ABSTRACTS

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TUMORS

Multiple surgeries as a way of approaching large brain tumors in the pediatric population

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Objective: To describe a case of a rare large pediatric brain tumor that underwent 3 surgeries to achieve complete resection.
Case Report: A 3-year-old male patient, weighing 11 kg, presented with vomiting, weight loss, headache, and strabismus. Magnetic resonance imaging (MRI) revealed a large expansile lesion in the occipital horn of the left lateral ventricle, with the largest diameter of 6.5 cm. Surgery was indicated and partial resection of the tumor was performed due to intraoperative instability related to tumor bleeding. After clinical stabilization, a new intervention was performed with additional resection of the tumor, but still partial, again interrupted due to hemodynamic instability. A third intervention was scheduled after clinical improvement, and en bloc resection of the remaining lesion was then performed.
Result: Postoperative MRI showed complete resection of the tumor. The pathological examination together with immunohistochemistry diagnosed choroid plexus carcinoma. The patient presented progressive neurological improvement and continued oncological treatment
Conclusion: Large tumors in small pediatric patients are a major challenge for the neurosurgeon, and may require multiple interventions to achieve complete resection

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Keywords: Oncology, Brain Tumor, Neurosurgery

BRAF Inhibitors in the Management of Low-Grade Gliomas in the Pediatric Population: A Systematic Review and Meta-Analysis

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Introduction: Gliomas are central nervous system tumors classified according to histological features, degree of aggressiveness, and molecular markers. The identification of BRAF V600 mutations in low-grade gliomas (LGG) has raised interest in the use of BRAF inhibitors (BRAFis) as a targeted therapy. This study aims to evaluate the efficacy of BRAFis in the treatment of LGG in the pediatric population.
Methods: A systematic search was conducted in PubMed and Embase to identify studies reporting the use of BRAFis in pediatric patients with LGG. Clinical trials, retrospective cohorts, case-control studies, and case series were included. Statistical analyses were performed using OpenMeta Analyst. Heterogeneity was assessed using the I^2 statistic, and a random-effects model was applied to all outcomes.
Results: A total of 604 patients from 9 studies were included in this systematic review. A subgroup of 387 patients under the age of 25 with LGG received treatment with BRAFis. The most commonly used drug was dabrafenib, followed by tovorafenib and vemurafenib. Statistical analysis revealed a significant Objective Response Rate (ORR) to BRAFi treatment, estimated at 45.9% (95% CI: 35.9%–56%; I^2 = 66.23%; $p < 0.001$). The 12-month Progression-Free Survival (PFS) rate was 85% (95% CI: 77.7%–92.3%; I^2 = 62.166%; $p < 0.01$).
Conclusion: The use of BRAF inhibitors in pediatric patients with LGG is associated with a significant ORR and a PFS comparable to other therapeutic approaches, suggesting a meaningful antitumor effect. Further studies, particularly those including control groups, are warranted to better assess the long-term benefits of this therapeutic strategy.

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Keywords: Low-grade glioma (LGG), BRAF inhibitors (BRAFis), Targeted therapies, Glioma

Disseminated alveolar rhabdomyosarcoma in the spinal column

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Alveolar rhabdomyosarcoma is a rare and aggressive malignant tumor, representing 5–8% of pediatric cancers, with spinal involvement being an uncommon manifestation. We report the case of a 15-year-old female who presented with cervicobrachialgia and progressive motor weakness of the left upper limb. During hospitalization, she developed paraparesis and complete plegia of the left upper extremity. Magnetic resonance imaging revealed multiple expansile lesions throughout the spinal column, with intradural extramedullary, epidural, and soft tissue extension from the atlantoaxial junction to the L3–L4 level. The patient underwent bilateral hemilaminectomy from T4 to T7 with gross total resection of the extradural lesion. Despite surgical intervention, she developed acute respiratory failure ten days after admission, progressing to cardiopulmonary arrest and death. Histopathological and immunohistochemical analysis confirmed the diagnosis of alveolar rhabdomyosarcoma. This case highlights the aggressive behavior and poor prognosis associated with spinal alveolar rhabdomyosarcoma, emphasizing the need for early recognition and multidisciplinary management.

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Keywords: Alveolar Rhabdomyosarcoma, Spine, Aggressive Tumor, Pediatric Malignancy

A Self-Supervised Artificial Intelligence Framework for Objective and Accessible Interpretation of Brain Tumor Histopathology

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Modern neuropathological classification of brain tumors increasingly relies on molecular profiling. However, histopathological interpretation of hematoxylin and eosin (HE)–stained slides remains foundational for diagnosis—yet is often poorly understood outside the community of practicing pathologists due to its highly specialized nature. Recent advances in artificial intelligence—particularly self-supervised foundation models—offer a novel approach to characterizing complex histomorphological features in a more objective and accessible manner. In this study, we investigated whether embeddings derived from a self-supervised foundation model, referred to as Virchow 2, could effectively cluster a large dataset of brain tumor images. Using the Euro eBrains dataset of approximately 3,000 whole-slide images encompassing 126 tumor types, we extracted 24 million image patches (256×256 pixels at 20× magnification). A random sample of 2.4 million patches was embedded into a 2,560-dimensional feature space by

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Virchow 2. We then applied UMAP for dimensionality reduction and visualized the resulting projections in Cosmograph, labeling points by tumor type and relevant metadata. Our findings revealed distinct clusters corresponding to certain tumor subtypes, suggesting that self-supervised embeddings capture meaningful histopathological features. However, regions of overlap in the embedding space indicate non-discriminatory features or shared morphologies. These results underscore the potential utility of foundation model embeddings for brain tumor classification and pave the way for more scalable, objective, and integrative histopathological analyses.

