



## 28th Congress of the European Society for Pediatric Neurosurgery (ESPN) Rome-Italy, 7-10 May 2023

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### Oral Presentations

#### Abstract

#### Session 1: Spine I

##### OP001

#### Neurosurgical Outcomes in Fetal Spina Bifida Repair: a Comparison of Fetoscopic, Open Fetal, and Postnatal Approaches at a Single Institution

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**Introduction:** Fetal spina bifida (SB) repair has evolved significantly. Technical innovation strives to reduce maternal and fetal risk. We aimed to characterize our experience and compare neurosurgical outcomes at a single institution for a 3-port/3-layer fetoscopic, open fetal, and postnatal approaches to spina bifida repair.

**Methods:** A retrospective study was conducted examining all cases of prenatal and postnatal SB repair at our institution from 2017-2022. Open fetal surgical repair was initially offered in 2017. In November 2020, fetoscopic closure was offered. In each case, surgical repair with a three-layer back closure was attempted regardless of timing of surgical intervention.

**Results:** 78 patients were included; of which 25 underwent fetoscopic, 27 open fetal, and 26 postnatal repair. Technically more challenging, fetoscopic intervention was longer in duration. Primary dural closure was feasible in open and fetoscopic closure; primary skin closure was difficult in fetoscopic closure, achieved in 64% of cases. 74% of fetoscopic patients underwent vaginal delivery; all open fetal patients underwent cesarean delivery. 35% of fetoscopic patients were born at term. An older mean gestational age was observed transitioning from open to fetoscopic closures. CSF diversion via ETV or VPS by 12-months was required in 48% (10/21) postnatal, 33% (9/27) open fetal, and 29% (2/7) fetoscopic repairs with 12-month follow-up. 84.2% (32/38) of those with hindbrain herniation, closed in utero, experienced resolution on postnatal imaging. Open and fetoscopic closures experienced improved lower extremity strength, as assessed by manual muscle test per PT, than anticipated based on anatomical level.

**Conclusion:** We demonstrate technical feasibility of three-layered closures in open fetal and fetoscopic SB repairs. Neurosurgical outcomes including functional level, need for CSF diversion, and brainstem function are consistent with MOMs data, but require continued assessment to determine the lifetime impact of technical innovations in fetal SB repair.

##### OP002

#### Global, regional and national epidemiological trends in neural tube defects between 1990-2019

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**Introduction:** Neural tube defects (NTD), such as spina bifida, are surgically treatable and primarily preventable non-communicable diseases. How the incidence, mortality and disability-adjusted life year (DALYs) rates of NTD have modulated over time is not well defined. **Methods:** A retrospective review of data from the Global Burden of Disease Study 2019 Database was performed. Global, regional and national outcomes for NTD were collected for incidence, mortality and DALY rates, and their age-standardized metrics analyzed.

**Results:** Globally, the latest age-standardized rates of incidence, mortality and DALYs of NTD were 2.1 per 100,000 population, 1.3 per 100,000 population and 117 per 100,000 population respectively. All rates demonstrated decreases in the last two decades to now. Regionally, Sub-Saharan Africa and North America demonstrated the highest and lowest age-standardized rates of incidence (4.0 vs 0.5 per 100,000 population), mortality (3.0 vs 0.4 per 100,000 population) and DALYs (266 vs 33 per 100,000) respectively. Similar to global trends, all regions demonstrated decrease in these rates over the last two decades. Nationally, the highest age-standardized rates were reported in African countries, Central African Republic with highest incidence rate (7.6 per 100,000), and Burkina Faso with highest mortality rate (5.8 per 100,000) and DALY rate (518 per 100,000). Between 1990-2019, 182/204 (89%), 188/204 (92%) and 188/204 (92%) countries and territories demonstrated a decrease in age-standardized incidence, mortality and DALY rates respectively, with the greatest decreases seen in Saudi Arabia for all statistics.

**Conclusions:** Between 1990 to 2019, overall trends in incidence, mortality and DALY rates of NTD have been favorably downtrending globally. Regionally, these rates in the highest Sub-Saharan Africa were 8 times greater compared to the lowest North America. Nationally, although the majority of countries showed decreases in these rates, multiple countries demonstrated high and uptrending rates of NTD.

##### OP003

#### One-year paediatric outcomes following prenatal or postnatal myelomeningocele repair in a contemporary UK cohort

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**Introduction:** To compare clinical outcomes following prenatal repair of myelomeningocele (MMC) by the two-center NHS England Commissioned service with a contemporary series of standard postnatal closures.

**Methods:** A retrospective cohort study of UK patients who underwent open prenatal or postnatal repair of MMC between February 2017 and June 2021. All patients were managed by the joint fetal surgery centers, including University College London Hospital, University Hospitals Leuven, and Great Ormond Street Hospital. Primary outcomes include the persistence of Chiari II malformation and the need for shunt placement by 12 months of age. Secondary outcomes include the use of clean intermittent catheterization and motor milestones.

**Results:** 67 patients underwent prenatal and 31 received postnatal closure (n=98). Preterm birth was higher after prenatal repair (median GA 35+2 weeks), hence lower birthweight (median 2528g) as compared to postnatal repair (median GA 38+0 weeks,  $p<0.01$ ; median birthweight 3125g;  $p<0.01$ ). Hindbrain herniation was confirmed on postnatal MRI in 16.9% of infants after prenatal repair compared to 74.2% after postnatal repair ( $p<0.001$ ). In the prenatal repair group, 24.6% required shunt placement compared to 70.9% in the postnatal group ( $p<0.001$ ). 72.7% of patients required intermittent catheterization in the prenatal cohort compared to 67.7% of patients in the postnatal cohort ( $p=0.03$ ). There was no difference in motor milestones.

**Conclusions:** An initial experience of open prenatal repair surgery for MMC in a UK cohort with a contemporaneous postnatal group confirms outcomes in keeping with the MOMS trial, i.e., the reduced presence of Chiari II malformation and the need for shunt placement at 12 months of age.

#### OP004

##### A review of the results and complications in the first 100 cases of fetal repair of myelomeningocele

**Frank Van Calenbergh**, Roland Devlieger, Luc De Catte, Liesbeth Lewi, Honorine Maes, Philippe De Vloot, Jan Deprest  
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**Introduction:** In 2012, a program for fetal surgery for spina bifida was established in the University Hospital of Leuven, Belgium, accepting patients from different countries in Europe and even from other continents. From the start, it was decided to treat the patients in a decentralized way, i.e. that delivery and care of the neonate would be performed in the referring unit in their own country. The program was not considered a prospective clinical study but measures were taken to register the maternal and fetal outcomes as much as possible, and both the mothers and the centers received detailed information and instructions to provide feedback.

**Material and Methods:** An audit of the first 100 treated cases was performed. In cases where the referring center did not spontaneously return all necessary information, additional requests were made to the physicians and then to the mothers, by telephone and by e-mail. Complications were graded according to the Maternal and Fetal Adverse Terminology 5-graded classification.

**Results:** Primary skin closure was done in 77, while a patch was used in 23. In 41, the placode was neurulated. One fetus died 24 hours after the procedure, one fetus died at 29 weeks from an unrelated umbilical knot, one child died postnatally because of complications of prematurity (29 weeks). There were no maternal deaths or grade 4 complications, 5 experienced grade 3 complications. Gestational age at delivery was 35,3 weeks.

The indication for VP shunting or ETV was made in the different local centers: 46% of the children were treated for hydrocephalus.

**Discussion:** The outcomes and the rate of complications in our series are similar to that of the MOMS and other large series. Spontaneous return of outcome data by referring centers is sometimes disappointing but patient empowerment highly improves data collection.

#### OP005

##### Detethering in ARM patients: The role of multidisciplinary team in the surgical decision

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**Introduction:** Anorectal malformation (ARM) occurs in approximately 1–3 in 5000 live birth. Up to 60–70% of patients with ARM have occult spinal dysraphism (OSD). In most cases, patients with ARM have underlying bladder and bowel change. For this reason, the surgical indication is not always clear and there is not uniformity in the indications for detethering in the literature and in the working groups.

**Methods:** Patients with ARM consecutively treated between January 1999 and August 2022 were included. Patient demographics data, type of ARM according to Wingspread classification, associated anomalies, presence of OSD, signs and symptoms of tethered cord syndrome (TCS), the urodynamic and MRI studies and the neurosurgical procedures were retrospectively analyzed.

**Results:** 358 patients with ARM were enrolled in the study, 345/458 performed spinal cord MRI and 202 presented spinal cord anomalies. 36/202 patients required neurosurgical intervention. Fatty filum was the most common type of OSD (n=25/36). Median age for the diagnosis and surgical procedure were 27 and 58 months respectively. Bladder dysfunction and back pain were the most common symptoms in patients with fatty filum (16/25) due to the tethered cord syndrome (TCS). In the remaining patients, the surgical procedure was made according to the radiological evaluation and complexity of spinal dysraphisms. In the post-operative period about 80% of patients had a clinical or radiological improvement.

**Conclusion:** Our study suggests the importance of a multidisciplinary and dedicated approach in patients with ARM, to early detect the presence of OSD and to manage TCS. The main goal is to reduce the risk of neurological evolution even in asymptomatic patients and to improve quality of life. The application of a multidisciplinary workflow makes it possible to decide when the patients need neurosurgical procedure and what kind of studies are needed in the pre- and post-operative period

#### OP006

##### Ventral Tethering - Is the prognosis worse than in Dorsal Tethering in dysraphic spine?

**Sandip Chatterjee**  
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**Introduction:** Ventral tethering of the cord is seen in complex dysraphism cases-mainly neuroenteric cysts, split cord malformations, spinal cord herniation, complex lipomas and anterior sacral meningoceles. The aim was to compare cases of ventral tethering with cases of dorsal tethering seen over a 10 year period to ascertain whether they carried a worse prognosis as these were more difficult in terms of surgical challenges.

**Method:** Retrospective review was done of 16 neuroenteric cysts, 28 split cord malformations, 3 spinal cord herniations, and 5 anterior sacral meningoceles leading to a total of 52 ventral tethering cases. These were compared with 108 cases of dorsal tethering operated upon at the same time. All cases were operated upon by the same surgeon. Neurological status at presentation, signal changes in the cord, IONM changes at surgery, postoperative neurological status and radiological evidence of re-tethering postoperatively (24–48hours after surgery) were compared between the two groups.

**Results:** Cases of ventral tethering presented with far more neurodeficit than those with dorsal tethering, IONM changes were more in these cases as baseline deficits, and although radiological untethering was appreciable in similar percentage of cases, postoperative neurodeficit seemed to occur more in these cases though this did not reach statistical significance.

**Conclusion:** Ventral tethering cases must be considered as a special subset which is technically more challenging and can produce worse results than cases of dorsal tethering which are more frequent.

#### OP007

##### Is bulbo cavernosus reflex predictive of urinary incontinence in children with spinal dysraphism?

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**Introduction:** Bulbo cavernosus reflex (BCR) monitoring has been used as a surrogate measure of urinary function in children undergoing surgery for spinal dysraphism. However, the reliability and predictive value are unproven.

In this study, we aimed to assess if preoperative continence was correlated with BCR before surgery and, secondly, whether intraoperative loss of BCR predicted late incontinence in a cohort of patients undergoing spinal dysraphism surgery.

**Methodology:** A retrospective observational analytical study of patients aged 4 to 18 with spinal dysraphism undergoing spinal cord untethering with intraoperative monitoring of BCR between 2015 and 2020. Evaluation of bladder function was performed preoperatively and at least one year postoperatively. In all cases, baseline BCR was recorded before the commencement of surgery during and at the end of the procedure. Incontinence was defined as involuntary voiding of urine  $\geq 2$  times/month during the day or night, loss of desire to urinate, or the need for intermittent urethral catheterization. Loss of BCR was defined as a persistent reduction in amplitude of more than 50% compared with the baseline at the end of the surgery.

**Results:** 50 patients satisfied the inclusion criteria. Absent bilateral or unilateral BCRs before starting the surgery ( $n=15$ ) were correlated with preoperative incontinence with a sensitivity of 42.9% (95% CI 21.8–66) and a specificity of 79.3% (95% CI 60.3–92). Loss of bilateral or unilateral BCRs at the end of the surgery ( $n=8$ ) was predictive of incontinence with a sensitivity of 14.3% (95% CI 3.1–36.3) and a specificity of 82.8% (95% CI 64.2–94.2).

**Conclusion:** BCR is correlated with urological function. Furthermore, loss of BCR during surgery was the most specific test to predict urinary incontinence at a one-year follow-up. However, loss of BCR preoperatively was most robust to predict baseline urinary incontinence since it had an increased specificity with satisfactory sensitivity.

#### Abstract Session 2: Craniosynostosis

#### OP008

##### Advancing management of craniosynostosis using computer simulations

**Mehran Moazen**

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**Introduction:** The physical-mechanical interactions that shape our skulls during growth are poorly understood. This is a significant engineering challenge due to the complexity of this system. The aim of this study is to outline the computational approach that we have developed to predict skull growth and the outcome of different treatment approaches for the management of sagittal craniosynostosis.

**Methods:** A detail morphological and material property characterisation of normal and a craniosynostotic mouse model during the growth was carried out. These data enabled us to develop a validated finite element (FE) model of calvarial growth in mouse. Here we used intracranial volume to drive the calvarial growth and developed a strain-based tissue differentiation algorithm to predict the bone formation at the cranial joints.

**Results:** We then applied the same approach to develop a validated FE model of normal calvarial growth in human, having characterised morphological changes during the human craniofacial growth in  $n=241$ , aged 0–4 years. We then applied our approach to 2 patient-specific models of craniosynostosis predicting calvarial growth following the surgery. Our methodology was then used to compare the biomechanics of 10 different treatment options for sagittal craniosynostosis. Our results highlight that different surgical techniques can constrain the growth of the brain in different ways that can potentially have an impact on the neurodevelopment of these children.

**Conclusion:** In summary the computational approach developed and described here can be applied to compare various treatment options for different forms of craniosynostosis. Nonetheless, combination of this approach with geometric morphometric and artificial intelligence can potentially transform the future of craniofacial surgery.

This work has been supported by the RAEng (10216/119), Rosetrees Trust (A1899 & PhD2021\100017) and EPSRC (EP/W008092/1).

#### OP009

##### Volumetric changes after endoscopic correction of scaphocephaly according to CSF dynamics

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**Introduction:** CSF spaces may be enlarged in scaphocephaly. However, the role that CSF dynamics may play in this craniosynostosis is still uncertain. Aim of this work was to analyse the early post-operative evolution in children with sagittal suture synostosis submitted to an endoscopic repair followed by helmet therapy according to their preoperative CSF spaces.

**Methods:** Children with a sagittal synostosis submitted to an endoscopic removal of the sagittal bar and occipital and parietal decompression followed by helmet remodelling therapy between 2020 and 2021, were prospectively studied with a 3D optic scan before surgery (pre op), after surgery before helmet therapy (post-surgery) and at the end of helmet therapy (posty-helmet). Children were divided in subgroups (large vs small ventricles, large pericerebral spaces vs small) based on preoperative CT scans.

**Results:** Twenty-one children were included. There were 2 girls. The mean age at surgery was 4 months. A significant change in morphology could be appreciated in the immediate post-operative period. The mean preoperative cranial index (CI) was 67, the mean CI after surgery was 74.6 and the mean CI after helmet therapy was 81. Large ventricles were found in 13 children and enlarged pericerebral CSF spaces in 11. Though preoperative CI were similar (ventricular spaces analysis: 68.8 vs 66.8,  $p : 0.29$ , pericerebral spaces analysis : 66.8 vs 69.1,  $p : 0.19$ ), post surgical CI

were significantly bigger in case of large ventricle (75.6 vs 71.8,  $p : 0.04$ ) and in case of large pericerebral spaces (76.1 vs 72.06,  $p : 0.02$ ).

**Discussion:** CSF dynamics may play a role in increasing the post-operative remodelling obtained by a surgical parietal decompression. Conversely, the post-operative helmet therapy might be especially needed in children with small CSF spaces.

## OP010

### H craniectomy 12-years review in isolated sagittal craniosynostosis: evidence of improvement in reintervention rate and Esthetic results

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H craniectomy is one of the most common technique for the treatment of isolated sagittal craniosynostosis. A long-term analysis of 12 years of experience in the same Centre in 456 patients is presented.

**Methods:** Between 2010 and 2021, 456 patients have been operated for isolated scaphocephaly in Necker Hopital Enfants Malades in Paris. Clinical, radiological, neuropsychological data of 233 patients in the period 2010-2015-15 (group 1) and 233 patients in the period 2015-2021 (group 2) have been analysed. Some differences in age at surgery and mild changes in surgical technique has been introduced in 2015.

**Results:** In the group 1, we record 108 patients – 48.4% - operated older than 160 days, while in the group 2 we note only 30 patients - 12.8%. The re-intervention rate is the 3.6% in group 1 (8 patients) and 0.8% in group 2 (2 patients). Considering the age at surgery, 7/8 of re-operated patients in the group 1 and 2/2 in the group 2 can be observed in the older subgroups of patients. In both groups, we observe environ 80% of secondary coronal synostosis, 20% of mild cognitive difficulties and 6% of autism spectrum problems.

**Discussion:** H craniectomy is the gold standard surgical procedure for isolated scaphocephaly in our department. Trying to reduce the re intervention rate and the esthetic outcome, some changes has been introduced from 2015: the evidence of increased risk in patients firstly operated older than 160 days has been demonstrated. No significative differences in neuropsychological problems have been found in the 2 groups of patients: a possible genetic substrate need to be assessed.

**Conclusion:** H craniectomy is a very well know technique. The strict respect of age at surgery inferior of 160 days and an enlarged length of craniectomy seems to reduce the re intervention rate.

## OP011

### Free flaps cranial vault remodelling for sagittal synostosis

**Laura Grazia Valentini**<sup>1</sup>, Alessandra Erbetta, Enrico Gambatesa, Ignazio Gaspare Vetrano, Mirella Seveso, Tommaso Galbiati, Veronica Saletti

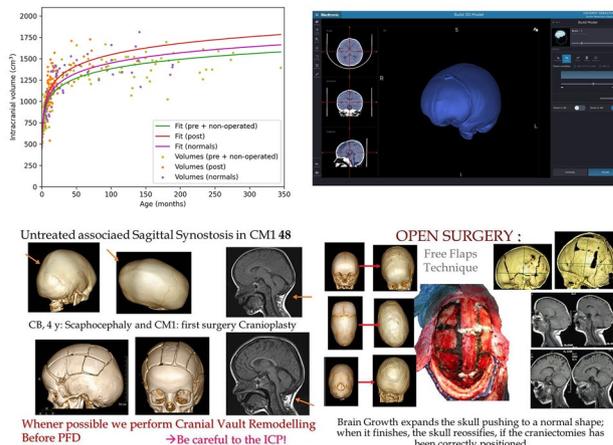
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We quantified the results of our technique. 175 children were operated on for sagittal synostosis as documented by 3DCT, with a male prevalence (136/39). At surgery 146 children were <1 (mean 6,8 months), 29 >1 year. The technique applied in the two groups was similar: multiple craniectomies were performed with high speed drill, by zig-zag bicoronal approach; bone flaps were programmed on preoperative-CT; two broad, symmetric fronto-parieto-temporal free flaps were created, and each one was divided, leaving a residual bone at vertex upon the sagittal sinus 2-4 cm wide, to prevent venous damage and late diastasis. To allow vertical growth, this bridge was interrupted by 2-4 transversal cuts, wo rigid fixation. Continuous absorbable suturing (“Denim”) was used to improve the scar and PIC was measured intraoperatively in the older. There was no mortality, nor major complications; 2 children were re-operated to improve cosmesis, 1 for progressive bone defect, one for CSF collection. Aesthetics was judged by doctors and parents in 150 children by serial photos, with a specific questionnaire. Neuropsychological results were evaluated on testing and school performance resulted in no delay in the early-surgery.

Volumes were measured on pre and postoperative 3DCT and compared with normal volumes, obtained in a comparable group of

children submitted to 3DCT for other reasons: preoperative volumes were lower than normal and this difference increased by age; post-operative volumes, on the contrary, were higher than normal, also improving in the “late surgery” group. At late MRI none of early operated children presented CM while it was documented in 9/29 of older children and regressed after surgery.

The present series confirms that CM1 may be due to sagittal synostosis, causing low intracranial volumes; our technique is highly repeatable, feasibility at any age, with low morbidity and high efficacy increasing intracranial volumes, so preventing CM.



## OP012

### When does the bregmatic fontanel close? A 3D-CT scanner study

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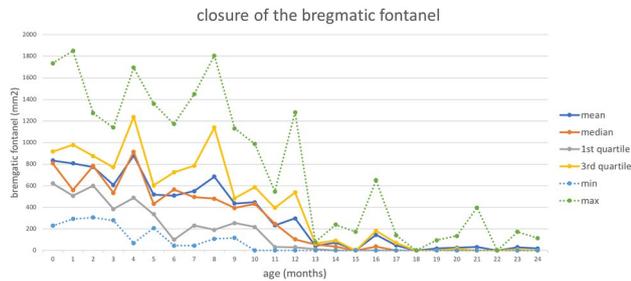
**Background:** The normal age of closure of the bregmatic fontanel is poorly documented in literature; as a result, the “small fontanel” is a common preoccupation for pediatrician colleagues who refer children for what turns out as unnecessary cranio-facial consultation. Data from literature are often old, based on limited numbers of dry skulls, and the measurements are fraught with problems of bone shrinkage and growing population standards. For many years now, imaging tools have been available, allowing the study of the anatomy and development in children, victims of accidental trauma being a priori drawn from a normal population.

**Material and Methods:** In order to evaluate the means and variations, we collected whenever possible 12 individuals from each of the 25 age groups between 0 and 24 months. Skull CT scanner of normal children victim of accidental trauma were studied using the Horos<sup>®</sup> freeware. CT were reconstructed in 3D-multiplanar reconstruction and the area of greater fontanel was traced manually using the “closed polygon” tool.

**Results:** We studied 207 skulls in 124 male and 83 female infants (M/F ratio 1.49). The results of area measurement showed a wide variety before the age of 12 months, and a steep decline in surface with loss of diversity after that age.

**Discussion:** The high variability of the size of the bregmatic fontanel during the first months of life, suggests that this variable is of little importance. Its rapid closure around the time of acquisition of the erect position also suggests that negative intracranial pressure is the stimulus for ossification of the calvaria.

**Conclusion:** Accidental traumas provide a large cohort for the study of normal anatomy in infants, and 3D CT database represent a rich trove of data, with sophisticated computer tools allowing easy and accurate measurements in live subjects.



### OP013

#### Long term follow up of the Leeds Fronto-Orbital Advancement and Reconstruction technique (reverse frontal bone graft without the use of the orbital bar)

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**Objective:** We present our long-term outcomes using a reverse frontal bone graft without the use of the orbital bar to perform FOAR surgery. Having previously used both the Marchac template technique and reversed frontal bone method utilizing the reshaped orbital bar we were often dissatisfied with the final shape- particularly with bitemporal narrowing.

**Methods:** Since mid 2015 we have performed 31 FOARs using this technique for non-syndromic metopic or unicoronal synostosis. Follow up ranges from 12-33 months.

In our technique the frontal bone flap is reversed and new orbital rims are fashioned. This is advanced in cases of coronal synostosis but not metopic. We remove the orbital bar & use it only as onlay grafts or bone dust, applying this at the temporal area or to any exposed dura.

**Results:** With this technique all 31 had good cosmetic outcomes. All constructs are stable and no revisions for cosmetic indications have been required. One patient does have bitemporal narrowing and is a candidate for future implants. 94% parents report they are either satisfied, pleased or very pleased with the outcomes. One patient has had raised ICP requiring posterior expansion, none have developed papilloedema and 81% have continued to have consistent head growth.

**Conclusion:** Our results show it is safe to remove the orbital bar all together and use its bone dust to fill gaps with good long term cosmetic results. Funding: N/A

### OP014

#### Novel noninvasive therapeutic approach for the treatment of craniosynostosis in Crouzon syndrome patients using nanoparticle-mediated siRNA delivery

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**Introduction:** Crouzon syndrome (CS) is a rare craniofacial malformation caused by gain-of-function mutations in the FGFR2 gene. Therapeutic protocols are based on multiple surgeries. The aim of this project is to develop a noninvasive therapy for CS by reducing the overactive FGFR2 signaling through allele-specific siRNAs targeting the mutant FGFR2 allele, delivered by highly biocompatible nanoparticles (NPs) in patient cells.

**Methods:** Calvarial mesenchymal stromal cells (CMSCs) were isolated from calvarial tissue of six FGFR2 mutation-positive CS patients. Specific siRNAs targeting the mutant FGFR2 allele of each patient were designed and evaluated in patient cells through Real time PCR and Western blot analyses. Ferritin-based nanocarrier and PLGA NPs have been evaluated for siRNAs delivery in CMSCs using fluorescence microscopy and Incucyte Live-cell analysis system.

**Results:** Gene expression analysis allowed identifying specific siRNAs that presented the strongest inhibitory effect on the expression of mutant FGFR2 allele without reducing the wild-type allele in CMSCs derived from CS patients. Fluorescence microscopy showed an efficient internalization of the tested NPs within CMSCs cytoplasm, thus demonstrating they are suitable delivery systems.

**Conclusion:** Our data suggested that allele-specific FGFR2 knockdown by siRNA represents a desirable strategy to silence FGFR2 mutant allele in CS patients. We are optimizing the intracellular trafficking and siRNAs release of nanoparticles-siRNAs constructs. Funding: AFM Telethon (Grant Ref. 23607).

### OP015

#### Role of BBS9 in the pathophysiology of Non-Syndromic Craniosynostosis

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**Introduction:** The physiopathology of nonsyndromic craniosynostosis (NCS) is yet to be clarified, though an aberrant osteogenic differentiation of calvarial MSCs (CSMCs) within sutures has been observed. We have previously demonstrated that CMSC from fused sutures produce less primary cilia and overexpress the Bardet Biedl syndrome gene 9 (BBS9) encoding a protein required for primary cilium assembly. Our aim is to clarify the role of the overexpressed BBS9 isoforms in NCS pathophysiology.

**Methods:** We performed in silico modeling of BBS9 assembly to the multiprotein BBSome complex and evaluated BBS9 expression during osteogenic differentiation in N- and P-CMSCs – respectively from unfused and fused sutures – isolated from surgical wastes from cranio-plastic surgery on NCS patients.

**Results:** The BBSome containing a BBS9 isoform lacking the C-term showed reduced stability. qPCR showed comparable expression trends across all the BBS9 mRNA variants, while BBS9 protein expression was higher in P- compared with N-CMSC.

**Conclusions:** BBS9 expression increases during osteogenic differentiation of CMSCs suggesting a functional implication in NCS etiopathogenesis. Further ongoing studies are aimed to identify the primary structure of the overexpressed isoforms and confirm their role in the BBSome assembly. We propose that BBS9 may represent a novel suitable biomarker for suture ossification in NCS, and a target for the design of innovative therapeutic strategies.

**OP016****MRI-based CAD-templates for cranosynostosis surgery****Matthias Krause***University Leipzig, Leipzig, Germany*

**Introduction:** Computer-assisted design and manufacturing (CAD/CAM) techniques have been implemented in cranosynostosis surgery to facilitate cranial remodeling. However, until now, computed tomography (CT) scans with ionizing radiation were necessary to plan the procedure and create guiding templates. The purpose of this study was to present our series using CAD/CAM techniques in planning and conducting fronto-orbital advancement surgery in patients with trigonocephaly with datasets acquired only by “black bone” magnetic resonance imaging (MRI).

**Methods:** Twelve consecutively operated cases from 2019 were included in this study. All patients had suffered from non-syndromic trigonocephaly with no primary surgeries. All patients received underwent cranial MRI including black bone sequences. Preoperative planning, and guides were created based on the DICOM datasets. We analyzed Demographic data, clinical data, and outcome measured by Whitaker score.

**Results:** In all cases, precise frontobasal advancement was possible with the CAD/CAM guides created by black bone MRI. The mean operation time, and planning time were 222 and 32 minutes. The time on intensive and intermediate care unit (ICU/IMC) time was 4.5 days, respectively. All but one case was classified as Whitaker I.

**Conclusion:** In trigonocephaly followed treatment by frontobasal advancement, black bone MRI-based CAD/CAM cranosynostosis surgery is safe and feasible. It offers the major advantage of completely avoiding CT scans and ionizing radiation with superior imaging quality of intracranial structures. Thus, it improves intraoperative safety and –at the same time– has the potential to reduce operating room (OR) time.

**Abstract Session 3: Technology****OP017****Robot-assisted endoscopy in pediatric neurosurgery****Davide Luglietto**, Alessandro De Benedictis, Antonella Bua, Alessandra Marasi, Andrea Carai, Carlotta Ginevra Nucci, Alessandra Savioli, Carlo Efsio Marras*Ospedale Pediatrico Bambino Gesù, Rome, Italy*

**Introduction:** The development of robotics in neurosurgery is improving the quality of surgical procedures requiring a high level of accuracy and safety [1],[2]. However, the combined use of standard stereotactic methods and robotic technology for endoscopy approaches has been rarely reported so far [3]. We report on a series of pediatric patients who underwent endoscopic procedures with the assistance of the ROSA (Robotized Surgical Assistant, Zimmer Biomet) system at the Neurosurgery Unit of the Bambino Gesù Children's Hospital, in Rome, Italy.

**Methods:** We retrospectively evaluated 168 pediatric patients who underwent 198 robot-assisted procedures from 2011 to 2022. Among these patients, we selected 80 endoscopy cases. The pre-operative neuroimaging planning included MR images and a volumetric CT scan. Registration was performed following the specific robot-guided workflow. A rigid 30° endoscope (medium size, Karl Storz), integrated with the robot arm, was used under navigation control.

**Results:** We performed 80 endoscopic procedures assisted by the ROSA system. They included: 5 ETVs, 5 endoscopic septostomies, 10

endoscopic cysto-ventriculostomies, 27 tumor biopsies and 1 tumor resection, 37 hypothalamic hamartomas (HH) disconnections. In all cases, the surgical assistant allowed to successfully complete the surgery. Transient postoperative complications included 1 frontal subdural hematoma for tumor biopsies (3,7%), and transient hypothalamic dysregulation in 3 HH cases.

**Conclusion:** Robotic technology may improve precision and safety of minimally invasive endoscopic approaches, optimizing the quality of treatment of several diseases, also extending the spectrum of indications to more complex conditions, such as small sized or distorted ventricles, multiple trajectories, narrower corridors, and long-lasting procedures.

[1] A. DB et al., “Robot-assisted procedures in pediatric neurosurgery,” *Neurosurg. Focus*, 2017

[2] J. J. D et al. “Robotics in neurosurgery: Evolution, current challenges, and compromises,” *Cancer Control*, 2015

[3] A. P. et al. “Expanding the spectrum of robotic assistance in cranial neurosurgery,” *Oper. Neurosurg.*, 2019

**OP018****Predictive value of sonographic parameters on the effects of cranial molding helmet therapy in infants with positional plagiocephaly****Maria Licci<sup>1</sup>**, Agnes Paasche<sup>2</sup>, Alexandru Szathmari<sup>2</sup>, Pierre Aurélien Beuriat<sup>2</sup>, Carmine Mottolose<sup>2</sup>, Federico Di Rocco<sup>2</sup><sup>1</sup>*University Hospital Basel, Basel, Switzerland*, <sup>2</sup>*Hôpital Femme Mère Enfant, Lyon, France*

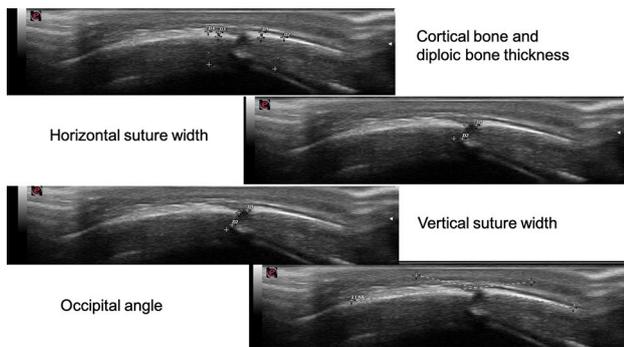
**Background and Aim:** In a population of infants with positional unilateral or asymmetric bilateral plagiocephaly - treated with cranial molding helmet orthosis at the pediatric neurosurgery department of the "Hôpital Femme Mère Enfant" - we aimed at investigating the predictive value of ultrasonography- parameters on the efficacy of orthotic helmet therapy for skull shape correction. Secondly, a correlative analysis of the correlative effect of both caliper cephalometry values and age on treatment efficacy was performed.

**Methods:** Prospective, non-randomized, non-invasive, single-center data collection over a six months period between January 2021 and June 2021 on a consecutive cohort of 50 patients <10 months of age at time of therapy onset. The following sonographic parameters were assessed on both the affected and unaffected side at onset of treatment and within 8-12 weeks: lambdoid suture width, adjacent cortical and diploic bone thickness and occipital angle. Caliper cephalometry value collection consisted in CVAI, CI and ear displacement. Clinical appearance was graded according to the classification of Argenta (I-V)

**Results:** Bone thickness at onset of orthotic helmet treatment and initial caliper cephalometry angles reflect radiologically

objectionable occipital angle changes over time. In fact, there is a correlation between bone thickness and improvement of occipital angle, with smaller bone thickness responding better to remodeling orthotic helmet therapy. (p=0.030). Moreover, both longer duration of helmet treatment/day and overall helmet treatment duration lead to reduction of CVAI Severity over time (p= 0.010 and p=0.042, respectively). Finally, efficacy of orthotic helmet treatment inversely depends on age at onset of treatment (p=0.035).

**Discussion:** In addition to the well-known diagnostic value in differentiation between positional and cranosynostotic head deformities, ultrasonographic measurements seem to add predictive information concerning the efficacy of orthotic helmet deformation and to relativize the sole impact of age at treatment onset.



#### Abstract Session 4: Trauma/Vascular

##### OP019

#### Suture associated extra-dural haemorrhage as possible new diagnostic sign in patients with history of non-accidental injury due to shaking mechanism

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**Introduction:** This study aims to assess the diagnostic utility of adjacent suture associated extra-dural haemorrhage (SAEDH) on brain Computed Tomography (CT) scan of children victim of physical abuse to corroborate a non-accidental injury (NAI) diagnosis with a shaken mechanism.

**Methods:** We undertook a retrospective study of all patients admitted to a supra-regional paediatric neurosurgical institution with suspected NAI and evidence of intracranial trauma on CT head referred between January 2011 and June 2022. Patients with subsequent NAI confirmation by the child protection team represented the main cohort of our study. This was compared to a non-NAI control group. All presenting CT scans were reviewed by a senior paediatric consultant radiologist to determine whether there was a SAEDH.

**Results:** 88 patients were identified (mean age six and a half months). 31 confirmed NAI (mean age two months old) and 57 non-NAI (mean age seven months). Six out of 31 patients (19.4%) with confirmed NAI had SAEDH on initial presenting imaging. Only one patient out of 57 (1.7%) in the non-NAI group had SAEDH. Thirteen patients were identified as shaken in nature with four of them presenting with SAEDH on radiological imaging (31%). There was a significant difference in the rate of the SAEDH between shaken a babies and NAI children ( $p$  0.0035).

**Conclusion:** In children with confirmed NAI and cranial trauma with mechanism of shaking injury we identified SAEDH in nearly a third of the patients. We believe this previously unpublished radiological finding, if confirmed by other larger studies, could be of diagnostic help when considering the possibility of NAI due to shaking mechanism.

##### OP020

#### School performance following traumatic brain injury – a nationwide register-based cohort study

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**Introduction:** Paediatric traumatic brain injury (TBI) is a social and global burden. Minor and mild TBI can, as well as moderate and severe TBI, be followed by cognitive and behavioural impairment. Sequela following head trauma varies and may not be visible before the child is facing higher academic demands. This nationwide study aimed to estimate long-term educational consequences following childhood TBI.

**Methods:** All living-born children in Denmark (1977–2002) were included and followed through Danish national health and educational registers until 2018. Children with no TBI were used as controls. There were no exclusion criteria.

**Results:** Of 1,527,513 children, 88,015 suffered from TBI (30,609 minor, 46,379 mild, 6,983 moderate, and 4,044 severe) before the age of 15 years. Children with TBI were 4% less likely to complete compulsory school than other children. Severity as well as accumulated number of TBI's throughout childhood had a negative effect on completing compulsory school, with a reduction range from 2% to 9% with increased severity and 4% to 15% with increased numbers. Of children with TBI, 89.7% achieved a final grade point average (GPA) when completing school, compared to 92.0% among other children (average 6.21 vs 6.51, respectively). Fifty-four percent of children with TBI had a GPA below that of the entire cohort, compared to 48% of other children. GPA decreased in children of both sexes with increasing numbers of TBI's, severity of TBI, and older age at time of TBI. Interaction analyses showed that neurological and psychiatric co-morbidities in the patients or parents did not markedly affect outcome.

**Conclusion:** TBI during childhood reduce the chance of completing compulsory school and receiving the same GPA as other children. The effect increases by numbers of TBI's and TBI severity. The findings highlight the importance of educational support, even in children with less severe TBI.

##### OP021

#### External lumbar drainage in the management of refractory intracranial hypertension in pediatric traumatic brain injury: a retrospective single-center study

**Lelio Guida**, Alissa Visentin, Kevin Beccaria, Sandro Benichi, Syril James, Giovanna Paternoster, Thomas Blauwblomme  
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**Introduction:** The management of refractory intracranial hypertension (rIH) in children with severe Traumatic Brain Injury is still challenging, as its treatment with burst-suppression drugs or decompressive craniectomy (DC) has a high morbidity rate. Cerebrospinal fluid (CSF) drainage represents a tier 1 treatment; however, External Ventricular Drainage (EVD) in children can be challenging because of the small size of lateral ventricles. External lumbar drainage (ELD) could be a useful alternative for CSF depletion, therefore we analyzed the outcome of a cohort of pediatric patients who underwent ELD to treat rIH.

**Methods:** This study retrospectively enrolled pediatric patients in rIH following severe TBI undergoing ELD. ICP and Cerebral Perfusion Pressure (CPP) curves were analyzed 12 hours before and after the procedure. Any change in medical therapy was recorded, as well as total volume and length of drainage. Cerebellar tonsillar position according to the McRae line was noted before and after ELD.

**Results:** 30 patients were included, with a mean age of  $8.3 \pm 4.3$  years, and a median GCS at admission 7 (range 3–13). ELD was performed after a median delay of 1 day (range 0–7), mean drainage volume/day was of  $296 \pm 129$  mL, median duration of drainage was 7 days (range 2–12). ICP significantly decreased after ELD (mean difference  $10.7 \pm 4.2$  mmHg,  $p < 0.005$ ), whereas CPP significantly increased (mean difference  $13.7 \pm 1.2$  mmHg). 12 patients (40%) did not need

any burst-suppression drug after ELD, and two patients underwent DC to control ICP (6%). No complication was noted, cerebellar tonsil level did not vary at imaging control before ELD ablation.

**Conclusion:** ELD is safe and effective to treat rIH in children, limiting the use of burst-suppression drugs and DC. This pilot study could pave the way to a multicentric European prospective trial.

#### OP022

##### **Avoiding decompressive craniectomy: Barbiturate coma as first tier treatment of refractory intracranial hypertension**

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**Introduction:** Refractory intracranial pressure hypertension (RICH) following traumatic brain injury (TBI) is a severe condition that requires potentially harmful treatment strategies such as barbiturate coma (BC) and/or decompressive craniectomy (DC). There are concerns regarding complications and outcome related to both treatments. The aim was to evaluate the outcome and occurrence of complications related to BC treatment and the need for DC in children with RICH due to TBI, using a standardized escalated management protocol. BC was used before DC in cases with RICH and no mass.

**Methods:** Ten year retrospective cohort of all children, age  $\leq 16$  years, treated at NICU Uppsala with BC due to RICH following TBI. Data were acquired from patient records and digital monitoring system files.

**Results:** Twenty-one children were studied, median age 14 years (range 2–16), median GCS 7 (range 4–8) and median Rotterdam CT brain score 4 (range 4–5) at admission. Sixteen patients (76%) did not require DC to control RICH. Three children had unilateral DC; two before and one during BC. Two children had bilateral DC; one during and one after BC. Another six children had mass lesions evacuated prior to BC. Average BC initiation 46 hours (range 5–198) after TBI and duration 107 hours (range 50–329). Median max S-thiopental 220  $\mu\text{mol/L}$  (range 30–460). The onset of BC resulted in lower ICP values, amplitudes and decreased incidence of A-waves. No major disturbances in blood gases, liver and kidney function or secondary insults were seen during this period. One year median outcome showed Glasgow outcome scale (GOS) 5 (good recovery) and Kings Outcome scale for head injury (KOSCHI) 4a (moderate disability).

**Conclusions:** The use of BC in TBI children with RICH as first tier treatment in cases without mass reduces the need for DC, without severe complications and with relatively good long-term outcome.

#### OP023

##### **Introducing the ESPN Pediatric Craniectomy and Cranioplasty Registry (PedCCR)**

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**Introduction:** In the pediatric age group the evidence regarding decompressive craniectomy (DC) is weak. Literature on pediatric cranioplasty is even more limited. Concerns are resorption of autologous bone flaps, skin breakdown and growth-related problems associated with synthetic implants. The relevance of DC and subsequent cranioplasty and the need for accurate and systematic data have led

to the creation of the Pediatric Craniectomy & Cranioplasty Registry (PedCCR) under the auspices of ESPN.

**Methods:** After thorough literature review, gaps in knowledge and evidence were identified and the following aims for PedCCR were defined: 1) Compare surgical practices regarding indication, timing and operative technique in a large multicenter cohort. 2) Provide outcome data in a large multicenter cohort. 3) Establish risk factors for complications and cranioplasty failure based on systematic data reaching back as far as the initial injury.

**Results:** The PedCCR is a prospective, multicenter, open registry enrolling children ( $\leq 16$  years) undergoing DC and/or cranioplasty for any pathology. A steering committee will assure good scientific practice. Standardized questionnaires were designed and data will be collected online in an electronic joint database, which is currently in the testing phase. The core set of data comprises case report forms for DC, cranioplasty and follow-up visits for at least 24 months, with follow-up forms being recommended until adulthood. Incident reporting forms can be submitted for any incident or unscheduled operation. All forms allow reporting of basic data even in the absence of the patient, i.e. by telephone interview or written contact to parents/carers/treating physicians/rehabilitation clinic.

**Conclusions:** The PedCCR will answer highly relevant open questions regarding pediatric DC and cranioplasty by using a high-quality prospective approach. The registry proposal was designed to maximize scientific yield at reasonable effort for participating centers. Collaboration is highly appreciated to advance this project together, as the registry is now recruiting.

#### OP024

##### **Predictive Factors of Surgical Revascularizations for Pediatric Moyamoya: A Meta-Regression Analysis**

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**Introduction:** There is no clear consensus regarding the technique of surgical revascularization for moyamoya disease and syndrome (MMD/MMS) in the pediatric population. This meta-analysis aimed to report a comparison between studies of indirect (IB), direct (DB) and combined bypasses (CB) in pediatric patients with MMD/MMS. Furthermore, we investigate predictors of outcomes.

**Methods:** In accordance with PRISMA guidelines, systematic searches of Medline, Embase and Cochrane Central were undertaken from database inception to 7th October 2022. Perioperative adverse events were the primary outcome measure. Secondary outcomes were rates of long-term revascularization, stroke recurrence, morbidity and mortality. Summary-level meta-regression was performed using the mixed-effects model to identify predictors of perioperative TIA, stroke, long-term revascularization, stroke and mortality. Predictors were year of publication, age, presence of MMS, presence of sickle cell disease, neurofibromatosis and Down syndrome, in accordance with literature.

**Results:** 37 studies reporting 4432 hemispheres were included in the meta-analysis. Overall pooled mean age was 9 years, and 45.0% were male. There was no significant difference in perioperative adverse events, stroke recurrence, morbidity and mortality between the DB/CB and IB groups. After meta-regression analysis, year of publication and age were significant predictors of outcomes. On meta-regression, age significantly predicted rates of perioperative stroke ( $p=0.048$ ), rates of perioperative TIA ( $p=0.005$ ). Furthermore, meta-regression identified age ( $p=0.010$ ) as a significant predictor

stroke recurrence. Meta-regression did not identify the presence of MMS nor its specific phenotypes to significantly affect outcomes, however this is likely to be a function of the limited number of studies reporting them.

**Conclusions:** Low-quality GRADE evidence suggest IB, DB/CB are relatively effective and safe revascularization options for pediatric MMD/MMS. The large number of studies enabled us to perform a meta-regression to explore possible confounders. Younger age is associated with higher risk of peri-operative stroke, TIA complications and long-term stroke recurrence.

#### OP025

**Pediatric brain arteriovenous malformation clustering and MRI atlas**  
**Sandro Benichi**<sup>1</sup>, Joseph Benzakoun<sup>2</sup>, Maxime Taverne<sup>3</sup>, Grégoire Boulouis<sup>4</sup>, Lélio Guida<sup>1</sup>, Marie Bourgeois<sup>1</sup>, Gabriel Morin<sup>5</sup>, Guillaume Canaud<sup>5</sup>, Manuelle Kossorotoff<sup>6</sup>, Olivier Naggara<sup>2</sup>, Thomas Blauwblomme<sup>1</sup>

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**Introduction:** Pediatric brain arteriovenous malformation (PbAVM) is a rare but life-threatening disease. PbAVM shows specific angio-architectural features and behaviors compared to adult onset. We aim at characterizing PbAVM in relation to shape and location.

**Methods:** Between 2014 and 2022, we retrospectively included 112 patients carrying 113 AVM that underwent MRI and DSA. We recorded purely angioarchitectural data on DSA and performed principal component analysis with hierarchical clustering to identify subgroups. We added 123 patients carrying 132 AVM using unsupervised machine learning cluster attribution on MRI data to create MRI atlas after manual nidus segmentation and special normalization procedure. We performed probabilistic analyses using voxel-based lesion symptom mapping technique.

**Results:** 245 AVM were included in the final analysis. Hierarchical clustering isolated 3 clusters of patients related to nidal volume and vessel structures. MRI atlas revealed that AVM distribution differed from randomness. We identified preferential location for clinical presentation and cluster.

**Conclusion:** We highlighted three clusters of PbAVM and preferential location according to clinical presentation and cluster. Further studies are needed to determine clinical and biological significance of our findings.

#### OP026

**Multi-modality management for pediatric AVMs: a single-center experience**

**Giuseppe Mirone**<sup>1</sup>, Alessia Imperato<sup>1</sup>, Nicola Onorini<sup>1</sup>, Claudio Ruggiero<sup>1</sup>, Domenico Cicala<sup>1</sup>, Mario Muto<sup>2</sup>, Giuseppe Cinalli<sup>1</sup>

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**Objective:** Intracranial arteriovenous malformations (AVMs) are a common cause of intracranial hemorrhage in children. The authors review their experience in the treatment of pediatric AVMs.

**Methods:** Demographic, clinical, and radiological data of pediatric patients with AVMs, treated in our institution from 2007 to 2022, were reviewed to compare the clinical and functional outcomes of patients managed with single or combined treatment.

**Results:** Fifty patients were diagnosed and treated during the study period. Six patients (12%) presented with non-bleeding AVMs, and forty-four (88%) presented with cerebral hemorrhage which onset symptom. Among the unruptured AVMs, the onset symptoms were headache in 2 cases, in 2 cases incidental (MRI performed for GH deficiency), and in 2 cases seizures. 50% of unruptured AVMs and 11% of ruptured AVMs were treated conservatively. Overall, eighteen (36%) AVMs were treated surgically. Only 18% of AVMs underwent radiation therapy (3 of which were non-bleeding at onset), reserving this type of treatment for vascular malformations that cannot be attacked surgically and/or by route endovascular or to those AVMs in which surgical and endovascular treatment had not produced a satisfactory result. Three patients (6% of the series) died because of major bleeding: a thalamic-midbrain, one from the posterior cranial fossa, and one patient with multiple AVMs. 78% of patients overall had a score between 0-2 on the modified Rankin Scale (mRS). The mean follow-up was 6.44 years (5 months-15 years).

**Conclusions:** The decision to treat any given AVM is based upon a critical evaluation of the balance of the implicit risks of the natural history of these lesions and the implicit risks of any therapeutic approach. Our data suggest that patients with lower grade AVM treated surgically and those with AVM of higher grade managed without surgery have good functional and clinical outcomes.

#### Abstract Session 5: Hydrocephalus I

##### OP027

**Transorbital point-of-care ultrasound versus fundoscopic papilledema to identify potentially elevated intracranial pressure in children**

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**Introduction:** Fundoscopic papilloedema is missing in approximately 50% of cases with raised ICP requiring treatment. Transorbital point-of-care-ultrasound techniques - optic nerve sheath diameter (ONSD) and optic disc elevation (ODE) - were compared to fundoscopic findings in children with symptoms rising suspicion of raised ICP.

**Methods:** 72 children were included; 50 with proven pathology associated with raised ICP (e.g. pseudotumor cerebri, tumor, hydrocephalus), 22 with pathology excluded. Bilateral ONSD and ODE were quantified by US using a 12 MHz linear array transducer. This was compared to fundoscopic optic disc findings and, in 28 cases, invasively measured ICP values.

**Results:** The diagnostic accuracy of cut-off values for potentially raised ICP for ONSD (5.73mm, AUROC 0.952) and ODE (0.43mm, AUROC 0.804) was considerably higher compared to fundoscopic papilledema (AUROC 0.730). Repeatability and observer-reliability of ODE examination was eminent (Cronbach's  $\alpha = 0.978-0.989$ ). Papilledema was only found in case ODE was  $> 0.67$ mm, an ODE  $> 0.43$  had a positive predictive value for potentially raised ICP of 90%. Sonographic examination results were more appropriate to indicate success / failure of treatment than fundoscopic papilledema.

**Conclusions:** In our cohort transorbital point-of-care US determination of ONSD and ODE could more reliably detect potentially elevated ICP in children than fundoscopy. Sonographic determination of ONSD and ODE did indicate ICP decrease after treatment earlier and more reliably than fundoscopy. The established cut-off values for US-ONSD and US-ODE and a newly developed US-based grading of ODE can be

used as an ideal first-line-screening tool to detect or exclude conditions with potentially elevated ICP in children.

#### OP028

##### Point-of-care ultrasound-based initial diagnosis and follow-up monitoring of pediatric idiopathic intracranial hypertension

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**Introduction:** Part of the signature of pediatric idiopathic intracranial hypertension (pIIH) is raised ICP and normal ventricular size. Both parameters can be assessed or measured, respectively, by point-of-care ultrasound techniques. We investigate whether a combination of optic nerve sheath diameter (US-ONSD) and third ventricular diameter (US-TVD) can be used as screening tool in pIIH to indicate elevated intracranial pressure (ICP) and exclude hydrocephalus as underlying cause. Further, we analyzed the potential of US-ONSD and US-TVD to monitor treatment effects and disease recurrence.

**Methods:** 888 ultrasound examinations were performed in 36 children with pIIH and 32 control patients. Using a 12-Mhz linear transducer and a 1-4-Mhz phased-array transducer, respectively, US-ONSD and US-TVD were determined repeatedly at baseline and during the course of treatment over a maximum period of 72 months.

**Results:** Mean baseline US-ONSD was significantly larger in pIIH patients ( $6.45 \pm 0.65$ mm) compared to control children ( $4.96 \pm 0.32$ mm); mean baseline US-TVD ( $1.69 \pm 0.65$ mm) of pIIH patients was significantly smaller compared to control ( $2.99 \pm 1.31$ mm),  $p < 0.001$ . During the course of successful treatment US-ONSD significantly decreased, while US-TVD increased divergently. Both, US-ONSD and US-TVD indicated relapse and repeat treatment success after relapse. The main co-morbid associations of pIIH were venous sinus abnormalities (96.2%), overweight/obesity (36.1%) and autism-spectrum disorder (8%).

**Conclusion:** The combined use of point-of-care US-ONSD and US-TVD is an ideal noninvasive screening approach to suspect elevated ICP and rule out hydrocephalus at the same time, which is regarding raised ICP especially important in patients without papilledema. Treatment success of pIIH can be effectively monitored by repeated US examinations, which also reliably indicate recurrence. This can avoid repeated invasive lumbar punctures after initial diagnosis.

#### OP029

##### The influence of the Covid-19 pandemic on the shunt revision rate

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**Introduction:** During the first year of the Covid-19 pandemic we observed a decrease of our shunt revision rate. In order to investigate a possible correlation with an assumingly lower general infection rate in children in times of lock down and homeschooling, we performed a detailed analysis of our shunt and general pediatric patient population.

**Methods:** Electronic patient charts retrieval for children admitted for shunt revision or infectious diseases was performed for four time periods (study period April 2020 – March 2021, control periods from three previous years). A detailed analysis of all shunt revision and infectious cases including age and season specific evaluation followed. Possible correlations were investigated.

**Results:** A total of 318 shunt revision and 13,919 pediatric cases have been evaluated. The shunt revision rate during the study period was 29% less compared to the average rate of three previous years ( $p = 0.061$ ), the number of pediatric cases with main diagnosis infection dropped significantly ( $p < 0.05$ ), whereas other pediatric admissions remained stable. Significant age or seasonal influences did not exist. The number of shunt revisions in association with a documented systemic infection or a primary shunt infection dropped significantly

during the study period ( $p < 0.05$  each). This was not the case for underdrainage, overdrainage ( $p > 0.05$  each) or other indications. In general, infections of upper and lower airways, the gastrointestinal and nervous system decreased during the pandemic, urinary infection rates remained stable.

**Conclusion:** The decreased shunt revision rate during the first year of the pandemic seems correlate with a decrease of the general infection rate in children and adolescents at the same time. Infection-associated shunt failures showed a significant decrease during this period compared to previous years.

#### OP030

##### The analysis of cerebrospinal flow dynamics in mouse fetuses - Cerebrospinal fluid may flow out from the brain through the frontal skull base and choroid plexus

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**Objectives:** Cerebrospinal fluid (CSF) has been believed to be produced in choroid plexus, and move to third and 4th ventricles, and drain into arachnoid granules to sagittal sinus in one-way. Recently, new CSF drainage tracts were proposed such as brain capillaries, meningeal and nasal lymphatic system. In this study, we investigated the extracranial outflow of particles in CSF. The analysis will contribute for the treatment of hydrocephalus, prevention of brain aging and cerebral degenerative diseases.

