensory conduction, along with motor conduction slowing and absent H-reflex. Miller Fisher syndrome (MFS) was diagnosed under clinical and neurophysiological features. Intravenous immunoglobulin was introduced, followed by slowly recovery of balance and cranial nerves palsies. Haemodialysis continued three times per week, without recurrence. **Discussion:** Chronic renal insufficiency leads to a myriad of neurological complications, of which one of the most common is peripheral neuropathy. Usually, gradual onset of a distal axonal neuropathy is the clinical picture. Haemodialysis and continuous peritoneal dialysis are thought to arrest progression or even recede uremic neuropathy. This view is, conversely, challenged by a few reports of acute or subacute demyelinating polyradiculoneuropathy associated to dialysis. Neurological involvement as a complication of dialysis has a scarce literature. It mainly manifests as Wernicke's encephalopathy, aluminum poisoning or electrolytes imbalance. A Guillain-Barré-like (GBS) illness after initiation of peritoneal dialysis has been eventually described since 1993. Small osmotic changes between interstitial tissue and nerve sheaths may predispose either to direct myelin injury or expose nerve tissues to immune vigilance. The clinical triad of ophthalmoplegia, ataxia and arreflexia

the hallmark of MFS, a variant of GBS described in 1956. Pathophysiological mechanisms involve the targeting of antibodies to peripheral nerves' gangliosides. Despite many reports of unusual triggers in MFS, association to haemodialysis had been reported only once. Similar to our patient, initial assessment led to misguided diagnosis focused on vascular complications of renal insufficiency. Sometimes forgotten clinical signs, such as deep tendon reflexes and gait exam, unveil directly the diagnosis. Despite positivity of anti-gangliosides in 83% of patients, diagnosis can be made on clinical grounds, as in our patient. **Final comments:** In conclusion, sudden onset of ataxic symptoms in patients in haemodialysis should not be overlooked and MFS should enter in the differential diagnosis.

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Unusual aseptic meningitis and cognitive syndrome in a patient under anti-TNF therapy for rheumatoid arthritis: a case report

Guilherme Lago Miranda[1], Isabela de Almeida Stella[1], Ingrid Lorena da Silva Gomes[1], Giovanni Caniçali Simões[1], Henrique de Moraes Bernal[1], Pedro Almeida Martins Pontes[1], Tainá Fátima Ramos Gonçalves[1], Marcele Schettini de Almeida[1], Herval Ribeiro Soares Neto[1], Sonia Maria Cesar de Azevedo Silva Moura Magalhães Gomes[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, São Paulo SP, Brazil.

Correspondence: Guilherme Lago Miranda; Email: lagomiranda.g@gmail.com.

Case presentation: A 66-year-old female retired lawyer, with a past medical history of rheumatoid arthritis (RA) treated with Golimumab for the last 10 months, with behavioral change in the past two months, after a single dose of intra-articular corticosteroid due to articular pain. Her family claims that she became progressively agitated, easily irritated and euphoric, and made excessive expenses. She was taken to a psychiatrist and started on olanzapine and valproic acid. Subsequently, her humor became depressed, with a feeling of anguish and loss of interest in her usual activities. Due to progression, she was taken to the emergency department. Her neurological exam was only remarkable for a cognitive impairment, scoring 21 out of 30 points in the Montreal Cognitive Assessment (MoCA), with losses in visuospatial function, attention, delayed recall and orientation. Her CSF showed a mild increase in cellularity of 7 and proteins of 58, and a brain MRI showed aseptic meningitis (AM). Considering her immunosuppressed status, she was started on empirical acyclovir, ceftriaxone and ampicillin, without improvement. A CSF metagenomic was negative. Paraneoplastic investigation was also unremarkable. As an alternative diagnosis, we considered the possibility of Golimumab being the trigger for the AM; therefore, the medication was stopped and she received a course of IVIg followed by IV methylprednisolone. Upon discharge, brain MRI and MoCa improved, and on outpatient follow-up, she no longer had remarkable findings on neuroimaging and her MoCA evaluation was 28/30. **Discussion:** RA can cause meningitis, but its treatment can also be linked to AM by direct effect or leading to infectious meningitis due to the immunosuppressive effect. 23 cases of RA patients developing AM due to the use of anti-TNF medications were found by Yıldırım, R. et al., with half of them within the first year of exposure. There are no specific findings that differentiate rheumatoid AM from anti-TNF-induced AM, but the development of the condition after starting treatment suggests the later. Since anti-TNF agents cannot cross the blood-brain barrier, it is thought that the inhibition of tumor necrosis factor (TNF) results in paradoxical increase in its concentration within the CNS. Additionally, it can lead to immune dysregulation by inhibiting the apoptosis of self-reactive T cells. There is no consensus on treatment, but discontinuation of the precipitating drug and high-dose corticosteroids are the most common initial choices. **Final comments:** AM gives rise to a series of possible differential diagnoses, so a thorough investigation is mandatory. Special attention was paid to infectious etiologies, and we only pursued immunotherapy after a negative study of her CSF. It's a known fact that medications can cause AM, and anti-TNF, already linked to a series of other neurological diseases, has to be on the list of causes when evaluating a patient.

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Unusual kleptomania in a Parkinson's disease patient treated with pramipexole: a case report

Maria Eduarda Vieira de Oliveira[1], Carlos Frederico Lima[2], Maria Clara Arcoverde[2], Maria Thayana Barros Bandeira[3], Maria Luiza Vasconcelos Montenegro[1], Laís Diniz Pessoa de Santana[1]

[1]Universidade de Pernambuco, Recife PE, Brazil.
[2]Hospital Universitário Oswaldo Cruz, Recife PE, Brazil.
[3]Faculdade de Medicina de Olinda, Olinda PE, Brazil.

Correspondence: Maria Eduarda Vieira de Oliveira; Email: eduardaoliveira0194@gmail.com.

Case presentation: A 49-year-old male with Parkinson's Disease (PD), diagnosed in 2017, initially presented with rigidity in the left upper limb, progressing to resting tremor in the same limb, later affecting the right upper limb and both lower limbs. He also reports chronic constipation, agitated sleep, festinating gait, difficulty swallowing, dysarthria, and various bodily pains. The patient was using Pramipexole 0.125 mg, Fluoxetine 20 mg, Clonazepam 2 mg, and Levodopa 800 mg daily. Initially, he was taking Pramipexole 0.125 mg three times a day. Due to nocturnal freezing and the short duration of Levodopa's effect, the dose of Pramipexole was increased to 1 mg three times a day. Later, he reported taking 1.5 mg of Pramipexole three times a day, resulting in significant improvement in motor symptoms. At the most recent consultation, he was advised to reduce Pramipexole to 1.5 mg per day due to significant dyskinesias and compulsive behaviors. Other medications remained unchanged. During the evaluation, the patient reported compulsive behaviors related to Pramipexole. He mentioned an uncontrollable urge to take items from supermarkets, a behavior never exhibited before, causing concern for his wife. He reported taking multiple small items from the market that he did not need, just to satisfy this urge. He has no diagnosis of conduct disorder, bipolar disorder, or antisocial personality disorder. **Discussion:** Kleptomania is characterized by a recurrent failure to resist the impulse to steal objects not needed for personal use or monetary value, with tension before the act and pleasure or relief during the theft (DSM-IV TR). Dopamine agonists like pramipexole are effective in treating PD motor symptoms but can induce impulse control disorders (ICDs) such as pathological gambling, hypersexuality, and compulsive shopping. The pathogenesis of ICDs involves hyperactivation of the mesolimbic dopaminergic system associated with reward behavior and impulse control. Pramipexole, by excessively stimulating D3 dopamine receptors, can dysregulate these circuits, leading to compulsive behaviors. In this case, the patient developed an uncontrollable compulsion to take items from the market, leading to a reduction in the pramipexole dose. Studies show that ICDs induced by dopamine agonists are relatively common, with prevalence ranging from 6.1% to 14% in PD patients. However, kleptomania as a side effect of pramipexole use has been reported only three times in the international literature. **Final comments:** This case illustrates the complexity of managing Parkinson's Disease, especially when medication side effects compromise the patient's quality of life. Identifying kleptomania as a rare side effect of pramipexole is crucial for appropriate therapeutic adjustment. Continuous studies on the adverse effects of dopamine agonists and regular patient monitoring are essential to improve outcomes and quality of life for these individuals.

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Unusual manifestation due to infection with viruses from the herpesviridae family: Guillain Barré syndrome associated with herpes simplex – a case report

Daniel Lacerda da Costa[1], Beatriz Carneiro Gondim Silva[1], Angelina Maria Martins Lino[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Daniel Lacerda da Costa; Email: dlacerdadacosta@gmail.com.

Case presentation: E.L.S, 72 years old, male, has a history of short-term paraparesis in 2021, pulmonary tuberculosis treated 8 years ago and benign prostatic hyperplasia. In April 2022, the patient developed non-specific flu-like symptoms, and after 20 days he developed signs of ascending weakness. He was admitted to our institution with significant limitations in his ability to walk. On neurological examination, the patient presented with flaccid tetraparesis, areflexia, in addition to appendicular ataxia of the upper limbs and facial diparesis. Upon admission, cerebrospinal fluid was collected, which resulted in: 90 cells, with a predominance of monocytes (45%) and lymphocytes (42%), proteinorrhaquia of 143 mg/dl and glucose 43 mg/dl. Herpes simplex I/II was identified in the viral panel. Electroneuromyographic examination demonstrated marked multifocal demyelinating neuropathy, fulfilling criteria for Guillain-Barré syndrome (GBS) in its AIDP form. Human immunoglobulin was initially administered at a dose of 2g/kg over 5 days with progressive worsening of the clinical condition. After the third day of hospitalization, with the results of the viral panel, IV acyclovir was added (dose of 10 mg/kg every 8 hours for 14 days), with significant improvement. The patient was discharged on May 16th to continue rehabilitation with the physiatry and multidisciplinary team. **Discussion:** The involvement of the nervous system by the herpes simplex virus is a common condition and can be serious if not identified early. Forms such as meningoencephalitis, Mollaret's meningitis, myeloradiculitis, Bell's palsy are extensively reported. Herpes virus-associated acute polyradiculoneuritis (PRN) is an uncommon manifestation. There are few reports in the literature related to the herpes family such as HSV-1, HSV-2, VZV, EBV and CMV and their association with GBS. The first report of the condition dates back to 1975, of a case that developed flaccid tetraparesis and areflexia due to HSV infection. Despite the use of corticosteroids and physiotherapy, the patient evolved well. There are reports of cases in which immunoglobulin and acyclovir were administered with good evolution. The patient in this case showed significant worsening in the first three days after starting immunoglobulin, with significant improvement after the introduction of acyclovir 10mg/kg/dose every 8 hours per day for 14 days, showing excellent evolution. **Final comments:** The recognition of unusual forms of common diseases is important, even more so in diseases that may compromise functionality and risk death, such as GBS. Increased cellularity in Guillain syndrome is one of the warning signs, and should list the possibility of differential diagnosis or associated disease, as in this case, the association with Herpes Simplex.

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Unusual oropharyngeal tremor in Parkinson's disease: a case report

Mariana de Almeida Cruz Silva[1], Gabriel Felipe Almeida de Barros[1], Bruno Henrique Carneiro Costa Filho[2], Luísa Couceiro de Albuquerque Macêdo[2], Marcela Maria Cabral Ribeiro[1], Aimê de Paula Santos[1], João Pedro Matos de Santana[2], Víctor de Moraes Chagas[2], Míriam Carvalho Soares[1], Marcos Eugenio Ramalho Bezerra[2]

[1]Universidade Federal de Pernambuco, Recife PE, Brazil.
[2]Universidade Federal de Pernambuco, Hospital das Clínicas, Recife PE, Brazil.

Correspondence: Bruno Henrique Carneiro Costa Filho; Email: brunohccosta97@gmail.com.

Case presentation: An 85-year-old patient with Parkinson's disease for 16 years began to exhibit vocal tremor in the last six years. A videolaryngoscopy was performed to better characterize the tremor and explore the option of treatment with botulinum toxin (BT) in the vocal cords. Meanwhile, considering the possibility that the oropharyngeal tremor could be due to comorbid essential tremor (ET), treatment with topiramate and primidone was attempted, but there was no benefit, leading to the discontinuation of these drugs. Videolaryngoscopy revealed the presence of resting tremor in the larynx, pharynx, base of the tongue and adjacent facial structures, consistent with the diagnosis of PD. It was decided not to administer BT due to the videolaryngoscopic findings, which suggested that the vocal simptons were mostly secondary to the pharyngeal and facial muscles tremor. Instead, the optimization of the dopaminergic therapy dose was chosen, along with continuing the speech therapy that the patient was already undergoing. **Discussion:** Resting tremor is one of the core motor symptoms of PD, but vocal tremor is less frequently observed and can significantly impair speech ability. It is usually due to laryngeal tremor, but palate, pharyngeal, and tongue tremor can also occur. Due to the uncommonness of this symptom, there are no treatment guidelines specifically for vocal tremor in PD, though optimization of antiparkinsonian medications are usually the first line of therapy. Anticonvulsants, benzodiazepines, and beta-blockers can also be tried, if comorbid ET is suspected. Specific cases may be managed with BT or deep brain stimulation. **Final comments:** We present a case of an elderly female with Parkinson's disease with an uncommon presentation of laryngeal, pharyngeal, tongue and facial resting tremor, resulting in significant speech difficulties. The case underscores the heterogeneous and progressive nature of the disease, as patients may develop additional symptoms and complications over time. There is a need for a personalized approach to effectively address these different presentations, as there is insufficient data to indicate the best treatment modalities for this specific symptom.

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Unveiling the fog: rapid cognitive decline in an elderly patient resolved with innovative therapies for seronegative autoimmune encephalitis

Ingrid Lorena da Silva Gomes[1], Sephora Sabrina Candido de Almeida[2], Guilherme Lago Miranda[1], Erick Dupont[3], Giovanni Caniçali Simões[1], Pedro Almeida Martins Pontes[1], Henrique de Moraes Bernal[1], Tainá Fátima Ramos Gonçalves[1], Herval Ribeiro Soares Neto[1], Sônia Maria César de Azevedo[1]

[1]Instituto de Assistência Médica ao Servidor Público Estadual, Hospital do Servidor Público Estadual, São Paulo SP, Brazil.
[2]Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.
[3]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Ingrid Lorena da Silva Gomes; Email: ilsgomes@yahoo.com.

Case presentation: An 82-year-old male patient presented with a 4-month history of gait disturbances leading up to an inability to stand or walk unassisted. Several episodes of forgetfulness, coupled with progressive and rapid changes in behavior and mood were also remarkable, prompting his family to seek specialized evaluation. His medical history included a thyroidectomy for thyroid malignancy at age 62, for which he was on levothyroxine. The neurological examination revealed a dysexecutive patient, with reduced fluency and a Mini-Mental State Examination (MMSE) score of 10/30, which deteriorated progressively, rendering further testing unfeasible. An investigation for rapidly progressive dementia was initiated. Serum levels of Vitamin B12, thyroid function, serology for HIV, hepatitis, and syphilis were normal. CSF analysis and neoplastic investigation also showed no abnormalities. Subsequent testing of an anti-neuronal autoantibodies panel (including cell surface and onconeural antibodies) yielded negative results prior to initiation of any immunotherapeutic treatment. Brain MRI evidenced symmetric signal alteration affecting the cortical/subcortical region of the insulae, thalami, and some central areas of the pons, with high signal intensity on T2/FLAIR and EEG was abnormal. Based on our primary suspicion of an autoimmune encephalitis, a course of IV methylprednisolone was started, with improvements in motor tasks and on his MMSE score of 17/30. Nevertheless, during steroid tapering, he started to deteriorate again. As he was responsive to immunotherapy, we requested a 5-day course of plasmapheresis (PLEX), which further established his progressive improvement: in learning abilities, a better performance on the clock test, and he achieved an MMSE score of 25/30. He also resumed playing the guitar, an activity he could no longer engage in, being back to his baseline. However, 4 months after complete cessation of prednisone, he experienced symptom recurrence, necessitating readmission and a new course of pulsed therapy, with subsequent improvement. We chose to maintain him on a minimal regimen of prednisone and started rituximab. **Discussion:** Seronegative autoimmune encephalitis comprises a group of encephalitides lacking identifiable specific antibodies, presenting a diagnostic challenge in neurological practice. Clinical presentation guides treatment decisions, as neuroimaging findings may not always be definitive. Initial therapies include high-dose corticosteroids, intravenous immunoglobulin, and PLEX. Long-term immunosuppressive therapy may be necessary in select cases. **Final comments:** Rapidly progressive cognitive decline requires thorough evaluation. Despite inconclusive complementary exams, acute immunotherapy should be considered for patients with progressive neurological impairment in the absence of suggestive degenerative signs. Treatment should be individualized and maintenance immunosuppression after a second relapse is essential.

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Unveiling the relationship between poliovirus vaccination rates and acute flaccid paralysis incidence: a data-driven analysis

Ariana Leal Borges da Cruz[1], Ana Flávia Novaes[1], Qesya Rodrigues Ferreira[1], Júlia Spínola Ávila[1], Davi Tanajura Costa[1]

[1]Universidade Federal da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Ariana Leal Borges da Cruz; Email: arianacruz@ufba.br.

Background: Poliovirus is a pathogen capable of causing poliomyelitis, commonly known as infantile paralysis. It has three serotypes, with type 1 most associated with paralysis development. Approximately 1% of infected individuals are estimated to develop acute flaccid paralysis (AFP). Brazil provides a cost-free vaccination schedule against the disease for all children. As a result of vaccination efforts, the country was certified free from wild poliovirus circulation in 1994. However, vaccine coverage has significantly declined over the past decade, re-emerging as a critical public health issue. **Objective:** In this sense, this study aims to analyze the incidence of Acute Flaccid Paralysis associated with vaccination coverage in Brazil. **Methods:** This study employs a quantitative cross-sectional approach. Data were sourced from the Department of Informatics of the Unified Health System (DATASUS), a division of the Ministry of Health of Brazil. Data from the Notifiable Diseases Information System (SINAN) are publicly accessible. The study analyzed vaccination coverage from 2011 to 2023 and incidence of Acute Flaccid Paralysis (AFP) among Brazilians up to 15 years old from 2011 to 2021, with 2021 being the latest year for which data are available in the system. **Results:** Between 2011 and 2021, there were 4663 reported cases of AFP, with 4662 occurring in individuals up to 15 years old. The year 2018 recorded the highest number of cases (520), with the Northeast region reporting the highest total notifications during this period (1761). Regarding ethnicity, 49.92% of cases occurred in individuals of brown skin color. Additionally, 55.75% of cases occurred in males. No cases were classified as "wild poliovirus", although three cases were characterized as "poliomyelitis-compatible" during the study period (one case each in 2011, 2018, and 2019). Moreover, 3935 cases of AFP were classified as "discarded poliomyelitis", six as "vaccine-associated", 99 as "inconclusive", and 620 as "ignored/blank". Confirmation criteria included 447 cases recorded as "ignored/blank", 353 as "clinical-epidemiological", 15 as "loss to follow-up", 27 as "death", 570 as "evolution", and 3252 as "laboratory". National vaccination coverage data indicated a rate of 101.33% in 2011, while the 2023 epidemiological bulletin reported coverage at 84.7%, marking a decline of 16.63%. The lowest vaccination rate occurred in 2021 (71.04%). **Conclusion:** There is no demonstrable association between cases of Acute Flaccid Paralysis and Poliomyelitis. However, the decline in vaccination coverage remains a significant concern in the country. Additionally, there is a pressing need for more detailed data regarding the final classification of AFP cases, particularly in light of the substantial number reported as "ignored/blank".

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Unveriling the intersection: cerebral venonus thrombosis in the context of ulcerative colitis

Gabriel Abrahão Stoliar[1], Catarina Dantas Corrêa[1], Valéria Cristina Scavasine[1], Juliano André Muzzio[1], Viviane Hiroki Flumignan Zetola[1], Marcos Christiano Lange[1]

[1]Universidade Federal do Paraná, Hospital de Clínicas, Curitiba PR, Brazil.

Correspondence: Gabriel Abrahão Stoliar; Email: gabrielstoliar@gmail.com.

AACB, male, 47 years, no comorbidities. Presented with sudden onset of severe periorbital and temporal headache. About four days later noticed blurred vision and speech impairment, while maintaining the headache pattern. The patient denied taking medication, smoking, alcohol or illicit drugs abuse. Systemic enquire revealed hematochezia for the last three months with episodes of mucus diarrheal stools. Neurological examination showed reduced comprehension, fluency, naming, repetition and reading associated with intelligible dysarthria, right homonymous hemianopsia and right nasolabial fold erasure. There were no other deficits. The patient was submitted to a CT scan that revealed intraparenchymal hemorrhage in the left temporal lobe with adjacent edema. Subsequently, CT angiography showed thrombosis of the sigmoid, transverse, straight venous sinuses and the left internal jugular. During hospital, stay AACB was diagnosed with active ulcerative colitis which was related to the prothrombotic status that caused the extend cerebral venous thrombosis with venous infarct complication. Anticoagulation with dabigatran was prescribed to treat CVT and mesalazine to colitis ulcerative's control. Inflammatory Bowel Disease (IBD) accounts for less than 2% of Cerebral Venous Thrombosis cases, with Ulcerative Colitis (UC)

being slightly more common than Crohn's disease (CD) among those cases. In these autoimmune diseases, the risk of developing thrombosis increases proportionally to disease activity. Systemic inflammation leads to excessive platelet and coagulation cascade activation and endothelial dysfunction, which culminates in an enhanced thrombosis tendency. Nevertheless, inflammatory diseases are not the only risk factors. Other predisposing factors, such as anemia, thrombocytosis, thrombophilia, deep venous thrombosis, oral contraceptives, infections and recent surgeries should also be considered. Clinical presentation of CVT related do IBD is similar to others CVT with headache present in 80% of the cases. Seizure, vomiting, hemi/tetraplegia, altered awareness, vision deficits and papilledema are also common. The involvement of several venous sinuses simultaneously is frequent, occurring in 50% of patients. The most affected sinus is the superior sagittal followed by straight sinus. The presence of venous infarct caused by CVT is also common. Treating CVT related to IBD is identical to CTV by other causes. Anticoagulation must be started as soon as possible, even in the presence of GI bleeding, unless it may be life-threatening. It is recommended that all the patients with IBD receive prophylactic anticoagulation when hospitalized due to its prothrombotic state. The use of steroidal anti-inflammatory drugs may be helpful reducing the hypercoagulable state and so the risk of thrombotic event. This case highlights the importance of a detailed medical history, with a thorough review of systems, in diagnosing the cause of cerebral venous thrombosis.