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Keywords: Artificial intelligence, Brain tumor histopathology classification, Self-supervised learning, Foundation models (Virchow 2), Embeddings and clustering

Impact of Preoperative Embolization in Choroid Plexus Lesions

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Objective: To report a rare case of choroid plexus carcinoma in a pediatric patient with a prior history of abdominal neuroblastoma, highlighting the role of preoperative embolization in surgical management.
Case Report: A 4-year-old male presented with mild abdominal pain persisting for 30 days. Physical examination revealed a palpable abdominal mass. Histopathological analysis following biopsy confirmed undifferentiated neuroblastoma. Staging investigations included whole-body imaging and brain MRI, which revealed a lesion in the choroid plexus near the atrium of the left lateral ventricle, as well as a second lesion with similar radiological characteristics in the cerebellopontine angle cisterns. The patient underwent microsurgical resection of the first lesion via a right-sided approach, complicated by significant intraoperative hemorrhage. Intraoperative frozen section analysis was consistent with choroid plexus carcinoma. Prior to resection of the contralateral lesion, preoperative embolization of the tumor's feeding arteries was performed using Onyx, significantly improving surgical visualization and hemostasis during the second procedure.
Results: Preoperative embolization provided substantial benefit in achieving hemostatic control, reducing intraoperative bleeding during both surgical interventions.
Conclusion: In patients with choroid plexus carcinoma, preoperative embolization of feeding vessels may facilitate safer surgical resection by minimizing intraoperative blood loss and improving operative conditions.

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Keywords: Pediatric, choroid plexus carcinoma, embolization

Papillary tumor of the pineal region in a child: Case report and literature review of a rare tumor.

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Objective: To report a rare case of papillary tumor of the pineal region in a pediatric patient, highlighting the challenges in diagnosis, management and long-term follow-up.
Case Report: A 10-year-old boy with persistent headache and vomiting for 2 months. MRI showed an expansive lobulated lesion in the pineal region, with discrete hyperintense signal on T1 and T2, contrast uptake and measuring 12 mm in the largest axis. The patient underwent resection of the lesion via a transventricular transcortical frontal approach, and the immunohistochemical results confirmed a papillary pineal tumor. No additional treatment was initially performed. After 3 years, the lesion recurred with hydrocephalus. A new resection and ventricular bypass were performed using the same previous approach. After discharge, outpatient follow-up without additional treatment was recommended.
Results: The papillary pineal tumor recurred late, requiring a new surgical intervention.
Conclusion: Papillary pineal tumors are rare neoplasms with a high recurrence rate. The lack of clinical trials and the scarcity of cases prevent the definition of a standardized therapeutic protocol. The extent of surgical resection remains the main prognostic factor, directly influencing survival and the risk of recurrence.

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Keywords: Papillary, tumor, pineal, pediatric

Meningioma of the Central Nervous System in the pediatric population: 7 years of experience in a single center and review of the literature

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Introduction:
Meningiomas account for 2% of childhood brain tumors. They are distinguished from adult meningiomas by the predominance of high-grade tumors, with high rates of proliferation and invasion of brain tissue. Many cases of pediatric meningiomas (PM) are associated with genetic syndromes, especially Neurofibromatosis type II (NFII).
Objective: To analyze the epidemiological, clinical, radiological and surgical profiles, in addition to histological types and therapeutic strategy employed in patients with PM treated in the last seven years; and to compare these results with the literature.
Materials and Methods: This was a retrospective study carried out at a pediatric oncology referral service. Data from January 2014 to December 2024 was collected from electronic medical records.
Results: Among the 775 patients who underwent surgery at the service, 10 were meningiomas. Seven were boys; the average age was 16.1 years (4-18 years). Epileptic seizures and headache were the most common symptoms. Eight patients had intracranial tumors and 2, with NFII, had spine tumors. Seven of them underwent two consecutive surgeries to completely excise the intracranial lesion. The average diameter in the longest axis was 6.5 cm (1-11 cm). Only one patient underwent preoperative embolization. Two anatomopathological findings corresponded to WHO grade II. There were no recurrences during a mean follow-up of 2.8 years.
Conclusion: MPs are rare, aggressive and highly vascularized lesions that reach large dimensions at diagnosis. Preoperative embolization should be considered to control tumor bleeding during surgery especially in chindren.

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Keywords: pediatric, meningioma, neurofibromatosis

A Pineal Gland Cyst with an Unusual Presentation: A Case Report

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Case Report: A.C.N.L., a previously healthy 16-year-old female patient, presented with a
brief episode of vertigo and diplopia after jumping on a trampoline. Neurological examination was unremarkable, fundoscopy revealed no signs of intracranial hypertension, and neuropsychological testing was within normal limits, with no evidence of cognitive
dysfunction. Neuroimaging revealed a cystic expansive lesion located posterior to the third
ventricle, with evidence of adjacent structure compression, associated with ventriculomegaly and ependymal transudation. The diagnosis was oligosymptomatic hydrocephalus secondary to an expansive lesion of the pineal gland. A neuroendoscopic approach was performed, during which biomarkers (Alpha-Fetoprotein and Beta-HCG) were collected from cerebrospinal fluid (CSF) and peripheral blood, both of which returned normal values. Additionally, a third ventriculostomy and biopsy of the expansive lesion were performed endoscopically. The lesion exhibited darkened colloidal content upon capsule rupture, and both the capsule and its contents were sent for pathological analysis. The immediate postoperative course was uneventful. Neurological monitoring and follow-up imaging demonstrated a reduction in ventricular dilation and normalization of cerebrospinal fluid flow. The patient remains under outpatient follow-up, with no residual neurological deficits, and has a favorable prognosis following neurosurgical intervention. Results and Conclusion: This case involved a pineal gland cyst that mimicked a neoplasm. Histopathological analysis of the collected tissue revealed the presence of histiocytes and hemosiderin deposits within a capsule lined by ependyma, with no evidence of neoplastic features. Follow-up magnetic resonance imaging (MRI) performed six months postoperatively confirmed the diagnosis of a pineal cyst, with no evidence of progression. Following cyst drainage, cerebrospinal fluid (CSF) flow through the cerebral aqueduct was restored. Pineal cysts are common incidental findings on imaging and typically do not require surgical intervention. However, the presence of hydrocephalus necessitates treatment, often via a neuroendoscopic approach. In this case, the cystic lesion

