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Complex craniosynostosis in Pitt-Hopkins Syndrome: Case report in twins.

José Roberto Tude Melo, Ana Rita de Luna Freire Peixoto, Danilo Marden de Lima Souza

Introduction: Pitt-Hopkins syndrome (PTHS) is a rare genetic syndrome associated with neurodevelopmental disorders and craniofacial dysmorphisms caused by variations in the TCF4 transition factor. The aim of this article is to report the case of two twin infants diagnosed with PTHS, confirmed by the identification of a heterozygous pathogenic variant in the TCF4 gene through DNA extracted from a buccal swab, with premature fusion of the left coronal and metopic sutures in both twins. **Case Presentation:** Both infants presented with craniofacial asymmetry with a metopic crest and cranial deformity. During the diagnostic investigation, computed tomography with three-dimensional reconstruction of the skull showed premature fusion of the left coronal and metopic sutures in both twins. They underwent craniofacial reconstruction at the 9th month of age using a combination of techniques. The postoperative outcomes were satisfactory in both cases. **Conclusion:** This is the first case

report to describe the occurrence of complex craniosynostosis (CCS) in children with PTHS. Further studies are needed to determine whether the co-occurrence of PTHS and CCS described here indicates an association or is explained by chance.

Keywords: congenital neurologic disorders, craniofacial surgery, craniosynostosis, neurosurgery, transcription factor 4.

Quantitative analysis of trigonocephaly: a literature review

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INTRODUCTION:Trigonocephaly is the second most common type of craniosynostosis, accounting for up to 25% of cases. Measuring its severity, determining surgery indication, and evaluating post-surgery outcomes still present challenges for neurosurgeons. The present study aims to conduct a comprehensive literature review on the main anthropometric measures that assist in measuring the anterior cranial fossa, diagnosing trigonocephaly and its severity, as well as evaluating post-surgery outcomes.



MATERIAL AND METHODS: Integrative literature review with data collected from MEDLINE/PubMed.

RESULTS: The measurement of the anterior cranial fossa and of the trigonocephaly can be performed through the analysis of several anthropometric measures. These include: intercoronal distance, interparietal distance, cranial length, anterior and lateral interorbital distance, interpupillary distance measurement of proptosis, intertemporal distance, length of lateral and medial walls of the orbit, metopic index, bifrontal endocranial angle, adjusted bifrontal endocranial angle, frontal angle, anterior cranial fossa angle, horizontal cone angle, and bitemporal/biparietal distance ratio. The studies reviewed showed that the metopic index, frontal angle, and adjusted bifrontal endocranial angle are the measures that best quantify the severity of the trigonocephaly and consequently assist in determining the need for surgical repair and providing a non-subjective evaluation of post-surgery results.

CONCLUSION: Cranial dysmorphisms can be quantified through anthropometric measures. The metopic index, the frontal angle, and the adjusted bifrontal endocranial angle are useful and appropriate for measuring the severity of the trigonocephaly and, consequently, to support its surgery indication. These measures can also be used to evaluate post-surgery outcomes.

Keywords: Craniosynostosis, Trigonocephaly, Anthropometry

Comparative analysis of surgical techniques for the treatment of scaphocephaly: an evaluation of aesthetic outcomes

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INTRODUCTION: The present study aims to compare two surgical techniques (with and without coronal and lambdoid preservation) for the treatment of scaphocephaly in children operated on under 6 months of age, by evaluating the aesthetic outcome 1 year after surgery. **METHODS:** This is a retrospective, observational, and longitudinal study of patients with scaphocephaly who underwent surgical treatment before 6 months of age between 2018 and 2022. To evaluate the aesthetic outcome one year after post-operative, a 10-item questionnaire based on the Whitaker classification was administered to parents. **RESULTS:** Regarding the parents' assessment, a superior evaluation was observed in the technique with suture preservations, with the majority rating the outcome as excellent (53.3%). In the evaluation of the surgeons, alopecia at the incision site (57,14%), strabismus (6%), and keloid at the surgical scar (6%). In the surgery without suture preservation alopecia at the incision site (57,14%), prominent crest (42%), and strabismus (28.5%) were observed. No patient showed facial asymmetry after either technique. The patients after surgery

without suture preservation presented grade 2 on the Whitaker scale (85%) and (14.2%) were classified as grade 4. The patients after surgery with suture preservation 33.3% as grade 2, 53.3% as grade 1, and 13.3% grade 3. **CONCLUSION AND DISCUSSION:** The method of preserving sutures led to significantly better aesthetic outcomes in comparison to the technique without suture preservation, resulting in a reduced number of bone defects, cranial prominences, and lower occurrence of strabismus.

Keywords: scaphocephaly, craniosynostosis

Cloverleaf skull (kleeblattschadel): a serious manifestation of pfeiffer syndrome

Julia Santana de Oliveira, Henrique Rodrigues Osorio, Leticia Tozzini Tavares da Silva, João Victor Rodrigues Bubicz

Pfeiffer Syndrome is a rare disease that occurs in one out of every 200,000 newborns. It is characterized by the premature closure of cranial sutures associated with facial malformations. This case reports a newborn (NB) with PS type 2, which represents the most severe presentation, consisting of a trilobed cranial deformity (cloverleaf skull), extreme proptosis, finger abnormalities, elbow ankylosis. The cloverleaf skull can lead to restricted brain growth, and extreme proptosis. The present case describes an NB diagnosed with fetal malformation during prenatal ultrasound, cesarean due to fetal malformation. Upon evaluation, there were noted wide sagittal and metopic sutures forming a broad non-tense median fontanelle, and temporal growth resulting in a cloverleaf head appearance. CT scan revealed partial closure of sagittal, coronal, and lambdoid sutures with cranial protrusion of brain through the anterior fontanelle, bulging of the cranial vault in the temporal regions consistent with a cloverleaf skull, and dilation near the choroid plexus of the lateral ventricles. Ultrasound examination partially visualized the corpus callosum and showed absence of the septum pellucidum. The decision was made to pursue expectant treatment. By 3 months, she developed tense fontanelles, and a tomographic exam revealed hydrocephalus, necessitating a ventriculoperitoneal shunt. The patient experienced several infectious complications and at 7 months, died due to COVID-19 infection-related complications. The prognosis is guarded and linked to the severity of anomalies, with early mortality being a common outcome. Further studies and reports are of great importance to arrive at better therapeutic options.

