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ABSTRACTS

26th Congress of the European Society for Pediatric Neurosurgery (ESPN) Bonn-Germany, 6-9 May 2018

Oral presentations

Abstract session 1: Craniovertebral junction

OP01

"Bony only" decompression for Chiari malformation type 1 and 1.5 – A safe fist-line option

Friederike Knerlich-Luksochus, Stephanie Jünger, Martina Messing- Jünger (Sankt Augustin, Germany)

Objective: Controversy concerning optimal surgical approaches for Chiari (CH) type I is ongoing. Recent studies imply decompression with duroplasty more effective than bony removal with dural splitting. We review our experience of treating CH 1/1.5 with decompression of the craniocervical junction without duroplasty.
Methods: The surgical database of our hospital was searched for patients who received surgery for CH 1/1.5(2015 to 2017). Cases were selected in whom suboccipital craniectomy plus C1-laminectomy without dural opening was performed. Patients' records were analyzed retrospectively regarding clinical history, neurological exams, polysomnography, electrophysiology. Pre- and post-surgical (after 3 months and most recently) MRI studies were reviewed for cerebellar tonsil herniation, syringomyelia, pre-syrinx state. Foramen magnum decompression measured 3-4 cm wide and 3-4 cm. The dural band at the foramen magnum was released and the superficial dural layer split. In all cases intraoperative ultrasound was performed.
Results: 22 patients were included (CH 1.5 n=8). Mean age at surgery was 13 y ±7 (plus one 60y old patient; 11 male, 11 female). Pre-operative mean tonsillar herniation was 15.47 ±7.29 mm. Symptoms included sleep apnea, suboccipital headaches, positively related with vasalva maneuver, sensorimotor deficits, nausa, vomiting, ataxia. 5 patients had scoliosis, 11 syringomyelia, one pre-syrinx state. In 11 cases surgery resulted in complete resolution of occipital headaches, nuchalgia, ataxia (for 5 patients, follow-up was not yet available). In 7 cases tonsils ascended after surgery. Mean tonsilar herniation after surgery was 11.55±7.13. Improvement in syringomyelia was seen in 3 patients. There were no postoperative complications. Repeat surgery was performed in 2 patients (one received second surgery with duroplasty; in the other bony decompression was expanded).
Conclusion: Bony decompression can be considered a safe fist-line option for CH 1/1.5 malformation. Also, patients with syringomyelia may profit from this approach.

OP02

Natural history and treatment failure of syrinx associated with Chiari I malformation after foramen magnum decompression: how should we proceed?

Jehuda Soleman, Andrea Bartoli, Akiva Korn, Shlomi Constantini, Jonathan Roth (Tel Aviv, Israel; Geneva, Switzerland)

Objective: The natural history of Chiari I (CMI)-associated-syrinx following foramen magnum decompression (FMD) is not fully defined. Surgical treatment of patients with persistent, recurrent, or progressive syrinx after FMD is debated. Some advocate re-decompression of the foramen magnum, while others suggest a shunting procedure (such as a syringopleural or syringo-subarachnoid shunt). Our aim is to describe the natural history and discuss the different treatment modalities for these patients.
Methods: We retrospectively collected data of CMI patients with persistent, recurrent, or progressive syrinx after FMD. In addition to baseline characteristic, surgical treatment options, neurological and radiological outcome, time between FMD and second surgery, morbidity, mortality, and overall follow up time were assessed. We provide a descriptive analysis of the natural history of failed FMD, and of the different treatment modalities and their outcome.
Results: At our department, between the years 1998 and 2017, 48 consecutive patients (35 females (72.9%), average age 16.8 years (±11.5 years)) underwent FMD for a CMI-syrinx complex. Of these, 24 patients (50%) underwent surgical treatment for a persistent (n=10), progressing (n=12), or recurrent (n=2)

syrinx on average 21.4 months (± 27.9 months, median 14.6 months, range 12 days - 134.9 months) after FMD. Two patients (8.3%) underwent redo FMD, 18 patients (75%) underwent 19 syringo-subarachnoid-shunt procedures, and 4 patients (16.7%) had 6 CSF diversion procedures. Overall follow up time was 40.1 months (±47.4 months, median 25 months, range 3 months - 230 months).

Conclusion: Based on our results it seems that 50% of the patients undergoing FMD for CMI syrinx complex will need surgical syrinx treatment due to persisting, progressing or recurrent syrinx. This treatment is tailored according to the suspected underlying pathology causing the FMD failure. A subgroup of patients can be managed conservatively; however, these patients need close clinical and radiological follow up.