**Materials and Methods:** Mouse fetuses at a gestational age of 13 days were used. Either Fluorescent probes with differing molecular weights (MW) or gold particles with different size or cadaverine were injected into the lateral ventricle. The probes movements were monitored in live. Gold particles deposition was studied under stereomicroscope or electron microscope (EM). Cadaverine showed its movement tracts.

**Results:** 1) Fluorescent study: The injected probes dispersed into the 3rd ventricle and aqueduct immediately, but did not move into the 4th ventricle and spinal canal. With an injection of low and high molecular weight (LMW, HMW) probes conjugate, both probes dispersed into the brain, but only the LMW probe dispersed into the whole body. 2) Gold particle study: The accumulation of gold particles on the frontal skull base was observed. EM study revealed gold particles deposition between the ependymal cells. And only small size particle (2 nm) was found in liver at 15 minutes after injection. 3) Cadaverine study: Its deposition was observed on the frontal skull base, choroid plexus, ependyme and peri-vascular space.

**Conclusion:** 1) The particles in CSF may drain to extracranial space via frontal skull base and choroid plexus through ependyme and peri-vascular space quickly. 2) This outflow of particles is regulated by its MW and size. This work is supported by a Grant, JSPS KAKENHI (18K08963).

#### OP031

##### Management of Abdominal Pseudocyst (APC) in Children with Shunted Hydrocephalus: A Hydrocephalus Clinical Research Network (HCRN) Study

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**Introduction:** Abdominal pseudocyst (APC) causes distal site failure in children with VP shunts. Management of children with APC in HCRN have not been assessed.

**Methods:** The HCRN Registry was queried for children diagnosed with APC and management strategies were compared. The primary outcome was shunt failure/infection after APC diagnosis/management.

**Results:** 147 children were analyzed for initial management of APC; shunt surgery prior to APC diagnosis was revision (71%), insertion (18%) or infection (11%). Time from previous shunt surgery to APC diagnosis was 3.9 months. CSF gram stain was positive in 14/140 (10%) and APC cultures were positive in 14/147 (9.5%). Initial management of APC: 67% were treated with externalization, 29% had shunt removal, and 4% underwent revision without removal (all underwent re-operation within 1 month). There was no association between initial management and gram stain/culture positivity. Children managed with externalization were more commonly re-implanted in an extraperitoneal site after treatment ( $p=0.01$ ). There was no difference in shunt survival (log-rank test,  $p=0.45$ ) between children who had shunt re-implantation in the abdomen vs. extraperitoneal location. The type of shunt failure after APC treatment was more commonly revision after extraperitoneal vs abdominal site re-implantation (43% vs. 23%,  $p<0.01$ ). Infection was a more common type of failure when shunts were re-implanted in the abdomen vs extraperitoneal (27% vs. 8%,  $p<0.01$ ): median follow up 1.3 years. Thirteen (8.8%) children suffered a recurrent APC.

**Conclusions:** APC is usually managed with externalization; revision without removal led to failure within 30 days. Although no differences were found in overall shunt failure rate between re-implantation in the abdominal cavity vs. extraperitoneal site, infection was a more common reason for failure after reimplantation in the abdomen.

### OP032

#### Setting ventricular shunt independence: a retrospective study of a single institution's 25-year experience among 529 pediatric patients with hydrocephalus having undergone VPS placement

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**Introduction:** Most patients having undergone ventriculoperitoneal shunts (VPS) placement will require for their entire lifetime a cerebrospinal fluid (CSF) diversion. In recent times, however, the dogma “once a shunt, always a shunt” has been questioned and pediatric neurosurgeons have demonstrated an increasing interest in the possibility to remove VPS, setting a shunt free life to hydrocephalic patient.

**Methods:** We have retrospectively analyzed our pediatric cohort of 529 patients with hydrocephalus having undergone VPS placement over 25 years at our single institution. In 104 patients, we have performed surgical strategies and reached the shunt independence, assessed by a long-term follow up.

**Results:** During the follow-up of patients with VPS, we have paid unceasing attention both to patients with shunt failure that gave an opportunity for shunt weaning and also to those long-time shunted patients in a good health condition to whom we then have performed shunt removal surgeries whenever possible. We have compared shunt-dependent patients both to shunt-free patients and to patients which shunt freedom surgery failed; we have retrospectively analyzed patients' clinical data and focused on diagnosis, time of shunting, number and type of surgical procedure in order to have an accurate overall view. In our experience, we have reached in our hydrocephalic pediatric shunted cohort the 20.03% rate of shunt independence.

**Conclusion:** Shunt free life represents a real benefit to pediatric patients in terms of re-surgery rate, quality of life and long-term outcome. The analysis of our cohort of patients confirms that is possible to stop the vicious cycle represented by the VPS.

### OP033

#### Comparison study of shortened and non-shortened distal catheters in ventriculo-peritoneal shunts in growing children – a paradigm shift

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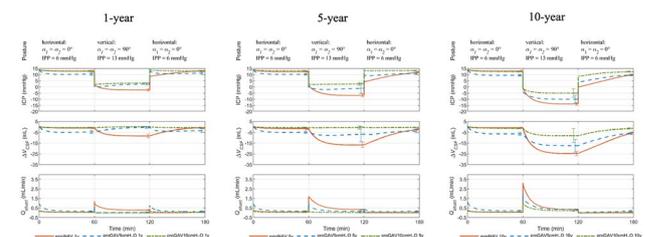
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**Introduction:** Hydrocephalic children treated with ventriculo-peritoneal shunts (VPS) face several developmental challenges. In addition to the modification of intracranial pressure with brain development, the increase in body length shifts the draining pattern towards more drainage. Furthermore, the distal catheter is often cut due to the smaller dimensions of the abdominal cavity of younger children. There is no data which directly compares the effect of shortened and non-shortened catheters with different valves in growing children. This study investigates the effects of shortened distal catheters in the shunt drainage behavior at different ages.

**Methods:** Two different distal catheters (60cm and 120cm in length) were tested using three differential-pressure Miethke valves (MINAV, 10 cmH<sub>2</sub>O; MN10), and three adjustable ProGAV2.0 valves at either 5 cmH<sub>2</sub>O (PG5) or 10 cmH<sub>2</sub>O (PG10) opening pressures. The hardware-in-the-loop test bed was adjusted for the physical length of one-, five- and ten-year old children. The software simulated the physiological conditions of intracranial and intra-abdominal pressures. All valves with both catheter-lengths were tested in five one hour runs in horizontal and vertical positions. Intracranial pressure (ICP), cerebrospinal fluid (CSF) volume changes and the flow through the VPS were measured.

**Results:** The shortened distal catheter led to a significant ICP drop in the vertical position on PG5 for all ages ( $p<0.05$ ), on MN10 for 10-y ( $p=0.002$ ) and on PG10 for 5-y ( $p=0.04$ ). There was a significant decrease in CSF volume with the 60cm catheter in vertical position for all ages on PG5 and MN10.

**Conclusions:** This study shows that shortening the distal catheter leads to increased drainage patterns in children as young as five. This is even more pronounced for low pressure opening valves and in valves without GU. This study suggests that to avoid significant changes in ICP and CSF volumes, the distal catheters should not be shortened.



### OP034

#### Cognitive profile and impact of the cyst on daily life in 100 children with a temporosylvian arachnoid cyst : the cyst shape has more impact than its size

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**Objective:** The authors' aim was to study the cognitive profile of children with a temporal arachnoid cyst (TAC) and its impact on daily life in relation to age and cyst characteristics, to facilitate surgical decision-making.

**Methods:** Authors prospectively analyzed the cognitive and psychological profiles of 100 consecutive children relative to age (3-16y) and cyst characteristics (side, cyst-size, and cyst-shape: convex/non-convex), and their outcome 4 years later.

**Results:** Children had varied profiles: 50% had no difficulties; 50% needed rehabilitation for learning difficulties; 26% had academic difficulties. Impairments were observed in language (31%), memory (25%), executive function (21%) and attention (24%); 29% had at least one impaired Wechsler index, particularly in speed and working-memory. One-third required psychotherapy, especially with a right-cyst and older children. Cyst size had very little neuropsychological impact. Convex-cysts were significantly associated with worse cognitive performance and more behavioral difficulties and psychotherapy than non-convex. Children with a ruptured or incidentally discovered cyst had a good cognitive level. Parents of non-operated children with neuropsychological disorders still reported difficulties four years later.

**Conclusion:** The profile of children with a TAC is variable. Some have no issues, especially in case of incidental discovery, and simply require medical follow-up; others may have relevant somatic or neuropsychological disorders, for which surgery may be considered.

#### OP035

##### Postoperative neuropsychological and clinical improvement in children with a temporosylvian arachnoid cyst

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**Objective:** The authors' aim was to study clinical, images, and neuropsychological changes in children who underwent surgery for a temporal arachnoid cyst (TAC).

**Methods:** Thirty-four children (age 3-16y) were prospectively assessed at diagnosis and postoperatively (mean 14 months) with clinic visits, images, cognitive tests, and parental questionnaires on daily-life. Parents fulfilled an outcome questionnaire 4 years postoperatively.

**Results:** The 34 children selected for surgery had signs of raised intracranial pressure (74%) and/or neuropsychological difficulties in learning (65%), cognitive functions (56%), and mood/behavior (47%). The majority had a convex cyst (85%) and underwent microsurgical fenestration (85%). TAC volume decreased  $\geq 50\%$  for 59%. The cohort significantly improved in IQs, language, working memory, episodic memory, and executive functions. Improvements were higher with a preoperative heterogeneous profile with isolated lower scores, and a left cyst. Parental questionnaires showed reduction in anxiety, aggressivity, social problems, and executive disorders in daily life. Individually, 76% of children improved, whatever the age. Four years later,

97% of parents described child's improvement, correlated with the postoperative cognitive improvements.

**Conclusion:** Some children with a TAC have no difficulties; others may show signs of raised intracranial pressure or specific neuropsychological disorders that impact daily life and require long-lasting rehabilitation. In these cases, surgery may be helpful: 76% of our cohort improved, regardless of the child's age, particularly in patients with selective disorders. We propose a surgical decision-making algorithm for TAC.

#### Abstract Session 6: Posterior Fossa Oncology I

##### OP036

##### The clinical course and characteristics of pediatric medulloblastoma in the posterior fossa: A database study

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**Introduction:** Pediatric medulloblastomas specific to the posterior fossa are rare tumors, with their clinical characterization yet to be completely profiled. Correspondingly, this study aimed to clinically characterize these tumors, and elucidate specific clinical predictors of survival from a large national database.

**Methods:** All relevant tumors in patients aged  $\leq 18$  year in the U.S. National Cancer Database (NCDB) between 2005-2016 were retrospectively reviewed. Data were summarized and overall survival (OS) was modeled using Kaplan-Meier and Cox regression analyses.

**Results:** A total of 844 cases of pediatric medulloblastoma in posterior fossa were identified. Overall, mean age at diagnosis was 7.7 years (range, 1-18) with there being 296 (35%) females and 548 (65%) males. All tumors showed undifferentiated histology on microscopic exam and deemed high grade. In terms of treatment, surgery was pursued in 818 (97%) patients, chemotherapy in 689 (82%) patients, and radiation therapy in 631 (75%) patients. Five-year overall survival was estimated to be 74% (95% CI, 70-78%). Multivariate regression revealed older age (HR 0.95,  $P < 0.01$ ), and surgery treatment (HR 0.29,  $P < 0.01$ ) both significantly and independently predicted longer overall survival. Within surgery treatment, gross total resection demonstrated statistical advantage over subtotal resection and biopsy in terms of longer overall survival (HR 0.67,  $P < 0.01$ ). Chemotherapy, radiation therapy and socioeconomic parameters did not demonstrate statistically significant association with overall survival.

**Conclusions:** Pediatric medulloblastomas specific to the posterior fossa are rare, and approximately three-quarters of patients will survive at five years after diagnosis. Older age and surgical treatment are both independently prognostic of longer overall survival. There remains a clear survival benefit in pursuing maximal safe resection in these patients. As such, judicious management including upfront surgical treatment by an expert multi-disciplinary team should be considered the standard of care for these tumors in children.

##### OP037

##### National multicentered retrospective review of clinical and intra-operative features associated with the development of cerebellar mutism after pediatric posterior fossa tumour resection

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**Background:** Cerebellar mutism (CM) is a condition characterized by a significant lack or loss of speech in children following posterior fossa (PF) surgery. The biological origin of CM remains largely unclear and remains the subject of ongoing debate. The more important observation is however, that despite multidisciplinary interventions, the outcome is less favorable than previously described.

**Methods:** A national multi-centered retrospective review of all the children undergoing PF resection in 4 mid-sized Canadian academic pediatric institutions was undertaken. Patient, tumour, and surgical factors associated with the post-operative development of CM was reviewed. The study retrospectively identified patients developing CM with an internal control population consisting of patients who did not develop post-operative CM.

**Results:** 258 patients across the 4 centers over a 10-year period (2010–2020) were identified. The average age of the cohort was 6.73 years and 42.2% of children were female. Frozen section was available in 90.3% of cases. Most of the final tumour histology in children developing CM was medulloblastoma (35.7%), pilocytic astrocytoma (32.6%), ependymoma (17.1%) and exophytic glioma (1.2%). Intraoperative impression of adherence to the floor of the 4th ventricle was negative in 47.7%, positive in 36.8% of cases. Intraoperative surgeon's assessment as gross total resection occurred in 69.8% of cases. Intraoperative abrupt changes in blood pressure and/or heart rate was identified in 19.4% and 17.8% of cases. CM was experienced in 19.5% of patients (N=50). The clinical resolution of CM as mainly assessed by a neurosurgeon (86%) and was complete, significant resolution, slight improvement, no improvement, and deterioration in 56.0%, 8.0%, 20.0%, 14.0%, 2.0% respectively.

**Conclusion:** As a devastating surgical complication, identifying and understanding the biological origin of CM is the first step to complication avoidance. Maximal safe resection irrespective of intraoperative pathology remains the goal to avoid the devastating complication of CM.

#### OP038

**A cerebral blood flow decrease in the left supplementary motor areas: New insight into postoperative pediatric cerebellar mutism syndrome using arterial spin labelling perfusion MRI**

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**Introduction:** Postoperative pediatric cerebellar mutism syndrome (pCMS) is a complication that may occur after posterior fossa tumors resection, in particular medulloblastoma (MB) in children. It is characterized by a transient mutism or a severe reduction of speech occurring in average 48 hours after surgery, associated with emotional lability. Proximal dentato-thalamo-cortical tract injury has been associated with pCMS, but perfusion-MRI abnormalities remain poorly investigated. Thus, our

aim was to investigate postoperative changes in arterial spin labelling (ASL) at rest in children who developed pCMS after MB resection.

**Method:** This study compared preoperative and postoperative T2-weighted MRI signal abnormalities and ASL at rest measuring cerebral blood flow (CBF) and using a whole-brain analysis in 27 children who had undergone MB resection, including 11 patients who developed mutism (mean age 7.0 years) and 16 who did not (mean age 8.9 years). Analysis were conducted with Statistical Parametric Mapping.

**Results:** In patients with pCMS, comparison of preoperative and postoperative CBF showed a significant postoperative decrease in CBF in the left pre-supplementary motor area (pre-SMA) (p=0.007) and left supplementary motor area (SMA) (p=0.009). In patients who did not develop pCMS, no significant differences were observed between preoperative and postoperative CBF. Analysis of T2-weighted signal abnormalities showed that patient with pCMS were significantly more likely to present with T2-weighted hyperintensities in the right dentate nucleus (DN) (p=0.02) postoperatively than those who did not.

**Conclusion:** This study provides the first evidence of an association between pCMS, damage to the right DN and left pre-SMA and SMA hypoperfusion. Evidence of post-operative left pre-SMA/SMA hypoperfusion, key areas involved in speech production, provides a new step towards a better understanding of the brain mechanisms involved in pCMS with mutism. Resting state fMRI studies is needed to describe functional connectivity pattern in this puzzling and intriguing syndrome.

#### OP039

**Place of neo/adjuvant chemotherapy and limited surgery in the treatment of non-metastatic childhood medulloblastoma**

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**Introduction:** Current treatment of childhood medulloblastoma is based on maximal surgery, followed by chemotherapy and radiotherapy. However maximal resection might induce neurological complications, especially posterior fossa syndrome. We aimed to review cases of non-metastatic childhood medulloblastoma treated by partial resection followed by treatment based on response to neo/adjuvant chemotherapy.

**Methods:** We retrospectively included patients less than 18 years old treated for a non-metastatic medulloblastoma between 2005 and 2020 with biopsy or partial resection, followed by neo/adjuvant chemotherapy (carboplatin and etoposide). A second look surgery was discussed after 2 courses of chemotherapy and the oncological treatment was based on the achievement of complete response. Clinical, radiological, histopathological data and outcome were collected.

**Results:** Ten patients (median age = 9.1; 5.2 - 17) were included; median follow-up was 5.3 years (2.7 - 15.8). Hydrocephalus was treated before initial tumor surgery in 6/10 cases (endoscopic third ventriculostomy=4; ventriculo-peritoneal shunt=2). Histological diagnosis was obtained by opened biopsy (n=2) or partial tumor resection (n=8). No post-operative complication was observed. The histological subtype was classic in 6 cases and desmoplastic in 4. The molecular subgroup was SHH in 2 cases, group 3 in 4 and group 4 in 1 (3 missing data). After neo/adjuvant chemotherapy, 3 complete responses were achieved, and disease was stable or in partial response in 7 cases. Among patients with residual disease, 6/7 had a second look surgery, with a complete resection in 3. One patient presented a cerebellar mutism after the

second surgery. The treatment was continued according to standard-risk in 6 cases and to high-risk in 4. At last follow-up, 6 patients are alive in complete response (4 in standard-risk, 2 in high-risk).

**Conclusion:** Limited surgery associated with neo/adjuvant chemotherapy may be an efficient strategy in the treatment of non-metastatic childhood medulloblastoma and may limit the long-term morbidity in survivors.

#### OP040

##### The site of origin of medulloblastoma: does the neurosurgical perspective support the current concept from molecular data?

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**Introduction:** Medulloblastoma (MB) is one of the most common malignant brain tumors in childhood.(Northcott et al., 2019) Developmental gene expression data supported by retrospective MR imaging studies suggest that WNT-MB originate from the region of the embryonic lower rhombic lip (LRL), whereas SHH-MB and non-WNT/non-SHH MB (Group 3\_4) arise from cerebellar precursor matrix regions. (Gibson et al., 2010; Hendrikse et al., 2022) This study aims to analyze detailed intraoperative data specifically with regard to tumor growth pattern and compare these with preoperative imaging and with reference to molecular grouping with the hypothesized regions of origin.

**Methods:** Clinical and molecular data from 58 pediatric patients operated on a histologically confirmed MB at our Department of Neurosurgery between 1990 and 2020 were retrospectively analyzed. The site of origin (STO) was assessed by two experienced neuroradiologists based on standard preoperative MR examinations and compared to the intraoperative findings of an expert neurosurgeon (surgical videos, detailed operative reports), all blinded to the molecular group allocation.

**Results:** Compared to the intraoperative findings, the imaging-based STO prediction was correct in 0% of WNT-MB, 43% of SHH-MB and 100% of non-WNT/non-SHH MB. Using the intraoperatively defined STO as the standard of reference, correct prediction of molecular groups was feasible in 30% of WNT-MB, 60% of SHH-MB and 71% of non-WNT/non-SHH MB.

**Conclusion:** Our findings challenge the current concept of the group-specific origins based on developmental gene expression data. In our series of 58 cases, neither neuroradiological review nor intraoperative assessment of the STO could reliably predict the molecular group. We conclude that for now, maximal safe gross-total resection should remain the aim of surgery irrespective of the observed STO.

#### OP041

##### The use of sodium fluorescein and YELLOW 560 nm filter for the resection of pediatric posterior fossa lesions

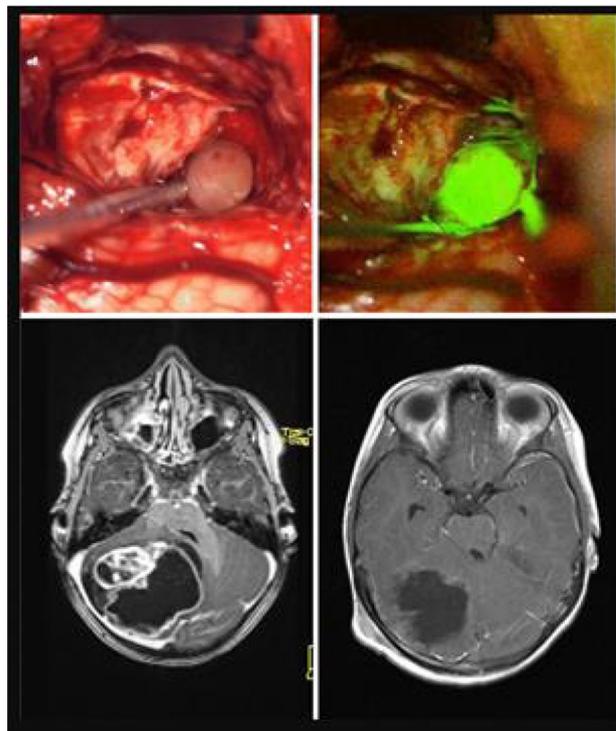
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**Purpose:** This study aimed to verify the feasibility, safety, and benefit of using fluorescein sodium (FL) and a YELLOW 560 nm filter in posterior fossa tumors in children.

**Methods:** All cases of pediatric posterior fossa tumors that have undergone surgery using fluorescein (2018–2022) have been included and were examined retrospectively. In those cases where resection of the tumor was planned, a blinded neuroradiologist distinguished gross total resection and subtotal resection according to the postoperative MRI findings. The surgical report and medical files were reviewed regarding the intraoperative staining grade and adverse events. The grade of fluorescent staining of the targeted lesion was assessed as described in the surgical reports. The screening was conducted for any reference to the degree of fluorescent staining: “intense,” “medium,” “slight,” and “no staining”.

**Results:** 19 cases have been included. In 14 cases, a complete resection was initially intended. In 11 of these cases, a gross total resection could be achieved (78.6%). Staining was described as intense in most cases (58.8%). Except for yellow-colored urine, no side effects obviously related to FL were found throughout the observation period.

**Conclusion:** In combination with a specific filter, FL is a reliable, safe, and feasible tool in posterior fossa surgery in children.



#### OP042

##### The role of reoperation in paediatric cerebellar pilocytic astrocytoma

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**Introduction:** Cerebellar pilocytic astrocytomas (cPAs) in childhood is associated with good prognosis after total resection but the final outcome after incomplete surgery remains largely unpredictable.

**Methods/Results:** We retrospectively identified 202 paediatric cPA. The majority of patients were between 1-10 years of age (72.8%) without gender predominance (M:F=101:101), usually presenting with 1 lesion (N=195, 96.5%) and rarely with disseminated disease at diagnosis

(3.4%). The majority of initial resective surgeries achieved GTR (76.1%). In patients with

**Conclusion:** Residual disease in cPA should be maintained under clinoradiological surveillance post-operatively with a more conservative approach when residual disease is not significantly changing over time.

#### OP043

##### Cerebellopontine angle tumours in the paediatric population – An original series of 43 patients

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**Introduction:** Posterior fossa tumours account for almost 40 to 50% of brain tumours in children, but less than 10 % are in the cerebellopontine angle (CPA). We aimed to report characteristics and surgical management of CPA tumours in a referral centre.

**Methods:** Patients less than 18 years old treated for a CPA tumour from 2010 to 2020 at Necker-Enfants Malades Hospital were retrospectively included. CPA tumours were defined as tumours entirely or predominantly developed in the CPA. Clinical, radiological, histological, and surgical data were collected. Statistics were performed with BiostaTGV.

**Results:** Forty-three patients were included (median age = 6.9 years; 0.03 - 16.5). Most frequent symptoms leading to diagnosis were facial nerve palsy (25%), hearing loss (25%), and intracranial hypertension (18.41%). Hydrocephalus was observed in 20 cases (46.5%) and treated before tumour resection in 14/20 cases (shunt, endoscopic third ventriculostomy, cyst puncture or debulking in emergency). Tumour resection was performed in 37/43 cases (partial = 23; complete = 14). Infiltration of cranial nerves or the brainstem was the main reason for limited tumour resection. The tumour was malignant in 20/37 cases. Most frequent histologies were ATRT (n=6), neurinoma (n=6) and pilocytic astrocytoma (n=5). Patients with a benign tumour were significantly older than those with a malignant tumour: 9.1 years vs 4.7 years, respectively (p=0.007). After surgery, improvement or stability of cranial nerve palsy was observed in 14/37 cases and permanent worsening in 2 cases. Most common long-term sequelae were facial palsy, hearing loss, and swallowing disorders.

**Conclusion:** CPA tumours are a rare entity in children with diverse histologies. Their location makes their resection complex. The surgical strategy must consider the age of patient, the existence of hydrocephalus at diagnosis and the histology. Monitoring of the cranial nerves should become systematic to limit complications and to optimise tumour resection.

#### OP044

##### Cerebellopontine angle tumors in children

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**Introduction:** Cerebellopontine angle (CPA) tumors represent 5-10% of intracranial tumors in adulthood and only for 1-3% in children. Subsequently, only a few pediatric series have been reported in the literature so far. Herein, we describe the experience of a single Institution on CPA tumors in children.

**Materials and Methods:** Children undergoing surgery for CPA tumors between January 2007 and December 2017 (minimum follow-up: 5 years) were reviewed by analyzing demographics, clinical presentation, surgical procedures, approaches, extent of resection and survival. Children with high-grade tumors underwent adjuvant treatments according to the disease and risk stratification-related protocols.

**Results:** Overall, 42 children with 45 lesions were included (3 patients had bilateral tumor), accounting for 4% of all surgically treated intracranial lesions in the same period. The mean age at diagnosis was 6.9 years (range: 3 months-18 years). Retro-sigmoid approach was utilized in 23 cases and midline suboccipital approach in the remaining 19. Histological analysis pointed out: ependymoma in 31% of cases (13 patients), medulloblastoma in 17% (7), AT/RT and astrocytoma in 14% each (6), schwannoma in 10% (4), chordoma in 5% (2), epidermoid cyst, primitive neuroectodermal tumor, ganglioglioma and glioblastoma in 2%, respectively (1). A gross total resection was obtained in 22/45 tumors (49%), a subtotal resection in 13 (29%) and a partial resection in 10 (22%). Repeated surgery for recurrence was performed in 11 patients. No surgical mortality was detected; permanent complications occurred in 14% of cases (cranial nerve palsy, dysmetria, hemiparesis, hydrocephalus). After a 7.2-year mean follow-up, 66% of patients are alive (28 cases).

**Conclusions:** CPA lesions are uncommon but not rare in the pediatric population. Although some of them may not offer significant surgical difficulties, often they require a specific surgical experience because of the specific characteristics of the CPA region in children, thus resulting in a real surgical challenge.

#### Abstract Session 7: Posterior Fossa Oncology I / Epilepsy

#### OP045

##### Intraoperative high field 3Tesla magnetic resonance imaging in pediatric epilepsy and tumor neurosurgery

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**Objective:** Intraoperative high field MR 3 Tesla magnetic resonance imaging (iopMRI) is an elaborate task, especially when used in children. On the other hand, advantages on prognosis may be significant, especially in pediatric epilepsy and tumor surgery.

**Methods:** Thus, we searched our intraoperative MRI database to identify all children operated at the Neurosurgical Department, Vienna Medical University since implementation in October 2020.

**Results:** Altogether, 73 children in 86 surgeries were operated on using the 3 Tesla intraoperative MRI suite. Of those, 39 epilepsy surgeries (patients median age 6, 0-16) and 34 tumor surgeries (patients mean age 8, 0-18) were performed. The epilepsy surgery cases consisted of 18 resections, 12 disconnections, 9 depths electrode implantations and 10 LITT procedures. For the tumor cases, 33 resections, 2 stereotactic biopsies and 2 open biopsies were performed in the iop MRI suite. Intraoperative assessment of the extension of lesional epilepsy (FCDs, LEATS, Cavernomas) and identification of tumor borders were feasible in every case. Due to the intraoperative MR imaging results, second look surgeries were performed in 20-30% of the cases, dependent on the specific diagnosis. Highest numbers of second look surgeries were found in diffuse gliomas and in FCDs. Additionally, intraoperative diffusion tensor imaging (DTI) was performed in disconnective epilepsy surgeries for

evaluating the target fibre disconnection. No adverse effects like intraoperative anaesthesia instability or postoperative infection occurred.

**Conclusion:** Intraoperative high field MRI in pediatric neurosurgery (especially in epilepsy and tumor surgery) seems to be beneficial at least in increasing extent of resection in 20–30% of the cases and may thus contribute to improve the outcome in pediatric tumor and epilepsy cases.

#### OP046

##### Lesional epilepsy surgical management inside the epilepsy surgery program at IRCCS Istituto Giannina Gaslini

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**Introduction:** The predominant seizure type in all ages is focal seizure, frequently associated with structural lesions. More than 30% of epilepsy patients may become drug-resistant, especially for some specific lesion type as focal cortical dysplasia type II, hippocampal sclerosis and LEATs. Epilepsy surgery is a fundamental treatment option, frequently associated with a favorable outcome, especially in cases of short epilepsy duration (Barba et al. *Epilepsia*, 2021). We present our institutional series, illustrating the management of lesional epilepsy surgery at our institution

**Methods:** The institutional epilepsy surgery database, updated in collaboration with the Child Neuropsychiatry Unit was retrospectively reviewed to identify the cases of epilepsy linked to the presence of a structural lesion on anatomical MRI sequences in which the same alteration were judged to be linked to clinical epileptic manifestation and EEG epileptic discharges. Cases treated with laser ablation were excluded.

**Results:** 73 cases meeting the selection criteria were identified from 2014 to 2021. Other 10 cases of lesional epilepsy were operated in 2022. Preclinical data were reported: history, frequency and semiology of seizures, neuroradiological diagnosis. The lesions linked to the epileptic manifestation were composed principally by different low grade neoplastic lesions (DNET, GG and LGG), followed by FCD IIb and other FCDs, vascular malformations and other non neoplastic lesions. Seizure outcome has been classified in Engel I or II in 55 cases (2014–2021).

Further, we present the cases in which advanced anatomical MRI imaging such as Voxel-based modeling of gray-white matter blurring and gray matter intensity increased sensitivity for the detection of alterations, most commonly FCD.

**Conclusion:** Anatomical-electrical-clinical concordance is one of the most important predictor of seizure outcome after epilepsy surgery. Advances in structural MRI analysis could help defining an epilepsy as lesional and designing the most correct surgical plan.

#### OP047

##### MRI-guided Laser Interstitial Thermal Therapy for Hypothalamic Hamartoma – A Norwegian Experience

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**Introduction:** Hypothalamic hamartoma (HH) is a rare, congenital, tumour-like growth in hypothalamus. It may be associated with precocious puberty, epilepsy with gelastic or dacrystic seizures and neurobehavioral symptoms. Depending on the anatomical location and morphology on the magnet resonance imaging (MRI), it may

be classified in four different types (Delalande and Fohlen, 2003). The treatment options for HH-associated epilepsy may include open surgery, endoscopic disconnection, radiofrequency ablation or radiosurgery, with varying degrees of treatment-associated morbidity. A novel treatment option, MRI-guided laser interstitial thermal therapy (MRgLITT), is aimed to minimize postsurgical complications and optimize the destruction of the pathological tissue. It was recently added to the surgical armamentarium in the treatment of pharmacoresistant epilepsy in Norway.

**Methods:** Clinical and radiological data of 5 patients with HH who underwent MRgLITT for pharmacoresistant epilepsy were prospectively collected. The placement of laser catheter was performed using frameless system (Varioguide) with intraoperative CT autoregistration and postoperative verification of catheter at the target.

**Results:** The median age at the time of treatment was 11 years (range, 4–35 years). Two patients had previous surgery, one underwent a partial endoscopic HH disconnection and the other an MRgLITT at another center. No surgical complications occurred during MRgLITT, however, all patients experienced transient electrolyte disturbances. Three patients were completely seizure free (two at 2-year and one at 6-month follow-up), and two patients had Engel class IIIA outcome (Engel et al. 1993).

**Conclusion:** In patients with pharmacoresistant epilepsy and HH, MRgLITT showed promising postsurgical results with low risk of complications.

#### OP048

##### Accuracy of Robot-Assisted Stereotactic Laser Ablation in Children with Epilepsy

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**Introduction:** Robot-assisted (RA) stereotactic laser ablation has been reported as a safe and effective technique for the treatment of epileptogenic foci in children and adults. This study aimed to assess the accuracy of RA stereotactic laser fiber placement in children, and to identify factors that might increase the risk of misplacement.

**Methods:** A retrospective single institution review of all children from 2019 to 2022 who underwent RA stereotactic laser ablation for epilepsy was undertaken. Placement error was calculated at the target by calculating the Euclidean distance between the implanted laser fiber and pre-operative plan. Age at surgery, date of robot calibration, entry angle, extracranial soft-tissue thickness, bone thickness, and intracranial length were collected. A systematic review of the literature was also performed using Ovid Medline, Ovid Embase, and Cochrane Central Register of Controlled Trials (CENTRAL).

**Results:** Twenty-eight children who underwent RA stereotactic placement of a total of 35 laser fibers were identified. Twenty (71.4%) had undergone ablation for hypothalamic hamartoma, seven (25.0%) for presumed insular focal cortical dysplasia and one (3.6%) for periventricular nodular heterotopia. Nineteen children were male (67.9%) and nine were female (32.1%). Median age at the time of the procedure was 7.67 years (IQR 4.58 – 12.26 years). Median target point localization error (TPLE) was 1.27 mm (IQR 0.76 – 1.71). Median offset error between the planned and actual trajectories was 1.04 degrees (IQR 0.73 – 1.46). Age, gender, time interval between date of surgery and robot calibration, entry angle, soft-tissue thickness, bone thickness, and intracranial length, were not associated with accuracy. There were no immediate surgical complications. From meta-analysis, overall pooled mean TPLE was 1.46 mm (95% CI: -0.58; 3.49).

**Conclusions:** RA stereotactic laser ablation for epilepsy in children is highly accurate. These data will aid surgical planning.

#### OP049

##### **Magnetic Resonance-Guided Laser Interstitial Thermal Therapy (MR-gLiTT) of different epileptogenic lesions: report of fifteen pediatric cases**

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**Introduction:** We tested the efficacy and safety of Magnetic Resonance-guided Laser Interstitial Thermal Therapy (MR-gLiTT) in a series of different epilepsy-determining brain lesions. The aim is to investigate seizure outcome and complications rate.

**Methods:** We describe a series of 15 children (mean age at MRg-LiTT was 7 years 33 months) suffering from epilepsy who underwent MRg-LiTT at our Centre between July 2020 and December 2022.

**Results:** All patients suffered from drug resistant epilepsy (DRE). The mean number of anti-seizure medications (ASMs) before MRg-LiTT was 3.8. Etiology was: HH–9 patients, TS–1 patient, DNET- patient, Focal Cortical Dysplasia–3 patients, Rasmussen Encephalitis (1 patient, who underwent hemispheric deconnection). The total number of lesions treated was 16 (patient affected by TS had 2 tubers). The total number of MRg-LiTT procedures is 18. Three of them underwent previous stereoEEG monitoring followed by standard resective surgery or SEEG-guided Rf-TC at other institution. Following seizure recrudescence, MRg-LiTT approach was indicated. One patient underwent two previous lesionectomies to treat a DNET at our Hospital. One patient underwent three previous endoscopic resections for HH at a third Center. One patient underwent previous left hemispheric deconnection to treat DRE in Rasmussen Encephalitis.

In this observational study, 4 patients suffered post-surgical complications, two of them suffered transitory complications (extradural hematoma and transitory worsening of pre-existent left arm weakness), two of them persistent complications (diabetes insipidus after third MRg-LiTT procedure and hypocorticosurrealism which needs medical correction).

The mean follow up is 12 months. Postoperative seizure outcome (Engel Class), ranged from class I to class IV. At last follow up, 9/15 (60%) patients are in Engel Class I and 12/15 (80%) patients experienced a reduction of seizures from baseline (Class I + Class II).

**Conclusion:** Our experience helps to confirm that MR-gLiTT is a highly safe and effective procedure for several epilepsy conditions in children.

#### OP050

##### **Factors influencing seizure outcome after ganglioglioma surgery in pediatric patients**

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**Background:** Gangliogliomas are glioneural tumors causing refractory epilepsy, often early in life. Neurosurgical resection can control or even cure seizures. However, postoperative recurrence of the epileptic seizures can occur. We therefore aimed to analyze factors influencing seizure outcomes after ganglioglioma removal.

**Methods:** We performed a retrospective analysis of pediatric patients with ganglioglioma and epileptic seizures treated at our center. Seizure outcomes are reported according to the ILAE classification. BRAF-V600E mutation status was detected immunohistochemically.

**Results:** Forty-one patients, mean age 10.2 (5.5) years, were included in the analysis and had a mean follow-up time of 6.8 (6.5) years. One year after surgery, 28 patients (68.3%) had an outcome of ILAE class

I. Residual tumor tissue was suspected in 15 cases and a second operation was performed in 11 patients, which led to 35 patients (85.4%) being seizure free at the last follow-up. Older age and longer duration of epilepsy showed increased risk for seizure relapses ( $p=0.017$  and  $p=0.043$  respectively). A BRAF-V600E mutation could be detected in 24 specimen (58.5%). Patients with BRAF mutations had higher seizure frequency preoperatively (4.6 vs. 3.9 per month,  $p=0.048$ ) and shorter average time until seizure relapses (1.2 vs. 3.4 years), but the latter finding was not significant due to high variance.

**Conclusion:** Resection of gangliogliomas can provide seizure control in a high percentage of patients. Early surgery appears to confer a benefit for patients. BRAF mutation seems to increase the epileptogenicity of gangliogliomas and should be studied as a possible target for treatment.

#### Abstract Session 8: Epilepsy

#### OP051

##### **Towards a better definition of a resection strategy in pediatric LEATs causing refractory epilepsy**

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**Introduction:** Resection strategy in pediatric long-term epilepsy associated tumors (LEATS) consists of pure lesionectomy, ECoG guided tailored resection or even partial/complete lobectomy. A retrospective data analysis was performed to clarify the most appropriate strategy.

**Methods:** Twenty-two children suffering from medically intractable seizures harbouring suspected LEATs were retrospectively investigated based on video EEG monitoring, extended MRI and FDG and Methionine (MET) PET data as well as resection strategies based on ECoG, neuronavigation and iopMRI. All children (mean age 8 yrs, from 2-18) were consecutively resected during a 3 years period.

**Results:** In temporal LEATS, mainly antero-temporal resections or temporal lobectomies were performed (15 patients), whereas in extratemporal LEATS lesionectomies or tailored resections were achieved (7 patients). Histological diagnosis was GG in 20 and DNET in 2 patients. Preoperative MRI contrast was present in 10 GG (45%) and FDG PET showed a hypo-metabolic area in 6 GG (27%). Intensive Methionine (MET) PET uptake was found in 13 GG, weak MET enhancement in 6 GG and no tracer uptake in 1 GG. One DNET showed MET PET uptake, the second one did not. However, MET PET uptake did not correlate neither with MR contrast, nor with extent of the tumor defined by FLAIR. In temporal resections, ILAE Class 1 seizure outcome was achieved in 75%, improved to ILAE Class 1 in 94% by performing 6 repeat surgeries with antero-temporal lobectomies. The extratemporal patients experienced ILAE Class 1 seizure outcome in 86%. (mean FU 28 months).

**Conclusion:** In childhood LEATs amino acid PET was found to have high diagnostic sensitivity for GGs. For extratemporal LEATs a pure lesionectomy or tailored resection may be an appropriate strategy, for temporally located LEATs an antero-temporal resection or even temporal lobectomy may be necessary to achieve seizure freedom and avoid recurrences.

#### OP052

##### **Preoperative Detection of Subtle Focal Cortical Dysplasia in Children by Combined ASL, VBM, EEG-fMRI, Resting-State Regional Homogeneity, and FDG-PET**

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**Introduction:** Focal cortical dysplasia (FCD) causes drug-resistant epilepsy in children that can be cured surgically but the lesions are often unseen by imaging. We aimed to assess the efficiency of arterial spin labeling (ASL), voxel-based morphometry (VBM), fMRI-EEG, resting-state regional homogeneity (ReHo), FDGPET, and their combination in detecting pediatric FCD.

**Methods:** We prospectively included 10 children for whom FCD was localized by surgical resection. They underwent 3T MR acquisition with concurrent EEG, including ASL perfusion, resting-state BOLD fMRI (allowing the processing of EEG-fMRI and ReHo), 3D T1-weighted images processed using VBM, and FDG PET-CT co-registered with MRI. Detection was assessed visually and by comparison with healthy controls (for ASL and VBM).

**Results:** Eight children had normal MRI and two had asymmetric sulci. Using MR techniques, FCD was accurately detected by ASL for 6/10, VBM for 5/10, EEG-fMRI for 5/8 (excluding 2 with uninterpretable results), and ReHo for 4/10 patients. The combination of ASL, VBM, and ReHo allowed correct FCD detection for 9/10 patients. FDG PET alone showed higher accuracy than the other techniques (7/9) and its combination with VBM allowed correct FCD detection for 8/9 patients. The detection efficiency was better for patients with asymmetric sulci (2/2 for all techniques), but advanced MR techniques and PET were useful for MR-negative patients (7/8).

**Conclusion:** A combination of multiple imaging techniques, including PET, ASL, and VBM analysis of T1-weighted images, is effective in detecting subtle FCD in children.

#### OP053

##### **Review of the targeting accuracy of robot-assisted stereo electroencephalography (SEEG) electrode implantation in pediatric epilepsy patients with use of Renishaw 3D neurolocate module**

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**Introduction:** Renishaw 3D neurolocate module is a frameless patient registration module that is designed for use with the Renishaw neuro-mate stereotactic robot. When noninvasive modalities fail to adequately localize the seizure onset zone (SOZ) in children with medically refractory epilepsy, invasive interrogation with stereo-electroencephalography (SEEG) may be required. This study aims to provide a review of targeting accuracy of SEEG electrode implantation, in a single centre, comparing the use of standard frame-based techniques versus the Renishaw 3D neurolocate module.

**Methods:** Eighteen (18) patients underwent implantation of SEEG electrodes under general anaesthesia during the period August 2019 through November 2022 at our center. All patients underwent robotic-assisted stereotactic implantation of the SEEG electrodes with intraoperative 3D scanner confirmation of the final electrode position. These coordinates were compared to the planned entry and target; with attention to depth, radial, directional and absolute errors, in addition to Euclidean distance.

**Results:** Of the 235 electrode implantations undertaken, 109 employed Renishaw 3D neurolocate technology. The mean age was 11.0 years (range from 4–18 years), of which 75% were male. The median absolute errors in x-, y-, z- axes were 0.45mm, 0.7mm and 0.8mm respectively with use of the Renishaw 3D neurolocate versus 0.4mm, 0.8mm and 1.3mm with standard techniques. The median Euclidean distance from the planned target to the actual electrode position with Renishaw 3D neurolocate module was 1.34 mm vs standard frame-based techniques at 1.70mm. No major perioperative complications occurred.

**Conclusion:** Stereo-electroencephalography (SEEG) electrode implantation with the Renishaw 3D neurolocate module is safe for use in the pediatric population with good surgical accuracy. The Renishaw 3D neurolocate module for SEEGs surgeries allows compatibility with the intraoperative 3D scanner and has the potential to improve surgical targeting accuracy and patient comfort.

#### OP054

##### **Short and Long-Term Outcomes after Resective Epilepsy Surgery in Pediatric Temporal Lobe Epilepsy**

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**Objective:** Resective surgery has been established as a curative treatment in pharmaco-resistant pediatric epilepsy. However, the amount of resection remains an individual decision, where possible impairments of cognitive development are weighed against the chance of seizure freedom. We therefore aimed to evaluate which factors influence the seizure outcome and relapse rates after different types of resective temporal lobe surgeries in a pediatric cohort.

**Methods:** We performed a retrospective analysis of our pediatric patient cohort that had undergone resective temporal lobe surgery for pharmaco-resistant epilepsy with a minimum follow up of one year. Surgery type, histological findings, and seizure outcome according to the ILAE classification were extracted from patient records. Surgeries were stratified into lesionectomy, anterior temporal lobectomy (TLR) and selective amygdalohippocampectomies (SAHE).

**Results:** In total, 97 pediatric patients were operated between 1993 and 2019 with 34 lesionectomies, 50 temporal lobectomies and 13 SAHEs. The most frequently diagnosed pathologies were tumors (45.4%) followed by hippocampal sclerosis (37.1%). Follow up ranged from 1.5 to 28 years, median 10.1 years. One year after surgery, 68 patients (70.1%) were seizure free corresponding to the ILAE outcome class 1a or 1. This outcome was lower in SAHE compared to lesionectomies and TLR. At later timepoints no difference between surgical groups was found. Relapses occurred in 25 patients (26.0%) that were initially seizure free. Younger age at surgery and the presence of generalized seizures increased the likelihood of experiencing relapses.

**Conclusions:** Pediatric patients with intractable epilepsy undergoing temporal lobe surgery have good rates of seizure outcome. Surgery type appears to be a modifiable prognostic factor for early seizure freedom but not necessarily for the long-term outcome. Caregivers of younger patients and those with generalized seizures should be counseled about higher chances of seizure relapses after initially successful surgery

#### OP055

##### **Interhemispheric transcallosal hemispherotomy: technique, outcome, pitfalls. A single center retrospective series of 31 cases**

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**Introduction:** In case of hemispheric seizures, hemispheric disconnection is efficient and safe. Lateral perisylvian or vertical parasagittal approaches have been described, and recent studies suggest a better outcome for the vertical technique. Here, we describe a variation of the vertical technique: the interhemispheric transcallosal disconnection. We describe its surgical steps, outcome, and discuss its limitations according to morphometric criteria.

**Methods:** Single center, retrospective study. Clinical electronic files were exhaustively extracted from a clinical data warehouse. MRI were reviewed, to determine if the disconnection was complete. Preoperative morphometric criteria were addressed: anterior and posterior angle of the corpus callosum, ventricular size, coronal and axial thalamic angles, hemispheric diameter. Epileptic outcome was evaluated according to the ILAE classification.

The surgical technique is a midline interhemispheric approach, followed by complete corpus callosum transection, laterothalamic and basifrontal disconnections.

**Results:** From 2013 to 2021, 31 children were operated: 18 males, with a median age of 5.5 years (range 4 months, 16 yo). Etiology was: Hemimegalencephaly (n=10), stroke (n=7), Rasmussen's encephalitis (n=7), multilobar MCD (n=4), Sturge Weber Syndrome (n=2). Radiological disconnection was complete in 25 cases. A second surgery was performed in 6 cases, because of seizure relapse (n=1), or based on MRI findings (n=5). Incomplete disconnection was more frequent in HMG (n=4) and was located around the thalamus (n=3), and the splenium (n=2). Interestingly, a higher coronal and axial thalamic angles were noted in those cases. Main complications were hydrodynamic, requiring a VPshunt in 3 cases. At last follow up, 27 patients were ILAE class I allowing AED withdrawal or major reduction in 20 cases.

**Conclusion:** Interhemispheric midline disconnection is a variant of the vertical paramedian approach that allows good control of the corpus callosum, and comparable seizure outcome. However, lateral extension can be restricted by the thalamic angle, notably in HMG patients.

#### OP056

##### Vertical, Parasagittal Transventricular Hemispherotomy in Pediatric Drug Resistant Epilepsy - Single Center Retrospective Study

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**Objective:** Drug resistant epilepsy in hemispheric pathology represent favorable candidates for epilepsy surgical treatment. For this group of patients, we report our experience with the minimal invasive technique for hemispherotomy in a single center pediatric case series.

**Patients and Methods:** In the time period between 1/2018 until 12/2022 a retrospective review according to our in-house data base was performed to collect clinical data from patients who underwent minimal invasive, AR guided hemispherotomy epilepsy surgery. Indication for surgery was given during an interdisciplinary case discussion in a tertiary pediatric medical center. The peri-operative data was evaluated for surgical time, intensive care unit and hospital stay, possible complications and seizure outcome during follow-up.

**Results:** A total of 32 patients (mean age: 6.6±4.5yrs; range: 0.2-15) received vertical parasagittal transventricular hemispherotomy at our institution with a mean follow up time of 2.1±1.3 yrs. were included in this study. Underlying disease for epilepsy was perinatal ischemia or hemorrhage (n=16), hemispheric dysplasia (n=10); Rasmussen encephalitis (n=4), Sturge-Weber Syndrom (n=2). Mean surgical time was 187±43 minutes (120-297). Median hospital stay was 9 (range: 6-63) days while ICU and IMC stay was 2±2.4 and 1.7±3.5 days, respectively. Six patients needed additional subsequent surgeries including 2 ETV, 3 shunt placements and 1 decubitus revision. New shunt rate was 9.4%. One of these patients had and previous premature IVH and another suffered from a coagulopathy with postoperative hemorrhage. Seizure outcome was generally favorable according to Engel classification (Engel Ia: 84%, Ib: 9.4%, Id: 3.1%, III: 3.1%).

**Conclusions:** Vertical, parasagittal transventricular hemispherotomy is a challenging but feasible and effective technique to beneficially control drug resistant epilepsy in unilateral hemispheric pathologies. The complication rate was experienced to be acceptable with a comparable secondary rate of hydrocephalus development. Future perspective investigations are under way to evaluate neurocognitive development after hemispherotomy.

#### OP057

##### Hippocampal resection during hemispherotomy – is it needed?

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**Introduction:** Hemispherotomy is an effective surgery for pediatric intractable hemispheric epilepsy. Over the years, the surgical goal has shifted from a complete hemispheric resection (anatomical hemispherectomy) to a functional hemispherotomy (FH). Multiple techniques for hemispherotomy have been described, and often hippocampal resection is concurrently performed. The goal of the current study is to assess the role of hippocampal resection in FH.

**Methods:** We retrospectively accrued all clinical data of children (<18 years old) that underwent FH between 2001-2022 at the Tel-Aviv Medical Center and at Texas Children's Hospital. Epilepsy, and surgical outcome was compared based on whether the hippocampus was resected or disconnected at the amygdala and atrial segment of the fornix.

**Results:** A total of 86 patients (32 females) were included. The most common epilepsy etiologies were stroke (31), Rasmussen's encephalitis (16), cortical dysplasia (10), and hemimegalencephaly (9). Mean age at surgery was 74.8 years. The median number of antiseizure medications (ASM) before surgery was 3. Hemispherotomy techniques included peri-insular (54), vertical (23=endoscopic 19 + parasagittal 4), and trans-sylvian (9). Mean follow-up was 41.538 months. Forty-three patients had hippocampal resection and 43 patients had a hippocampal disconnection. Both groups had a mean Engel outcome scale of 1.2 (p=0.85), and there was no significant difference in the functional outcome status of the groups.

**Conclusions:** Functional hemispherotomy is highly effective for pediatric intractable hemispheric epilepsy. Our data suggest that there is no need for hippocampal resection if a complete disconnection is performed.

#### OP058

##### Vagus Nerve Stimulation in Children: Outcomes and Future Directions

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**Background:** Vagus Nerve Stimulation (VNS) is a neuromodulatory procedure most extensively studied as an adjunct to medically refractory epilepsy. Despite widespread adoption and decades of clinical experience, clinical predictors of response to VNS remain unclear.

**Objectives:** To evaluate a retrospective cohort of pediatric patients undergoing VNS at our institution to better understand who may benefit from VNS and identify factors which may predict response to VNS

**Methods:** We conducted a retrospective cohort study examining pediatric patients undergoing VNS over nearly a 20-year span at a single institution. Presurgical evaluation, including demographics, clinical history, and diagnostic EEG and imaging findings were examined. Primary outcomes included VNS response.

**Results:** 297 subjects were studied. Mean age at surgery was 10.1 (SD=4.9, Range=0.8-25.3); length of follow-up was a mean of 4.6 years (SD=3.5, median=3.9 years, range 1 day-16.1 years). There was no association between demographic factors, epilepsy etiology, or genetic basis and VNS outcomes. There was an association between reduction in main seizure type with positive MRI finding. Of all MRI findings analyzed, brain atrophy was significantly associated with worse VNS outcomes, while dysplastic hippocampus and chronic periventricular leukomalacia findings were found to be associated with improved

outcomes. Increased seizure semiology variability and seizure type were also associated with improved seizure outcomes.

**Conclusions:** Predicting response to VNS remains difficult, leading to incompletely realized benefits as well as suboptimal resource utilization. Specific MRI findings and increased seizure semiology variability and type can help guide clinical decision-making and patient counseling.

## Abstract Session 9: Hydrocephalus II

### OP059

#### Changes in CSF Surface Tension in Relation to Surfactant Proteins in Children with Intraventricular Hemorrhage

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**Introduction:** The regulation of surface tension (ST) by surfactants plays an important role in the human respiratory system but is largely unexplored in brain homeostasis. The aim of this study was to evaluate changes in ST in relation to surfactant proteins (SPs) in children with intraventricular hemorrhage (IVH).

**Patients and Methods:** CSF samples from 93 patients were analyzed for ST with a force tensiometer and SP-A-D and -G with ELISA assays. Patients belonged to six groups: (i) IVH before primary intervention (PI), (ii) IVH 4-28 days after PI, (iii) IVH 44-357 days after PI, (iv) hydrocephalus, (v) sepsis and (vi) controls.

**Results:** We found indirect correlations and significant differences in ST and SPs (all  $p < 0.001$ ; except for SP-C,  $p = 0.007$ ). Post hoc analyses showed significantly decreased ST in IVH patients before PI compared with patients with hydrocephalus, sepsis or controls ( $p < 0.001$ ), but it increased in IVH patients over time. All SPs were significantly elevated when comparing IVH patients before PI with controls (all  $p < 0.001$ ; except for SP-C,  $p = 0.003$ ).

**Conclusion:** Children suffering from IVH displayed an increase in SPs and a decrease in ST as coping mechanisms to preserve CSF flow. The increase in ST over time could serve as prognostic marker for the healing process.

### OP060

#### The genetic background of hydrocephalus in a population-based cohort: implication of ciliary involvement

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**Introduction:** The disease mechanisms behind hydrocephalus are complex and not well understood, but some association with dysfunctional cilia in the brain ventricles and subarachnoid space has been indicated. We aimed to investigate potential involvement of the ciliome in hydrocephalus and describe genotype-phenotype associations with autism spectrum disorder (ASD).