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Update on new pharmacological treatments for motor symptoms of Parkinson's disease: a systematic review

érica Otoni Pereira Miranda[1], Lukas Santos Freire[1], Priscila Santos Oliveira[1], Davi Tanajura Costa[1], Philip George Glass Andrade[1], Emmanuel Santos Trindade[1], Giovana Andrade de Oliveira[1]

[1]Universidade Estadual do Sudoeste da Bahia, Vitória da Conquista BA, Brazil.

Correspondence: Érica Otoni Pereira Miranda; Email: ericaotoni15@gmail.com.

Background: Parkinson's disease (PD) is a neurodegenerative disorder caused by the loss of dopaminergic neurons in the substantia nigra and the presence of misfolded α-synuclein in Lewy bodies. This disorder is present in at least 1% of the population over 60 years of age and is characterized by bradykinesia and at least one other symptom of rigidity and resting tremor, so that the manifestations are progressive and significantly affect quality of life. Currently available treatments aim to control motor symptoms, but do not affect the natural course of the disease. **Objective:** To provide an update on new pharmacological treatments for motor symptoms of Parkinson's Disease. **Methods:** This is a systematic literature review carried out in PubMed and BVS databases, using the descriptors "Pharmacological treatment" and "Parkinson's disease". Randomized controlled trials and meta-analyses published between 2020 and 2024, in English, Portuguese and Spanish were considered. The studies included were those addressing new drug treatments for motor symptoms of PD, with free access to the full text. Duplicates or those that did not correspond to these criteria were excluded. **Results:** Of the 708 articles initially found, 42 were included in this review. In general, those studies evaluate drugs according to PD scales and ON/OFF periods. Among the scales, the most used is the Unified Parkinson's Disease Rating Scale (UPDRS), a classification tool in 4 parts, with Part III - motor subscale - being the most relevant in this study. The ON and OFF states depict periods in which the drug has an adequate motor response or not, respectively. The most consolidated adjuvant drugs to levodopa for the motor treatment of PD were: apomorphine, pramipexole, selegiline, safinamide and opicapone, the last two being quite elucidated and effective in reducing OFF periods time and motor fluctuations, with improvement in UPDRS Part III. Safinamide (a modulator of dopaminergic and glutamatergic systems) was effective, in the medium to late stages of the disease, in doses of 50-100mg, and opicapone (a catechol-O-methyltransferase inhibitor), in doses of 25-50mg. Other drugs lack robust evidence or are controversial, such as isradipine (a dihydropyridine calcium channel blocker), effective, in one trial, at a dose of 10mg, but ineffective in the motor sphere in another, at the same dose. Also controversial, nilotinib (a tyrosine kinase inhibitor) - despite being beneficial in several studies, in doses of 150-300mg - was symptomatically ineffective and worsened motor function in the arms, in one trial, with the same dose. **Conclusion:** Few drugs have shown benefits in controlling the motor symptoms of Parkinson's disease, particularly when associated with levodopa. There is a lack of evidence for the use of these drugs as monotherapy in the management of the disease. Therefore, robust randomized controlled trials are needed to evaluate the real effectiveness of these new drugs.

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Urban mobility through the special assistance service (Atende) for people with autism in the city of São Paulo

Andréia Maria Souza Cardoso[1], Gabriela Vitória da Costa Barbosa[1], Lissandra Andrea Nadalini Mauá[1], Paula Santos Justino[1], Soraia Micaela Silva[1], Alessandra Cristina Guedes Pellini[1], Isaac Lucas Camargo Arruda[1], João Carlos Pina Faria[2], Éric Edmur Camargo Arruda[1]

[1]Universidade Nove de Julho, São Paulo SP, Brazil. [2]Faculdade de Medicina do ABC, Santo André SP, Brazil.

Correspondence: Andréia Maria Souza Cardoso; Email: andreiamaria@uni9.edu.br.

Background: The Special Assistance Service - Atende, was created through decree No. 36,071 of May 9, 1996 and is currently governed by Municipal Law No. 16,337, of December 30, 2015. It is a type of free, door-to-door transportation, with its own regulations, offered by the City of São Paulo, managed by São Paulo Transporte S.A. and operated by public transport companies in the city of São Paulo and accessible taxi cooperatives. **Objective:** It is aimed at people with autism, deafblindness or physical disabilities with a high degree of severity and dependence, from 7am to 8pm, from Monday to Sunday, excluding holidays. **Methods:** Data were obtained using an electronic form sent to the Electronic Citizen Information System (e-SIC) (https://esic.prefeitura.sp.gov.br/) at the São Paulo City Hall Public Transparency Portal, requested on 08/31/2023, identified with protocol 076111 and forwarded to SP TRANS (São Paulo Transporte S/A) requesting information on the quantity, age, region and neighborhood of the autistic users of the Special Attention Service (ATENDE) residing in the city of São Paulo. **Results:** By August 2023, 2761 autistic users were contemplated through ATENDE, of which 2138 were male, 566 were female, 1 was the other and 56 were not registered with their gender, 906 users reside in the east zone, 588 reside in the west zone, 650 reside in the north zone and 617 reside in the south zone of the city of São Paulo. In the city of São Paulo in the ATENDE service, ages 2 to 10 represent 76.74% of users, the minimum age was 2 years and the maximum of 84 years, the average age is 9.82 years and standard deviation of 6.93, the east zone has an average age of users of 8.87 years and standard deviation of 7.37 years, the west zone has an average of users' age of 8.31 years and standard deviation of 5.80, the north zone has an average age of users of 8.50 years and standard deviation of 6.40 and the south zone has an average age of users of 9 .56 years and standard deviation of 7.70. The neighborhoods with the most users are Americanopolis (south zone), Jardim Peri (north zone), Itaquera (east zone), Conjunto Residencial Jose Bonifácio (east zone), Parque Paulistano (east zone), Cidade Líder (east zone) and Jardim São Luiz (West Zone). **Conclusion:** The Special Care Service - Atende assists 2761 autistic people in the city of São Paulo by taking them to consultations and treatments. More studies on this population are necessary so that primary care can map and guarantee universal and equitable care for all.

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Use of antisense oligonucleotides as a therapeutic approach for Huntington's disease: a systematic review

Matheus Jarek[1], Anna Caroline Ulson da Costa[1], Eduarda Nakoula[1], Mohamad Ali Hussein[2]

[1]Universidade Positivo, Curitiba PR, Brazil. [2]Instituto de Neurologia de Curitiba, Curitiba PR, Brazil.

Correspondence: Matheus Jarek; Email: matheusjarek.up@gmail.com.

Background: Huntington's disease (HD) is a neurodegenerative disorder caused by the expansion of the CAG trinucleotide repeat in the HTT gene, which leads to the coding of mutant huntingtin (mHTT). Therapies for HD are still restricted to treating the symptoms. Recent clinical trials have evaluated the use of antisense oligonucleotides (ASOs) to reduce mHTT levels. ASOs are small single-stranded nucleic acid sequences that bind via Watson-Crick hybridization to the target RNA; regulating gene expression especially by acting on messenger RNA (mRNA) through cleavage and degradation of the ASO-mRNA complex mediated by RNase H. **Objective:** Evaluate the evidence on the efficacy of using ASOs as a therapeutic approach for HD. **Methods:** A systematic review was carried out according to the PRISMA protocol. PubMed, Embase, and ClinicalTrials.gov databases were used, and studies were selected up to January 2024. Inclusion criteria were defined based on study design, language, intervention performed, and outcomes of interest. Animal studies, in vitro studies, and literature reviews were excluded. **Results:** From 143 studies identified, 4 were included after the selection and eligibility stages, which totaled 986 participants in clinical trials evaluating the use of ASOs in HD. Three ASOs were identified: Tominersen, WVE-120101, and WVE-120102. Tominersen was associated with significant reductions in mHTT concentration levels in cerebrospinal fluid (CSF) (reductions of more than 40% were observed in patients on a 120 mg dose); however, there was a worse performance on the Composite Unified Huntington's Disease Rating Scale (cUHDRS) compared to placebo in patients who received tominersen intrathecally every 8 weeks. In addition, transient increases in CSF neurofilament light chain (NfL) levels, increases in ventricular volume, and the occurrence of serious adverse effects were observed; including 3 cases of hydrocephalus. As for WVE-120101 and WVE-120102, both are allele-selective ASOs, selectively decreasing mHTT and preserving HTT expression through targeting based on the single nucleotide polymorphisms (SNPs) rs362307 (SNP1) and rs362331 (SNP2), respectively. Patients on a 16 mg dose of WVE-120101 showed an average reduction of 11.6% in the concentration of mHTT in the CSF, while those treated with a 32 mg dose of WVE-120102 had an average decrease of 9.9%. No significant changes in CSF NfL levels were observed in either ASO. As for adverse effects, most were mild to moderate in intensity, but there was an increase in the number of serious adverse effects, such as ataxia, meningitis, and delirium, in patients taking the 32 mg dose of WVE-120102. **Conclusion:** Despite being a promising therapeutic approach for HD, ASOs are not yet completely effective for treating the disease. Promoting a substantial reduction in mHTT levels as well as reducing the occurrence of adverse effects represent challenges to be overcome in the development of effective ASOs for HD.

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Use of anxiolytics and antidepressants in medical students

Clara Alves Caram[1], Lucas Seri Iijima[1], Mércia Tancredo[1]

[1]Pontifícia Universidade Católica de São Paulo, Sorocaba SP, Brazil.

Correspondence: Clara Alves Caram; Email: claracaram@outlook.com.

Background: Medical students are constantly subjected to an intense routine from the beginning of their degree, with many activities and demands, which can lead to emotional and physical exhaustion. Consequently, the feeling of incapacity, which favors symptoms of anxiety and depression. Emotional signs and symptoms include depressive mood, apathy, pessimism, highlighting the delay in thinking and action, which has a direct impact on students' academic performance. Anxiety disorders and depression affect university students in general and this rate is even higher in medical students, due to the high study workload and the emotional stress faced by these students. **Objective:** To identify the prevalence of anxiety and depression among medical students and the presence of anxiolytics and/or antidepressants. **Methods:** After approval by the Ethics and Research Committee and acceptance of the Free and Informed Consent Form, medical students responded to a semi-structured, self-administered questionnaire developed for this research, created using Google Forms. **Results:** Of the 102 students surveyed, the majority (55.9%) used medication for depression or anxiety. When segmenting for clinical indication, most participants (82.5%) cited anxiety, followed by depression (47.4%), insomnia (17.5%), premenstrual tension (5.3%), obsessive compulsive disorder (3.5%) and one bipolar disorder student (1.8%). Among users of antidepressants and anxiolytics, they use the medications as prescribed by psychiatrists (84.3%), followed by neurologists (11.8%). It is important to highlight that 49.1% have been using it for more than 2 years. **Conclusion:** The vast majority of medical students reported a diagnosis of anxiety and/or depression and the continuous use of anxiolytics and antidepressants for more than two years, demonstrating that even when using these psychotropic drugs continuously, the signs/symptoms of anxiety and depression remain, characterizing the chronicity and resistance of this pathology, which highlights the need for reassessment of treatment methods.

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Use of artificial intelligence for brain mapping and surgical planning

Julio Cerqueira Sampaio[1], Bruna Rios Beserra[1], Lara de Oliveira Boaventura[1]

[1]Escola Bahiana de Medicina e Saúde Pública, Salvador BA, Brazil.

Correspondence: Bruna Rios Beserra; Email: brunabeserra23.1@bahiana.edu.br.

Case presentation: In 2022, a 44-year-old female patient from Salvador, Bahia, with a pre-diagnosed tumor in the posteromedial region of the left cerebral lobe, was admitted to the hospital due to a significant worsening of the neurological condition, in which the patient presented aphasia, low level of consciousness, intense headache, emesis, paresthesia in the right hemibody and difficulty processing working memory. At the time, the patient tested positive for COVID-19, which led to the neurological symptoms being associated with a viral infection. However, a new computed tomography scan showed significant tumor growth, with signs of degeneration suggestive of high-grade glioma. **Discussion:** Given the situation exposed, the surgical procedure to remove the tumor was performed, so a mapping of brain networks was used using magnetic resonance imaging with Omniscient Neurotechnology technology, the Quicktome. This resource assisted in surgical planning provided a clear visualization of brain areas affected by the tumor and brain areas that were at risk of being injured during resection, such as the limbic/paralympic system and the medial temporal system. **Final comments:** At this time, brain mapping carried out using Quicktome, based on studies from the Human Connectome Project, emerged as the safest method for performing the surgery, as it was this resource that provided a safe corticectomy without significant sequelae for the young patient. Therefore, the present work aims to highlight the importance of knowing individual brain tracts, as well as the importance of using software such as Omniscient Technology, which can bring greater safety to the procedure and a better prognosis for the patient.

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Use of audiovisual resources in the teaching of the photomotor reflex

Akson Isaac Oliveira de Souza[1], Giovana de Sousa Ferro Barbosa[1], Giulia Veloso Matias Santos[1], Luciana dos Santos Viana[1], Carlos Eugênio Rangel Vieira[1], Caio Vinicius Seixas Freitas[1], Helena Vieira Goudard[1], Jussara Mathias Netto Khouri[1]

[1]Universidade Federal do Rio de Janeiro, Macaé RJ, Brazil.

Correspondence: Akson Isaac Oliveira de Souza; Email: aksonisaac5@gmail.com.

The photomotor reflex, or pupillary light reflex, is characterized by the contraction of the pupils in response to a light stimulus that penetrates the eye. This mechanism involves connections between the retina and brainstem neurons, mediated by cranial nerves. Assessment of this reflex is a crucial step during the neurological examination, both in alert and comatose patients (Bear et al., 2017). Information and Communication Technologies (ICT) inserted in the educational environment offer teachers different ways of streamlining classes, in addition to stimulating new learning skills. Such resources are facilitating tools in the teaching of neurology, whose physiological and anatomical bases are often challenging and real barriers for medical students, due to their complexity (Schon et al., 2002). Didactic videos, for example, allow students a practical visualization of the topic, helping to avoid these unknown knowledge gaps during graduation (Nascimento et al., 2022). This work aims to create an animation as a teaching tool to demonstrate the neural pathway of the pupillary light reflex and its clinical importance. The animation will address the anatomical, physiological and clinical aspects of the photomotor reflex arc exam in a dynamic way, improving the learning process. It is concluded that knowledge of the afferent and efferent pathways of the photomotor reflex is fundamental for the physician, given its clinical relevance in the evaluation, diagnosis and monitoring of neurological patients.

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Use of benzodiazepines in sleep disorders in the elderly population: a systematic review

Kerolayne Lisa Thimoteo da Costa[1], Daniel Lima Varela[1]

[1]Atitus, Passo Fundo RS, Brazil.

Correspondence: Kerolayne Lisa Thimoteo da Costa; Email: kerolayne_thimoteo32@outlook.com.

Objective: This study sought to investigate in a generalized and rigorous way, through a systematic review, the use of benzodiazepines in the treatment of insomnia in the elderly, verifying their efficacy, effectiveness, possible benefits, and potential risks with chronic use. **Methods:** In this study, we considered high-qua- lity scientific works published in journals with a high impact factor, and that discussed the use of benzodiazepines in the elderly population as a method of treatment for insomnia. This research includes experimental, randomized clinical trials, preferably double-blind and with a control group, as well- conducted prospective observational studies on the proposed subject matter, and published between January 2013 and January 2022. Studies carried out on animals, adul- ts, and elderly people who are not taking benzodiazepines were not included in the analysis. Case studies and reports, literature reviews, systematic reviews, and meta- analyses were also excluded from this study. This process of elaborating and conducting the systematic review was guided by the guidelines of the PRISMA protocol. **Results:** The use of benzodiazepines in old age is controversial, as these drugs can cause a series of side effects, including congnitive impairment, excessive sedation, and the risk of falls. Tolerance and dependence on these drugs must also be taken into account. Therefore, the use of benzodiazepines should be monitored safely and restricted to a certain time period. Considering the risks and side effects associated with the use of benzodiazepines, it is crucial to consider alternative therapeutic measures, such as Sleep Hygiene and Cognitive Behavioral Therapy. **Conclusion:** This review provides key insights into the impact of the use of benzodiazepines in the elderly population for the treatment of insomnia, consolidating significant evidence that highlights the importance of sleep in health during aging. This work highlights the importance of a holistic approach in the treatment of insomnia in the elderly. Considering clinical, psychosocial and behavioral aspects, it is possible to develop care plans that improve the quality of life of the elderly, promoting healthy sleep and, consequently, healthy aging.

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Use of cannabidiol in refractory epilepsy with positive outcome: case report

Karen Arissa Iwase[1], Rafael Gustavo de Mello Couto[1], Barbara Franco Martins[1], Anna Claudia Lavoratti[1], Livia Ceolin Valério[2], Maria Julia de Souza Padulla[1], Laura Regovichi[2], Bianca Gomes Suckel[1], Carolina Brun Ferrarini[1]

[1]Centro Universitário Fundação Assis Gurgacz, Cascavel PR, Brazil.
[2]Unicesumar.

Correspondence: Karen Arissa Iwase; Email: karen.arissa@outlook.com.

Case presentation: Patient R.S, a 24-year-old female, was referred to a Reference Center in the West of Paraná for investigation of refractory epilepsy in 2018 following a status epilepticus (SE), during which she experienced recurring convulsive seizures without regaining consciousness between them. She reports suffering from generalized tonic-clonic and absence seizures, as well as focal seizures, for the past 10 years. Her first convulsive seizure occurred at the age of 8, and since then, she has not achieved adequate control of this pathological condition with medication, fitting the diagnosis of difficult-to-control refractory epilepsy. **Discussion:** In 2018, during the patient's first consultation at the Reference Center, she was already on several medications, including topiramate, lamotrigine, valproic acid, and carbamazepine. Subsequent consultations involved dose increases of these medications, yet the patient continued to experience frequent generalized seizures. Levetiracetam was added progressively to the regimen, along with increased doses of the other anticonvulsants, reaching maximum doses. However, the patient still experienced frequent seizures. In 2023, progressive doses of cannabidiol were introduced (current dose 200mg every 12 hours), and since then, the control has been very

satisfactory. As a result, the patient reports brief absence seizures, approximately 4 per month, and has not experienced any further episodes of generalized tonic-clonic seizures or the need for hospitalization. There has been a significant improvement in the quality of life perceived by the patient and her family. Currently, there has been a reduction in the dose of the current anticonvulsant regimen, with the patient currently taking Levetiracetam 750mg, two tablets every 12 hours; lamotrigine 100mg every 12 hours; Valproic acid 500mg every 12 hours, in addition to cannabidiol 200mg/ml, 1 ml every 12 hours. **Final comments:** Epilepsy is one of the major neurological diseases affecting young adults worldwide, and treatment with anticonvulsants fails to control seizures satisfactorily in over 30% of the population, directly impacting patients' quality of life, autonomy, and self-esteem. There is a need to explore alternative treatments, such as Cannabis derivatives, in which one of the main active components is cannabidiol (CBD), responsible for its anticonvulsant properties and the absence of typical psychoactive effects of Δ-9-tetrahydrocannabinol (THC). Furthermore, this derivative has been shown to be safe over a wide dosage range and has not produced significant adverse effects.

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Use of FilmArray® ME in the diagnosis of suspected central nervous system infections: experience in 2,502 cases

Renan Domingues[1], Irineu Massaia[1], Daiane Salomão[1], Márcio Vega[1], Myrna Monteiro[1], Camila Spinelli[1], Lais Guerra[1], Carlos Senne[1]

[1]Senne Liquor Diagnóstico, São Paulo SP, Brazil.

Correspondence: Renan Barros Domingues; Email: contato@renandomingues.med.br.

Background: The FilmArray® meningitis/encephalitis is a real time multiplex PCR for identifying 14 CNS infection agents, including bacteria, Cryptococcus and viruses. **Objective:** In this study, we retrospectively evaluated the clinical utility of using this method, in cerebrospinal fluid (CSF), in samples from patients with suspected neuroinfection. **Methods:** We retrospectively evaluated data from 2,502 CSF samples from patients with suspected CNS infection in whom CSF was submitted to the FilmArray®. The diagnostic utility of this panel was evaluated. **Results:** 888 tested CSF samples (35.5%) were positive with FilmArray®. Enterovirus was the most common agent, being present in 539 (60.7%) of the positive samples. 86 samples (9.7%) were positive for bacteria. Streptococcus pneumoniae was the most common bacterial agent identified by FilmArray, in 35 of the 86 CSF samples positive for bacterial infection (40.7%). In 21 out of the 2502 samples (21.4%) the etiological agent was identified by a microbiological method other than FilmArray, including bacterioscopy and culture. Of the 1614 FilmArray negative samples, an infectious agent was identified by conventional microbiology method in 4 cases, namely: Micrococcus sp, Escherichia coli, Streptococcus intermedius and Pseudomonas aeruginosa. Three out of these 4 samples were diagnosed as meningitis based on cytological and biochemical analysis of the CSF. **Conclusion:** FilmArray® was able to identify the etiology of neuroinfection in 867 cases where other etiological identification methods did not reveal the causative agent. In only 3 cases of meningitis, FilmArray was negative while a conventional microbiological test was positive. Therefore, FilmArray considerably increases the likelihood of an etiological diagnosis in cases of neuroinfection.