Keywords: Craniosynostosis, kleeblattschadel, Pfeiffer

Postural plagiocephaly, cranial orthoses and conservative therapy: a literature review

Amanda Jhully Rodrigues Lopes

Objective: To carry out a literature review on the indication and benefit of using cranial orthoses versus conservative therapy in postural plagiocephaly. **Method:** The review was carried out in the PubMed, Scielo and Lilacs databases, in the last 5 years without language restrictions, with the descriptors cranial orthoses AND postural plagiocephaly. 18 articles were found, of which 2 were excluded. **Result:** The number of cranial deformities has increased in recent years, since the beginning of international recommendations on the prevention of sudden death. In that case postural plagiocephaly has been very recurrent in the neurosurgeon's office. The search for the indication of the use of cranial orthosis is a common theme, however with a lot of divergence in relation to its indication. Despite the controversy about the indication, most studies indicate conservative therapy and the use of the orthosis, not making much distinction between the two alternatives, except in very specific cases. Some studies that put the use of the orthosis as indicated, had some conflict of interest and/or low evidence. This does not exclude the valid indication of the orthosis, given that studies with a high level of evidence also prove its benefit, provided that the indication protocols are strictly followed. **Discussion and Conclusion:** In view of this, we can conclude that most of these patients can receive the indication of conservative therapy without significant differences in their long-term results, considering that impeding factors such as failure to adhere to protocols and non-availability in the SUS favor the conduct based on conservative therapy.

Keywords: Plagiocephaly, Cranial Orthosis, Conservative therapy

Kyphectomy in patients with myelomeningocele. Is there a better age for surgery correction? A systematic review of literature

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Background: Myelomeningocele can be associated with spine curve deformities, including kyphosis. These deformities can be progressive and impair patients' quality of life. Surgery for correction is often necessary. However, there is no consensus on when the surgical procedure should be performed. Thus, the objective of this analysis is to compare kyphectomy in different age groups.

Methods: This study is a systematic review of literature, performed by PRISMA protocol. The PICOT strategy where P - patients with myelomeningocele associated with kyphosis, I - for neonatal kyphectomy, C - patients who did not undergo neonatal kyphectomy but who underwent it at another age, and T- follow-up period.

Results: Overall, 21 articles reporting 142 patients were analyzed. The patients were subdivided into three groups according to the age of kyphosis surgery: neonatal, up to ten years old, and over to ten years old. The mean follow-up was 3.8 years (2.3-5.9), and the mean kyphosis corrected angle was 84° (SD: 34°). All populations studied showed statistical significance in the comparison between pre-and-post-operative kyphosis angles, reinforcing the effectiveness of surgical therapy for this deformity. In our results, all groups presented with an improvement in the quality of life (QoL), nevertheless the neonates had a higher rate of QoL. In our analysis, there was no difference between groups during the safety assessment. No neonatal deaths were reported. In comparison, the up to 10 years and over to 10 years groups had a mortality rate of 1.4% and 0.7%, respectively.

Conclusion: The present study shows that kyphectomy is the surgical procedure of choice for treating kyphosis in patients with myelomeningocele and it is safe and effective in all age groups. In addition, the procedure performed on neonates seems to improve the quality of life and if necessary a late second surgery will be more straightforward.

Keywords: Myelomeningocele; neonate; kyphosis correction angle; quality of life; kyphosis.

Discordant lumbar myelomeningocele in a dizygotic twin and literature review

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Introduction: Myelomeningocele (MMC) is a congenital malformation of the central nervous system resulting from a neural tube defect (NTD). The incidence of MMC ranges from 0.5 to 1.0 per 1,000 pregnancies. In twins, the incidence of NTD is even higher, at 5.38%. However, only a few studies have reported discordant MMC in dizygotic and dichorionic twins, which highlights the importance of describing the clinical course and outcome of this case.

Case Report: A newborn girl, the first of dichorionic and dizygotic twins to be delivered by elective cesarean at 37 weeks of gestation, was diagnosed with discordant MMC at birth. The 26-year-old mother, a smoker and recreational drinker who did not take folic acid during her pregnancy, had no family history of neurological malformations. The mother started prenatal care at 11 weeks of gestation, with no diagnosis of fetal pathologies. The infant underwent surgery for MMC at two days of life but developed persistent cerebrospinal fluid (CSF) leakage from the surgical wound after 14 days, which led to ventriculoperitoneal shunt placement at 19 days of life.

Discussion: MMC is a multifactorial malformation. As an NTD, its incidence is reduced by up to 70% with regular prenatal folic acid intake. MMC can be diagnosed through

prenatal screening tests and ultrasound. Surgical repair may be indicated during the fetal period or within 48 hours of birth.

Conclusion: Twin pregnancies have a higher risk of malformations than singleton pregnancies. Because the incidence of twin pregnancies is increasing, this study will help specialists and the scientific community better diagnose and manage such cases.

Keywords: Myelomeningocele; dysraphism; twins

Epidemiological aspects of patients with myelomeningocele in a university hospital - a case series

Júlia Santana de Oliveira, Henrique Rodrigues Osório, Bruno Henrique Mota Segnori, Georgia Varaschin Debes, Luca de Carvalho Contieri

Among the malformations of the Central Nervous System (CNS), myelomeningocele accounts for 85% of cases of neural tube non-closure, resulting in the exposure of spinal cord contents. It is present in Brazil in 1 in every 1000 live births. The present study conducts a comparative analysis between 7 cases of myelomeningocele in a hospital, examining risk factors, diagnostic methods, treatment, and complications. A literature review was conducted using the PubMed database. Patient records with myelomeningocele born between the years 2020 and 2023 were analyzed. Six of the babies were delivered via cesarean section, and one through vaginal delivery. Five of them had prenatal diagnoses. Information about the use of folic acid during pregnancy was only available for three of the newborns, all of whom did not take it. Regarding surgery, four underwent the procedure on the same day as birth, three on the following and one on the 13th day after birth (referred externally). The length of hospital stay ranged from 18 to 30 days. Among the reported complications, five presented hydrocephalus, with one of them also having hydranencephaly. Concerning comorbidities, two newborns had none, one had hypospadias and syndactyly, another had PCA and non-palpable testicle, another had congenital clubfoot and hydrocephalus, another had neurogenic bladder, and another had congenital clubfoot. Ventriculoperitoneal shunting was necessary in four cases. The profile of patients with myelomeningocele is consistent[d] with the literature. The lack of consistent information observed in the medical records hindered further refinement of the study..

Keywords: myelomeningocele, malformations, Dysraphism

Interhemispheric arachnoid cysts: case series.