**Methods:** A total of 121 hydrocephalus candidate genes were screened in a whole-exome sequenced cohort, comprising 72 hydrocephalus patients and 4,181 background population controls. Candidate genes containing likely pathogenic variants were systematically evaluated for their involvement in ciliary function and ASD.

**Results:** Median age at diagnosis for the hydrocephalus patients: 0 years of age (range 0-27 years), 70.5 % were males. Median age for controls: 18 years (range 11-26 years), 53.3 % were males. Fifty-two likely pathogenic variants in 34 genes, were identified in 42 patients (58.3 %). In hydrocephalus patients, we found increased, but not significant, enrichment of likely pathogenic variants (OR 1.51, 95% CI

0.92-2.51,  $P = 0.096$ ), which was driven by a significant enrichment of rare protein truncating variants (OR 2.71, 95% CI 1.17-5.58,  $P = 0.011$ ). Fourteen of the genes with likely pathogenic variants are part of the ciliome, whereas another six genes affect cilia-dependent processes during neurogenesis. Furthermore, 15/34 genes with likely pathogenic variants, and 3/8 genes with protein truncating variants were associated with ASD.

**Conclusion:** Our results point to the significance of hydrocephalus as a ciliary disease in some cases. Future studies in brain ciliopathies may not only reveal new insights into hydrocephalus, but also, brain disease in the broadest sense, given the essential role of cilia for neurodevelopment.

**Funding:** Lundbeck Foundation Initiative for Integrative Psychiatric Research ([www.iPSYCH.au.dk](http://www.iPSYCH.au.dk)). The Danish National Biobank resource, which is supported by the Novo Nordisk Foundation. Independent Research Fund Denmark (grant no: 1030-00239B)

### OP061

#### Transition of hydrocephalus patients from paediatric to adult neurosurgery: a single institution experience

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**Introduction:** The transition process from paediatric to adult neurosurgery is an essential part of patient's care. This study aims to report our institution's experience and obtain information about this patient subgroup.

**Methods:** This is a single centre retrospective case series study. Data regarding current age, hydrocephalus etiology, number of shunt revisions in childhood and after transition, type of presentation (acute vs routine follow up), type of valve, length of follow up and outcome (including school attendance, highest degree, work and living conditions, mRS, Karnofsky and HUI index) was collected.

**Results:** 227 patients were included (mean 32 years, min 16- max 68 years; SD+/-11.9). 122 were transitioned from the nearest paediatric center, Great Ormond Street Hospital (GOSH); 105 from other national and international hospitals. Common hydrocephalus etiologies were: post haemorrhagic 19.8%, Chiari type 2 12.7%, Aqueduct stenosis 12.7%, congenital 8.81%, post infectious 7% and tumor 5.2%. Hydrocephalus following hemispherectomy, syndromic craniosynostosis or idiopathic intracranial hypertension was rare. In 16.7% (38 cases) no cause could be identified, due to insufficient documentation. 171 (75.3%) presented electively, whilst 37 (16.2%) were referred with suspicion of acute shunt dysfunction without being transitioned. 19 (8.3%) were lost to follow up. Patients presenting acutely were more likely to come from other centers 28/105 (26.7%) compared to 9/122 (7.37%) GOSH ( $p < 0.005$ ; Wilcoxon Signed Rank). 69/227 (30%) patients underwent at least a CSF diversion intervention in adulthood. 16/21 (76%) conservatively followed up cases required surgery in adulthood. Outcome data was collected from 108 patients. Significant sequelae such as headaches and fatigue remain persistent in up to 70% regardless of hydrocephalus etiology with just around a half of the patients being able to have a fully independent life.

**Conclusion:** Hydrocephalus is a condition that requires lifelong neurosurgical follow up. A more structural approach towards transition is needed.

### OP062

#### Jugular foramen stenosis in external hydrocephalus

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**Introduction:** Significant restrictions of dural venous sinuses at the level of the posterior fossa have been repeatedly described and observed in children affected by external hydrocephalus. The reasons for these dural sinuses alterations remains unknown. The anatomy and size of jugular foramina (JF) has never been studied in this group of patients. The object of this study is to measure the size of both jugular foramina in a group of patients and compare these results with a control group.

**Methods:** Studies from 82 patients with diagnosis of external hydrocephalus and 56 controls (CTRL) in whom MR venous (MRV) angiography data were available were selected for the analysis. All MRI studies were performed on the same 1.5 Tesla MRI scanner under pharmacological sedation. Sequences analyzed were axial T2 and sagittal 3D PC-MRA. Jugular foramen (JF) areas, as well as intracranial, subarachnoid space (SAS) and ventricular volumes, were measured. For each patient, the surface areas of right and left jugular foramina were measured on axial MPR reconstructions between the petrous temporal bone and the jugular tubercle of the exocciput of the occipital bone. Venous Obstruction Grading Score (VOGS) was assessed on 3D MIP reconstructions of PC-MRA sequences. Differences between patients and controls in the three venous outflow efficiency measures (minimum and mean of JF areas and venous grading), were tested on the whole dataset by stepwise binary logistic regression.

**Results:** Compared to the CTRL group, VOGS was significantly higher in patients ( $p < 5 \cdot 10^{-8}$ ) as expected, while mean ( $p = 0.019$ ) and minimum ( $p < 0.0005$ ) JF areas were significantly smaller.

**Conclusions:** Jugular foramen stenosis was observed in this group of patients with external hydrocephalus when compared with a control group. A pathophysiological mechanism comparable to that observed for Achondroplasia is postulated to explain the external hydrocephalus and dural venous sinus anomalies observed in these patients.

#### OP063

##### **Predicting endoscopic third ventriculostomy success in pediatric shunt dysfunction: a monocentric retrospective case-series of 70 consecutive children**

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**Introduction:** The role of Endoscopic Third Ventriculostomy in previously shunted children experiencing shunt dysfunction is still discussed in terms of efficacy (success rate from 40 to 80%) and safety (0-32,5% of complications). Reported predictive factors of failure of secondary ETV are age, early onset of hydrocephalus and prematurity. There is no consensus about the best surgical strategy in the different subgroups of patients with shunt dysfunction. We aimed to identify subgroups of shunted patients associated with favorable outcome of ETV and to define the role of ETV in patients with Global Rostral Midbrain Dysfunction Syndrome (GRDMS).

**Methods:** Single center retrospective case series of previously shunted children who underwent secondary ETV for shunt dysfunction between 2012 and 2022. Clinical and MRI features were examined, as well as surgical outcome, etiology of hydrocephalus and preoperative ETV Success Score. Univariate and multivariate analysis were performed to find predictors of outcome of secondary ETV. Youden's index J was calculated on age distribution to find optimal age cut-off.

**Results:** Seventy consecutive patients were included. The overall success rate of secondary ETV was 63%. Primary obstructive hydrocephalus, age > 36 months and the presence of aqueductal obstruction were predictors of ETV success. Multivariable analysis found that age < 36 months, primary inflammatory hydrocephalus, and presence of Isolated Fourth Ventricle were associated with ETV failure. All patients with GRDMS experienced clinical and radiological improvement after ETV.

**Conclusion:** ETV is a safe and effective treatment option for children experiencing shunt dysfunction notably in cases of primary obstructive hydrocephalus with aqueductal stenosis, post inflammatory hydrocephalus and among children aged > 36 months.

#### OP064

##### **Neuroendoscopic Lavage in preterm infants with IVH**

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**Introduction:** Intraventricular hemorrhage (IVH) is a well-known sequelae of preterm infants and associated with a high rate of complication treatment, morbidity and mortality. Over the last few years new treatment concepts like neuroendoscopic lavage have been established. We present our single center experience of multimodal treatment of preterm infants with posthemorrhagic hydrocephalus (PHH).

**Methods:** We present all preterm infants with IVH and PHH treated since 2011 at our University Hospital in Linz. We prospectively recorded data on sex, gestational age, birth weight and as outcome parameters complications and shunt dependency.

**Results:** Forty seven infants were treated surgically. 14 patients received NEL plus ventricular access device (VAD), and 33 patients were treated with a VAD without lavage. Both groups had comparable patients' characteristics. Mean gestational age was  $28 + 4$  ( $23 + 5 - 37 + 6$ ), median birth weight was 1403g ( $564 - 3070$ ).

Shunt dependency after 12 months was 50 % ( $n = 7$ ) in the NEL and 81 % ( $n = 27$ ) in the VAD group. The revision rate in the patients who have been treated with a VP -Shunt due to PHH in the first year was 43 % ( $n = 3$ ) in the NEL and 70 % ( $n = 19$ ) in the VAD group ( $p = 0.02$ ). There was one (6 %) re-bleeding and 2 infections (13%) in the NEL group and 8 infection (20 %) in reservoir-only group.

**Discussion:** NEL offers an alternative treatment option and reduces shunt dependency after 12 months by almost 40 % compared to infants treated with VAD. Further evaluation is necessary to evaluate long-term safety and neurodevelopmental improvement in this group of patients.

#### OP065

##### **The optimal treatment of hydrocephalus in patients with Dandy Walker Malformation and other midline posterior fossa cysts: a single-center study on 54 patients**

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**Introduction:** Hydrocephalus frequently occurs with midline posterior fossa cystic collections (mPFCCs). The classification of this heterogeneous group of developmental anomalies including Dandy-Walker malformation (DWM), persisting Blake's pouch (PBP), retrocerebellar arachnoid cysts (RAC) and mega-cisterna magna (MCM), is subject of debate. The absence of diagnostic criteria is confusing regarding the ideal management of PFCC related hydrocephalus. We aimed to decipher the surgical strategy for the treatment of children with PFCC related hydrocephalus through a retrospective analysis of the surgical outcome driven by their clinical and radiological presentation.

**Methods:** This study enrolled patients operated of symptomatic PFCC-related hydrocephalus. Clinical and MRI features were examined, as well as the surgical outcome. Unbiased subgroup classification of the patients was performed with multiple component analysis (MCA) as a function of imaging characteristics and Hierarchical Clustering on Principal Component (HCPC). Outcome was assessed with binomial logistic regression and Kaplan Meier analysis.

**Results:** Fifty-four patients were included between 2007 and 2021. MCA suggested that cerebellar and vermian hypoplasia, vermian rotation, basal-tentorial angle, and fastigial angle were strongly correlated. Hierarchical clustering and the distribution of the patients in the bidimensional-plot showed the clear segregation of 3 major clusters, which correlated with the radiologic diagnosis ( $p < 0.001$ ). Binomial logistic regression and survival analysis showed that ETV was an effective treatment for patients with PBP, while failing to control hydrocephalus in most of patients with DWM.

**Conclusion:** Preoperative MRI in patients with PFCC-related hydrocephalus is essential to better define the diagnosis. PBP is a under diagnosed entity. ETV appears to be a very effective treatment in these patients, notably in cases of coexisting Lushka foramina obstruction. On the other hand, DWM is associated to low success rate of ETV, probably because of multifactorial nature of hydrocephalus.

#### OP066

##### Incidence and clinical relevance of the association between trigonocephaly and sylvian fissure arachnoid cysts

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**Introduction:** The association between trigonocephaly and sylvian fissure arachnoid cysts (ACs) has been occasionally reported in the literature (1, 2). However, the real incidence of this association and its clinical relevance is unknown.

**Methods:** Clinical charts and CT scans of patients surgically treated for trigonocephaly at the Pediatric Neurosurgical Department of the Fondazione Policlinico Universitario "Agostino Gemelli" IRCCS in the period January 2014 - December 2022 were collected and retrospectively reviewed.

**Results:** 129 patients underwent surgery for trigonocephaly during the study period. A sylvian fissure arachnoid cyst (AC) was found on the preoperative CT scan in forty cases (31%); the AC was bilateral in 19/40 cases (47.5%), and unilateral in 21 cases (52.5%), all located on the left side. According to the modified Galassi classification, a Grade I AC was documented in 38 cases and a Grade II AC was found in 2 cases. All children underwent a comprehensive neurocognitive evaluation before surgery; thirteen of them showed a neurocognitive delay (10.2%); no significant difference was found in terms of cognitive development between children with arachnoid cyst/s and those without this association. Surgery for the arachnoid cyst was needed during the follow-up in one patient harbouring a bilateral Grade I sylvian AC at the first observation.

**Conclusion:** Even if there is no clear evidence of its clinical impact, the coexistence of either bilateral or unilateral ACs in children with metopic synostosis should be taken in mind and followed up, because of the possibility of evolution of the cysts, in spite of the correction of the craniosynostosis, which in selective cases might need surgical treatment.

References: 1) <https://doi.org/10.1007/s00381-005-0003-5>, 2) <https://doi.org/10.1159/000516207>

#### Abstract Session 10: HC / Spine II

#### OP067

##### Predictive outcome value of angular craniometry in surgical treatment of cerebellar tonsillar herniation

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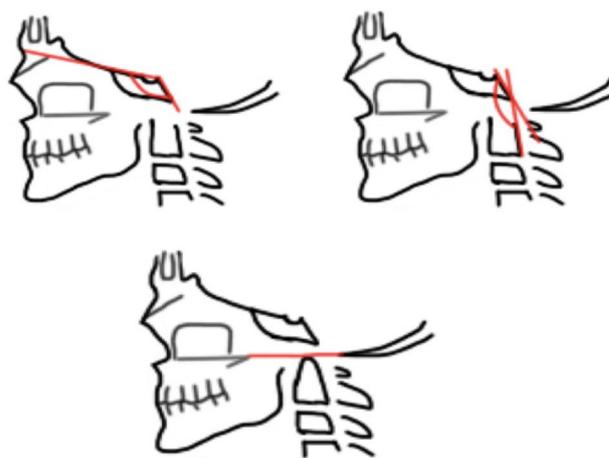
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**Background:** Treatment of isolated, symptomatic tonsillar herniation through the foramen magnum classically consists in decompressive surgery of the posterior fossa (PFD) and expansile duroplasty. The authors aimed at assessing the role of abnormal primary cranial and secondary craniocervical angular craniometric parameters in patients undergoing PFD on the postoperative clinical outcome and complication rate.

**Method:** A retrospective review of data of pediatric patients (<18 years) that underwent PFD at the Pediatric Neurosurgery Department of the Hospital "Femme Mère Enfant" in Lyon, France, over a 20-year study period from 2001 to 2020 were analyzed. Preoperative T1w MRI images were screened for: basal angle, Mc Rae line, Chamberlain line and clivoaxial angle. The association of angular craniometric parameters with clinical outcome criteria were investigated and the CCOS (Chicago Chiari Outcome Scale) was assessed.

**Results:** Of 101 formally eligible patients for data analysis, 55 patients were identified with isolated tonsillar herniation. 25.5% (n=14) % showed a basal angle >133° consistent with platybasia. Basilar Invagination was confirmed in 20% (n=11), with a higher rate in the platybasia group (64.2%). An abnormal clivoaxial angle <145° was found in 62% (n=34). A linear correlation between primary and secondary craniometric angles was confirmed. The CCOS-pain component was significantly higher in case of platybasia and BI. Conversely, there was no statistically difference in overall CCOS score at one year follow-up. Complications were higher in BI, platybasia and in a subgroup of patients with an abnormal CXA < 145° associated with BI. The rate of revision surgeries showed a positive correlation with patients presenting both BI and reduced CXA.

**Conclusions:** Our results confirm the correlation between craniometric angular parameters, show a relative high rate of these findings in patients with apparently isolated tonsillar herniation and suggest that abnormal craniocervical angular geometrics may have a negative predictive outcome value in patients undergoing PFD surgery alone.



**OP068****Posterior cranial encephaloceles**

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**Introduction:** Cephaloceles are a congenital herniation of intracranial structures through a skull defect which occur approximately once in every 2000 births.

**Methods:** We analyzed radiological findings and post-operative outcome of 112 patients diagnosed with posterior encephalocele at Hôpital Femme Mère Enfant of Lyon, Hôpital Roger Salengro of Lille and Charité Universitätsmedizin of Berlin, from 2001 to 2022: 67 patients were diagnosed with parietal (or atretic) cephalocele (PC), and 45 with occipital cephalocele (OC).

**Results:** Antenatal diagnosis was more common in the OC than PC (60% vs 16%;  $p=0.00001$ ). All but 1 patient (diagnosed with giant OC) presented a skin covered lesion at birth. The mean age of surgical correction was 6.2 months (1 day – 33 months). The mean follow-up was 35 months (1–175 months). Brain anomalies were more common in OC than PC (47% vs 28%;  $p=0.04$ ): they were represented from cerebellar vermis dysgenesis, corpus callosum hypoplasia, arachnoid cyst, complex occipito-cervical malformation, Chiari type 3 malformation, Chiari type 1 malformation, ventriculomegaly, hydrocephalus and tentorial displacement. About half of patient presented different vascular anomalies: persistence of falcine sinus, duplicated rectus sinus, duplicated falcine sinus, anomalies of transverse sinus and sinus thrombosis. Polymalformative syndromes were present in 27% of patients diagnosed with OC and in 13% of PC (NS). Developmental outcome was characterized by various degree of psychomotor impairment in 44% of OC and in 16% of PC ( $p=0.01$ ). Five patients died during the follow-up: 1 with PC and 4 with OC.

**Conclusion:** Parietal and occipital encephaloceles represent two different entities: the clinical outcome is significantly more unfavorable in case of OC, while, with few exceptions, PC have a post-operative more favorable clinical course. Their diagnosis and distinction during the antenatal period are fundamental to provide accurate information and adapted management for the patient and for the parents.

**OP069****Intraoperative neuromonitoring in pediatric occult spinal dysraphism surgery - focus: sacral function and BCR**

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**Objective:** A main goal in occult spinal dysraphism surgery is to maintain or improve neurological function, with urogenital function being of strong interest. To assess the functional integrity during neurological surgery, intraoperative neuromonitoring (IONM) is well accepted. Due to anatomical and physiological differences the use of IONM in children is challenging. Monitoring of the bulbocavernosus reflex (BCR) is used to assess the integrity of sacral nerve roots and S2–4 spinal cord segments. Bladder function may be presumed to be closely associated with intraoperative BCR response, but data is sparse. Feasibility and value of BCR in pediatric IONM for occult spinal dysraphism is still under investigation and is addressed in our study.

**Methods:** A retrospective review of pediatric patients undergoing untethering surgery for occult spinal dysraphism in our institution with available intraoperative baseline BCR, pre- and postoperative testing of urogenital function (6 m) was conducted (2017–2022). All patients received total intravenous anesthesia and IONM (NIM-Eclipse® system, Medtronic). BCR waveforms were recorded and free-run electromyography, motor and sensory evoked potentials were obtained. Charts were reviewed and statistical analysis was performed.

**Results:** 51 patients were identified meeting the inclusion criteria (mean age 9+/-30m, F:30). The diagnosis included congenital dermal sinus (3), low lying conus/tight filum (9), fatty filum (15) lumbosacral lipoma (3), lipomyelomeningocele (20) and split cord malformation (1). Baseline BCR waveforms were successfully recorded in 49 of 51 patients (96%). A significant decrease in BCR amplitude was seen in 2 patients, though sacral function at follow up remained stable.

**Conclusion:** IONM recordings of BCR were stable and feasible in our young cohort. A true predictive value regarding loss of BCR and sacral function impairment could not be concluded in our cohort, but observation of BCR amplitude growth after detethering might have further implications for monitoring strategies. Details and correlations will require investigation.

**OP070****Lipomas of the cono-cauda region: value of surgery and gross-total resection in the preservation of long-term neurological outcome**

**Claudia Pasquali**, Francesco Sala

AOUI Borgo Trento, Verona

**Introduction:** The surgical indication for spinal cord untethering in case of spinal dysraphism is shared in the case of symptomatic patients, while in the case of asymptomatic ones there is no agreement: in this analysis we aim to evaluate the presence of predictive variables of a better neurological outcome.

**Methods:** We included 33 patients surgically treated for tethered cord secondary to cono-cauda region lipomas, with the aid of intraoperative neurophysiological monitoring from 2011 to 2021 at our hospital (20 children, 13 adults; mean age 17.7 years (1 day - 64 years)). The clinical evaluation was made using the "Necker score" (good quality of life up to 15 points).

**Results:** The medium-long term outcome (mean follow-up 6 years (8 months – 11 years)) was evaluated in relation to age, pre-operative status and the degree of resection.

No adult patient was asymptomatic at the time of surgery and pre-operative pediatric scores were on average higher than those of adults (16.6 vs 14.4pt). The same was true for post-operative Necker scores (16 vs 13.7pt;  $p=0.009$ ).

Among asymptomatic patients (score 18) and among patients with good Quality of Life (QoL) (score 15–17), there was a certain degree of worsening, but post-operative scores remained in the "good QoL" class (mean 16.3 pt), while the patients who pre-operatively belong to the "poor QoL" class presented a certain degree of improvement (mean 13.1pt), but their scores remained significantly lower than those in the first group ( $p=0.02$ ).

Conus lipomas subjected to subtotal resection (STR) had significantly lower scores at follow-up than lipomas subjected to gross total resection (GTR) and filum lipomas ((14.2 vs 16.7 vs 16.3pt;  $p=0.04$ ).

**Conclusion:** The recovery of pre-operative deficits is difficult and surgery in asymptomatic patients appears to offer protection from severe clinical deterioration. Gross-total excision seems to allow a better clinical outcome at follow-up.

**OP071****New insights in craniovertebral junction MR changes leading to stenosis in children with achondroplasia**

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**Introduction:** We aim to characterize natural history and early changes of craniovertebral junction (CVJ) stenosis in achondroplasia correlating with clinical and radiological outcome.

**Methods:** Retrospective measures on CVJ were performed blindly, on sagittal T2-weighted images, in 21 patients with achondroplasia referred from 2008 to 2020. Clinical and polysomnography data were retrospectively collected. Each patient was paired for age and gender with four controls.

**Results:** Twenty-one patients (11 females, from 0.1 to 39 years of age) were analyzed and paired with 84 controls. A CVJ stenosis was found in 11/21 patients, all before the age of 2 years. Despite a significant reduction of the foramen magnum diameter (mean  $\pm$  SD: patients  $13.6 \pm 6.2$  mm, controls  $28.5 \pm 4.7$  mm,  $p < .001$ ), CVJ stenosis resulted from the narrowing of C2 dens-opisthion antero-posterior diameter ( $8.7 \pm 3.9$  mm vs  $24.6 \pm 5.1$  mm,  $p < .001$ ). Other significant changes were opisthion anterior placement ( $-0.4 \pm 2.8$  mm vs  $9.4 \pm 2.3$  mm,  $p < .001$ ), posterior tilt of C2 ( $46.2 \pm 13.7^\circ$  vs  $31.6 \pm 7.9^\circ$ ,  $p < .001$ ) and of C1 ( $15.1 \pm 4.3^\circ$  vs  $11.9 \pm 5.0^\circ$ ,  $p = 0.01$ ), and dens thickening ( $9.4 \pm 2.2$  mm vs  $8.5 \pm 2.1$  mm,  $p = 0.03$ ), allowing to define three distinguishable early CVJ patterns in achondroplasia.

**Conclusion:** CVJ in achondroplasia results from narrowing between C2 dens and opisthion related to anterior placement of opisthion, thickening of C2 dens, and posterior tilt of C1-C2.

**OP072****Growth trajectories of the occipito-cervical junction in healthy children: geometry, anatomy, and biomechanics**

**Maxime Taverne**, Angèle Ganet, Juliette Raoul-Duval, Roman Hossein Khonsari, Sandro Benichi

*Hôpital Necker Enfants Malades, Paris, France*

**Introduction:** Decision to operate children presenting with anomalies of the occipito-cervical junction (OCJ) is difficult, mostly because so little is known about the normal growth of this region, and regarding the natural history of the conditions affecting it. The present study aims at quantifying the growth of the OCJ in a cohort of healthy children to pave the way for future studies that will provide objective insights to surgeons about whether and when to operate.

**Materials and Methods:** The CT-scans and MRIs of 90 control children aged 0-18 years were included to reconstruct 3D surfaces of the skull base, the first two cervical vertebra, and the ligament network. Their 3D shape was quantified using geometric morphometrics. Growth trajectories of morphological variation were computed using 2-blocks partial least-squares regressions. The evolution of the biomechanics of OCJ was quantified based on the congruence of articular surfaces, and on density variations of bones and ligaments.

**Results:** Here, we showed that morphology strongly covaried with age, allowing us to propose a robust predictive model. This model was very accurate, with less than a millimeter of local discrepancy between predicted shapes and real bone shape for any given age. Our model also precisely predicted the timing of closure of synchondroses.

Finally, we were able to estimate the biomechanical properties of the OCJ throughout age.

**Conclusions:** We believe this work provides some objective information on the evolution of healthy patients and paves the way for further research on cohorts including patients with congenital anomalies of the OCJ. We hope that this will help to better understand their anatomical specificities, and to anticipate their morphological and functional evolution, should they undergo surgery or not.

**OP073****Cranio-cervical mobility assessment in dynamic magnetic resonance imaging in children**

**Flavie Grenier-Chartrand**, Sandro Benichi, Maxime Taverne, Giovanna Paternoster, Raphaël Levy, Kévin Beccaria, Timothée de Saint Denis, Nathalie Boddart, Roman H. Khonsari, Syril James, Thomas Blauwblomme

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**Introduction:** Many pediatric disorders can present with cranio-cervical junction (CCJ) anomalies and subsequent hypermobility that can lead to spinal cord damage and the need for cervical stabilization surgery. We aimed to classify patients according to their CCJ mobility and identify morphometric hypermobility criteria that could be helpful to diagnose cranio-cervical instability.

**Methods:** We retrospectively analyzed cervical dynamic MRI (dMRI) of 47 children with CCJ anomalies performed between 2017 and 2021. For each patient, we used 3D morphometric geometry by applying 13 landmarks (4 for skull base, 4 for C1 and 5 for C2) on sagittal T2-weighted MRI in each position of the head (neutral, flexion and extension) to distinguish hypermobile and normal mobile patients. Delta values (absolute difference between flexion and extension) of 7 morphometric parameters were then measured: basion-dens interval, basion-axis interval (BAI), clivo-axial angle, C1-C2 Cobb angle, pBC2 line (pBC2), Klaus index and C2-opisthion distance, and ROC curves were obtained to define cut-off values of CCJ hypermobility.

**Results:** Using 3D morphometric analyses, two distinct groups of patients were identified: group 1 corresponding to patients with normal CCJ mobility, and group 2 corresponding to patients with CCJ hypermobility. Cut-off values with high predictability of hypermobility were established for pBC2 ( $\text{delta} > 2\text{mm}$ , area under the curve of 0.97) and BAI ( $\text{delta} > 3\text{mm}$ , area under the curve of 0.98).

**Conclusion:** This study identified hypermobile patients using morphometric analyses on dMRI. Furthermore, pBC2 and BAI are morphometric parameters that could be routinely used to diagnose CCJ hypermobility on dMRI. An ongoing study using CT-scan segmentation of the CCJ is being conducted in order to compare our study population to an age-adjusted normal growth model to determine anatomical features of hypermobility.

**OP074****Controversies in Closed Spinal Dysraphisms' surgery: delayed diagnosis and surgery**

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At our institution > 300 patients have been operated for Closed Spinal Dysraphisms, more of 2/3 of them pediatrics, with an increasing percentage of infants. We organized a "one-stop" multidisciplinary clinic for Closed Spinal Dysraphisms with a pediatric neurosurgeon, urologist, neurologist; a pediatric orthopedic or physiatric consultation is added, when needed; MRI are discussed with a neuroradiological team devoted to pediatric cases; a bowel-management team cooperates with the clinic in children needing Clear Intermittent Catheterizations

(CIC). At the same time the analysis of long term results offers new insights into solution of neurosurgical controversies. The surgical policy changed from the beginning Symptomatic-Partial Removal of conus Lipomas to Prophylactic Surgery-Radical Lipoma Removal after 2010. Despite the indications, many children were diagnosed after 24 months; the present study analyzes this patient cohort. A radicular Mapping was performed since 1996 and complete Intraoperative Neurophysiological Monitoring (IOM) comprehensive of anal Meps and BCR since 2010. 78 operated children older than 24 months were reviewed. Mean age at surgery was 7 years (84 months, ranging from 25 up to 216 months); 40 were females and 38 males; reclassified by Pang's subtype they were: 10 transitional, 10 caudal, 6 dorsal 1 caudal conus lipomas; 4 Terminal Myelocystoceles, 6 Retained Medullary Cord, 15 Limited Dorsal Myeloschisis, 21 Filar Lipomas, and 5 Split Cord Malformations. 45/78 patients had already altered preoperative Urodynamic Studies and 28/45 remained CIC dependent after surgery. Our experience supports the early surgery for Closed Dysraphisms: natural history leads to neurourological deficits by adulthood in all the cases and prophylactic detethering with radical lipoma removal is the only technique able to prevent this deterioration.

## Abstract Session 11: Oncology II

### OP075

#### Meta-Analysis and Comparison of Transcranial and Endoscopic Endonasal Approaches for Craniopharyngiomas in Children. Is there an Evolution?

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**Objective:** The purpose of this study is to evaluate the current role of endoscopic endonasal approach (EEA) in the treatment of pediatric craniopharyngiomas, as compared to transcranial approaches (TCs).

**Background:** Historically, comparison between EEAs and TCs has been impaired by patients and lesions selection biases. Recently, there has been a rapid expansion of the EEAs, which have become a valid alternative to the TCs in the adult population, whereas their role remains uncertain among pediatric patients.

**Methods:** We performed a systematic review and meta-analysis of english-language articles published from 2010 to 2021, pooling patients baseline characteristics and outcomes. After screening 617 articles, 20 articles describing 1087 patients undergoing TCs and 18 articles describing 491 patients undergoing EEAs were included. Statistical analyses were done via R and RStudio. Demographic characteristics and outcomes were compared using "metaprop" function in R.

**Results:** Number of patients undergoing EEAs increased over time. Before surgery, patients undergoing TCs had more hydrocephalus ( $p < 0,001$ ), increased ICP ( $p < 0,001$ ), visual impairment ( $p < 0,001$ ), and diabetes insipidus ( $p = 0,025$ ), as compared to EEAs. There was no statistical difference for tumor dimension ( $p = 0,49$ ). Suprasellar localization was more frequent among TCs (92,6% versus 78% in EEAs). EEAs were associated to significantly higher tumor gross total resection rate and lower recurrence rates. CSF leak rate was lower in TC surgeries ( $p < 0,001$ ). After EEAs, neurological ( $p = 0,003$ ), diabetes insipidus ( $p < 0,001$ ), visual ( $p = 0,002$ ), and hypothalamic outcomes were better, whereas pituitary dysfunction was more frequent as compared to TCs. No differences were observed in perioperative mortality, follow-up length, and overall survival.

**Conclusion:** We didn't find any baseline differences regarding tumor dimension and localization between groups, allowing for a direct comparison between the surgical techniques. With an expansion in indications, EEAs are becoming increasingly used for the removal of craniopharyngiomas among children, including bigger tumors, also with supradiaphragmatic extension.

### OP076

#### Extent of resection determines progression rate and growth velocity of residual tumor after resection of pediatric low grade gliomas

David Gorodezki, Julian Zipfel, Manon Queudeville, Jordana Sosa, Ursula Holzer, Jan Kern, Andrea Bevot, Jens Schittenhelm, Thomas Nägele, Martin Ebinger, **Martin U. Schuhmann**  
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**Introduction:** Pediatric low-grade gliomas (pLGG) residuals following incomplete resection (IR) show a high variety of clinical behavior regarding progress or senescence. We retrospectively analyzed tumor growth velocity (TGV) of pLGG before surgery and after IR to investigate the impact of surgical extent, tumor location and molecular BRAF status on postoperative residual tumor growth behavior.

**Methods:** Of 172 patients with pLGG receiving surgical treatment, 107 had IR (66%). 53 and 94 patients could be included in the pre- and post-operative cohort, respectively, mean follow-up time was 40.2 vs 60.1 months. Sequential three-dimensional MRI-based tumor volumetry of a total of 407 MRI scans was performed to calculate pre- and postoperative TGV.

**Results:** ROC analysis identified a residual cut-off tumor volume  $> 2.03 \text{ cm}^3$  associated with a higher a risk of progression post IR (sensitivity 78,6%, specificity 76,3%, AUC 0.88). Mean preoperative TGV of  $0.264 \text{ cm}^3/\text{month}$  showed significant deceleration of tumor growth to  $0.085 \text{ cm}^3/\text{month}$ ,  $0.024 \text{ cm}^3/\text{month}$  and  $-0.016 \text{ cm}^3/\text{month}$  after 1st, 2nd, and 3rd IR, respectively ( $p < 0.001$ ). Results remained significant after excluding patients undergoing (neo)adjuvant treatment. Resection extent showed correlation with postoperative reduction of TGV ( $R = 0.97$ ,  $p < 0.001$ ). Postoperative TGV of BRAF V600E-mutant LGG was significantly higher than of BRAF wild-type LGG ( $0.123 \text{ cm}^3/\text{month}$  vs.  $0.016 \text{ cm}^3/\text{month}$ ,  $p = 0.047$ ).

**Conclusion:** These data strongly suggest that the extent of surgical resection impacts pediatric LGG growth kinetics post incomplete resection by either inducing senescence (no progression) or inducing a significant deceleration of tumor growth. BRAF-V600E mutation was confirmed as a risk factor for higher postoperative TGV. Larger multi-center cohorts should be explored to confirm these institutional results. Maximal safe resection trying to reduce residual tumor volume to the minimum remains therefore the primary treatment options in pediatric LGG.

### OP077

#### Childhood Tumours in Adults – a single centre analysis

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**Introduction:** Paediatric brain tumours constitute a particular oncological entity and differ from those found in adults. Rarely, adults present with these tumours. The tumour's behaviour and response to treatment is not well known in adults and to date treatment has been largely determined by following the paediatric experience. We aimed to review the literature on paediatric-like tumours in adults, to analyse such patients presenting to our Institution and compare our results to the data from the Surveillance, Epidemiology, and End Results (SEER) program.

**Methods:** We searched for adult patients diagnosed with paediatric-like brain tumours between January 2012 and August 2022. The following histopathology-diagnosis were included: medulloblastoma, pylocytic

astrocytoma (PA), craniopharyngioma, choroid plexus papilloma, pleomorphic xanthoastrocytoma (PXA), atypical teratoid/rhabdoid tumour (AT/RT), ependymoma, pineal region tumours, germinoma, subependymal giant cell astrocytoma (SEGA) and embryonal tumour NOS WHO 9473/3. Patient's demographics, age at diagnosis, tumour location, molecular diagnosis, presence of multifocal disease or metastasis, type of surgical treatment and adjuvant therapy were collected. Survival at 6-m, 5-y and 10-y was registered. The data was thereafter compared with the SEER's data.

**Results:** Overall 36 patients presented with PA, 14 with medulloblastoma, 1 with AT/RT, 9 with choroid plexus papilloma, 4 with germinoma, 28 with ependymoma, 25 with craniopharyngioma, 10 with pineal region tumours, 5 with PXA, 3 with SEGA and 3 with embryonal tumour WHO 9473/3. Concerning survival, 5.7% of the patients with PA and 20% with ependymoma died within 1-year due to disease progression. 35.7% of the medulloblastoma patients did not reach 5-year survival. Further analysis is in progress.

**Conclusions:** We present one of the first series regarding the incidence of paediatric-like brain tumours in adults. Further collaborative studies will allow a better overview of disease's natural course of these tumours in adults and help us to optimise treatments and patient's outcomes.

#### OP078

##### Evaluating the need of early post-operative CCT imaging after brain surgery in children

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**Introduction:** In many institutions, early post-operative CCT (EPOCT) is routinely performed after elective intracranial procedures. Considering the lack of evidence for the clinical value of EPOCT esp. in pediatric patients, we sought to determine the impact of planned EPOCT on post-operative management of children who underwent brain tumor procedures at our institution.

**Methods:** Patients 0 to 18 y of age who underwent brain tumor surgeries and received EPOCT (i.e. within 4 hours after surgery) were included. Demographic data, pre-op Lansky score, early post-op neurological status, histological diagnosis, tumor location, extend of resection, surgical positioning, pre-operative ventricular size (Evans ratio and FOHR), duration of intubation/surgery, unexpected findings (UF) on EPOCT, early return to OR (RTOR), hemoglobin count, and need of blood products were retrospectively analyzed to identify risk factors for unexpected imaging findings.

**Results:** 247 patients were included (mean age 9 +/- 5y; female to male ratio 44,1%/55,9%). In 20 cases (8%) EPOCT revealed unexpected findings. These included—among others—significant pneumatocephalus (after sitting position) (n=8), new hydrocephalus (n=3), significant blood clot on the resection site (n=1). RTOR occurred in 9 of these cases. The relative risk of RTOR was elevated after surgery in a sitting position (OR 12.21; CI 1,55 – 96,06). Patients with new neurological (26,7%) deficits were more likely to have UF on EPOCT (OR 7.85; CI 2.87 to 21.45). Patients with severe neurological deficits presented UF findings that necessitated further surgical intervention. UF occurred slightly more after resection of infra-tentorial tumors (OR 1.76; CI 0,7 – 4.4).

**Conclusion:** Based on these data, pediatric patients with no or mild post-operative changes of their initial neurological status after brain surgeries can be monitored clinically. Severe post-operative neurological deficits justified EPOCT. These preliminary data provide a basis to further prospectively develop clinical scores to better define the indication for EPOCT in children.

#### OP079

##### Management of orbital tumors in children

**Grazia Menna**, Paolo Frassanito, Federico Bianchi, Ludovico Agostini, Angela Musarra, Gianpiero Tamburrini, Luca Massimi  
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**Introduction:** Traditionally, invasive surgical approaches have been the mainstay of treatment for orbital tumors. Minimally invasive approaches are aimed at minimizing surgical morbidity and facilitate recovery. Aim of the present, single center, retrospective study is to evaluate whether the employment of minimally invasive approaches to orbital lesion could effectively improve surgical, clinical, and aesthetic outcomes compared to invasive ones.

**Methods:** All consecutive children admitted to our Institution from January 2010 to January 2020 for orbital tumors were enrolled for the present study. Thirty patients were collected and were divided into two groups: group A, 14 children (46.66%), treated with traditional craniotomic approaches; group B, 16 children (53.44%), managed by mini-invasive transorbital ones.

**Results:** There were no significant differences between the two groups in terms of anagraphical data and extent of tumor resection. Mean surgical times (40 min vs 70 min,  $p < 0.0001$ ), surgical complications such as periorbital edema (37% vs 78%,  $p 0.02$ ) and dural tear (0 vs 21%,  $p 0.05$ ), and procedures costs ( $p < 0.0001$ ) were significantly reduced in group B patients. In terms of clinical outcomes, group B showed a significant reduction both in terms of post-operative pain (mean score based on visual pain scale was 2.9 vs 4.1  $p 0.003$ ) and mean hospitalization time (4.5 days vs 5.5 days,  $p 0.0004$ ). The aesthetic outcome according to Sloan classification was significantly improved in this group as well. (81% vs 36% class I patients,  $p.005$ ).

**Conclusions:** In spite of the limitations related of a single Center, retrospective study, this analysis shows that the employment of mini-invasive approaches to orbital tumor has clear advantages in terms of surgical, clinical, and aesthetic outcomes, therefore should be preferred whenever feasible.

#### OP080

##### Sodium Fluorescein-guided surgery for low-grade gliomas in pediatrics: a single-center experience

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**Introduction:** Low-grade gliomas (LGGs) account for about 40% of all CNS tumors in pediatrics. The first-line treatment remains surgery, both providing pathological samples and reduction in tumor volume, which may anyway present some challenges. A certain number of adjuncts have been studied to reach for the maximal safe resection in neuro-oncology, and among these some intraoperative dyes, like Sodium Fluorescein (SF), demonstrated their utility in high-grade neoplasms. However, the role of SF in pediatrics and in lower grade lesions has not been fully elucidated yet.

**Methods:** A retrospective review of charts, radiological findings, and intraoperative images of children operated for low grade gliomas (WHO grade I and II) at our Institution from September 2021 to October 2022 was conducted. We analyzed intraoperative subjective surgeon's experience with SF, we made a comparison between intensity of fluorescence and gadolinium enhancement at preoperative MRI and evaluated the relationship of SF to intraoperative pathological sampling.

**Results:** A group of 22 patients operated for a CNS low-grade glioma was found. Globally, 86% of lesions showed intraoperative fluorescence, which was considered as “helpful” by the surgeon in 74% of cases. Intensity of fluorescence and gadolinium enhancement (GE) at the preoperative MRI seem similar: 94% of the LGGs with GE at preoperative MRI showed intraoperative SF uptake (perfect intensity correspondence in 59%, brighter than GE in 27%). Intraoperative pathological sampling was efficient in every case showing SF uptake (88%), while it was not when SF uptake was absent (12%), although the definitive examination confirmed the tissue as tumor.

**Conclusion:** SF has a promising role especially in gadolinium enhancing pediatric LGGs, as it is often considered helpful by the surgeon, and can become part not only in tumor removal but also in the macroscopic diagnostic, being integrated into the intraoperative immediate pathological evaluation.

## Flash Presentations

### Abstract Session 1: Spine I

#### FP001

#### Short and Long-Term outcomes of post-natal repair of myelomeningocele: a UK case series

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**Introduction:** Open myelomeningocele (MMC) is a congenital abnormality frequently associated with hydrocephalus and Chiari II malformation (CM). The aim of this study was to evaluate the short and long-term outcomes of babies undergoing post-natal closure of MMC in our institution.

**Methods:** We conducted a retrospective review of all babies born with open MMC in our institution from 2013-2022. Demographics, time to closure, need for CSF diversion, presence of CM, ambulation, continence and cognitive long-term outcomes were recorded (range 1-8 years).

**Results:** A total of 62 babies underwent primary closure of the MMC (M:F ratio 1:1) with 96% being diagnosed antenatally. The most common type of delivery was caesarean section (n=35). A total of 46 babies were born at term and 11 pre-term (<37 weeks). Repair of MMC was carried out within 48 hours in 95% of cases. CM was present in 38 babies (61%). Of the 59 babies that survived, a total of n=45 (75%) required a VP shunt. Average time to shunt was 33 days post MMC repair (range 1-260 days). The most common indication for shunting was increasing OFC, followed by CSF leak from the MMC wound. Long-term follow up indicated that 48% of patients were ambulant, 93% had some degree of neurogenic bladder and 39% had cognitive/language developmental delay, while 65% of patients were in mainstream schooling.

**Conclusion:** The majority of babies born with MMC were diagnosed antenatally and underwent a primary closure within 48 hours of birth. We report a slightly lower rate of shunt requirement compared to the reported literature, and no significant change in practice or shunting rate over the 9 year observation period. Around half of the children were ambulant and able to engage in mainstream education.

#### FP002

#### Successful operative management of anterior sacral meningocele by ligating the fistulous connection via posterior approach: Experience of 5 Cases

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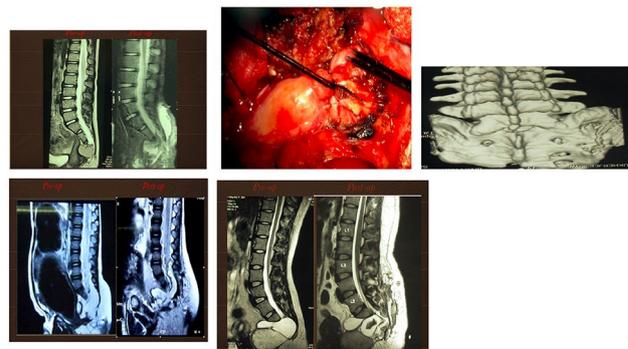
**Introduction:** Anterior sacral meningocele (ASM) is a rare neurulation disorder with defect in anterior sacral wall causing dural outpouching

in the pelvis. They occur in third decade of life and more common in females. Mechanism of pathogenesis can be congenital, ischemic, traumatic or iatrogenic. These are commonly associated with spina bifida, bicornuate uterus, epidermoid, vaginal duplication. Most commonly they are asymptomatic and when increases in size, present with constipation urinary incontinence, low back pain, menstrual symptoms and bacterial meningitis. Familial cases with autosomal dominant transmission have also been described. Diagnosis can be made by X ray of sacrum which shows sickle shaped bony defect. MRI pelvis is the diagnostic modality of choice. If not surgically intervened, these have 30 % mortality. Though transabdominal, endoscopic, laparoscopic approach has been described in literature, posterior approach remains the standard.

**Methods:** 5 cases of ASM who were managed in our institute since 2014.

**Results:** We had 5 cases of ASM, out of which 3 were children who presented with foot drop, gynecological problems, constipation, discharging sinus and scoliosis. To our knowledge foot drop and scoliosis in ASM has not been described earlier in literature. Two of our patients had associated dermoid cyst and one had tethered cord with intramedullary abscess. All cases were managed via posterior approach, where the fistulous connection was identified and ligated. Post operative MRI Spine of cases showed residual sac but the connection was obliterated. In all our cases, there was significant post operative clinical improvement.

**Conclusion:** ASM can be effectively managed by posterior approach and complete excision of sac is usually not required.



#### FP003

#### Spectrum of neural/cutaneous ectodermal disjunction failures: the limited dorsal myeloschisis-myelocystocele riddle

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**Background:** Limited dorsal myeloschisis (LDM) is a relatively recent addition to the classification of spinal dysraphisms and is defined as a focal lack of neurulation of the primary neural tube, resulting into a tethering stalk that originates from the posterior aspect of the spinal cord, traverses a focal posterior spina bifida, and terminates with a superficial skin tag. On the other hand, myelocystoceles (MCC) are classically defined as extrusions of a focal syrinx into a skin-covered meningocele and are categorized into terminal and nonterminal forms depending on their location at the terminal end of, or along the length of, the spinal cord, respectively.

**Methods:** We retrospectively review our surgical series of 30 consecutive patients with a neuroradiological diagnosis of either LDM or MCC referred to our tertiary care pediatric institution between 2005 and 2022. All patients were successfully operated on and improved clinically thereafter. No patient required second-look surgery for rethetering.

**Discussion:** LDM and MCC are characterized by a focal connection between the spinal cord and the overlying skin, which can occur in a nonsaccular (LDM) or saccular (MCC) fashion; the latter is further complicated by the extrusion of a focal syrinx when the lack of separation between the cord and the skin is sizable enough to allow CSF extrusion from the central canal. It is not necessary to invoke incomplete neurulation as an explanation for LDM, hence the term “myeloschisis” is probably a misnomer in that no open spinal dysraphism is present, and a neural placode cannot be identified on surgery. **Conclusion:** Both LDM and MMC represent morphological clusters along a spectrum of saccular and nonsaccular abnormalities caused by focal lack of disjunction between the neural and cutaneous ectoderm at the end of the primary neurulation process. For classification purposes, the term “LDM spectrum” can probably be accepted.

#### FP004

**Outcomes Of Untethering Of Asymptomatic And Symptomatic Patients Presenting With Tethered Cord Syndrome: Experience From A Tertiary Care Center And Review Of The Literature**  
**Marwan Najjar**, Charbel Moussallem, Sarah Kawtharani, Nour Najjar  
*American University Of Beirut, Beirut, Lebanon*

**Introduction:** Tethered cord syndrome is a rare neurologic entity characterized by varying signs and symptoms, ranging from asymptomatic to a significant impairment of quality of life. The present study showcases the presentation and outcomes of 3 subgroups undergoing spinal cord untethering and aims to highlight the importance of early surgery for untethering of the spinal cord, along with review of the literature.

**Methods:** A retrospective review from a tertiary care center in the Middle East was conducted between 2011 and 2020. The studied population was classified into three groups depending on the age at presentation and whether the operation consisted of primary or secondary untethering. Patients with underlying myelomeningocele were excluded. The patients were followed postoperatively for a variable period and were monitored for symptom development, regression, or progression.

**Results:** The sample consisted of 28 patients, divided into 3 groups. The age ranged between 5 days and 27 years and 64% were females. The first (39%) and second (25%) groups involved patients presenting before 1 year and past 1 year of age for primary untethering, respectively. The third group (36%) involved symptomatic patients presenting for secondary untethering after having had spina bifida surgery without untethering at an early age (mostly in another institution). The most common symptom was lower extremity deformity (43%), followed by neurogenic bladder (32%). Children in the first group were mostly asymptomatic (88%) and remained asymptomatic at last follow up. Amongst the 19 symptomatic patients, mostly from the second and the third groups, 13 (68%) had persistent symptoms, whereas 5 (26%) patients noted improvements in their symptomatology, and none were completely asymptomatic at last follow-up.

**Conclusion:** We recommend early untethering of the spinal cord, even before symptom development. Untethering for symptomatic patients should be done for symptomatic relief, halting symptom progression, and preserving baseline functions.

#### Abstract Session 2: Craniosynostosis

##### FP005

**Oxycephaly: clinical phenotype descriptions and outcome differences depending on the surgical treatment**  
**Giovanna Paternoster**, Michela Vita, Eric Arnaud, Federico Di Rocco, Dominic Renier, Roman Hossein Khonsari  
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**Introduction:** Oxycephaly is a rare condition of premature fusion of both coronal sutures and the sagittal suture, with or without fusion of the lambdoid. Two types of treatment are more frequently performed: 1) vault decompression (biparietal flaps) and 2) fronto-orbital advancement with fronto-parietal remodeling. Nevertheless, because of the rarity of oxycephaly, there is no real evidence about the natural history of this condition, the best surgical management and outcome.

**Methods:** Twenty-eight cases of oxycephaly managed and operated by the Necker Enfants Malades craniofacial team since 2000 were retrospectively examined. Clinical, radiological and esthetical data has been recorded and a statistical analysis has been performed.

**Results:** Three groups of main different morphologic types have been observed: A (forehead tilted backward, opening of the frontonasal angle), B1 (harmonious ovoid) and B2 (dolichocephalic). Increased intracranial pressure, cognitive difficulties and developmental delay is present in several patients. We analyzed 2 groups of patients: group 1 - submitted to FOA and group 2, submitted to vault decompression with biparietal flaps. A significant improvement in cranial volume when a FOA was performed instead of vault decompression has been statistically demonstrated (p 0.0128).

**Discussion and Conclusion:** Oxycephaly is a rare and heterogenous condition and due to the high risk of intracranial hypertension and cognitive deficits, an early recognition of this craniosynostosis should be mandatory. The high frequency of increased intracranial pressure, cognitive impairment and developmental delay confirmed that surgery is not optional in these patients because these sequelae should be completely prevented or reduced by an early surgical treatment. Regardless of age at surgery, the fronto-orbital advancement (FOA) permits to achieve bigger cranial volume, better aesthetic results and seems to reduce the risk of reintervention

##### FP006

#### Posterior Cranial Vault Distraction for Craniosynostosis

**Laura Grazia Valentini**<sup>1</sup>, Veronica Saletti, Enrico Gambatesa, Alessandra Erbetta, Enrico Gambatesa, Tommaso Galbiati, Alberto Sommariva, Ignazio Gaspare Vetrano, Fabio Mazzoleni  
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Polysynostosis are correlated with low cranial volumes and frequent association with secondary Chiari Malformation Type 1 (CM1). Cranial Vault remodeling has the side effect to cause a relevant stress to the cranial scars. Since 2014 a selected group of children affected by polysynostosis were operated by Posterior Cranial Vault Distraction to increase intracranial volumes.

One child had Pfeiffer Syndrome, 4 Brachicephaly due to bicoronal Synostosis without any midface or orbital problems, 2 had hypophosphatemic polysynostosis, 2 had oxycephaly and 1 frontonasal Dysplasia. Volumes were measured on pre and postoperative 3DCT and compared with normal volumes, obtained in a comparable group of children submitted to the 3DCT at our institution for other reasons (mainly skull small tumors). The distraction was performed from a minimum of 2.5 cm up to 3 cm; than the distractors were left in place for another month of consolidation, then after removed; there were no major complications, neither lack of ossification; in just 1 child we observed a transient alopecia; in all cases CM1 improved and Syringomyelia regressed.

The present series confirms that Cranial Vault Osteogenic Distraction is a safe treatment, able to increase intracranial volumes, so curing CM1 and improving aesthetics. Another advantage was feasibility at any age, also in older children.

##### FP007

#### Extracellular vesicles in nonsyndromic craniosynostosis: potential key players in the pathogenesis of the disease

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**Introduction:** The physiopathology of nonsyndromic craniosynostosis (CS) is still unclear. In a highly dynamic microenvironment like the osteogenic niche, the study of extracellular vesicles (EVs) has become essential to decode cellular messages. In this study, we isolated EVs from CS patients' sutures and established an in vitro model to evaluate their possible implication in the disease pathogenesis.

**Methods:** Specimens from normal and pathological sutures were obtained as surgical waste from nonsyndromic CS patients' cranioplastic surgery. Tissues and their primary cells were grown in serum EVs-free culture medium to isolate tissue- and cell-EVs, respectively. Nanoparticle Tracking and Dynamic Light Scattering analyses were done to evaluate EVs quantity and size among the different samples.

**Results:** We successfully isolated tissue-EVs from CS patients' sutures. Pathological sutures displayed a high rate of EVs production with a different range size, compared to their normal counterpart. Similarly, pathological cell-EVs retained a high rate of EVs production in vitro, with a range size more comparable to normal samples.

**Conclusions:** The premature ossification of skull bone sutures in CS arises from a dysregulated ossification process. EVs are important players in the crosstalk within the different osteogenic cells. Our data showed that skull bone tissues can release EVs whose number and size vary in relation to the pathological/physiological state of the suture of origin. The same secretory properties were retained by the primary cells giving the chance of a functional validation, in the view of our ongoing experiments on the EVs cargo. Funding: Lazio Innova A0375-202036576

#### FP008

**Does the participation of a senior plastic surgeon improve perioperative conditions in craniosynostosis repair surgery?**

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**Introduction:** The real impact on perioperative care of the participation of other surgical specialties together with neurosurgeons during craniosynostosis repair surgery has not yet been demonstrated. The purpose of this study was to determine whether the participation of a second senior surgeon (plastic surgeon) during surgical repair of pediatric monosutural craniosynostoses improved perioperative medical care.

**Material and Methods:** The authors retrospectively reviewed 2 cohorts of patients who had consecutively undergone primary repair surgery for trigonocephaly and unicoronal craniosynostosis. Infants were operated on by a single senior pediatric neurosurgeon before December 2017, and with the collaboration of a senior plastic surgeon after January 2018.