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Use of fingolimod in patients at a reference center for demyelinating diseases of Recife-PE: epidemiological characteristics and association with HLA-G

Bruno Paulo Teles Chaves[1], Vanessa Cristina Fragoso Cassiano Alencar[1], Marcílio José de Oliveira Filho[1], Mauricéa Novaes Costa Pereira[1], Lidiane Alves de Macedo Souza[1], Nathalia Saraiva Ramos e Cruz[1], Maria Iris Moraes Machado[1], Álvaro J P Silva Moreira[1], Norma Lucena Silva[1], Maria Lucia Brito Ferreira[1]

[1]Hospital da Restauração, Recife PE, Brazil.

Correspondence: Bruno Paulo Teles Chaves; Email: brunopaulo.c@gmail.com.

Background: Multiple sclerosis (MS) is an inflammatory disease with associated neurodegeneration affecting the central nervous system (CNS). In the preclinical and clinical studies performed to date in MS, fingolimod has been proven to exert different protective effects, including inhibition of lymphocyte egress, inhibition of microglial and astroglial activation, reduction of neuronal death, restoration of lost synapsis, and reduction of dendritic spine loss, among others. HLA-G is an immunological checkpoint molecule, which is immunoprotective in some physiological situations, such as pregnancy. he role of the HLA-G molecule in multiple sclerosis has been studied considering the specific characteristic of immune mediation, related to reactivation of T lymphocytes, presence of meningeal inflammatory cell niches B and Th17 lymphocyte populations. The expression of HLA-G or sHLA-G has been associated with poor survival, poor prognosis, therapeutic response, clinical and outcomes in various malignancies. **Objective:** To evaluate the profile of patients using fingolimod in a health center reference in demyelinating diseases in Recife-PE. Assess whether there is any variable that determines better or worse outcome related to the use of fingolimod, establishing sHLA-G levels in individuals who used the treatment. **Methods:** Ninety patients who underwent treatment with fingolimod at some stage of treatment were selected in an institutions database. **Results:** Seventy-one patients were female (78.8%). Fingolimod was initiated as first-line treatment in 14 patients (15.6%), 61 as second line (67.8%), 13 as third line (14.4%) and 2 as fourth line (2.2%). Of the patients who started fingolimod as second-line treatment, eight were using glatiramer acetate (13.1%) and 53 were using interferon (86.9%). Therapeutic failure ocurred in 44 patients (72.1%), with radiological worsening in 35 patients (57.4%). Of the patients who used fingolimod as second-line treatment, eight patients experienced therapeutic failure (13.1%), requiring a change to third-line medication. Therapeutic failure ocurred in 3 patients (23%) who used fingolimod as third-line treatment. The analysis of sHLA-G levels was carried out in 17 patients using fingolimod (N=2) and interferon (N=15) as first-line treatment. Patients using interferon had the highest levels of sHLA-G and resistance to the drug, being eligible for the use of fingolimod as a treatment for second (N=11) and third (N=4) lines. **Conclusion:** It's possible to observe that the majority of patients use fingolimod as second-line therapy, with a low rate of therapeutic failure. To clarify a possible connection between the use of fingolimod and low sHLA-G levels and better clinical response, it's necessary measuring sHLA-G levels after switching interferon treatment protocol for fingolimod and expanding the number of tests.

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Use of fingolimod in pediatric multiple sclerosis: assessment of patients from a specialized center

Mariana de Oliveira Trintinalha[1], Danielle Karla Feitosa Alves Feitosa[1], Alfredo Damasceno[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Mariana de Oliveira Trintinalha; Email: marianaatrintinalha@hotmail.com.

Background: Multiple sclerosis (MS) is a chronic inflammatory disease of the central nervous system, which mainly affects young women, however 2-5% of cases occur in children under 18 years of age, pediatric MS. Due to the greater risk of progression at this stage, the choice of treatment is very important. Two main strategies: scaling and high effectiveness. Among the highly effective medications is Fingolimod (non-specific sphingosine 1 phosphate receptor agonist), the first medication studied and released in pediatric MS. There is a scarcity of data in this population. **Objective:** To evaluate the characteristics of patients using Fingolimod in the outpatient clinic. **Methods:** Retrospective observational study with medical record analysis. Included patients diagnosed with pediatric MS, under 18 years of age at diagnosis, using BRANDO (ColaBoRative LAtin AmericaN Database for Multipe SclerOsis). **Results:** 7 patients in the pediatric age group using Fingolimod. The average age was 17 years old, 86% girls and 14% boys. All had RRMS. EDSS at diagnosis: 4 patients with unknown prior EDSS, three with EDSS of 1, 1.5, and 4. EDSS currently: 1 patient with EDSS of 1, 3 patients with EDSS 1.5, 2 with EDSS 2.5, and 1 with EDSS 0. The history of outbreaks before Fingolimod: 28.57% had 6 outbreaks and 14.29% had 1, 3 and 4 outbreaks. None had clinical flare-ups after fingolimod at the time of evaluation. Children are closer to the biological onset of the disease, so treatment at this stage determines long-term results. An important factor is the immaturity of the immune system, which can increase susceptibility to autoimmune diseases. There are several therapies for adults, however the safety of many has not been evaluated in children. Interferons and Glatiramer are started more frequently, approximately one third of these patients fail early and change treatment within the first 6 months. The PARADIGMS study demonstrated that Fingolimod was superior to Interferon β-1a in patients with RRMS aged 10 to 17 years, with lower rates of attacks and lesions on Magnetic Resonance Imaging (MRI). Immovilli et al., 2022 described a 13-year-old patient, after 2 years of using Interferon β-1a, he presented new lesions and was switched to Natalizumab, interrupted due to adverse effects, opting for Fingolimod, after 2.5 years, which patient was in NEDA 3 (absence of flare-ups, active lesions and no worsening in EDSS). The results from our study are consistent with the literature, 7 patients have shown no activity or progression of the disease to date. **Conclusion:** Fingolimod can be a suitable option, with a good tolerability and disease control profile, demonstrated in our patients and in the literature.

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Use of immunophenotyping and cytomorphology in cerebrospinal fluid (CSF) samples for diagnosis of neurological involvement in patients with leukemia

Renan Barros Domingues[1], Irineu Massaia[1], Myrna Monteiro[1], Márcio Vega[1], Camila Spinelli[1], Lais Guerra[1], Daiane Salomão[1], Carlos Senne[1]

[1]Senne Liquor Diagnóstico, São Paulo SP, Brazil.

Correspondence: Renan Barros Domingues; Email: contato@renandomingues.med.br.

Background: Lymphoid and myeloid leukemias are hematological neoplasms with the highest risk of central nervous system infiltration. The presence of neurological involvement in these conditions has very important therapeutic consequences. The diagnosis of this infiltration can be made through cytomorphology and immunophenotyping in CSF samples. **Objective:** To evaluated the results obtained with both methods in CSF samples obtained from patients with acute and chronic, lymphoid and myeloid leukemias, investigating the diagnostic performance of both methods. **Methods:** We retrospectively evaluated immunophenotyping and cytology data of CSF samples, assessed by two examiners, from 797 CSF samples from

patients with acute lymphoid leukemia (ALL), 19 cases of chronic lymphoid leukemia (CLL), 120 samples from patients with acute myeloid leukemia (AML), and 15 with chronic myeloid leukemia (CML). We also recorded the total cell count in the CSF of these patients and the percentage of samples with pleocytosis among those with confirmed neurological involvement. **Results:** Positivity with at least one method was obtained in:

- 163 cases of ALL (20.5%), with 79 (48.5%) positive with both methods, 6 (3.4%) positive only with cytology, and 78 (47.8%) positive only with immunophenotyping.
- 6 out of the 19 cases of CLL were positive with both methods and 8 were positive only with immunophenotyping.
- Forty cases of AML (33.3%), with 28 (70%) positive with both methods, 7 (17.5%) only with cytology, and 5 cases (12.5%) positive only with immunophenotyping.
- Nine out of 15 cases of CML (60%) were positive, with 7 being positive with both methods (77.8%), 1 case (11.1%) positive only with immunophenotyping, and 1 case positive only with cytology. Increased CSF cellularity among patients with CNS infiltration was found in:
- 42 (25.8%) cases of lymphoid leukemias
- 20 (50%) cases of AML, and all 9 cases of CML.

Conclusion: Immunophenotyping significantly increased diagnostic sensitivity for CNS infiltration, particularly in lymphoid leukemias. Immunophenotyping and cytology had similar diagnostic sensitivity in cases of CNS infiltration by AML and CML. The results confirm the importance of paired testing with both examinations. Only one in four cases of lymphoid leukemias and only half of the cases of myeloid leukemias with meningeal neoplastic infiltration showed increased CSF cellularity, highlighting the need for specific tests to increase the chances of detecting an infiltrative process.

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Use of telemedicine as a tool in the evaluation of traumatic tomographic changes for emergency referrals in secondary care hospitals in Rio Grande do Sul

Guilherme Nobre Nogueira[1], Rafaela Fernandes Gonçalves[2], Gustavo Rassier Isolan[2]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.
[2]Faculdade Evangélica Mackenzie do Paraná, Curitiba PR, Brazil.

Correspondence: Guilherme Nobre Nogueira; Email: guiermenobre@gmail.com.

Background: Neurotrauma represents a significant medical emergency, with the proper management of these patients being a major concern for healthcare professionals. In this context, telemedicine emerges as a significant advancement in the rapid diagnosis and treatment of individuals who have suffered neurological trauma. Delayed management of cranioencephalic and spinal cord injuries can result in a worsening prognosis for patients. **Objective:** Tto evaluate the use of telemedicine in assisting with traumatic tomographic changes, based on the different clinical histories of each patient. **Methods:** This is a cross-sectional, retrospective study involving 355 patients from 14 hospitals in the states of Rio Grande do Sul and São Paulo, conducted from April 2019 to October 2022. All patients signed an informed consent form for this research, and the data collection adhered to the General Data Protection Law. **Results:** The association between tomographic changes and the clinical history of the patients (categorized as follows: spinal cord injury (SCI) due to fall, SCI due to assault, traumatic brain injury (TBI) due to fall, TBI due to assault, TBI due to motor vehicle accident, progressive neurological symptoms, headache, and sudden neurological symptoms) was analyzed for the 355 patients. Of these, 23 (6.48%) suffered SCI due to fall, 23 (6.48%) suffered SCI due to assault, 88 (24.79%) suffered TBI due to fall, 46 (12.96%) suffered TBI due to assault, 46 (12.96%) suffered TBI due to motor vehicle accident, 90 (25.35%) presented with progressive neurological symptoms, 38 (10.7%) with headache, and 1 (0.28%) with sudden neurological symptoms. The analysis of the relative frequency underscores the clinical importance of each cause and aids in prioritizing care and resource allocation in emergency services. These data are crucial for guiding appropriate medical approaches in emergency cases and implementing effective prevention strategies to reduce the incidence of traumatic injuries. **Conclusion:** Telemedicine effectively addresses traumatic tomographic changes due to the increased likelihood of treatment within short care windows. Thus, telemedicine emerges as a significant approach to improving the survival and quality of life of post-traumatic individuals.

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Use of the erasmus GBS respiratory in sufficiency score in predicting mechanical ventilation: a study of Brazilian patients with Guillain-Barré syndrome

Miguel Inácio Silva Gomes[1], Fátima de Menezes Dantas[1], José Víctor Carvalho Dantas Gualberto[1], Heric Weverton dos Santos[1], Mário Emílio Teixeira Dourado Júnior[1], Selma Maria Bezerra Jerônimo[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Miguel Inácio Silva Gomes; Email: miguel.inaciosg@gmail.com.

Background: mEGRIS is a score that predicts the risk of a patient with Guillain-Barré Syndrome requiring mechanical ventilation during the first week of hospitalization. This score is a simplified version of the EGRIS score, but it has not yet been validated in Brazil. **Objective:** To evaluate the effectiveness of mEGRIS in predicting the risk of mechanical ventilation after 1 week of hospital admission in Brazilian patients with Guillain-Barré syndrome. **Methods:** A retrospective study was conducted, in which the medical records of 50 Brazilian patients diagnosed with Guillain-Barré syndrome between 2017 and 2024 were evaluated. The following data were collected to calculate mEGRIS: time from onset of weakness to hospital admission (in days), bulbar weakness (yes or no), cervical weakness (MRC 0 to 5) and bilateral psoas weakness (0 to 10). Each patient's classification on the Hughes Disability Scale was also recorded after 1 week of onset of motor symptoms. A Mann-Whitney U test was used to compare the mEGRIS of patients who underwent mechanical ventilation (Hughes 5) with those who did not (Hughes < 5). The variables were correlated using a Spearman test. The study was approved by the Local Ethics Committee (CAAE: 75882923.7.0000.5292) and all participants signed an informed consent form. **Results:** The median mEGRIS of patients undergoing mechanical ventilation was 25.0 (18.5 - 27.0), while that of those not undergoing mechanical ventilation was 4.0 (1.0 - 7.0), p<0.0001. As none of the patients died in the 1st week, there were no cases of Hughes 6, which would be a possible confounding factor in the analysis. The median time from disease onset to hospital admission was 4 days for patients undergoing mechanical ventilation (2 - 7 days), whereas those not undergoing mechanical ventilation had a median of 6 days (4 - 10 days), p= 0.231. The mEGRIS and Hughes variables showed a strong positive correlation (r=0.7857). **Conclusion:** Final analysis suggests that mEGRIS is effective in predicting the risk of mechanical ventilation after 7 days of hospitalization in Brazilian patients diagnosed with Guillain-Barré syndrome. Since it has similar accuracy to EGRIS, even with the assessment of a smaller number of muscle groups, mEGRIS is a valuable tool for emergency contexts. General practitioners can be trained to apply it, in order to identify patients at risk of developing respiratory failure at an early stage. To confirm our results, prospective studies with larger cohorts need to be carried out.

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Use of the palliative care screening tool for patients admitted for ischemic stroke

Luciana de Oliveira Neves[1,2], Igor Bessa Santiago[1], Vitor Silva Abreu[1], Lara Carvalho de Oliveira[1], Carmem Rachel Alves de Paula Flôr[1], Benevides José Silva Santos[1], Clarissa Rocha Montenegro[1], João Renato Figueiredo Souza[1], Karla Maria Carneiro Rolim[2], Paulo Goberlânio[3]

[1]Hospital São Carlos, Fortaleza CE, Brazil.
[2]Unichristus, Fortaleza CE, Brazil.
[3]Universidade de Fortaleza, Fortaleza CE, Brazil.

Correspondence: Luciana de Oliveira Neves; Email: dra.luciananeves@gmail.com.

Background: Stroke is an important cause of morbidity and mortality in Brazil and worldwide and one of the main pathologies to consider Palliative Care. Many advances have been made over the years, and there is no doubt about the importance of effective acute care for therapies such as thrombolysis and thrombectomy in acute ischemic stroke. The Palliative Care Screening Tool (PCST) is a tool that can be useful for screening patients to be referred to Palliative Care. **Objective:** To evaluate the usefulness of the PCST for patients admitted in the acute phase of stroke, undergoing thrombolysis and/or thrombectomy in a high complexity hospital during 2021. **Methods:** This is an observational, retrospective study, with data obtained through the analysis of medical records of patients hospitalized for acute ischemic stroke and undergoing thrombolysis and/or thrombectomy, in 2021. The study is part of the project approved by the Ethics and Research Committee of hospital itself. Two neurologist researchers evaluated the admission note and two others, the last evolution of hospitalization, using Google Forms. The following variables were evaluated: age, sex, NIH Scale (National Institutes of Health Stroke Scale) and Rankin on admission and discharge, outcome and whether Palliative Care was addressed. Upon admission, the PCST scale was applied, with its score categorized as up to 2 points: no indication of Palliative Care; up to 3 points: clinical observation; ≥4 points: consider Palliative Care. Data analysis was carried out using the SPSS program, obtaining the frequencies and means of the variables and the kappa index between the evaluators. **Results:** 14 medical records were evaluated. 57.14% of patients were male and the average age was 74.71 years. 71.4% underwent intravenous thrombolysis, 21.42% thrombolysis and mechanical thrombectomy and 7.14% thrombectomy alone. The average NIH admission among the researchers was 12 and 11 and the Rankin was 0 and 2. When using the PCST, for one researcher, 14.28% and for another 21.42% of patients should be referred to Palliative Care. Regarding the outcome, the average NIH of patients who were discharged was 5 and 3 and the average Rankin was 2.71 for both. Three patients (21.14%) died during hospitalization, one of which was mentioned Palliative Care in the medical record. The PCST Kappa index among the researchers who evaluated the admission was 0.759, with a significance of 0.003. **Conclusion:** The study demonstrated that the PCST has a good agreement between evaluators and could have been useful in detecting patients with a profile for a palliative approach. The use of assessment instruments can help with more assertive and person-centered decision-making. A characteristic of the place studied is that the Palliative Care team was only active at the end of the 2nd semester. As an improvement, the authors suggest a new prospective study with a larger number of participants.

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Use of the Wobble Chair to evaluate trunk postural control in individuals with Parkison's disease

Thayná Lye Ishii[1], Hayslenne Andressa Gonçalves de Oliveira Araújo[1], Sarah Possetti Godoi[1], Natália Mariano Barboza[1], Andressa Leticia Miri[1], Renata Pasquarelli Volpe[1], Suhaila Mahmoud Smaili[1]

[1]Universidade Estadual de Londrina, Londrina PR, Brazil.

Correspondence: Sarah Possetti Godoi; Email: sarahpossetti@gmail.com.

Background: Trunk postural control is crucial for our transfers and is an essential part of providing postural stability, especially in individuals with Parkinson's disease (PD). **Objective:** To evaluate trunk postural control in individuals with PD in the seated position using a Wobble Chair and to analyze the influence of cognitive and motor demands on their postural stability. **Methods:** A cross-sectional study with a sample of 15 individuals with idiopathic PD in mild to moderate stages. The Mini-Mental State Examination was used for cognitive assessment, and the modified Hoehn & Yahr and the Unified Parkinson's Disease Rating Scales were used to assess disease staging and progression, respectively. Trunk mobility and functional independence were measured using the Trunk Mobility Scale, and the Schwab and England Activities of Daily Living Scale. Postural control was assessed using the Wobble Chair, an instrument based on a force platform with a pivot and spring system that creates an unstable base for evaluating trunk control while in the seated position. Participants underwent four different conditions: a) eyes open (EO) - control position; b) eyes closed (EC); c) dual-task cognitive (DT); and d) no foot support (NF). The conditions were randomized, repeated twice for 30 seconds each, with a 30-second interval between each of them. The outcome measures collected were center of pressure (COP) sway area, anteroposterior (AP) and mediolateral (ML) amplitudes, as well as AP and ML velocities. Friedman's test was used for comparison between test conditions, considering $p<0.05$. Ethical approval is under opinion 31775420.8.0000.0108. **Results:** A larger COP area, amplitudes, and velocities were observed in the DT condition compared to the EO condition. The DT condition also showed greater AP amplitude and higher AP and ML velocities compared to the NF condition. Furthermore, the DT condition exhibited higher postural control values compared to the EC condition for the variables of AP and ML velocity. Thus, the DT condition was the most challenging position with increased activity of postural control indicators. There was no significant correlation between any trunk postural control variables and trunk mobility and functional independence assessments. **Conclusion:** The evaluation of trunk control while in the seated position in individuals with PD showed greater postural instability in more challenging positions, specifically when adding dual-task cognitive demands and removing visual stimuli, in all balance variables measured using the Wobble Chair, especially AP and ML velocities. Assessing postural control in the seated position under different conditions can aid in understanding neuromuscular strategies and treating postural control in individuals with PD, as most studies are conducted on individuals in the standing position.

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Use of transcranial doppler and optic nerve sheath diameter ultrasonography in acute idiopathic intracranial hypertension: a case report

Thamires Marx da Silva Santos[1], Rodrigo Santiago Gomez[1], Sabrina Gesteira Souza[1], André Luis de Oliveira Silveira[1], Italo Pena Guimarães[1], Nathália Luísa Carlos Ferreira[1], Gustavo Carvalho Oliveira Gonçalves Machado[1], Maria Luiza Nóbrega Lima de Albuquerque[1], Bernardo Gomes Muffato[1]

[1]Universidade Federal de Minas Gerais, Hospital das Clínicas, Belo Horizonte MG, Brazil.

Correspondence: Thamires Marx da Silva Santos; Email: marxthamires@gmail.com.