José Roberto Tude Melo, Leonardo Conrado Silva Lima

Objective: Interhemispheric arachnoid cysts (IHACs) are a rare congenital malformation arising from the duplication of the arachnoid membrane of the cerebral meninges associated with malformations of the corpus callosum. The scope of this study is to describe a series of cases of IHACs treated via neuroendoscopy in a reference center. Method: description of a case series of children diagnosed with IHAC undergoing neuroendoscopic treatment (cystoventriculostomy) between 2015 and 2022, describing the main clinical manifestations, the surgical indication, and the neuroendoscopic technique performed. Results: Four children diagnosed with IHACs, with a predominance of males (3:1), were included in the study. Considering the Mori and Barkovich classification systems, magnetic resonance imaging of the brain showed multiseptate extra-axial interhemispheric cysts without communication with the ventricular system in all cases. Macrocrania and bulging fontanel were the clinical manifestations that indicated urgent hospitalization for the proposed neurosurgical treatment. Conclusion: Signs of increased intracranial pressure were observed in all cases of IHACs in the first months of life. Endoscopic cystoventriculostomy was performed in all cases with satisfactory results, showing that this is an option to be evaluated for the treatment of IHACs classified as multiseptate extra-axial and without communication with the ventricular system or brain cisterns, according to Mori and Barkovich classification systems.

Keywords: arachnoid cysts, cysts of the central nervous system, neuroendoscopy, neurosurgery, congenital malformations.

Schizophrenic symptoms in patients with cerebral gliomas

Ana beatriz sales vieira, Eder Cruz de Sousa

Introduction: Glioblastoma (GBM) mainly affects the frontal lobe and may show signs and symptoms of neurological deficits, changes in personality, memory or seizures. In this same aspect, schizophrenia can also present functional deficits, delusions, hallucinations, thought disorders and behavioral changes. Development: this work presents an analysis of primary tumors located in the frontal lobe, which occur with endogenous psychoses, as well as in cases of GBM and schizophrenia. Final considerations: it is important to have a correct diagnosis in order to initiate adequate treatment and alleviate both psychiatric and neurological symptoms.

Keywords: Gliomas. Glioblastoma. Schizophrenia. Cerebral cortex. Behavioral symptoms.

Corpus callosotomy in the treatment of refractory epilepsy in pediatric patients

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OBJECTIVES: The presenting literature review aims to evaluate the efficacy and reliability of corpus callosotomy (CC) as a palliative treatment in pediatric patients with refractory epilepsy. **METHODS:** Relevant studies were searched in the databases Pubmed, Scopus, and Lilacs. For such, the keywords "corpus callosotomy" AND "epilepsy" AND "pediatric" OR "child" were used. The following eligibility criteria were taken into consideration in the selection: descriptive studies published in English in the last 5 years; ≥ 20 patients from 0 to 19 years old presenting refractory epilepsy who underwent total or partial CC; and a minimum of 6 months of post-operative follow-up. The procedure's efficacy and safety, in addition to improvement in quality of life, were evaluated. **RESULTS:** In general, the most used surgical approach was total CC and the majority of patients presented more than one type of epilepsy. The percentage of seizure-free patients after CC was significant in two studies (21,7% and 35%) and more than half attained a reduction of at least 50% in epileptic crises. In one study, crisis reduction had a mean of 90% and 72,7% of patients reported improved quality of life. The number of direct complications in two studies was $\leq 10\%$ and deaths due to CC were rare. Neurological deficits were relatively common in one study, mostly transitory. **CONCLUSION:** Corpus callosotomy is an efficient palliative resource in the treatment of refractory epilepsy in pediatric patients. Furthermore, the number of direct complications and deaths are rare and neurological deficits are mostly temporary. **Keywords:** corpus callosotomy, palliative surgery, refractory epilepsy.

Post-trauma management of epilepsy in pediatric patients

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Objective: To analyze the different management of pediatric patients with epilepsy in a post-traumatic context, considering pathophysiological and social issues. **Method/materials:** The data search comprised the databases BVS, Scielo and Cochrane Library and used the combination of the descriptors "pediatric epilepsy", "trauma" and "management", as well as their respective ones in English language ("pediatric epilepsy", "trauma" and "management") and use of the Boolean operator AND. The inclusion criteria were articles published in English and Portuguese within the last 5 years. After eligibility analysis by reading the titles and respective abstracts, 18 articles were selected and read in full. **Results:** Approximately half of children with a history of head trauma develop epilepsy, with a greater number of seizures, and, on average, 3 years earlier than patients with epilepsy of non-traumatic origin. After

diagnosing epilepsy in pediatric patients, it is important to assess the responsiveness of less aggressive treatments, such as non-pharmacological management of the condition. Faced with the lack of response to the initial treatment, it is worth incorporating medication. In cases of pharmacological resistance, management for epilepsy through vagal stimulation is recommended, associated with a reduction in the frequency of seizures in 50% of cases. **Discussion and conclusion:** Childhood head injury (CBI) affects approximately 475,000 children under 15 years of age annually, mainly between 0 and 4 years of age. Due to the formation process of the cranial bone framework and intense childhood activity, the prevalence of cases of post-traumatic epilepsy in this age group is due to a greater risk of traumatic injuries, which can have serious consequences when affecting a system that is still under development. . After recognition, identification and diagnosis, therapeutic measures should be initiated in order to curb new crises, often by non-pharmacological means, in addition to the possibility of performing vagus nerve stimulation and other surgical procedures when there is poor response to drugs. This condition can lead to several neurological and psychiatric consequences, such as depression and neuropsychomotor delay, so early diagnosis and effective treatment of epilepsy are crucial to avoid permanent damage. children against these serious consequences.

Keywords: Pediatric epilepsy, Trauma, Management

Does maternal COVID-19 affect the neuromotor development of children in the first year after birth? A systematic review

Flávia Martins Gervásio, Daniela de Souza Silva, Aline Helena Nascimento Veloso

Objective: To identify in the literature about maternal contamination by SARS-CoV-2 the impact on the neuromotor development of babies in the first postnatal year. **Methods:** Systematic review registered in the International Prospective Register of Systematic Reviews (PROSPERO). **Databases:** Pubmed, Virtual Health Library and Embase. **Search strategy:** (SARS-CoV-2 OR coronavirus OR COVID-19 OR Severe Acute Respiratory Syndrome) AND (embryogenesis OR fetal medicine OR maternal infection OR intrauterine) AND (Infant OR newborn OR toddler OR neonate) AND (follow-up OR neurodevelopmental disorders OR neurodevelopmental outcomes OR motor outcome OR motor development OR motor function assessment OR neural impairment). **Inclusion criteria:** studies that evaluate the neurodevelopment from zero to one year of age, prospective or retrospective and case studies. Reviews, protocols, editorials and studies that do not use standardized instruments were excluded. After complete reading, articles were grouped by main outcomes, period and assessment instruments. **Results:** A total of 288 articles,

63 selected after reading the title and abstract and 14 articles were included after screening based on inclusion and exclusion criteria. There were 12 prospective longitudinal studies, one retrospective and one case report, one published in 2020, two in 2021, seven in 2022 and four in 2023, all in English. Instruments used were: Ages & Stages Questionnaires, Developmental Profile, Precht's General Movement Assessment, Denver II Screening, Alberta Motor Assessment Scale, Neonatal Behavioral Assessment Scale and head circumference. Preterm birth outcomes and longer periods of hospitalization have been related to severe forms of COVID-19. With low mortality rates, the babies were reassessed between the 3rd and 6th postnatal month, with normal development, even when compared to non-exposed babies, and mild delays in fine motor, social and language domains. Discussion and Conclusion: There was no significant delay in neurodevelopment in early childhood among neonates of mothers with gestational COVID-19 and preterm birth was an important associated condition. The pandemic context was considered by the authors as a stressor for development, regardless of exposure to the virus. There are limitations in the somatosensory investigation and malformations of other systems, themes that should be investigated in future studies.