**Results:** Overall, 60 infants were included in the study: 29 infants in group 1 (single surgeon, 2011-2017), and 31 in group 2 (pair of surgeons, 2018-2021). The median duration of surgical procedures was significantly lower in group 2 compared with group 1: 180 vs 167 minutes;  $p = 0.0045$ . There was no statistical difference between 2 groups regarding estimated blood volume lost or intra/postoperative packed erythrocytes transfusion. Postoperative drain output was significantly lower in group 2. Median lengths of stay in hospital and in intensive care were statistically lower in the group of pair surgeons: median timesaving was estimated respectively as one and two days;  $p \leq 0.0001$ .

Volume of solution infused, diuresis, immediately postoperative hemoglobin, hematocrit, hemostasis (platelet count, fibrinogen, prothrombin time and activated partial thromboplastin time), and return to receiving food orally did not differ from one group to the other.

**Conclusion:** Results confirmed our impression of an improvement in perioperative medical care. However, the role of surgical experience and the influence of the medical/nursing staff must not be minimized in these complex surgical procedures.

#### FP009

**Craniosynostosis Repair: Cases series and results of a novel team at Brazilian countryside**

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**Introduction:** Craniosynostosis surgery demands a perfect experience curve to reach the best results. This paper depicts a series of 45 cases operated on by the same neurosurgeon and his novel team among the most experienced specialists in the city.

**Methods:** Between January 2018 and November 2022Thirty-four34 boys (75,5%) and 11 girls (24,5%) were operated on for craniosynostosis. The cases were divided into Scaphocephaly (46,67%); Trigonocephaly (40%), Anterior Plagiocephaly (8,89%); Posterior Plagiocephaly (2,22%), and Brachycephaly (2,22%). Age at surgery ranged from 3 - 42 months; most of them, 71%, were approached before one year of age, (13%) were operated on between 12 and 24 months, and seven children (16%) were approached lately, between 24 and 42 months.

**Results:** The time elapsed for each surgery ranged between 40 minutes and 3 hours, and 30 minutes. Scaphocephaly cases were the fastest ones. Blood transfusion was done in 27 cases (38%). Those who needed orbitofrontal advancement and the brachycephaly and posterior plagiocephaly cases were transfused. The mean days of hospitalization were 3,8 days (range 3-7). Intraoperative complications were divided into minor, such as dural tears (13,3%), and major ones, like air embolism (1/45 - 2,22%) and superior sagittal sinus (SSS) lesion (1/45 - 2,22%). All the dural tears were promptly stitched. The air embolism was quickly aspirated from the central line without any consequences. The SSS lesion was sutured without problems. Late complications were nihil. Restenosis was seen in only one trigonocephaly case, and the parents had already had their first child operated on for trigonocephaly ten years ago. They refused cosmetic forehead repair.

**Conclusions:** This paper shows that even in the inner of developing countries, achieving the same results as well-stated centers worldwide is possible. Scaphocephaly was the most common one, and trigonocephaly followed. Intraoperative complications can always be quickly handled with experienced hands.

#### FP010

**Long Term Cosmetic, Ophthalmological, Cognitive, and Parental Satisfactory Outcome after Open Sagittal Craniosynostosis Repair**

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**Aim:** The aim of this study was to investigate the long-term cosmetic, ophthalmological, cognitive, and parental satisfactory outcome of children after open surgical correction for non-syndromic sagittal craniosynostosis.

**Methods:** Patients under the age of twelve months, who underwent surgical reconstruction with a follow up period of at least 3 years after surgery at the Department of Pediatric Neurosurgery, University Hospital of Basel, Switzerland were included in this single center study. The

primary outcome was parent-reported satisfaction of the surgery (PRSS) assessed through a parental questionnaire. Secondary outcomes were neurocognitive development and health-related quality of life (HRQoL), measured using the Vineland Adaptive Behavior Score (VABS) and the KINDL® questionnaire, ophthalmological outcome, cosmetic outcome based on 3D imaging of the patients and clinical evaluation.

**Results:** We included 25 children with a mean age of 5.94 months at surgery (16% girls). Based on PRSS 96% of the parents are satisfied with the head shape of the surgical outcome. VABS showed a mean of 104.52, corresponding to an overall age-adequate adaptive level. The patients showed a mean score of 84.42/100 for HRQoL, which is in the norm. The mean Cephalic index is with 78% in the mesocephalic range. The ophthalmological outcome is for all children unremarkable.

**Conclusion:** The results from our series demonstrate that parents satisfaction is high after surgery, the cosmetic outcome is satisfying subjectively and objectively. In our cohort we could not find any developmental or ophthalmological disabilities above the average of normal population and the HRQoL is described within the norm.

#### FP011

##### The incidence of sagittal synostosis in normocephalic children undergoing craniovertebral decompression for Chiari 1 malformation

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**Introduction:** The presence of an underlying sagittal synostosis (USS) in children with a dolichocephalic head presenting with Chiari 1 malformation (CM-1) has been reported in the literature to be around 15%. However, little is known about the incidence of USS in normocephalic CM1 patients undergoing craniovertebral decompression (CVD).

**Methods:** A retrospective review of the first author's prospective database was performed. Patients less than 17 years old with CM-1 who underwent CVD were reviewed in detail, and their clinical and radiological outcomes were examined. USS was also searched in a control group of normocephalic children who underwent CT head in the same institution for reasons different than craniosynostosis, hydrocephalus, or shunt-related issues.

**Results:** Between October 2013 and October 2022, 50 children underwent CVD for CM-1. 36 patients (72%) underwent a CVD with dura opening and duroplasty (DOPD), 32 patients as the first treatment, and 4 patients following a previous unsuccessful osteo-ligament decompression (OLD). In the DOPD group, three patients developed hydrocephalus (8.3%), in one patient preceded by CSF leakage; in two patients (6.6%) the syringomyelia did not respond to the CVD. All these children underwent CT head scans before undergoing further surgery, revealing in 4 out of 5 cases the presence of USS. The incidence of USS in the control group of 250 children was 3.6%.

**Conclusion:** An USS could be found in at least 8% of normocephalic CM-1 patients, and in this series accounted for 80% of the complications requiring further surgery. This experience indicates that a 3D CT scan head should be performed in all patients undergoing CVD. Furthermore, the high percentage of complications seen in this subgroup suggests cranial vault augmentation (fixed or dynamic) as the treatment of choice in patients with CM1 secondary to USS irrespective of age and head shape.

#### Abstract Session 3: Technology

#### FP012

##### Novel workflow in craniosynostosis: Virtual surgical planning based in statistical shape modeling and intraoperative navigation

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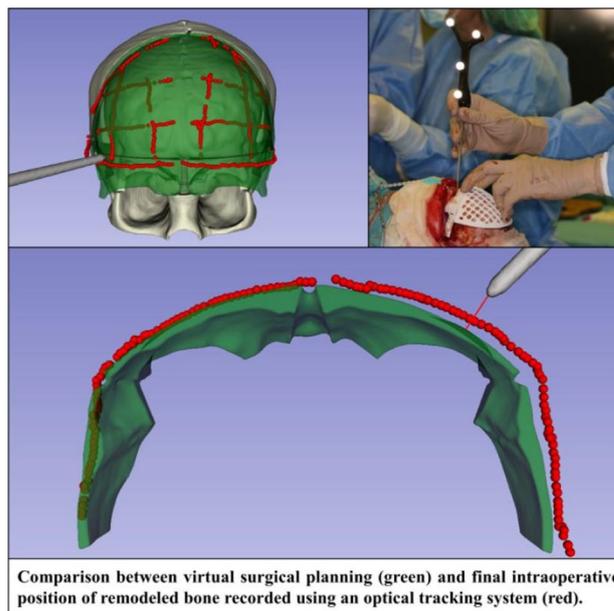
**Introduction:** In craniosynostosis, virtual planning software manipulate preoperative images to recontour the patient's anatomy and to develop stereolithographic models to guide osteotomy and bone fragment remodeling.

Despite the positive impact of these technologies, there is a lack of objective methods to quantify cranial malformations in clinical practice, which makes diagnosis, the decision to go for a surgery, and the intervention planning very dependent on the surgeon's expertise.

**Methods:** The proposed workflow was followed in eleven patients presenting craniosynostosis. For each patient, a multi-atlas of healthy cases was used to compute a normative skull model from the preoperative CT scans. These normative models were used as a reference for preoperative virtual surgical planning. Finally, an intraoperative navigation system, based on optical tracking and developed in our center, was used to accurately position remodeled bone fragments and ensure an accurate translation of virtual planning through real-time 3D metrics. Our navigation system does not require head immobilization or the attachment of markers prior to surgical intervention.

**Results:** The Navigation System confirmed intraoperatively the correct performance of the planned cranial reshaping and its exact three-dimensional positioning on the child. The use of the surface scanner allowed an accurate evaluation of the surgical outcomes. The estimation of navigational accuracy using intraoperative scanning yielded an average error of less than 0.5 mm in the remodeled frontal bone and the supraorbital bar.

**Conclusion:** This novel workflow for craniosynostosis reconstruction enables an accurate translation of preoperative virtual surgical planning into the operating room, thus improving reproducibility and reducing inter-surgeon variability. Moreover, the use of a normative skull model as reference for preoperative planning allows the surgeons to achieve optimal outcomes.



Comparison between virtual surgical planning (green) and final intraoperative position of remodeled bone recorded using an optical tracking system (red).

#### FP013

##### Planning of spring-assisted cranioplasty using patient-specific simulation

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**Introduction:** To enhance clinical outcomes of spring-assisted cranioplasty a simulation tool would be useful to calculate individualized osteotomy parameters for each patient. This study aimed to explore the feasibility of patient-specific finite element (FE) simulation for surgical planning. It included evaluation of applicable modeling approaches [1], sensitivity analysis of surgical parameters and investigation of optimization strategies.

**Methods:** Cranial geometries were derived from three patient cases and used as inputs for FE patient models [1]. Cranioplasty was virtually reproduced with representation of the CranioXpand system (KLS Martin) simplified as linear springs calibrated with test data. Each model was parameterized with respect to cut placement, spring characteristics and positioning. Simulation outcomes were compared with respect to deformation using standard surgical practices. Sensitivity of surgical parameters was evaluated for potential optimization criteria (cranial index and symmetry). Finally, an optimization study was conducted to identify cut and spring parameters for an optimal post-operative outcome.

**Results:** Post-operative deformation could be successfully generated for all three patient models consistent with previous works [1]. Sensitivity analysis highlighted the impact of spring positioning besides spring characteristics on cranial index, whereas cut placement was found to be more crucial for obtaining cranial symmetry. Optimization confirmed that a higher cranial index and better symmetry can be achieved by optimal selection of surgical parameters.

**Conclusion:** The obtained results demonstrated feasibility and applicability of patient-specific FE simulation for surgical planning with reservation of rigor validation. The identified surgical parameters impact the postoperative outcome such that the defined optimization criteria and techniques can be utilized within a simulation tool to improve surgical intervention.

**References:** [1] Borghi et al. *Biomech Model Mechanobiol.* 2020 19(4):1319-1329

#### FP014

##### Skull bone thickness assessment using ultrasonography - a validation study

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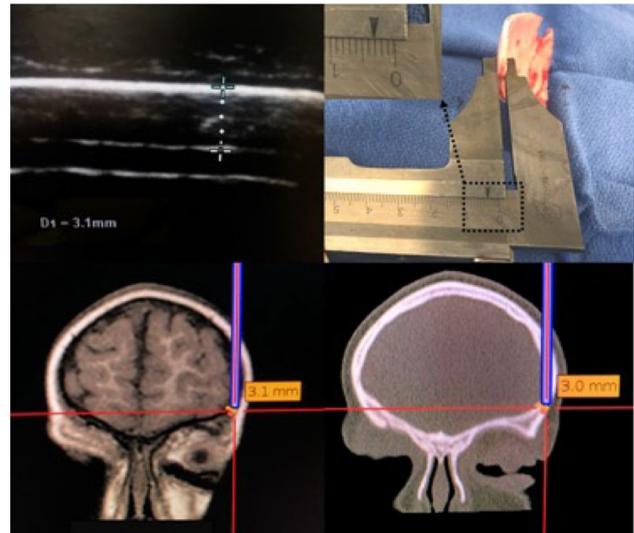
**Background and Aim:** Data regarding the accuracy of skull bone thickness assessment using ultrasonography are scarce in literature, although knowledge about these parameters could be helpful for interventional planning and surveillance of physiologic growth and development. Our study aimed at investigating the validity of the ultrasound technique, comparing the measurements with an intraoperative assessment of bicortical bone thickness.

**Method:** Using a high linear high frequency transducer probe (13MhZ), we performed 24 ultrasonographic measurements of skull thickness prior to incision, taken on two adjacent target points in 12 consecutive children undergoing planned craniotomies with neuronavigation at our department of pediatric neurosurgery at the « Femme Mère Enfant » Hospital in Lyon, France. The bone flap was performed according to the preoperative craniotomy plan defined by neuronavigation and an intraoperative direct measurement of the skull thickness was repeated at the same points. In addition, measurements were taken on MRI Studies and on CT Scan studies and the measurement techniques were compared.

**Results:** Compared to direct intraoperative skull thickness measurement performed on the bone flap, linear regression analysis showed a statistically significant accuracy of the ultrasonographic method ( $p < 0.001$ , 95% CI 0.853 – 1.080). Moreover, ultrasonography showed a good correlation not only with the intraoperative measurement

performed on the bone specimen, but was also comparable to the measurements that were taken both on MR and CT studies ( $p < 0.001$ ).

**Conclusions:** Ultrasonography is a valid tool to assess skull bone thickness. In patients undergoing head surgery, this approach could represent an useful, easily available, real-time instrument to facilitate operative planning, minimize complications and allow for non-invasive follow-up, e.g. in craniostylosis surgery and head pin placement.



#### FP015

##### A feasibility study for augmented reality applied in paediatric craniofacial surgery

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**Introduction:** Augmented Reality (AR) is an advanced technology which allows overlapping and integrating the virtual information with the reality related to subject. It has been widely applied to medical sciences recently and has a great potential in order to realize a full concept of intraoperative navigation.

**Materials and Methods:** In this study the authors aim to assess the feasibility of HoloLens 2 head mounted display in craniofacial surgery.

The selected procedure to test in vitro is fronto orbital remodeling (FOR). The authors tested the AR application for HoloLens 2 by performing selected FOR osteotomies (i.e. the nasal osteotomy and the frontal osteotomy) under its guide. Each user repeated the procedure six times on the same 3D printed phantom. Accuracy was then measured by means of calibrated CAD CAM cutting guides.

**Results:** With HoloLens 2, 97% of the users were able to successfully trace the osteotomy trajectory with an accuracy level of  $\pm 1.5$  mm for the nasal cut and 80% the frontal cut. For accuracy level  $\pm 1$  mm and  $\pm 0.5$  mm, for nasal cut, we reported 80% and 61% respectively. Regarding the frontal cut, the reported success rate for accuracy level  $\pm 1$  mm was 52% and for  $\pm 0.5$  mm accuracy level was 33%.

**Conclusions:** This is an in vitro study, the encouraging results suggest that the technology might be suitable for craniofacial surgery.

#### FP016

##### **3-Dimensional operative simulation improves residents' surgical skills: a randomized trial in pediatric neurosurgery**

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**Objectives:** There has always a challenge in training residents to perform a surgical procedure. Many tools can be used for training. Simulation may be able to provide procedural training. The aim of this study was to evaluate the effect of simulation-based education on the knowledge and skills of neurosurgical residents in the design of the advancement of the fronto-orbital bandeau.

**Materials and Methods:** This was a cluster randomized controlled trial of a simulation-based educational intervention designed to increase residents' skills on a particular aspect of a complex surgical procedure (design osteotomy lines of a fronto-orbital bandeau). Residents were randomized in groups to an intervention group (simulation trained, n = 4) or a control group (traditionally trained, n = 4). These groups were randomly assigned to receive either traditional training and the educational intervention or traditional training alone. Evaluation of residents' will be done by using the Objective Structured Assessment of Technical Skills (OSATS) instrument. Performance assessment between simulator-trained and traditionally-trained residents was compared to measure the effectiveness of the simulation-based intervention. Performance assessment was done by a senior neurosurgeon blinded of the resident randomization

**Results:** There were no differences in the training previously received by the residents. We found a significant difference ( $p < 0.05$ ) in the osteotomy design between both groups with a better performance of the students in the intervention group. There was a trend ( $p < 0.1$ ) in the time used by the resident to draw the osteotomy lines with a shorter time in the intervention group.

**Conclusions:** To our knowledge, this is the first cluster randomized controlled trial of a simulation-based educational intervention designed to increase residents' skills on a particular aspect of a complex surgical procedure in neurosurgery.

Our study proved that when using the right teaching tool, simulation-based education improved residents' training compared to a standard education lecture.

#### FP017

##### **Implementation of 3D modeling to improve understanding and conceptualisation of arteriovenous malformations (AVM) morphology for execution of safe surgical excision of complex pediatric AVMs**

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**Introduction:** A prerequisite to arteriovenous malformation (AVM) surgery is a thorough understanding of lesion anatomy, including feeding arteries, nidus, draining veins, and their spatial relationship with the surrounding brain.

AVMs are rare. In an era where experience with open vascular cases decreases, 3D models help deconstruct AVM morphology, and

provide tactile and visual conceptualization, thereby guiding safe surgical excision of these complex and often unforgiven lesions.

The application of virtual reality technology to neurosurgical training has been reported in recent years. However, the use of 3D printing technology in AVM surgery for pediatric patients has yet to be explored.

Our aim is to evaluate the feasibility of utilizing 3D AVM models in pediatric patients and validate their use intraoperatively.

**Methods:** Skull and vascular models of AVMs in Pediatric patients were constructed in 3D utilizing multimodality imaging: magnetic resonance imaging (MRI), computed tomography (CT), and cerebral catheter angiography. Intraoperative validation of model fidelity was performed using perioperative imaging and post hoc analysis of models with intraoperative photography. Outcomes include clinical and postoperative angiographic results.

**Results:** This approach was utilized in seven consecutive patients. The mean age was 10.4 years old. Four cases were Spetzler-Martin (SM) 4 (Lawton-Young [LY] supplemented 6), two were SM2 LY4, and one was SM3 LY5. Four patients underwent microsurgical excision alone and three microsurgical excisions combined with preoperative adjunctive endovascular embolization. An angiographic cure was confirmed on 6/7 patients, with one patient having a small residual lesion that required Gamma Knife stereotactic radiosurgery. The intraoperative evaluation confirmed excellent fidelity in the printed models to the AVMs.

**Conclusion:** 3D printed models of AVMs in pediatric patients can be constructed with high fidelity and supplement other tools (such as navigation systems) for planning and execution of safe AVM surgery.

#### FP018

##### **Augmented-Reality Telementoring System for EVD Placement Supervision**

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**Introduction:** In cases of trauma or acute hydrocephalus, the placement of an External Ventricular Drain is commonly performed, often by junior residents and even at the bedside. While the procedure is relatively easy and not particularly technical, multiple attempts are sometimes needed before targeting the ventricle. The localization of the entry point and the angle of insertion are critical factors for a successful insertion.

**Methods:** We have developed an (Augmented Reality) AR-based platform that can be used both as a training tool and as a remote telestration tool to allow a consultant to guide a trainee remotely. In a typical Use Case, the trainee would wear an AR headset, and while viewing the patient, their volumetric scan data can be attached using a drag-and-drop to a secure portal where a cloud-based processing pipeline registers, segments, and renders a graphical representation that is overlaid to the patient. The consultant is then able to telestrate on the shared view using fingertip controls on a tablet or mobile phone.

We have tested the platform using our virtual EVD simulator, consisting of a haptic device where the resident, after scrolling through the CT of the patient indicates an entry point and estimates the insertion angle.

**Results:** The system is able to save the session data allowing us to evaluate the speed and accuracy of the targeting. Trainees were able to correctly position of EVD making use of remote supervision.

**FP019****Decreased Need for Anesthesia during Ultra-Fast Cranial MRI in Young Children: One-Year Summary**

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**Purpose:** Rapid volume coverage sequences based on real-time MRI allow for scanning of the entire brain within a few seconds. Movements of children become almost irrelevant due to the ultra-fast acquisition of 30 ms per slice. The adoption of these sequences in a real-time cranial MRI protocol (RT-cMRI) is expected to reduce the frequency of examinations requiring anesthesia in infants and toddlers. The aim of the study was to quantify the reduction in the number of anesthesia examinations in young children after the implementation of the new RT-cMRI protocol.

**Materials and Methods:** All cMRI studies of children up to 6 years in the first 12 months after the establishment of the RT-cMRI 2019/2020 were retrospectively compared to a matched group of the same period in 2017/2018. The frequency of examinations under anesthesia vs. non-sedation examinations was analyzed. In addition, the number of follow-up examinations and the effectiveness of RT-cMRI was determined.

**Results:** The launch of RT-cMRI led to a significant decrease in the proportion of cMRI under anesthesia from 92 % to 55 %. Only 2 % of the RT-cMRI failed and required conventional MRI under sedation in the follow-up. The speed and ease of use of RT-cMRI increased the number of follow-up examinations from 1.3 to 1.4 examinations per child.

**Conclusion:** This innovative real-time MRI examination allows a drastic reduction in the number of studies under anesthesia for suitable cranial pathologies in children under 6 years. However, cautious selection of indications as well as adjustments to the workflow in the radiological department are required.

**Key points:** · Real-time MRI sequences are almost unaffected by patient movement. · The application of real-time cranial MRI can spare children from sedation. · Low-threshold access results in more frequent follow-up examinations.

**Abstract Session 4: Trauma/Vascular****FP020****Preliminary Results of the Dutch Pediatric Craniectomy and Cranioplasty Registry (DPCCR) - A Prospective Multicenter Open Registry**

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**Objective:** Based on existing literature, consensus cannot be reached regarding optimal timing, implant material and peroperative management of children undergoing cranioplasty following craniectomy. To overcome this, systematically collected data are necessary. Moreover,

the importance of surveillance of implants has been imposed upon by governmental regulations. Therefore, we established the Dutch Pediatric Craniectomy and Cranioplasty Registry (DPCCR).

**Methods:** The DPCCR is a prospective multicenter registry that started December 2019 and includes academic hospitals with a pediatric neurosurgical service in the Netherlands. Patients are eligible for inclusion if they are younger than 18 years and undergo cranioplasty following craniectomy. Data is collected at time of craniectomy, cranioplasty and follow-up. Follow-up takes place 6 weeks and 1 year after cranioplasty, and every 2 years until age 18. Then, the patient fills in a questionnaire regarding cosmetic satisfaction and self-esteem. The DPCCR was approved by our institution's Medical Ethics Research Committee (MERC) and all patients provided informed consent.

**Results:** As of December 2022, 8 patients have been included (6 males, mean age  $9.1 \pm 5.6$  years). Indications for craniectomy were trauma (62.5%), vascular disease (25.0%) and Langerhans cell histiocytosis (12.5%). Mean cranial defect size was  $83.5 \pm 31.3$  cm<sup>2</sup> (range 32.0 - 122.1). Mean time interval between craniectomy and cranioplasty was  $154.3 \pm 77.7$  days (range 59 - 240). Glass fiber-reinforced composite was used in 6 patients (75.0%) and polymethyl methacrylate in 1 patient (12.5%). One patient has not had a cranioplasty implant yet. In two patients (25.0%) a cerebrospinal fluid drain was inserted. Mean skull circumference after cranioplasty was  $52.5 \pm 3.8$  cm. The mean follow-up after cranioplasty is  $1.30 \pm 1.04$  years.

**Conclusion:** The DPCCR aims to answer clinically relevant questions regarding cranioplasty following craniectomy in children by collecting long-term, high-quality and uniform data using a prospective multicenter approach.

**FP021****Reconstructive cranioplasty after large craniectomies: a retrospective evaluation of clinical outcome**

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**Introduction:** Cranioplasty after large craniectomy is mandatory to protect the brain and to restore the physiological conditions for CSF dynamics and brain perfusion. Main factors affecting the cranioplasty outcome in pediatric population are the growing skull, the lower bone thickness and the high incidence of CSF disorders or brain swelling. No consensus on the ideal material has been reached. Autologous bone has been the "gold standard" but has significant incidence of side effects such as bone resorption and infection.

**Methods:** A retrospective review of all pediatric patients from 2002 to 2020 undergoing a large cranioplasty at a single institution (HUS) was performed.

**Results:** Over the 19-year period, 16 cranioplasties were done. 11 were males and median age at cranioplasty was 13 years (range 2-16 yrs). Initial etiology was trauma in 14, subarachnoid hemorrhage in 1, and large benign extra-axial tumor in 1. 12/16 had history of hemicraniectomy, 3 with bifrontal hemicraniectomy and 1 with large frontotemporal craniectomy. The median size of the craniectomy was 102,5 cm<sup>2</sup> (range 85-130cm<sup>2</sup>). 13/16 were primary operations and 3 secondary after bone resorption.

10 cranioplasties were done with autologous bone. 4 had to be replaced after bone resorption, giving the rate of resorption as high as 40%. 1 had to be removed 112 days later because of the enterococcus fecalis infection, giving the rate of infection 10%.

As an alloplastic implant, 3 had a Glace bioglass implant, 2 had PEEK implant and 1 PE implant. None of these 6 got infected nor had to be replaced for other reasons. 1 patient who had a hydrocephalus already before cranioplasty surgery had a VP-shunt after cranioplasty. 3 patients had a VP-shunt done before autologous bone cranioplasty, but 2 of them were removed during the cranioplasty surgery.

**Conclusion:** Alloplastic implants are a good choice for large pediatric cranioplasties.

#### FP022

##### Effects of barbiturate coma on intracranial pressure and compliance in children with traumatic brain injury

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**Introduction:** The aim was to study how barbiturate coma treatment (BCT) affected intracranial pressure (ICP) and intracranial compliance (RAP index) in children (<18 years of age) with severe traumatic brain injury (TBI) and refractory intracranial hypertension (RICH).

**Methods:** High frequency ICP and mean arterial pressure (MAP) monitoring data were collected during neurointensive care of children who developed RICH due to TBI. Cerebral perfusion pressure (CPP) and RAP index (ICP amplitude/ICP, a surrogate marker of compliance) were calculated. To study the temporal effects four half hour long periods were analyzed: before bolus injection and at 5, 10, and 24 hours thereafter, respectively, and a fifth tapering period with S-thiopental between <100 and <30  $\mu\text{mol/L}$ . S-thiopental concentrations and administered doses were registered.

**Results:** Seventeen children (2007-2017) were included; median age 15 (range 6-17) and median Glasgow coma motor score 5 (range 2-5). Median time from trauma to start of BCT was 44.5 h (range 2.5-197.5) and from start to stop 99.0 h (range 21.0-329.0). Median ICP was 22 (IQR 20-25) in the half hour period before onset of BCT and 16 (IQR 11-20) in the half hour period 5 h later ( $p=0.011$ ). The corresponding figures for CPP were 65 (IQR 62-71) and 63 (57-71) ( $p>0.05$ ). The RAP index was in the half hour period before onset of BCT 0.6 (IQR 0.1-0.7), in the half hour period 5 h later 0.3 (IQR 0.1-0.7) ( $p=0.331$ ) and in the whole BCT period 0.3 (IQR 0.2-0.4) ( $p=0.004$ ). According to Glasgow outcome scale at six months, 14/17 children (82%) had favorable outcome (good recovery=8 patients and moderate disability = 6 patients).

**Conclusion:** BCT due to RICH significantly reduced ICP and RAP index (improved compliance) with preserved CPP. Clinical outcome was relatively favorable using BCT for RICH.

#### FP023

##### Cerebral pressure autoregulation and optimal cerebral perfusion pressure during neurocritical care of children with traumatic brain injury

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**Introduction:** The most appropriate cerebral perfusion pressure (CPP) during treatment of children with traumatic brain injury (TBI) is not completely elucidated since normal blood pressure is age dependent and the role of cerebral pressure autoregulation (CPA) is unclear. The aims were to examine PRx (pressure reactivity index), CPP, CPPopt (optimal CPP), deviations from CPPopt ( $\Delta\text{CPPopt}$ ) in a series of children with severe and moderate TBI, and relate the findings to age, time after trauma and clinical outcome.

**Methods:** Intracranial pressure (ICP) and mean arterial pressure (MAP) monitoring data were collected during neurointensive care of 57 TBI children <18 years. CPP, PRx, CPPopt and  $\Delta\text{CPP}$  (actual CPP - CPPopt) were calculated. Glasgow Outcome Scale (GOS) was assessed 6 months after injury and dichotomized into favorable outcome (GOS 4-5) and unfavorable (GOS 1-3).

**Results:** The median age was 15 (range 0.5-17) and median Glasgow coma scale motor score 5 (range 2-5) at admittance. 49/57 (86%) had favorable outcome. There was a tendency for the whole group that lower PRx (better preserved CPA) was associated with more favorable outcome ( $p=0.052$ ). This finding was statistically significant in children <15 years ( $p=0.016$ ) but not in children >16 years ( $p=0.528$ ). In children < 15 years, lower proportion of time with  $\Delta\text{CPPopt}<-10$  was significantly associated to favorable outcome ( $p=0.038$ ) but not in the older age group. Temporal analysis indicated that PRx was higher (more impaired CPA) from day 4 and CPPopt higher from day 6 in the unfavorable outcome group compared to the favorable outcome group, although those findings were not significant.

**Conclusion:** Impaired CPA is related to poor outcome, particularly in children <15 years of age. In that age group, actual CPP below the optimal CPP level contributed significantly to unfavorable outcome. CPPopt appears to be higher during the time period when CPA is most impaired.

#### FP024

##### Brain arteriovenous malformations in children – institutional experience from two decades of treatment

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**Introduction:** Treatment of brain arteriovenous malformations (bAVM) in children is still challenging. The objective of bAVM obliteration and reducing the life-long risk of haemorrhage must be outweighed by the acceptable risk associated with treatment. The long-term outcome data are scarce, and the optimal treatment strategy is not firmly established.

**Methods:** We reviewed clinical and radiological data from children treated for bAVM from 2000 to 2022. The clinical outcome data, radiological obliteration rate, complication rate and incidence of de novo lesions were the primary follow-up parameters.

**Results:** Data from 86 consecutive patients were included (46 boys and 40 girls, mean age  $13.3\pm 4.5$  years). Most patients harboured Spetzler Martin Grade 1 bAVM (20%), Grade 2 (35%) or Grade 3 (31%). 13% of bAVMs were located in the posterior cranial fossa. 54% of children presented with intracerebral haemorrhage, while another 6% experienced bleeding during the observation or ongoing treatment; 14% were asymptomatic. First-line treatment was microsurgical in 40% (out of which acute removal in 44%), radiosurgical in 26%, endovascular in 15% and conservative (including removal of haemorrhage only) in 19%. Combined multimodal treatment was necessary in 51% of cases. The obliteration rate was 68%; the frequency of de novo lesions was 4%, occurring 3-7 years after the completed treatment (long-term data available from 74 patients). A favorable clinical outcome (modified Rankin Scale score 0-3) was achieved in 92% of children, while 5% died due to haemorrhage. Despite frequent haemorrhagic presentation and complex pathology requiring multimodal treatment in over half of the cases, favorable clinical outcomes may be expected in the majority of patients. In our experience, microsurgery and radiosurgery result in the highest obliteration rates, while the role of endovascular treatment is only supplementary in a few selected cases. The study also confirmed de novo formation of bAVM after documented obliteration.

#### FP025

##### Outcome assessment by single photon emission computed tomography in patients with Moya Moya disease

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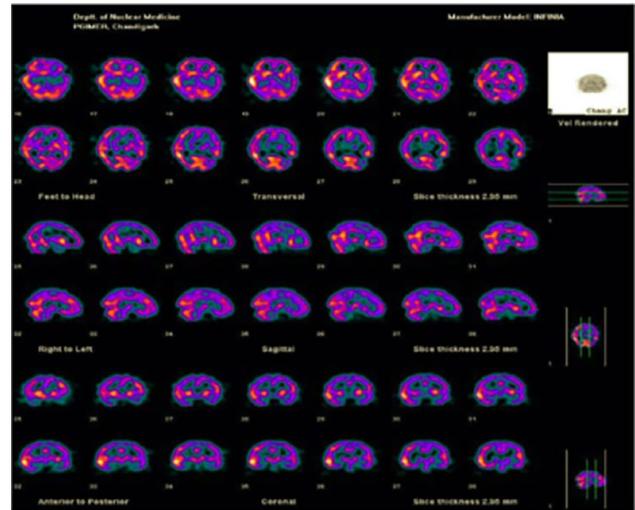
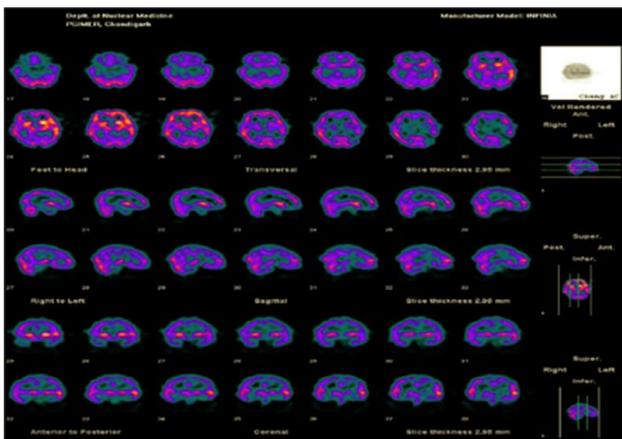
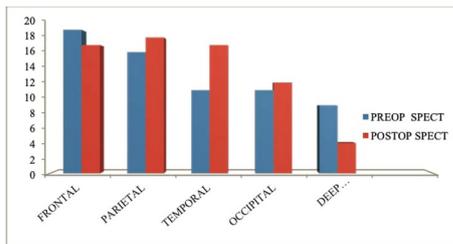
**Introduction:** Moyamoya disease is chronic progressive cerebrovascular disease involving bilateral stenosis or occlusion of terminal portion

of internal carotid artery and proximal portion of anterior cerebral artery and middle cerebral artery. The purpose of the study was to assess brain perfusion on SPECT in preop period. The area of hypoperfusion was then compared on postop SPECT thus assessing the functional usefulness of revascularization procedures.

**Method:** This was an ambi directional observational study .22 patients were enrolled in the study. The hospital records were used for analysis and patients were contacted and followed up for 6 months. The pre and post op SPECT findings were compared and correlated with clinical outcome.

**Results:** 22 patients of moya moya disease who underwent revascularization procedure were included. Results STA MCA anastomosis along with EDAMS was done in 21 patients and EDAMS alone was done in one patient. Post op clinical improvement was seen in 63.6 % of patients and no improvement in 36.3 % of patients. It was found that 63.6 % patients showed improvement in clinical outcome and there was no improvement in 36.3 % patients. All patients underwent SPECT after three months of revascularization procedure and it was observed that 90.9% patients had perfusion defect on left side, 77.3% patients had on the right side and 68.1% patients had bilateral perfusion defects. 59.1% patients had improvement on SPECT postoperatively. There was statistically significant improvement in right anterior circulation and when analyzed further statistically significant improvement was noted in right parietal region.

**Conclusion:** This study demonstrates that perfusion defect in moyamoya disease involve both anterior and posterior circulation unilaterally or bilaterally which is better delineated in perfusion imaging such as SPECT compared to anatomical imaging modalities. Our study proves that surgical revascularization which consisted of STA–MCA anastomosis and indirect synangiosis improved perfusion in anterior circulation.



**FP026**

**Multiple Burr-Hole Surgery for treating Moyamoya in Children**  
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**Objective:** Moyamoya disease (MMD) is characterized by progressive spontaneous occlusion of distal internal carotid arteries and their main branches. It is necessary to promptly diagnose and treat this condition, especially in children, because of the high risk of stroke and consequent severe disability. In this study, we examine the role of multiple burr-hole surgeries (MBHS) in treating pediatric patients with MMD and Moyamoya-like.

**Methods:** Prospective collection of the clinical/radiological data of a consecutive series of pediatric patients with symptomatic MMD/ Moyamoya-like who underwent at least one MBHS surgery since 2009.

**Results:** Thirty-eight MBHS procedures were performed in 28 children. MMD was diagnosed in 7 patients. Moyamoya syndrome was associated with NF1 in 10 patients, with Down Syndrome in 3 patients, with radiotherapy for OPG in 5 patients, and with a rare disease in 3 patients (PHACE syndrome, Encefalocraniolipomatosis, and Lenz Majewski ) Clinical and radiologic follow-up for all patients ranges from 3 months to 13 years. One patient had a massive stroke in the postoperative period. Other 2 patients experienced transient ischemic attacks during the follow-up and underwent additional revascularization surgeries. Postoperative digital subtraction angiography showed good revascularization around the burr-hole sites in all patients. The functional efficacy of the surgery was confirmed by angiography and perfusion MRI in 25/28 patients during follow-up.

**Conclusions:** Our data underline the excellent rate of success of MBHS in pediatric MMD and quasi-MMD. This technique results in good collateral revascularization, improved cerebral perfusion, and excellent short and long-term symptom control, with low perioperative risk. Postoperative changes in perfusion parameters and ivy sign MRI findings seem to show a positive correlation.

## FP027

**Intracranial aneurysms in the pediatric population. Analysis of a series of cases**

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**Objective:** Pediatric cerebral aneurysms are rare and differ from their adult counterparts. Data on the long-term follow up after treatment are lacking in the literature. We present our retrospective 35-years series focusing on the outcome.

**Methods:** A retrospective database of 15 pediatric patients with intracranial aneurysms managed in our unit between 1988 and 2021 was reviewed. Patient medical records were reviewed for: age, clinical presentation, type and location of aneurysm(s), surgical approach, complications, clinical and angiographic outcomes.

**Results:** Fifteen patients with 16 aneurysms with male predominance (8 male patients and 7 female patients; age range, 3 years to 17 years), were admitted with subarachnoid hemorrhage (60%), infectious cause (6%) stroke (18%), traumatic subarachnoid hemorrhage (6%), and incidental aneurysm (10%). Anterior circulation location was seen in 93% patients with middle cerebral artery aneurysm predominance (36%). An equal number of patients presented fusiform or saccular type (45% each), two patients presented with giant aneurysm and 1 patient with mycotic type. Aneurysm sizes ranged from 3 to 26 mm. Most aneurysms were clipped directly (80%). Eighty five percent of the patients were independent at follow-up. There was one death post endovascular coiling technique. The long-term morbidity rate was 20%, and the mortality rate was 9%. Clinical outcome was favorable in 81%. the mean angiographic follow-up was of 48 months. Only one patient treated by the endovascular technique showed progression.

**Conclusion:** Pediatric aneurysms require complex microsurgical techniques to achieve favorable outcomes. A team of skilled neurovascular radiologist and surgeon are required to achieve good outcomes. In children, microsurgical treatment may be better than endovascular management. We advocate at least one angiographic follow-up at five-years, with angio-MRI each year.

## FP028

**Supratentorial cavernous malformations in children - indications for surgery and analysis of outcome**

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**Introduction:** Cerebral cavernous malformations (CMs) are most commonly localized in the supratentorial compartment. The initial presentation may be due to hemorrhage, epilepsy and/or focal neurological deficit, but a considerable group of these malformations is diagnosed accidentally.

**Methods:** A retrospective analysis of patients who underwent surgical treatment for supratentorial CMs in the Department of Pediatric Neurosurgery in Katowice between 2010 and 2021 was performed.

**Results:** Study population consisted of 22 patients (7 girls and 15 boys, average age: 11 years). The most common initial symptoms were seizures (n=12), followed by focal neurological deficits (n=7),

headaches (n=6), nausea/vomiting (n=5), and impaired consciousness (n=4). In 7 cases resected CMs were localized in temporal lobe. Other localizations included frontal (n=6), parietal (n=6) and occipital lobes (n=2), but in one patient CM was situated in basal ganglia. Three patients had multiple CMs. Patients were qualified for surgical removal of CMs due to symptomatic hemorrhage (n=10), drug-resistant epilepsy (n=6) and progressive neurological deficits (n=3). Three children were asymptomatic. Control MRI confirmed a complete resection of the CM in all but one case (95.5%). The mean follow up was 48 months. In studied population no patient experienced rebleeding from CM. None of the patients with CM bleeding associated with first time seizure (n = 6) had developed an epilepsy in the postoperative period. All patients who were operated due to long-lasting drug-resistant epilepsy (n=6) had experienced an improvement (complete lack of seizures in 4 cases).

**Conclusion:** Surgical treatment of supratentorial CMs is an effective method of preventing recurrent bleeding from the lesion and dealing with the concomitant epileptic seizures.

**Abstract Session 5: Hydrocephalus I**

## FP029

**Evaluation of long-term distal shunt catheter failures in pediatric hydrocephalus patients**

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**Introduction:** Ventriculoperitoneal shunt implantation carries complications, including distal shunt catheter ruptures (2.1-15.3%). These may be particularly frequent in growing children and due to possible subcutaneous calcifications. Aim of this work is to analyze the occurrence of distal catheter alterations and disconnections in different catheter types.

**Patients and Methods:** In the time-period between 4/2009-12/2022, 429 shunt surgeries were performed in 293 individuals. Patients bearing barium impregnated catheters (Miethke-Aesculap, Germany/ Bactiseal, Codman, Integra, USA) or barium striped catheters (Miethke-Aesculap, Germany/ Medtronic, USA) with at least 5 year follow-up were included. Revision cause and survival rate of distal catheters as well as radiographic alterations were evaluated.

**Results:** Barium impregnated catheters were used in 258 individuals (mean age: 26.4±4.3 months) and barium striped catheters were implanted in 35 patients (mean age: 62.7±5.9 months) with a total follow up time of 13.5 years. In radiographic evaluation, no alterations were found in barium striped, while barium catheters showed a secondary thickening (>2.5mm) of catheter diameter in 44/215 cases (20.5%). The distal barium impregnated catheters were subjected to 72 revisions (tensions/radiographic calcifications, (n=23), disconnections (n=15), others (n=33)). The barium striped group underwent 7 revisions (no tensions, disconnections (n=2), and others (n=5); p=0.09). Overall survival rate of distal barium striped catheters at 10 years follow up was 53.6% and 64.3% in barium impregnated catheters. Acknowledging disconnections and tension related revisions only the survival was 54.2% and 94.1%, respectively.

## FP030

**Ventriculo-Sinus Shunts (VSS): The Role in Selected Patients**

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**Objective:** Ventriculoperitoneal shunts (VPS) are the most common treatment for hydrocephalus. However, most patients will require

multiple shunt revisions over time, usually distal. Nevertheless, there have not generally been better alternatives for peritoneal placement; the pleura and heart are suboptimal locations, and often fail. Sinus shunts have been described yet are not commonly used. The potential vascular complications, as well as inexperience with these shunts, make them less attractive. The goal of this study is to share our experience with VSS in selected patients.

**Methods:** Between December 2018 and February 2022, a total of 7 patients (1 adult, 6 children) underwent 11 surgeries for placement of a VSS at our center. Data regarding complications and shunt function were collected.

**Results:** Age at surgery was 1.3 - 38years ( $11.9 \pm 12.8$ ). All patients had previous shunt systems that had failed, and had systemic conditions that made other distal sites less desirable. Both retrograde and antero-grade techniques were used. In all cases, a low-pressure differential valve was installed. Over a follow-up period of 1 - 26 months (mean  $10 \pm 7.5$ ), 4 of 7 patients underwent additional shunt revisions, of which 2 were placed to other sites. One patient had symptomatic partial sinus thrombosis, managed conservatively. During follow-up, five patients showed improvement in the symptoms of high intracranial pressure using the VSS. There were no severe complications of air embolism, bleeding, or infection.

**Conclusions:** VSS may have a role to play for selected patients who have failed more commonly used distal shunt locations. Significant complications are rare. Additional experience is needed to better understand the ideal catheter and placement locations.

### FP031

#### Retrospective Study comparing Ultrasound-guided VPS placement and Freehand insertion

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**Introduction:** Ventriculoperitoneal shunt (VPS) insertion is a common neurosurgical procedure. In this retrospective cohort study, we primarily aim to evaluate the accuracy of VPS placement under ultrasound (US) guidance and freehand technique. Our secondary purpose is to evaluate the learning curve for surgeons to adopt the method of US-guided VPS insertion.

**Method:** This study followed patients who underwent VPS placement over the period of January 2018 to December 2021 at a single-centered tertiary hospital, the National University Hospital of Singapore. This study involved both the adult and paediatric populations. Patients who underwent VPS placement via neuro-navigation were excluded. Accuracy of shunt placement were determined by evaluating tip of the shunt catheter in the first post-operative imaging and the shunt revision rate. Shunts that were revised within 30 days were likely to be attributed to sub-optimal placement. The relevant data for the study will be collected using the hospital's database (de-identified Neurosurgery VPS audit). Data collected will be analysed using R software version 4.2.0.

**Results:** Our study data includes 223 patients, of which 45 patients underwent US-guided VPS insertion. Rates of optimally placed and functioning shunts were higher using US-guidance (79.5% vs 50.6%,  $p = <0.01$ ) compared to freehand insertion. There were no shunt revisions indicated within 30 days for those in the US cohort, compared to 5.62% ( $p = 0.18$ ) in the freehand cohort. In the univariate analysis for potential confounders, none of the factors demonstrated significant relationship with accuracy of shunt placement. Accuracy of placement in the US cohort was comparable between residents and consultants.

**Conclusion:** To our knowledge, this study is the first of its kind to be conducted in Singapore. The results of this study suggest that patients

with US-guided VPS insertion can eventually have better outcomes and provide insight into the training of future neurosurgeons.

### FP032

#### Symptomatic chronic overdrainage

#### The role of high-pressure adjustable valves and gravitational devices

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**Introduction:** Symptomatic chronic overdrainage (SCO) is extremely difficult to treat and recommended sequenced strategies can be risky for patients. Admitted first recommendation is upgrading the valve and in refractory cases shunt removal protocols or cranial expansion procedures have been proposed. We present our experience with the placement of programmable high-pressure valves (HPV) (over 200 mmH2O) and gravitational devices (GD) in refractory SCO.

**Methods:** Eighteen children of 302 paediatric patients shunted between 2000 and 2022 at our institution were treated with HPV and GD between 2018-2022. Variables related to patient characteristics, hydrocephalus aetiology, HPV response peculiarities and follow-up were recorded and a descriptive analysis of the data was performed.

**Results:** Post-hemorrhagic hydrocephalus was the most frequent aetiology and most of the patients were shunted at first months of life. Seven of 18 patients treated with HPV and GD had previously undergone failed shunt withdrawal protocols and one of them had also been treated with cranial expansion and posterior fossa decompression. Shunt open pressures previous to HPV were around 200 mmH2O and GD were between 0/20-0/30. HPV were routinely set at 150 mmH2O with increased GD resistance. During the follow-up, 8/18 patients required programming changes due to overdrainage (6/18) or underdrainage (2/18) One patient required change of ventricular catheter with neuronavigation. Refractory SCO was successfully treated in 17/18 patients with open pressures of 150-220 mmH2O and GD between 0/30-0/35. Reservoir collapse and radiological ventricular size improved in most of them.

**Conclusion:** The use of HPV and GD for the treatment of refractory SCO was favourable in most cases, managing to control the symptoms and improve radiological collapse.

### FP033

#### Effect of antisiphon devices on ventriculo-peritoneal shunt drainage dynamics in growing children

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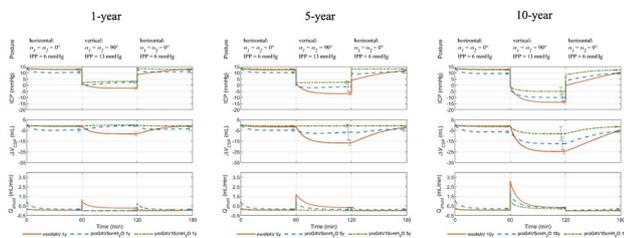
**Introduction:** Infants and small children face changing boundary conditions when treated with a ventriculo-peritoneal shunt (VPS) for hydrocephalus. There are no systematic data describing shunt drainage-behavior in a growing child. This study investigates with a child-adapted patient-simulator the drainage behavior of fixed differential-pressure (DP) valves and adjustable valves with overdrainage protection mechanisms at different ages.

**Methods:** Three DP valves (Miethke MiniNAV, 10 cmH2O; MN10) and three adjustable Miethke ProGAV2.0 valves with a 25 cmH2O gravitational unit (GU) at low (5 cmH2O; PG5) and medium opening

pressure (10 cmH<sub>2</sub>O; PG10) were investigated in a hardware-in-the-loop test bed. Body orientation and length were set according to child's age. The software simulated the physiological situation of a one-, five- and ten-year-old child. All valves were tested per specification with five runs for 1-h each in horizontal, vertical and horizontal positions. Intracranial pressure (ICP) and VPS flow were measured, and respective CSF volume changes and ICP set value computed.

**Results:** Drainage parameters in vertical position increased with age in all valves, pronounced with MN10, here already at 1-y. GUs in PG5 and PG10 substantially reduced drainage compared to MN10. In the 1-y and 5-y setups, a PG10 prevented drainage, but there was some drainage at physiological ICPs in the 10-y setup. In contrast, MN10 produced the largest ICP drops across all ages and positions, and overdrainage resulting in insufficient ICP recovery in the subsequent horizontal position. ICP levels were most constant with PG10 at all ages.

**Conclusions:** This study shows that unprotected DP valves might lead to overdrainage already in infants, while low-pressure GU valves can prevent overdrainage until 5y and medium-pressure GU valves admit physiological pressures until at least 10y. Therefore, overdrainage preventing mechanisms should be included in the first shunt implanted and opening pressures should be adjusted with the child's growth.



#### FP034

##### Influence of gender and pathology on development of papilledema as opposed to point-of-care ultrasound techniques in case of raised ICP requiring treatment in children

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**Objective:** To analyse if gender and type of pathology influence the development of papilledema and compare fundoscopic and point-of-care-ultrasound techniques (optic nerve sheath diameter (ONSD) and optic disc (OD) elevation) to detect pathologies with ICP increase in children.

**Methods:** 72 children were included; 46 male, 26 female; 50 with proven pathology (e.g. pseudotumor cerebri, tumor, hydrocephalus), 22 with pathology excluded. Bilateral ONSD and OD were quantified by US using a 12 MHz linear array transducer. This was compared to ophthalmoscopic optic disc findings and in some patients with invasive ICP values.

**Results:** In patients with proven pathology, significant more girls (69%) had fundoscopic papilledema compared to boys (37%,  $p < 0.05$ ). Girls had also larger US-OD values ( $0.88 \pm 0.36$  mm vs.  $0.65 \pm 0.4$  mm in boys). US ONSD revealed no gender-specificity. In tumor patients 80% had initial papilledema (100% girls, 79% boys), compared to 50% in pseudotumor cerebri (PTC) (83% girls, 30% boys). Initial US OD was larger in tumor compared to PTC. ONSD had no pathology specificity.

**Conclusion:** Papilledema seems to be a gender- and pathology-specific feature, whereas US ONSD is not. Girls rather seem to develop papilledema under similar conditions. Male sex and PTC are risk factors to remain undetected using

ophthalmoscopic OD findings for diagnostics. US ONSD and OD can be useful additions to detect pathologies with possible raised ICP in children.

#### Abstract Session 6: Posterior Fossa Oncology I

##### FP035

##### Treatment of cerebellopontine angle tumors in infants and children: case series and literature review

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**Introduction:** About 1-3% of all pediatric intracranial tumors is represented by Cerebellopontine-angle (CPA) and cerebellomedullary fissure (CMF). The management of these lesion is still controversy in literature, without consensus of surgical approach. We retrospectively reviewed our series and performed a wide literature analysis, with the aim of describe clinical presentation, surgical approach, correct timing of hydrocephalus treatment and adjuvant therapy planning.

**Materials and methods:** During the period 2007-2022, 28 children with CPA-CMF were treated in the pediatric neurosurgery department of "Santobono-Pausilipon" Children's Hospital of Naples. Medical records, histology features, operative approach and outcome were retrospectively collected and analyzed. A systematic review of English-language articles published from 2010 to 2021 was performed, pooling patients baseline characteristics and outcomes. After screening 274 articles, 53 describing 301 patients (331 surgical procedures) were included.

**Results:** There were 17 males and 11 females (mean age  $6,3 \text{ y} \pm 5,3 \text{ y}$ ). 15 out of 29 lesions were low-grade tumors (dermoid, meningiomas, and neurinomas), while 13 were high-grade (AT/RTs, GBM, medulloblastomas, ependymomas, choroid plexus carcinomas). Hydrocephalus was present in 60,7% of patients, and was treated before the tumour resection in 35,7% of child (7 DVP, 5 ETV and 3 EVD). A lateral suboccipital approach was performed in 41,3% of cases, a midline suboccipital in 23,9%; a retrosigmoid in 23,9% and a subtemporal in 10,8. 53,6% of child had 1 surgery; 28,6% 2 procedures; 3,6% 3 surgery and 14,3% more than 3, with a 71,4% of GTR. 12 patients developed a recurrence of disease and 9 child died in a mean follow-up period of 37,4 months.

**Conclusion:** Adequate choice of surgical approach is necessary to achieve GTR and a satisfactory outcome in CPA-CMF tumors. GTR in the mainstay of treatment in low-grade tumors, while prognosis in high-grade lesions, although after gross total resection, remains unfavorable.

##### FP036

##### National multicentered retrospective review of salvage therapy for pediatric medulloblastoma: A Canadian Experience

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**Background:** Children diagnosed with medulloblastoma (MB) that are refractory to upfront therapy or experience recurrence have very poor prognoses. Though phase I and II trials exist, these treatments bear significant treatment-related morbidity and mortality.

**Methods:** We retrospectively reviewed children diagnosed with treatment refractory or recurrent MB from 2000–2020 across 5 Canadian Pediatric Neurosurgical Centers.

**Results:** 137 patients were identified where 135 patients had sufficient data for analysis. The mean age of first operation was 7.09 years with 17.8% (n=17) presenting <3 years of age with a male predominance (64.4%). The upfront therapy provided was GTR in 65.2% and debulking in 29.6% followed by autologous stem cell transplant (34.1%), chemotherapy (100%) and radiotherapy (72.6%). The clinical course was no disease progression (64.4%), initial response to therapy with recurrence (22%), progressive disease prior to finishing adjuvant therapy (4.4%), treatment refractory progressive disease (5.2%) with the majority a local recurrence (53.5%) followed by leptomeningeal dissemination (46.7%) at a mean time interval of 18.5+11.3 months. Forty-five children (33.3%) received salvage therapy at disease progression as re-resection (20%), steroid (35.6%), chemotherapy (77.8%), radiotherapy (40%). Of the 45, 9 progressed a second time at an interval of 19.7+23.0 months with children received a second salvage regimen with 6 neurosurgical procedures (Biopsy: 6, GTR: 1) with steroid administration (85.7%) and palliative radiation (22.2%). The 5-year overall survival for the cohort was 69.1%.