Case presentation: We report the case of an 18-year-old female patient admitted with a 25-day history of headache and visual blurring with progressive worsening. She was obese, having gained 25 kg in the last 4 months, without other comorbidities. Upon admission, she had a perception of movement in the right eye, with restriction of ocular movement in all directions, and in the left eye, she could count fingers at 10 cm and had restricted abduction. Bilateral papilledema was noted on fundoscopic examination. Cranial tomography revealed intraocular protrusion of the optic discs, thickened optic nerves bilaterally, and a partially empty sella turcica. A lumbar puncture was performed, with cerebrospinal fluid analysis within normal limits and an opening pressure of 85 cmH2O, determining, according to the International Headache Society criteria, the diagnosis of idiopathic intracranial hypertension (IIH). In the initial transcranial Doppler (TCD) study, signs of intracranial hypertension were observed with a mild to moderate increase in the pulsatility index (PI) in the middle cerebral artery bilaterally and the right vertebral artery, and an increase in mean velocity (MV) in these vessels. The ultrasonography of the optic nerve sheath diameter (ONSD) showed a significant bilateral increase, also indicating intracranial hypertension. A lumboperitoneal shunt was chosen, along with treatment with acetazolamide, topiramate, and five days of pulse therapy with methylprednisolone. The patient showed significant improvement in headache and partial improvement in visual acuity in both eyes (20/200 on the Rosenbaum chart), improvement in right eye ophthalmoparesis, with persistent left eye abduction restriction. The TCD performed after therapeutic measures showed normalization of MV and PI in the studied arterial segments, indicating improvement after intracranial pressure (ICP) reduction. **Discussion:** IIH is a disease of unknown etiology, presenting with signs and symptoms of elevated ICP in the absence of structural lesions or other identifiable causes. One of the main risk factors is obesity, and with the current global obesity epidemic, the incidence of IIH has increased considerably, making diagnostic, treatment, and follow-up measures increasingly important. Currently, ICP measurement is performed via lumbar puncture, an invasive and uncomfortable procedure for the patient. Non-invasive methods such as TCD and ONSD ultrasonography are being studied for indirect ICP assessment in these cases. Literature shows that increased ICP significantly affects cerebral blood flow, altering parameters evaluated by TCD, such as MV and PI, which tend to normalize after adequate treatment and ICP reduction, and that ONSD measurements have a positive correlation with ICP. **Final comments:** TCD and ONSD ultrasonography are useful, non-invasive, and low-cost tools that can potentially be implemented for the diagnosis, monitoring, and clinical management of patients with IIH.

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Use of virtual reality as a treatment for motor functions in patients with Parkinson's disease: a systematic review and meta-analysis

Luiz Carlos Figueiredo Filho[1], Walter Oliveira Verissimo[2], Ricardo Massoud Ormanes[1], Rafael Augusto Silva Cabeça[1], Ingrid Jade Muniz Wanderley[1], Alexandre Marques da Rocha[1], Luanna de Melo Pereira Fernandades[1], Leonardo Yuji Nihira Alencar[2]

[1]Universidade do Estado do Pará, Belém PA, Brazil.
[2]Universidade Federal do Pará, Belém PA, Brazil.

Correspondence: Leonardo Yuji Nihira Alencar; Email: leo1820.alencar@gmail.com.

Background: Parkinson's disease (PD) is degenerative, causes tremors, bradykinesia and muscle rigidity, and affects almost 1% of the world's population. Diagnosis is clinical, but accuracy is challenging due to overlapping symptoms and lack of a specific cause. In this sense, virtual reality (VR) can improve the treatment of the disease, complementing physiotherapy and developing physical, mental and emotional skills. **Objective:** To evaluate the use of virtual reality in rehabilitating Parkinson's patients, focusing on motor prognosis, balance, gait, coordination and quality of life. **Methods:** This is a systematic review with meta-analysis, defined using the PICOS strategy, where (P) patients with Parkinson's disease; (I) virtual reality; (C) conventional treatment; (O) improvement in motor symptoms, gait, balance and daily activities. The Preferred Reporting in Systematic Reviews and Meta-Analyses (PRISMA) guidelines were used. Scientific articles from randomized clinical trials (RCTs) and quasi-randomized clinical trials (QRCTs) were included. The articles were collected from the PUBMED/MEDLINE, VHL, Embase, Cochrane Database, Web of Science and Clinical Trial.gov databases. The results were arranged in tables using the Microsoft Excel 2016 program, according to the variables: author, year, age of participants, gender, type of intervention, evaluation protocol, outcomes analyzed and supervision by professionals during the test application. The meta-analysis was carried out using Review Manager 5.4 software. Heterogeneity between studies was assessed using the Chocran Q test and I2 statistic and interpreted following the Cochrane Handbook for Systematic Reviews of Interventions. **Results:** We identified 492 articles in the databases. After removing duplicates, 323 were selected, 119 full articles were assessed for eligibility, and 25 were included in the final synthesis of the review. It was observed that VR induced beneficial changes in balance when using the BBS scale MD=2.22 (95%CI 1.58-3.63, $p< 0.00001$) and improved motor function using the UPDRSIII index MD=-4.4 (95%CI -7.97; -1.71, $p=0.002$) when compared to conventional treatment. However, no substantial difference was found between the physiotherapy and VR groups regarding gait, balance, activities of daily living and motor function, indicating that the interventions are partially effective. The funnel plot showed a symmetrical dispersion in the analysis of clinical heterogeneity and publication bias. **Conclusion:** VR can improve the treatment of PD, it showed comparable efficacy to traditional physiotherapy in balance and motor functions, however, it does not improve all aspects of treatment. Further studies are needed to evaluate the complete integration of VR exercises to overcome the limitations of the few studies available for precise analysis and to definitively establish improvements in the treatment of PD.

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Using artificial intelligence to analyze praxic-phonic errors in acquired apraxia of speech

Marta Maria Silva Lira-Batista[1], Rogério da Silva Batista[2], Matheus Levi da Silva Barbosa[2], Werton Pereira de Silva[2], Romero Antônio Ramos de Mendonça[2], Marcela Lima Silagi de Siqueira[3], Lara Liane de Queiroz Pereira

[1]Universidade Federal do Piauí, Hospital Universitário, Teresina PI, Brazil.
[2]Instituto Federal do Piauí, Teresina PI, Brazil.
[3]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Marta Maria da Silva Lira Batista; Email: fonointensivista@gmail.com.

Background: Apraxia of speech is a neurological disorder that affects a person's ability to sequence and coordinate the voluntary movements necessary for the production of

speech sounds, while preserving the cognitive linguistic level. The unpredictability and inconsistency of speech errors are a major challenge for rehabilitation. **Objective:** To identify the most efficient and effective computer model for speech recognition in people with acquired apraxia of speech (AOS). **Methods:** Cross-sectional, prospective, quantitative study. Ten people with apraxia were asked to read a list of words (from monosyllables to polysyllables). The audios were evaluated by 11 speech therapists. The final score per audio considered the phonetic and phonological accuracy of the target word, giving a score between 0 and 100. The data was processed by averaging the scores obtained from the form based on the differences between the averages obtained in the descriptive statistics. The 5 main similarity algorithms for speech recognition (Levenshtein; Jaccard; Jaro-Winkler; Hamming; Caracter Error Rate (CER)) have different characteristics for obtaining similarity between two strings. To evaluate the models, the audio and its correct transcription (string) were used; each model used its own algorithm to translate the audio and obtain its correlated result. Statistical correlation (student's t-test) was used to compare the means obtained by the AOS with the expected means. The study was approved by the CEP under report no. 5111083. **Results:** The average of the judges' results was 80%. The average word analysis of the results evaluated by each algorithm was: Hamming (39.1%); CER (41.1%); Jaccard (53.7%); Levenshtein (60.6%): Jaro-Winkler (75.8%). When analyzing the performance of each algorithm separately, it was possible to determine that the least assertive was CER (660.27 variance), which was considered the least accurate and reliable in the analysis of apaxic speech. CER is widely used in automatic speech recognition systems and therefore does not take into account the general intelligibility of the string, which is highly impaired in AOS. However, the Jaro-Winkler algorithm showed the greatest stability, the lowest variance (n=114.90) and the highest accuracy, capturing well the similarity between strings with small transpositions of characters. In the Person correlation test, Jaccard showed a negative correlation when compared to the judges' averages; this can be explained by the phonological changes that occurred between the speakers, and is a limitation of the algorithm (ineffective in analyzing word sequence order; phonological changes). **Conclusion:** Once an efficient model of apraxic speech has been determined, it will be possible to develop more effective applications aimed at speech rehabilitation for this audience.

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Vaccination against polio in Brazilian federal regions from 2014 to 2024: causes, consequences and goals

Luana Cristina de Oliveira Lima[1], Arthur Vinícius Feitosa Santos[1], Vagner Costa da Cunha Júnior[1], Sabrina Sthéphanie Ataíde Martins[1], Maria Cecília Leal Menezes[1]

[1]Universidade Tiradentes, Aracaju SE, Brazil.

Correspondence: Luana Cristina de Oliveira Lima; Email: luana.cristina0410@gmail.com.

Background: Poliomyelitis is an acute contagious disease that can infect children and adults, through direct contact with feces or secretions eliminated through the mouth of sick people and can cause paralysis in the lower limbs in severe cases. Due to declining vaccination coverage over the past 10 years, the Brazil has been classified as very high risk (2022) and high risk (2023) for the reintroduction of wild poliovirus and the emergence of vaccine-derived poliovirus. **Objective:** To describe the drop in vaccinations over a 10-year period, its possible causes, consequences and new national goals. **Methods:** Cross-sectional ecological study, with secondary data from the IT Department of the Unified Health System (DATASUS), the National Vaccination Program (SI-PNI),the SUS Hospital Information System (SIH/SUS) and the Brazilian Institute of Geography and Statistics (IBGE). The variables analyzed are region, Vaccination Coverage (VC) and the human development index (HDI). **Results:** In 2014, national VC reached 96.76%, however the North region was already below the target established by the PNI (95%),with coverage at 90.05%. From 2015 to 2021, the five regions had a decrease in VC, reaching the mark of 71.04%, with the North having the lowest index (62.29%) and the South having the highest (79.98%). Moreover, in the following years, there was a slight increase in vaccination, reaching a coverage of 84.7% in 2023, despite the North, Southeast and Central-West regions maintaining the worst immunization rates, respectively. Regarding the human development index (HDI), of the 10 Brazilian federative units with the highest rates, 7 belong to the regions with the lowest vaccination coverage in 2023, while the Northeast - second highest vaccination coverage index - has the states with the lowest human development indices. **Conclusion:** Therefore,it is noted that,from 2015-2021, vaccination targets were not achieved in any region, a period marked by COVID-19, the reduction of some preventive health services and anti-vaccination movements. From 2022 onwards, the VC increased,however, the targets have not yet been achieved, as the confidence of segments of the population has been affected by the vast circulation of health misinformation on social media. Moreover, on the other hand, it was observed that the regions with the highest HDI had a low VC, which may be related to the strong adherence to the denialist and anti-vaccine movement experienced in recent years, especially in these locations. Regarding regions with lower HDI (North and Northeast), it can be pointed out that there is a correlation between poor immunization and difficulty in accessing public health services. Therefore, it is necessary to raise awareness among the people about the effectiveness and safety of vaccination,informing them about its schedule and the consequences of non-compliance with it, in order to prevent previously eradicated diseases from returning and bringing consequences to the well-being of the society.

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Validation of alternative technologies for gait analysis compared to the reference system in hereditary spastic paraplegia

Diana Maria Cubillos Arcila[1], André Ivaniski Mello[1], Edilson Fernando de Borba[2], Leonardo Alexandre Peyré Tartaruga[3], Jonas Alex Morales Saute[1]

[1]Universidade Federal do Rio Grande do Sul, Porto Alegre RS, Brazil.
[2]Universidade Federal do Paraná, Curitiba PR, Brazil.
[3]Universidade de Pavia, Pavia, Italy.

Correspondence: Jonas Alex Morales Saute; Email: jsaute@hcpa.edu.br.

Background: Hereditary Spastic Paraplegias (HSP) are a group of neurogenetic diseases. The main symptoms are characterized by hyperreflexia, muscle spasticity and weakness that compromises the walking ability. The heterogeneity of these diseases complicates the identification of progression biomarkers; however, spatiotemporal gait parameters are crucial for understanding both normal and pathological gait, making them potential progression biomarkers. Furthermore, Inertial Measurement Units (IMUs), with their portability and ease of use, offer a promising motion capture technology that could aid in identifying the best parameters as progression biomarkers. **Objective:** To evaluate the reliability of an inertial system and a mobile application, compared to a reference motion capture system to capture spatiotemporal parameters in patients with HSP. **Methods:** Twelve patients with PEH were evaluated, 11 of them with SPG4 and 7 with gait independent of assistive devices. Each patient´s gait was assessed using simultaneously a system composed of seven IMUs, a mobile application, and the optoelectronic reference system thought of the timed up and go test at a distance of 5 meters. Each participant performed two trials at self-selected and maximum walking speed. The agreement between systems was assessed using the Bland-Altman statistical method. The parameters were stratified by right and left leg steps, and the effect of using walking aids was analyzed with three groups: general group, patients with independent walking and patients requiring walking aids. **Results:** The system with seven IMUs compared with the optoelectronic reference system showed statistically significant agreement for the overall group when analyzing steps from both right and left sides together on the spatiotemporal parameters. However, when a separate analysis of right and left steps on the spatiotemporal parameters for the overall group was conducted, the inertial systems showed statistically significant agreement only for patients who were independently ambulant. The systems did not agree on the step length parameter for patients needing walking aids. The mobile application was not reliable for evaluating spatiotemporal gait parameters in patients with HSP. **Conclusion:** The inertial system utilized in this study effectively measured spatiotemporal gait parameters in independently ambulant patients with HSP. This portable device has the potential to be used in future clinical trials for evaluating gait in patients with the disease.

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Vascular Parkinsonism due to cerebral cavernomatosis associated with a heterozygous mutation in the KRT1 gene: a case report from a healthcare unit in Amazonas

Emanuelle Campos Amaral[1], Maria Esther Lima Rios Evangelista[2], David França Ferreira Cruz[1], Danilo Lemes Reis[1], Fernanda Almeida Carvalho[1], Manuela Brandão Ferreira Teles[1], Júlia Ágata Cardoso Barbosa[2], Frederico de Sousa Marinho Mendes Filho[2], Paulo Roberto Moss Lopes[2], Marcus Vinícius Della Coletta[1]

[1]Universidade do Estado do Amazonas, Manaus AM, Brazil.
[2]Universidade Federal do Amazonas, Manaus AM, Brazil.

Correspondence: Emanuelle Campos Amaral; Email: eca.med22@uea.edu.br.

Case presentation: Female patient, 70 years old, previously diagnosed with idiopathic Parkinson's disease, with a slow progression of symptoms for about 2 years and intestinal constipation, with a history of sleep disorders for 8 years. On physical examination, the patient presented with bilateral bradykinesia, with a slight predominance on the right, mild bilateral postural tremor, action tremor during line and spiral tests, and mild rigidity on the left and moderate on the right. The first MRI analysis showed multiple low-signal foci on the gradient sequence, with suspected microhemorrhages related to amyloid angiopathy. Upon repeating the MRI for a more recent analysis, multiple and countless supratentorial and infratentorial cavernous angiomas were found bilaterally, as well as in the brainstem, suggesting familial multiple cavernomatosis syndrome as the main diagnostic hypothesis (KRT1 gene mutation). **Discussion:** Cerebral vascular malformations refer to a group of conditions characterized by abnormal vascular configurations occurring within the brain (and spinal cord). Familial CM cases have an autosomal dominant inheritance pattern and are estimated to account for approximately 20 percent of all cases of CMs. Most familial cases have multiple CMs. Pathogenic variants are known to cause familial CMs, with KRIT1 being the variant identified in the examination of the patient described here.Supratentorial CMs commonly present with hemorrhage, seizures, and progressive neurologic deficits. Seizures and progressive neurologic deficits may result from mass effect and secondary compromise of the microcirculation or from microhemorrhages with local hemosiderin deposition irritating cortical or subcortical

tissue. Infratentorial CMs commonly present with hemorrhage and progressive neurologic deficits. Lesions in the brainstem present with cranial neuropathies and long-tract signs that cause progressive neurologic decline due to the high volume of critical nuclei and fiber tracts in this area. Notwithstanding these common symptoms, the patient in this case presented the evolution of the main complaint with a classic picture of idiopathic Parkinson's syndrome. With the typical exome of cerebral cavernomatosis due to an alteration in the KRIT1 gene, the patient demonstrates an atypical clinical picture rarely found in the literature regarding the correlation of this pathology with Parkinsonian syndrome, highlighting the rarity and relevance of the case. **Final comments:** As demonstrated in Neurology, there are clinical cases that can present with atypical manifestations. It is important not to limit oneself and to seek differential diagnoses, highlighting the importance of complementary examinations such as magnetic resonance imaging and, especially in this case, genetic testing. Despite the limitations within the state of Amazonas, it was possible to report an atypical manifestation of parkinsonism of vascular origin due to cerebral cavernomatosis.

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VCP gene mutation: muscle disorder or nerve disorder?

Luma Santos de Oliveira[1], Fabio Victor Vieira Rocha[1], Emanuelly Victoria Andrade Fonseca[1], Igor Flavio Gonçalves do Vale[1], Daniel Mariani Favalessa[1], Bruno Batitucci Castrillo[1], Renann Nunes Pirola[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Luma Santos de Oliveira; Email: luma.oliveira@edu.ufes.br.

Case presentation: A 64-year-old male patient with a decade-long history of progressive and asymmetric proximal and distal crural paraparesis, along with foot paresthesias and significant lower back pain, without any cognitive complaints. Neurological examination showed muscle strength (D/E) of II/I for hip flexion, IV/III for knee extension and flexion, and IV/III for ankle dorsiflexion. Achilles reflexes were absent, with painful hypoesthesia and hypopallesthesia in a glove-and-stocking pattern. Lab results indicated a CPK of 293 and alkaline phosphatase of 259. Electroneuromyography revealed severe, length-dependent axonal sensory-motor polyneuropathy. The patient declined a muscle biopsy. MRI and CT of the lumbar spine showed vertebral cortical and trabecular thickening with uptake. Bone scintigraphy indicated increased bone uptake with high osteoblastic activity. Genetic testing confirmed a mutation in the VCP gene (R93C), compatible with Charcot-Marie-Tooth disease type 2Y (CMT2Y). **Discussion:** Autosomal dominant mutations in the VCP gene lead to a multisystem proteinopathy that can present with inclusion body myositis, Paget's disease of bone, and frontotemporal dementia. Ubiquitin-positive protein aggregates are a key pathological characteristic. In rare cases, mutations in this gene are also linked to CMT2Y. The patient shows confirmed polyneuropathy and Paget's disease of bone. Although myopathy hasn't been definitively confirmed, the clinical presentation of initial proximal weakness and slightly elevated CPK levels suggests a possible overlapping myopathy. The genotype-phenotype correlation shows significant clinical variability, even among family members with the same mutation. Until now, CMT2Y has been linked only to the E185K mutation, while this patient has the R93C mutation. **Final comments:** This case illustrates the complex genotype-phenotype relationships in VCP gene mutations. The patient's progressive crural paraparesis, polyneuropathy, and Paget's disease of bone are consistent with VCP-related conditions. This emphasizes the need for comprehensive genetic testing and clinical assessment in patients with diverse multisystem symptoms to fully understand the range of VCP-related disorders

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Velpeau's Law in practice: consecutive diagnoses of inclusion body myositis on the same day

Igor Flavio Gonçalves do Vale[1], Fernanda Bulhões Pagotto Pádua[1], Lucas Brumatti Setubal[1], Renann Nunes Pirola[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Igor Flavio Gonçalves do Vale; Email: igor.f.vale@edu.ufes.br.

Case presentation: Velpeau's Law, described by Alfred Velpeau, posits that rare diseases tend to manifest two or more times in a short interval. This observation was corroborated by the evaluation of two female patients, with an average age of 52 years, who presented with progressive asymmetric weakness in the proximal muscles of the lower limbs (LL), worse on the left, and varying degrees of bulbar involvement, progressing for more than five years, who were seen on the same day. They also reported distal weakness in the upper limbs (UL), with an emphasis on the deep flexors of the fingers (DFPs), associated with atrophy of the flexor muscles of the forearm. Magnetic resonance imaging (MRI) of the LL revealed predominant hypotrophy and fatty replacement in the bilateral quadriceps femoris in the first patient, while the second showed muscle atrophy with adipose degeneration in the posterior compartment of the thigh.Creatine kinase (CK) levels were 449 U/L and 3220 U/L, respectively. Neuromuscular ultrasound (NMUS) revealed a hyperechoic pattern in the quadriceps femoris muscles in both patients, indicating atrophy and fatty replacement in this region, sparing the rectus femoris. **Discussion:** Inclusion body myositis (IBM), with a prevalence of 24.8 to 46 cases per million inhabitants and a male predominance (2:1) after age 45, is a subtype of idiopathic inflammatory myopathy characterized by asymmetric and progressive weakness of the appendicular muscles. The weakness is predominantly proximal in the LL, primarily affecting the quadriceps femoris muscle, and distal in the UL, especially the deep flexors of the fingers (DFPs) and flexor pollicis longus (FPL). Additionally, there is the presence of bulbar dysfunction, which progresses to dysphagia in 70% of cases. Due to the insidious and progressive nature of the disease, the diagnosis, which is based on clinical presentation, elevated CK levels, less than 12 times the reference value, and histopathological findings such as rimmed vacuoles and p62 aggregates, or MRI findings of muscle atrophy and fatty replacement of the quadriceps femoris, usually takes three to five years to establish. However, NMUS has emerged as a promising diagnostic tool, with 98% specificity and 82% sensitivity. It allows the detection of severely affected muscles, such as the gastrocnemius and, primarily, the DFP, which is the most discriminative muscle for IBM diagnosis. This technique also shows relative preservation of the adjacent flexor carpi ulnaris muscle, enabling a useful comparison in a single image, minimizing the need for invasive and high-cost procedures. **Final comments:** In the context of a rare disease with a long diagnostic window, NMUS has the potential to become an extension of the bedside physical examination, strengthening clinical reasoning and enabling a faster diagnosis. In line with Velpeau's Law, two women were consecutively seen and diagnosed with inclusion body myositis, facilitated by NMUS.