Keywords: SARS-CoV-2, Pregnancy, Child Development.

Evaluation of motor function after selective dorsal rhizotomy in pediatric patients with spastic cerebral palsy, associated with GMFCS IV OR V

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Objective: To analyze post-surgical results of motor function in pediatric patients with spastic cerebral palsy using the Gross Motor Function Classification System (GMFCS) IV and V who underwent Selective Dorsal Rhizotomy (SDR).

Materials and Methods: This study carried out an integrative literature review in the PubMed(29), Scielo(0) and Lilacs(0) databases using the English descriptors "Selective dorsal rhizotomy", "GMFCS" and "Pediatric". Articles related to pediatric patients with spastic cerebral palsy grades IV and V on the GMFCS scale, over a period of 15 years (2008-2023), were included.

Results: A study developed by D'Aquino et al., 2017 and Nordmark et al (2008) demonstrated that patients undergoing SDR achieved significant improvement in lower (LLLL) and upper (MMSS) limbs over a period of 2, 8 and 14 months. The mean reduction in the Ashworth score for GMFCS IV and V patients was, respectively, 2.8 and 3.2 in the LL and 2.8 and 2.9 in the UL. The analysis revealed a high degree of safety for the procedure and indicates that, if associated with physiotherapy, it reduces the spastic condition in patients with GMFCS grades IV and V (D'AQUINO

et al., 2017). The mean number of rootlets cut between S2-L2 was 40%, resulting in an immediate reduction in muscle tone in adductors, hamstrings and dorsiflexors, with no recurrence of spasticity over 5 years.

Conclusion: it was concluded that the SDR showed positive results in improving the motor function of pediatric patients with spastic cerebral palsy of GMFCS grade IV and V with a significant improvement in spasticity.

Keywords: Selective dorsal rhizotomy, spastic cerebral palsy, GMFCS IV-V

Current perspectives on vagus nerve stimulation in Lennox-Gastaut syndrome in children

Francisco Teles de Macedo Filho

INTRODUCTION: Lennox-Gastaut syndrome (LGS) is characterized by various seizures (tonic, atonic, atypical absences), behavioral and cognitive changes. Refractoriness to multiple medications is also a characteristic. Thus, on July 16, 1997, the VNS (Vagus Nerve Stimulation) Therapy System™ (Cyberonics, Inc) was approved by the Food and Drug Administration (FDA) as a therapy, with refractory LGS being a specific indication. **OBJECTIVES:** The authors conducted a systematic review to evaluate published outcomes of VNS for medically refractory epilepsy from the perspective of LGS in children according to evidence-based criteria in published articles. **METHODS:** A systematic review was performed using the databases Scielo, Uptodate, Bireme, Pubmed, and Medscape. This search was further limited to articles published between 1980 and 2022. Accordingly, articles were examined for inclusion or exclusion in the final analysis based on the following criteria: diagnosis of LGS, age from newborn to 15 years old, average follow-up of 1 year or more, inclusion of medically refractory epilepsy, and preoperative consistency with surgical evaluation. When assessing the effectiveness of VNS therapy, seizure severity was considered, along with rates of seizure frequency reduction. **RESULTS:** The search of the cited references resulted in 28 articles that met the search criteria. The vast majority of articles, around 75%, reported results in terms of a reduction of 50% or more in seizures, three studies reported seizure freedom through a follow-up of 1 year or more, and two articles reported reduction in medication dosage to control seizures, within a follow-up period of over 2 years. In all analyzed articles, it was also observed that VNS is associated with lower rates of complications. **CONCLUSION:** VNS is a safe and effective alternative for patients with medically refractory epilepsy in LGS. However, the evidence for these results is still limited by the small number of studies present in the literature. Further research is needed to assess the efficacy and tolerability of VNS from the perspective of refractory LGS, as

well as its indications for greater effectiveness in the pediatric population.

Keywords: Lennox-Gastaut syndrome, Vagus nerve, Epilepsy

Costs of pediatric hydrocephalus treatment for the Brazilian public health system in the Northeast of Brazil.

Luan Guanais Soriano, José Roberto Tude Melo

Purpose: To estimate the costs of the surgical treatment of pediatric hydrocephalus, specifically ventriculoperitoneal shunt (VPS) and endoscopic third ventriculostomy (ETV), for the Brazilian public health system (SUS). **Methods:** Retrospective cohort study of health records of patients < 14 years of age with a diagnosis of hydrocephalus who underwent VPS or ETV between September 2009 and June 2016, regularly followed up for 24 months. **Results:** Seventy-six medical records were included. The groups of children who underwent VPS and ETV consisted of 60 and 16 patients, respectively. Complications during two years of follow-up were identified in 56% of the children undergoing VPS and in 18% of those undergoing ETV ($p = 0.0103$). The initial cost of VPS was lower than that of ETV up to approximately 1 year of post-surgical follow-up. After that, VPS generated higher expenses for the SUS due to higher rates of late post-surgical complications and repeated readmissions. **Conclusion:** Higher public expenditures were observed in the group of children undergoing VPS due to higher rates of infectious and mechanical complications requiring repeated hospitalizations and prosthesis replacements. Public policies must be tailored to offer the best treatment to children with hydrocephalus and to make judicious use of public resources without compromising the quality of treatment.

Keywords: Public Health, Health Systems, Neurosurgery, Pediatrics, Public policy.

Epidemiological analysis of absolute mortality in up to 1 year-old live births with hydrocephalus in Brazil from 2010 to 2020

Lauro Arthur Benvneutti, Matheus Felipe Kuhn Urnau, Leonardo Diehl Filippio, Charles Andre Carazzo

Objective: Hydrocephalus is a disorder in which an excessive quantity of cerebrospinal fluid accumulates inside the cerebral ventricles and/or subarachnoid space. Given its elevated morbimortality and this study aims to epidemiologically analyze the prevalence of hydrocephalus in live births (HLB) and absolute mortality in children under 1-year-old due to hydrocephalus in Brazil between 2010 and 2020. **Methods:** A descriptive retrospective study was conducted using data from the Brazilian National Health System (DATASUS) to determine the number of HLB and absolute mortality in children under 1-year-old. Variables included HLB, mortality rate, sex, race, and region. **Results:**

In 11 years, Brazil reported 6,985 cases of HLB and 894 deaths in children under 1 year old due to hydrocephalus. The Southeast (36.93%) and Northeast (30.95%) regions had the highest HLB numbers, while the Northeast (41.61%) had the highest mortality rate. Male cases accounted for 53.99% of HLB and 53.57% of deaths. Brown children constituted the largest portion of HLB (54.20%), and white children had the highest mortality rate (45.41%). The mortality rate due to hydrocephalus was 12.79%, with the high rate in the Northeast. **Conclusion:** Between 2010 and 2020, Brazil experienced 6,985 cases of HLB and 894 deaths in children under 1 year old due to hydrocephalus, resulting in a mortality rate of 12.79%. The Southeast exhibited the highest HLB count, while the Northeast had a high mortality rate. Male patients and white children had the highest number of deaths.