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exhibited imaging characteristics suggestive of a pineal region tumor. Nevertheless, cyst formation was likely secondary to an old hemorrhage, which triggered an inflammatory response mimicking a neoplastic process. This hypothesis was further supported by the absence of contrast enhancement on the follow-up MRI six months after surgery.

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Keywords: Pineal gland cyst , Hydrocephalus , Neuroendoscopy , Differential diagnosis , Pineal region tumor.

The influence of BRAF-KIAA and BRAF-V600E genetic alterations on the clinical-radiological outcome of pediatric patients with low-grade gliomas.

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Abstract
Pediatric low-grade gliomas (LGGs; WHO grade 1–2) encompass diverse subtypes—27 pilocytic astrocytomas, 2 subependymal giant cell astrocytomas, 3 gangliogliomas, 1 pilomyxoid astrocytoma, 1 diffuse astrocytoma, and 1 choroid plexus papilloma. Molecular activation of the BRAF proto-oncogene, via KIAA1549-BRAF fusions or the BRAF-V600E point mutation, underlies distinct tumor behaviors.
In this retrospective single-center study, 35 pediatric LGGs underwent resection between 2018 and 2023. Tumor DNA from 24 cases was analyzed by real-time PCR for BRAF alterations. Clinical records and serial MRI (mean follow-up 79.5 months) were reviewed by molecular subgroup.
Of 24 sequenced tumors (mean age 6.6 years), 12 (50 %) showed no BRAF alteration, 9 (37.5 %) harbored KIAA1549-BRAF fusions, and 3 (12.5 %) carried the BRAF-V600E mutation. All BRAF-V600E and fusion-positive cases maintained stable residual lesions without recurrence or progression. No perioperative morbidity correlated with molecular status.
Pediatric LGGs exhibit distinct molecular profiles but, in our cohort, neither BRAF-V600E nor KIAA1549-BRAF fusions were associated with early recurrence. Nonetheless, V600E cases warrant prolonged surveillance given reports of late progression. Preoperative BRAF screening should guide surgical extent, imaging schedules, and consideration of targeted therapies, supporting precision neuro-oncology in children.

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Keywords: Pediatric gliomas; Molecular profile; Prognosis.

TRAUMATIC BRAIN INJURY (TBI)

Ideal timing for tracheostomy in pediatric patients with severe traumatic brain injury: systematic review and meta-analysis

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OBJECTIVE: We conducted a systematic review with meta-analysis to evaluate the effects of early tracheostomy (ET) versus late tracheostomy in pediatric patients with severe traumatic brain injury (sTBI).
METHODS: A systematic search was performed in the PubMed, Embase, and Web of Science databases. ET was defined as tracheostomy performed within 10 days, while sTBI was characterized by Abbreviated Injury Scale scores greater than 2 or Glasgow Coma Scale scores lower than 8. Outcomes analyzed included hospital length of stay, intensive care unit (ICU) stay, incidence of pneumonia, and mortality. Statistical analysis was conducted using RevMan 5.4.1.
RESULTS: The initial search yielded 217 results, of which 25 were removed due to duplication and 180 were excluded based on title/abstract screening. Of the remaining 12 studies, 8 did not adequately address the topic, resulting in the inclusion of 3 retrospective cohort studies in the final review. A total of 2,518 pediatric patients with sTBI were analyzed. ET was performed in 1,025 (40.7%) participants, with a median age of 16.03 years (IQR: 4–18). The incidence of pneumonia (RR: 0.73; 95% CI: 0.66 to 0.81; $p < 0.00001$) and hospital length of stay (MD: -14.3; 95% CI: -24.29 to -4.31; $p = 0.005$) were significantly lower in the ET group. Other outcomes were not consistently reported across the three studies, precluding adequate analysis.

CONCLUSION: Tracheostomy performed within 10 days is associated with a reduced incidence of pneumonia and shorter hospital stay in pediatric patients with sTBI. Further studies are needed to assess other relevant outcomes.

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Keywords: Length of stay, Pediatric neurosurgery, Pneumonia, Tracheostomy

Epidural Hematoma Presenting as Jaundice in a Newborn

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Objective: To report a case of epidural hematoma in a patient with progressive jaundice.
Case Report: A male patient was admitted at 21 days of life, brought in by his mother due to progressive jaundice. There was a previous report of a subgaleal hematoma with suspected skull fracture due to birth trauma on initial ultrasound, along with prolonged hospitalization for phototherapy. On physical examination, the patient was active and reactive, with adequate tone, moving all four limbs, pupils equal and reactive to light, anterior fontanelle open and normotensive, presenting with jaundice at Kramer zone 3/4 and a right-sided subgaleal hematoma. Cranial MRI confirmed the hypothesis of a chronic epidural hematoma secondary to birth trauma, measuring 33 mm in thickness, with an estimated 7 mm contralateral midline shift. The patient underwent craniotomy for epidural hematoma drainage at 26 days of life, with no intraoperative complications. Postoperatively, the patient showed clinical improvement along with progressive resolution of jaundice. Follow-up CT scan revealed a small residual hematoma and significant re-expansion of the brain parenchyma.
Results: To evaluate different presentations of intracranial hematomas in newborns.
Conclusion: Jaundice is a clinical sign associated with hyperbilirubinemia; the presence of intracranial hematomas may be associated with prolonged jaundice.