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Venlafaxine versus amitriptyline in the prophylactic treatment of migraine: an updated systematic review

Pedro Vitor Ferreira Rodrigues[1], Leonardo José Rodrigues de Araújo Melo[1], Francisco Luciano Honório Barreto Cavalcante[1], Paulo Victor de Santiago Gonçalves[1], Cássia Caroline Aguiar da Ponte[1], Pedro Lucas Grangeiro de Sá Barreto Lima[1]

[1]Universidade Federal do Ceará, Fortaleza CE, Brazil.

Correspondence: Pedro Vitor Ferreira Rodrigues; Email: pedrovitorf7@gmail.com.

Background: The current migraine pharmacological prophylaxis consists mainly in the use of beta-blockers, tricyclics antidepressants and anticonvulsants as first-class drugs for the reduction in the number and severity of headache events. However, many physicians had used Venlafaxine (VLF), a selective serotonin and norepinephrine reuptake inhibitors, as option in the treatment of this headache type without a relevant quantity of data about the benefit of this medication compared with the traditional treatment. **Objective:** To evaluate and compilate the data about the efficacy of Venlafaxine versus Amitriptyline (AMT) in migraine prophylaxis treatment. **Methods:** This study was conducted using the Preferred Reporting Items for Systematic Review and Meta-analyses (PRISMA) guidelines. The search was conducted in the PUBMED/MEDLINE, EMBASE and COCHRANE databases. Only articles comparing directly Venlafaxine with amitriptyline were included. Any papers in languages other than English were excluded. The screening was done by 2 independent authors, which selected the articles for this study based on the PICO process (Migraine; Venlafaxine; Amitriptyline; Frequency, Severity, Pain and Adverse effects) in which 1083 articles were selected for screening, with 289 of them being excluded as duplicates and 790 for not meeting the eligibility criteria defined in the formulation of the study. At the end, 3 articles were selected for compose this systematic-review. **Results:** A total of 284 patients were included in this review, with 142 in the venlafaxine group. The follow-up time ranged from 8 to 24 weeks. The mean dose of venlafaxine was of 75mg, and 25mg of Amitriptyline was used as comparison. Only 2 articles showed a statistical significance in the difference between the two drugs, with a mean reduction of 5,4 attacks per month ($p< 0.0001$) for reduction of migraine frequency in the venlafaxine group. There was no statistical difference between the drugs in the headache duration ($p> 0.05$). One study showed a remarkable reduction in the pain by the Visual Analog Scale in the VLF group. Two studies had no difference between the severity decrease in the drugs comparison. Most of the adverse effects cases were of patients using venlafaxine, with sleep disturbances being the principal side effect related. A meta-analysis was not done by the heterogenicity of the data. **Conclusion:** In our analysis there was no real difference ($p>0.05$) between the two drugs in the duration and severity of the migraine attacks, showing no likely vantage in the choice of one drug instead of the other in terms of efficacy. However, the benefit expressed by one of the articles in the reduction of pain by venlafaxine and the high incidence of adverse effects, shows the necessity of more studies comparing the two pharmacological therapies for a better analysis and efficacy study.

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Ventricular papilloma in a pregnant patient: case report

Lianna da Silva Facco[1], Luidia Giacomini[2], Milena Moraes[1], Ana Flávia Orlandini[1], Israel Farias Silva[1], Giovanni Cândido Volino[1], Guilherme Weimer Vilanova Audino[1], Laura Martens Fischer[1], Matheus Felipe Kuhn Urnau[1], Charles André Carazzo[2]

[1]Universidade de Passo Fundo, Passo Fundo RS, Brazil.
[2]Hospital São Vicente de Paulo, Passo Fundo RS, Brazil.

Correspondence: Lianna da Silva Facco; Email: lianna.facco@gmail.com.

Case presentation: A 28-year-old woman, 38 weeks pregnant and with a history of migraines without attacks for 2 years, was transferred to our institution complaining of a severe occipital headache that started the day before. She reported intense unilateral pain in the occipital region, which started suddenly in the early hours of the day of admission, accompanied by vomiting and prostration. The patient had photophobia and blurred vision. The pain worsened with movement and improved with pressure applied to the back of the neck. There have been reports of seizures at the hospital of origin. On physical examination, the patient was alert but prostrate, with no signs of fever, preserved pupillary reflexes, a positive bilateral Hoffmann reflex, and pain on palpation in the cervical region. Owing to the warning signs (sudden and intense headaches in pregnant women), magnetic resonance imaging (MRI) with TOF sequence was requested for investigation. The following day, the test results showed an expansive lesion in the fourth ventricle, with signs of acute bleeding and increased dimensions compared with previous tests. A cesarean section was performed on the same day due to concerns about the initial signs of cranial hypertension, with a contraindication to spinal anesthesia. On the days following her hospitalization, the patient was diagnosed with ventricular papilloma and underwent surgery to resect the tumor of the fourth ventricle with intratumoral hemorrhage. After the procedure, the patient showed progressive improvement without recurrence of headaches or other symptoms. Subsequent exams confirmed tumor removal and a reduction in intratumoral bleeding. The patient was discharged 12 days after hospitalization, with outpatient follow-up instructions. **Discussion:** Ventricular papillomas are a rare condition, being more common in children, representing 1-4% of all pediatric brain tumors, even though they can appear at any age. In children, they normally affect the internal regions of the lateral ventricles. In adults, there is a preference for the fourth ventricle. They are divided into choroid plexus papilloma (CPP), atypical choroid plexus papillomas (APP), and choroid plexus carcinoma (CPC). The presence of simian virus 40 in the coding regions of some genes appears to produce a tumor antigen (TAG) that can interact with tumor suppressors, p53 and pRb, forming a complex, which appears to be an important factor in the formation of these types of tumors. **Final comments:** This case illustrates the complexity of the differential diagnosis of headaches in pregnant women, emphasizing the importance of thorough investigation in the face of serious symptoms, such as sudden and intense headaches. Early identification of warning signs and prompt intervention are crucial to avoid serious complications for both the mother and the fetus.

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Verheij syndrome: a case report of a differential diagnosis of autism

Rachel Leirner Argelazi[1], Gabriel Salmen Lobo Gonçalves[2], Santhiago Calvelo Graça[1], Marcella Gonçalves Ferreira[2], Hugo Raphael Resende Cruz[2], Gabriela Ribeiro do Val[2], Yasmim Mendes Silva[2], Raquel Monico Cavedo[2], Hugo José de Carvalho Garcia dos Santos[2], Laís Russo Carneiro Peruzzi[2]

[1]Santa Casa de São Paulo, Faculdade de Ciências Médicas, São Paulo SP, Brazil.
[2]Irmandade da Santa Casa de Misericórdia de São Paulo, São Paulo SP, Brazil.

Correspondence: Rachel Leirner Argelazi; Email: racheleirner@gmail.com.

Case presentation: HBS, a 4-year-old, attended his first pediatric neurology consultation in May 2024 at Hospital Santa Casa de Misericórdia in São Paulo for neuropsychomotor development delay. He is the only child of a healthy, non-consanguineous couple with no prenatal complications. At birth, he had dysphagia and laryngomalacia, requiring speech therapy. Milestones included head control at 3 months, sitting with support at 9 months, walking at 1 year and 6 months, and speaking first words at 2 years. Despite delays, he has attended school for a year and a half, participates in physical activities, sleeps well, and maintains good family relationships. Mild autism was suspected. During evaluation, he was alert, cooperative, age-appropriate cognitively, oriented, and had coherent speech. His gait was atypical, and muscle strength, coordination, balance, tone, and trophism were adequate, with no involuntary movements or neurological issues. He had dysmorphic facial features, including large ears, a thin face, and almond-shaped eyes, leading to consideration of a genetic syndrome. Exome sequencing revealed a PUF60 gene alteration, diagnosing Verheij Syndrome, a new mutation with a mild presentation. **Discussion:** Verheij syndrome, or PUF60-related developmental disorder, is a rare genetic condition caused by mutations in the PUF60 gene, identified through genetic testing like whole exome sequencing. This gene is crucial for proper mRNA processing and protein synthesis. The syndrome is usually linked to de novo heterozygous loss-of-function mutations in the PUF60 gene. In our study, the patient had the c.245_246del; p.(Val82Alafs*57) variant found in heterozygosity, which causes a premature stop codon and likely results in protein degradation by the Nonsense Mediated Decay (NMD) pathway. Verheij syndrome is characterized by a range of congenital anomalies and developmental issues, including ophthalmic coloboma, short stature, skeletal abnormalities, congenital abnormalities, developmental delay, palatal abnormalities, congenital heart and kidney defects and facial dysmorphims. There is significant variability in the presentation of Verheij syndrome. Some individuals may have milder forms with fewer features, while others have more severe manifestations. The reported patient presents a mild form of the syndrome. Despite developmental delay and typical phenotype, the child is able to develop and engage in school, sports, and social activities. **Final comments:** This is one of the first cases of Verheij syndrome in the state of São Paulo, and it is even rarer due to its mild presentation. The differential diagnosis of genetic syndromes in suspected autism cases is essential for proper diagnosis and management.

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Vertebral artery dissection as an etiology of intense headache in the post-operative surgery of elective plastic surgery: a case report

Pedro Igor Lustosa Roriz[1], Gabriela Sebastiana Barbosa Sá[1], Eloismaile Leite Ferraz[1], Ádrya Aryelle Ferreira[2], Cleilson Barbosa de Freitas[2], Victor Ribeiro Neves[2]

[1]Instituto de Educação Médica, Juazeiro BA, Brazil.
[2]Universidade de Pernambuco, Petrolina PE, Brazil.

Correspondence: Pedro Igor Lustosa Roriz; Email: pedroilroriz@gmail.com.

Case presentation: Female patient, 32 years old, with no history of previous chronic illness or continuous use of medication. No history of smoking or alcohol consumption. On the first day after elective aesthetic plastic surgery on her abdomen and breasts, shortly after being discharged from the hospital, she developed a severe headache, which reached its peak after about 15 minutes. The headache was bilateral, pulsating, intense and worsened with any effort. In addition, she presented photophobia, phonophobia, nausea and mild and persistent neck pain. The headache was even more exacerbated in the supine position and there was a slight improvement in orthostasis. The patient was hospitalized again to control her symptoms. Initially, a head tomography was requested, which showed no changes and was treated only with symptomatic medication when a migraine was suspected. The next day, she was evaluated by the hospital's neurology team for persistent headaches and diplopia. The neurological examination revealed only abduction deficit in the right eye, suggesting possible involvement of the right sixth cranial nerve. The examination did not highlight neck stiffness, fundus changes, or other cranial nerve involvement. Although the brain resonance showed no changes that would justify the pain (also without an area of restriction to water diffusion), the angioresonance showed an image suggestive of dissection in the right vertebral artery. **Discussion:** The importance of remembering the warning signs in the evaluation of headaches must be highlighted to exclude differential diagnoses of more severe syndromes that may pose more serious risks to patients. At the beginning of the case, the patient had no previous history of headache refractory and high-intensity pain as warning signs. **Final comments:** In this case, the surgeon performed the aesthetic procedure away from the cervical region (breast and abdomen). It is possible to relate the appearance of cervical vascular injury to patient manipulation in bed or to the spontaneous occurrence of the disease. However, it cannot be ruled out that the surgical procedure contributed to the emergence of the problem.

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Vertigo induced by unilateral hippocampal infarction: a case report

Maria Eduarda Andreatta[1], André Luiz Pereira Martins[1], Joana Laurindo da Silva[1], Olga Cassol Silva[1], Carolina Aparecida Donofrio Santos Costa[1], Mariana Costa Borges[1], Cristine Koehler[1], Rafael Marques Mendes[1], Gabriela da Silva Cremonese[1], Gladys Lentz Martins[1]

[1]Hospital Governador Celso Ramos, Florianópolis SC, Brazil.

Correspondence: André Luiz Pereira Martins; Email: martinsandrelp@gmail.com.

Case presentation: A 64-year-old female presented to the emergency department with sudden-onset vertigo. There were no previous similar episodes, nor any history of weakness, speech changes, tinnitus or hearing loss. She denied recent head or neck trauma. Her medical history included systemic arterial hypertension. On examination, she displayed horizontal nystagmus with a rightward fast phase induced by rightward gaze, asymmetric between both eyes, non-fatigable and with no latency, upbeat vertical nystagmus and dysmetric rightward saccades. Positive skew deviation and a negative head impulse test were noted. Also, she had a positive Romberg test and cerebellar ataxic gait. An angiotomography revealed an old ischemic lacuna in the right cerebellar hemisphere and ruled out occlusion and stenosis. A follow-up magnetic resonance showed two foci of diffusion restriction in the right hippocampus, confirming a subacute ischemic event. Treatment included aspirin, clopidogrel and metoclopramide, leading to symptomatic improvement. Transthoracic echocardiogram and electrocardiogram were unremarkable. Laboratory findings showed dyslipidemia. The patient was discharged for outpatient follow-up with further investigations for secondary prevention. **Discussion:** Acute vestibular syndrome is characterized by the rapid onset of vertigo accompanied by nausea, gait unsteadiness, and nystagmus lasting a day or more. Its primary cause is usually peripheral vestibulopathy, such as vestibular neuritis. Nonetheless, vertigo can manifest as early symptoms of acute stroke, often involving the posterior circulation. In the central nervous system, lesions typically responsible for vertigo are often found in the brainstem. Conversely, there is limited research on whether lesions in the supratentorial hemispheres can lead to vertigo. Brain studies in humans have revealed several vestibular cortex areas that includes the insular area, the superior temporal gyrus, the precuneus, the anterior cingulate gyrus and the hipoccampus. Although not uncommon, literature on hippocampal infarction remains sparse. Increasing

evidence supports the existence of a lateralized vestibular cortex, suggesting that cortical strokes in the right hemisphere are more likely to cause vestibular symptoms compared to the left hemisphere. Hippocampal infarctions can lead to persistent anterograde and retrograde amnesia, mental status changes and, more rarely, vertigo. Treatment options differ; anticoagulants or antiplatelet agents are prescribed for secondary prevention of hippocampal infarctions, based on the underlying cause. **Final comments:** It is essential to remember that acute central vestibular syndrome can arise from not only the brainstem but also supratentorial structures. We present a rare case of a woman who developed an acute central vertiginous syndrome due to a hippocampal infarction. To our knowledge, this is the first reported case of a hippocampal stroke presenting with vertigo.

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Very late-onset myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD): a case report

Lara Santos Machado[1], Larissa Braga Mendes[1], Fernanda Lustosa Cabral Gomez[1], Luiz José Malta Gaia Ferreira Júnior[1], Lucas Toshio Uenishi[1], Nilton Amorim de Souza[1], Enedina Maria Lobato de Oliveira[1]

[1]Universidade Federal de São Paulo, São Paulo SP, Brazil.

Correspondence: Lara Santos Machado; Email: laramachado231@gmail.com.

Case presentation: A 73-year-old woman sought care for vision loss in the right eye, which progressed to the left eye in one day. She presented at the same time with paresthesia and progressive weakness of her legs, becoming unable to walk in 15 days, associated with urinary incontinence and constipation. At presentation, she had right RAPD, had a visual acuity of hand movement in the right eye, and could count fingers at 10 meters with the left eye. She had weakness of both legs, hypoesthesia with sensory level at T4, and diffuse pathologically brisk reflexes, with bilateral Babinski sign. She had been diagnosed with chronic lymphocytic leukemia (CLL) without treatment 5 years before. T2 FLAIR brain and spinal cord MRI showed a subcortical hyperintense signal in the right parietal lobe and the cervicothoracic region, with gadolinium enhancement and a cerebellar lesion. The cerebrospinal fluid analysis presented 7.3 white blood cells/μL with 95% lymphocytes and an increased protein concentration of 51 mg/ dL. Oligoclonal bands and immunophenotyping were not available. Infectious, rheumatology and a paraneoplastic investigation were negative. She tested negative for anti-AQP-4 and positive for anti-MOG antibodies. The patient was treated with intravenous methylprednisolone, followed by intravenous immunoglobulin and high daily doses of corticosteroids, with partial improvement of symptoms. The patient presented a recurrence of symptoms after weaning from corticosteroid therapy. Azathioprine, rituximab, and venetoclax treatment were started due to the possibility of autoimmune complications of CLL. **Discussion:** A very late-onset inflammatory demyelinating event (VLO-IDE) of the central nervous system is an onset after age 70. Degenerative, neoplastic, paraneoplastic, granulomatous, rheumatologic, and infectious lesions must be excluded. Given the possibility of an inflammatory etiology, the institution of immunotherapy treatment should not be delayed. Demyelinating diseases include very late-onset clinically isolated syndrome (VLO-CIS), multiple sclerosis (VLO-MS), neuromyelitis optica (VLO-NMO), or myelin oligodendrocyte glycoprotein antibody-associated disease (VLO-MOGAD). MOGAD can present with optic neuritis, transverse myelitis, acute disseminated encephalomyelitis, cortical encephalitis, brainstem encephalitis, seizures, and meningitis. Elderly patients present subacute evolution and decreased risk of recurrence. There is no report of MOGAD associated with CLL, but an NMO-like presentation as a CNS complication of CLL has been described. **Final comments:** A clinical inflammatory demyelinating event in elderly patients is rare and must be extensively investigated. Neoplastic, paraneoplastic, and infectious etiologies must be discarded. With population aging, attention must be paid to the possibility of VLO-IDE, including MS, NMO, and MOGAD.

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Very long-chain acyl-CoA dehydrogenase deficiency: an illustrative case of late manifestation inborn error of metabolism

Helena Fussiger[1], Giuseppe Dick Bonato[1], William Alves Martins[1], Marina Scop Medeiros[1], Rodrigo Rezende Arantes, Lilia Farret Refosco, Bibiana Mello Oliveira

[1]Pontifícia Universidade Católica do Rio Grande do Sul, Hospital São Lucas, Porto Alegre RS, Brazil.

Correspondence: Giuseppe Dick Bonato; Email: g.d.bonato@hotmail.com.

Case presentation: A 28-year-old female with a history of recurrent nausea and vomiting since childhood exacerbated by physical exertion, temperature changes, and prolonged fasting, notably worsened over the past year. At age 27, she experienced three episodes spaced approximately 20 days apart, preceded by fasting, characterized by vomiting, lethargy, and acute epileptic encephalopathy necessitating ICU admission. Episodes also featured hypoglycemia, muscle weakness, deteriorating renal function, metabolic acidosis, elevated transaminases, hyperCKemia, and hyperammonemia. The unusual clinical presentation led to consideration of psychiatric diagnoses, including suicidal attempts, prompting outpatient psychiatric evaluation. There was no parental consanguinity, but a maternal and paternal cousin died at 22 years old from gastroenteritis and fulminant hepatic failure. Physical examination revealed proximal muscle weakness and predominantly distal hyporeflexia. During the third episode of acute encephalopathy, investigations revealed cardiomyopathy, metabolic acidosis, hyperlactatemia, hypoglycemia, elevated CPK, ammonia, transaminases, and bilirubin. Liver biopsy indicated cirrhosis with a cholestatic pattern; EMG suggested axonal sensory-motor neuropathy. Brain MRI and cardiovascular assessment were normal. Acylcarnitine profiling showed deficiencies in C0 and elevated organic acids, specifically 4-OH-phenylacetic and 4-OH-phenylpyruvic acids. Molecular analysis using a treatable diseases panel identified likely pathogenic variants p.Lys382Serfs*11/p.Glu285Gln in the ACADVL gene, confirming VLCAD deficiency. Treatment recommendations included avoiding prolonged fasting, limiting long-chain lipids, consuming complex carbohydrates, and initiating L-carnitine supplementation along with triheptanoin. These interventions led to clinical improvement with no acute episode recurrence within 4 months post-treatment initiation. An emergency management plan for clinical incidents was established, and family investigation continues. **Discussion:** Inborn errors of metabolism (IEM) are rare genetic disorders typically appearing in childhood due to enzymatic or transport deficiencies affecting specific metabolic pathways. Very long-chain acyl-CoA dehydrogenase deficiency (VLCAD) is a disorder within fatty acid beta-oxidation, impairing the conversion of very long-chain fatty acids into energy. This condition predisposes individuals to encephalopathy, cardiomyopathy, hepatopathy, and hypoglycemia, inherited in an autosomal recessive manner. **Final comments:** This case underscores the severe manifestation of IEM in adulthood, presenting significant diagnostic and therapeutic challenges. The definitive diagnosis followed a prolonged investigative journey, enabling targeted therapies aimed at reducing disease-related morbidity and mortality.

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Vestibular paroxysmia: a case report

Bibiana Mayer[1], Chadi Emil Adamo[2]

[1]Universidade de Ribeirão Preto, Ribeirão Preto SP, Brazil.
[2]Universidade de São Paulo, São Paulo SP, Brazil.

Correspondence: Bibiana Mayer; Email: bibiana_mayer@hotmail.com.