Keywords: Hydrocephalus, absolute mortality, epidemiological analysis, children

Study of 48 cases of patients submitted to ventriculoperitoneal bypass using a particular technique for surgical field assembly

Júlia Santana de Oliveira, Henrique Rodrigues Osorio, João Victor Rodrigues Bubicz, Georgia Varaschin Debes, Lohana Pompelli Scapatici

Ventriculoperitoneal shunt (VPS) is a modern and efficient treatment, but it still presents morbidity and mortality due to complications related to the procedure. The medical records of 48 patients were reviewed and an epidemiological analysis was carried out. Among the patients 42,2% were newborns, 16% were between 28 days and 6 months, 15.6% between 6 months and 2 years, 11.1% between 2 and 12 years and 16% over 12 years, predominantly males (69%). The main etiologies included malformations (21%), neoplasia (19%) and myelomeningocele (10%). The most common symptoms before surgery were vomiting (42%), lowered level of consciousness (35%), headache (27%) and macrocrania (51% of patients younger than 2 years). The technique that precedes the procedure consists of degerming the skin, followed by placing compresses around the surgical field with OPSITE dressing and then the sterile drapes are positioned. It is believed that the method positively influences the infection rate of surgeries, which was 14.6%, consistent with literary findings in other hospitals, of 15% (take into account that this is a university training hospital, that not all surgeries were performed in the first hour and among other limitations of the study). Among the main infectious symptoms, fever (33%) and valve malfunction (44%) were observed. In addition, 42% of the patients underwent PVD revision and 31% underwent device replacement. death rate found was 10.4%, also similar to the bibliography. Longer follow-up time would be necessary for better evaluation

Keywords: Shunt, Hydrocephalus, SURGICAL TECHNIQUE

The pediatric Traumatic Brain Injury approach

Jeanne Beatriz Nunes da Silva, Lucas Amaral da Silveira, Lara Kaiulani Laounier, Laís Emanuelle Lamounier, Daniela de Stefani Marquez

Objective: This work aims to discuss the approach to pediatric traumatic brain injuries. **Materials and methods:** This integrative literature review was based on the search for articles published in the last 5 years, in the BVS database, using the descriptors "TBI" and "Pediatric", combined by the Boolean Operator AND. It resulted in 105 articles, of which 2 were selected that presented information relevant to the objective of the work. **Results:** Childhood TBIs represent a high number of emergency calls, an alarming fact, considering that major neurological changes during the period from 0 to 4 years represent an important cognitive and behavioral deficit for the patient's life. The use of treatments directed to adults in children is a widely discussed and relevant subject, since studies indicate that the propaedeutics should not be similar given the anatomical and physiological differences. Changes such as increased intracranial pressure (ICP), cerebral pressure perfusion, systemic arterial pressure and Average arterial pressure should be related to infant parameters and their corrections. Furthermore, the increase in infant ICP is relative, since the presence of sutures and fontanelles not yet closed may cause the cranial vault to become malleable and the brain to accommodate. The complete analysis of the child's condition, with the assistance of tools to quantify the commitment is important and widely implemented, in order to correct the affected areas in an agile and skillful manner, since the treatment time influences the presence or absence of neurological sequelae. Therefore, research is needed to describe and implement standards for the management of children who have suffered TBI, with possible clinical and behavioral changes, and to avoid changes in neuropsychomotor development.

Keywords: Neurosurgery, Pediatrics, TBI

Morbidity and public health expenditures in traumatic brain injury in childhood-adolescence in the northern Brazilian region: a retrospective ecological study.

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Objective: To describe, in the child and adolescent age group, injuries of Intracranial and/or Cranioencephalic Trauma (TBI), North-Brazil region, whose outcomes were morbidity and costs of medical-hospital care. **Method:** Ecological time series study, descriptive, January 2017 to January 2023, data from the Department of Informatics of

the Unified Health System (SUS) and Hospital Information Systems of SUS: morbidity, hospitalization rates and costs in the conservative treatment of medium and severe TBI. **Results:** Age range 1 to 14 years, with 1 to 4 years, the highest number of cases (2648), followed by 5 to 9 years (1709), male sex (4645) higher than female (2924). There were 7569 hospitalizations, mortality 134 cases (mortality rate 1.77), being spent by the Federal Government R \$ 6,030,722.43 reais, in cases that evolved to death, whose average value of authorization for hospitalization R \$ 796.77 reais, with an average of 30 days of hospitalization. It is noteworthy that the age group analyzed represents 5.3% of all deaths compared to any age group with this diagnosis. **Discussion:** The costs and the high rate of death in childhood-adolescence by TBI is a worldwide concern. The adoption of screening protocols / early diagnosis of injuries and assertive treatment are measures adopted in Europe and the United States to minimize human and financial damage, unlike the region studied. **Conclusion:** High morbidity and health expenditures were identified in the Northern Region of Brazil due to TBI with a high degree of risk of sequelae, since they are moderate or severe injuries.

Keywords: Traumatic Brain Injury, Public Health Expenditure, Childhood Mortality.

Incidence of shaken baby in the midwest of Brazil

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Objective: The objective is to analyze quantitative data on the incidence of intracranial trauma (TBI) resulting from maltreatment in the age group of 12 months to 4 years in the states of the Midwest of Brazil, with the purpose of tracing measures of tracking for cases of risk in the most incident locations. **Method:** This is a quantitative study with analysis of data collected by the Department of Informatics of the SUS (DATASUS), through the SUS Hospital Information System database (SIH/SUS) by place of residence, by the number of hospitalizations according to the central-west federative region, selecting Intracranial Trauma and Maltreatment Syndrome in the ICD-10 Morbidity List and restricting the age group to 12 months to 4 years, during the periods from October 2020 to October 2022, making state percentage of referrals CID, one should pay attention to the population number of these states. **Results:** The number of 693 cases of trauma due to abuse resulted. When we evaluate these data by quantity and states we have: Mato Grosso do Sul 14.2%, Goiás 21.5%, Mato Grosso 30.1% and Distrito Federal 34%. **Discussion and Conclusion:** When quantitatively analyzing the incidence of TBI related to abuse, it is noted that the highest concentration of cases

occurs in the DF in terms of number of occurrences. It is concluded that, despite being a predictable type of trauma, there is a high incidence rate in the region and, therefore, it is essential that there is greater awareness and mitigation in relation to this type of tragedy, involving both legal guardians.