**Conclusion:** Recurrent MB carries a devastating prognosis despite the administration of salvage therapy.

#### FP037

##### **Patterns and Outcomes in Relapsed Paediatric Intracranial Ependymoma: A Single Centre 20-year Experience**

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**Introduction:** Ependymoma is the third most common paediatric brain tumour. High rates of relapse are reported, often at a late stage. Initial management is primarily surgical, aimed at Gross Total Resection (GTR), typically followed by radiotherapy. GTR has been reliably associated with favourable outcome, however other factors are unknown. Here we report outcomes from our tertiary unit.

**Method:** We retrospectively reviewed all intracranial paediatric ependymoma patients treated between 2000–22. We assessed patient, disease and treatment related factors associated with relapse and overall survival.

**Results:** A total of 30 patients were identified over a 20-year period. 58.3% were male with a median age of 5 years at diagnosis. Infratentorial tumour location was commonest (62.5%), with only 1 patient found to have drop metastasis at presentation. All patients were primarily managed surgically with attempted GTR, 79.2% achieving this after one surgery, a further 12.5% achieving this with a 'second-look' surgery within 10 days.

Overall relapse rate was found to be 65%. The median time to initial relapse was found to be 699 days. 26 redo operations across this subgroup were performed. The median interval between subsequent relapses reduced sequentially. 73.9% of relapses occurred local to the initial tumour location.

Overall mortality was 25%. The median interval from initial operation to death was 1602 days. Higher rates of hydrocephalus at initial presentation, subtotal resection at initial surgery and transformation to higher grade were found in the deceased group. The time from initial surgery to repeat surgery was close to half in this subgroup compared to overall average, 655 and 1170 days respectively.

**Conclusion:** Long-term survival in this cohort of patients is poor with high rates of relapse despite good rates of GTR. Those with subtotal resection, hydrocephalus at presentation and a shorter interval to relapse appear to have a less favourable outcome.

#### FP038

##### **Venous Air Embolism during Semisitting Position in Pediatric Posterior Fossa Tumour Surgery: A Single Center Retrospective Study**

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**Introduction:** Literature is inconclusive about sitting position in pediatric posterior cranial fossa tumor (PCF) surgery and the associated incidence of significant venous air embolism (VAE) and subsequent hypotension.

**Methods:** Clinical data, charts, neuroradiological images, intraoperative notes of 68 children harboring a PCF tumor operated at our Hospital from 2008 to 2022 were collected.

Clinically significant VAE was defined by experienced neuroanesthesiologists as a decrease in endtidal CO<sub>2</sub> levels by more than 4 mmHg, associated with a characteristic sound in the thoracic Doppler, and severe related complications (decline of pO<sub>2</sub> or hypotension).

**Results:** 64/68 (94%) was operated in the semisitting position, 4 patients were operated in prone position. Mean age was 8,12years. 27 children were under 8 and only two children, were under 1 year. All patients were preoperatively screened for patent foramen oval with Transcranial Doppler. We found an overall incidence of clinically significant intraoperative VAE of 14%. The surgery was completed in all cases. A change in position, was required in one case. 32 / 68 (50%) patients were extubated early after surgery. No pneumocephalus required intervention. 4/68 (5.8%) required reintubation. The median duration of ICU and hospital stay was respectively 2.7 and 16 days. Definitive histology was obtained in 67 out of 68 patients: 31 pylocytic astrocytoma; 10 ependimomas (grade II/III); 8 Medulloblastomas; 2 DIPG; 2 dermoid/epidermoid Cysts; 2 Embryonal Tumors with Multilayered Rosettes; 2 Atypic Theratoid Rhabdoid Tumors; 1 ependimoblastoma; 1 High Grade Glioma; 1 Haemangioblastoma; 1 Schwannoma; 1 Extraventricular Neurocytoma; 1 Papillary Tumor; 1 Ganglioglioma; 1 Germinoma; 1 Pineocytoma; 1 Mature Teratoma

**Conclusions:** Semisitting position could be used in pediatric PCF tumors if appropriately chosen. Prospective studies are lacking. Key points to consider in children are different body proportions, large head and short neck, pins size, variability in anatomical and functional closure of oval foramen.

#### FP039

##### **Sitting position for posterior fossa lesion surgery: technical considerations and position-related complications in a retrospective monocentric series of 210 patients**

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**Introduction:** Surgery for posterior fossa (PF) tumors in sitting position is reported to be challenging, because of the risk of air embolism and related morbidity and mortality, especially in the pediatric population. Undeniable advantages include lower intraoperative bleeding, decreased intracranial pressure and reduced need for cerebellar retraction. We analyzed position-related complications in a cohort of patients undergoing posterior fossa surgery in sitting position.

**Methods:** All patients undergoing PF surgery in sitting position in the last 5 years in Necker Enfants Malades Hospital were included in the present study. Patient files, operative reports and imaging studies were analyzed to extract the rate and the gravity of position-related complications.

**Results:** 210 consecutive patients were included, with a median age of 4 years-old (range 0–16 years-old). Echocardiography was not performed systematically to exclude patent foramen ovale before surgery. Surgical procedures were performed for fourth ventricle tumors (72%) via telovelar approach, vermian and cerebellar hemispheric tumors (20%), and pineal region lesions (8%) via supracerebellar infratentorial approach. Military anti-shock trousers were used to rise venous pressure. Seven (3.3%) cases of air embolism occurred, diagnosed through perioperative hypoxemia. Three (1.4%) of them induced visible lesions on postoperative MRI and one (0.5%) was also clinically relevant. Postoperative pneumocephalus (subdural or intraventricular) was a common finding, and it led to delayed extubation and drainage in 5 cases. One case of transitory peripheral nerve paralysis (external popliteal sciatic nerve) was noted. No position-related death was observed.

**Conclusion:** In highly specialized centers with dedicated anesthesiologists and trained surgeons, sitting position can be performed routinely, without major morbidity and mortality.

#### Abstract Session 7: Posterior Fossa Oncology I / Epilepsy

##### FP040

##### Tumor sample collection as a quality indicator in a certified pediatric oncology center – the neurosurgical perspective

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**Objective:** Pediatric brain tumors require histological and molecular diagnostics to reach an integrated diagnosis. Molecular data is used for risk stratification of current therapies. Tumor samples are further needed for patient-specific experimental approaches. Thus extensive collection of samples is important, but requires substantial infrastructure and staffing. Here we review the experience at a pediatric oncology center.

**Methods:** We report the results of an internal audit of neurosurgical performance with regard to intraoperative biological material collection from 2015 to 2019. Data was collected from electronic records in a retrospective anonymized fashion. Types of biological material collected intraoperatively were: a) tumor tissue obtained with grasping forceps, b) surgical aspirate obtained with filters from suction or ultrasound aspiration and c) 5ml EDTA blood samples. For technical reasons, cerebrospinal fluid and normal CNS tissue were rarely obtained during surgery and are thus not included in the audit. However, CSF specimens are routinely obtained by lumbar puncture approximately 14 days after surgery.

**Results:** In the audit period, 184 pediatric CNS lesions underwent resection or biopsy (17% pilocytic astrocytoma, 16% medulloblastoma, 15% high-grade glioma, 12% ependymoma, 40% other). Tissue for routine diagnostics was obtained in all cases, mostly along with ETDA blood samples, corresponding to SIOPE priority 1 (= highest). Tissue and/or surgical aspirate for internal and external research was obtained in 164 cases (89%), corresponding to SIOPE priority 2 (= highly important). In 107 cases (58%) tumor cell cultures were established, potentially enabling patient-specific treatment approaches.

**Conclusion:** Collection according to SIOPE categories 1 and 2 was achieved in 100% and 90% of neurosurgical operations within the framework of a certified pediatric oncology center. This should be

regarded an essential prerequisite for precision medicine and access to new therapies. We consider biomaterial collection a neurosurgical quality indicator, which should be subject to active auditing and benchmarking.

##### FP041

##### Blood brain barrier characteristics in diffuse midline glioma and impact of the opening with ultrasound on delivery of neoplastic drugs

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**Objectives:** The poor prognosis of pediatric diffuse midline gliomas (DMGs), including diffuse intrinsic pontine gliomas (DIPGs), is partly due to the blood-brain barrier (BBB). The BBB can be safely opened by ultrasound (US), but few studies have focused on its specificities in pediatric DMGs, especially when submitted to US. Moreover, the effect on DMGs of AsiDNA™, a DNA repair inhibitor, and the influence of the BBB on its efficacy are unknown. The objectives were to characterize the BBB in DIPGs and the influence of US on its specificities, and to evaluate the effect of US-induced BBB opening on the kinetics of AsiDNA™ in DIPG murine models.

**Material and Methods:** US-induced BBB opening was performed with the SonoCloud® device. Permeability of DIPGs' BBB was assessed with fluorescent dextrans. An histological evaluation of BBB markers was carried out by immunohistochemistry and immunofluorescence, including endothelial markers (CD34, GLUT1 and PLAVP), tight junctions proteins (claudin 5 and Zona occludens 1), and the efflux transporter ABCB1. An in vivo and ex vivo kinetic study of AsiDNA™ was performed with fluorescence.

**Results:** In vivo, 3 kDa and 70 kDa dextrans crossed the BBB after US-opening, while 500 kDa dextrans did not. Ultrasounds seems to increase the passage of these dextrans. Transcripts of ABCB1 showed a decrease in certain DIPGs and were downregulated by US. The spontaneous passage of AsiDNA™ through the BBB was limited but seems potentiated by US.

**Conclusion:** We have shown that the BBB was specific in DIPG murine models, particularly in the expression of ABCB1. This BBB can be modulated and permeabilized by US, a technique that could allow for an improved delivery of AsiDNA. This study is a preliminary step before the introduction of a such technique and treatment in future clinical trials in children and adolescents with DMGs.

##### FP042

##### Retrospective study of biopsies taken from caudal brainstem tumors in pediatric patients

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**Objective:** Biopsies in patients with lesions in the caudal brainstem remains to be somehow controversial. Possible risks needs to weight against possible benefits in the context of novel molecular biology diagnostic means. We aim to study the different biopsies of the caudal brainstem retrospectively in a single center patient cohort.

**Methods:** We included children (age < 18 years) who received biopsies in the pons or medulla oblongata due to different lesions as performed at our pediatric neurosurgical center in the time period between 2009 and 2022.

**Results:** 27 children were included in this study. 12 navigated Varioguide, 5 robotic Autoguide, 3 endoscopic and 8 open biopsies were performed. In two patients transient neurological deficit was seen immediately after biopsy, while none of the children showed intervention

related permanent morbidity without any intervention related mortality. Histopathological diagnosis was given in all patients. Molecular genetic characterization was feasible in 97% of requested specimen (n=23). Most common diagnosis (60 %) was diffuse midline glioma (H3K27M mutated). Low grade gliomas were identified in 14%. Overall survival was 37.5% after a mean follow up of 24 months.

**Conclusion:** Biopsies of the caudal brainstem in children are experienced to be feasible and safe in the presented setting. Surgical techniques are chosen according to the patient condition and tumor location. Acquired material was sufficient for histopathological diagnosis and molecular pathological profiling in most cases. These challenging interventions in an eloquent CNS region in children should be performed following interdisciplinary decision making at specialized center to possibly enable targeted therapies in future perspective.

#### FP043

##### **Molecular Subgroup of Medulloblastoma: Evaluation of contribution to CSF diversion Following Tumour Resection**

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**Introduction:** Medulloblastoma is the commonest malignant brain tumour in children. Pre-operative hydrocephalus is present in up to 90% of these patients at presentation. Following posterior fossa surgery, despite resolution of fourth ventricular obstruction, a proportion of these children will still require cerebrospinal fluid (CSF) diversion for management of persistent or new hydrocephalus. Currently no accurate tool exists regarding which pathological subset or group of medulloblastoma patients will require a shunt post-operatively. In this study we investigated the impact of molecular subgroup of medulloblastoma on shunt dependency post-operatively in paediatric patients.

**Methods:** We undertook a retrospective multi-centre study of children with medulloblastoma who underwent tumour resection. Those with available molecular subgroup were identified. Demographic data and clinical parameters including age, sex, presence of pre-operative hydrocephalus, extent of surgical resection, evidence of metastasis/leptomeningeal disease and need for CSF diversion post-operatively were further analyzed.

**Results:** Sixty-nine children with medulloblastoma with available molecular data were identified during the study period with male to female ratio of 1.5:1 (42M:27F). Twelve patients (17.4%) belonged to SHH, 10 (14.5%) Wnt, 19 (27.5%) Group 3 and 15 (22.0%) Group 4; 13 (19%) were non-specified Group 3 or 4. A total of 18 (26%) patients had evidence of leptomeningeal disease at presentation (20% Wnt, 42% Group 3, 33% group 4, 23% group 3/4, 0% in SHH). Fifteen patients (22%) underwent post-operative ventriculoperitoneal (VP) shunt insertion. No patient in the Wnt group required ventriculoperitoneal shunt post-operatively in this cohort. Need for shunt was associated with pre-operative hydrocephalus, leptomeningeal disease, with molecular group 3 or 4 demonstrating higher rate of leptomeningeal disease, and pre-operative hydrocephalus.

**Conclusion:** All patients requiring permanent post-operative ventriculoperitoneal shunt belonged to non-Wnt groups particularly group 3 and 4.

#### FP044

##### **Enhancing the reliability of Intraoperative ultrasounds in space-occupying brain lesions**

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**Introduction:** Intraoperative ultrasound (IOUS) may aid the resection of space-occupying brain lesions, though technical limits may hinder its reliability.

**Methods:** IOUS (MyLabTwice, Esaote, Italy) with a microconvex probe was utilized in 45 consecutive cases of children with supratentorial space-occupying lesions aiming to localize the lesion (Pre-IOUS) and evaluate the extent of resection (EOR, Post-IOUS). Technical limits were carefully assessed and strategies to enhance the reliability of real-time imaging were accordingly proposed.

**Results:** Pre-IOUS allowed to localize the lesion accurately in all the cases (5 cavernomas, 7 DNT, 28 gliomas/gangliogliomas, 5 other tumors). In 10 deeply located lesions coupling with neuronavigation was useful to plan the surgical route. In 7 cases the administration of contrast warranted a better definition of vascular pattern of the tumor. Post-IOUS allowed to evaluate EOR reliably in small lesions (< 2cm). In large lesions (>2cm) assessing EOR is hindered by the collapsed surgical cavity, especially when the ventricular system is opened, and artifacts that may simulate or hide residual tumors. Main strategies to overcome the former limit are: inflation of the surgical cavity through pressure irrigation while insonating, closure of the ventricular opening with gelfoam before insonating. To overcome the latter: avoid the use of hemostatic agents before IOUS, insonating through normal adjacent brain instead of corticotomy.

These technical nuances enhanced the reliability of Post-IOUS, with a total concordance to postoperative MRI. Indeed, the surgical plan was changed in about 30% of cases, as IOUS showed a residual tumor that was left behind.

**Conclusion:** IOUS warrants a reliable real-time imaging in the surgery of space-occupying brain lesions. Limits may be overcome with technical nuances and proper training.

#### FP045

##### **Navigated transcranial magnetic stimulation motor mapping and DTI tractography for diencephalic tumour in pediatric patients**

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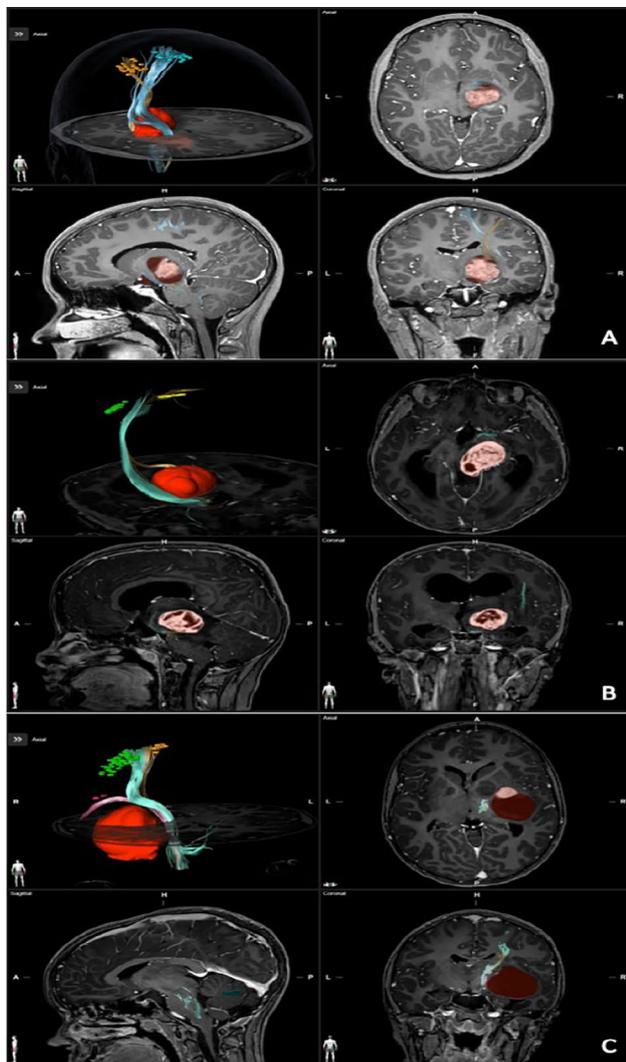
**Introduction and Goals:** In deep seated brain tumours, adequate pre-operative planning is mandatory for assessing the best surgical corridor to obtain maximal safe resection. Functional diffusor tensor imaging (DTI) tractography based on navigated transcranial magnetic stimulation (nTMS) motor mapping has proven to be a valid preoperative examination method in adults. However, its use on pediatric population is still uncommon. Our aim is to present the application of nTMS and functional DTI tractography in a series of pediatric diencephalic tumours.

**Discussion and Conclusion:** Three patients affected by thalamic (one) and thalamopeduncular tumour (two) were successfully examined with nTMS motor mapping and DTI tractography between October 2020 and October 2021 (F:M 3:0, mean age 12 years 0.8). Cortical representation of leg, hand and mouth were determined in the affected hemisphere and the positive stimulation spots were set as seeds point for tractography.

**Results:** Mapping of the motor cortex and tracts reconstruction for leg and hand were successful in all patients, while facial function was properly mapped in one patient only. In all the cases the procedure was well

tolerated and no adverse events were recorded. Spatial relation between tumour and functional tissue guided the surgical planning (Figure). All patients underwent surgery with IONM control and the white matter stimulation during tumour removal confirmed the results of tractography i.e. the sub-fascicles position and depth. Extent of resection varies from 96.1% to 100% with a postoperative new motor deficit in one patient.

**Material and Methods:** nTMS and DTI fiber tracking is a feasible, effective and well tolerated method to identify motor pathway in deep seated lesion in pediatric population. Moreover, it enables a more precise preoperative risk assessment for postoperative motor deficit, improving preoperative counseling.



#### FP046

**Laser Interstitial Thermal Therapy for the treatment of deep seated brain tumors and focal epilepsies. Monocentric retrospective study of 23 procedures in 20 children**

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**Objective:** Laser interstitial Thermal Therapy has emerged in the last decade as an innovative, successful minimally invasive surgical tool for the

ablation of brain lesions. However, it has been available in Europe only 3 years ago. Here, we wish to analyze our preliminary results in 23 procedures in 20 children with deeply seated WHO grade I brain tumors or malformations of cortical development in term of clinical and radiological outcome.

**Material and Methods:** Monocentric retrospective study of a prospectively acquired database of 20 children consecutively treated with LITT in Necker Hospital. Surgery was performed with the VISUALASE system (Medtronic) under MRI control, after robotic stereotactic placement of the laser fiber. We reviewed medical files and radiological imaging.

**Results:** 20 children (23 procedures) were operated between February 2021 and December 2022 (M/F ratio=1, mean age 11 yo ;sd 3.9y). Etiologies were as follows: WHO grade I tumors (n=9), FCD (n=11), Hypothalamic hamartoma (n=1). 14 children had previous surgery: SEEG recording (n=8), stereotactic biopsy (1), incomplete resection (1), LITT(2), indirect revascularization for moyamoya (2). Lesion localization was: left insula (7), left mesial temporo occipital (4), left cingulum (n=3), central region (2) basal ganglia/mesencephalon (2), hypothalamus (n=1), left temporal neocortex (1). There were 2 perioperative complications: transient hemiparesis (insular case) and hydrocephalus (midbrain tumor) however no patients had permanent deficits. With a mean follow up of 10.7 months: 13/16 patients were Sz free, and tumor relapse was noted in 1/10 case after 6 months. Radiological course depends on histological diagnosis.

**Conclusion:** This preliminary series shows that LITT is safe and efficient for children with deeply seated brain lesions both for oncologic and epilepsy treatments. We highlight in this series specific indications, with scarce available literature, notably in insular epilepsies, and in NF1 patient harboring tumors after cerebral revascularization.

#### FP047

**Laser ablation for deep brain tumors in children**

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**Introduction:** Laser ablation (LA) is a minimal-invasive treatment option for deep-seated tumors introduced in Europe in 2018. This study reports tumor control, radiological outcome, complications, days in hospital after LA.

**Methods:** Patients aged 0-20 years treated with LA at the Sahlgrenska University Hospital were followed at least 6 months after treatment. Three- and six-months radiological outcome and complications were registered.

**Results:** Six patients with tumor were treated with seven procedures, age 3-20 y (mean 11.8 y). Biopsy was done in 5 patients. Pathology included pilocytic astrocytoma, opticohypothalamic glioma, medulloblastoma, DNET and low-grade tumor of unknown type. All tumors were in deep locations. One patient had a transient hemiparesis and visual field defect, resolving after 5 months. Patients where no complication was seen were discharged 1-2 days after surgery. Good tumor control was achieved in the ablated region in all except in medulloblastoma.

**Conclusion:** LA is a good option for deeply located brain tumors. Hospital stay was short and patients recovered quickly. Neurological deficits occurred in 1 of 6 patients, but the neurological deficit resolved within 5 months. Tumor control was good in low-grade tumors.

#### FP048

**Results of Stereo-EEG Electrode Replacement Following LITT for Treatment of Refractory Epilepsy**

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**Introduction:** In some complex cases of medically refractory epilepsy LITT can change the epileptic network resulting in continued

seizure activity. We present our experience with post LITT stereo EEG monitoring.

**Methods:** From 2016 to 2021 we replaced stereo EEG electrodes in 28 patients following LITT.

**Results:** There were no adverse events related to electrode replacement. In 10 patients the post LITT monitoring did not reveal additional seizure activity and the electrodes were removed without incident. In 18 patients additional surgery including resection, additional LITT or RNS was performed. Twelve subjects achieved ILAE 1 outcome, one ILAE 2, two ILAE

**Conclusions:** Stereo EEG monitoring can be safely performed after LITT. Post LITT monitoring can sometimes lead to improved seizure control through additional surgery

## Abstract Session 8: Epilepsy

### FP049

**Seizure and neuropsychological outcome after surgery of low-grade associated-epilepsy tumors: a single center, retrospective, observational study**

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**Introduction:** Low-grade epilepsy-associated neuroepithelial tumors (LEATs) are increasingly recognized as a cause of focal epilepsy, particularly in children. Epilepsy surgery should be considered an early treatment option for LEATs. Gross total resection may be the most important prognostic factor; however, the timing of epilepsy surgery, the role of epileptological presurgical evaluation, post-operative seizure outcome and medication withdrawal as well as neuropsychological outcomes, remain manner of debate. In this retrospective, observational study, we reviewed focused on what might make surgery "resolutive" in terms of being seizure and medication free, and the impact on neuropsychological profile.

**Methods:** The study population included paediatric subjects who had undergone resection of LEATs between 2015 and 2021 at our institution. Extensive clinical, surgical data and neuropsychological assessments have been reviewed.

**Results:** 47 patients (25 temporal lobe, 22 extra-temporal) were assessed. Age range of seizure onset was  $70 \pm 54.4$  months, whereas at surgery was  $91.5$  months  $\pm 52$  months. Epilepsy until surgery was  $18.8 \pm 22.7$  months. 48,9% of (23) patients had preoperative development impairment. Leading histologies were dysembryoplastic neuroepithelial tumour (DNT) in 26 cases and ganglioglioma in 14 cases. Anterior and extended temporo-mesial lobectomy accounted together 44.6 % (21) of surgical approaches. Following surgery, 76,5% (36) of patients remained seizure-free (Engel IA/ILAE 1), and half were also medication free. Within the age range studied, the improvement documented was statistically significant ( $p$ -value  $< 0,004$ ) in verbal communication and attention domains.

**Conclusions:** Duration of epilepsy, younger age at surgery, and extended resection of temporal lobe tumours have been suggested as favourable prognostic factors for seizure control, discontinuation of anti-seizure drugs and neuropsychological outcome. Therefore, we advocate early epilepsy surgery without requiring proof of drug resistance, especially to lesions associated with the non-eloquent cortex.

This may represent a resolutive approach, potentially avoiding the introduction of antiseizure medications in LEATs.

### FP050

**Chloride deregulation in Focal Cortical Dysplasia type 2**

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**Background/Objectives:** Focal cortical dysplasia type II (FCDII) are emblematic malformations of cerebral cortical development associated with epilepsy in children. Somatic mutations in the PI3/AKT/mTOR pathways have been identified in FCD Type II, and recently we showed GABAergic dysfunction secondary to abnormal expression of chloride co-transporters (CCC) NKCC1 and KCC2. However, the pathways linking the genetic alterations to functional abnormalities and altered physiological excitability are missing.

Here, we investigate a possible link between mTOR activation, and CCC deregulation through abnormal control of the WNK1/SPAK/OSR pathway.

**Methods:** Ex vivo electrophysiological recordings were performed with Microelectrode arrays (MEA) in acute human cortical slices from pediatric patients operated of FCDII. The physical interactions between PI3K/AKT/mTOR and WNK1/SPAK/OSR1 proteins were investigated by co-immunoprecipitation. Expression studies were completed by western blot.

**Results:** First, we recorded spontaneous interictal spikes from human FCDII slices incubated with artificial cerebrospinal fluid (ACSF); they were abolished after administration of mTOR inhibitors demonstrating the involvement of mTOR pathway in the occurrence of epileptic activity. Second, we revealed a physical interaction between mTOR and mSIN1 of mTORC2 with WNK1 and SPAK/OSR1 respectively; consequently, mTORC2 increases the activation and the degree of phosphorylation of WNK1/SPAK/OSR1. Third, western blot analysis showed reduced levels of KCC2/NKCC1 ratio in FCD cortical slices as compared to the controls. Interestingly, inhibition of WNK-SPAK kinases with Staurosporine and N-ethylmaleimide (dephosphorylate the Thr233 and Ser373 of SPAK) induced an increased KCC2 expression and rescued the KCC2/NKCC1 ratio in FCDII. Eventually, treatment with mTOR inhibitors restored the KCC2/NKCC1 ratio along with a decreased activity of WNK/SPAK/OSR.

**Conclusion:** This study shows that mutations of mTOR pathway activates WNK/SPAK/OSR kinases, resulting in deregulation of cation chloride cotransporters in FCD, supporting epileptogenesis through dysfunction of GABA inhibition.

### FP051

**Deep brain stimulation in paediatric dystonia:therapeutic realism over nihilism**

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**Background:** Pediatric dystonia (PD) has a significant negative impact on the growth and development of the child.

**Objective:** This study was done retrospectively to analyze functional outcomes in pediatric patients with dystonia who underwent deep brain stimulation.

**Methods:** In this retrospective analytical study, all the patients of age less than 18 years undergoing DBS for dystonia between 2012 and 2020 in a single center were analyzed and their functional outcomes were measured by the Burke–Fahn–Marsden-dystonia-rating-scale (BFMDRS).

**Results:** A total of 11 pediatric patients were included with a mean age of onset, duration of disease, and age at surgery being 5.75 years, 7.36 years, and 13.11 years respectively with a mean follow-up of 23.22 months. The

mean pre-DBS motor score was  $75.44 \pm 23.53$  which improved significantly at 6 months and 12 months follow up to 57.27 (p value 0.004) and 50.38 (p value <0.001) respectively. Limbs sub-scores improved significantly at both the scheduled intervals. There was a significant improvement in disability at 1-year follow-up with significant improvement in feeding, dressing, and walking components. There was 27.34 % and 36.64 % improvement in dystonia with 17.37 % and 28.86 % reduction in disability at 6 months and 12 months respectively. There was a positive correlation between the absolute reduction of the motor score and improvement in disability of the patients at 6 months ( $\rho = 0.865$ , p value 0.003). **Conclusions:** This is one of the largest series in world literature on DBS in Dystonia. It has an enormous role in reducing disease burden and achieving a sustainable therapeutic goal.

#### FP052

##### **Role of resting state functional MRI in preoperative language mapping in children with therapy resistant epilepsy: ready for clinical use?**

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**Introduction:** Preoperative language mapping is a necessary task in patients with therapy-resistant epilepsy (TRE) to assess role of language in the epileptic network and to determine the risk of developing deficits following surgery. Task-based fMRI (tb-fMRI) is the only clinically available tool for non-invasive language mapping. However, children's cooperation during the study is low and they get easily distracted. Resting-state fMRI emerges as a valid alternative that may cope with some of these issues, but further clinical validation is required.

**Methods:** Twenty-seven children with TRE preselected for epilepsy surgery had tb-fMRI (2 tasks, verb generation and object naming in block design) and rs-fMRI (short video projection) scans as part of the preoperative study. Standard pre-processing steps were carried out. For rs-fMRI, an independent component analysis approach combined with a template-matching procedure were used to identify language networks. Outputs for both techniques were compared for lateralization with laterality index and for localization with Dice coefficients. Overlapping of language networks was analyzed.

**Results:** Language lateralization results from task-based fMRI and rs-fMRI mapping had an acceptable concordance. Resting-state fMRI and task-based language maps were positively correlated in terms of lateralization of language. In some cases, rs-fMRI indicated a more extensive language representation. Localization yielded broader areas with rs-fMRI derived networks than with tb-fMRI.

**Conclusion:** High incidence of atypical language location was observed. Resting-state analysis was acceptably concordant compared with tb-fMRI. Funding: The present study was self-funded.

#### Abstract Session 9: Hydrocephalus II

#### FP053

##### **Relation of third ventricular diameter (TVD) and ventricular indices to cerebrospinal fluid volumes and brain volume in pediatric hydrocephalus**

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**Introduction:** Treatment decision and monitoring of treatment success in pediatric hydrocephalus is often based on changes in ventricular

width. After closure of the fontanel, the ventricular width can be estimated either by determining the third ventricle diameter (TVD) by transtemporal ultrasound (US) or by measuring the common ventricular indices in cranial CT or MRI. The aim of our study was to investigate whether and to what extent TVD and the ventricular indices correlate with the actual volume of the inner and outer cerebrospinal fluid (CSF) spaces and brain volume.

**Methods:** 83 children (newborn to 18 years) diagnosed with hydrocephalus were investigated. All patients underwent cranial MRI and using an automated segmentation algorithm the volumes of lateral ventricles, third and fourth ventricle as well as total internal/external CSF and brain volume were determined. Volumes were compared to TVD, Cella Media Index (CMI), Evans Index (EI) and frontal-occipital-horn-ratio (FOHR).

**Results:** TVD and all 3 ventricular indices correlated excellently and highly significant ( $p < 0.01$ ) to the volume of the lateral ventricles ( $r = 0.666-0.859$ ), the third ventricle ( $r = 0.447-0.849$ ) and the total internal CSF spaces ( $0.668-0.871$ ). All linear parameters correlated significantly negative ( $r = -0.618 - -0.272$ ;  $p < 0.01$ ) with external CSF volume. Brain volume and CSF volumes correlated negatively only depending on pathology. CMI was the only linear ventricular parameter to correlate significantly negative with brain volume ( $r = -0.324$ ,  $p < 0.01$ ).

**Conclusion:** TVD and all three lateral ventricular indices reliably mirror the inner CSF volume and correlate negatively to the outer CSF volume. TVD was not inferior in its correlation to CSF volumes compared to lateral ventricular indices. Since TVD can be determined easily, precisely and reliably by transtemporal US, in contrast to ventricular indices demanding CT or MRI, it becomes a superior, reliable, non-invasive diagnostic bedside tool in pediatric hydrocephalus.

#### FP054

##### **Noninvasive quantitative approximation of ICP in pediatric idiopathic intracranial hypertension based on point-of-care ultrasound optic nerve sheath diameter**

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**Introduction:** Diagnosis and follow-up in children with idiopathic intracranial hypertension (IIH) often require repetitive lumbar punctures (LPs). This study investigates the inter- and intraindividual relationships between ONSD and ICP in childhood IIH and evaluates whether an individualized mathematical regression equation obtained from two paired US-ONSD/ICP values can be used to approximate ICP directly from ONSD values to avoid further LPs.

**Methods:** 159 ultrasound examinations and 53 invasive ICP measures via lumbar puncture were performed in 28 children with IIH. US-ONSD was measured using a 12-Mhz linear transducer and compared to ICP values. In 15 children a minimum of 2 paired US-ONSD/ICP measures were performed and repeated-measures correlation (rmcorr) as well as intraindividual correlations were analyzed.

**Results:** Over the entire cohort, the correlation of ONSD and ICP was moderately good ( $r = 0.504$ ,  $p < 0.01$ ,  $Rmcorr = 0.91$ ,  $p < 0.01$ ). In contrast, intra-individual correlations of ONSD and ICP were outstanding ( $r$  ranging from 0.956-1). A mathematical regression equation obtained from two paired US-ONSD/ICP values can be calculated and applied for the individual patient to approximate ICP from US-ONSD.

**Conclusions:** Point-of-care US-ONSD is a noninvasive, quick and reliable method to control treatment efficacy in pediatric IIH. An individualized regression formula can be established from two US-ONSD/ICP value pairs for each patient, for example during initial lumbar puncture before and after withdrawal of CSF; This formula can then be used to directly approximate ICP from ONSD values. Thus, invasive follow-up methods can potentially be avoided.

## FP055

**Endoscopic Third Ventriculostomy for Shunt Malfunction in the Pediatric Population: A Systematic Review, Meta-analysis and Meta-Regression Analysis**

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**Introduction:** Despite growing published evidence of the merits of endoscopic third ventriculostomy (ETV) instead of shunt revision at the time of shunt malfunction (secondary ETV), concerns about its efficacy and complications remain and ETV is still not used widely in this context. This study aimed to carry out a comprehensive meta-analysis and reports on the success and safety of secondary ETV in the pediatric age group.

**Methods:** In accordance with PRISMA guidelines, systematic searches of Medline, Embase and Cochrane Central were undertaken from database inception to 7th September 2022. ETV success was defined as the lack of need for a shunt and was the primary outcome measure. Secondary outcome measures were rates of complications and mortality. A random effects model was used. Summary-level meta-regression was performed to identify predictors for success in accordance with the ETV Success Score (ETVSS).

**Results:** Sixteen studies reporting 584 patients who underwent a secondary ETV for shunt malfunction were included in the meta-analysis. The overall pooled mean age was 6.1 years, and 57.0% were male. Pooled prevalence of hydrocephalus etiologies was as follows: aqueduct stenosis (39.3%); myelomeningocele (27.6%) post-infectious (17.1%); post-hemorrhagic (13.0%); neoplasm (13.0%); malformation (11.3%). Overall pooled success rates of ETV for shunt malfunction at 3 months, 6 months and 12 months were: 65.69% (95%CI: 52; 77); 63.25% (95%CI: 54; 72); 53.37% (95%CI: 24; 81). Overall pooled prevalence of intraoperative bleeding was 4.96% (95%CI: 0; 64). Overall rates of complications were low with new neurological deficit (transient or permanent) the highest at 1.61% (95%CI: 0.68; 3.72). On meta-regression, age, proportion of post-infectious hydrocephalus, number of shunt revisions were not statistically significant predictors of secondary ETV success at 6 months.

**Conclusions:** This meta-analysis demonstrates that secondary ETV after shunt malfunction in pediatric patients is a feasible option with acceptable success rates and low complication rates

## FP056

**Efficacy of Endoscopic Third Ventriculostomy in the management of shunt failure and predictive factors**

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**Introduction:** Endoscopic third ventriculostomy (ETV) is known as a safe procedure for obstructive hydrocephalus and it is gaining popularity as an alternative to shunt revision in the management of shunt malfunction. We evaluate the ETV effectiveness in terms of ventriculoperitoneal (VP) shunt removal in pediatric patients with shunt failure.

**Methods:** We retrospectively reviewed the medical records of 76 pediatric patients who underwent ETV because of ventriculoperitoneal shunt malfunction at Meyer Children Hospital between 1998 and 2022.

**Results:** Median age at secondary ETV was 10 yo (10 ± 9,6).

The hydrocephalus was post-hemorrhagic in 30% of patients, congenital in 17%, neural tube defects in 13%, aqueduct stenosis in 11%, post-infectious in 11%, tumor associated in 5%, cyst associated in 5%, Chiari I associated in 3%, and post-traumatic in 1%. Follow-up was 5,1 years (5,1 ± 4,8). The overall success rate for ETV after shunt malfunction was 61% with 46 patients without shunt at the last follow-up.

**Conclusions:** ETV can be a viable option even after shunt malfunction. The temporary external ventricular drainage (EVD) was a negative prognostic factor whereas the time between first surgery and secondary ETV >12 years was a positive prognostic factor (P<0.01). The neural tube defects, age at secondary ETV >9 yo and first surgery performed as VP shunt were positive prognostic factors affecting the good outcome of the ETV. Unfortunately, these positive factors did not reach statistical significance.

## FP057

**Tailored concept for accurate neuroendoscopy: a comparative retrospective single-center study on image-guided neuroendoscopic procedures**

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**Objective:** Neuroendoscopy utilizes several techniques for precise endoscope guidance, ranging from coordinate planning to computer-assisted technologies. We report a case-based stratification system for tailored application of the available technologies.

**Methods:** The neuroendoscopic cases at our department over 36 months (2016-2018) were retrospectively evaluated regarding their complication, outcome, and revision rate within 6 months follow-up according to the used techniques (coordinate-based [CB], guide-based [GB], ultrasound-assisted [UG] or augmented reality-navigated [NAR]).

**Results:** 309 neuroendoscopic interventions in 243 patients were included in the study. Hydrocephalic conditions (81.6%) due to intraventricular hemorrhage and/or infection (34%), intracranial tumors (20.1%) or cysts (20.7%) and aqueductal stenosis (10.7%) were the major indications. In 62.5% interventions additional guidance beyond anatomical landmarks (CB) was used, most frequently provided by navigation (NAR, 30.4%), followed by ultrasound (UB, 18.4%) and ventricular access guide (GB, 13.6%). Predominant procedures differed according to mean age (UG: 0.42±2.15 yrs, CB: 6.64±7.49 yrs, NAR 8.14±7.08 yrs, GB 9.31±9.53 yrs). UG was preferably employed in NEL and ventriculostomies, whereas navigation was primarily used for fenestrations, biopsies and tumor resections. Revision rate was highest in the age group <1 year (83.8%). The analysis confirmed significant differences regarding the UG group including mean age at surgery (p<.001), type of diagnosis (p<.001) and revision surgery rate (p<.015). Also, the prevalence of hydrocephalus in NAR (p<.001), missing association of lesion with ventricle system in the NAR group (p<.001), surgical time between GB and NAR groups (p=.029) and total number of surgeries in the CB group (p<.05) were significant.

**Conclusions:** Depending on the anatomical calvarial status, entry point localization, ventricular size, and targets a stratified approach towards image-guided neuroendoscopy prove to be safe and feasible. We suggest this four-tier strategy for intraoperative ultrasound, coordinate calculation, ventricular access guide and neuronavigation to be applied for accurate neuroendoscopy. **Keywords:** augmented reality; image-guided neuroendoscopy; hydrocephalus; intraventricular hemorrhage; presurgical planning; intraoperative ultrasound; ventriculostomy; neuronavigation

## FP058

**Endoscopic third ventriculostomy for the management of children with cerebrospinal fluid disorders, ventriculomegaly and associated Chiari I Malformation**

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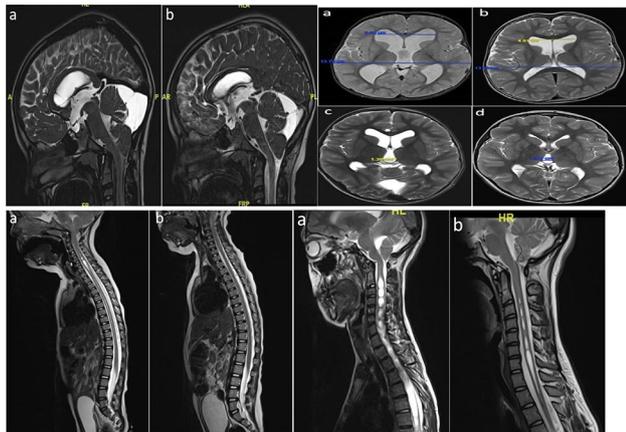
**Introduction:** Our objective was to review the outcomes of children with CIM and associated cerebrospinal fluid (CSF) disorders and ventriculomegaly undergoing endoscopic third ventriculostomy (ETV) as a primary intervention.

**Materials and methods:** A retrospective, single-centre, observational cohort study was conducted, of consecutive children with CIM with associated CSF disorders and ventriculomegaly treated first by ETV between January 2014 and December 2020.

**Results:** Raised intracranial pressure symptoms were the most frequent with ten patients followed by posterior fossa and syrinx symptoms in three cases.

One patient had a later closure of the stoma and required a shunt insertion. The success rate of the ETV in the cohort was 92% (11/12). There was no surgical mortality in our series. No other complications were reported. The median herniation of the tonsils was not statistically different in the pre- vs post-operative MRI (1.14 vs. 0.94,  $p=0.1$ ). However, the median Evan's index (0.4 vs 0.36,  $p<0.01$ ) and the median diameter of the third ventricle (1.35 vs. 0.76,  $p<0.01$ ) were statistically different between the two measurements. The preoperative length of the syrinx did not change significantly when compared with the postoperative (5 vs. 1;  $p=0.052$ ), nevertheless, the median transverse diameter of the syrinx did improve significantly after the surgery (0.75 vs. 0.32,  $p=0.03$ ).

**Conclusions:** Our study supports the safety and effectiveness of ETV for the management of children with CSF disorders, ventriculomegaly and associated CIM.



## FP059

**Placement of EVD in pediatric posterior fossa tumors: safe and efficient or old-fashioned? The Vienna experience**

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**Purpose:** The perioperative treatment of hydrocephalus in pediatric posterior fossa tumors with an external ventricular drain (EVD) is the treatment of choice in our center. We analyzed our experience in using EVD concerning safety and effectivity.

**Methods:** This is a single-center retrospective cohort study of 100 consecutive pediatric patients who underwent resection for a newly diagnosed tumor in the posterior fossa between 2011 and 2022.

**Results:** 49 patients (49%) underwent placement of an EVD. In 40 patients the EVD was inserted at a mean of 2.25 days prior to the tumor resection, 9 had the EVD inserted during tumor resection (frontal trajectory in 7 patients, occipital trajectory in 2 patients). The mean number of total days with the EVD in place was 8.61 $\pm$ 3.82 (range 2-16 days). The mean number of days with an EVD after tumor resection was 6.35 $\pm$ 3.8 (range 0-16 days). EVD associated complications were seen in 6 patients (12.24%) including one infection. None of these resulted in a worse clinical course or any long-term sequelae. Permanent CSF diversion at six months after surgery was necessary in 13 patients (13%), including two VP-shunt, two SD-shunt, six endoscopic third ventriculostomy (ETV), and three combined VP-Shunt and ETV procedures. Patients with a medulloblastoma or ependymoma had a higher rate of permanent CSF diversion needed than the group of pilocytic astrocytoma patients (27.9% versus 2.13%,  $p<0.001$ ). In patients with metastatic disease 7 of 17 patients (41.18%) needed a permanent CSF diversion, compared to 6 of 83 patients (7.23%) in the group without metastasis ( $p=0.001$ ).

**Conclusion:** The treatment of hydrocephalus in pediatric posterior fossa tumors with an EVD as a temporary measure is safe and effective, provided that a multi-professional understanding for its handling is given and there is no need for a long transport of the children.

## FP060

**Predictive models for postoperative hydrocephalus in pediatric patients with posterior fossa tumors**

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**Introduction:** Persistent hydrocephalus after posterior fossa tumor resection is common with incidences up to 40%. The (modified) Canadian Preoperative Prediction Rule for Hydrocephalus (CPPRH) was developed to identify children at risk for persistent hydrocephalus. These children likely benefit from an internal cerebrospinal fluid (CSF) diversion procedure prior to tumor resection which reduces the risk of persistent hydrocephalus. External validation is essential before implementing the CPPRH clinically. We analyzed external validation studies addressing the CPPRH.

**Methods:** Interpretation of external validation can be guided in three steps. Step 1 investigates the extent of relatedness of the development and validation sets. The degree of relatedness between sets of patients is determined by case-mix including outcome incidence and patient characteristics. Step 2 assesses model performance in the validation set. Calibration and discrimination are key performance measures. Step 3 addresses the interpretation of the model validation results. If a model performs poorly, model updating should be considered.

**Results:** Three studies were identified. At step 1 we found different case-mixes and outcome incidence (11% to 31%), Table 1. This implies that the different populations have a low degree of relatedness, and the validation study is a difficult bar for the CPPRH. At step 2 we found that discrimination measures were reported. However, calibration plots were missing. Consequently, the magnitude and direction of possible model miscalibration cannot be derived. At step 3 we observed that the CPPRH performed well at external validation in Northern America. In contrast, the European report concluded that the CPPRH had failed. However, model updating techniques were not considered.

**Conclusion:** The CPPRH has been validated with promising results in Northern America. However, to guide decision-making on prophylactic internal CSF diversion surgeries, external validation of the CPPRH in

European children strongly needs to be further tested. This work was recently published in JNS Pediatrics.

TABLE 1. Summary measures of the baseline study characteristics used to evaluate case-mix heterogeneity

Table with 4 columns: Study Characteristic, Riva-Cambrin et al., 2009, Foreman et al., 2013, Pitsika et al., 2021. Rows include Total number of patients, Incidence of postop hydrocephalus, Age <2 yrs, Papilloedema present, Transpendymal edema present, Moderate to severe hydrocephalus, Metastasis present, Tumor diagnosis (Juvenile pilocytic astrocytoma, Medulloblastoma, Ependymoma, Dorsally exophytic brainstem glioma, Other).

Values are shown as number (%) unless indicated otherwise. Statistical calculations have been proposed—provided that individual patient data are available—for assessment of the degree of relatedness between the development and validation set. Foreman et al. substituted transpendymal edema from the CPRRH for papilloedema and accordingly created the modified CPRRH. Therefore, this strategy could be seen as an updating study. Adequate sample size and outcome incidence are vital in prediction studies. Traditionally, external validation studies require a minimum of 100 children with the outcome and 100 without the outcome in order to reliably study the external validity of tools such as the CPRRH. Although more tailored sample size calculations have been proposed, the sample sizes of the current validation studies are likely suboptimal. In the study by Riva-Cambrin et al., radiological characteristics were reported for 331 patients in the development cohort and 108 patients in the validation cohort because of the lack of preoperative imaging for the remainder of the patients.

(n=7). Occurrence of de novo hydrocephalus in Group I after surgery was not observed in our cohort.

Age and histology were no confounding factor for EVD placement between Group IIa and IIb (p=0.34). The use of EVD did not result in better control of hydrocephalus compared to no-EVD patients considering pre- and postoperative MRI ventricular indices (p=0.4). Perioperative placement of an EVD resulted in a three-fold risk for subsequent VPS or ETV (Group IIa 55.5% vs IIb 16.6%): Relative risk for EVD patients compared to no-EVD patients with hydrocephalus was 3.3 (CI = 1.06 - 13.43, p=0.09).

Conclusion: Perioperative EVD placement appears to harbour a three-fold relative risk of requiring subsequent permanent CSF diversion in children above 2 years. EVD was not more effective to control ventricular enlargement compared to tumor removal alone. The noEVD strategy was safe and did not result in postoperative complications. Thus, because EVD might have an adverse effect on hydrocephalus outcome, a prospective study is warranted to falsify the results.

Abstract Session 10: HC / Spine II

FP062

Neonatal kyphectomy in the child with myelomeningocele

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Background: Kyphotic deformity of variable severity occurs in children with myelomeningocele at the upper lumbar or thoracolumbar area.

Objectives: To describe the feasibility and outcome of correction of the kyphosis in the neonatal period at the time of dural closure.

Methods: From 2018 to 2021 a series of 15 children with myelomeningocele and kyphosis operated in the first two days of life was studied.

Results: The kyphectomy was performed with resection of one to three vertebral bodies. There were no deaths or serious complications. The wound closure was successful in all patients. The correction of the kyphosis was considered satisfactory in all. The follow up ranged 1-3 years. In most of the children the late results were good, but in 7/15 the kyphotic deformity recurred and progressed. This recurrence of the kyphosis was noted after the child started to be positioned in the sitting position.

Conclusions: Neonatal kyphectomy at the time of mielomeningocele correction is a safe procedure with excellent initial correction. It promotes good skin closure and prevents skin ulcers. Late deformity is common and may be progressive.

FP063

Growth trajectories and postoperative outcomes of the occipitocervical junction in subgroups of pediatric Chiari malformation type 1

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JNS PEDIATRICS External validation of predictive models for postoperative hydrocephalus in pediatric patients with posterior fossa tumors. Includes abstract text and a table of model performance metrics.

FP061 Perioperative external ventricular drainage vs. no-EVD strategy in pediatric posterior fossa tumors – pilot study results Matthias Krause University Leipzig, Leipzig, Germany

Introduction: Pediatric brain tumors of the posterior fossa often present with occlusive hydrocephalus. Endoscopic third ventriculostomy (ETV) or ventriculoperitoneal shunting (VPS) have been established for definite hydrocephalus treatment. Aim of the study was to analyze the impact and safety of perioperative temporary external ventricular CSF drainage (EVD) placement on postoperative hydrocephalus outcome compared to a no-EVD strategy.

Patients and Methods: In a prospective database 36 posterior fossa tumor patients of 2 - 18 years were included with a follow-up of 1 year. 58% presented with preoperative hydrocephalus. Patients were assigned to non-hydrocephalus group Group I (n=15) and to preoperative hydrocephalus Group IIa with EVD placement (n=9) and Group IIb without EVD (n=12).

Results: Median age of patients was 8.1 years (range 3.17 to 16.58 years). One third of 21 hydrocephalus patients required ETV or VPS

**Introduction:** Chiari malformation type 1 (CM1) can lead to headaches, syringomyelia, and sleep disordered breathing. We recently described three subgroups of CM1, each related to a putative pathophysiological condition (bony compression, basilar invagination and foramen magnum obstruction) that correlate with specific surgical management. We aim to describe morphometric characteristics of each subgroup and to evaluate postoperative outcome.

**Methods:** Between 2010 and 2021, we retrospectively included 96 children aged 0 to 17 years that underwent posterior surgical procedure for CM1 at Necker Hospital. We used 3D morphometric geometry by applying 13 landmarks (4 for cranial base, 4 for C1 and 5 for C2) on T2 sagittal MRI to characterize occipitocervical joint morphology of patients and generate predictive growth model for each subgroup. Finally, we compared age-corrected morphological patterns for preoperative morphology and postoperative outcome across subgroups.

**Results:** We confirmed that subgroup morphology and growth trajectory differ from one another. More specifically, subgroup 1 worthen with age whereas subgroup 3 tends to mimic control group approaching adolescence, confirming that the underlying pathological process differs from one group to another.

**Conclusion:** Morphometric analyses confirmed the clinical relevance of CM1 subgroups. Our results may be applied in the clinical practice for surgical decision making. Further effort is needed to define if morphological features influence symptoms severity inside subgroups.

#### FP064

##### Scoliosis and vertebral anomalies in patients with anorectal malformation

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**Introduction:** Anorectal malformation (ARM) occurs in approximately 1–3 in 5000 live births. About 50% of ARM patients have associated congenital anomalies, involving other organs or systems. Sacral and vertebral anomalies, easily detected with spine X-ray in the neonatal period, have been suggested to be sensitive indicators of SCA in patients with ARM. Sacral and vertebral anomalies are rare in the general population, while their prevalence increases in patients with ARM, mainly in the setting of VACTERL association.

**Methods:** Patients with ARM consecutively treated between January 1999 and August 2018 were included in the study and retrospectively analyzed. Presence, topography and grade of scoliosis (according to SOSORT Consensus Guidelines 2016) were correlated to the type of ARM, according to Wingspread classification, to clinical and radiological evolutions, MRI findings, and spinal and skeletal x-ray.

**Results:** 64 (18%) out of 347 patients with ARM, treated during the study period, were followed by orthopedics and physiatrist. 29 patients (45%) had at least one vertebral anomaly. Multiple vertebral anomalies, were found in 24/29 patients (83%). Sixteen patients (25%) had isolated or multiple rib deformities. 47/64 (73%) had scoliosis, surgically

treated in 10/47 cases. A significant correlation was found for the presence of vertebral anomalies and syndromes related to scoliosis. No significant association was found between the presence of scoliosis and the level of vertebral anomalies

**Conclusion:** Our data showed a strict association between scoliosis and ARM. However, in our series, only a minority of patients (18%) have been followed up by a multidisciplinary team. This suggests the importance of a multidisciplinary and dedicated approach to patients with ARM, with appropriate radiological, physiatrist and orthopedic assessment at long-term follow up, to early detect possible scoliosis.

#### FP065

##### Cervical spine surgery for osteoblastoma in a pediatric population. Can the stability of the cervical spine be preserved? Analysis of 4 cases and review of the literature

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**Background and Aim:** Pediatric cervical osteoid osteoma and osteoblastoma are rare primary bone tumors. Resective surgery is of great importance but may be challenging. We report 4 cases emphasizing diagnostic and surgical management for different locations with complete resection without late instability.

**Material and Methods:** We report 4 pediatric cases of osteoblastoma managed in our unit, with different location at the level of the cervical spine. All 4 patients had a common complaint of persistent and progressive neck pain, 2 reported worsening pain at night, and 3 were attempted NSAID treatment without significant response. In the radiological evaluation, a lytic lesion of bone origin in the cervical spine was present in all cases: 2 at the pediculolaminar complex, one laminar and one at the level of the C2 body and odontoid. A comprehensive review of the literature was carried out.