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Keywords: Epidural, Hematoma, Newborn, Jaundice, Pediatric

MISCELLANEOUS

Fetal surgery for myelomeningocele: is it reliable in developing country settings?

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Objective: To evaluate the epidemiological and surgical aspects of fetal surgery for myelomeningocele (FSM) in an obstetric referral center in Brazil. Material and method: A series of FSMs was reviewed in an obstetric referral center in a developing country, from May 2019 to March 2024. Maternal and neonatal data were collected. Results: The study included 35 pregnant women, aged between 19 and 46 years (mean: 27.5 years) and gestational age at FSM between 22 weeks and 5 days and 27 weeks. Among them, 51.4% were nulliparous and 34.2% had undergone cesarean section. Cervical measurement ranged from 28 to 45 cm. Fetal neurological lesions were from T12 to S5, predominantly high (34.2%) and low (25.7%) lumbar. Clubfoot occurred in 37.1% of cases. FSM was performed by open hysterotomy, with a mean time of 179 minutes. Complications included amniotic abruption, premature rupture of membranes, and preterm birth. The interval between FSM and delivery was 63.2 days. Gestational age at birth ranged from 27 weeks and 2 days to 37 weeks and 5 days, with 77.1% of premature infants. The mean weight was 2.115 kg, and 28.5% required brainstem hernia reversal. Ventriculoperitoneal shunt was necessary in 17.1% of cases, and there was one perinatal death. Conclusion: Despite maternal risks, FSM showed good neonatal outcomes, reducing VPS. However, structural limitations and lack of specialized teams in developing countries reinforce the need for further research and investment.

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Keywords: Myelomeningocele, Prenatal surgery, Fetal surgery.

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Grisel's Syndrome Secondary to Recurrent Upper Respiratory Tract Infections: A Case Report

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Objective:
To recognize a syndrome with high morbidity potential in children
Case Description:
An 8-year-old male patient presented with recurrent complaints of cervical pain, progressively worsening limitation in the range of motion of the head, and cervical muscle stiffness over a period of 5 months. During anamnesis, no traumatic history or other relevant factors were identified, except for frequent upper respiratory tract infections (URTIs).
Clinical evaluation revealed a patient in good general condition, with hypertrophied tonsils and significant cervical rigidity associated with rotation of the head to the right, along with restricted cervical movements. Kernig's and Lasegue's signs were negative, and cranial nerves, muscle strength, sensory examination, and cerebellar tests were all within normal limits.
Radiologic examinations were performed for differential diagnosis. Cervical X-ray revealed a rotational deformity of the C1-C2 vertebrae and an increased atlantodental interval. Cranial CT showed no abnormalities; however, cervical CT and MRI revealed atlantoaxial subluxation with rightward rotation of C1, widening of the atlantodental interval, and degenerative changes in the C1-C2 articulations.
Based on clinical and radiological findings, a diagnosis of Grisel's syndrome was proposed.
Outcome:
The chosen treatment was conservative, utilizing a cervical collar, which resulted in the patient's complete recovery.
Conclusion:
Due to its rarity, Grisel's syndrome must be promptly recognized to ensure appropriate treatment and minimize the consequences of this condition—particularly in children and adolescents, who are at high risk for associated comorbidities. Early diagnosis and conservative management typically lead to favorable outcomes.

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Keywords: Grisel's syndrome, Subluxation, Atlantoaxial, Pediatric patients, Cervical contracture

Occipital Dermoid Cyst Mimicking Meningoencephalocele: A Case Report

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We report the case of an 11-month-old female infant presenting with a progressively enlarging midline occipital mass over a six-month period. Magnetic resonance imaging (MRI) of the brain revealed a sessile extracranial lesion located in the midline occipital region, with imaging findings suggestive of an atretic meningoencephalocele. The patient exhibited no neurological deficits and had appropriate neuropsychomotor development. Surgical resection of the lesion was performed. Intraoperatively, a sessile extracranial mass with a small bony defect connecting it to the intracranial space was identified. Given that the defect terminated within the diploic space, craniotomy was deemed unnecessary. Histopathological analysis confirmed the diagnosis of a dermoid cyst (DC).

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Keywords: Dermoid cyst; Occipital mass; Infant; Meningoencephalocele; Neuroimaging; Case report

Sinus Pericranii Undergoing Embolization: A Case Report and Literature Review

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Objective:
To report a case of sinus pericranii treated with embolization.
Case Report:
A 9-year-old female patient with Noonan syndrome, characterized by typical facies and neuropsychomotor developmental delay, underwent cranial magnetic resonance imaging (MRI) due to a palpable, hardened mass in the right parietal region. On physical examination, she presented with an asymmetric skull, turricephaly, an apparently open lambdoid fontanelle, ocular hypertelorism, downward-slanting palpebral fissures, and a palpable hardened lesion in the right parietal region. Cranial MRI revealed a venous vascular anomaly in the parieto-occipital soft tissues that connected to the superior sagittal sinus via an osseous defect, while computed tomography angiography (CTA) demonstrated a midline parieto-occipital osseous defect with a dural venous vascular structure connecting to the superior sagittal sinus, in addition to a folded vascular lesion in the adjacent extracranial soft tissues measuring approximately 24 x 8 x 18 mm. The patient underwent an endovascular procedure with successful embolization of the arteriovenous malformation.
Results:
A review of the literature was conducted, discussing the various intervention possibilities for sinus pericranii.
Conclusion:
Sinus pericranii is a rare vascular malformation with a challenging diagnosis that can lead to severe complications and aesthetic deformities. Early surgical or endovascular treatment is necessary.