Keywords: Pediatric neurosurgery, midwest, shaken baby

Epidemiology of the number of hospitalizations and absolute mortality by malignant neoplasm of the encephalon and other central nervous system structures from 0 to 14 years old in Brazil from 2010 to 2020.

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OBJECTIVES: Malignant neoplasms of the encephalon and other central nervous system structures (MNECNS) present significant prevalence in children. Due to scientific scarcity and the subject's importance, this study aims to epidemiologically analyze the number of hospitalizations and absolute mortality by MNECNS in children. **METHODS:** Descriptive retrospective study about the number of hospitalizations and absolute mortality by MNECNS in Brazil from 2010 to 2020 in children from 0-14 years old. Data were obtained through the Department of Informatics of the Brazilian National Health System (DATASUS). This system shows limitations, such as the underreporting and the non-differentiation of the neoplasm's histologic types. The variables utilized were the number of absolute deaths, hospitalizations, age group, sex, race, and region. There was no need for approval of the Ethics Committee for data collection since this study uses publicly available data. **FINDINGS:** In Brazil, 5424 absolute deaths and 32,458 hospitalizations due to MNECNS were reported in children from 0-14 years old, from 2010 to 2020. Southeast region had highest numbers of absolute deaths (39,14%) and hospitalizations (45,17%). White children represented the majority of absolute deaths (51,07%) and hospitalizations (40,76%). The age with most cases of either absolute deaths (35,9%) and hospitalizations (35,96%) was 5-9 years old. Boys represented 52,64% of absolute deaths and 55,99% of hospitalizations **CONCLUSION:** Between 0-14 years old, the hospitalizations and absolute deaths caused by MNECNS were more common in children of 5-9 years old, white individuals, boys, and in the Southeast of Brazil (which presented a higher overall number of cases).

Keywords: Malignant neoplasms, encephalon and other central nervous system structures, children, absolute mortality, hospitalizations

H3K27-altered diffuse midline glioma: report of a case with atypical clinical and radiological presentation

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This is a case report of a 3-year-old patient, at first manifesting symptoms of an ischemic stroke, MRI indicating a potential meningioma, but with posterior anatomopathological confirmation of a midline diffuse glioma. To evaluate this type of glioma and its implications, it was performed a review of medical records and literature with articles between 2015 and 2023. Research included the databases: SCIELO and PUBMED.

The high-grade gliomas constitute 20% of childhood gliomas, including anaplastic astrocytomas, diffuse gliomas and glioblastoma multiformes. The clinical picture is usually progressive, because it rises in the pons and infiltrates the brainstem. This situation may cause bilateral neurological deficits, signs of brainstem dysfunction or liquoric obstruction.

The case is about a diffuse midline glioma, with the H3K27M mutation, classified as IV grade of OMS. This type of glioma represents approximately 10% of all childhood cerebral tumors. The presence of this mutation indicates a bad prognosis, with a median survival of a year. About the treatment, a surgical resection has a main roll at the increase of the median survival of the patient. Intraoperatively was found an intra-axial lesion, hardened and well-vascularized. Partial resection carried out due to hemodynamic instability. Intraoperative impression indicated a possible astrocytoma. Good postoperative progress with no other deficits. Developed hydrocephalus and underwent subsequent placement of a VP shunt. Following the histopathological result, the diagnosis of altered H3K27 diffuse midline glioma was confirmed, requiring chemotherapy with temozolomide and etoposide. The importance of understanding possible different clinical and radiological manifestations of pediatric gliomas is reaffirmed, where early diagnosis is essential.

Keywords: midline glioma, supratentorial, H3K27-altered

Conservative and surgical treatment of arachnoid cyst: literature review

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Objective: The aim of this study is to present the clinical and surgical treatment of arachnoid cysts in pediatric patients. **Method/materials:** The research was carried out from the BVS, Scielo and Cochrane Library databases and the descriptors used were "Tratamento cisto aracnóideo" and "Manejo cisto aracnóideo", as well as their respective in English ("arachnoid cyst treatment" and "arachnoid cyst management"). The applied criteria included articles in

Portuguese and English and published in the last 5 years. From the eligibility analysis by reading the title and their respective abstracts, 27 articles were selected, which were read in full. Results: Arachnoid cysts (AC) are prevalent in children aged between 1 and 10 years. Although conservative management is the most applied, headache, vomiting and increased intracranial pressure (ICP) were the most frequent clinical presentations and the most indication for surgery. Obstructive hydrocephalus was also a common finding in the studies. Increased ICP was observed even during pregnancy in most cases of patients younger than 1 year old, with the diagnosis made in utero, and is related to developmental delay. Endoscopic fenestration was the most frequently performed procedure, with the best postoperative outcome for CA, leaving cyst bypass as the second option. The need for a revision surgery is strictly associated with 1 year after the initial operation, and in these cases, refenestration and/or placement of an additional shunt were performed. Midline interhemispheric cysts usually require a second intervention and cause higher rates of neurodevelopmental deficit. Discussion: AC in pediatric patients have different anatomical locations, which is why there are different clinical manifestations. The most affected sites are the middle cranial fossa (40.5%), posterior fossa (24.3%) and interhemispheric fossa (13.5%), leading to clinical alterations such as: increased intracranial pressure (34.2%), obstructive hydrocephalus (28.9%) and AC rupture (21.1%). Treatments consist of clinical and surgical approaches (craniotomies, endoscopic fenestrations, cystoperitoneal shunts, drainage operations with trepanning, and spinal CA excision). Conclusion: The therapeutic approach for pediatric patients points to an individualized approach due to the wide spectrum of anatomical locations and clinical manifestations associated with the condition.

Keywords: Arachnoid cyst, Conservative treatment, Surgery

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Júlia Santana de Oliveira, Valentina Lima Cartaxo da Silva, Júlia Morais Zandavali

This is a case report of a 3-year-old patient, at first manifesting symptoms of an ischemic stroke, MRI indicating a meningioma, but with an anatomopathological confirmation of a midline diffuse glioma. To evaluate this type of glioma and its implications, it was performed a review of records and literature with articles between 2015 and 2023. Research included the databases: SCIELO and PUBMED.

The high-grade gliomas constitute 20% of childhood gliomas, including anaplastic astrocytomas, diffuse gliomas and glioblastoma. The clinical picture is usually progressive, because it rises in the pons and infiltrates the brainstem. This situation may cause bilateral neurological deficits, signs of brainstem dysfunction or liquoric obstruction.