**Results:** Complete surgical resection was performed by a posterior approach in 3 patients and a transoral approach in one. Postoperative stability was ensured by a hard neck collar under molding. All 4 were diagnosed with osteoblastoma based on clinical, radiological and histopathological criteria. Control imaging studies demonstrated satisfactory lesion resection with good late bone reconstruction. The mean follow-up time is 26.5 (6-47) months. None had a recurrence nor late instability requiring fixation.

**Conclusion:** Treatment of pediatric cervical spine of osteoid osteoma and osteoblastoma should seek a complete resection. In regions with risk for secondary instability, the preservation of capsule-ligament structures and temporary orthopedic immobilization can allow bone regeneration with no need for fixation. Extended follow-up is recommended especially in very young children.

#### Abstract Session 11: Oncology II

#### FP066

##### The use of the apparent diffusion coefficient for quantifying the utility of sodium-fluorescein guided-resection in the surgical management of central nervous system paediatric tumours

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**Introduction:** In the paediatric population, central nervous system (CNS) tumours represent the most common solid neoplasm. In the majority of cases, surgical treatment is of utmost importance, and the extent of resection correlates with the patient's prognosis. In this context, several fluorescent markers, such as Sodium fluorescein (SF), have been used to improve the intraoperative visualization of these lesions. However, at the moment, there are no means to assess its utility prior to surgery. This prospective study aimed to assess whether intraoperative fluorescence intensity can be predicted by analyzing the apparent diffusion coefficient (ADC) parameter on diffusion-weighted imaging (DWI) preoperative sequences.

**Methods:** Patient data were prospectively collected at our Institution. The whole lesion mean value of the ADC parameter was analysed on preoperative DWI sequences. The combined mean and standard deviation of ADC were calculated using a random effects model for each tumour type. Preoperative data and intraoperative findings were afterwards correlated to determine if an easy radiological parameter such as ADC can be predictive and estimate the utility of the fluorescein in CNS paediatric tumours before surgery.

**Results:** Ninety-eight patients were included in the study. In 67 (68.3%) procedures, fluorescence was reported as moderate or intense, in 9 of 98 (9.1%) absent and in the remaining was considered as slightly bright. No adverse effect to SF was registered. Intraoperative fluorescence was reported in 90.9% of surgeries whose corresponding preoperative MRI had shown contrast enhancement. A certain degree of correlation between ADC mean values on the whole lesion and intraoperative fluorescence has been found. In case of ADC mean values inferior to 1, a low or absent intraoperative fluorescence was documented.

**Conclusion:** Our preliminary results show that the ADC parameter correlates with the intraoperative fluorescence to a certain extent, although the contrast enhancement seemed a better reliable discriminant.

**FP067**

**Demographic profile and outcome in primary pediatric brain tumors. A single tertiary institute experience in India**

**Supriya Chauhan**  
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**Introduction:** Pediatric brain tumors are the most common tumor in children after hematological malignancies. There is very few data about the epidemiology of pediatric brain tumors in India.

**Methods -** This was a prospective and retrospective study in pediatric patients who had undergone surgery in our institute (JIPMER,Pondicherry). 80 cases were recruited and followed up for minimum follow up period of 1 year. The demographic profile was analysed and IHC markers were done for embryonal tumors and glioma.

**Results:** Pediatric brain tumors was equally distributed among male and females. (1:1) .Mean age of presentation was 10 years . 27.5 % of our cases were embryonal tumors,low grade glioma (16.25 % ),high grade glioma ( 12.5 % ) ,ependymoma and craniopharyngioma comprised 15 % of our cases each. Medulloblastoma comprised 23.75 % of cases Out of which 31.5 % had craniospinal metastasis at time of diagnosis. The most common location of SHH pathway medulloblastoma was cerebellar hemisphere and non WNT/non SHH was fourth ventricle. 45.45 % of patients with high grade glioma had recurrence .50 % of ependymoma cases were infratentorial. we had 2 cases of ganglioglioma ,one in the midbrain and other in temporal lobe .Gross total resection was achieved in 30 % ,Subtotal resection in 46.25 % and partial resection in 20 % of our cases. Outcome of patients at the end of 1 year for low and high grade glioma, ependymoma and craniopharyngioma were similar to western literature. Two patients acquired COVID 19 and died while undergoing treatment. Molecular markers like INI1, LIN28 A was highly sensitive and specific for diagnosing atypical teratoid rhabdoid tumor (ATRT) and embryonal tumor with multilayered rosettes (ETMR )respectively.

**Conclusion:** Our study emphasizes the need of standardized and systemic cancer registries in India.

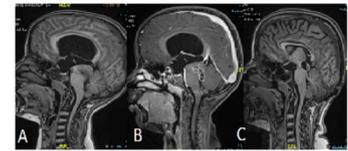
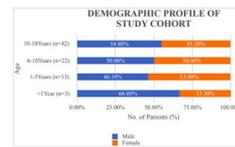
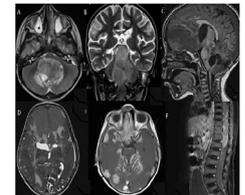


Table 4 - Histopathological subtypes of pediatric brain tumors and associated outcome (N=80)

Tumor	Histopathological Subtype	Total Number of Cases	Number of Cases at End of Follow-up	Survival Rate (%)
Astrocytoma	Low-grade Astrocytoma	12	10	83.33%
	High-grade Astrocytoma	2	1	50%
Ependymoma	Low-grade Ependymoma	10	8	80%
	High-grade Ependymoma	2	1	50%
Medulloblastoma	Medulloblastoma	10	8	80%
	Medulloblastoma	2	1	50%
Craniopharyngioma	Craniopharyngioma	12	10	83.33%
	Craniopharyngioma	2	1	50%
Ganglioglioma	Ganglioglioma	2	1	50%
	Ganglioglioma	2	1	50%
Teratoma	Teratoma	10	8	80%
	Teratoma	2	1	50%
Pilocytic Astrocytoma	Pilocytic Astrocytoma	10	8	80%
	Pilocytic Astrocytoma	2	1	50%
Oligodendroglioma	Oligodendroglioma	10	8	80%
	Oligodendroglioma	2	1	50%
Meningeal Tumor	Meningeal Tumor	10	8	80%
	Meningeal Tumor	2	1	50%
Pilo-cystic Astrocytoma	Pilo-cystic Astrocytoma	10	8	80%
	Pilo-cystic Astrocytoma	2	1	50%
Craniopharyngioma	Craniopharyngioma	10	8	80%
	Craniopharyngioma	2	1	50%
Teratoma	Teratoma	10	8	80%
	Teratoma	2	1	50%

Table 1 - Distribution of cases based on histopathological subtypes (N=80)

Histopathological Subtype	No. of Patients (%)
Low-grade Glioma (LGG)	30 (37.5%)
High-grade Glioma (HGG)	12 (15%)
Medulloblastoma (MB)	10 (12.5%)
Craniopharyngioma (CP)	12 (15%)
Ganglioglioma (GG)	2 (2.5%)
Teratoma (T)	10 (12.5%)
Pilo-cystic Astrocytoma (PCA)	10 (12.5%)
Oligodendroglioma (ODG)	10 (12.5%)
Meningeal Tumor (MT)	10 (12.5%)
Pilo-cystic Astrocytoma (PCA)	10 (12.5%)
Craniopharyngioma (CP)	10 (12.5%)
Teratoma (T)	10 (12.5%)
Pilo-cystic Astrocytoma (PCA)	10 (12.5%)
Oligodendroglioma (ODG)	10 (12.5%)
Meningeal Tumor (MT)	10 (12.5%)
Pilo-cystic Astrocytoma (PCA)	10 (12.5%)
Craniopharyngioma (CP)	10 (12.5%)
Teratoma (T)	10 (12.5%)



**FP068**

**A comprehensive analysis of infantile tumors to improve differential criteria between DIG/DIA, IHG and their potential overlapping diagnoses**

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**Introduction:** Recent epigenomic analyses have pointed out the existence of a new DNA methylation class of infant-type hemispheric glioma (IHG). As desmoplastic infantile ganglioglioma/astrocytoma (DIG/DIA), these supratentorial tumors mainly affect infants. While DIG/DIA are characterized by BRAF or RAF1 alterations, IHG are enriched for receptor tyrosine kinase genes' fusions. In this rapidly evolving field, a more comprehensive analysis of infantile glial/glioneuronal tumors including clinical, radiological, histopathological, and molecular data is needed.

**Methods:** We retrospectively investigated data from 30 infantile glial/glioneuronal tumors consecutively compiled from our center. They were analyzed by two experienced pediatric neuroradiologists, without former knowledge of the molecular data. We performed a clinical, histopathological and molecular evaluation (next generation sequencing, RNA sequencing, FISH analyses), as well as DNA methylation profiling (using the v12.5) for those samples with sufficient material available.

**Results:** The integrative histopathological, genetic and epigenetic analyses, including t-SNE analyses segregated tumors into: ten DIG/DIA (33.3%), six IHG (30.0%), three gangliogliomas (10.0%), two pleomorphic xanthoastrocytomas (6.7%), two pilocytic astrocytomas (6.7%), two supratentorial ependymomas, ZFTA fusion-positive (6.7%), two supratentorial ependymomas, YAPI fusion-positive (6.7%), two embryonal tumors with PLAGL2-family amplification (6.7%), and one diffuse low-grade glioma, MAPK-pathway altered. The current work highlights significant differential features in terms of histopathology (leptomeningeal infiltration, intense desmoplasia and ganglion cells in DIG/DIA and necrosis, microvascular proliferation and siderophages in IHG), and radiology between DIG/DIA and IHG. Moreover, these results are in line with the literature data concerning the molecular dichotomy (BRAF/RAF1 alterations vs. RTK genes' fusions) between DIG/DIA and IHG.

**Conclusion:** Infantile tumors correspond to a large spectrum of histobiomolecular entities, DIG/DIA and HIG being the more frequent ones. DIG/DIA and HIG can be differentiated by histological and radiological features.

This study was funded by the Association pour la Recherche en Neurochirurgie Pédiatrique (ARNP). Authors want to thank the Integragen and the RENOCLIP-LOC.

#### FP069

##### Against the overuse of MRI in neurofibromatosis type 1

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**Introduction:** Optic pathway glioma (OPG) is the most common central nervous system tumor in children with neurofibromatosis type 1 (NF1). Management with visual and neurologic examinations alone while ophthalmologic symptoms are absent remains controversial.

**Methods:** We retrospectively reviewed the medical records of 152 children with NF1 followed in Lille University Hospital from January 2017 to January 2022. We quantified the numbers of MRI examinations and their influence on the management of these children.

**Results:** Twenty-three patients had confirmed OPG (15.1%) on MRI. The mean number of MRI for the non OPG group was 2.43 (1.46 under general anesthesia (GA)). In the OPG group, number of MRI was 5.3 (3 under GA) for untreated patients and 13.47 (9.14) for the treated ones. Five patients with OPG (22%) had visual symptoms at the time of diagnosis. Radiological progression was observed in fourteen cases (60%). Six children (26%) were treated with chemotherapy due to worsening visual function and one (4%) due to significant tumor progression on MRI without visual impairment. After treatment, one child had worse visual function (14%), four remained stable (57%), and two had visual improvement (29%).

**Conclusion:** Systematic MRI screening changed our therapeutic management for one patient (0.7% of NF1 and 4% of OPG patients), with stable visual examination after chemotherapy. Regarding the limited therapeutic implications, the risks (GA), the stress for children and families and the costs (procedure, hospitalization), MRI should be realized to find an OPG only if neuro-ophthalmologic symptoms are present. Once the diagnosis is established, repeat MRI is useless if careful ophthalmologic exams do not show clinical worsening.

#### FP070

##### Pediatric thalamopeduncular tumors: advanced preoperative imaging to define a safe surgical planning. A multicentric experience

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**Introduction:** Thalamopeduncular tumors are challenging lesions occurring within the first two decades of life with progressive spastic hemiparesis. The optimal treatment for these tumors remains debated. However, new microsurgical and neuroimaging techniques allow subtotal or gross total resection with a good outcome.

**Methods:** We present a retrospective review of 8 children undergoing surgery for thalamopeduncular tumors in the Pediatric Neurosurgery Units of Padua and Verona from 2005 to 2020. For each patient we collected demographic, clinical and radiological data and postoperative results.

**Results:** We reported a series of 8 patients, M:F 1:1, average of 9 y.o. Pre-operative cerebral MRI showed a single thalamopeduncular lesion in 6 cases, multiple confluent lesions in a NF1 patient and an important contralateral extension in another case. All histological diagnosis were pilocytic astrocytoma. The main symptoms presented were progressive spastic hemiparesis, VII cranial nerve palsy, headache, ataxia and tremor. Three patients had associated hydrocephalus. The corticospinal tract was studied with diffusion-tensor imaging in all patients and with navigated transcranial magnetic stimulation in 2 patients. Intraoperative neurophysiological monitoring assisted surgery in 7 cases. We used a trans-sylvian approach in five patients, a transtemporal approach in two and a posterior parietal approach in one case. We achieved STR in 4 patients, GTR in 3 cases and in one patient only a partial resection. After surgery 5 patients maintained stable their neurological status, two patients improved, 3 patients worsened. Medium follow-up is 64,5 months. Five patients have a stable residual, two are disease-free and one patient is deceased.

**Conclusion:** Trans-sylvian and transtemporal routes with IONM are the principal approaches that allow a gross total or subtotal resection. An accurate presurgical planning including DTI tractography and nTMS enables to preserve the neurological functions of these patients obtaining a long-term control of disease.

#### FP071

##### Establishing a dedicated children's Gamma Knife service - experience from 65 cases

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**Introduction:** Gamma Knife stereotactic radiosurgery (GKSRS) is a well-established neurosurgical technique with neurovascular, oncological and functional indications. In most centres, children who require GKSRS are treated alongside adult patients by neurosurgeons without paediatric specialisms. We report our experience of developing a dedicated paediatric GKSRS programme in partnership with a busy adult centre. **Methods:** in 2016 a paediatric neurosurgeon (GJ) underwent specialist training in GKSRS and under the mentorship of an experienced GKSRS surgeon (NK) established a paediatric GKSRS service commissioned to serve the southern half of England. This service involves a paediatric neurosurgeon and paediatric anaesthetic and nursing teams working in conjunction with an experienced adult GKSRS team. Dedicated radiologists with paediatric experience supervised treatments and performed all angiography, and a dedicated advanced nurse practitioner dealt with all pre- and post-operative assessment.

**Results:** From October 2016 the service has undertaken 65 treatments in 59 children aged 5-17 years. 64 treatments were for vascular lesions (56 brain AVM, 8 residual vein of Galen malformation) and 1 treatment for tumour (recurrent ependymoma). 6 children required volume staged treatment (all high grade AVM). All treatments were undertaken under general anaesthesia. For those children with 5 year follow up data available (17), 13 had complete angiographic cure,

and a further 3 had significant reduction in their AV shunt. There were no intra-procedural complications, although 4 children required oral steroids for cerebral oedema during the treatment period. There was 1 re-bleed and no permanent neurological effects and no mortality. Patient feedback has been excellent.

**Conclusion:** by working in collaboration with an adult service, a paediatric neurosurgeon can manage a dedicated children's GKSRS service with highly acceptable results. Multidisciplinary teamwork is critical for the success of such a service.

#### FP072

##### Quality indicators for evaluating the 30-day postoperative outcome in pediatric brain tumor surgery: a 10-year single-center study and systematic review of the literature

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**Introduction:** Surgery is the cornerstone in the management of pediatric brain tumors. There is a need for indicators reflecting quality of the delivered care. The objective of this study was to analyze currently applied quality indicators in pediatric brain tumor surgery and identify factors associated with poor outcome at a tertiary neurosurgical referral center in western Norway.

**Methods:** All patients younger than 18 years of age who underwent surgery for an intracranial tumor between 2009 and 2020 were included. The primary outcomes of interest were classic quality indicators: 30-day- readmission, -reoperation, -mortality, -nosocomial infection, and -surgical site infection (SSI) rates; and length of stay. A systematic literature review of articles concerning pediatric brain tumor surgery reporting at least two quality indicators were of interest were conducted.

**Results:** 82 patients aged 0-17 years included. The 30-day outcomes for unplanned reoperation, readmission, mortality, nosocomial infection, and SSI were 9.8%, 14.6%, 0%, 6.1%, and 3.7%, respectively. Unplanned reoperation was associated with eloquent localization ( $p = 0.009$ ), primary emergency surgery ( $p = 0.003$ ), and CSF diversion procedures ( $p = 0.002$ ). Greater tumor volume was associated with unplanned readmission ( $p = 0.008$ ), nosocomial infection ( $p = 0.004$ ), and CSF leakage ( $p = 0.005$ ). In the systematic review, 16 articles were included and provided outcome data for 1856 procedures. Overall, the 30-day mortality and reoperations rate varied from 0% to 9.3% and 1.5% to 12% respectively. The SSI rate ranged between 0% and 3.9%, and 0% to 17.4% of patients developed CSF leakage. Four studies reported infratentorial tumor location as a risk factor for postoperative CSF leakage.

**Conclusions:** The 30-day outcomes were comparable to published outcomes. Tumor volume and location were related to unfavorable outcomes. Standardized outcome definitions in a large and multicentric database are needed in the future for evaluation of quality indicators.

#### FP073

##### Relaxation technique of imagery based story telling reduces manifestation of anxiety in children and adolescents undergoing brain tumor surgery

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**Introduction:** Children and adolescents undergoing surgical resection of brain tumors are prone to marked psychologic burden. Especially fear of surgery and its consequences like pain or neurologic sequelae is an important issue. Techniques of relaxation might reduce the intensity of the experience of anxiety and therefore might improve quality of life.

**Methods:** In this study, we aimed at determining the effect of a standardized imagery story telling on experiencing anxiety as quantified by the questionnaire KAT III and cardiac frequency (CF) before and after intervention at two time points (before and after surgery). 12 patients (age: 6-17 years) undergoing craniotomy due to brain tumor resection were included to the study. KAT III-scores and CF were determined and compared before and after interventions using a dependent t-test.

**Results:** Mean KAT III-score before first intervention was 0.23 (SD: 0.23), while it was 0.15 (SD: 0.21) after ( $p = 0.11$ ). Mean KAT III-score before second intervention was 0.11 (SD: 0.13), after: 0.05 (SD: 0.9),  $p = 0.07$ . Mean CF before first intervention was 77.1 (SD: 10.3), after: 68.36 (SD: 6.8),  $p = 0.003$ . Mean CF before second intervention was 71.67 (SD: 9.57), after: 65 (SD: 8.72),  $p = 0.003$ . CF was significantly lower post-interventionally after the first and the second intervention. KAT III-score showed a trend to be lower post-interventionally after the second intervention.

**Conclusion:** Our study points at an efficacy of the deployed relaxation technique of imagery story telling in children and adolescents undergoing brain tumor surgery, although it was limited by a small patient number. Further studies with larger patient numbers and a comparison of randomized intervention vs. non-intervention groups are warranted.

#### FP074

##### Brainstem tumors in children: a retrospective monocentric experience in the light of genetic and bio-molecular progress

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Brainstem tumors represent a challenge for pediatric neurosurgeons. Their management and prognosis vary according to their anatomic-pathological finding and their genetic and bio-molecular fingerprints. We report our experience with a retrospective analysis of pediatric patients. **Material and methods:** All patients with a brainstem tumor admitted at the Pediatric Neurosurgical Service of Lyon between January 1997 and December 2019 were considered for this study. 112 patients were included with an age from 6 months to 18 years and a median age of 8,9 years. The male/female ratio was of 0,94.

At diagnosis, 14 % of patients presented a deficit of the mixed cranial nerves, 21% cerebellar troubles with balance disorders, 16% nystagmus and torticollis, 10% impaired consciousness while 38% presented with a mild intracranial hypertension.

According to the classification of Choux 59 patients were classified as belonging to the group I (52,67 %), 25 to the group II (22,32 %), 26 to group III (23,21 %), and 2 to group IV (1,8%).

The main location was at level of the pons (53%), tectal region (12,5%), and the bulbo medullary region (14,3%). Thirty-five patients were treated for hydrocephalus with forty-eight surgical procedures. Forty-four of the surgical procedures were biopsies either open (23 patients), frame based (16 patients) or frameless (5 patients). Forty-two patients had surgery for tumor removal with a total surgical resection in 20 cases, a subtotal resection in 13 patients, and a partial resection in 9 patients. 56 patients could have biomolecular studies.

Sixty-six patients (59%), had postoperative chemotherapy according our neuro oncological team and according to the protocol of the SFOP. Twenty-six patients were treated with only chemotherapy and 31 patients were treated with chemotherapy followed by radiotherapy. Three patients received chemotherapy after a new surgical procedure realized for a recurrence, six patients were included in a protocol of chemotherapy for a tumor progression. Five patients with a DIPG received only radiotherapy for the severe evolving clinical neurologic aggravation.

**Results:** At last follow-up, the overall survival rate was 45% with a median follow-up of 7,5 years (range from 1 to 23 year. For DIPG the median survival is inferior to one years with a range between 4 months and 36 months. The survival rate of the non-DIPG tumors

was 90.6% (only 5 deaths over 53 patients). The median survival for all patients with benign focalized or exophytic brainstem tumors were of 13 years with a range varying from two years to 23 years. 11 patients had a survival superior to fifteen years with a range between fifteen years and twenty-three years.

**Conclusions:** Our series confirms that benign histological tumors have a good survival with surgical removal and complementary RT/CHEMO treatment. The diffuse pontine gliomas have a dismal prognosis also at the light of the more recent biomolecular progress. The new protocols have not yet reported significant improvement of the survival rate.

The future depends mainly on the discover of more effective drugs that could change the dismal evolution of infiltrating brain stem tumors.

## Poster Presentations

### Poster Track 1.1: Craniosynostosis / Malformation

#### PP001

#### Endoscopic treatment of sagittal craniosynostosis. Single institution experience

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**Introduction:** In recent years there is a tendency towards less invasive endoscopic suturectomy techniques in the management of sagittal synostosis, combined with postoperative external orthosis, to avoid open cranial reconstruction. We present our experience of endoscopic treatment of sagittal synostosis.

**Methods:** A single institution retrospective review was performed from 2016 to 2022 for patients with sagittal synostosis (scaphocephaly) who underwent endoscopic suturectomy. Some patients received postoperative helmet therapy. All patients had preoperative 3DCT scans for planning. Outcomes were assessed with pre- and postoperative 2D photographs.

**Results:** There were a total of 16 patients (13 males) who underwent endoscopic suturectomy for correction of sagittal synostosis at a mean age of 4.8 months (range: 2,8-10). The mean follow up period was 24 months (range: 2-79). There were no surgical complications. All patients were discharged from hospital the second postoperative day. Four patients received helmet postoperatively for a period of 3 months. All but two patients, even the patient operated at 10 months of age, achieved good aesthetic results. There was no significant difference between those who had helmet treatment and those who did not. Two patients developed mild oxycephaly at nine months postoperatively. They were considered for reoperation but this was not performed eventually.

**Conclusion:** Minimally invasive endoscopic suturectomy technique is good alternative to open skull release procedures for scaphocephaly, when performed early in the first few months of life. The benefits of minimally invasive surgery include shorter surgical time, less blood loss, and small incisions and short hospital stay.

#### PP002

#### Smad6 in patients with midline craniosynostosis

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**Introduction:** Smad6 was found in our population to be involved in midline craniosynostosis. We reviewed our cases of patient with craniosynostosis and a genetically confirmed Smad6 variant.

**Method:** We have retrospectively reviewed the files of 8 children had a genetically confirmed Smad6 variant.

All of them received a multidisciplinary craniofacial management as well as a cardiologic evaluation and neuropsychological testing.

**Results:** There were 4 boys and 4 girls with different types of synostosis: 6 patients with trigonocephaly, 1 with scaphocephaly, 1 with oxycephaly. They all had been operated on (mean age at surgery: 11.6 months).

They were aged between 18 months and 49 months at the time of neuropsychological testing.

A great variability among individuals was found. We observed:

- a cognitive impairment in executive functions in all children over 30 months of age.

- a normal development with a weakness for global motor in the scaphocephalic patient.

- an early detectable and specific delayed expression of language and cognitive development related with executive functions difficulties in the trigonocephalic patients group and the oxycephalic patient.

Only the patient with oxycephaly showed an abnormal cardiac ultrasonography examination.

**Discussion:** Our results confirm the impact of Smad6 on the functional outcome of patients with craniosynostosis. An early recognition of neurodevelopmental issues, which can be obtained with appropriate tools, as well as long neuropsychological follow-up in craniosynostosis, are of paramount importance for the overall outcome of the patients highlighting the importance of a systematic genetic screening in these patients.

#### PP003

#### Biparietal remodelling or total vault remodelling in scaphocephaly? A comparative study using 3D Stereophotogrammetry

**Peter Spazzapan**, Tomaž Velnar

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**Introduction:** Scaphocephaly is the most commonly encountered type of nonsyndromic craniosynostosis. Many different surgical techniques have been developed through the last decades. 3D stereophotogrammetry allows to clearly define the outcomes of different surgical procedures.

**Methods:** From 2016 to 2020 two different techniques have been used to treat children with scaphocephaly: the Renier "H" technique and the total cranial vault remodeling technique. Stereophotogrammetry has been used to compare the outcomes of these two groups.

**Results:** 28 children have been operated with each technique. The first group was operated from 2016 to 2018, the second from 2018 to 2020. At the age of three years we analyzed the children of each group and compared their results with those obtained from a control group of 28 children without any cranial pathology. The measurements were performed in the axial, sagittal and coronal plane, to allow the determination of the outcome in all three planes.

**Conclusion:** 3D stereophotogrammetry is a non-invasive method that allows to define precisely the craniometric outcomes in craniosynostosis surgery.

#### PP004

#### A comparison of minimal invasive suturectomy with postoperative helmet therapy and extensive cranial vault remodelling for treatment of sagittal craniosynostosis

**Leonie Witters**, Mania De Praeter, Herman Vercruysee

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**Introduction:** So far, there has been no surgical technique proven to be superior regarding cosmetic outcome in the treatment of sagittal craniosynostosis. Surgical approach is primarily based on surgeon's

experience. In the authors' institution, total cranial vault remodeling (TCVR) was the standard of care. To minimise blood loss and morbidity, extended strip craniectomy without postoperative helmet therapy was performed for a while. However, a previous study showed no significant advantages of this technique over TCVR. Striving to further minimise morbidity while improving cosmetic outcome, minimally invasive suturectomy with postoperative helmet moulding was performed. The goal of this study is to compare morbidity and cosmetic outcome in TCVR and minimally invasive suturectomy with postoperative helmet moulding in patients with scaphocephaly. **Methods:** We performed a retrospective comparative cohort study. Forty-three patients with isolated sagittal craniosynostosis who underwent surgery at the Antwerp University Hospital between April 2008 and December 2022 were included. So far, TCVR was performed in 17 patients between 2008 and 2016, suturectomy was performed in 26 patients between 2016 and 2022. Data of interest were gender, age, preoperative cephalic index (CI) and head circumference (HC), duration of surgery, blood loss, transfusion needs, complications, length of stay, postoperative evolution of CI and HC, as well as cosmetic outcome according to the Cosmetic Outcome Scale. Data between the treatment groups were compared using SPSS software.

**Results:** Since the study is enrolling patients until December 2022, data are currently still being collected. Results will therefore be available in January 2023.

**Conclusion:** As to our knowledge, there have been no studies comparing minimal invasive suturectomy with postoperative helmet moulding to TCVR in scaphocephaly. As this procedure might offer a less invasive alternative to TCVR, it is worth comparing the two techniques. Results and conclusions of the study will be available in January 2023.

#### PP005

##### Bilateral fronto-orbital advancement vs a two-step surgery to treat unicoronal craniosynostosis

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**Introduction:** Unicoronal craniosynostosis (UCS) is characterized by premature fusion of the coronal suture, leading to retrusion of the forehead and orbital rim on the affected side and contralateral frontal bossing, orbital asymmetry and nasal deviation. Surgery is the standard treatment and aims at the correction of the craniofacial scoliosis. Our goal was to compare two surgical techniques: a standard one-stage surgery with bilateral fronto-orbital advancement (FOA) versus a two-stage surgery including early distraction with springs, followed by bilateral FOA. **Methods:** We retrospectively reviewed the clinical files of 23 pediatric patients who underwent surgical correction for UCS at Centro Hospitalar Universitário Lisboa Norte (CHULN) between January 2012 and December 2021. We compared the outcomes of two surgical procedures using computed tomography (CT). The craniometric measurements used were skull base twist (SBT) and midface twist (MFT). **Results:** 13 children were included in the study (7 submitted to one-stage surgery and 6 submitted to a two-stage surgery). In the first group, the mean SBT changed from 12,12 to 10,52 degrees, and the mean MFT improved from 13,16 degrees to 9,16. Children treated with a two-stage surgery had a more significant improvement in craniofacial deviation, even after the first procedure with spring-mediated distraction, without further complications. Mean SBT changed from 10,76 to 5,09 degrees and mean MFT changed from 12 degrees to 4,58.

**Conclusion:** Our study demonstrates that children who were treated with a two-stage surgery achieved greater improvement of craniofacial scoliosis.

#### PP006

##### Cranial Meningo-encephaloceles: Management, Associated Anomalies and Outcomes

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**Introduction:** Meningo-encephaloceles (MEC) are considered to be simple congenital neural tube defects. They are rare, and there is relatively limited understanding regarding their correlation with other associated anomalies, as well as the long-term outcomes.

**Methods:** A retrospective review of all cranial MEC surgically treated in our unit in the period 2013-2022 was conducted. Electronic records and radiological investigations were reviewed. Demographics, location and size of the lesion, associated CNS and other abnormalities, as well as long term outcomes were recorded.

**Results:** A total of n=16 patients underwent surgery for a cranial MEC. Average age at time of treatment was 8 months (range 1 day- 2.8 years). Location was infratentorial for n=11, and all but one were in the midline. Average maximum diameter was 4.3 cm (range 1-10cm). Larger MCs were treated earlier in life in all but one case. In n=7 (43%) there were other associated CNS abnormalities (30% for meningoceles and 75% for encephaloceles), with the most common one being a Dandy-Walker malformation variant. Less significant anomalies were seen in two patients (tethered tent, split superior sagittal sinus). Ventriculomegaly was present in four patients at time of presentation, while two developed hydrocephalus following the MEC repair. Four required a ventriculoperitoneal shunt. Surgical treatment was uncomplicated in all but one patient with the largest MEC, with a large bony defect who suffered a cardiorespiratory arrest intraoperatively, likely due to air embolus when the MC was opened. All children with other associated CNS abnormalities had significant developmental delay.

**Conclusion:** Cranial MECs can be associated with other significant CNS or systemic abnormalities and developmental delay, more frequently in cases that the MEC contains neural tissue. Surgical treatment is usually not challenging, but in cases of very large bone defects around the venous sinuses, morbidity can be increased due to intra-operative complications

#### PP007

##### Endoscopic ultrasonic aspiration of posterior fossa abscess in Dandy Walker Malformation. Case report

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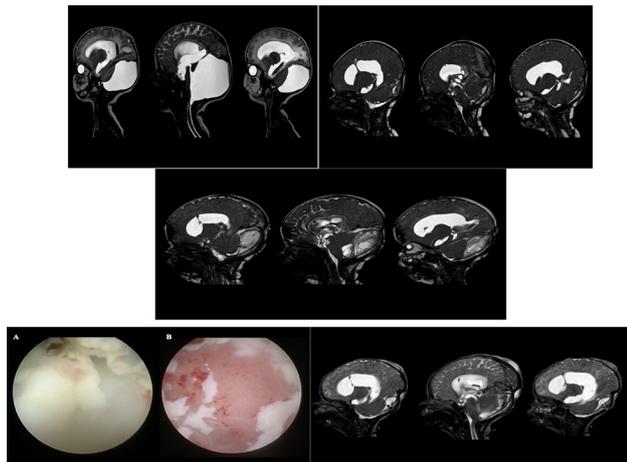
**Introduction:** Dandy-Walker malformation (DWM) is a posterior fossa malformation characterized by a huge posterior fossa cyst in communication with the fourth ventricle. Hydrocephalus is associated with more than 80% of cases and is usually treated by shunting. Despite infection being a common complication of the shunt, abscess formation within the cyst was reported only once.

**Methods:** A neonate affected by DWM developed a posterior fossa abscess following a shunt infection. The purulent collection was refractory to standard treatment (antibiotics and burr hole drainage); therefore, an endoscopic approach was performed in order to remove the purulent collection under direct vision. This material was aspirated with the help of an endoscopic ultrasonic aspirator.

**Results:** The outcome was favorable, with a resolution of infection and re-implantation of the ventriculo-peritoneal shunt. Surprisingly,

post-operative radiological examination showed substantial modification of the anatomy of the posterior fossa with disappearing of the Dandy-Walker cyst. To the best of our knowledge, this is the first documented report of a true Dandy-Walker malformation that modified its anatomical appearance over time.

**Conclusion:** Endoscopic aspiration of intracranial purulent collection should be considered a valid option to manage complicated cases. An endoscopic ultrasonic aspirator may make the procedure more effective and faster.



#### PP008

##### Erdheim-Chester Disease from diagnosis to management: case report and literature review

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**Introduction:** Erdheim-Chester Disease (ECD) is a rare multisystemic disease of unknown etiology, classified by the WHO as a histiocytic neoplasm.

**Methods:** ECD organ lesions usually result from local and systemic chronic inflammation and they typically manifest with multifocal sclerosis and foamy histiocytes on biopsy. The excessive production and accumulation of histiocytes result in dense fibrotic tissues, which can lead to organ failure if left without treatment.

In some patients, Erdheim-Chester Disease presents with an indolent course involving a limited number of systems, while in others it can be aggressive and affect multiple organs.

The disease involvement mainly includes the brain, skin, heart, long bones of the legs and arms, lungs, kidney, abdominal cavity, blood vessels, and adrenal glands. In this scenario, the most frequent causes of death are to be found in respiratory distress, pulmonary fibrosis, and heart failure.

However, the disease prognosis is variable due to the variety of clinical presentations and systems involved.

**Results:** ECD is considered a “non-Langerhans histiocytic multisystem disorder” and is thought to originate from the monocyte-macrophage cell lineage. Nevertheless, it is frequently associated with other systemic histiocytosis derived from Langerhans cells, as Langerhans Cell histiocytosis (LHC) occurs in 10 to 20% of ECD patients.

**Conclusion:** We report a case of Erdheim-Chester disease in a 20-year-old male.

#### PP009

##### The challenging treatment of a giant anterior meningoencephalocele in neonate: About one case

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 Ali Ait Idir Specialized Hospital, Algiers, Algeria

**Introduction:** Meningoencephalocele is an uncommon neural tube defect, called giant when the size of the lesion is greater than the head. The fronto-ethmoidal location is rare with possible involvement of anterior floor of the skull base. Surgical repair is complex with a functional and aesthetic interest.

**Clinical Case:** We present the case of a newborn with a giant congenital fronto-ethmoidal meningoencephalocele. The indication for urgent surgery in the neonatal period is motivated by breathing and feeding difficulties caused by the evolving mass. The exploration was done by MRI and a 3D cerebral CT-scan.

A puncture of the meningoencephalocele sac allowed the emptying of 1300 cc of hematic CSF. The closure of the defect and the cosmetic result were correct. However, despite transfusion and resuscitation measures, the patient presented hemodynamic instability complicated by death.

**Discussion:** The complexity of this procedure is due to difficulties with anesthesia and surgical aspects related to the neonatal period. The surgery includes 2 stages: the resection of the huge bulky sac, and a cosmetic step for the reconstruction of the bone defect. These 2 steps are laborious and life-threatening for a patient who has a precarious general preoperative condition due to poor diet and iterative cyanosis access. The early operative timing was imposed by the compression of the upper airways, the risk of sac rupture, and the difficulty of feeding and nursing. It seems more relevant to us to proceed in stages, an operative stage for the removal of the meningoencephalocele sac and the second stage is deferred for the repair of the bone defect.

**Conclusion:** Giant meningoencephalocele represents a therapeutic challenge in the neonatal period. The large volume of the mass will require emergency surgery because of the vital prognosis. Planning the surgical approach in stages will reduce the morbidity and mortality of this malformation.



**PP010**

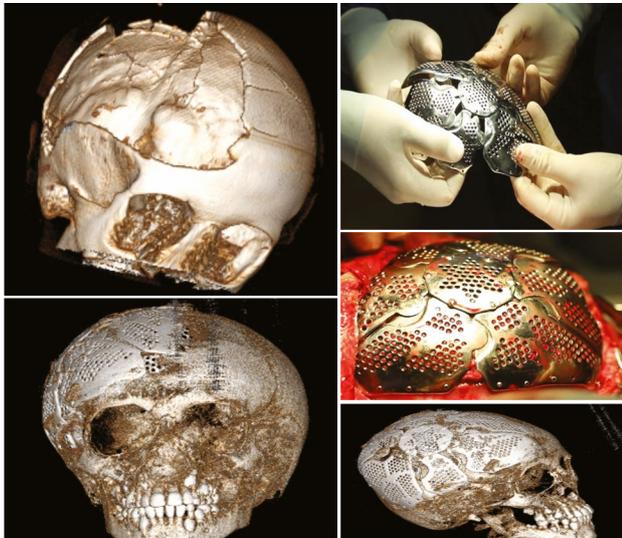
**Pediatric cranioplasty using an expandable titanium construction**  
**Paylak Sujyan<sup>1</sup>**, Lusine Avagyan<sup>2</sup>, Bagrat Baghdasaryan<sup>3</sup>, Mariam Avetisyan<sup>1</sup>, Margar Martirosyan<sup>1</sup>, Manya Sargsyan<sup>2</sup>  
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**Introduction:** Cranioplasty is a surgical repair of skull defects, which mainly arise from decompressive or resection craniectomy. It is important from the point of brain protection and satisfying cosmetic results. Cranioplasty with autologous bone flap is generally considered the reconstructive material of choice in pediatric patients. Nevertheless, there is a high risk of delayed bone resorption and infective complications. Artificial and synthetic cranioplasties may be considered as an alternative to initial bone flap reimplantation in the growing child. Another major factor is the implant's lack of ability to grow in parallel with the skull development, which can lead to skull asymmetry. Thus, we suggest a novel solution to the issue - using an expandable metal construction for cranioplasty.

**Methods:** In our study we noted that the ratio of the brain and facial parts of the skull is 8/1 in infants, and in adults it is about 1/1. It should be noted that the formation of the neurocranium is completed by 95% at the age of 10-12 years. To predict the development of the skull, was performed CT scan of the patient's parents.

**Results:** Using all the above factors, we produced an expandable titanium construction which has the possibility of asymmetric growth in all 360° throughout the skull growth in a large time interval. Fragmentation of the implant is designed along the line of the suture of the skull bones, as well as in difficult relief areas.

**Conclusion:** Cranioplasty using an expandable titanium construction is an effective option for correcting pediatric skull defects. It has more impact when used in the 6-12 year old children group, related to skull growth in the mentioned age group. Follow-up monitoring will be continued for these patients. Furthermore, expandable metal constructs are novel and promising alternatives for the skull defect correction of growing children.

**PP011**

**An illustrated clinico-radiological study of skull base disease in children with McCune-Albright syndrome**  
**Ian Coulter**, Marina Pitsika, Claire Wood, Tim Cheetham  
 Royal Victoria Infirmary, Newcastle Upon Tyne, United Kingdom

**Introduction:** McCune-Albright syndrome (MAS) is a rare multi-system condition, which typically includes fibrous dysplasia (FD) affecting the skeletal system, including the skull base. Such skull base disease may threaten neurological function. We aimed to review clinico-radiological aspects of the cranial disease in a cohort of paediatric patients managed by a specialist multidisciplinary clinic at our institution.

**Methods:** For this institutional service evaluation study, data were retrieved from patient records and radiological imaging investigations undertaken as part of the patient's management. Regions of skull disease were recorded as well as involvement of the cranial nerve foraminae on computerised tomography (CT) scans.

**Results:** Of the five children with a diagnosis of MAS managed at our institution, three cases have been diagnosed with skull base FD (Aged 4,5 & 7 years) thus far. Initial Isotope bone scans revealed polycystotic FD, including skull base/facial involvement. Subsequent CT imaging has demonstrated principal involvement of the clivus (3/3), temporal bones (2/3) and sphenoid bone (2/3) with sclerotic and expansile bony lesions evident. A single case has mild stenosis of a hypoglossal canal, otherwise significant cranial nerve foraminae involvement was not observed. One child underwent a cranial MRI scan which did not reveal any additional pathology. During the observed follow-up period (1-8 years), one patient has experienced a very mild facial asymmetry, but none have suffered from functional neurological impairment. As such, surgical treatment has not been required.

**Conclusions:** Once radiologically confirmed, we have employed clinical monitoring to manage our paediatric patients with MAS related skull base FD, rather than routine radiological surveillance. Herein we have illustrated some of the skull base bony pathology identified on imaging studies of patients with this rare disease, which can be managed conservatively if not causing functional impairment.

**PP012**

**Big data approach on the impact of COVID-19 lockdown on pediatric neurosurgery**

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**Objective:** COVID-19 lockdowns in 2020 limited activities of daily life. Data on the impact on healthcare systems is ambiguous. So far no data has been published for pediatric neurosurgery in Germany. We present the results of a big data approach.

**Methods:** We obtained anonymous data from the nationwide database of hospital statistics (German Federal Statistical Office, [www.destatis.de](http://www.destatis.de)) in August 2022. For the age group <18 years in 2016-2020, the following diagnoses (ICD-10 code) were analysed: Intracranial injury (S06), malignant (C71), benign (D33) and unspecified (D43) neoplasia, abuse (T74), violent attack (Y09) and, for comparison, forearm fracture (S52). The following operations (OPS code) were chosen: Operation of the central nervous system (CNS, 5-01-05), excision of intracranial lesion (5-015), shunt insertion (5-023),

shunt revision (5-024), plastic operations of the spinal cord (5-036) and, for comparison, overall number of operations (OPS-5), spondylodesis (5-836) and functional neurosurgery (5-028) at all ages.

**Results:** The number of operations declined from 17.23 million in 2019 to 15.82 million in 2020 (8.2%). CNS operations declined by 5%. Shunt insertion declined by 8.1%, revision by 5.1% and spinal cord operations by 6.6%, whereas excisions of intracranial lesions increased by 1.7%. Spondylodesis and functional operations both decreased by 11.9%. The hospitalisations for intracranial neoplasia declined by 8.8% compared to previous years. The number of inpatients with intracranial injury declined by 16%. Violent attack was also documented less often (-13.2%), but child abuse increased by 3.3%. Forearm fractures declined by 5.6%.

**Conclusion:** The COVID-19 lockdown in 2020 led to reduction of operations. Neuro-oncological operations were not affected. Intracranial injury even declined by 16%, reflecting limited activities and mobility. An increase in documented child abuse by 3.3% compared to the average of previous years was observed. These results help understand the impact of pandemics and political decisions and guide future decision-making.

## Poster Track 1.2: Spine / Vascular

### PP013

#### Infected intramedullary dermoid cyst in a toddler

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**Introduction:** Intramedullary Dermoid cysts are infrequently encountered while an infected Dermoid cyst in a toddler is extremely rare. These are difficult to recognize since presentation may be nonspecific and patients are unable to communicate effectively yet.

**Method and Results:** A review of medical records was done for a 15-month-old female who presented with decrease in sensorium, hemiplegia, and a cutaneous finding. Diagnosis shifted from Spina Bifida Occulta to a long segment infected intramedullary Dermoid Cyst. She underwent 3 surgeries which included CSF diversion, excision of the dermal sinus tract, and drainage of the abscess. Culture revealed heavy growth of *Bacillus subtilis* and appropriate antibiotic therapy was given.

**Conclusion:** Due to the paucity of cases and limited resources, recognition and delay in diagnosis may lead to catastrophe. A multidisciplinary approach ensures that optimal care is given in a timely fashion. Fortunately, at one year follow-up, motor strength has improved and milestones are at par with age.

### PP014

#### Isolated Intraspinal Juvenile Xanthogranuloma in a Toddler Presented with Acute Paraparesis

**Lusine Avagyan**<sup>1</sup>, Paylak Sujyan<sup>2</sup>, Eduard Asatryan<sup>2</sup>, Margar Martirosyan<sup>2</sup>, M Manya Sargsyan<sup>1</sup>, Mariam Avetisyan<sup>2</sup>

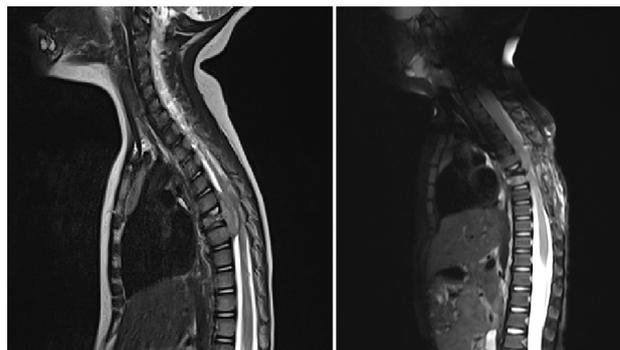
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**Introduction:** Juvenile xanthogranuloma was first described in 1905 by Adamson as a “congenital xanthoma multiplex”. It is a benign, non-Langerhans, histiocytic proliferative disease that predominantly affects the skin and mainly occurs in childhood. There are only about 100 cases reported. Central nervous system involvement is very rare.

**Methods:** 2-year-old girl was admitted to the hospital complaining of acute onset walking difficulties which worsened over 5 days. The child's anamnesis was remarkable with a history of being from a vegan family and not receiving any supplementation. She was hospitalized in the Neurology department. Blood labs reveal megaloblastic anemia with B12 deficit (< 50.00 pg/mL). Spine MRI with contrast enhancement showed pathological compression fracture of Th7 vertebral body, proliferative and structural changes with mainly intravertebral growth (approximately 28x20x9.5 mm) and spinal cord compression. The brain MRI was unremarkable. Tuberculosis excluded, differential diagnosis also includes schwannoma, meningioma, lymphoma, etc. Patient is treated with intramuscular B12. Th5-Th9 laminectomy and complete resection of the lesion was done. Postoperative immunohistochemical diagnosis was juvenile xanthogranuloma (CD68+, S100+, CD45+, CD56-, PHOX2V-). The lesion

**Results:** In consultation with an oncologist, we decided that chemotherapy and radiotherapy is not necessary at this point. No tumor recurrence or metastases were revealed on the follow-up spine MRI after one month. Dynamic MRI control is necessary. After surgery and rehabilitation course, the kid started to walk with individual spinal fixator.

**Conclusion:** This was the first case of intraspinal juvenile xanthogranuloma described in Armenia. A multidisciplinary approach, including neurosurgeons, ICU specialists, oncologists, is necessary for the diagnosis and management of these patients. More information is needed to understand the possible connection between vitamin B12 deficiency and neurological manifest in such cases.



### PP015

#### Management of tethered cord syndrome (TCS) in adult patients with previously untethered spinal lipoma

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**Introduction:** Management of patients born with spinal lipoma in most occasions involves early surgical untethering. Nevertheless, there are adult patients who were born a few decades ago and did not have the benefit of surgical untethering early in life. These patients continue to have problems from tethered cord syndrome throughout life. We analysed our experience with the neurosurgical management of adult patients who presented late with progressive neurological deficits due to spinal lipoma, who did not have untethering procedures early in life.

**Methods:** We performed a retrospective review of adult patients with previously untethered spinal lipoma, who were operated on between 2015-2022 and had an attempt to untether the spinal cord.

**Results:** There were 7 patients with previously untethered spinal lipoma (5 males) who presented at a mean age of 27,5 years (range:

19,7-46,4). Symptoms included leg pain (6 patients), leg weakness (5 patients), and bowel/bladder dysfunction (6 patients). All patients had orthopedic procedures in the legs before. Spinal cord partial untethering was performed in all. Complete untethering was not achieved in any patient. One patient had a second untethering procedure nine months after the first operation. At a mean follow up of 4,2 years (range: 1-7,6) all patients experienced pain relief, improvement of gait and balance. There was only one patient in whom bladder function improved.

**Conclusion:** Even late spinal cord untethering in patient with untreated spinal lipoma can achieve neurologic improvement significant enough to have a positive impact on daily life. Bladder function is difficult to improved after years of deterioration.

#### PP016

##### **Tethered cord syndrome secondary to type I diastematomyelia**

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**Introduction:** Diastematomyelia is a rare form of occult spinal dysraphism in which the spinal cord is separated into two parts. It is more frequent in the female sex (3:1). There are two types: type 1, the two parts of the spinal cord are in two different dural tubes, separated by a bony or cartilaginous septum; type 2, there is a single dural tube containing the two parts of the spinal cord, separated by a fibrous septum.

**Methods:** 15-month-old girl diagnosed with congenital clubfoot.

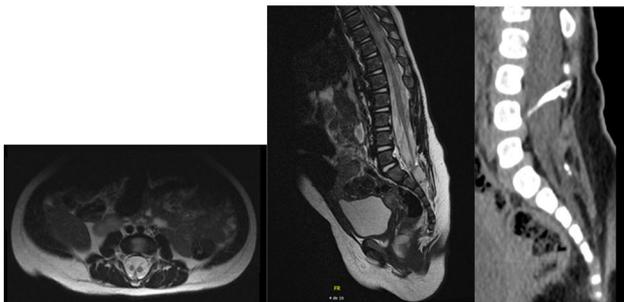
**Examination:** motor and sensory function preserved in the lower extremities. No sphincter alteration. No cutaneous stigma at dorsolumbar-sacral level.

**MRI/CT spine:** complete duplication of the spinal cord at L2- L4. Bony septum at the level of the posterior arch of L3 that runs in an anterior direction. Tethered cord at L5 level. Fusion defect of posterior L2-sacral arches (Image 1 and 2).

**Results:** Surgical procedure: extradural removal of the bony septum that divides the spinal cord at the level of L3. Untethering of the two parts of the spinal cord is performed resecting the dura around the bony septum (Image 3).

**Follow up:** the patient has a favorable postoperative evolution with no motor or sensory alteration in the lower extremities and no sphincter alteration.

**Conclusions:** Diastematomyelia type 1 is a very rare entity that belongs to the group of occult spinal dysraphisms. Early surgery could prevent irreversible long-term neurological sequelae.



#### PP017

##### **Multiple and laterally located dermal sinus tracts**

**Peter Spazzapan**

*University Medical Centre Ljubljana, Ljubljana, Slovenia*

Dermal sinus tracts (DST) are congenital lesions that connect the cutaneous ectoderm with the underlying neuroectodermal tissues. They are

usually solitary lesions located in the midline. Multiple and laterally located DST have been only rarely described.

**Methods:** Among all DST operated between 2015 and 2022 we have selected all multiple and laterally located DST.

**Results:** Two cases of multiple and laterally located DST have been treated. The first presented with meningitis and two tracts in the right buttock, one of which entered the spinal canal through the right S3 neuroforamen. The second child had three midline lumbar DSTs, one subcutaneous dermoid cyst and one intradural epidermoid cyst. In both cases surgical treatment was successful and the tracts have been completely excised.

**Conclusions:** Multiple and laterally located DST are rare congenital lesions that carry the same risks as the more common midline, single tracts. They need to be completely excised to avoid the risks of neurological deterioration, in particular due to infection. An incomplete process of disjunction does not sufficiently explain the embryogenesis of these lesions and different mechanisms, including a failure during secondary neurulation must be implicated.

#### PP018

##### **Surgical resection of conus lipomas - Results of an early Follow-Up**

**Peter Spazzapan**, Tomaž Velnar, Borut Prestor

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**Introduction:** Surgery of conus lipomas has been radically modified by the **Introduction** of intraoperative neuromonitoring and by the long-term results of radical resection, which has shown to guarantee a long term protection against the development of tethered cord syndrome. The technique of radical resection of conus lipomas has been adopted at our Institution in 2017 and the early results of our cohort of patients are here presented.

**Methods:** We analyzed retrospectively all children operated from 2017 to 2022 for a conus lipoma. The surgical technique adopted was based on the principles of complete untethering, complete removal of the lipoma, neurulation of the placode and creation of a wide dural sac.

**Results:** From 2017 to 2022 20 children have been surgically treated for a conus lipoma. Based on the Pang Classification, 10/20 were transitional, 8/20 were dorsal and 2/20 were chaotic. 17/20 were asymptomatic, 3/20 were symptomatic (urological deficits were present in 3 cases, motoric deficits and orthopedic deformations in 2 cases).

Median age at surgery was 6.4 years. An early postoperative worsening of clinical conditions has been experienced by 3/20 children: 1/20 developed a neurogenic bladder, 2/20 developed sensory deficits. In all these three cases the lipomas were of transitional type. We achieved a gross total removal in 17/20 cases, while in 3/20 a remnant was visible on postoperative MR.

Within a median follow up of 2.5 years no clear neurologic or neurological deterioration occurred, apart from the 3 children who had an immediate postoperative worsening.

**Conclusions:** Despite the short follow up of our series, the early results of radical resection of conus lipomas are promising. Complex transitional and chaotic lipomas represent a surgical challenge.

#### PP019

##### **Os odontoideum and craniovertebral junction instability secondary to dystonia: A case series**

**Michelle Kameda-Smith**, Dominic Thompson

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**Introduction:** Os odontoideum is a rare radiological description of a round ossicle detached from a hypoplastic odontoid process at the

body of the axis. The aetiology has been hotly debated and believed to either be congenital or result from trauma. Acute traumatic cervical cord injury has been observed in children with myelopathic instability secondary to os odontoideum and some authors have advocated for prophylactic treatment (i.e., fusion).

**Methods/Results:** We retrospectively reviewed the imaging features of 3 children presenting with severe dystonic cerebral palsy with clinical myelopathic deterioration secondary to atlantoaxial instability due to os odontoideum. This observation in a small cohort of children with dystonic cerebral palsy recommends radiological investigation of the cervicovertebral junction for os odontoideum as the cause of progressive myelopathy secondary to atlantoaxial instability. In fact one child had previous imaging prior to the dystonic episodes providing evidence for a non-congenital traumatic cause for development of os odontoideum.

**Conclusion:** In children developing progressive myelopathy with a history of severe dystonia, complete radiological investigation is critical in making the diagnosis and developing the appropriate management.