V.T.D., male, 50 years old, electrical engineer. He reported acute pain in the cervical region followed by episodic, non-positional subjective vertigo lasting about 30 seconds over 2 weeks. He underwent an otolaryngological examination, which did not detect any peripheral causes. He received antivertigo medication without improvement. Neurological examination revealed no cranial nerve abnormalities except for a Weber test lateralized to the left with a positive Rinne test. The head impulse test was negative. The Fukuda test showed lateralization to the left and slight asymmetry of reflexes, with the left reflex stronger than the right. Deep tendon reflexes were normal, with no posterior cord abnormalities. Cerebellar, motor, and sensory tests were preserved. During hyperventilation maneuvers, the vertigo episode was reproduced after about 45 seconds. He underwent cranio-cervical MRI and vascular studies, which showed no areas of ischemia and ruled out vertebral artery dissection and abnormalities in the cranio-cervical junction (i.e., foraminal stenosis). The investigation was complemented with an MRI using FIESTA sequences and a mastoid MRI. During this period, he experienced additional episodes of paroxysmal vertigo and worsening of unilateral pulsatile tinnitus, not synchronous on the right. Upon re-evaluation, tortuosity of the V4 segment of the left vertebral artery was observed, conflicting with the emergence of the eighth cranial nerve on the same side, compressing the superolateral portion of the pons on the left. The right vertebral artery was hypoplastic (2.2 mm in diameter) and joined the basilar artery (4.45 mm in diameter), projecting axially to the right. The mastoid MRI showed dehiscence of the right superior semicircular canal. The patient was treated with carbamazepine, optimized to a dose of 400 mg/day, leading to symptom resolution. Antivertigo medications were discontinued. Given the above situation, the patient met the criteria for a condition known as Vestibular Paroxysmia, in this case, secondary to compression of the vestibulocochlear nerve by dolichoectasia of the basilar arterial system. The neurophysiopathology of this disease is attributed to neurovascular compression, similar to trigeminal neuralgia; ephaptic transmission has also been proposed as a mechanism. Dedicated MRI of the internal auditory canal may reveal unilateral cross-compression of the anterior inferior cerebellar artery (AICA) or its branch and the eighth cranial nerve complex, but clinical and radiological correlation is not always evident. Certain characteristics define the syndrome, such as: short, very frequent, and intense attacks of rotational or to-and-fro vertigo lasting seconds, sometimes milliseconds; attacks often triggered by specific head positions; permanent hearing loss or hearing loss during an attack (often associated with "typewriter tinnitus"); audiovestibular deficits on testing; and exclusion of a central cause.

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vHIT: a new method for vestibular function testing

Carolina Oldoni[1], Marcelo Tohoru Nakamura[1], Fernando Henrique Magri Alves[1], Emanuelle Roberta da Silva Aquino[1], Cristiana Borges Pereira[1]

[1]Universidade de São Paulo, Faculdade de Medicina, Hospital das Clínicas, São Paulo SP, Brazil.

Correspondence: Carolina Oldoni; Email: carololdoni15@gmail.com.

Case presentation: The video Head Impulse Test (vHIT) began to gain popularity around 2009 as a possible method to evaluate the function of each semicircular canal (SCC) individually (in pairs, right and left sides) non-invasively, and it is increasingly being used for the assessment of peripheral vestibular disorders. Prior to this, the expensive, complex, ingenious, and semi-invasive scleral search-coil systems were used. In many places, the vHIT has become the preferred initial test over caloric testing for patients suspected of having peripheral vestibular lesions, considering that caloric testing is quite uncomfortable for the patient and its results can vary depending on the examiner and the technique used (air versus water). The vHIT, on the other hand, is a quick, non-invasive test where the patient is subjected to rapid, passive, unpredictable head movements (about 15º, 100º/s) while looking at a fixed target. A device attached to the patient's head measures the head movements performed in the planes corresponding to the SCC action planes and the associated eye movement through a laser directed at the pupil's center of mass. These devices, linked to the supplier's software, generate graphs showing SCC function (by measuring the "gain") and the presence or absence of compensatory saccades, with the added advantage of recording covert saccades, i.e., eye movements that are not visually noticeable as they occur during head movement but are captured by the eye movement sensor. These covert saccades provide additional information, especially about the recovery of vestibular function. **Discussion:** This case aims to illustrate the use of a new complementary examination method for vestibular disorders, the vHIT. Here we describe the application of this test in patient A.M.R., a 50-year-old man diagnosed with superior branch right vestibular neuritis, with a vHIT showing reduced gain in the right anterior and lateral canals and the presence of both covert and overt compensatory saccades (test performed about 40 days after the ictus). Additionally, patient M.C.T., a 60-year-old woman diagnosed with a right posterior medullary stroke (likely involving the inferior vestibular nucleus), had a normal vHIT. The vHIT plays a primary (but not exclusive) role in differentiating peripheral from central vestibular diseases (when there are no other neurological examination findings aiding clinically in this differentiation), in conditions such as vestibular neuritis. **Final comments:** The vHIT emerges as another option for complementary vestibular function testing, this time in a non-invasive, portable manner, with minimal discomfort for the patient. There is still much to be understood about the alteration patterns presented in the vHIT, not only for diagnosis but also for prognosis and evaluation of vestibular function recovery.

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Vibratory stimulation system for reducing diabetic neuropathy associated sequelae

Laura Caroline Felipe de Freitas[1], Sérgio Ricardo de Jesus Oliveira[1], Mariana de Oliveira[1], Samuel de Oliveira Lino[1], Leonardo Bongiovani Batista de Souza Figueiredo[1], Felipe Rodrigues Moreira dos Santos[1], Daniel Botelho Silveira[1], João Vitor Silva[1], Pedro Luís Mendonça Silva[1]

[1]Universidade Federal de Uberlândia, Uberlândia MG, Brazil.

Correspondence: Laura Caroline Felipe de Freitas; Email: lauracffreitas03@gmail.com.

Background: Diabetic neuropathy is a Diabetes Mellitus complication associated with disorders of the nervous system, which can affect motor, sensory and autonomic functions. As a result of sensory loss in the foot region, people with diabetic neuropathy are at greater risk for developing ulcerative lesions. The impairment of tactile feedback associated with the increased vulnerability of people with diabetes to infectious processes suggests the need for strategies capable of helping the patient to perceive the lesions, so diligent treatment can be scheduled. **Objective:** To test a system composed of tactile insoles, which will be used to monitor the level of pressure on the plantar surface of the feet in volunteers with diabetic neuropathy. In conjunction with the insole, a mechanical vibration stimulator will convert the signals received from the tactile sensors into a pattern of vibratory stimuli. The intensity and location of the stimuli will be defined depending on the location and severity of pre-existing injuries. **Methods:** This study is a clinical trial and will longitudinally and prospectively follow an experimental and a control group. Both will be defined based on the following inclusion criteria: being 25 years old or over, having diabetic neuropathy and presenting symptoms compatible with the syndromic condition of diabetic foot. Volunteers who are absent from experimental sessions and who do not use the device consistently enough will be excluded from the study. The follow-up rate for each participant will be 10 days and data collection will be carried out every 5 days. The data obtained from the collections will be useful in evaluating the history of pressure values at specific points on the insole, in order to observe whether the stimulation system can be effective in preventing the worsening of previously existing injuries due to mechanical improvements. The normality of the data obtained for each group will be verified using the Kolmogorov-Smirnov test. Appropriate parametric or non-parametric statistical tests will be used according to the normality of the variables, considering a significance level of 5%. **Results:** This is an ongoing study. It is speculated that increasing the perception of stimuli through the use of the device during the time of segments of the volunteers in the experimental group could be effective in n reducing pressure values over ulcerative lesions, enhancing the healing process and predisposing to an optimization of the time required for resolution. **Conclusion:** From this study, we hope to find an alternative to improve, in a non-invasive way, diabetic neuropathy carriers quality of life. Thus, it would be possible to contribute to reducing the morbidity associated with the disease and its complications.

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Viral encephalitis: incidence and hospital morbidity in Brazil

Larissa Bianca Borsone[1], Deborah Araujo Silva[1], Rynele Almeida de Fonseca[1], Annayza de Oliveira Bessa[1], Amanda Oliva Spaziani[1], João Carlos Bizinotto Leal de Lima[1], Vanessa Maria Gonçalves de Souza[1], Francieli Gomes Lima[2], Luis Carlos Spaziani[3], Gustavo Henrique da Silva[4]

[1]Universidade Brasil, Fernandópolis SP, Brazil.
[2]AME Fernandópolis, Fernandópolis SP, Brazil.
[3]Fundação Educacional de Fernandópolis, Fernandópolis SP, Brazil.
[4]UPA Santa Fé do Sul, Santa Fé do Sul SP, Brazil.

Correspondence: Larissa Bianca Borsone; Email: larissa.borsone@hotmail.com.

Background: Viral encephalitis is an inflammatory process of the brain parenchyma associated with clinically manifested neurological dysfunction. Viral encephalitis generally occurs following hematogenous viral dissemination into the central nervous system. The most common causative agents are herpes viruses, non-polio enteroviruses, and arboviruses. Although relatively rare, it is a clinical condition with high morbidity and mortality rates. Symptoms and signs of viral encephalitis can range from fever, headache, cerebrospinal fluid pleocytosis, and abnormal MRI findings to altered mental status, severe encephalopathy, seizures, and permanent neurological deficits. **Objective:** To evaluate the profile of hospitalizations due to viral encephalitis in Brazil between the years 2019 and 2023. **Methods:** Data collection was conducted using publicly available information from the Hospital Information System of the Brazilian Unified Health System (SIH/SUS) in the Tabnet/DATASUS database between January 15 and May 15, 2024. This study employed a retrospective longitudinal design with a quantitative descriptive approach. Data were grouped by Brazilian macro-regions, type of care, sex, age group, and race/ethnicity. Statistical inference was performed using BioEstat 5.3 software, utilizing paired T-tests for related samples to compare groups, and descriptive statistics, measures of central tendency, and dispersion were used to present the data. **Results:** Between 2019 and 2023, there were 9,486 hospitalizations for viral encephalitis in Brazil. The majority, 53.61% (n=5,085), were male (p<0.0038), and 51.03% (n=4,841) were of mixed race (p=0.0039). The age group most affected was 1-4 years, accounting for 17.37% (n=1,648; p<0.0001) of the cases. The Northeast region had the highest concentration of cases (n=3,890), corresponding to 40.99% (p=0.0004), with an average annual treatment cost of R$ 7,658,211.31. Mortality was most prevalent in the Northeast, with 38.13% (n=188) of deaths (p=0.0016), and a mortality rate of 10.16 (p=0.2311) in the South region. The number of hospitalizations peaked in 2019, accounting for 25.68% (n=2,436) of the total, compared to only 21.79% (n=2,067) in 2023. The majority of hospitalizations, 51.03% (n=4,841; p<0.0039), were classified as urgent. **Conclusion:** Hospitalizations due to viral encephalitis are predominantly concentrated in the Northeast region of Brazil, with a higher proportion of deaths and treatment costs during the study period. Most cases occurred among children aged 1-4 years who were male and of mixed race, with urgent care being the most common type of hospitalization.

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Virtual reality for improving social skills in children and adolescents with autism spectrum disorder: a systematic review and meta-analysis

Mayara Bottentuit Nogueira[1], Arthur Duarte de Sousa[1], Murilo Freitas e Silva Filho[1], Lanna Marry Silva de Morais[1], Patrícia Mendes Viana[2]

[1]Universidade Federal do Maranhão, São Luís MA, Brazil.
[2]Universidade do Extremo Sul Catarinense, Criciúma SC, Brazil.

Correspondence: Arthur Duarte de Sousa; Email: arthurddesousa@gmail.com.

Background: Although Autism Spectrum Disorder (ASD) is characterized by its persistent social deficits, the emotional processing abilities of individuals with ASD can improve over time with training. Virtual Reality (VR) technology is often reported as a tool that might help children and adolescents with ASD develop various social and emotional skills. However, despite several randomized controlled trials (RCTs) addressing its use, its efficacy in social and communication skills remains to be seen and will therefore be further investigated in this meta-analysis. **Objective:** To conduct a systematic review and meta-analysis to compare the effectiveness of virtual reality on social skills improvement compared to non-VR treatment in patients with ASD. **Methods:** We systematically searched Medline, Cochrane, and EMBASE databases for randomized controlled trials (RCTs) published until May 16th, 2024. Outcomes of interest were changes in social and communication skills, measured by changes from baseline in the validated scores: Autism Behavior Checklist (ABC), Psychoeducational Profile (PEP-3) and Vineland Adaptive Behavior Scale (VABS). All analyses were performed using Review Manager Software (v5.4). The effect sizes across studies were combined using the fixed-effect model, and the standardized mean difference (SMD) was used to combine the continuous outcomes. **Results:** Three RCTs were included in the meta-analysis, with a total of 167 patients, aged 3 to 12 years, of whom 77,2% were male and 54,5% underwent VR, which had a duration range of 3 to 5 months. Compared with the control groups (conventional therapy), the intervention group (adjunctive VR therapy) showed significant improvements

in social skills (SMD 0.57, 95% CI 0.24 to 0.89, p= 0,0006). The studies did not report any major side effects. In the qualitative assessment, four studies were included, and they suggested that adjunctive virtual reality therapy in children and adolescents with ASD is associated with significant improvement in social communication skills, but not with imitation, social interaction, language, or primary emotion recognition skills. **Conclusion:** Our study suggests that adjunctive VR therapy improves social skills when compared to conventional therapy. Due to the limited sample size, further research should be considered to strengthen the best available options for improving social skills in children and adolescents with ASD.

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Visual and hearing loss in a patient after electric shock: Cogan syndrome and differential diagnosis

Giovani Zago Borges[1], Daniela Aparecida Lima Viana[1], Ana Luisa Rufino de Sousa[1], João Henrique da Matta Clementino[1], Elisangela Ferraz Pazinato[1], Amanda Soares Pimenta[1], Alex Eduardo da Silva[1]

[1]Universidade Federal do Triângulo Mineiro, Uberaba MG, Brazil.

Correspondence: Giovani Zago Borges; Email: gzagob@gmail.com.

Case presentation: Male, 46 years old, admitted to the emergency room due to sudden low visual acuity, tearing, pain in the right eye and holocranial headache with rapid evolution of symptoms 1 day after an accident at work with an electric shock (220V) in right upper limb, with no history of local trauma or other previous neurological complications. Upon admission, laboratory tests were carried out and toxic-metabolic-infectious causes that could justify the condition were ruled out, in addition to showing no changes on Magnetic Resonance Imaging (MRI) of the brain and orbits or on Optical Coherence Tomography (OCT), with visual field examination confirming low visual acuity of the affected eye, without other ophthalmological changes. It was decided to start pulse therapy with methylprednisolone for 5 days without satisfactory response. In an outpatient follow-up consultation, after 3 months, his condition worsened, with bilateral progressive hearing loss worse on the right, profound and moderately severe on the left, confirmed by audiometry and evoked potentials of optic nerves indicating reproducible, asymmetric, central responses. with prolonged absolute latencies and reduced amplitudes on the right side. A new investigation was carried out during hospitalization, with no changes in the cerebrospinal fluid examination, with a new normal MRI and OCT, a hypothesis was raised based on clinical and exclusion criteria of Cogan syndrome, immunoglobulin was administered followed by pulse therapy, both for 5 days, prednisolone and methotrexate were prescribed at discharge. with stabilization of the evolution of symptoms 4 months later. **Discussion:** Cogan syndrome is a rare autoimmune disease that affects the eyes and inner ear, resulting in eye inflammation (interstitial keratitis) and hearing problems (vertigo, tinnitus and hearing loss). The prevalence is unknown, but it is considered very rare. The typical form presents interstitial keratitis and auditory vestibule symptoms. The atypical form includes other ocular and systemic manifestations. The diagnosis is clinical, based on symptoms and exclusion of other conditions, such as SUSAC syndrome, stroke and other central nervous system vasculitis. Treatment involves corticosteroids and immunosuppressants to control inflammation, and, in severe cases, biological therapy may be necessary. **Final comments:** Understanding Cogan syndrome is crucial for early diagnosis and effective treatment of an autoimmune disease, which, despite being very rare and underdiagnosed, can cause permanent hearing loss and severe visual complications. Correct identification of characteristic symptoms, such as interstitial keratitis and auditory vestibule problems, allows immediate medical interventions, reducing disease progression and improving patients' quality of life. Furthermore, knowledge about this syndrome helps to exclude other conditions with similar symptoms, ensuring adequate clinical management.

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Visual evoked potentials in spinocerebellar ataxias type 3 and 10

Otto Jesus Hernandez Fustes[1], Francisco Branco Germiniani[1], Cláudia Suemi Kamoi Kay[1], Paulo José Lorenzoni[1], Renata Dal-Prá Ducci[1], Paula Raquel do Vale Pascoal Rodrigues[1], Leo Coutinho[1], Rosana Herminia Scola[1], Hélio Afonso Ghizoni Teive[1]

[1]Universidade Federal do Paraná, Curitiba PR, Brazil.

Correspondence: Otto Jesus Hernandez Fustes; Email: otto.fustes@hc.ufpr.br.

Background: Visual evoked potentials (VEP) is a neurophysiological test that evaluates the function of the visual system, including the optic nerve, brain stem and cerebral cortex, through the electrical response to visual stimulation. Spinocerebellar ataxias (SCA) are heterogeneous diseases, with more than 50 types described, they present with different phenotypes, varying speeds of progression and clinical manifestations of different severity. Considering the presence of neuropathy, myelopathy, epilepsy and oculomotor changes among the SCAs, several studies have used electrophysiological methods to better characterize such manifestations. **Objective:** To report the electrophysiological findings presented in VEP in a cohort of patients with spinocerebellar ataxias type 3 and 10, comparing them. **Methods:** This was a cross-sectional study, in which we evaluated the VEP in a cohort of patients with SCA 3 (n=15) and SCA 10 (n=9). The patients underwent clinical evaluation in which demographic and clinical data, such as sex, age, duration of disease, and SARA scores, were collected. The VEP exam followed the guidelines from the American Clinical Neurophysiology Society. Both amplitudes and latency for the P100 potential were recorded and analysed. For statistical analysis we used a t-test and Wilcoxon test to compare normally and non-normally distributed numerical data respectively, whereas Spearman's correlation test was used to search for correlations between duration of disease/SARA scores and VEP abnormality. **Results:** 25 patients were enrolled in our study, 16 with SCA 3 and 9 with SCA 10. The clinical and demographic data did not present any statistically significant difference between the two groups. SARA scores and age of onset were similar between both groups. Conversely, VEP abnormalities differed between the two groups, as increased P100 latency and amplitude decrease were the most frequent change in the SCA 3 group (n=4). However, the occurrence of VEP abnormalities did not have a statistically significant correlation to either duration of disease, or SARA scores. **Conclusion:** Our data indicate that patients with SCA 3 have greater impairment of the visual pathway than those with SCA 10. More data are needed to confirm these findings, as well as to determine differences in the electrophysiological profile between Brazilian patients and other populations of patients with SCA 10. VEP can be a useful test in the monitoring of SCAs, as VEP can be used to detect changes in visual function that may be a sign of disease progression.

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Visual loss: beyond demyelinating diseases

Fernanda Nicoli Broch[1], Cândida Driemeyer[1], Laís Elizabete Favaretto[1], Alexandre Laisson Prado Taschetto[1], Marlise de Castro Ribeiro[1], Liselotte Menke Barea[1]

[1]Universidade Federal de Ciências da Saúde de Porto Alegre, Hospital Santa Casa de Misericórdia de Porto Alegre, Porto Alegre RS, Brazil.

Correspondence: Fernanda Nicoli Broch; Email: fernandanbroch@gmail.com.

Case presentation: A 32-year-old female patient was referred to the emergency department by the ophthalmologist due to the complaint of low visual acuity in the left eye. The patient reported progressive blurred vision in the left eye which had begun one year ago. Two months ago she noticed significant worsening of the symptoms leading to unilateral amaurosis. She was then hospitalized in another institution, where it was performed a skull computed tomography, which showed infiltration of the optic nerve. She then received a presumed diagnosis of neuromyelitis optica (NMO). It was prescribed pulse therapy and the patient was discharged with prednisone 80 mg/day. One month later, she was evaluated in our hospital presenting a persistent headache, pain when moving the left eye and blurred vision in the right eye. An investigation was carried out, and evidenced pachymeningitis on skull MRI and normal rheumatological tests, except for ANA, which was 1:640. Lumbar puncture had no abnormalities. Because of these findings, it was suspected of a granulomatosis disease and it was necessary to perform a meningeal biopsy. The immunohistochemical analysis confirmed pachymeningitis for IgG4-related disease (IgG4-RD). **Discussion:** IgG4-RD is an inflammatory disorder, immune mediated and it can affect multiple organs and systems, which can lead to fibrosis and tissue destruction. Any organ might be involved, but pancreas, biliary tract and lacrimal glands are most often affected. Neurologically, the disease is relevant due to the possibility of affecting the orbits, pituitary gland, meninges, peripheral nerves and even the brain. It usually affects individuals between 50-80 years old. Autoimmune pancreatitis affects both man and women in a proportion of 4:1. IgG4-RD is usually indolent, although acute and subacute manifestations are common when there is vascular involvement. In this case, significant involvement of the pachymeninges was observed with infiltration at the level of the optic foramen bilaterally, more evidently on the left, in addition to the orbital apex on the left and the retropharyngeal space. Infectious hypotheses were ruled out, serum IgG4 level was normal. CSF IgG4 level was not available. In the biopsy, lymphoplasmacytic predominance was noted with an IgG4:IgG ratio > 60%. Treatment with corticosteroids was started and continued with a biannual infusion of Rituximab. **Final comments:** Early consideration of IgG4-RD in differential diagnosis is crucial, as prompt diagnosis improves response to corticosteroid treatment. Delayed diagnosis can lead to irreversible fibrotic changes and permanent disabilities. Our patient spent over a year from the onset of symptoms until diagnosis. Despite the use of anti-CD20 therapy, the patient continues to show disease progression, highlighting concerns regarding limited therapeutic options and evidence.

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Weight loss at the time of diagnosis is associated with a shorter survival in ALS

Laura Carvalheira Dourado[1], Emanuela Coriolano Fidelix[1], Lucia Leite Lais[1], Mario Emílio Teixeira Dourado[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Mario Emilio Teixeira Dourado Junior; Email: medourado03@gmail.com.