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Keywords: Sinus pericranii, embolization, Noonan, Pediatric

Treatment of recurrent aneurysmal bone cyst with RANK-inhibiting monoclonal antibody

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Introduction: Aneurysmal bone cyst (ABC) is a benign but locally aggressive tumor that occurs predominantly in children and adolescents. The most common locations are in the long bones and secondly in the spine. The clinical symptons usually begins with pain and reduced mobility. The imaging exam of choice is magnetic resonance imaging showing cavities filled with blood with a liquid-liquid level. Initial treatment is en bloc surgical resection. In addition, sclerotherapy and embolization can be performed. To date, there have been no reports of an association between medulloblastoma and ABC, but as already described, radiotherapy can lead to an increased risk of bone lesions.
Case presentation: A 6-year-old female patient followed up at the neurosurgery outpatient clinic due to a previous history of SHH-type medulloblastoma. She was diagnosed at the age of 1 and treated with surgical resection and autologous hematopoietic stem cell transplantation. She has been out of treatment for 6 years (2018), with regular follow-up. She had a sequela of VI cranial nerve palsy on the left eye and mild ataxia. She was admitted reporting low back pain 6 months ago after a fall from her own height, with progressive worsening 1 month ago. On physical examination, the patient's strength was preserved, with no sensory or sphincter alterations. In order to investigate the low back pain, CT and MRI scans were carried out showing lesion in L1 with expansive aspect, hypervascularized, with liquid-liquid levels with hyperintensity in T2, with compression of the dural sac and medullary cone. Patient underwent L1 corpectomy via lateral access and T11-L3 arthrodesis via posterior approach. Four months after the surgery, the patient returned to the ER with a history of low back pain associated with urinary and fecal incontinence. A new approach to the lesion was indicated just to decompress the spinal canal without removing the surgical instrumentation. The patient continued to complain during outpatient follow-up. The decision was made to introduce Denosumab. In a 10-month follow-up after the introduction of the medication, the patient had no new recurrence of the lesion or worsening of symptoms. Serial imaging exams were carried out with no evidence of lesion growth
Discussion and conclusion: Denosumab is a RANK L inhibitor with a central role in bone remodeling by stimulating osteoclastogenesis and bone reabsorption. It is a well-established treatment for giant cell bone tumors, so due to the histopathological similarities between these diseases, some authors have selected patients with aneurysmal bone cysts for use. In this case, we can identify aneurysmal bone cyst as a differential diagnosis of medulloblastoma dissemination, as well as the benefit of using the RANK L inhibitor, which is an excellent approach for these patients with recurrent lesions.

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Keywords: Aneurysmal bone cyst, denosumab

Pediatric Patients with Holoprosencephaly: A 10-Year Experience at a Reference Center

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Purpose: To outline the epidemiological profile, therapeutic management, and outcomes of pediatric patients with holoprosencephaly who received treatment at the Hospital of Clinics of the Ribeirão Preto Medical School, University of São Paulo (HC-FMRP USP), over a 10-year period. Methods: A retrospective analysis was conducted on medical records of pediatric patients diagnosed with holoprosencephaly who received treatment at HC-FMRP USP between January 1, 2014, and January 1, 2024. Demographic, clinical, and outcome data were collected and systematically described. Results: Twenty-five patients with holoprosencephaly were treated. One-third were diagnosed via prenatal ultrasound. The most common subtype was alobar (40%). Over half (52%) were born at the hospital, with 52% being female. The most frequent surgery was external ventricular drain (17 procedures in 5 patients), followed by ventriculoperitoneal shunt (13 procedures in 11 patients). Six patients needed shunt revisions, and five developed ventriculitis. Seven patients (28%) died, with a median age of 1.4 months. Pediatric palliative care consultations were provided to 8 patients (32%). Patients with alobar subtypes had worse outcomes overall. Conclusions: In most cases within this series, the diagnosis was established postnatally, with alobar being identified as the predominant subtype. Less than fifty percent of the patients underwent ventriculoperitoneal shunt placement. The observed mortality rate stood at 28%, with 4 out of the 7 deceased patients having an advance care plan prioritizing symptom relief.

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Keywords: Holoprosencephaly, Pediatric Palliative Care

Hydrocephalus

Endoscopic Choroid Plexus Coagulation as an Adjuvant Therapy for Refractory Hydrocephalus in Pediatric Patients: A Systematic Review