The case is about a diffuse midline glioma, with the H3K27M mutation, classified as IV grade of OMS. This type of glioma represents approximately 10% of all childhood cerebral tumors. The presence of this mutation indicates a bad prognosis. About the treatment, a surgical resection has a main roll at the increase of the median survival. In intraoperative was found a Intra-axial lesion, hardened and well-vascularized. Partial resection carried out due to hemodynamic instability. Intraoperative impression indicated a possible astrocytoma. Good postoperative progress with no other deficits. Developed hydrocephalus and underwent subsequent placement of a VP shunt. Following the histopathological result, the diagnosis of altered H3K27 diffuse midline glioma was confirmed, requiring chemotherapy with temozolomide and etoposide. The importance of understanding possible different clinical and radiological manifestations of pediatric gliomas is reaffirmed, where early diagnosis is essential.

Keywords: midline glioma, supratentorial, H3K27-altered

Schmahmann's disease and posterior fossa tumors in children: service analysis of the last 05 years

Tiago Paiva Cavalcante, Rilton Marcus Morais

Infratentorial tumors in pediatrics account for 50% of those in the central nervous system. Of these, about 70% originate in the cerebellum-pontine-mesencephalic tract. In addition to the complications related to hydrocephalus, cerebellar motor syndrome and vestibulocerebellar syndrome, Schmahmann's syndrome or cognitive-affective cerebellar syndrome is responsible for serious behavioral sequelae that impact the social and family life of these patients. We present our experience over the last 05 years in the diagnosis of Schmahmann's syndrome, etiological factors involved in cerebellar anatomical location, age, nature of tumors involved and type of surgery performed.

We included 18 patients (9M: 10F), mean age 5.6 years (2 - 14y), not submitted to previous oncological surgeries or tumors in other parts of the body, who developed neurobehavioral symptoms during the post-surgery outpatient follow-up, operated in the Pediatric Neurosurgery service between January 2019 and March 2023.

Cerebellar affective cognitive symptoms occurred in 70% of the cases, more related to solid pilocytic astrocytomas of the posterior fossa, bilateral paramedian location. Male patients were more affected and there was no specific age group. Symptoms such as mental rigidity, social isolation, loss of language intention ability, irritability, personality change occur frequently in pediatric cancer patients and are attributed to post-traumatic stress from surgeries, long hospital stays, need for radiochemotherapy. However, injury to the frontopontocerebellar tract in infratentorial tumors should be objectively treated as a sequel to the

surgical process with neuropsychological support, scales and specific medications.

Keywords: Brain tumors, cognitive sequelae, posterior fossa tumors

Mortality from neoplasms in the pediatric population in Brazil

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Objective: Malignant brain tumors are the leading cause of mortality from malignant neoplasms in children. The goal of this study is to characterize the national epidemiological profile of mortality in pediatric patients with malignant neoplasms in the brain between 2018 and 2022. **Methods:** An epidemiological study was conducted using the Informatics Department of the Brazilian Unified Health System (DATASUS). Data were extracted on mortality from malignant brain neoplasms in patients between 0 and 19 years old in Brazil between 2018 and 2022, stratifying by year, age group, sex, ethnicity, and region. Statistical analysis was performed using Microsoft Excel. **Results:** Between 0 and 19 years, a total of 877 deaths were found, with a higher incidence in children in the 5-9 age group, with 292 deaths (33%). Patients with 1-4 years old were the least affected, with 35 deaths in total. The Southeast region had the highest number of cases, with 39% (N=345) of deaths, followed by the Northeast with 29% (N=258), South with 14% (N=127), North with 9% (N=83), and Midwest with 7% (N=64). In the Southeast region, there was no significant discrepancy between genders, with 50.4% females and 49.6% males, and this slight disparity between genders is maintained in the other regions. When comparing the year with region and age group, cases were more predominant in 2021 with 180 deaths (20.5%), with higher rates in the Southeast region. In terms of age, there was a hegemony in the 5-9 age group, accounting for 35% of mortality, followed by 10-14 years with 22%, 1-4 years with 21%, 15-19 years with 18%, and under 1 year old with 3%. **Discussion and Conclusion:** The high mortality rate from malignant brain tumors in the pediatric population requires attention to issues that may hinder diagnosis, making it late, and treatment, such as interruption due to social isolation during the Covid-19 pandemic that began in 2020. Sex appears not to be a determinant to indicate higher mortality, since the discrepancy is small, and mortality appears to be directly proportional to the population size, increasing in the most populous regions. Considering the importance of having constant and updated epidemiological studies, identifying the age group of 5-9 years old as having the highest number of deaths from malignant neoplasms in the brain is relevant

information for characterizing and conducting possible associations on the population profile, as well as directing health planning actions.

Keywords: Brain tumors; Mortality; Pediatric Neurosurgery; Children

The role of preoperative tractography in posterior fossa tumors in the prevention of cerebellar mutism in childhood

Brigida Schembida de Oliveira, Marcos Vinicius Calfat Maldaun, Nelci Zanon Collange

Introduction: Tractography is the only method currently available to indirectly demonstrate white fiber tracts in the central nervous system in vivo. However, its usefulness in terms of use for posterior fossa pathologies hasn't been proven, especially when applied to the analysis of tracts in cerebellar mutism syndrome. **Methodology:** We systematically reviewed studies carried out for posterior fossa tractography applied to cerebellar mutism published on the last ten years, comparing results of alterations seen in fraction of anisotropy at the dentato-rubro-thalamo-cortical tract of patients submitted to the exam in the pre and/or in the postoperative period. We aimed to verify if positive alterations found could be consistent with the posterior fossa syndrome symptoms. **Results:** After exclusion criteria we found four articles in which 33 patients had posterior fossa syndrome against 96 not affected; mean age was variable between studies, ranging from 8,4 +/- 4,4 to 10,3 +/-4. Main tumors were medulloblastoma from the fourth ventricle. Fractional anisotropy changes weren't statistically significant in patients with postoperative cerebellar syndrome compared with controls. **Conclusion:** It's not yet possible to make objective conclusions about the validity of the test in cerebellar mutism based on FA. Because of the complexity of neuronal connections in human tracts, we can't consider only fractional anisotropy as an isolated index for assessing the integrity of these connections nor as the main factor for predicting cerebellar mutism even if it was altered preoperatively during tractography.

Keywords: Tractography; Posterior Fossa Syndrome; Cerebellar Mutism; Fractional Anisotropy (FA)

James Tait Goodrich and the pioneering separation of craniopagus conjoined twins: a bibliography

Carolina Guimarães Moura, Ana Laura Fernandes Scarelli, Nathália Bononi Candido Mendes, Lara Samanta Wagner, Nicollas Nunes Rabelo

Introduction: James T. Goodrich was a pediatric neurosurgeon who introduced staged surgery in craniopagus twins separations. Thus, this article explains the sequence of events that led him to be a pioneer.