Background: Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease, and the nutritional status of ALS patients is associated with survival. **Objective:** We retrospectively analyzed the effect of weight

loss (≥ 10%) at diagnosis and its association on patient survival. **Methods:** We reviewed a cohort of 132 ALS patients in our outpatient clinic at the hospital between January 2008 and March 2022. Clinical and demographic data were compiled. To assess predictors of survival, we analyzed weight loss ≥ 10% at time of diagnosis. Survival was defined as the time, in months, from the onset of symptoms to death or tracheostomy. **Results:** 54 (40.9%) patients had weight loss ≥ 10% at time of diagnosis. This weight loss was associated with a shorter survival time, comparing 24.0 months (95% CI: 19.2 - 28.8) vs. 34 months (95% CI: 30.9-37.1), p<0.010. In addition, 61 (46.3%) patients had significant weight loss at diagnosis without suffering from dysphagia. **Conclusion:** Weight loss at diagnosis is a significant concern in ALS and cannot always be attributed solely to dysphagia. Treatment for weight loss, including a high-calorie diet, oral nutritional supplements, and/or percutaneous endoscopic gastrostomy, should be discussed and considered during the patient's follow up.

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Wernekink Commissure syndrome with pseudo abducens palsy: a case report

Laura Alonso Matheus Montouro[1,2], Guilherme Spuri Martins[1], Giselle Fernandes de Oliveira[1], Nathalia Mike Moura Nakai[1], Rafaella Ferrisse Galbeiro[1], Matheus Vieira Santili[1], Eliana Meire Melhado[1], Natália Prando[1], Renan Cenize Guardia[1], Rodrigo Fellipe Rodrigues[1]

[1]Centro Universitário Padre Albino, Faculdade de Medicina de Catanduva, Catanduva SP, Brazil. [2]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: Guilherme Spuri Martins; Email: gui.spuri.martins@gmail.com.

Case presentation: A 77 years-old woman was admitted to the emergency presenting sudden onset of vertigo, diplopia, strabismus, ataxic gait, profuse vomiting, dysarthria. She was independent for daily activities and had a history of arterial hypertension, atrial fibrillation and a previous stroke without sequelae. Physical examination: right ptosis with ipsilateral eye exotropia, ophthalmoparesis, preserving just abduction movement of right eye, left eye has a neutral position, adduction and lower gaze preserved with abduction, conjugate vertical gaze palsy, bilateral appendicular ataxia, right body lateropulsion. MRI with high-signal T2/FLAIR lesions with real diffusion restriction (DWI/ADC map) in the right thalamus, mesial portion of the right temporal lobe and right paramedian midbrain. Hypoplasia of left vertebral and occlusion of right P2 on MR Angiography. On electrocardiogram, no visible P waves and an irregularly irregular QRS complex. On Echocardiogram, left atrial enlargement. Rivaroxaban prescribed for cardioembolic stroke prevention. **Discussion:** Midbrain infarctions are unusual, 0.9% of strokes, which is explained by the complex arterial blood supply of midbrain, supplied by posterior cerebral artery, upper basilar artery and the superior cerebellar artery. First described in 1958 by Lhermitte, the Wernekink Commissure syndrome (WCS) is very rare (69 cases), caused by an infarction of the paramedian midbrain, affecting wernekink commissure (brachium conjunctivum), medial longitudinal fasciculus, oculomotor nucleus and nerve, reticular formation, red nucleus. Clinical signs are bilateral cerebelar syndrome, variable eye movement anomalies and can also have variable movement disorders. This case has ipsilateral III nerve palsy, bilateral cerebelar syndrome, vertical gaze palsy and a contralateral reverse internuclear ophthalmoplegia, also called pseudo abducens palsy or pseudo-sixth phenomenon, a uncommon condition. This is the first report of a Brazilian patient with clinical and radiological WCS. **Final comments:** The midbrain has a complex arterial supply, which explains the rarity of WCS, and neuroanatomy, which can cause challenger neurological examination.

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What do we know about posterior reversible encephalopathy syndrome and burns? A systematic review

João Felipe Pissolito[1], Wilson Falco Neto[1], Beatrice David de Freitas[1], Felipe Bergamo Silva[1], Eliana Meire Melhado[1], Laura Alonso Matheus Montouro[1,2]

[1]Centro Universitário Padre Albino, Faculdade de Medicina de Catanduva, Catanduva SP, Brazil. [2]Faculdade de Medicina de São José do Rio Preto, São José do Rio Preto SP, Brazil.

Correspondence: João Felipe Pissolito; Email: felipe.pissolito@gmail.com.

Background: Reversible Posterior Encephalopathy syndrome (PRES) is an acute neurological condition characterized by a combination of clinical symptoms and radiological findings. It can occur in various clinical situations, but the incidence and characteristics of this syndrome in burn victims are unknown. **Objective:** Although no systematic review of this theme was previously conducted, the following study carried out a systematic search of the literature in order to clarify the clinical presentation of the syndrome. **Methods:** the following study consists of a systematic review following the PRISMA protocol. The search was carried out in three databases: Pubmed, Embase and BVS Saúde. In addition, the selected studies had their references checked in order to obtain new articles. The strategy was "Burned Patients" (Population) and "Posterior Reversible Encephalopathy Syndrome" (Outcome), however no terms referring to the population were used in order to obtain more articles. A search strategy was devised for each of the databases and the data from each was exported to the Rayyan platform for screening, which was carried out independently (blinded) by two researchers (FBS and BDF). Articles describing patients with PRESS after burns were included, regardless of the type of study or type of publication and language. Articles were excluded if they were not full-text or if they reported a case of the same patient. Data extraction was carried out by two researchers (WFN and JFP) and synthesis was carried out using the Google Sheets tool, obtaining information on demographics, diagnosis and clinical management. The quality of the articles included was assessed using the Joanna Briggs Institute Critical Appraisal tools' 8-item "Checklist for Case Reports" by two independent assessors (WFN and JFP). **Results:** once the databases had been searched, 14118 articles were found, of which 3276 were duplicates, totalling 10842 articles, which were scanned by reading the title and abstract. In the end, after reading the full text, a total of 5 articles were found, totalling 5 patients. Regarding clinical findings, the main findings were fever (n=3), visual alteration (n=2), convulsion (n=3), and hypertension (n=2), other findings include weakness and increased inflammatory response. With regard to imaging tests, the patients underwent CT and MRI of the skull, and bilateral vasogenic edema was found involving the white matter, predominantly in the temporal and occipital lobes.. In addition, two of the patients underwent EEG, showing generalized slowing, but no epileptiform activity. The findings converge with the classic characteristics of PRES. **Conclusion:** Even using a systematic search, only one case was located in relation to the last literature review. The low number of cases may be associated with the rarity of the condition and poor diagnosis. Neurological assessment of burn patients is essential for rapid identification and appropriate management.

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What is the influence of fatigue, global function, cognitive state, and pain in individuals with amyotrophic lateral sclerosis? A cross-sectional study

Tatiana Souza Ribeiro[1], Stephano Tomaz da Silva[1], Diogo Neres Aires[1], Aline Alves de Souza[1], Luciana Protásio de Melo[1], Saionara Maria Aires da Câmara[1], Ricardo Alexsandro de Medeiros Valentim[1], Ana Raquel Rodrigues Lindquist[1], Giane Amorim Ribeiro Samora[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil.

Correspondence: Tatiana Souza Ribeiro; Email: tatiana.ribeiro@ufrn.br.

Background: Fatigue is still an incompletely understood symptom in individuals with Amyotrophic Lateral Sclerosis (ALS), and little is known about its relationship with other symptoms such as global function, pain, and cognition. **Objective:** To identify the levels of fatigue, pain, global function, and cognition in a Brazilian group of individuals with ALS, in order to investigate potential influences between the aforementioned symptoms and fatigue. **Methods:** This is a cross-sectional study derived from a longitudinal (cohort) study. Individuals with ALS were recruited intentionally and non-probabilistically. After agreeing to participate in the research, the individuals were assessed using a custom evaluation form, collecting data regarding the level of fatigue (independent variable), global function, pain, and cognition (dependent variables). To assess fatigue, we used the Fatigue Severity Scale. For the dependent variables, we used the Amyotrophic Lateral Sclerosis Functional Rating Scale - Revised, the Visual Analogue Scale along with a body diagram, and the Amyotrophic Lateral Sclerosis Cognitive Behavioral Screen to measure global function, pain, and cognition, respectively. The covariates considered were sex, disease duration, type of symptom onset, age, and education level. Data were analysed by categorising fatigue (with and without fatigue) and considering the covariates. Comparisons, bivariate analyses, and multiple linear regression analyses were then performed. **Results:** We collected data from 72 individuals with ALS. Inferential statistics indicated differences between the fatigue categories in terms of global function, pain, and cognition. Bivariate analyses showed correlations between the independent variable and all dependent variables, as well as correlations between the covariates (except for type of symptom onset) and some of the dependent variables (P<0.20). Multiple linear regression analyses indicated a relationship between fatigue and the dependent variables. **Conclusion:** Fatigue is associated with lower levels of global function, poorer cognitive status, and higher levels of pain in a Brazilian sample of ALS patients. Additionally, age, sex, education level, and disease duration were identified as potential factors for the occurrence of fatigue, with a higher frequency observed in women compared to men with the condition.

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What is the role of telerehabilitation in motor symptoms, gait and balance of individuals with Parkinson's disease? A systematic review and meta-analysis

Liliane Santos de Vasconcelos[1], Lorenna Raquel Dantas de Macêdo Borges[1], Raíssa Souza Taveira[1], Larissa Araújo Gomes[1], Maria Amanda Ferreira Quirino[1], Larissa Coutinho de Lucena[2] , Lorenna Marques de Melo Santiago[2], Tatiana Souza Ribeiro[1]

[1]Universidade Federal do Rio Grande do Norte, Natal RN, Brazil. [2]Instituto Santos Dumont, Macaíba RN, Brazil.

Correspondence: Larissa Coutinho de Lucena; Email: larissa.lucena@isd.org.br.

Background: Parkinson's disease (PD) is a common neurological condition characterized by motor symptoms affecting mobility, social interaction, and mental health. In this context, Telerehabilitation (TR) uses information

and communication technologies to support continuity of care, reduce costs, and improve accessibility for this kind of patients. While studies indicate the feasibility and efficacy of TR in PD, extensive research is still ongoing post-pandemic, and existing reviews are not yet conclusive. **Objective:** To observe the effects of telerehabilitation in individuals with PD. **Methods:** This systematic review with meta-analysis follows PRISMA guidelines and included randomized and quasi-randomized controlled trials, and crossover studies with separed data for each phase. These studies involved individuals with PD at any stage of disease and aged over 18 years old, and the considered intervention was TR through physiotherapy. The primary outcomes were considered motor function (Movement Disorders Society - Unified Parkinson's disease Rating Scale: MDS-UPDRS); gait speed (10 Meter Walk Test - 10 MWT); gait stability (Dynamic Gait Index - DGI or by the Functional Gait Assessment - FGA); and postural balance (Berg Balance Scale - BBS or by the Mini-BESTest). For continuous outcomes with consistent instruments and units, the weighted mean difference was used. When instruments and units varied, the standardized mean difference was applied, along with 95% confidence intervals. For dichotomous outcomes, such as adverse events, the odds ratio and 95% confidence intervals were used. A random-effects model was adopted for the meta-analyses, and a narrative synthesis was conducted when meta-analysis was not feasible. **Results:** A total of eight randomized clinical trials were selected, conducted between 2016 and 2021, encompassing 486 individuals (average age of 68.59 years). The therapeutic modalities applied included resistance exercises (muscle strengthening), stretching, postural balance exercises, walking, and fall prevention strategies through TR. The studies varied in session duration from 15 to 60 minutes, and in frequency from 1 to 3 times per week, and for 3 consecutive days. A meta-analysis was conducted, and TR did not have a greater effect than control group on motor function (standardized mean difference = - 1.85, 95% CI -19.08 to 15.38; p=0.03; I^2=56%), on gait speed (standardized mean difference = -0.00 95% CI -0.31 to 0.31; p=0.75; I^2=0%), on gait stability (standardized mean difference = -0.01 95% CI -0.31 to 0.33; p=0.95; I^2=0%) and postural balance (standardized mean difference=0.09; 95% CI=-0.27 to 0.45; p=0.12; I^2=48%). **Conclusion:** TR did not demonstrate to be an effective physical therapy intervention to promote improvement in the outcomes analyzed in the population of individuals with PD in this study.

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Whole exome sequencing identifies SIN3A variant in a atypical Witteveen-Kolk syndrome patient: a case report

Lucas Scárdua Silva[1], Luciana Cardoso Bonadia[1], Alberto Rolim Muro Martinez[1], Clarissa Lin Yasuda[1], Iscia Lopes-cendes[1], Fernando Cendes[1]

[1]Universidade Estadual de Campinas, Campinas SP, Brazil.

Correspondence: Lucas Scárdua Silva; Email: scardua@unicamp.br.

Background: The Witteveen-Kolk syndrome is usually characterized by growth or feeding difficulties and possible brain malformation with developmental delay or intellectual disability. Epilepsy, psychiatric illnesses and dysmorphic facial features may accompany it, like a tall, broad forehead, down-slanting palpebral fissures, triangular face, pointed chin, and thin upper lip. Witteveen-Kolk is typically associated with variants of the SIN3A gene. **Objective:** To present a case of an adult male patient with an atypical Witteveen-Kolk syndrome, diagnosed with clinical and molecular methods. **Methods:** We performed whole exome sequencing (WES) in the patient's DNA extracted from peripheral blood. Variants of interest were selected and classified according to the ACMG recommendation. **Results:** The patient presented temporal lobe epilepsy, without brain malformation, associated with scapular, pectoralis major and sural bilateral muscular atrophy and weakness. He also presented some genetic marks as down-slanting palpebral fissures, a broad forehead and a triangular chin. Two maternal uncles presented with epilepsy, although his siblings and parents did not have epilepsy. He presented an electroneuromyography from 2008, which yielded a possible metabolic or muscular pattern. We identified a variant in the molecular analysis, c.1609C>T, p.R537* (NM_001145358.2) in SIN3A gene after analysis of the patient´s WES data. The variant was classified as pathogenic according to ACMG criteria: PM2- absent in population databases; PVS1-Null variant in a gene where the loss of function is a known disease mechanism and PS4- the prevalence of the variant in affected individuals is significantly increased compared to the prevalence in controls, Sanger sequencing confirmed the presence of the variant in a heterozygous state. No other pathogenic or likely pathogenic variants that could explain the phenotype were found. **Conclusion:** We report here a patient with a pathogenic variant in the SIN3A gene. The SIN3A gene encodes a transcriptional regulatory protein and has been definitely associated with intellectual disability syndrome. The Witteveen-Kolk syndrome has few muscular dystrophy-associated cases in the literature; future cases may help to better describe this additional phenotype.

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Wilson's disease mimicking essential tremor: a case report

Rafael Bastos Delgado[1], Maria Julia dos Santos[2], Regina Maria Papais Alvarenga[1], Marina Papais Alvarenga[1]

[1]Universidade de Vassouras, Vassouras RJ, Brazil. [2]Faculdade de Medicina de Catanduva, Catanduva SP, Brazil.

Correspondence: Rafael Bastos Delgado; Email: 16rafaelbastos@gmail.com.

Case presentation: A 43-year-old mixed-race woman from Rio de Janeiro was referred to the demyelinating diseases center in January 2024 with suspected multiple sclerosis. She reported starting a depressed mood in 2020, which she attributed to her husband's death from COVID-19. Simultaneously, she noticed hand tremors, weakness in all four limbs, and dizziness. She consulted an orthopedist who ordered electromyography (normal). In 2021, she was evaluated by a neurologist who first requested brain magnetic resonance imaging (MRI), showing scattered T2 and FLAIR hypersignal focus in the white matter adjacent to the lateral ventricles, with a nonspecific appearance. Additionally, there was T2 hypersignal without venous contrast enhancement or signs of diffusion restriction along the corticospinal tract extending from the thalamus through the midbrain to the pons bilaterally, also with a nonspecific appearance. Subsequent MRI scans in 2022 and 2023 showed the same images, suggesting metabolic changes such as copper accumulation disease. The patient denied prior neurological or systemic diseases, family history, and parental consanguinity. Neurological examination revealed postural and action tremors in the upper limbs, asymmetrically more pronounced in the left upper limb. Head tremors were also observed in denial. Tone, strength, reflexes, cranial nerves, and sensation were normal. The patient was lucid and oriented in time and space. Given the patient's extrapyramidal syndrome and MRI findings with a "panda face sign", Wilson's disease was suspected. Test results indicated reduced ceruloplasmin (13.3 mg/dL; normal range = 20-60 mg/dL), elevated urinary copper (209.1 mcg/L; normal range = up to 80 mcg/L), normal serum copper (91 mcg/dL; normal range = 76-152 mcg/dL), and presence of Kayser-Fleischer rings. Total abdominal ultrasound and liver function tests were normal. **Discussion:** We report a rare case of late-onset Wilson's disease with exclusive neurological presentation and essential tremor phenotype. The average age of onset in Brazil is 19 years, with onset after 40 years being rare. The patient presented with action and postural tremors typically associated with essential tremor, highlighting the importance of MRI in apparently benign cases. The absence of hepatic manifestations is uncommon, as most patients initially have liver involvement. **Final comments:** This case illustrates the phenotypic variability of Wilson's disease and the importance of differential diagnosis, especially in late-onset neurological presentations without a family history. Given that effective treatment exists for this disease, delays in diagnosis in atypical clinical presentations can be detrimental to quality of life and lead to severe neurological and hepatic complications.

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Wolfram syndrome: a rare multisystemic autosomal recessive disease case report

Lucas Araujo Limongi Horta[1], Breno Sousa Paiva[1], Mackeyle Siqueira de Souza[1], Sávio Cesar Rodrigues Pestana[1], Lucas Grobério Moulim de Moraes[1]

[1]Universidade Federal do Espírito Santo, Vitória ES, Brazil.

Correspondence: Sávio Cesar Rodrigues Pestana; Email: saviocesar05@gmail.com.

Case presentation: A 51-year-old Caucasian male with persistent dysglycemia was diagnosed with insulin-dependent diabetes mellitus at age six. During adolescence, he experienced a progressive decline in visual acuity over months to years, resulting in bilateral blindness. Ophthalmologic evaluation revealed bilateral optic nerve atrophy. In his second decade of life, the patient developed gradual, asymmetric sensorineural hearing loss of neurodegenerative origin. At age 20, a severe hypoglycemic event caused by inadvertent exogenous insulin administration led to hypoglycemic encephalopathy, resulting in transient coma. Due to a family history of Wolfram Syndrome (two siblings), this diagnosis was considered and confirmed through genetic testing. In his fourth decade, he reported neurological complaints, notably forgetfulness related to daily activities, and displayed progressive ambulatory difficulties, including impaired balance and a wide-based gait, requiring wheelchair use by age 45. In subsequent years, following an acute ischemic stroke, the patient developed dysphagia, REM sleep behavior disorder, urinary incontinence, and constipation. Additionally, he experienced focal seizures with secondary generalization, with presumed structural origin. **Discussion:** Wolfram syndrome (WFS) or DIDMOAD (Diabetes Insipidus, Diabetes Mellitus, Optic Atrophy, and Deafness), is a rare genetic disorder with these primary features. It can also involve neurological and psychiatric disorders and progressive neurodegeneration. The disease's severe neurological progression often leads to premature death, typically due to respiratory failure. WFS is an autosomal recessive condition associated with mutations in two genes, defining its subtypes: Wolfram syndrome-1 (WFS1) and Wolfram syndrome-2 (WFS2). WFS1, the classical form, is caused by mutations in the WFS1 gene, encoding the wolframin protein. WFS2, caused by mutations in the CISD2 gene, differs from WFS1 by presenting with bleeding, upper intestinal ulcers, defective platelet aggregation, and the absence of diabetes insipidus and psychiatric disorders. In the presented case, typical of WFS1, the initial symptom was diabetes mellitus, followed by visual acuity loss. By the first half of the second decade of life, the patient developed optic nerve atrophy progressing to blindness, consistent with most case reports. During this period, hearing loss unrelated to diabetes mellitus and diabetes insipidus may occur, potentially leading to hydronephrosis and atony of the bladder and ureter. Eventually, neurological symptoms such as reduced or abolished reflexes, dysphagia, and seizures may appear. **Final comments:** Despite limited understanding of WFS, advancements have been made in understanding the variability in onset and progression.

WFS should be suspected in cases of optic nerve atrophy following a diagnosis of diabetes mellitus in children and adolescents, with genetic testing used for confirmation.

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Worsening in attentional performance during presentation of medium-speed visual stimuli after a dialysis session

Carolina Corrêa Abramovicz[1], Gabriel da Silva Senges[1], Mario Meyer Rodrigues Fernandes[1], Ana Luiza Caldas Coutinho[1], Sergio Luiz Schmidt[1]

[1]Universidade Federal do Estado do Rio de Janeiro, Rio de Janeiro RJ, Brazil.

Correspondence: Carolina Corrêa Abramovicz; Email: carolinaca0226@gmail.com.