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Objective: To assess the efficacy of endoscopic choroid plexus coagulation (CPC) as an adjunctive treatment for refractory hydrocephalus in pediatric patients, both as a standalone procedure and in combination with endoscopic third ventriculostomy (ETV). Methods: A systematic review was conducted following PRISMA guidelines, using the PubMed, ScienceDirect, and BMC Neurology databases. Clinical trials, observational studies, and systematic reviews from the past 23 years were included. Studies enrolled patients under 18 years old with refractory hydrocephalus treated via CPC or CPC combined with ETV. Studies that were incomplete, outside the target age group, or lacking relevant data were excluded. Results: Out of 20 articles reviewed, 13 met the inclusion criteria, involving a total of 1,058 patients. CPC was found to significantly reduce cerebrospinal fluid (CSF) production and demonstrated effectiveness in managing neonatal post-hemorrhagic hydrocephalus, severe hydrocephalus, and hydranencephaly. Higher success rates were observed in the CPC+ETV group (60–80%) compared to CPC alone (40–50%), particularly in decreasing the need for ventricular shunting and reducing complication rates. The use of flexible endoscopes improved access to anatomically challenging areas within the lateral ventricles, enhancing surgical precision and outcomes. Conclusion: Endoscopic CPC is a promising adjunctive option in the management of refractory hydrocephalus in pediatric populations. The combination with ETV enhances success rates and decreases complications, although outcomes remain dependent on patient-specific factors such as etiology and age. Advances in neuroendoscopic techniques, notably the implementation of flexible endoscopes, have minimized surgical trauma and facilitated improved recovery and prognosis.

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Keywords: Choroid Plexus Coagulation; Refractory Hydrocephalus; Neuroendoscopy; Flexible Endoscope; Ventricular Shunting

EPILEPSY

The usefulness of electrocorticography (ECoG) in pediatric temporal lobe epilepsy surgery

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Introduction: Resective surgery is the main therapeutic alternative for drug-resistant temporal lobe epilepsy (TLE) in the pediatric population. Defining a clear anatomo-electro-clinical hypothesis to delineate the epileptogenic zone (EZ) is a crucial step for planning the surgical treatment.
Objective: The aim of this study is to evaluate the usefulness of electrocorticography (ECoG) in providing a more accurate surgical strategy and thus yielding seizure-freedom following resective surgery in children with TLE.
Materials and Methods: The authors conducted a retrospective review of pediatric patients with drug-resistant TLE due to various etiologies, excluding those with short follow-up (range 8.5 to 11.5 years). Patients were divided into two groups based on the use of invasive monitoring (ECoG). The efficacy of surgical treatment was assessed using Engel's classification. Seizure-freedom rate for each etiology was compared between groups using Fisher's exact test with a 95% confidence interval.
Results: A total of 81 patients were included in the study (mean age 11.8 years, range 1 to 18 years), of whom 63 (77.8%) achieved Engel I status after 10 years . The main etiology was hippocampal sclerosis (34/81, 41.9%), followed by tumors (25/81, 30.8%) and cortical dysplasia (22/81, 27.1%). Invasive monitoring was performed in 29/81 (35.8%) patients. Overall, there were no significant differences in the proportion of Engel I (p=0.10) among those who performed ECoG (22/29, 75.9%) and did not perform ECoG (n = 41/53, 78.8%). Among tumor-associated cases, Engel I was achieved in (8/8, 100%) of patients with ECoG compared to (13/17, 76.5%) without ECoG (p = 0.07). This difference, although not statistically significant, may reflect improved identification of adjacent type IIIb FCD. No significant differences were observed in cortical dysplasia (p = 1.00) or hippocampal sclerosis (p = 0.07).
Conclusion: The use of intraoperative electrocorticography did not yield higher rates of seizure freedom for pediatric TLE, and both groups showed satisfactory seizure control post-operatively. However, patients with tumor-related TLE that underwent surgery with ECoG showed a tendency towards higher seizure freedom rates as compared to their counterparts, so that ECoG could be beneficial in this scenario.

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Keywords: intraoperative electrocorticography, mesial temporal lobe sclerosis, temporal lobe epilepsy, tumor, cortical dysplasia.

DYSRAPHISM

Intrauterine myelomeningocele repair – a case series from the University of Campinas

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INTRODUCTION: The myelomeningocele persists as one of the main types of spinal dysraphisms and an important focus of the pediatric neurosurgeon. The intrauterine repair has shown promising results in the medical literature. This paper aims to describe the initial experience of this technique in a reference terciary hospital in Campinas, São Paulo, Brazil.
METHODS: This is a prospective and longitudinal case series of 11 patients who underwent myelomeningocele intrauterine repair through histerotomy (open technique) between 2021 and 2025 in a single-center (University of Campinas), performed by the same team of obstetricians and neurosurgeons.
RESULTS: 11 intrauterine myelomeningocele repairs have been performed in the time period, 7 (63,6%) of them being male and 4 (36,4%) being female. The median of the gestational age of the surgery was 26 weeks (ranging from 23 to 27 weeks). Until march 2025, 3 (27,2%) of the babies haven't been born yet, 3 (27,2%) were early preterm (<34 weeks), 3 (27,2%) were late preterm (34-36 weeks) and 2

(18,2%) were term. Analysing the hydrocephalus rate, none of the patients needed ventricular derivation as of yet.
CONCLUSION: This case series exhibited promising results, regarding both the obstetric complications (prematurity) and neurological outcomes (hydrocephalus and motor dysfunctions), reinforcing the medical literature propensity to foment the intrauterine timing for the myelomeningocele repair.