Objectives: This article was developed in order to seek original scientific and bibliographic information to clarify the life path of Dr. Goodrich.

Discussion and Conclusion: James Tait Goodrich was born in Portland, Oregon in 1946. During the Vietnam War, James served in the U.S. Navy. He completed his undergraduate degree at the University of California, Irvine and his graduate degree from Columbia University's (1978-1980). In 1986, he became a neurosurgeon, later becoming director of the Division of Pediatric Neurosurgery at Montefiore Medical Center.

Sadly, on 2020, Goodrich passed away due to complications from the coronavirus after a 30-year career in neurosurgery. However, his legacy still lingers in the methodology of craniopagus separation.

Before Goodrich, the separation was performed in a single long surgery with high mortality and morbidity rates due to the difficulty in forming collateral circulation. Therefore, Goodrich developed a pioneering multi-stage separation protocol, including adequate time between surgical approaches for recovery of the cerebral vasculature. He also highlighted the role of plastic surgery in preventing postoperative complications and the anesthetic difficulties in separating fused brains.

Thus, Goodrich became known worldwide as the leading expert in the separation of craniopagus. In Brazil, he guided several procedures, including the separation of Maria Ysabelle and Maria Ysadora, in Ribeirão Preto-SP (2018), and also of Lis and Mel, in Brasília-DF (2019).

Keywords: Craniopagus, Neurosurgery, Goodrich

Intracranial arachnoid cysts and their familial relationship: case report and literature review

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Objetives: to investigate the genetic relationship of intracranial arachnoid cysts (IAC) occurrence in pediatric patients from the same family.

Methods: PubMed was searched for studies addressing the genetic explanation of familial incidence of IAC in pediatric patients. The medical records of two brothers with IAC at Hospital UDI, São Luis-MA were reviewed.

Results: Nine selected articles demonstrated possible genetic evidence in familial IAC, with mutations being described in several genes and proteins, such as in SPG4/GPSM2/KIF7/FOXC2. Reported clinical cases that both brothers had a large fronto-temporal ACI, with the 4-year-old brother having seizures. The 1 year and 6 month old child showed only asymptomatic increase in head circumference, with alterations only in the EEG. Both were operated on, uneventfully and with improvement in the exams.

Discussion and Conclusion: IACs are usually asymptomatic but can cause headache, epilepsy, vertigo, nausea, cognitive impairment, being diagnosed by CT/MRI and treated conservatively or surgically in symptomatic cases. Patients with genetic and Mendelian syndromes, such as Chudley-McCullough/Acrocallosa/Down or Noonan syndrome, have a higher frequency of familial IAC. Although most IACs are sporadic, there is a likely pathophysiological relationship with mutations involved in microtubular dynamics, transcriptional regulation, and signaling pathways in brain embryogenesis. Future studies are needed to understand the genetics involved in IACs and their familial manifestation. The search for genetic patterns of the reported siblings should also be carried out in the future.

Keywords: Intracranial; Arachnoid cyst; Tumor; Relatives; Genetics

Surgical management of brain arteriovenous malformation in the pediatric patients: experience of a regional reference center.

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Objective: To present the experience in intracranial arteriovenous malformation (AVM) in pediatric patients treated at the Hospital da Criança de Brasília in a period of 5 years and the results after surgical intervention.

Material and Methods: All patients with radiological diagnosis of AVM identified between March 2018 and March 2023 were included. The inclusion criteria were: age below 18 years, confirmed radiological investigation. The clinical presentation, malformation characteristics (size, location, nutrition, drainage and classifications), and treatment performed were analyzed. The extent of resection, trans- and post-operative complications, the recurrence rate and the functional status (modified Rankin Scale-mRS) were the outcomes evaluated. The extent of resection was established primarily by angiotomography from 3 months postoperatively, with doubtful cases undergoing arteriography.

Results: Seventeen patients met the inclusion criteria: 15 cases (84,5%) had intracranial hemorrhage as the main manifestation, followed by epilepsy (2 cases) and headache (1 case). All patients were investigated with angiography. The decision for surgery were taken with endovascular section. The main indication for microsurgical treatment were the feasibility and safety of the procedure. Nine patients had 10 surgeries (1 after recurrence), and 88% of gross total resection rate. There were no trans- and post-operative complications in the surgical group. There is no significant change in functional performance in modified Rankin scale.

Conclusion: AVMs are a prevalent cause of intracranial hemorrhage in pediatric patients. Treatment should be as comprehensive as possible. The surgical procedure proved to be a safe and efficient option, without functional impairment and with adequate disease control.

Keywords: Vascular malformation, brain arteriovenous malformation, brain hemorrhage

children are challenging, due not only to its correlation to the device's failure and bigger incidence in the first months of life but to the diagnostics imprecision. Prospective studies that focus on the patient's medical, microbiological and surgical risk factors are extremely necessary for an improvement in the management, diagnosis and prophylaxis of infections.

Keywords: CSF infection, CSF derivation, hydrocephalus

Infection in Cerebrospinal Fluid Derivations in Pediatric Patients

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Objective: Cerebrospinal fluid (CSF) derivations are used in hydrocephalus to divert accumulated CSF. Infections in derivations frequently develop through skin flora colonization, which may occur in the surgery or the first weeks afterwards, due to a wound or skin rupture, and *Staphylococcus* is the most common pathogen. The goal of this study is to describe the current literature about the diagnosis and approach of CSF derivations infections in the pediatric population.

Methods: Systematic reviews or meta-analyses published between 2019 and 2023 were included and studies that were not about the pediatric population were excluded. The keywords "cerebrospinal fluid shunt infection in children" and "pediatric cerebrospinal fluid shunt infection" were used in the PubMed platform. 3 studies out of 17 were selected.

Results: Infection is an important cause of complications related to CSF derivations implantation in pediatric patients. This condition is responsible for 45% of failures in the first month after surgery, occurring between 3 and 15% of implantations, where 90% of infections occur in the first 6 months. Risk factors for this infection in children involve premature birth, younger age, previous neurosurgery, gastrostomy tube and previous infections, the number of people in the operating room in the shunt placement, exposure to perforated surgical gloves, intraoperative neuroendoscope usage, and surgical and antiseptic techniques less than meticulous.

Discussion and Conclusion: The derivation infection is oligosymptomatic, with or without unspecific symptoms and infection signs. If clinical manifestations suggest a possible infection, a diagnostic evaluation begins with CSF analysis, with pleocytosis as the main alteration. Management of this condition includes the device's removal, external drainage, parenteral antibiotics and derivation substitution when the CSF is sterile. Prevention measures include meticulous surgical and antiseptic techniques, time of surgical procedures and preoperative antibiotic prophylaxis. Diagnosis and management of CSF derivations infections in