#### PP020

##### **Bulbocavernosus Reflex (BCR) & Clinical Outcomes in Children Under 2 Years during Tethered Cord Surgery (TCS)**

**Audrey Tan**<sup>1</sup>, Widayana Hanafi<sup>3</sup>, Bindu Govindarajan<sup>3</sup>, Chin Lik Tan<sup>1</sup>, Nivedh Dinesh<sup>1</sup>, Li Xuan Wong<sup>2</sup>, Noemia Pereira<sup>4</sup>, Tseng Tsai Yeo<sup>1,2</sup>, Vincent Nga<sup>1,2</sup>

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**Objective:** Cord de-tethering surgery may put patients at risk of developing bowel or urinary incontinence. BCR has been advocated for use to monitor the sacral reflex arc during TCS to reduce risk of bowel/urinary incontinence. We evaluate the usefulness of intraoperative BCR monitoring and its correlation with clinical outcomes in children < 2 years of age.

**Methods:** Retrospective evaluation of patients that underwent TCS from 2021-2022. Amplitude/latency of the BCR response (R1) and clinical assessment of patient's urinary/bowel function at post-op and 6-months follow-up were analysed.

**Results:** The patients (male=3, female=3), mean age of 7.5 months at surgery, were diagnosed with spinal cord lipoma (n=4) or lipomyelocoele with associated anorectal malformation (n=2). They presented with urinary dysfunction (50%) or bowel dysfunction (33.3%). Patients with reduced anal tone (latency:74.85ms, amplitude:18uv) has delayed latency and lower amplitude at baseline than those with urine dysfunction (latency: 40.17ms, amplitude: 57.7uv).

At the end of surgery, 5 patients had improvements in BCR with no worsening of BCR post-op. Clinically, 50% of patients had improved urinary function while it remained unchanged for the rest. 5/6 (83.3%) had no change in bowel function; 1 patient requires fleet enemas for bowel movement. Patients with anal tone reduction had baseline BCR readings of lower amplitude and increased latency. Patients with improved clinical urinary function had robust intraop R1 amplitude improvement (>20%) from baseline. Improvements in urinary function persisted at 6 months follow up.

**Conclusion:** BCR can be monitored in patients <2 years old adequately. Preliminary findings suggest improved R1 amplitude is associated with improved urinary function. Reduced latency on the other hand did not translate into improved clinical function.

#### PP021

##### **The use of bovine pericardium as dural substitute in pediatric neurosurgery: experience with 108 cases**

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<sup>1</sup>Hospital da Criança Santo, Complexo Hospitalar Santa Casa, Porto Alegre, Brazil, <sup>2</sup>Medical School, Hospital de Clínicas de Porto Alegre (HCPA) Universidade Federal do Rio Grande do Sul (UFRGS), Porto Alegre, Brazil, <sup>3</sup>Hospital Ernesto Dornelles, Porto Alegre, Brazil

**Introduction:** The lack of adequate closure of the dura-mater in neurosurgical procedures is essential. If it is not possible to perform a primary dural, the use of dural substitute graft for duroplasty is necessary. Grafts from the patient himself are preferred. Several synthetic materials are available, usually more expensive. Bovine pericardium (BP) is an option, even for large defects, widely available and at a low cost. The aim of the study is to evaluate the use of BP as a dural graft in cranial and spinal neurosurgical procedures in pediatric patients.

**Methods:** 108 patients were studied retrospectively from August 2012 to December 2021. Variables collected were: sex, age, location of the procedure, intracranial infra or supratentorial and spinal. Also, complications were accessed, especially CSF leak and meningitis, use of external ventricular drainage (EVD) and necessity to remove the graft.

**Results:** Intracranial procedures were 96 (89%) with 71 infratentorial and 25 supratentorial, and 12 (11%) intraspinal procedures. CSF leak was the most common complication (n=34), as expected, 23/71 (32%) in infratentorial, 4/25 (16%) in supratentorial procedures and 7/12 (58%) in intraspinal procedures. 12/34 (35%) patients needed additional surgery to repair the CSF leak with reinforcing sutures in the graft. Meningitis occurred in 16/108 (15%). In patients with meningitis 15/16 occurred in infratentorial, including all the 5 cases confirmed by culture. From 71 patients with infratentorial approach, 34 (48%) received an EVD, 11 had a CSF leak and 9 received treatment for a presumed/proved meningitis. This suggests that the leak and infection were more related to location infratentorial and use of EVD than the use of BP (p<0,05). None of the 108 patients needed to have the BP graft removed.

**Conclusion:** The use of BP as graft for dural substitute is an effective alternative for duroplasty in neurosurgical pediatric patients, both in intracranial and intraspinal procedures.

#### PP022

##### **A ventral brainstem neurenteric cyst - case report and surgical video**

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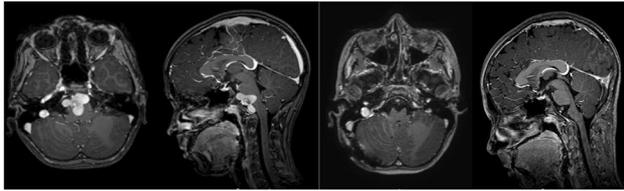
**Introduction:** Neurenteric cysts are uncommon, benign lesions that result from aberrant embryologic development of the notochord. They are typically located ventral to the spinal cord and rarely located intracranially. We present a case of a 10-year-old with a ventral brainstem neurenteric cyst.

**Methods:** The clinical information was retrieved from the patient clinical file and the video edited from the surgical procedure.

**Results:** 10-year-old healthy female child presenting with recurrent headaches. The physical exam was unremarkable. CT imaging showed a cystic lesion anterior to the brainstem. MRI showed a T1 and T2 hyperintense mass anterior to the pontomedullary junction occupying the pre-bulbar and pre-pontine cistern with no enhancement or restricted diffusion. The prepontine vasculature was anteriorly displaced and not encased. Due to progressive enlargement of the lesion with mass effect on the brainstem, a surgical treatment was considered. The patient underwent a right retrosigmoid craniotomy with extension to the foramen magnum. The cyst capsule was fenestrated and a yellow thick fluid was drained. Part of the cyst wall was adherent to the brainstem and basilar artery and was not removed. Histology revealed

a pseudo-stratified epithelium with no anaplasia, consistent with the diagnosis of a benign endodermal cyst. In the postoperative period, the patient experienced no neurological alterations.

**Conclusion:** We report a case of a rare congenital cyst in the ventral brainstem with a surgical video. Complete resection is ideal to reduce the risk of recurrence, although this may be precluded by adherence to critical structures.



### Poster Track 1.3: Trauma/ Hydrocephalus

#### PP023

#### Traumatic isolated skull fractures in infant: a 5 years retrospective study in a level I pediatric trauma center

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**Introduction:** Head trauma is the most frequent pediatric trauma. 95% are minor. Of these, 6% are complicated with a skull fracture. In France, there is no guidelines for the management of isolated skull fracture. Large series of children have shown few complications and suggest that patient can be discharged home after clinical observation at the emergency. At the Hospital Femme Mère Enfant, the current protocol is to hospitalize the child few days and request ophthalmologic (fundus) and electroencephalography (EEG) examinations. The aim of this study was to evaluate the complication rate of infants with isolated skull fracture at our institution.

**Methods:** This was a 5-year retrospective study. Criteria for complication were as follow: death, surgery, modification of the neurological status. For all patient, we included their results of the fundus and EEG.

**Results:** The complication rate was 4.3% during the hospitalization (4 of 93 patients). Three patients underwent surgery for a depressed skull fracture seen on the first computerized tomography (CT). One patient really got complicated within the first 24h after the trauma. He required a transfer to the ICU and underwent emergency surgery for an acute obstructive hydrocephalus due to a posterior fossa hematoma. A diagnosis of severe haemophilia A was made. For all patients, the fundus and EEG were normal. In the subgroup of patients consulting  $\geq 24$  hours after the head trauma, none had any complications during hospitalization.

**Conclusions:** The rate of life-threatening complication after an isolated skull fracture is very low. Systematic fundus and EEG examination seem not to be indicated in absence of a suspicion of epileptic seizure for the EEG or non-accidental head trauma for the fundus. However, hospitalization surveillance for at least 24h seems to be compulsory for trauma that happened less than 24h before the consultation at the emergency unit.

#### PP024

#### Growing Skull Fracture of the Posterior Fossa - case report and review of the literature

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**Introduction:** Head trauma with skull fracture is frequent in children. Of these, less than 0,1 % develop growing skull fractures, which are rare in the posterior fossa.

**Methods:** Case report and literature review.

**Results:** We report the clinical case of a 3 year-old boy who fell from a second floor window presenting with a moderate traumatic brain injury. At admission, he presented a pediatric Glasgow Coma Scale (GCS) of 10, without focal neurological deficits. The brain CT scan showed a right parieto-occipital subarachnoid hemorrhage and cortical contusion, a linear fracture extending from the occipital bone until the foramen magnum on the right, and small bone depression adjacent to a right cerebellar contusion. The child was admitted in the ICU, improved to a GCS 15 and was treated conservatively. He was discharged a week after admission, with a significant improvement of the head trauma lesions and without neurological deficits.

During follow up in the outpatient clinic, approximately 1 year after the head trauma, a small skull depression was noticed in the right occipital bone. The head CT scan revealed a growing skull fracture and an adjacent hypodense area corresponding to the previous right cerebellar contusion. The patient underwent surgical repair of an extensive dural tear underneath the right occipital fracture and cranioplasty. The post-operative period was uneventful.

**Conclusion:** We report a rare case of a posterior fossa growing skull fracture, discuss the surgical technique and provide a literature review.

#### PP025

#### Circumscribed thinning of the calvaria - a long-term sequela after vacuum extraction delivery

**Benjamin Liniger**

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**Introduction:** Vacuum extraction deliveries are associated with a number of complications like injuries to the skin, subperiosteal, subgaleal and intracranial hemorrhages. Even skull fractures have been described, but data on persistent changes in the bone structure of the skull in terms of thinning of the calvaria is lacking in the literature.

**Methods:** We present a series of 5 children, who were referred because of an irregularity of the cranial surface without other symptoms. The investigations revealed a circumscribed thinning of the calvaria in all 5 cases. Detailed history and the specific clinical findings were crucial in the assessment. Various imaging techniques were used to visualize the lesions and rule out some differential diagnoses.

**Results:** All children had undergone a vacuum-assisted delivery. All irregularities of the cranial surface were discovered incidentally. Clinical examination revealed a round (3) or linear (1) channel-shaped indentation of the calvaria in 4 cases. The shape and size of the annular thinning of the calvaria matched devices commonly used in vacuum extraction very well. In one patient, a localized protrusion of the calvaria was investigated, but an indentation more posteriorly was detected in the CT scan. CT scan was performed in 4 patients, in one, parents refused this examination. In the follow-up, no progression or symptoms was observed in any of the patients.

**Conclusion:** Such long-term sequelae after vacuum-assisted delivery have not been reported yet and the underlying mechanism is unclear. In children who present with irregularities of the cranium, an accurate history, including the mode of birth, and detailed clinical examination should be performed. Knowledge about the presented pathology may help to avoid extensive investigations, particularly CT scan. The

publication of further cases could help to propagate the circumscribed thinning of the calvaria after vacuum extraction as a distinctive clinical entity.

#### PP026

##### **Sleep disorders in young male patients after craniocerebral injuries**

**Nikolaos Syrmos**

*Aristotle University of Thessaloniki, Greece*

**Introduction:** Sleep disorders are serious post traumatic conditions. The signs and symptoms of sleep may include excessive daytime sleepiness, irregular breathing, increased movement during sleep, irregular sleep, abnormal wake cycle, difficulty falling asleep and also others situations.

**Material and Methods:** 20 cases of sleep disorders in young male (<30 years old) patients after craniocerebral injuries are presented and discussed. 5 cases of insomnia (25%), 5 cases of sleep apnea (25%), 4 cases of restless legs syndrome (RLS) (20%), 6 cases of narcolepsy (30%).

**Results:** We perform appropriate neurological, psychiatric and radiological evaluation with ct and mri studies in all 20 patients. In 4 cases we observe amelioration through six months period

**Conclusions:** Disordered sleep is a common phenomenon after craniocerebral injuries. Sleep disruption contributes to morbidity, development of neurocognitive - neurobehavioral deficits, and prolongs the recovery phase after the initial traumatic situation. Appropriate recognition and correction of these problems may limit the secondary effects of craniocerebral injuries and improve neuro recovery/patient outcomes. Collaboration with other medical disciplines is necessary in order to achieve optimal resorders in order to ameliorate the quality of life of young patients.

#### PP027

##### **Traumatic brain injuries in young male soccer players**

**Nikolaos Syrmos**, Georgios Ampatzidis

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Aim of this study was to evaluate cases with traumatic brain injuries in young male soccer players. 12 young players, range of age 14-24 years old, mean age 18,5 years old were participated in this study. 6 cases of brain concussions (50%), 4 cases of skull fractures (33,3%) and 2 small intracranial hematomas (16,7%) are presented, documented and discussed. Traumatic brain injuries are emergency sports situations and they need accurate management and specific health care approach.

#### PP028

##### **Endoscopic endonasal approach for skull base lesions in children**

**Amets Sagarrabay**, Dalila Forte, Miguel Correia, Jorge Mexia, Heredito Sousa, Mário Matos

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**Introduction:** Endoscopic endonasal approaches for pituitary tumors and skull base have gained importance in the last 25 years in the field of skull base surgery, mostly in adults. In pediatric population, due to relevant differences and inherent difficulties, these approaches grew in a slower rate but, nowadays, there is an increasing interest and many groups are working and improving this area of Neurosurgery.

**Methods:** the authors reviewed a consecutive serie of 18 endoscopic endonasal approaches in and review literature to analyze surgical technique, anatomical corridors and its differences depending on age and diseases.

**Results:** a consecutive serie of 18 endoscopic endonasal approaches in 14 pediatric patients were analyzed. There were 11 cases of level 2, 4 cases of level 3 and 3 cases of level 4 endoscopic endonasal complexity

approaches, respectively. The diseases treated by these approaches were, in order of frequency: craniopharyngiomas (33%), CSF leaks (22%), pituitary adenomas (17%), dermoid cyst (11%), xanthogranuloma (5%), and occipito-cervical instability (5%). The authors analyze the serie and show surgical technique and nuances in pediatric population

**Conclusions:** Endoscopic endonasal approaches are suitable and safe in pediatric population and may be the gold standard in many pediatric skull base diseases.

#### PP029

##### **A rare manifestation of infant cerebral venous outflow obstruction: a case report**

**Mohammed Ashraf Patel**, Mohsen Yaish, Shungu Ushewokunze

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**Introduction:** Cerebral venous outflow obstruction has been linked to enlarged subarachnoid spaces on neuro-imaging (Sainz et al., 2018), however these findings have largely been termed “benign” with or without the label of “external hydrocephalus” (Marino et al., 2014). Also examined in the literature is anatomical variation in dural venous sinuses for these patients (Sainz et al., 2021; Cinalli et al., 2021). We report on an exceedingly rare case where the combination of these was anything but benign.

**Case Presentation:** A term Caucasian male was born via planned Caesarean section. On day-1-of-life, he was found to have a tracheo-oesophageal fistula and oesophageal atresia. This was corrected surgically, and subsequent management involved oesophageal dilatation for a residual stricture. Of note, he was also on the spectrum of VACTERL association. He developed a squint at 10 weeks and was referred routinely to Ophthalmology clinic. During a General Surgical admission in October 2022 for vomiting and cyanotic episodes, an incidental review by a Neurosurgeon raised concerns of raised ICP. On examination, he had a marked convergent gaze and bulging anterior fontanelle with prominent scalp veins. Head circumference had crossed centiles (now above the 91st). Urgent CT scan was reported as consistent with “benign external hydrocephalus”. Urgent Ophthalmology review confirmed bilateral abducens palsies and papilloedema. MRI/MR venography revealed complete non-visualisation of venous sinuses except a small portion of patent superior sagittal and right transverse sinuses, and all venous outflow was via collaterals to the external jugulars.

An emergent VP shunt was inserted resolving the papilloedema, and patch retraining has corrected the abducens palsies. He remains under Neurosurgery follow-up with anticoagulation initiated by Neurology.

**Discussion:** We have encountered multiple patients with radiological appearance of “benign external hydrocephalus” who have required CSF diversion to manage raised ICP. MRV and Ophthalmology should be performed for this cohort.

#### PP030

##### **Communicating Hydrocephalus and Raised Intracranial Pressure in Association with Multi-Systemic Smooth Muscle Dysfunction Syndrome (MSMDS)**

**Anthony Roylance**<sup>1</sup>, Miss Benedetta Pettorini

<sup>1</sup>*Alder Hey NHS Foundation Trust and Charity, Liverpool, United Kingdom*

**Introduction:** Multi-System Smooth Muscle Dysfunction Syndrome (MSMDS) is a condition resulting in reduced smooth muscle contractility with less than 50 cases documented worldwide. As of yet, there are no cases with associated hydrocephalus. Smooth muscle contraction relies on calcium mediated calmodulin activation, actin-myosin interaction and subsequent cross-bridging. Alpha-smooth muscle actin isotype 2 (ACTA2) is one of six actin isoforms, which along with gamma-smooth muscle actin (ACTG), is preferentially expressed in

smooth muscle. Mutations involving arginine lead to more widespread involvement; Arg179His being the most common and severe, and demonstrating a predilection for neurovascular phenotype.

**Methods:** The first case of a patient with MSMDs and hydrocephalus is presented. Investigations, management and proposed follow up is discussed.

**Results:** Ventriculomegaly on MR was seen with raised intracranial pressure on ICP monitoring. Ventriculoperitoneal shunt insertion was performed without issue.

**Discussion:** Peri-ventricular white matter small vessel disease is seen in 95% patients with MSMDs. Segmental intracranial artery stenosis is seen in 77%. Pathological sequences similar to Moyamoya disease are described, yet certain studies demonstrate de-novo ACTA2 missense mutations lead to vascular changes distinct from Moyamoya with an absence of stereotypical basal collaterals. The pathological process in this case likely represents a gradually worsening communicating hydrocephalus secondary to compromised re-absorption, occurring at the level of the arachnoid granulations due to poor direct trans-ependymal reabsorption at the periventricular venous network, and globally compromised venous return as a result of cardiopulmonary disease. Periventricular leukomalacia culminates in a reduction of the reciprocal pressure to physiological ventricular CSF hydrostatic pressure normally provided by healthy white matter. Ependymal scarring secondary to peri-ventricular gliosis has been associated with this phenomenon and is a potential factor in this case. However, these factors suggest MSMDs patients would exhibit a normal pressure or ex-vacuo dilatation of the ventricles, which has been disproven by this case.

#### PP031

##### CSF Surfactant Protein Changes in Preterm Infants After Intraventricular Hemorrhage

**Matthias Krause**

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**Introduction:** Surfactant proteins (SP) have been shown to be inherent proteins of the human CNS and are altered during acute and chronic disturbances of CSF circulation. Aim of the study was to examine the changes of surfactant protein concentrations in CSF of preterm babies suffering from intraventricular hemorrhage.

**Patients and Methods:** Consecutive CSF samples of 21 preterm infants with intraventricular hemorrhages (IVH) and posthemorrhagic hydrocephalus (PHHC) were collected at primary intervention, after 5-10 days and at time of shunt insertion ~50 days after hemorrhage. Samples were analyzed for surfactant proteins A, B, C, and G by ELISA assays and the results were compared to 35 hydrocephalus patients (HC) without hemorrhage and 6 newborn control patients.

**Results and Discussion:** Premature patients with IVH showed a significant elevation of surfactant proteins SP-A, C, and G compared to HC and control groups: mean values for the respective groups were SP-A 4.19 vs. 1.08 vs. 0.38 ng/ml. Mean SP-C 3.63 vs. 1.47 vs. 0.48 ng/ml. Mean SP-G 3.86 vs. 0.17 vs. 0.2 ng/ml. SP-A and G concentrations were slowly falling over time without reaching normal values. SP-C levels declined faster following neurosurgical interventions and reached levels comparable to those of hydrocephalus patients without hemorrhage.

**Conclusion:** Intraventricular hemorrhages of premature infants cause posthemorrhagic CSF flow disturbance and are associated with highly significant elevations of surfactant proteins A, C, and G independent of total CSF protein concentrations.

#### PP032

##### The Cerebral Surfactant System and Its Alteration in Hydrocephalic Conditions

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**Introduction:** Pulmonary Surfactant reduces surface tension in the terminal airways thus facilitating breathing and contributes to host's innate immunity. Surfactant Proteins (SP) A, B, C and D were recently identified as inherent proteins of the CNS. Aim of the study was to investigate cerebrospinal fluid (CSF) SP levels in hydrocephalus patients compared to normal subjects.

**Patients and methods:** CSF SP A-D levels were quantified using commercially available ELISA kits in 126 patients (0-84 years, mean 39 years). 60 patients without CNS pathologies served as a control group. Hydrocephalus patients were separated in aqueductal stenosis (AQS, n = 24), acute hydrocephalus without aqueductal stenosis (acute HC w/o AQS, n = 16) and idiopathic normal pressure hydrocephalus (NPH, n = 20). Furthermore, six patients with pseudotumor cerebri were investigated.

**Results:** SP A-D are present under physiological conditions in human CSF. SP-A is elevated in diseases accompanied by ventricular enlargement (AQS, acute HC w/o AQS) in a significant manner (0.67, 1.21 vs 0.38 ng/ml in control, p<0.001). SP-C is also elevated in hydrocephalic conditions (AQS, acute HC w/o AQS; 0.87, 1.71 vs. 0.48 ng/ml in controls, p<0.001) and in Pseudotumor cerebri (1.26 vs. 0.48 ng/ml in controls, p<0.01). SP-B and SP-D did not show significant alterations.

**Conclusion:** The present study confirms the presence of SPs in human CSF. There are significant changes of SP-A and SP-C levels in diseases affecting brain water circulation and elevation of intracranial pressure. Cause of the alterations, underlying regulatory mechanisms, as well as diagnostic and therapeutic consequences of cerebral SP's requires further thorough investigations.

#### PP033

##### Surgical management and genetic analysis in monozygotic female twins with hydrocephalus and suprasellar cysts

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**Introduction:** We report virtually identical clinical and radiological presentations of supratentorial hydrocephalus and cystic formations from the suprasellar cistern in monozygotic female twins. Hydrocephalus affecting both twins is rare and genetic predispositions and possible exposures during pregnancy should be evaluated. Moreover, the surgical treatment of this complex condition constitutes a point of discussion.

**Methods:** Information of pregnancy-related exposures were retrieved from the parents and the patient files. Whole-exome sequencing (trio analyses) were performed with screening for pathogenic variants in 121 hydrocephalus candidate genes, and for pathogenic variants in all known disease-associated genes. Variants were classified according to ACMG criteria. A two-step surgical management was chosen with initial placement of ventriculoperitoneal shunts in both children with optional later fenestration of the cysts.

**Results:** Maternal epilepsy and treatment thereof were known exposures during pregnancy but are not established risk factors for CNS malformations. The extensive genetic analyses revealed no pathogenic variants in known disease-associated genes nor in candidate genes for hydrocephalus. Due to increasingly enlarged head circumference and regurgitation at 9 months of age, both children were treated with a ventriculoperitoneal shunt. Both children displayed close to normal development thereafter, but with minor visual impairment and slight motor delays. Brain MRI's were repeated at 4 years of age and revealed enlargement of the suprasellar cyst in both children, which have led to planning of endoscopic fenestration.

**Conclusion:** Surgical management of complex hydrocephalus must be based on tailored solutions. Genetic analysis is of importance to rule out inherited traits and to understand disease mechanisms. Funding: None

## Poster Track 2.1: Epilepsy

### PP034

#### Drug-resistant epilepsy secondary to Delalande type IV giant hypothalamic hamartoma

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<sup>1</sup>Complejo Asistencial Universitario De Salamanca, Salamanca, Spain,

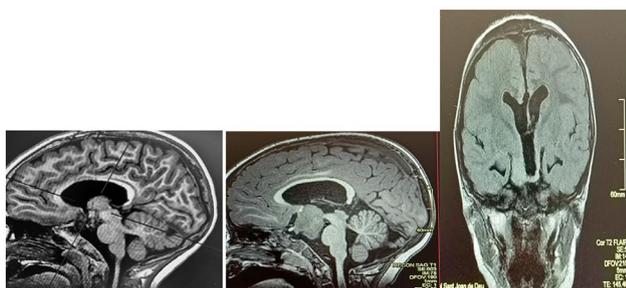
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**Introduction:** We present a case of a right-handed 5-year-old patient suffering from structural focal epilepsy with gelastic focal emotional seizures, starting at 14 months in the context of Delalande type IV giant hypothalamic hamartoma.

**Methods:** As this is a drug-resistant epilepsy, several epilepsy surgeries were performed: first, a resection surgery in 2019, followed by laser ablation surgery in 2020, and laser ablation surgery in 2021 (2 fibers).

**Results:** Epilepsy crises have not been completely controlled and some complications have occurred, such as third cranial nerve palsy of the left eye and placement of a ventriculoperitoneal shunt set to an opening pressure of 110 mm H<sub>2</sub>O after the first surgery. In addition, the patient presents a delay in cognitive development with associated ASD symptoms.

**Conclusions:** Given the symptoms of persistent seizures after the last surgery, with episodes of guttural sounds, babbling, initially of short duration and low frequency and subsequently longer and more frequent, on some occasions with ocular retroversion, accompanied by stereotyped head movements, without predominance of one side and without any other accompanying movement, and on some occasions with an unmotivated smile, with possible disconnection from his surroundings. Prolonged video-EEG monitoring shows epileptic seizures. In the neuropsychological evaluation, a delay in neurodevelopment was observed. Compared to the assessment from a year before, a slow evolution and a decrease in typical scores are observed, which imply a greater cognitive distancing from his peers. At the 3T brain MRI post-surgical changes can be seen due to laser treatment, with persistence of the right rear and lateral connections and smaller, laminar, left lateral connections. When an attempt was made to reduce the dose of carbamazepine the episodes increased in frequency. An attempt has been made to make a therapeutic adjustment with lacosamide that has not been effective, and reoperation has been indicated.



### PP035

#### Non-pharmacological treatment options of drug-resistant epilepsy in subcortical band heterotopia

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**Introduction:** Doublecortin (DCX) is a neuronal migration protein encoded by the DCX gene.

DCX-related and X-linked neuronal migration disorders comprise classic lissencephaly and subcortical band heterotopia (SBH) which primarily affects females and is often associated with drug-resistant epilepsy. SBH is also known as double cortex and subcortical laminar heterotopia

The aim of this study is to review the literature for non-pharmacological treatment options of drug-resistant epilepsy in SBH.

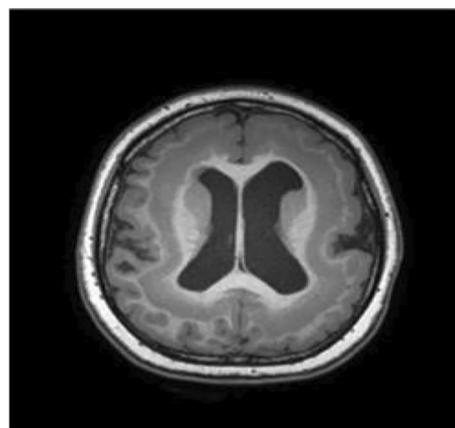
**Methods:** Following PRISMA guidelines we performed a systematic review in Scopus and PubMed databases and describe a case report, managed at our institution.

**Results:** Twenty-six patients who underwent 29 non-pharmacological treatments were reported.

Mean age at seizure onset: 6.5 years (range 0.2-23) with female sex predilection (5.25:1). Mean age at invasive or non-invasive treatment: 21.5 years (range 6.5-51).

Ten patients underwent corpus callosotomy (one two-staged), 8 had temporal lobectomy with amygdalohippocampectomy. Two patients respectively had focal cortical resection, multiple subpial transections, insertion of a vagal nerve stimulator, or deep brain stimulation of the bilateral anterior nuclei of the thalamus. One patient underwent responsive focal neurostimulation, another had transcutaneous stimulation of the vagal nerve. Nineteen patients reported a reduction or the disappearance of the seizures, 1 patient had no improvement. The outcome of 2 patients was class I, of 1 patient class II, of 4 patients class III, and of 6 patients class IV according to the Engel Epilepsy Surgery Outcome Scale.

**Conclusion:** Corpus callosotomy and temporal lobectomy have been the predominant non-pharmacological treatment. The small number of cases reported precludes drawing a firm conclusion as to which non-pharmacological treatment is the superior option for drug-resistant epilepsy in patients with SBH. The future relevance of procedures such as deep brain stimulation, responsive focal neurostimulation, transcutaneous stimulation of the vagal nerve, and also stereotactic laser interstitial thermal therapy remain to be seen.



### PP036

#### Laser ablation for focal epilepsy from deep lesions - outcome and complications

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**Introduction:** Laser ablation (LA) is a minimal-invasive treatment option for focal epilepsy, introduced in Europe in 2018. This study reports seizure outcome, complications and hospital stay after LA.

**Methods:** Patients aged 0–30 years treated with LA for epilepsy at Sahl-grenska University Hospital were followed for at least 6 months after treatment. Seizure outcome (Engel class) and complications were noted.

**Results:** Eight treatments were done for epilepsy in 7 patients, age 2–30 y (mean 16.5 y). A single catheter was used in all patients, 5/8 catheters had a 3-mm window, 3/8 had 10-mm window. Lesions included two hypothalamic hamartomas, four insular dysplasias on (4/4 left) and one low grade tumor in amygdala/hippocampus. Seizure outcome was Engel I in 5/7 after 1 month, and 3/7 after 6 months. At six months, Engel II was seen in two patients and Engel III in two. Complications were seen in three patients, one mild hemiparesis, one oculomotor nerve palsy and one short-term memory deficit, all resolved in three months. Total hospital stay was 3.3 days (range 3–5).

**Conclusion:** LA is a safe minimally invasive option for deeply located epileptic lesions, 5/7 patients had >75% seizure reduction, 3/7 were seizure-free. Hospital stay was short and patients recovered quickly. Neurological deficits occurred in 3/7 patients, all resolved within 3 months. LA can be staged and repeated if needed to create a more extended lesion.

Source of funding: The study was financed by grants from the Swedish state under the agreement between the Swedish government and the county councils, the ALF-agreement.

#### PP037

##### Vagus nerve stimulation in children – 15 years experience in Brno Epilepsy Center

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**Introduction:** Vagus nerve stimulation (VNS) is a standard lege artis palliative surgical treatment method in patients with pharmacoresistant epilepsy, in whom resection with curative intent is not possible. VNS is fully established in adult epileptology and is increasingly used in the treatment of pediatric patients, even in the youngest age categories.

**Methods:** A total of 55 pediatric patients have been implanted in the Brno Epilepsy Center since 2005 (average age at the time of implantation 10.85 years), while at least 1-year outcome can be evaluated in 46 and 3-year outcome in 33 patients. The number of responders is 52.17% (24/46 pts) one year after surgery, and 57.58% three years after surgery (19/33 pts).

**Results:** "Response-rate" depending on VNS implantation before and after the tenth year of age does not show a statistically significant difference (p value = 0.7675, respectively 0.296), also a comparison of the effectiveness of VNS in patients with focal (22 pts) versus multifocal epilepsy, respectively epileptic encephalopathy (23 pts, of which 15 with Lennox-Gastaut syndrome) is not statistically significant (59.09% versus 47.82%; p value = 0.5544), which gives a promising chance for improvement even for patients with focal epilepsy with extensive epileptogenic zone or patients after resection with an unsatisfactory result. Reported improvement in the quality of life in 73.17% of treated patients need to be considered as indisputable benefit, reflecting the disappearance of the most limiting type of seizure, shortening of seizures or post-seizure fatigue, reduction of the number of acute hospitalizations, etc. positive influence on condition, attention and mood.

**Conclusion:** Vagal stimulation is therefore a safe and effective treatment even for children under 10 years of age; the effectiveness increases with the duration of chronic stimulation. In addition,

technical advances bring new possibilities, which mean a simplification of the entire management and better treatment results.

#### PP038

##### VNS - Vagus Nerve Stimulatuon in Epilepsy

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**Introduction.** Epilepsy resistant to the medical treatment and other surgical procedures are also impossible, could be indication for the vagus nerve stimulation implant (VNS). History of the VNS started in humans in 1988 with the first implantation. Presentation of the study was in 1992. In 1994 was done EU registration and in 1997 US. In 2007 started the history of VNS in Slovakia.

**Methods.** VNS consist of generator and electrode and it is implanted at the left nervus vagus. We present our first experiences in the group of 59 children with VNS. The age at the time of implantation was in the average 9,8 years (2,2–17,8). Duration of epilepsy was similar - in average 6,7 years (1,5–16,7).

Effectiveness of the VNS after the implantation is to reduce the seizures more than 50%.

**Results:** In our series the effectiveness is 64%. It vary after operation and the best is within the first 12 months. Than it goes slightly down. It was better in the patients with early indication for surgery. 5% of our patients are seizures free. Complications are not so high. In two patients local with wound and in two other patients we have to remove the generator and electrode.

**Conclusion:** Vagus nerve stimulation - VNS remains suitable method in the management of Epilepsy in children. Earlier implantation seems to be more effective.

#### PP039

##### Use of antiepileptic drugs as prophylaxis against posttraumatic seizures in the pediatric population, a systematic review and meta-analysis

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**Objective:** We aim to assess the effect of anti-epileptic drug, "AED", prophylaxis for early or late post traumatic seizures, targeting the pediatric population with traumatic brain injury, "TBI".

**Design/Methods:** We systematically searched for studies reporting the incidence of posttraumatic seizures in pediatric patients who suffered from TBI and received AEDs prophylactically following their TBI incident. Studies that included adult patients, adult and pediatric patients but did not report results for the pediatric population separately and patients who did not suffer from a TBI, were excluded. Studies that did not indicate the use of antiepileptic drugs prophylactically following TBI were excluded.

**Results:** A total of 10 studies were included involving 4621 post traumatic brain injury patients of the pediatric age population (<18).

Five studies assessed the effect of prophylaxis on early seizures, 4 on late seizures, and 1 on any seizure. The mean incidence of post traumatic seizures with AED prophylaxis was 8% for early seizures and 7.1% for late seizures. Moreover, one study revealed no benefit of AED prophylaxis for early post traumatic seizures.

Meta-Analysis revealed a significant difference in the incidence of early post traumatic seizures with antiepileptic prophylaxis. However, no significant difference for late post traumatic seizures has been shown.

**Conclusion:** In conclusion, AED prophylaxis seems to be effective against early post traumatic seizures for the pediatric population, with

Levetiracetam possibly being more effective. Also, there is no observed benefit for late post traumatic seizures.

#### PP040

### Can neuropsychology and neurosurgery advance knowledge and treatment in pediatric lesional epilepsy? A critical review of the literature about working on working together

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**Introduction:** Although pediatric lesional epilepsy is a complex and multifactorial condition, neuropsychological pre-surgical assessment and post-surgical rehabilitation as well as neurosurgical interventions can assist children as patients and their families by improving the precision of diagnosis, treatment and planning of care regarding brain pathology, comorbidities, cognition and behavior at the time of the diagnosis, but also longitudinally. This review aims to explore the fields of collaboration between these two specialties.

**Methods:** PubMed database and Google Scholar were searched using the keywords 'neuropsychology', 'neurosurgery', 'pediatric', and 'lesional epilepsy' up to October 3, 2022. Full texts of the retrieved research articles and reviews were accessed.

**Results:** There is strong evidence that the collaboration of clinical neuropsychologists and neurosurgeons focuses on diagnostics and prognostics and improves cognitive and developmental outcome of the child-patient, as well as the quality of life for both children and other family members/caregivers. This is accompanied by advancements in technology and research, but a point that is still important and remains open is to identify certain types of children patients that are overlooked for surgical management of epilepsy due to individual and social characteristics (such as ethnicity, socioeconomic status of family, race, educational level of family).

**Conclusion:** Neuropsychology makes a highly valuable contribution to the care of individual pediatric neurosurgical patients by contributing to the diagnostic process and by serving as a tool for the monitoring of disease and treatment, thereby improving the quality and safety of care. Still worldwide standardized collaboration protocols and awareness programs informing parents and caregivers about these topics and how they can facilitate the abovementioned processes are missing.

#### PP041

### Ventriculo-gallbladder shunting as an alternative treatment for complicated shunt dysfunction

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**Objective:** Diversion of the cerebro-spinal fluid into extracranial spaces is the most frequently performed operation for the treatment of hydrocephalus. The intraperitoneal space is the preferred site for distal shunt catheter placement, usually followed by pleural cavity and cardiac atrium. In cases where the use of these regions is not suitable due to reasons such as malabsorption or infection, placing the distal catheter into gallbladder may provide an alternative solution.

**Material and Method:** In the study, the outcomes of the surgery, together with the clinical and radiological follow-up, are presented in 3 cases who underwent ventriculo-gallbladder shunt surgery between 2011-2021. All the cases were operated and followed up in cooperation with the Department of Pediatric Surgery.

**Results:** Three cases with a mean age of 15(7-24) months were presented with dysfunction and migration of a previously placed ventriculoperitoneal shunt system. All cases had the shunt removed and were treated with an external ventricular drainage system initially. Ventriculo-gallbladder shunt was applied when CSF cell count was found to be within normal range and culture results were negative. For the first patient, a 24-month-old boy, an additional ventriculo-atrial shunt was also placed 1 week later, and no problem was detected at one-year follow-up. Second patient, a 14-month-old girl had also adequate drainage at one-year follow-up. Third patient, a 7-month-old girl, developed shunt dysfunction 10 years after the placement and the system was made operational again by replacing the valve without revising the distal catheter.

**Conclusion:** In cases where CSF diversion into peritoneal or pleural cavity or right atrium is not possible for any reason, placing the distal end of the shunt catheter into the gallbladder may be preferred as an alternative treatment method. Although our study included a small number of cases, it is noteworthy that none of the patients had dysfunction of the catheter placed in the gallbladder.

#### PP042

### Topical instillation of vancomycin minimizes the rate of CSF shunt infections in children

**Matthias Krause**

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**Introduction:** Shunt surgery in pediatric patients still bears a significant risk of infection, in addition to mechanical complications of the shunt system. Antibiotic-impregnated systems and perioperative antibiotics have been extensively studied for their potential to reduce shunt infections. We examined the effectiveness of intrawound application of vancomycin powder during shunt surgery.

**Patients and methods:** Patient records of 78 primary shunt implantations at a mean age of 40 months were reviewed. In total, 52 patients (mean age 50 months) had been treated according to standard surgical and perioperative procedures (Std), whereas 26 patients (mean age 20 months) additionally had received topical application of vancomycin powder before wound closure (Vmc). Overall infection rate was 3.8%, in Std patients 5.8%, and in Vmc patients 0%. The rates of CSF fistula and revision surgery were similar in both groups (5.8% vs 8% and 23.1% vs. 30%, respectively).

**Conclusion:** To the best of our knowledge, this is the first report on topical vancomycin instillation, indicating its efficacy for the prevention of shunt infection in pediatric patients. Further studies with a higher number of patients are needed to verify this finding.

#### Poster Track 2.2: Hydrocephalus

#### PP043

### Normal Sonographic Anatomy of the Ventricles in the Healthy Newborn

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**Introduction:** The ultrasound phenomenon has aided diagnostics in medicine since 1942. More advanced techniques have made its use less frequent in the neurosurgical field. Computed tomography and MRI are the most accurate in diagnosing brain pathology, but what happens when the neurosurgeon does not have access to these tools? Ultrasound is simple and accessible, its widely present in most hospitals, and can be used to accurately diagnose many neurosurgical pathologies, even more in the newborn. The purpose of this study is to create an anatomical baseline for what is the normal anatomy in the healthy newborn.

**Methods:** We performed a transfontanelar ultrasound using a standardized technique to the healthy term newborns born during a period of one month in our institution. 87 ultrasounds were performed, 81 were used for statistical analysis. We defined 9 key measurements using known landmarks in the ventricular anatomy.

**Results:** We adequately measured the anterior horn, atrium, occipital horn, temporal horn and Levene index of the third ventricle (image1) Identified ventricular variants such as cavum septum pellucidum and cavum velum interpositum. Run correlation test between all variables and age at birth and weight at birth, found weak correlation with the size of the anterior horn and the age at birth.

**Conclusion:** Our results correlated with those of a similar population and methodology (Scherle et al) found mild similarities with Levene et al, contrasting that they included preterm neonates, hence there a statistical variation that is explained by the population used. Our study was limited by the equipment that was used and was performed by one observer (first author). We successfully identified anatomical variations and was able to safely characterize the anatomy of the ventricles in the healthy newborn.

Variable	n ins.	n est. anat. vis.	Min	Max	Media	DE	Cuartil 1	Media na	Cuartil 3
Longitud AP asta anterior D (mm)	81	80	14.43	28.35	20.21	2.64	18.23	20.27	21.7
Longitud AP asta anterior I (mm)	81	79	14.20	26.64	20.05	2.43	18.39	20.34	21.56
Longitud AP cuerpo ventricular y asta occipital D (mm)	81	80	17.19	34.54	24.50	3.36	22.3	24.72	26.66
Longitud AP cuerpo ventricular y asta occipital I (mm)	81	80	16.62	31.31	24.45	2.92	22.48	24.37	26.14
Longitud LM asta anterior D (mm)	81	80	2.63	13.67	7.98	2.15	6.72	7.76	8.6
Longitud LM asta anterior I (mm)	81	80	4.34	14.51	8.01	2.07	6.77	7.64	8.6
Longitud LM asta temporal D (mm)	81	54	1.08	7.03	3.71	1.67	2.39	3.71	4.99
Longitud LM asta temporal I (mm)	81	53	1.20	10.13	4.10	2.16	2.65	3.81	5.16
Distancia tálamo-occipital (mm)	81	79	4.72	14.50	8.52	2.06	7.15	8.25	9.6
Distancia cortico-ventricular (mm)	81	80	18.87	27.70	23.51	1.88	22.18	23.51	24.9
Distancia núcleo caudado a cuerpo calloso D (mm)	67	61	0.76	2.96	1.88	0.42	1.67	1.78	2.21
Distancia núcleo caudado a cuerpo calloso I (mm)	67	61	0.96	3.47	2.02	0.47	1.72	1.89	2.38
Tamaño plexos coroideos (mm)	81	76	15.45	29.98	22.67	4.09	19.1	22.71	26.46
LLL tercer ventrículo (mm)	81	45	1.01	3.13	1.79	0.60	1.32	1.69	2.25
Longitud CC asta temporal D (mm)	22	17	0.50	6.91	2.31	1.45	1.36	2.09	3
Longitud CC asta temporal I (mm)	22	17	0.51	6.91	2.29	1.55	1.2	1.6	3.13

**PP044**  
**Elements of neuroendoscopic anatomy of the fourth ventricle in young patients-cadaveric studies**  
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**Introduction:** Neuroendoscopic anatomy of the fourth ventricle has been reported infrequently in the literature, most of the times based on laboratory experiences. Neuroendoscopy offers a quite different outlook of the anatomy of the fourth ventricle and compared with the microsurgical descriptions it seems to provide a superior and detailed visualization of the superior part.

**Aim:** Regarding the upcoming neuroendoscopic techniques the fourth ventricle was examined.

**Material-Methods-**In a series of 10 anatomically cadaveric specimens (<20 years) the fourth ventricle was investigated endoscopically. We are

able to reach the fourth ventricle via the aqueductus cerebri and coming via the cerebromedullary cistern through the foramen of Magendie.

**Results-Conclusions:** Using principally rigid and sometimes flexible endoscopes the basic anatomic details seen under the neuroendoscope and the topographical landmarks of the approaches were investigated presented and discussed. It seems that neuroendoscopy is a unique and very useful tool that it has a place in the armamentum of the modern neurosurgeon, its application will only broaden as we gain experience in using neuroendoscopic systems. Neurosurgery may have been slow to accept the use of modern neuroendoscopic instruments in the management of neurological diseases but with advent of new scopes, illumination and instruments it is a fast-growing arena for minimally invasive neurosurgery.

**PP045**  
**Treatment of intracranial arachnoid cysts in children during 10 year period**  
**Bojana Zivkovic,** Nikola Jovicevic, Vladimir Bascarevic, Jelena Kostic, Aleksandar Jančićević  
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**Objective:** In this paper we compare different treatment modalities – microsurgical/endoscopic fenestration of intracranial arachnoid cyst vs. shunting.

**Material and Methods:** This is partly retrospective, partly prospective cohort study that included all patients under 18 years of age, that have been diagnosed with an intracranial arachnoid cyst and treated surgically in our institution during a period of last ten years.

**Results:** The data about 44 patients were collected. The most common clinical symptoms were headache (52.2%) and vomiting (54.5%). There was an apparent neurological deficit in nearly 39% of patients, among which the most frequent was cranial nerve palsy (27.2%).

The most used surgical technique was endoscopic fenestration (38.6%), followed by shunting (31.8%). Significant improvement was found in all the cases, however the majority of patients without any symptoms after surgery were treated endoscopically. The recovery was noted in all patients with cranial nerve palsy, except for the paresis of the abducens nerve in one case. Hemiparesis improved after treatment in all cases, however slight hemiparesis remained in 3/44 cases. Complications occurred in 40.9% of operated patients, with almost half of these related to shunt dysfunction. Other complications occurred rarely - hygromas (2/44), intracystic hemorrhage (1/44), wound infection (1/44), and one conversion to shunt.

Comparison was made between patients treated with fenestration vs. drainage procedures, there were no statistically significant difference regarding majority of postoperative symptoms, however, there was statistically significant advantage of fenestration regarding postoperative headache, vertigo, and especially complication rate.

**Conclusion:** This study includes a period of treatment of intracranial arachnoid cysts before the shift in thinking of majority of neurosurgeons, when shunting was used in the majority of cases. This resulted in the quite high rate of complications in our series. Fenestration of arachnoid cysts is an effective treatment and less burdened with complications compared to shunting.

**PP046**  
**Early neuroendoscopic washout for the treatment of Post-Hemorrhagic IVH causing Hydrocephalus in premature neonates**  
**Christos Chamilos,** Panagiotis Kokkalis, Andreas Mitsios, Spyros Sgouros  
*IASO Childrens' Hospital, Marousi, Greece*

**Introduction:** In recent years early endoscopic lavage is gaining momentum in the treatment of neonatal intraventricular haemorrhage

(IVH) of prematurity, expecting that it will reduce the rate for permanent shunting. We analyze the results of our first five cases.

**Methods:** Between September 2017 and September 2021, 5 premature neonates with post-hemorrhagic hydrocephalus were treated with neuroendoscopic lavage for removal of intraventricular blood clot remnants. The range of age at the operation was between 10 and 42 days. One patient had both ventricles cleared from the right side through a septostomy, two patients had a second operation on the other side to clear the contralateral ventricle and the other two had only unilateral intraventricular clot. All patients had insertion of ventricular access device and required postoperative ventricular tapping.

**Results:** All patients tolerated all procedures very well with no systemic or procedural complications. Two of the five patients did not require a shunt insertion at latest follow up (18 and 42 months respectively) and have normal size ventricular system without localizations. One patient developed a postop infection due to CSF leak from the wound which led eventually to shunting and 4th ventricle loculation. The other two shunted patients have not developed loculated ventricles (latest follow up 33 months) but required shunt revision.

**Conclusion:** Early neuroendoscopic lavage of intraventricular blood degradation products and residual hematoma is a valid alternative for the treatment of PHH in neonates with IVH. Early results are promising regarding shunt avoidance.

#### PP047

##### Management of post-haemorrhagic hydrocephalus in neonates

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**Introduction:** Intracranial haemorrhage occur in all gestational ages newborns. In the literature, the intracranial haemorrhage is estimated to 15% of all premature births, under 32 weeks. The association of hydrocephaly has frequency of 24%, being directly related to the severity of haemorrhage. The definitive treatment of post-haemorrhagic hydrocephalus remains ventriculo-peritoneal shunt, but in many cases may be necessary, the temporary procedures, until the normalization of cerebrospinal fluid. The management of posthaemorrhagic hydrocephalus, in newborns, includes numerous temporary options: external ventricle drainage, reservoirs, ventriculo-subgaleal shunt or lumbar punctures.

**Methods:** This paper is a retrospective study that includes 12 patients, admitted in the department of Neurosurgery of the MedLife Polissano Hospital, between January 2018 - December 2022.

The management of these patients consisted in the insertion of the external drainage for 1 month, then Ommaya reservoir, that was punctured and drained periodically and, finally, ventriculo-peritoneal shunt.

**Results:** Our Study included 12 patients, for whom the management consisted of external ventricular drainage, Ommaya reservoir and ventriculo-peritoneal shunt. The surgeries were uneventful in all the cases. The Ommaya reservoirs were maintained for 4 to 12 weeks, depending on the severity of the bleeding. The reservoirs were tapped variably, depending on the clinical progress, ranging from 1 to 3 taps/week. The volume of CSF drained per tap varied between 20 and 120 ml.

**Conclusions:** In our experience, the management of the posthaemorrhagic hydrocephalus consists of external ventricular drainage as the first step, which can bring the benefits of a faster bleeding evacuation and the optimal control of the intracranial pressure and head circumference. The Ommaya reservoir implantation, as a second step of the management of posthaemorrhagic hydrocephalus, is safe and extremely useful and may be beneficial for improving their neurodevelopmental outcomes. More studies may be needed to continue to validate the results of this report.

#### PP048

##### Valiation of the ETVSS - A single centre experience

**Peter Spazzapan**, Tomaž Velnar

University Medical Centre Ljubljana, Ljubljana, Slovenia

**Introduction:** Endoscopic third ventriculostomy (ETV) is a wellknown and effective treatment for selected patients with hydrocephalus.

**Methods:** The aim of our study was to retrospectively analyze all pediatric patients treated at our Institution from 2016 to 2021 with ETV. We compared our data with previously reported studies, in particular with the ETV scoring system (ETVSS).

**Results:** Our results confirm age as the most important single predictive factor for ETV success. The estimates for success were 33.3% for patients younger than 6 months and 94.4% for patients older than 6 months. The overall success rate in our series was 71%, which is similar to previously reported results.

**Conclusions:** A good patient selection remains of paramount importance for ETV success.

#### PP049

##### Complications of hydrocephalus treatment in pediatric patients with Miethke M.blue plus® Valve

**Nina Junold-Podlogar**

Helios Klinik Krefeld, Krefeld, Germany

In this abstract/presentation we want to point out the difficulties and complications we experienced in hydrocephalus treatment in pediatric patients with the Miethke M.blue plus® valve. From 2019 until 2022 we implanted the Miethke M blue plus valve in children and also adults. Decisive was the idea, that the valve allows an adjustment of opening pressure in upright position of the patient which is the most dynamic position with the highest risk of over drainage. 46 pediatric patients received this valve. The main diagnosis was posthemorrhagic hydrocephalus in the context of prematurity. Other diagnoses were posthemorrhagic hydrocephalus on the basis of trauma, congenital hydrocephalus, myelomeningocele, hydrocephalus malresorptivus due to an oncological disease and idiopathic intracranial hypertension. 15 patients had to be revised by reason of oftentimes dysfunction of the valve. This could be problems to adjust the valve to the point of impossibility to adjust the valve or occlusion of the valve.

Occlusion of the valve mostly occurred in children with posthemorrhagic hydrocephalus and prematurity which in this regard could depend on the higher level of protein in the cerebrospinal fluid.

Because of this we tried to figure out the best point in time with the optimum lab constellation when to implant the valve in children with posthemorrhagic hydrocephalus to prevent complications.

#### PP050

##### The role of ProGav adjustable valves and related adjustable anti-siphon devices in reducing the number of surgeries in children

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<sup>1</sup>Department of Pediatric Neurosurgery, Fondazione Policlinico Universitario A. Gemelli IRCCS, Università Cattolica del Sacro Cuore, Rome, Italy, Rome, Italy, <sup>2</sup>Department of Neuroscience, Neurosurgery Section, Università Cattolica del Sacro Cuore, Rome, Italy, Rome, Italy

**Introduction:** Slit ventricle syndrome (SVS) is a iatrogenic disease occurring in patients with ventriculoperitoneal shunt secondary to over-drainage. There are few evidence in literature showing the long-term efficacy of gravitational valves or anti-siphon devices in addressing this issue. However, literature is inconclusive and poorly systematic

on the topic. Aim of the present, single center, retrospective study is to evaluate whether and how the use of gravitational shunt valves could reduce over-drainage complications.

**Methods:** We reviewed a series of 47 patients who underwent to ventriculoperitoneal shunt implantation with antisiphon devices (MIETHKE ProGAV2.0 or MIETHKE M.blue) between March 2017 and December 2022 at our institution. We examined initial symptoms, radiological findings in preoperative, postoperative and long-term follow-up MRI and CT scans and the recurrence of symptoms requiring adjustments of the setting pressure or surgical revisions in case of malfunction.

**Results:** Eighteen patients (38%) already harbored a programmable or non-programmable fixed differential pressure valve and underwent surgical treatment to implant an antisiphon device after developing SVS. Overdrainage symptoms were managed without surgery in 15 (83%) of them; two patients needed to change the antisiphon device. Twenty-nine patients (62%) with hydrocephalus underwent primary ventriculoperitoneal shunt insertion. Five of them adjusted the valve without surgery. One patient became shunt-free after positioning a ProGav2.0. Shunt malfunction occurred in 8 cases of the whole series (17%).

**Conclusions:** Our results demonstrate antisiphon devices are effective in treating overdrainage symptoms in children with SVS and in preventing them in case of primary shunt insertion. Though there was no significant difference in the overall rate of shunt malfunction compared with other kinds of shunt systems, the possibility to adjust externally the valve allowed to avoid additional surgical procedures in 78% of the patients.

**PP051**

**Long-tunneled versus short-tunneled external ventricular drain: a quasi-experimental study in a cohort of pediatric patients**

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<sup>1</sup>Hospital de Pediatría Juan P. Garrahan, Buenos Aires, Argentina

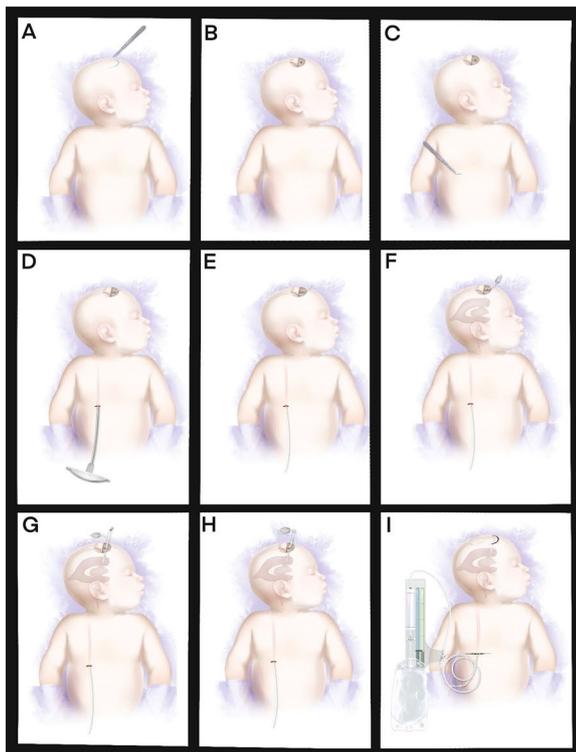
**Objective:** The primary aim of this study was to compare external ventricular drain (EVD)-related infection rates and mechanical complications between long-tunneled EVD (LTEVD) and short-tunneled EVD (STEVD) in a cohort of pediatric patients.