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Keywords: myelomeningocele, spinal dysraphism, intrauterine

Resection and arthrodesis of lumbar hemivertebra via posterior approach with unilateral hook and counter-hook fixation in a pediatric patient

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OBJECTIVE: To report the case of a pediatric patient who underwent resection of a lumbar hemivertebra (HV).
CASE REPORT: Female, 2 years old, presenting with lumbar deviation. No developmental deficits. History of esophageal atresia. Magnetic resonance imaging (MRI) revealed mild syringomyelia at the T7-T8 level and a fully segmented HV at the
L2-L3 levels, causing a leftward spinal deviation of 19° (Figures 1, 2 and 3). Resection and arthrodesis were performed in
posterior approach by compression, using hook and counter-hook system for fixation, under microscopy and neuromonitoring.
Uneventful postoperative course, with deformity reduced from 19° to 4°. RESULTS: Hemivertebra is a congenital malformation with an incidence of 0.03%, occurring either in isolation or, more
commonly, in association with other deformities. Prognosis depends on its location and growth pattern. Segmented HVs, multiple anomalies, or contralateral bony bars carry a higher risk of severe curvature and neurological involvement. Deviation
typically progresses at a rate of 1°–3.5° per year, worsening during growth spurts. It is usually identified prenatally, though up
to one-third of cases are only detected after birth. Surgical intervention is the most effective approach, which can be performed via posterior approach (preferred due to lower invasiveness, shorter duration, and reduced blood loss) or anteroposterior approach (used in complex cases, especially in the cervical region). The use of hook and counter-hook is indicated for milder curvatures and offers less invasiveness and lower cost. There is no consensus on the ideal age for surgery, but intervention before the age of 6 has shown better outcomes.
CONCLUSION: Hemivertebrae are rare deformities with high risk of progression and significant implications. Early diagnosis
is crucial for optimal outcomes. When surgery is indicated, early management demonstrates greater efficacy.

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Keywords: Hemivertebra, Spinal deviation, Surgical resection, Posterior arthrodesis, Early intervention

Analysis of Motor Prognosis in Prenatal Versus Postnatal Surgical Correction in Children and Adolescents with Myelomeningocele Followed in the Municipality of Blumenau, Brazil

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Objective: To analyze motor prognosis following prenatal versus postnatal surgical correction in children and adolescents with myelomeningocele in Blumenau, Brazil.
Materials and Methods: A prospective cohort study was conducted with 46 patients diagnosed with myelomeningocele. Participants were divided into two groups: those who underwent intrauterine surgery and those who underwent postnatal surgery. Data were collected through medical record review and a demographic/gestational questionnaire. Motor function was assessed using the Denver Developmental Screening Test and the. American Spinal Injury Association (ASIA) scale.
Results: The sample included 16 patients in the intrauterine group and 30 in the postnatal group. The most common lesion level was lumbar (41.3%). Neurogenic bladder was present in all patients(100%), and hydrocephalus was observed in 30 cases. Dependence on a ventriculoperitoneal shunt (VPS) was noted in 18.8% of the intrauterine group and

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50% of the postnatal group. Among the 24 patients assessed using the ASIA scale, most individuals in the postnatal group were classified as AIS D, indicating incomplete motor impairment. The intrauterine group showed broader distribution of classifications, with higher prevalence of AIS C and D.
Conclusion: Motor function impairment was observed in both groups; however, patients who underwent intrauterine surgery exhibited a lower rate of functional abnormalities.These findings suggest the need for further investigation.

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Keywords: Myelomeningocele, Motor Prognosis, Intrauterine Surgery

IMPACT OF INTRAUTERINE SURGERY ON THE REDUCTION OF HYDROCEPHALUS IN PATIENTS WITH MYELOMENINGOCELE: A COMPARATIVE ANALYSIS

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Objective:
To analyze the incidence of hydrocephalus secondary to myelomeningocele, comparing intrauterine versus postnatal surgical repair approaches.
Methods:
A retrospective cohort study included patients who underwent either intrauterine or postnatal surgery for myelomeningocele between 2019 and 2024. Inclusion criteria consisted of a head circumference at birth below a Z-score of +3 and at least six months of postnatal outpatient follow-up.
Results:
Within the cohort, 26 patients underwent intrauterine surgery, and 81 had postnatal repair. According to the inclusion criteria, 18 intrauterine and 45 postnatal cases were analyzed. Among intrauterine surgery patients, 33.3% were born preterm, with a median gestational age (GA) of 35 weeks (range: 30 weeks and 1 day to 37 weeks and 6 days). There was one case (5.6%) of wound dehiscence and no other surgical complications. Only one patient (5.6%) developed hydrocephalus requiring ventriculoperitoneal shunting (VPS).
Among postnatal surgery patients, 15.6% were preterm, with a median GA of 38 weeks and 2 days (range: 31 weeks and 5 days to 40 weeks and 2 days). There were 7 cases (15.6%) of wound dehiscence and 10 cases (22.2%) of cerebrospinal fluid fistulas. Hydrocephalus occurred in 31 patients (68.9%), with 90.3% developing the condition during their initial hospitalization. All required VPS, with 12 cases (38.7%) experiencing infections and 11 cases (35.5%) subsequently undergoing endoscopic third ventriculostomy.
Conclusion:
Intrauterine surgery, despite a higher rate of preterm births, proved superior to postnatal repair in reducing the incidence of hydrocephalus and associated complications.

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Keywords: hydrocephalus, myelomeningocele, fetal surgery.

Myelomeningocele repair using the "H" technique: a case series from a public hospital in Minas Gerais

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Objective: The objective is to describe a technique for closing myelomeningocele in newborns, using joint flaps with resulting "H" healing; Material and Method / Case series: Description of the myelomeningocele closure technique in four cases of children born between 2017 and 2019 in a public hospital in the state of Minas Gerais, with outpatient follow-up until 2024. According to the technique, variations in surgical time, advanced outcomes and risk of complications, such as infections and cerebrospinal fluid fistula, are proven; Results: Of the four serial cases, all newborns were full-term gestational age and were approached after the first hours of birth, and the average surgical time to correct the congenital defect was approximately two hours. Of the cases listed, three were lumbosacral myelomeningocele and one was thoracic myelomeningocele, and in all cases, a malformation was previously known in the prenatal period. After identification and dissection of the placode, rotation flaps and skin sutures were made in layers, resulting in "H" healing. No occurrence of infection related to the surgical site or fluid fistula was observed in the subsequent period; Conclusion: Given the clinical cases presented, it is possible to conclude that the use of the rotation flap with a resulting "H" scar was effective in preventing the occurrence of infections or cerebrospinal fluid fistulas possibly associated with the procedure. It is also possible to conclude that, with adequate training, it is possible to replicate the technique with reasonable surgical time and intraoperative safety.