Background: The Continuous visual attention test (CVAT) is a 15-minute Go/No-go task. Previously, we demonstrated that attentional deficit in patients with End-stage kidney disease (ESKD) is restricted to high-speed presentation of stimuli [Interstimulus time intervals (ISI)] of 1 second and their RTs do not increase as tests progress (68 hours without dialysis). **Objective:** To evaluate the distribution of the RTs across the 15-minute test after a dialytic session. **Methods:** 109 ESKD and 109 controls participated in the study. The CVAT is divided into three blocks (5 minutes each). Each block presents two different target frequencies [low, 20% and high, 80% (LF and HF)] and three Interstimulus time intervals (1, 2, and 4 seconds). We conducted repeated-measures MANCOVA. Within-subject factors: Target frequency (TF) - 2 levels; ISI - 3 levels; Block - 6 levels; Time-length - 3 levels; Test/Retest - 2 levels. Between-subject factor: Group (ESKD and controls). In case of a significant interaction between the Group and one of the within-subjects factors, subsequent ANCOVAs were performed. **Results:** After a dialysis session, the patients' RTs showed a statistically and clinically significant increase during ISI = 2 s (F=7.541, df=1.524, P=0.002). This result did not depend on the TF. Additionally, their RTs increased throughout the 15-min CVAT. **Conclusion:** ESKD patients' RTs remained impaired during ISI = 1 s (high-speed) and worsened during ISI = 2 s (middle-speed). These findings suggest a fatigue effect caused by the dialysis session.

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Years lived with migraine disability in the fourth decade of life in Brazil: a five-year analysis

Hálisson Ferreira Freitas Filho[1], Felipe José Silva e Silva[1], Nathália Coimbra Coêlho[1], Izabella Carolline Menescal Ferreira[1], Luan Víctor Silva Ribeiro[2], João Victor Souza de Barros[1], Igor Moreira Miguez Godoy[3], Kaliana Kennedy Oliveira Calixto[1], Antônio Marcos da Silva Henriques[1], Larissa Suzan Basilio e Silva[1]

[1]Universidade Federal do Pará, Belém PA, Brazil.
[2]Afya Faculdade de Ciências Médicas, Cabedelo PB, Brazil.
[3]Centro Universitário Metropolitano da Amazônia, Belém PA, Brazil.

Correspondence: Hálisson Ferreira Freitas Filho; Email: halisson.filho@ics.ufpa.br.

Background: Migraine is a neurological disease characterized by repeated attacks of pain with a very variable frequency, usually associated with nausea, vomiting and sensitivity to light. With the growth of the 30-39 year-old population in Brazil, which represented 15.9% of the population in 2022, the high prevalence and great impairment in the quality of life of this comorbidity is reducing the healthy life expectancy in this age group, which can be characterized as the average number of years a person can expect to live in good health, without difficulties in carrying out certain functional activities. **Objective:** To evaluate the evolution of the rate of years lived with disability (YLDs) due to migraine in the fourth decade of life in Brazil from 2017 to 2021, identifying trends and differences in order to subsidize effective public policies. **Methods:** This is an observational, descriptive epidemiological study in which data on YLDs due to migraine from 2017 to 2021 were obtained from the Institute for Health Metrics and Evaluation (IHME) platform, in the Global Burden of Disease (GBD) section. Information on neurological diseases was selected and migraine data was filtered annually. The YLDs were calculated by multiplying the prevalence of the disease by the corresponding degree of disability. In order to carry out the appropriate correlation with the years of the fourth decade of life and the calculation of the rate per 100,000 inhabitants, the age groups, of both sexes, 30 to 34 years and 35 to 39 years, classified respectively as age group 1 and age group 2, were selected. **Results:** In the first year of analysis, 2017, for males, the highest rate was 626.34, in age group 1, and the lowest was 596.61, in age group 2. In 2018, the highest rate was in age group 1 (632.46) and the lowest in age group 2 (602.33). In 2019, the highest rate was 635.10 (age group 1) and the lowest 605.52 (age group 2). In 2020, the highest rate was 627.35 (age group 1) and the lowest 601.84 (age group 2). In 2021, the highest rate was 632.73 (age group 1) and the lowest 610.48 (age group 2). In the first year of analysis, 2017, with regard to females only, the highest rate was 1,106.94, corresponding to age group 2, and the lowest rate was 1,106.75, in age group 1. In 2018, the highest rate was found in age group 2 (1,111.08), and the lowest rate was in age group 1 (1,108.79). In 2019, the highest rate was 1,112.57, in age group 2. In 2020, the highest rate was 1,111.63 (age group 2), and the lowest rate was 1,103.17 (age group 1). In 2021, the highest rate was 1,122.01, continuing for age group 2. In age group 1, the rate was 1,109.24. **Conclusion:** In men, age group 1 had the highest rates in the five years analyzed, with an increase of 1.02% in the highest rates and 2.32% in the lowest rates from 2017 to 2021. In women, age group 2 had the highest rates, with an increase of 1.36% in the highest rates and 0.22% in the lowest rates over the same period.

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Young patient with third cranial nerve palsy due to midbrain stroke: a case report

Giuliana Moro[1], Caio de Lima Ferreira[1], Mayara Tszesnioski Maçaneiro[1], Beatriz Heloisa Born[1], Maria Eduarda Dobner e Silva[1], Letícia Sonza Brun[1], Aléxia Zattar[2], Felipe Ibiapina dos Reis[1]

[1]Universidade da Região de Joinville, Joinville SC, Brazil.
[2]Hospital Municipal São José, Joinville SC, Brazil.

Correspondence: Giuliana Moro; Email: gmgiulianamoro@gmail.com.

Case presentation: A female patient, 34 years old, was admitted in a hospital with changes in left ocular motility and left hemiparesthesia in the last 24 hours, suggesting partial paralysis of the third pair of cranial nerve. Patient had a history of stroke 3 years ago, but was unable to detail the condition. Neurological examination revealed the patient to be lucid and oriented in time and space, with mRankin and previous NIHSS of 0, paralysis of the left third nerve with pupillary involvement and partial eyelid drooping. A brain Computed Tomography (CT) was performed, which only revealed the presence of arachnoid cysts in the left frontal region. The magnetic resonance imaging showed a lesion with restricted diffusion in the midbrain, suggesting an Ischemic Stroke (IS). The transthoracic echocardiogram showed microbubbles on the left side of the heart after the fourth heartbeat following the saline injection, suggestive of extracardiac shunt. Finally, the Venous Doppler ultrasound of the lower limbs showed no signs of venous thrombosis and the CT of the chest with contrast also showed no significant changes. After 8 days of admission, an IS was determined in the territory of midbrain perforators, suggestive of Patent Foramen Ovale (PFO), with no indication for further investigation or invasive procedures and no other conditions associated. According to the results, there was no cardiological indication for anticoagulation, but it suggests the possibility of complementing the diagnosis with Cardiac catheterization. The follow-up treatment and complete investigation was based in advising the patient about warning signs and symptoms, prescription of antiplatelet aggregation and analgesia and return to the monitoring results of cerebral and cervical angiography. **Discussion:** Isolated paralysis of the third cranial nerve is an uncommon condition with various differential diagnoses, such as infectious, vascular, or inflammatory diseases. The oculomotor nerve is responsible for the innervation and movement of ocular muscles and its nuclei are located in the upper midbrain region. The occurrence of an IS in the midbrain region can cause isolated paralysis of the oculomotor nerve and consequently lead to diplopia, ptosis, and mydriasis. IS can have different causes, the most common being cardioembolic. Patients with a PFO may suffer IS of cardioembolic origin and, in rarer cases, affect the circulation of the midbrain. **Final comments:** In conclusion, our case underscores the diagnostic challenges posed by the presentation of third cranial nerve palsy. The identification of a PFO in this patient shows the importance of comprehensive evaluation in cases of cryptogenic stroke. The rare association of PFO with midbrain ischemia causing third cranial nerve palsy underscores the need for comprehensive evaluation and multidisciplinary care, including cardiac catheterization, for optimal management.

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Z-drugs effects for the treatment of sleep disorders

Letícia Fernandes de Freitas[1], Paulo Souza Oliveira[1], Júlia Oliveira Caldas[1], Sophia Carnavale Maciel de Lima[1], Yuri Ogi de Lima Barbosa[1], Caroline Chaves Bittencourt[1]

[1]Universidade Nove de Julho, Guarulhos SP, Brazil.

Correspondence: Leticia Fernandes de Freitas; Email: lecaff1984@gmail.com.

Background: It is known that sleep disorders are exponentially prevalent throughout the world, ranging from 37.2% to 69.4% in adults, which is the majority affected population. Many studies have been carried out to investigate the etiology and adequate treatment of these disorders, but it has only been found that their origin is multifactorial and, currently, the most prescribed treatment is the Z drugs, represented by zolpidem, zopiclone and zaleplon. However, the effects of these medications in the treatment of sleep disorders must be analyzed, considering their benefits, but also their dangers for neurophysiology in the long term, compared with other medications, such as benzodiazepines. **Objective:** This research is up to analyze the effectiveness of Z drugs in the treatment of sleep disorders. Furthermore, the mechanisms of action of Z drugs on the central nervous system and their impact on sleep patterns should be investigated. **Methods:** The research was made through databases on sites such as BMC Medicine, Springer Links and PubMed/MEDLINE about the effects of Z drugs in the treatment of sleep disorders, following the Guidelines of Preferential Report Items for Systematic Reviews and Meta- Analysis [PRISMA], as proposed by Moher et al. (2009) and Higgins and Green (2011). **Results:** When carrying out the study, it was observed that prescriptions for Z drugs were greater than prescriptions for other

hypnotics, such as benzodiazepines, for the treatment of insomnia. These drugs were mostly prescribed by non-psychiatric doctors and for a longer period than recommended, as many patients were advised to continue the medication for a month or more. From this perspective, harmful effects were observed in patients using these drugs, who reported at least one side effect,. Thus, although Z drugs are currently considered the best choice for treating insomnia, caution and better knowledge on the part of health professionals about their effects are still necessary. **Conclusion:** Based on the analyzed information for insomnia treatment, it is deemed necessary to consider the risks and benefits of the medications used. For older adults, controlled-release melatonin and doxepin are recommended as first-line agents, while Z-drugs should be reserved for use if first-line agents are ineffective. It is important to avoid the use of benzodiazepines in insomnia treatment due to factors such as high tolerance and dependence rates, potential cognitive side effects, lack of long-term efficacy, and the availability of safer options for patients. Individuals with sleep apnea or chronic lung disease are advised to seek evaluation by a sleep specialist before taking sedative medications. These guidelines aim to ensure effective and safe treatment for patients with insomnia while emphasizing the importance of considering patients' experiences and preferences when prescribing hypnotic medications for insomnia.

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Lumbar fistula and spontaneous intracranial hypotension - case report

Welton Gomes de Paula[1], Rodrigo Braga Pacheco[1], Ana Claudia Dias Malta[1], Laura Mariana Araújo do Amaral[1], Rachel Tiago Ligório[2,3]

[1]FAMINAS, Belo Horizonte BH, Brazil.
[2]Santa Casa de Misericórdia, Belo Horizonte BH, Brazil.
[3]Rede Mater Dei, Belo Horizonte BH, Brazil.

Correspondence: Laura Mariana Araújo do Amaral; Email:laurithia@hotmail.com

Case presentation: A 53-year-old man with previous history of hypertension and diabetes started with a moderate-severe throbbing headache in October 2022. The pain was located in frontal head and worsened immediately after standing up and after Valsalva maneuver, accompanied by nausea, visual blurring, diplopia, gait ataxia and tinnitus. No trauma history. The patient presented to an emergency neurological care in February 2023 and cranial and spinal magnetic resonance imaging (MRI) showed indirect signs of cerebrospinal fluid (CSF) hypotension, including reduced mammillopontine distance and the descent of cerebellar tonsils through the foramen magnum. Cranial arterial and venous magnetic resonance angiography and CSF analysis revealed no abnormalities. Conservative treatment was unsuccessful. In April 2023, a blind epidural blood patch was performed and was also refractory. The patient continued to have significant orthostatic headaches and a cisternal scintigraphy in August 2023 showed leakage of radioactive material in the lumbar region. A targeted epidural blood patch was performed at the location of the lumbar CSF fistula, resulting in a good response and stability since then. **Discussion:** Spontaneous intracranial hypotension is a disorder caused by spinal CSF leakage and it typically occurs in patients in their midthirties to midfifties. The classic presentation is acute orthostatic headache, usually bilateral, which can present with migrainelike features, such as nausea and photophobia. The headache presents with moderate to severe intensity and can worse with the Valsalva maneuver. Other symptoms may include vestibulocochlear manifestations such as hypoacusis, dizziness and tinnitus, gait disturbances and cranial nerve palsies. According to the International Classification of Headache Disorders, Third Edition (ICHD-3), a diagnosis of spontaneous intracranial hypotension requires the presence of a headache that developed in temporal relation to low CSF pressure (<60mm) and/or evidence of CSF leak on imaging, in the absence of procedures or trauma. The most common complication is subdural hematoma. Several neuroimaging modalities can aid in diagnosis. Brain MRI may reveal typical findings, but CT-myelography or cisternal scintigraphy can be necessary to locate the CSF leakage. The treatment consists of conservative management such as strict bed rest, hydration and caffeine, epidural blood patches and, in refractory cases, surgical repair of spinal fistula. **Final comments:** Spontaneous CSF hypotension is rare but debilitating and should be promptly recognized. It typically presents with acute orthostatic headache but can have another associated symptoms. Currently, the epidural blood patch is the treatment of choice, especially when targeted to the exact location of the CSF leakage, resulting in good pain control and improved quality of life.

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Complexity of diagnosis and clinical management regarding neurological infection by the fungus paracoccidioides: a case report

Rodrigo Braga Pacheco[1], Laura Mariana Araújo do Amaral[1], Gabriella Bretas Alves[1], Gustavo Barros Loureiro[2,3]

[1]FAMINAS, Belo Horizonte BH, Brazil.
[2]Faculdade Ciências Médicas de Minas Gerais, Belo Horizonte BH, Brazil.
[3]Hospital Santa Casa de Belo Horizonte, Belo Horizonte BH, Brazil.

Correspondence: Laura Mariana Araújo do Amaral; Email: laurithia@hotmail.com.

Case presentation: A 51-year-old man, R.C.L.J, smoker, drinker, without comorbidities, a porter, and gardener. He was admitted to the hospital with a holocranial headache lasting for 02 months, with no precedents and gait alteration/difficulty; admission due to loss of strength in the left half of the body. Cranial nerve alterations with paresis and deviation of the tongue to the right, exophthalmos, ptosis, Babinski and Chaddock signs on the left, and signs of upper motor neuron syndrome on physical examination. Whole-body CT revealed lung lesions and a left occipitoparietal nodular lesion, both with characteristics suggestive of neoplasms or infection.Additionally, cranial CT showed multiple expansive lesions in the brain parenchyma and brainstem, with ring enhancement after contrast injection.Histopathology showed lymph nodes with architectural subversion and the presence of several granulomas with areas of necrosis compatible with the fungus causing PCM.Finally, brain MRI revealed cortical granulomas in both hemispheres, a lesion in the brainstem affecting the medulla, pons, and midbrain, a larger temporo-occipital lesion, and a left frontal lesion. The association of histological findings, previously presented symptoms with similar involvement of the CNS and lungs led to the diagnosis of NP.A 4-week treatment with intravenous amphotericin B was performed with subsequent clinical improvement, followed by maintenance itraconazole (200 mg oral tablet once daily) proposed for 12 months. **Discussion:** Paracoccidioidomycosis (PCM) is a systemic disease of fungal etiology caused by thermally dimorphic microorganisms of the genus Paracoccidioides, and its prevalence is significant in Latin America, especially in Brazil, with over 70% of cases.One of the main predictors of infection is middle-aged men and farmers, given that the fungus is saprophytic in soil.A systematic review demonstrated that the most affected site is the lungs, although cutaneous, lymph node, adrenal, and even central nervous system (CNS) lesions can also occur, characterizing neuroparacoccidioidomycosis (NP).The latter can present with intense headache, seizures, sensory, motor, cognitive, and cerebellar changes.The definitive diagnosis of PCM is achieved by isolating the fungus through biopsy or necropsy, but the complexity and applicability of these methods converge on a diagnosis based on evidence of involvement of other organs associated with symptoms and neurological imaging such as CT and/or MRI.In this context, a case report is presented aiming to clarify concrete and effective diagnostic methods, given the difficulty in managing patients with NP. **Final comments:** PCM is a systemic disease with relatively low CNS prevalence; thus, NP, despite low mortality, presents high morbidity with long-term sequelae, generating health burden. Therefore, the relevance of understanding the pathology and the importance of diagnostic efficacy for appropriate management are perceived.

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Immunotherapy in the treatment of Alzheimer's disease: a systematic review

Danilo Golfeto Dourado[1], Laura Dellagnesi Depieri[1], Felipe Iankelevich Baracat[1], Igor Nobre Cruz[1],

[1]Universidade do Oeste Paulista, Presidente Prudente SP, Brazil.

Correspondence: Igor Nobre Cruz; Email: igornobrecruz@gmail.com.

Background: Alzheimer's disease (AD) is a progressive and irreversible neurodegenerative disorder characterized by multiple cognitive deficits. It is marked by the accumulation of neuritic plaques and neurofibrillary tangles in the brain, primarily due to the build-up of beta-amyloid peptide (Aβ42) and tau protein (p-tau). AD affects millions of people and is a leading cause of disability and morbidity in the elderly population. Immunotherapy involves monoclonal antibodies, which are proteins that bind to Aβ42 to facilitate its removal from the brains of patients with AD. Current studies on immunotherapy are promising; however, there is no consensus on its therapeutic potential. Therefore, this review aims to elucidate the effects of this intervention on modifying therapies for Alzheimer's Disease. **Objective:** To compare the efficacy of monoclonal antibody immunotherapy versus placebo in the treatment of patients with AD through randomized clinical trials. **Methods:** The systematic review protocol was conducted according to the recommendations of the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA). Searches were conducted in the PubMed and Embase databases. Randomized clinical trials comparing the efficacy of monoclonal antibody immunotherapy and placebo in the treatment of AD patients were included. The outcomes evaluated were changes in the Clinical Dementia Rating Scale Sum of Boxes (CDR-SB) and Alzheimer's Disease Assessment Scale–Cognitive Subscale (ADAS-Cog13). Two independent reviewers performed study selection, data extraction, and risk of bias assessment, with a third reviewer resolving any discrepancies. The JADAD scale was applied to assess the methodological quality and risk of bias of the studies. **Results:** The search identified 2738 studies, of which 2709 were excluded based on title and abstract evaluation, while 29 were read in full. The main exclusion criteria were study design, thematic inadequacy, duplicates, and the absence of a comparison between monoclonal antibody use and placebo. To date, 22 clinical trials have been included in this review. Among these, 6 studies demonstrated a reduction in p-tau levels associated with immunotherapy, and 2 of them also showed a slower clinical decline compared to placebo; 2 studies independently associated monoclonal antibodies with a better composite score for cognition. Fourteen other studies did not present significant differences. However, interpretation should be cautious, as these results are partial. **Conclusion:** To date, the effects of immunotherapy in the treatment of AD, derived from the results, suggest a positive but limited impact compared to placebo. Nonetheless, even modest improvements justify further exploration of immunotherapy for AD.

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Neurosyphilis tumefactive lesions mimicking a tumor: a case report

Ana Clara Vlakov Isper[1], Letícia Isper[1], Guilherme Bussola Carazzatto[1], Igor Nobre Cruz[1], Ana Júlia Machado Parente[2], Maria Teresa Fernandes Castilho Garcia[1], Guilherme Menezes Mescolotte[1]

[1]Universidade do Oeste Paulista, Presidente Prudente SP, Brazil.
[2]Hospital Regional de Presidente Prudente, Presidente Prudente SP, Brazil.

Correspondence: Igor Nobre Cruz; Email: igornobrecruz@gmail.com.

Case presentation: Male, 50 years old, smoker, was transferred to our emergency service after initiated, 10 days ago, a progressive and asymmetric paraparesis. After 30 days of evolution the patient had loss of sensation at T6 level, with no upper motor neuron signs, only areflexy and hypotonic finds. The patient denied recent infectious history. Cranial and spinal MRI was performed revealing bilateral solidocystic expansive injuries in the temporal lobes with significant vasogenic edema. Extensive transverse myelitis affecting from cervical to medullary cone, including a well-circumscribed intramedullary nodular formation at the T6 level. Both with contrast enhancement. Cerebrospinal fluid (CSF) analysis demonstrated high cellularity with 228/ mm^3 lymphomononuclear cells, as well as hyperproteinorrhaquia with (150mg/dL) and VDRL titers above 1/16 in CSF. Bacterial and fungal cultures of the CSF were negative, as well as the viral panel. Following confirmation of Neurosyphilis, treatment with Crystalline Penicillin was launched. Screening for Paraneoplastic Syndrome was performed with Esophagogastroduodenoscopy, Colonoscopy, CT scans of the Abdomen, Pelvis, and Chest, revealing only mediastinal and retroperitoneal lymphadenopathy. Given the mazed diagnostic, a biopsy of the temporal lesion was performed, and the tissue showed leukocytic proliferation with heterogeneous density involving intra and extra-axial compartments and a polymorphic infiltrate, predominantly lymphomononuclear. Additionally, areas of non-suppurative necrosis with peripheral granulomatous outline were found. **Discussion:** Neurosyphilis can manifest at any point in the natural history of the onset, with the most severe forms appearing in the tertiary period. This involvement was more common in HIV-positive patients, however, this evidence could not be observed in the presented case. Histologically, there is evidence of chronic inflammatory process, predominantly lymphocytes and plasma cells. Gummas formation may be observed, mimicking expansive lesionsor syphilitic lesions forming masses, which in CT or MRI images may trick primary or metastatic malignant tumors. In the reported case, arranged with clinical record, laboratory and radiological findings, and foreclosure of other possible diagnoses, it is confirmed Neurosyphilis. **Final comments:** We thus demonstrate that syphilis may pose a diagnostic challenge, due to it's wide range of manifestations and it's potential imitator. Furthermore, the clinical findings associated to it's complex management finds out to be rarer than the others, adding medical knowledge.



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