The second objective was to compare hospital resources used between LTEVD and STEVD in the same cohort of patients and study period.

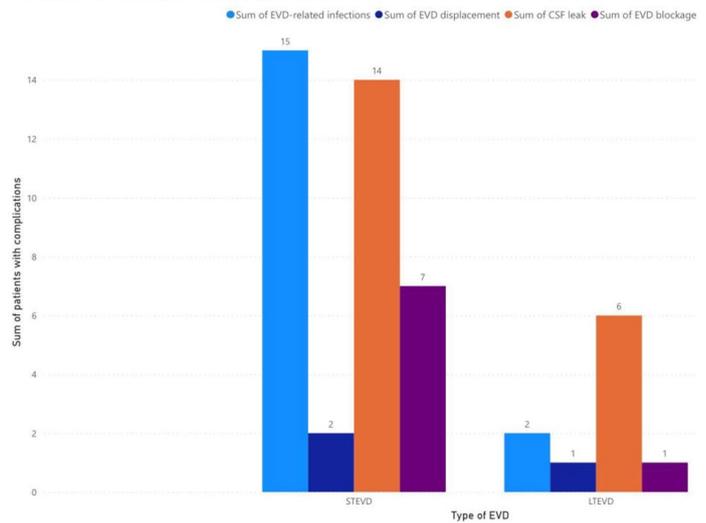
**Methods:** The study consisted of a quasi-experimental study comparing a prospective group of patients with LTEVD with a retrospective (historic) cohort of STEVD. The prospective non-randomized quasi-experimental protocol of LTEVD included patients that needed an EVD for more than three days. Data were recorded prospectively as the patients were added to the study until reaching the sample size established by the protocol. The comparison group of the STEVD cohort was retrospectively collected from patients' records. We included patients consecutively, from newest to oldest, starting with the last STEVD inserted at our hospital until reaching the sample size established in the protocol. Inclusion and exclusion criteria for both groups were the same. STEVD allocated more resources than LTEVD in most areas considered in this study.

**Results:** One-hundred thirty-four patients were included in this quasi-experimental study, sixty-seven in each group. LTEVDs reduced the odds of having an EVD-related infection by 92% (OR 0.08; 95% CI 0.01-0.39; p=0.002). LTEVDs reduced by 69% the odds of having a CSF leak compared to STEVD (OD 0.31; 95% CI 0.10-0.91; p=0.03). Neither CSF blockage (p=0.06) nor displacement (p=0.69) showed a statistically significant difference between groups. STEVD allocated more resources than LTEVD in most areas.

**Conclusions:** LTEVD, compared to STEVD, is a cost-effective and safe method to reduce EVD-related infection rates and other complications in pediatric patients. We believe that reducing the infection rate and complications and giving the patient more independence outweighs the additional costs that this new technique may yield.



Total number of complications by type of EVD



**PP052****Occipital lobe infarction in shunt malfunction****Peter Spazzapan***University Medical Centre Ljubljana, Ljubljana, Slovenia*

**Introduction:** Hypoxic ischemic encephalopathy is a rare and dramatic complication that can occur in cases of VP shunt malfunction. Occipital lobe infarction is a result of tentorial herniation and compression of the posterior cerebral arteries in the tentorial incisura. This complication can cause minor visual field defects, blindness or even severe impairments of the level of consciousness.

**Methods:** All pediatric cases of shunt malfunction in the period between 2013 and 2022 that have resulted in an ischemic event in the occipital lobes have been included in this series. An attempt has been made to identify the risk factors of this complication.

**Results:** Four children experienced this dramatic complication after an episode of VP shunt malfunction. The age of these patients was variable between 2 and 14. Three of them had a low pressure valve. Two children developed a visual field deficit, two other children developed a severe impairment of the level of consciousness.

**Conclusions:** Prompt and early shunt revision is of paramount importance to avoid these important complications. Dedicated pediatric neurosurgeons are indeed more used to recognize these situations and act correctly. In our opinion a low pressure valve represents a potential risk factor for this complication, since the rise of intracranial pressure in case of VP shunt malfunction can be fast.

**PP053****Is early shunt implantation a risk factor for shunt infection in preterm infants with posthemorrhagic hydrocephalus?****Aygül Wurzer**, Luisa Früh, Andrea Reinprecht*Medical University of Vienna, Vienna, Austria*

**Introduction:** Intraventricular hemorrhage (IVH) in preterm infants often leads to posthemorrhagic hydrocephalus (PHH) requiring neurosurgical intervention. The common treatment options include external ventricular drainage (EVD), ventricular reservoir (VR), and peritoneal or atrial shunting. However, early shunt failure and postoperative shunt infections are frequent complications. The aim of the study was to investigate the risk factors for postoperative shunt infection in preterm infants with PHH.

**Methods:** This retrospective study included all preterm infants born between January 2013 and August 2019 diagnosed with IVH who needed neurosurgical interventions. The patients were treated on the neonatal intensive care unit of the Medical University of Vienna in collaboration with the neurosurgical department. According to a standardized treatment protocol, first an EVD was placed in case of PHH. Shunt implantation was performed if the following criteria were met: (1) patients' weight >2000g, (2) cerebrospinal fluid (CSF) total protein <200mg/dl, and (3) CSF cell count <100. Otherwise, further EVDs were placed or, rarely, a VR was implanted.

**Results:** Fifty preterm infants with IVH were identified. The mean gestational age was 28.23 weeks and the mean gestational weight 1204.76g. The IVH was classified as grade II in 5 (10%), grade III in 29 (58%) and as grade IV in 16 (32%) patients. At least one EVD was placed in 45 (90%) patients and a VR in 9 (18%). A total of 38 (76%) patients received a ventriculoperitoneal (VP) and 5 (10%) a ventriculoatrial (VA) shunt. Postoperative infections occurred in 5 patients with VP shunt. The mean age of these patients was 76.6 days compared to 146.3 days in patients without shunt infection.

**Conclusion:** According to our study, the risk of shunt infections in preterm infants with PHH might be reduced if not only higher weight but also higher age is considered for timing of shunt implantation.

**PP054****A comparison of survival and overdrainage rate between pressure and flow regulated ventriculoperitoneal shunts for the management of hydrocephalus in the first year of life****Bilal Khaleel, Mohsen Yaish**, Stuart Stokes, Hesham Zaki, John McMullan, Shungu Ushewokunze, Patricia De Lacy, Veejay Bagga  
*Sheffield children's NHS trust., Sheffield, United Kingdom*

**Introduction:** Ventriculoperitoneal shunts are the mainstay of treatment of hydrocephalus in the first year of life; shunt valves can be pressure or flow regulated, and there is limited evidence suggesting which system is more appropriate in this population. A 2020 study at Sheffield Children's Hospital suggested that flow regulated OSV II valves were less likely than pressure regulated ProGAV 2 valves to cause overdrainage in neonates. We sought to compare the shunt survival and overdrainage rate between OSV II and ProGAV 2 valves inserted at any point in the first year of life.

**Methods:** A retrospective case series of primary ventriculoperitoneal shunt insertions in the first year of life at Sheffield Children's Hospital was carried out. 20 patients were fitted with ProGAV 2 valves, and 41 patients with OSV II valves. Overdrainage rate was assessed through head circumference measurements.

**Results:** The mean 6 month shunt survival was 45% for the ProGAV 2 valves and 71% for the OSV II valves ( $p=0.089$ ). Mean shunt survival at 1 year was 30% and 54% for ProGAV 2 and OSV II valves respectively ( $p=0.134$ ). The overdrainage rate at 1 year was 33% for patients with the ProGAV 2 valves and 36% for patients with the OSV II valves, with no significant difference between the valve types.

**Conclusion:** We found no significant difference in shunt survival or overdrainage rate between pressure regulated and flow regulated valve systems in the first year of life.

**Poster Track 2.3:Oncology****PP055****Rurality index score and pediatric neuro-oncological outcome in Ontario, Canada****Michelle Kameda-Smith**<sup>1</sup>, Gregory Pond<sup>2</sup>, Hsien Seow<sup>2</sup><sup>1</sup>*Great Ormond Street Hospital for Sick Children, London, United Kingdom*, <sup>2</sup>*McMaster University, Hamilton Health Sciences, Hamilton, Canada*

**Introduction:** Rapid access to neurosurgical decisions and definitive management are vital for the outcome of neurocritical patients. There are unique challenges associated with the provision of services required to maintain critical infrastructure for rural citizens. As a relationship between rurality, marginalization and health outcomes has been identified as associated with higher mortality rates and higher rates of many diseases we studied if poorer clinical outcome were associated with rurality in pediatric neuro-oncological disease.

**Methods:** Employing linked administrative databases, we retrospectively analyzed a population-based cohort of patients diagnosed with a pediatric brain tumor between 1996 to 2017 in Ontario, Canada. Our main variable of interest was Rurality Index for Ontario (RIO, larger denotes more rural); our main outcome was survival whilst controlling for surgery and tumor type. Results: Of the 1457 patients included 54.0% were male). Overall survival of all the children controlling for surgery and tumor type at 1, 5 and 10- years was 84.7%, 65.1% and 58.4% respectively. 11.5% classified as living in a rural area of Ontario. The distance to the nearest pediatric neurosurgical hospital ranged from 25.6 to 167.4 km. The RIO was 0 in 38.8% of children with the majority of patients with a RIO score of <39. Higher RIO score was not a significant factor (Continuous  $p=0.12$ /Ordinal  $p=0.18$ ) associated with

length of follow up, indicating rurality was not significantly linked with compliance to clinical follow-up.

**Conclusion:** Rurality of the region pediatric neuro-oncological patients reside were not associated with patient outcome (HR 0.83,  $p=0.39$ ).

#### PP056

##### Primary intracranial sarcoma in children. A case presentation of a rare entity

**Savvas Melissaris**, Georgios Markogiannakis, Rafailia-Nina Karela, Spyridoula Akakiou, Dimitrios Panagopoulos  
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**Introduction:** Primary Intracranial Sarcoma is an uncommon disease, with estimated incidence less than 0.2 % of primary brain tumors, the majority of which corresponds to the pediatric population (Lafay-Cousin, 2016). Despite limited knowledge about best treatment approaches, surgical resection of the tumor remains essential for the management of such an aggressive entity.

**Methods:** We describe the case of a 6-year-old boy who presented in the ER with impaired level of consciousness, multiple episodes of vomiting during the past 24 hours, a progressive deterioration of headache over one week and anisocoria (the left pupil was larger than the right). Also, the patient has been complaining for mild headaches for 6 months. Café-au-lait macules were identified as birthmarks, but no further investigation was performed. The CT scan revealed an intraparenchymal hemorrhage in the left fronto-parietal area of the brain, a lesion inside the hemorrhage and a >1cm midline shift. Immediately, the patient was intubated and taken to the OR. Using the microscope, a gross total resection of the tumor with the adjacent, infiltrated dura was achieved.

**Results:** The histological investigation revealed a malignant pleomorphic, spindle cell neoplasm with compact fascicular pattern involving the co-existed dura. The diagnosis of a Primary Intracranial Sarcoma with DICER-1 mutation was established and the patient underwent adjuvant chemotherapy and radiotherapy. The post-surgical MRI showed high intensity white matter regions, described as hamartomatous lesions compatible with NF 1.

**Conclusion:** This case illustrates a rare CNS sarcoma, presented in a boy with findings reminiscent of NF 1. Although NF 1 has been linked with this malignancy in literature, only a small number of patients develop the disease. Further research of similar cases could improve the therapeutic planning and reveal significant pathophysiological pathways.

#### PP057

##### Epidemiology, Investigation, Treatment and Outcome of Pediatric Cushing's Disease: A Case-series

**Santanu Bora**, Asish Suri  
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**Introduction:** Paediatric Cushing disease is a rare entity with its unique sets of challenges in diagnosis and management. This study aims to share our experience of 24 pediatric patients with Cushing disease treated surgically with special emphasis on outcome and remission.

**Methods:** Pediatric patients of Cushing disease treated from 2009 to 2022 at a tertiary research and teaching centre were retrospectively reviewed. Medical records, imaging and details of surgery were reviewed to extract the relevant information.

**Results:** 24 consecutive patients of pediatric Cushing disease treated were included. Mean age at presentation was 14.91 years (range 9-18 years) with a male: female of 1:1.4. Majority of these patients (n=19, 79.16%) had microadenoma, two had macroadenoma (8.3%) and three (n=3, 12.5%) did not have tumour visualised on the MRI. The mean

preoperative cortisol levels were 39.2  $\mu\text{g}/\text{dl}$  (range 12-212). Mean preoperative ACTH levels were 62.93  $\text{pgm}/\text{dl}$  (29-126). Endoscopic TNS (Trans nasal trans sphenoidal) was done in 13, microscopic TNS in 7 and sublabial trans sphenoidal (SLTS) was done in 4 cases. Average blood loss was 196 ml. Intraoperative CSF leak was present in 6 cases (25%). 6 patients lost to followup. Out of 18 patients, remission was achieved in 12 patients (66%). Remission in endoscopic surgery is 75% while in microscopic surgery remission is 50%. Out of six cases with persistence redo pituitary surgery was done in 4 cases with remission achieved in two (50%). One of the patients with persistence after redo surgery was given gamma knife with successful remission.

**Conclusion:** Transsphenoidal surgery remains the treatment of choice for pediatric Cushing disease. It has promising results with low complications. Early diagnosis and treatment are important to reduce morbidity and mortality. Day 1 cortisol levels correlate with the remission and high initial cortisol levels reduce the remission rate.

#### PP058

##### Papillary tumor of the pineal region in pediatric patients: A rare entity

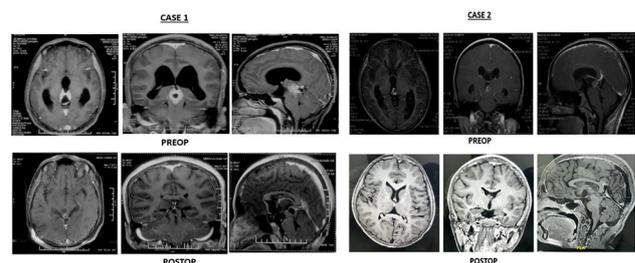
**Santanu Bora**, Amandeep Kumar  
All India Institute of Medical Sciences, New Delhi, New Delhi, India

**Introduction:** Papillary tumors of the pineal region (PTPR)s are rare neuroepithelial tumors of the pineal region. It was described first in 2003 by Jouvett et al and was included in 2007 WHO classification of central nervous system tumors. Only thirty cases of pediatric PTPR have been reported till date. This presentation aims to share our experience of two pediatric cases with PTPR.

**Method:** Pediatric patients with PTPR treated from 2016 to 2022 at a tertiary research and teaching center were reviewed. Medical records, imaging and details of surgery were reviewed to extract the relevant information.

**Results:** 2 patients, with male: female ratio of 1:1, operated during this period for pineal region tumors were found to have PTPR on histopathology and immunohistochemistry. Average age at presentation was 13.5 years. Headache and visual deterioration were the commonest presentation while upgaze paresis and incontinence were also present in the female patient. Average duration of symptoms before presentation was 5 months. In preoperative imaging hydrocephalus was present in both the cases but no CSF diversion was done prior to definitive surgery. Average volume of tumor was 3.5 ml. Krause approach was used in sitting position and near total excision was achieved in both cases. Hydrocephalus persisted in both cases after excision, so treated with endoscopic third ventriculostomy in one and ventriculoperitoneal shunt in the other. None has received adjuvant chemoradiotherapy. Both patients are doing fine without recurrence at 6 and 1 year follow up.

**Conclusion:** PTPRs are rare tumors of the brain. A definitive diagnosis of PTPR needs characteristic histological and immunohistochemical features as clinical and radiological presentation is highly nonspecific. Surgery is the mainstay of treatment. Adequate surgical resection may prevent recurrence. Treatment of associated hydrocephalus is equally important.



**PP059****Long survivor ETANTR: a singular case of a rare childhood brain tumor with an excellent outcome**

**Sergio Capelli**, Gianluca Agresta, Bianca Bossi, Davide Locatelli  
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**Introduction:** Embryonal tumors with multilayer rosettes (ETMR) is a histologically heterogeneous entity that includes variants identified in the previous WHO classification, such as embryonal tumors with abundant neuropil and true rosettes (ETANTR).

Because this is one of the childhood CNS tumors with the worst prognosis, developing an optimal treatment strategy is crucial.

**Methods:** We present a case of ETANTR in a two-year-old child, localized at the right parietal periventricular level, who underwent microscopic removal surgery and subsequent multimodal adjuvant therapy at the Circolo Varese Hospital (IT). The prognostic outcome was then placed within the context of the cases presented in the recent literature.

**Results:** Following a postoperative brain MRI that revealed a GTR, the patient underwent cycles of high-dose sequential chemotherapy (HDC), soon afterward by haematopoietic stem cell rescue and radiotherapy, and he showed no signs of disease recurrence after an 8-year follow-up. According to the recent literature, this finding falls outside of the known disease-free survival in ETANTR cases undergoing adjuvant HDC treatment, radiotherapy, or tandem HDC - autologous stem cell transplantation.

**Conclusion:** ETANTR is a childhood brain tumor with a poor prognosis and no known treatment regimen.

To our knowledge, this is the first case of long-term follow-up without recurrences of this kind of tumor, by combining all the adjuvant treatment without side effects. Thus highlights the importance of multimodal treatment in this type of pathology in which long term survival is possible.

**PP060****Periventricular and periependymal rosary-like pattern leading to diagnosis of neurosarcoidosis**

Patricia Alejandra Garrido<sup>1,2</sup>, **José Hinojosa Mena-Bernal**<sup>2</sup>, M<sup>a</sup> Victoria Becerra<sup>2</sup>, Santiago Candela<sup>2</sup>, Diego Culebras<sup>2</sup>, Mariana Alamar<sup>2</sup>, Daniel Pascual<sup>1</sup>, Laura Ruiz<sup>1</sup>, Andoni García<sup>1</sup>

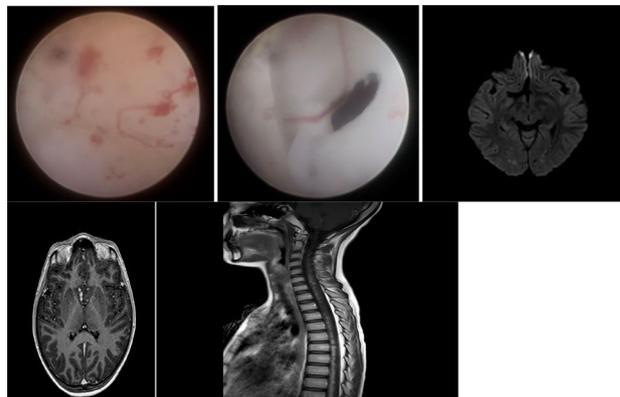
<sup>1</sup>Complejo Asistencial Universitario De Salamanca, Salamanca, Spain, <sup>2</sup>Hospital Sant Joan de Déu, Barcelona, Spain

**Introduction:** We present a case of a 12-year-old patient suffering from progressive spastic paraparesis secondary to multiple contrast-enhancing ependymal spinal cord lesions. The patient has been presenting varied symptoms: patellar hyperreflexia, urinary and fecal incontinence without sensory disorder, motor deficit in the right lower limb, posterior uveitis with retinal vasculitis, abdominal pain and vomiting of four years of evolution, arthralgia and deformity in one finger of the hand, hypogammaglobulinemia and mild hereditary thrombocytopenia. MRI revealed extensive vascular enhancement, beaded-shaped, at the medullary and cerebral periependymal level, bilateral subcortical, cerebellar peduncles, and right cerebellar hemisphere; and punctate enhancement at the medullary level with extensive medullary myelopathy involving the conus medullaris. All complementary studies to date had been negative.

**Methods:** The differential diagnosis was between an inflammatory disease or a tumor (ganglioneuroma as a feasible possibility), with infectious suspicion being the least likely. It was decided to perform a neuronavigation-guided biopsy and endoscopy of ventricular lesions in the left frontal horn with septostomy and biopsy in the contralateral caudate in which the uptake area can be seen.

**Results:** Neurosarcoidosis was the anatomopathological finding. Corticosteroids and immunosuppressants were prescribed and the patient's condition improved.

**Conclusions:** Neurosarcoidosis is a rare complication of sarcoidosis. Patients undergo several tests until the diagnosis is reached. When periependymal beaded-shaped lesions are observed in the MRI and a broad spectrum of symptoms appears in addition to neurological focality, it should be suspected. Some cases disappear spontaneously within 4 to 6 months, other cases continue intermittently. Prognosis is acceptably favorable with treatment, so early diagnosis is important. The treatment of choice should be individualized but a high dose of steroids and suppression of the immune system with chemotherapy, and immunotherapy drugs, such as infliximab, are usually included. Treatment is necessary for at least 5 years.

**PP061****A rare presentation of an intraventricular pilocytic astrocytoma in a 14-month-old child**

**Hajira Iftikhar**<sup>1</sup>, T Kapadia<sup>2</sup>, J Manfield<sup>2</sup>, Ian Kamaly-Asl<sup>2</sup>, D Vartholitis<sup>2</sup>

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**Introduction:** Intraventricular tumours of the cerebral hemispheres account for less than 1% of all intracranial tumours. This illustrative case report describes a rare case of an intraventricular pilocytic astrocytoma in a 14-month-old child who presented to the emergency department with a 4-week history of irritability, increased agitation, and worsening lethargy.

**Methods:** Illustrative case report highlighting unusual imaging features.

**Results:** Examination revealed hypertonicity in the right upper arm with flexion at the elbow. A CT head demonstrated moderate dilatation of both lateral and 3rd ventricles with a large mass lesion isodense to the cortex (no calcification) in the left lateral ventricle adhered to septum pellucidum. An MRI head subsequently demonstrated a large well-demarcated and highly vascularised intraventricular mass. These images were reported by neuroradiologists as most likely an atypical teratoid rhabdoid tumour (ATRT) with a choroid plexus tumour being a possible differential. Morphological, immunohistochemical, and molecular properties were in keeping with a pilocytic astrocytoma (WHO grade I).

Pilocytic astrocytomas account for most central nervous system tumours in children and often confer an excellent prognosis. Intraventricular tumours of the cerebral hemispheres account for less than 1% of all intracranial tumours (Louis et al., 2016). In this patient, this diagnosis was not considered radiologically at presentation but was confirmed on histology. The tumour was highly vascular which complicated surgery and the child

developed a cardiac arrest intraoperatively due to hypotension. Despite achieving gross total resection, the tumour progressed rapidly.

**Conclusion:** Intraventricular juvenile pilocytic astrocytomas (IV JPAs) are incredibly rare in toddlers and should be considered as a differential in patients presenting with an intraventricular mass. They should also be assumed to be extremely vascular during surgical planning. Haemorrhage appears to be a recurring theme amongst reported IV JPAs, with those with calcification bleeding the least (Pasricha et al., 2018).

#### PP062

##### No impact of perioperative dexamethasone treatment on tumor growth rates after incomplete resection in pediatric low-grade gliomas

**Martin U. Schuhmann**, David Gorodezki, Julian Zipfel, Manon Queudville, Ursula Holzer, Andrea Bevot, Jens Schittenhelm, Thomas Nägele, Martin Ebinger

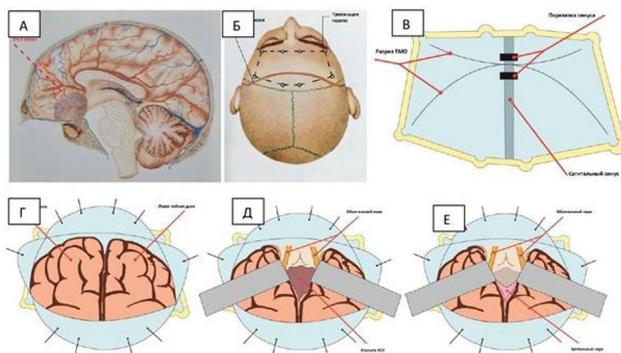
University Hospital Of Tuebingen, Tübingen, Germany

**Introduction:** Corticosteroids have proven to be beneficial in the management of acute symptoms related to CNS tumors, and have been widely used for many decades, with dexamethasone (DM) being used most commonly. However, in-vitro data suggested that DM might induced suppression of oncogene-induced senescence in a preclinical pediatric low-grade glioma (pLGG) model. This raised questions whether perioperative DM treatment in pLGG patients might have negative effects on the long-term postoperative course in case of incomplete resection.

**Methods:** 172 patients with pLGG were retrospectively analyzed concerning the impact of perioperative DM application on postoperative short- and long-term tumor growth velocity and PFS. Three-dimensional volumetric analyses of sequential MRI follow-up examinations were used for assessment of tumor growth velocity (TGV).

**Results:** Mean follow-up period accounted for 60.1 months. 65 patients (45 %) were perioperatively treated with DM. 5-year-PFS was 93% following gross-total resection (GTR) and 57% post incomplete resection (IR). Comparison of short- and long-term postoperative TGV in patients with vs without perioperative DM application showed no significant differences (short-term: 0.022 vs 0.023 cm3/mo, respectively; long-term: 0.019 vs 0.023 cm3/mo, respectively). PFS following IR (5-year-PFS: 65 % vs 55 %, respectively; 10-year-PFS: 52 % vs 53 %, respectively) and following GTR (5- and 10-years-PFS: 91 % vs 92 %, respectively) showed no differences.

**Conclusion:** The often-used perioperative DM application in case of pLGG seems to be safe without any impact on postoperative tumor behavior and TGV in case of incomplete resection.



#### PP063

##### Disseminated PEComa masquerading as NF-2

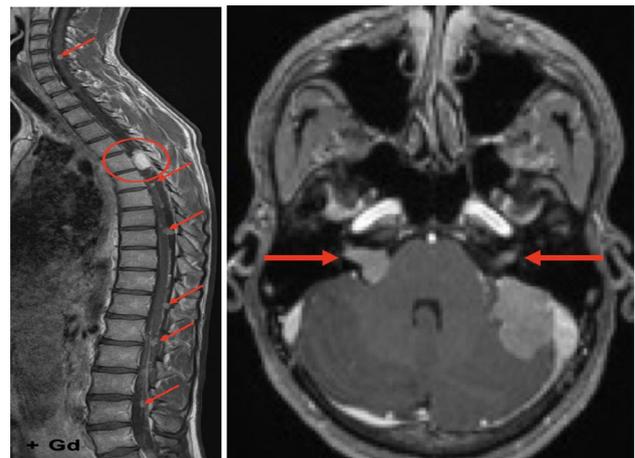
**Frank Van Calenberg**<sup>1</sup>, Rik Demaerel<sup>1</sup>, Pegah Masrori<sup>2</sup>, Laurens Dobbels<sup>2</sup>, Raf Sciote<sup>1</sup>, Patrick Schöffski<sup>1</sup>,

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**Introduction:** PEComas are rare mesenchymal-related tumors that share a distinctive cell type, the perivascular epithelioid cell or "PEC". The tumor is histologically (with epithelioid as well as spindle cells), immunohistochemically (melanocytic and smooth muscle markers) and genetically (TFE3 gene rearrangements and TSC2 mutations) very diverse. These tumors are usually described in abdominal locations. We present a patient with disseminated PEComa of the neuraxis, with cranial and spinal nerve lesions and meningeal lesions, mimicking NF-2.

**Case description:** A 17-year old boy presented with thoracic pain during sport activities and ascending, progressive thermoanalgesia with a sensory level of T5. Imaging showed multiple lesions: meningeal lesions supra- and infratentorial, lesions on both vestibulocochlear nerves, a large symptomatic intradural, extramedullary tumor at T5 and multiple other spinal tumors. Neurofibromatosis type II, Constitutional Mismatch Repair Deficiency and droplet metastases of melanotic PEComa were considered. He underwent a successful resection of the lesion at the level of T5. Intraoperatively the lesion was remarkably pigmented and vascularized. Histology showed atypical, clear cell epithelioid cells. Immunohistochemical staining was positive for melanine and HMB45. Methylation array was inconclusive and 'next generation genetic sequencing' demonstrated a NONO-TEF3 fusion, confirming the diagnosis of melanotic PEComa. New imaging after 6 weeks showed progression of all tumors and systemic chemotherapy with VIA/VIP was initiated.

**Conclusion:** We present a case of a very rare tumor, PEComa, which has never been described before in an craniospinal location. The tumor presented with neurological symptoms and was disseminated at diagnosis. The aggressive evolution is remarkable.



#### PP064

##### Hemostasis in giant intraventricular meningioma in child: use of saline-cooled radiofrequency coagulation

**Francesca Vitulli**<sup>1</sup>, Pietro Spennato<sup>1</sup>, Giuseppe Mirone<sup>1</sup>, Nicola Onorini<sup>1</sup>, Giuseppe Cinalli<sup>1</sup>

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**Introduction:** Meningiomas are relatively rare in children, and are more likely to be intraventricular and cystic, with malignant behavior and multiplicity. Gross total removal is associated with the most favorable outcome; furthermore, size and extension of these lesions often make total removal impossible in one step. The risk of intraoperative death from uncontrollable hemorrhage is not negligible. Aquamantys<sup>®</sup> is a novel bipolar coagulation device that incorporates a new bipolar coagulation technique, combining radiofrequency energy and saline to provide hemostatic sealing by collagen fibers denaturation.

**Methods:** A 10-year-old girl was admitted with a history of headache for the last 3-months. She had no other neurological deficits and there were no neurocutaneous markers. MRI revealed giant left intraventricular lesion with a volume of 166,63 cm<sup>3</sup> which was intensely enhancing, causing dilatation of lateral ventricle and significant mass effect. Very large draining veins were evident inside the tumor draining in the thalamostriate and internal cerebral vein complex. Cerebral angiography revealed multiple afferences coming mainly from branches of the posterior left choroidal artery with distal afferences, that could not be embolized. A neuronavigation guided parietal craniotomy flap was raised. Navigation guided posterior parietal corticectomy was done. Tumor was completely intraventricular and highly vascularized. The tumor bleeding has been managed with Saline-cooled radiofrequency coagulation, that allowed extensive coagulation of the outer surface of the tumor and progressive bloodless deepening of surgical cavity in alternance with ultrasonic aspiration. Bipolar coagulation and CUSA were used to achieve tumor resection. Results: Gross total excision was performed. The patient had no neurological deficits in the postoperative period. Postoperative contrast-enhanced MRI done revealed no residual tumor. Histopathology report a Grade-1 transitional meningioma.

**Conclusion:** Saline-cooled radiofrequency coagulation offers the possibility to achieve adequate hemostasis also in giant intraventricular tumor in young children, in order to obtain GTR resection minimizing blood loss.

#### Poster Track 2.4:Posterior Fossa

##### PP065

#### Zolpidem's paradoxical restorative action: A case study of functional brain imaging

**Jennifer Boisgontier**<sup>1</sup>, Kévin Beccaria<sup>2</sup>, Thomas Blauwblomme<sup>2</sup>, Ana Saitovitch<sup>3</sup>, Lelio Guida<sup>2</sup>, Ludovic Fillon<sup>3</sup>, Christelle Dufour<sup>4</sup>, Jacques Grill<sup>4</sup>, Hervé Lemaître<sup>5</sup>, Stéphanie Puget<sup>6</sup>, Alice Vinçon-Leite<sup>1</sup>, Volodia Dangouloff-Ros<sup>1</sup>, Mme Sarah Charpy<sup>1</sup>, Sandro Benichi<sup>2</sup>, Raphaël Levy<sup>1</sup>, Charles-Joris Roux<sup>1</sup>, David Grévent<sup>1</sup>, Marie Bourgeois<sup>2</sup>, Lila Saidoun<sup>4</sup>, Raphaël Gaillard<sup>7</sup>, Monica Zilbovicius<sup>8</sup>, Nathalie Boddaert<sup>1</sup>  
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**Introduction:** Zolpidem, a sedative drug, has shown to induce paradoxical effects promoting clinical recovery in a wide range of brain disorders. However, the underlying functional brain mechanism of such paradoxical effect remains poorly understood. Thus, we aimed to investigate functional changes in one patient who developed a postoperative pediatric cerebellar mutism syndrome (pCMS), a postoperative complication characterized by delayed onset transient mutism/reduced speech that may occur after posterior fossa tumor resection, and who experienced zolpidem-induced symptoms improvement within 24h. Indeed, even though there is no consensus regarding the therapeutic approach, a number of clinicians empirically support the use of zolpidem in the context of pCMS.

**Methods:** Brain function was investigated using arterial spin labeling MRI and resting-state functional MRI. Imaging was performed at three time-points: preoperative, postoperative during symptoms and after zolpidem intake. Whole brain rest cerebral blood flow (CBF) was compared two-by-two time points. Functional connectivity was investigated using Pearson correlation coefficient between pairs of regions of interest and compared to values from a control group.

**Results:** In postoperative images, compared to preoperative images, we observed a significant decrease in rest CBF in left supplementary motor area, Broca's area and left striatum and a decrease in functional connectivity within the dentato-thalamo-cortical and cortico-striato-pallido-thalamo-cortical loops. In postoperative images after zolpidem intake, we observed increase of CBF in the left striatum and back-to-normal values of functional connectivity within the disrupted loops.

**Conclusion:** Our results suggest that zolpidem acts alleviating pCMS symptoms through a restoration of disrupted brain function within thalamo-cortical loop. These findings help to extend our understanding regarding the functional brain mechanism of zolpidem in clinical improvements. Moreover, they strongly support thus the relevance of future rest functional investigations, based on randomized controlled clinical trial in large cohort, to provide further input into zolpidem mechanisms acting on neurological recovery.

##### PP066

#### The telovelar approach for IV. ventricular tumors in children: How we do it without removing C1

**Anna Cho**<sup>1</sup>, Cora Hedrich<sup>2</sup>, Muhammet Dogan<sup>1</sup>, Aygül Wurzer<sup>1</sup>, Amedeo Azizi<sup>2</sup>, Thomas Czech<sup>1</sup>, Christian Dorfer<sup>1</sup>

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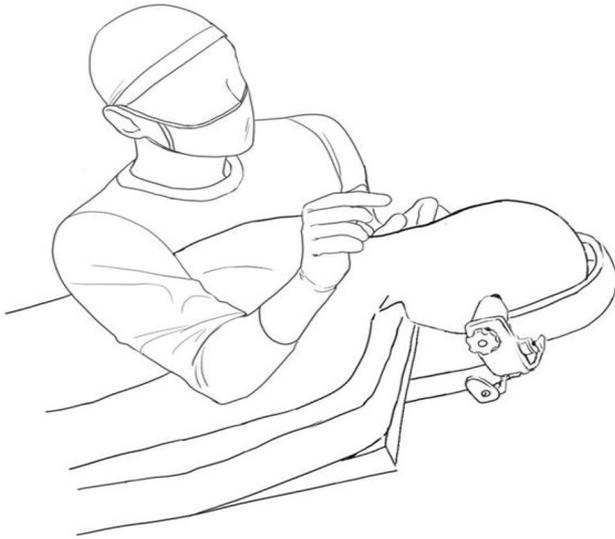
**Introduction:** Various surgical nuances of the telovelar approach have been suggested. The necessity of removing of the posterior arch of C1 to accomplish optimal tumor exposure is still debated. Therefore, we report on our experience and technical details of cerebellar tumor resection in a modified prone position aiming at not removing C1.

**Methods:** A retrospective analysis of all pediatric patients, who underwent a cerebellar tumor resection in the modified prone position between 2012 and 2021 was performed.

**Results:** We identified 61 patients with a median age of 7 (IQR 4-10) and a m:f ratio of 37:24. A telovelar approach was performed in all cases. In 57/61 (93%) of the study population, the posterior arch of C1 was not removed. In the remaining 4 patients the reason for removing C1 was tumor infiltration below the level of C2 or C3. Gross total resection/Near total resection could be achieved in 51/57 (90%), and partial resection in 6/57 (10%) patients. In none of the patients a limited exposure or side of view caused by the posterior arch of C1 was encountered necessitating an unplanned removal of C1. Furthermore, in none of the cases the surgeon had the impression of a limited side of view to the most rostral parts of the IV. ventricle.

Histology revealed 27 pilocytic astrocytomas, 24 medulloblastomas, 5 ependymomas, and 1 high grade astrocytoma.

**Conclusion:** A telovelar approach without the removal of C1 allows for an optimal exposure of the IV ventricle provided that critical nuances in patient positioning are considered.



#### PP067

##### **Management of obstructive hydrocephalus secondary to a posterior fossa tumor in children: about 24 cases operated at the Central Hospital of Army**

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**Introduction:** It's a retrospective study carried out in our institution from 2017 to 2022. The objective is to demonstrate the best surgical method for the management of obstructive hydrocephalus secondary to a posterior fossa tumor in children.

**Patients and Methods:** During this period, 24 children (over 1 year old) were admitted for treatment of the posterior fossa tumor, with obstructive secondary hydrocephalus, the majority benefited from VCS followed by surgery and excision, the rest, received after evacuation from other hospital structures, were managed there beforehand with ventriculoperitoneal diversion or external drainage. We found that the way hydrocephalus is managed has implications for subsequent management, hence the need for this study to determine the effectiveness of different surgical procedures used in the treatment of hydrocephalus in these patients. The evaluation is made on the basis of revision rate, complications, survival and radiological evaluation.

**Results:** 6 patients (25%) received a ventriculoperitoneal shunt (VPD), 15 patients (62%) underwent a ventriculocysternostomy (VCS), and 3 patients (12.5%) received temporary ventricular drainage before or during tumor excision. The postoperative results were almost similar. Nevertheless, a high failure rate (25%) was observed. no deaths are recorded. In total, 75% of children who had a DVP were reoperated. The revision by VCS was performed, in addition to the 4 patients benefiting from a DVP, in 1 patient having received an external drainage and only one revision of a VCS was recorded. In the two patients who received external drainage, a restoration of CSF outflow was observed following tumor resection.

**Conclusion:** VCS is indicated in first intention in the treatment of hydrocephalus secondary to a posterior fossa tumor, regarding satisfactory results obtained, and the high failure rate in DVP especially with the

presence of metastatic cells in the peritoneum but can be considered as a second-line treatment.

#### PP068

##### **Robot-assisted transcerebellar stereotactic approach to the posterior fossa in children**

**Andrea Di Rita**<sup>1</sup>, Matteo Lenge<sup>2</sup>, Simone Peraio<sup>1</sup>, Alice Noris<sup>1</sup>, Giorgio Mantovani<sup>1,3</sup>, Luca Emanuele<sup>1,4</sup>, Maddalena Dardo<sup>1,5</sup>, Flavio Giordano<sup>1</sup>

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**Introduction:** During the last decade there has been renewed interest in stereotactic approaches to the posterior fossa in children, due to the development of new concepts in molecular biology and management of diffuse intrinsic pontine gliomas (DIPGs) and the subsequent need for tissue sampling. Frame-based techniques and robotic assistance are associated with reduced target error in stereotaxy and have been incorporated into standard practice at our institution.

**Methods:** 4 children (age 2-7 years) underwent a robot-assisted frame-based transcerebellar approach using the Leksell G frame and the NeuroMate robot. Procedures included 3 biopsies (two brainstem tumors and one cerebellar hemispheric lesion) and 1 deep electrode implantation for a low-grade tumor of the middle cerebellar peduncle in an epileptic girl. Targeting was based on MRI and in one case 18F-FET-PET was coregistered to MRI to improve sampling accuracy. The frame was applied 180° rotated compared to standard orientation and patients were positioned prone during operation. The posterior post ipsilateral to the approach was omitted. Preoperative stereotactic CT and postoperative CT were obtained. The latter was coregistered to preoperative MRI to check accuracy of targeting.

**Results:** No complications occurred and targeting was accurate in all cases. Tissue sampling was diagnostic and in the epilepsy case deep EEG recording was effective and guided further management.

**Conclusion:** According to our experience a transcerebellar frame-based robotic stereotactic approach to the cerebellum and the brainstem is feasible, safe and effective even in young children.

#### PP069

##### **Management of Hydrocephalus in Paediatric H3K27-Mutant Diffuse Midline Glioma: A Single Centre Retrospective Review**

**Kristy Kehoe**<sup>1</sup>, Lucie Ferguson, Marina Pitsika, Christopher Cowie, Ian Coulter

<sup>1</sup>Royal Victoria Infirmary, Newcastle Upon Tyne, United Kingdom

**Introduction:** H3K27M-mutant Diffuse Midline Glioma (DMG) is a paediatric intracranial tumour with dismal prognosis. The complexity in managing this aggressive and often treatment refractory tumour is compounded by tumour location, typically involving the brainstem or thalami, presenting unfavourable surgical targets. Obstructive hydrocephalus can occur as a result. No optimal management is reported for hydrocephalus in such patients. We present a review of management in our centre.

**Methods:** We retrospectively reviewed paediatric cases of H3K27M-Mutant DMG in our paediatric tertiary centre. We assessed the incidence, clinical and radiological markers of hydrocephalus and subsequent management.

**Results:** 9 patients with a diagnosis of DMG following biopsy were identified over an 8-year period. Mode of biopsy varied by tumour location and clinician preference, and included endoscopic, image guided and stereotactic. Tumour location was found to be brainstem

in 77.7% followed by thalamus (22.2%) and pineal (11.1%). 55.5% underwent surgery for obstructive hydrocephalus – 1 via ETV in the first instance at the time of endoscopic biopsy, 4 via VPS insertion. Clinical symptoms of raised intracranial pressure and radiological features of hydrocephalus were reliable indicators of need for CSF diversion in all cases requiring intervention. Fronto-Occipital Horn Ratio (FOHR), Third Ventricular Bowing and transependymal odema were also reliable predictors. Overall survival was on average 8.5 months from diagnosis.

**Conclusion:** The incidence of hydrocephalus in this sub-group of patients is relatively high and may contribute significantly to morbidity. Clinical and radiological features are reliable predictors for need for CSF diversion and careful consideration at the time of biopsy should be given to prevent further surgery and anaesthetic in this unfortunate cohort.

#### PP070

##### Paediatric intracranial ependymoma: a single centre review over 20 years

**Howra Ktayen**, Pierce Burr, Andrew Croall, John Goodden, Atul Tyagi, Gnanamurthy Sivakumar, Susan Picton, Paul Chumas  
*Leeds Teaching Hospitals NHS Trust, Leeds, United Kingdom*

**Introduction:** Paediatric ependymomas are often difficult to treat. Many factors are considered prognostic, including GTR. We aim to review management and outcome over 20 years and compare this to the literature.

**Methods:** We reviewed all paediatrics patients, under the age of 17, with intracranial ependymoma treated in our centre, between 1.1.2000 to 31.12.2019. Electronic patient records and imaging were reviewed, and clinical characteristics of these patients were retrospectively analysed.

**Results:** We identified 34 patients, with a mean age of 4 years. Female to male ratio was 1:2 The mean age for supratentorial tumours was 4 years and mean age for infratentorial tumours was 4.5 years. 14 patients were under the age of 3. Tumour location: 10 supratentorial and 24 infratentorial. Histological grading: WHO grade II (15) and WHO grade III (19). Two patients had metastatic disease on presentation. With regards to initial symptoms and presentation, nausea and vomiting was the most common and seen in 44% of patients. This was followed by headaches (32%), ataxia (21%), abnormal eye movements (12%), increasing head circumference/motor weakness/papilloedema/seizure/torticollis/visual disturbance (17%), collapse/drowsiness/developmental regression (6%), confusion/developmental delay/dizziness/tremor (3%). We operated on all patients, and first-time gross total resection (GTR) was achieved in 20 patients (59%). Of those with first time GTR, recurrence was seen in 8 patients (40%). All patients received adjuvant therapy, in the form of chemotherapy or proton radiotherapy, except one child whose neurological status was too poor. Overall, 5-year survival in this sub-set for our cohort was 70%.

**Conclusion:** The SIOP Ependymoma 1 study recently published their results in children aged 3–21 years with non-metastatic disease. In this international study GTR was accomplished in 33/74 (44.6%) and 5 year survival was 69.3%. Our results are in keeping with those and include patients under 3 years and with metastatic disease.

#### PP071

##### Endoscopic Transventricular Biopsy in Brainstem Diffuse Midline Glioma

**Emanuele La Corte**<sup>1</sup>, Micol Babini<sup>1</sup>, Beatrice Gardenghi<sup>1</sup>, Mariella Lefosse<sup>1</sup>, Mino Zucchelli<sup>1</sup>

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**Introduction:** Brainstem diffuse midline glioma (DMG) is a predominantly pediatric central nervous system malignant tumor. While historically the diagnosis has been typically made based upon MR imaging, modern progress in surgical techniques and technology lead to incorporate biopsy sampling to advance our understanding of the molecular biology behind DMG.

**Methods:** We describe the key steps of an endoscopic trans-ventricular biopsy of a brainstem diffuse midline glioma and associated ventriculomegaly.

**Results:** The endoscopic transventricular approach for brainstem DMG is indicated when there is ventriculomegaly and/or hydrocephalus and the need for tissue for diagnostic and research purposes. An endoscopic third ventriculostomy associated with biopsy of the exophytic portion of tumor within the interpeduncular and prepontine cistern is performed. The choice of the surgical approach side is dictated by the most representative lateral ventricle volumetry and the best trajectory to target the third ventricle floor and brainstem tumor.

**Conclusion:** The endoscopic third ventriculostomy combined with a punch biopsy of a brainstem diffuse midline glioma represent a feasible and low-risk procedure to simultaneously treat incipient hydrocephalus and achieve a molecular diagnosis for future treatment and research.

#### PP072

##### Case-control study on the occurrence of postoperative posterior fossa syndrome in children - dependent on intraoperative factors?

**Stefanie Ott**<sup>1</sup>, Matthias Belau<sup>2</sup>, Martin Mynarek<sup>3</sup>, Gertrud Kammler<sup>1</sup>, Jens Gempt<sup>1</sup>, Friederike Groth<sup>1</sup>

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**Introduction:** Posterior fossa syndrome (PFS) is a complex syndrome of great neurological impairment occurring most commonly after neurosurgical resection of infratentorial lesions in children. Literature reveals the histology of the lesion, midline location, infiltration of the brainstem or superior cerebellar peduncle associated with the development of an PFS. However, there is shortage of knowledge on the surgeon's role and intraoperative factors in this context. Thus, we aimed to examine the association between intraoperative features and PFS.

**Methods:** The study population included pediatric patients at the University Medical Center Hamburg-Eppendorf, Germany, between 2013–2022 who had a space-occupying lesion in the cerebellum and required surgical therapy. Patients were included if they developed PFS after surgery. Controls were matched to cases for age months at diagnosis using coarsened exact matching. We fitted logistic regression models using weights assigned to the observations' stratum for each intraoperative variable using PFS (yes/no) as the outcome variable and case-control indicator variable as the independent variable adjusting for age months at diagnosis. As intraoperative features we examined the administration of dexamethasone, coagulative factors, mannitol, usage of preoperative CSF drain, either by ventricular drainage or by ventriculocisternostomy, the operative approach and the extend of resection.

**Results:** Of 168 eligible pediatric patients (mean age (SD) months 91.9 (62.4), female 38.8%), PFS occurred in 26 patients. We found a significant association between PFS and intraoperative use of dexamethasone (OR=3.73, 95% CI=[1.01, 13.82]), coagulation factors (OR=4.50, 95% CI=[1.02, 19.92]), and CSF drainage (OR=3.91, 95% CI=[1.19, 12.83]). Other features did not show any significant differences between the groups.

**Conclusion:** Despite the small sample size, the results suggest a relationship between the intraoperative procedure and the occurrence of PFS. Further studies are needed to investigate relevant homeostatic variables and possible influences of the intraoperative methods to name reversible and influenceable causes of PFS.

**PP073****A rare surgical case of Coats Plus Syndrome**

**Amets Sagarrabay<sup>1</sup>**, Dalila Forte<sup>1</sup>, Miguel Correia<sup>1</sup>, Carlos Pontinha, Manuela Mafra, Carla Conceição, Inês Carvalho, Mário Matos<sup>1</sup>  
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**Introduction:** Coats Plus syndrome or cerebroretinal microangiopathy with calcifications and cysts (CRMCC) is a very rare disease with an autosomal recessive pattern caused by a mutation in the encoding conserved telomere maintenance component 1 (CTC1) gene. The CTC1 gene is located on chromosome 17p13.1 and plays an important role in telomere replication. It is a multisystem disorder characterized by retinal telangiectasia and subretinal exudate, intracranial calcification, leukodystrophy, prenatal and postnatal growth retardation, parenchymal brain cysts, osteopenia with predisposition to fractures, bleeding in gastrointestinal tract and portal hypertension can be observed in the patients. CT features are extensive calcifications involving bilateral basal ganglia, bilateral deep cerebellar nuclei and brain stem. MRI demonstrates extensive abnormal white matter hyperintensity suggesting leukodystrophy.

**Methods:** The authors describe the case of an adolescent diagnosed with CRMCC that underwent surgical cyst removal and review of literature.

**Results:** The authors show a surgical case of Coats Plus syndrome in a 14 years old patient with multiple cyst and calcifications in supratentorial and posterior fossa regions and a progressive asymmetric spastic tetraparesis. A pontine cyst was described some years before surgery and it grew in the last two so we decided to remove in order to relieve signs and symptoms of tetraparesis. The postoperative period was uneventful and was discharged without complications. He gain some autonomy in positioning and transferring. The authors show surgical and histopathology description. The revision of literature show less than 100 cases described and few of them underwent brain surgery, so histopathology description is very rare in Coats Plus syndrome.

**Conclusion:** Coats Plus syndrome or cerebroretinal microangiopathy with calcifications and cysts (CRMCC) is a very rare degenerative disease that may be included in the differential diagnosis of cerebral calcification and cysts and may need neurosurgical intervention.

**PP074****Differential expression of markers of inflammatory microenvironment in medulloblastoma allow for classification according to molecular subgroups**

Julian Zipfel, **Martin U. Schuhmann**

*Department of Neurosurgery, Tuebingen, Tuebingen, Germany*

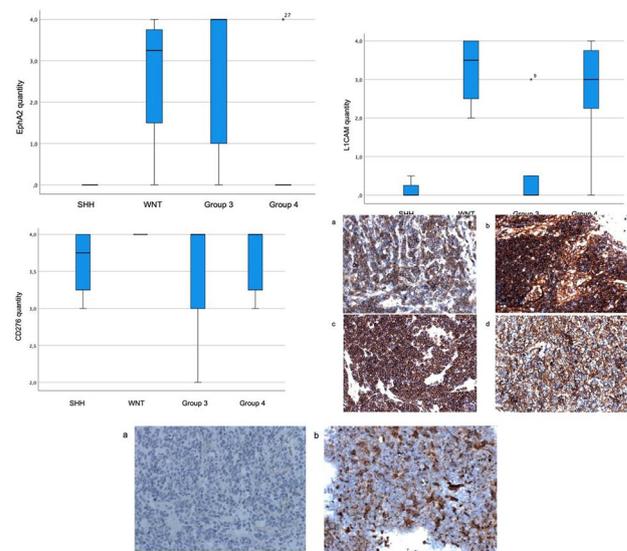
**Introduction:** Medulloblastomas (MB) are the most common malignant primary brain tumors in children. The prognosis of patients diagnosed with these tumors varies greatly and is mainly dependent on molecular subgroups. The exact reasons for clinically distinct behavior of those groups are not yet known. Inflammatory microenvironment may contribute to local infiltration and metastasis as well as resistance to therapy. In this study, the impact of several immunohistochemical markers and the association with molecular subgroups was assessed in a cohort of children with brain tumors.

**Materials and Methods:** In this study, 27 tumor samples underwent tissue microarray (TMA) processing and subsequent immunohistochemical staining. The expression of GD2, CD276, EphA2, L1CAM and Her2 was assessed. Results of (epi)genetic testing and molecular subgroup classification were compared with immunostaining.

**Results:** TMA-based immunostaining yielded stable and repeatable results with a high throughput. WNT-MB as well as Group 3-MB showed significant expression of EphA2. CD276 is expressed in MB

in all molecular subgroups. Thus this might serve as a good target for further evaluation.

**Conclusion:** TMA is an optimal tool for assessment of multiple antigens in large patients populations and samples. It is a cheap method with a high throughput and yields results consistent with literature on classic immunostaining. Expression of EphA2 might be associated with migratory properties in MB. Association to outcome has to be assessed in larger cohorts. CD276 is expressed universally in all molecular subgroups and might serve as a promising target for individualized therapy.

**PP075****Pediatric posterior fossa tumors in Slovenia 2018-2022**

**Peter Spazzapan**, Borut Prestor, Roman Bošnjak

*University Medical Centre Ljubljana, Ljubljana, Slovenia*

**Introduction:** The incidence of pediatric brain tumors varies among different countries, from 1.15 to 5.14 cases per 100,000 children. The rate of posterior fossa tumors is higher in children compared to adults.

**Methods:** We analyzed all pediatric posterior fossa tumors that have been treated surgically at the University Medical Center Ljubljana between October 2018 and April 2022. We included all cases aged between 0 and 18 years who needed a surgical treatment, defined as biopsy, subtotal and total excision.

**Results:** Among 60 pediatric tumors treated at our Institution, 18 were located in the posterior fossa. The median age of the patients was 5 years. The tumor locations were as follows: 4th ventricle in 10 cases, cerebellar hemisphere in 6 cases, pontocerebellar angle in 2 cases. The histopathological examinations revealed the diagnosis of: medulloblastoma in 9 cases, pilocytic astrocytoma in 6 cases, ependymoma in 2 cases and schwannoma in 1 case. In 15 cases the resection was total, in 3 cases subtotal. 2 children had a wound dehiscence, one had a CSF fistula. We observed 4 cases of cerebellar mutism, which showed a long-term improvement in 3 cases.

**Conclusions:** Posterior fossa tumors represented 30% of all pediatric brain tumors, which arose in the Slovenian population with an incidence of 3.8 per 100,000 children a year. Cerebellar mutism and wound dehiscence represented the most common complications.

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