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Keywords: Myelomeningocele, Spina bifida, Neural tube defects

Filum terminale lipoma in childhood - analysis of patients treated in a reference service: should refractory constipation be considered a warning sympton?

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Introduction: Filum terminale lipoma (FTL) is a type of occult spinal dysraphism, characterized by infiltration of adipose tissue at some level of the filum, determining tethered cord syndrome (TCS). Clinical suspicion occurs in the first year of life, in the presence of cutaneous stigmata and perineal malformations. In older children, sphincter dysfunctions are more common and intestinal constipation is found in 15 to 45% of cases. Objective: To analyze the epidemiological, clinical and radiological characteristics of patients with FTL who underwent surgery at a reference hospital in the city of São Paulo from January 2022 to December 2024. Materials and methods: This is an ecological study; data were collected through electronic medical records. Results: Among the 5 boys and 5 girls, the average age was 9.12 years. 3 patients had cutaneous stigmata. The main symptoms were constipation (75%), difficulty walking (62.5%) and low back pain (50%). The average diagnostic interval was 3.3 years. The conus medullaris was at the level of S1 in one patient. All patients were submitted to microsurgical section of the FTL under neurophysiological monitoring. 66% of patients showed improvement in constipation; and 100% exhibited improvement in gait and low back pain. There were no complications or reoperations. Conclusion: The surprisingly high proportion of constipation symptoms prompted the establishment of a multidisciplinary protocol for the early diagnosis and treatment of FTL in patients diagnosed with refractory constipation or retentive fecal incontinence.

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Keywords: Filum terminale lipoma; Tethered cord syndrome; Constipation

CRANIOSYNOSTOSIS

Total Cranial Remodeling Using the Modified Melbourne Technique for the Treatment of Late Scaphocephaly: Case Report

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OBJECTIVES: To report a case of late-diagnosed scaphocephaly surgically treated with total cranial remodeling using the modified Melbourne technique.
CASE REPORT: A 5-year-old female patient, with no previous medical conditions, was diagnosed late with scaphocephaly. She underwent the modified Melbourne technique, involving a zigzag biparietal incision and subperiosteal dissection of flaps from the supraorbital region to the occipital protuberance. To reduce the anteroposterior cranial length, a strip craniotomy was performed at the point of maximum biparietal width. The resulting bone strip was divided for remodeling of the posterior calvaria and rotated 90 degrees, then fixed to the occipital structure to increase occipital height. Frontal and parietal segments were remodeled to expand the intertemporal and interparietal distances. The forehead was reshaped to correct frontal bossing. Bone defects were filled with particulate autologous bone graft.
RESULTS: The modified Melbourne technique effectively reduced the anteroposterior cranial length and increased occipital height. No intraoperative or postoperative complications occurred. Clinical progression was satisfactory, with appropriate wound healing and no signs of infection or bone instability.
CONCLUSION: The modified Melbourne technique proved effective in correcting severe cranial deformities, such as in cases of late scaphocephaly, restoring cranial proportions.

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Keywords: Craniosynostosis, Scaphocephaly, Melbourne, Total Cranial Remodeling.

Predictors od Epilepsy in Syndromic Craniosynostosis

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Epilepsy in patients with craniofacial syndromes seems to be uncommon in craniofacial practice, and there are few reports in the literature devoted to identifying predictors of epilepsy in these patients. The objective of this study is to compare syndromic patients with and without epilepsy and identify predictors of epilepsy in those patients diagnosed with syndromic craniosynostosis.

An observational retrospective multicentric study was performed on 426 consecutive patients with Apert, Crouzon or Pfeiffer syndromes and underwent any type of surgery between 2007 and 2022. Demographic data, diagnosis, surgery-related data, were verified using medical records, radiologic examination, and interviews. The International League Against Epilepsy, Classification of Epilepsies questionnaire was handed out to all syndromic patients or families. Bivariate and multiple linear regression analyses were performed to determine which independent variables were significantly predictive of epilepsy. Groups were divided in patients with epilepsy (n=24) and patients without epilepsy (n=402).

The model demonstrated that the independent variable "Previous surgery elsewhere" showed a p-value <0.001 with an OR = 853. Patients with previous surgery elsewhere are 853 times more likely to have epilepsy than those who have not had prior surgery elsewhere and were treated at ours Centers since the first primary consultation. Patients were divided in two groups: with and without epilepsy. The variables that were prevalently observed in the epilepsy group were patients with Chiari malformation type I, prior surgery performed elsewhere, intracranial complication, encephalomalacia, hidrocephalus, family history of epilepsy, Le Fort III, Meningitis, and placement of a VP Shunt.

Such findings are suggestive that epilepsy in syndromic craniosynostosis patients seems to be acquired and not related to underlying congenital cerebral malformations. What is in accordance with the late presentation of epilepsy in this population. The findings of the logistic regression analysis showed that when prior surgery was performed elsewhere, it represented a predictive risk for epilepsy.

We can assume that the underlying physiopathology of the acquired epilepsy in syndromic craniosynostosis patients can be like post-traumatic epilepsy: epilepsy related to brain contusion and subsequent inflammation, leading to a late epilepsy presentation.

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Keywords: Epilepsy, Craniosynostosis

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