OP03

Complete reversibility of the Chiari type II malformation following post-natal repair of myelomeningocele

Federico Di Rocco, Pierre-Aurélien Beuriat, Alexandru Szathmari, Carmine Mottolese (Bron, France)

Introduction: Chiari type-II malformation (CM-II) has been associated with spina bifida (SB) and especially myelomeningocele (MMC) for a long time. It is reported that more than 80% of MMC patient at birth will have tonsillar herniation. Moreover, CM-II can become symptomatic and require a surgery. We report a large group of MMC patient treated at birth for the MMC and focused on the group of patients that presented a complete ascent of the cerebellar tonsils after the MMC repair.

Material and Methods: Sixty-one patients were included in this study. They were operated from October 1997 to September 2016. The CM-II was considered present when the tip of the cerebellar tonsil was more than 5 millimeters below the McRae line (occipito-clivus line) according to the MRI. The presence or absence of the CM-II after the closure of the spinal dysraphism was assessed on the most recent brain imaging available for the patient.

Results: Forty-seven (77%) patient had a CM-II at birth. On the last cerebral MRI, only 28 (46%) had a remaining CM-II. Statistical analysis showed that patients with higher vertebral lesion of the MMC had significantly more risk to have a CM-II at birth (p=0,006). The reversibility is also significantly more frequent in lower lesion patient (p=0,004). There is significantly more patient with a remaining CM-II that required surgery for hydrocephalus (p=0,048).

Conclusions: The post-natal management of MMC allowed the reversibility of the CM-II. Modern techniques with surgical microscope permit the good restauration of the CSF dynamic. This is confirmed by the fact that more patients with a remaining CM-II will required a CFS diversion procedure.

OP04

Grisel syndrome in pediatric age: an Italian single center experience and review of the literature

Gianluca Piatelli, Pasquale Anania, Mattia Pacetti, Alessandro Consales, Marco Pavanello, Marcello Ravegnani, Armando Cama (Genoa, Italy)

Introduction: Grisel syndrome is a non-traumatic subluxation of atlanto-axial joint. The most frequent cuases described are infections, followed by surgery. Pediatric population has the higher frequency of presentation, probably for the greater cervical ligamentous laxity. Inflammatory process directly spreads from the ENT (eyes, nose, throat) area to the adjacent paravertebral cervical muscles, increasing the hypermobility and laxity of ligaments surrounding the articulation.

Methods: We retrospectively reviewed all the clinical records of patients presenting at our Institute with a sudden onset torticollis in the last few years.

Results: 5 patients were found to be diagnosed of non-traumatic subluxation of the atlanto-axial joint. 4/5 (80%) patients had experienced different inflammatory process of the neck and the mean time from the inflammatory episode to torticollis onset was 11 days, but the mean time from the onset to when they come to our attention was 172 days (3/5 patients had an inveterate luxation). 3/5 (60%) patients were found to have radiological signs of inflammation in the neck. 5/5 patients (100%) underwent positioning of Halo jacket, with progressive reduction of rotator subluxation. Halo jacket was the single treatment for only 1 patient (20%) who was treated 20 days after the onset. The other patient with recent subluxation was treated in another center primarily with Philadelphia collar and recurred after its removal (after 2,5 months); thus, he needed Halo fixation and reduction with subsequently surgical arthrodesis. 3/3 (100%) patients with inveterate subluxation needed surgical arthrodesis.

Conclusion: There is no a real gold standard for the management of this condition; the data analysis in our center shows that in case ofinveterate subluxation the best treatment is Halo fixation with progressive reduction and surgical arthrodesis. For recent subluxation we suggest primarily Halo fixation and reduction with immobilization for at least 3 months.

Abstract session 2: Craniofacial I

OP05

Intracranial hypertension and sleep-disordered breathing in children with craniosynostosis

Kylie Russo, Martin Samuels, Francois Abel, Justine O'Hara, David Dunaway, Owase Jeelani, Gregory James (London, United Kingdom)

Introduction: Sleep-disordered breathing (SDB), by means of hypercapnia-related vasodilatation, can be a contributor to

raised intracranial pressure (rICP). Many children with craniosynostosis have SDB – either obstructive sleep apnoea (OSA) related to midface hypoplasia and airway problems, and/or due to central causes such as Chiari I- in addition to rICP. In our institution, children who have unequioval findings of rICP such as frank papilloedema progress directly to treatment (usually vault expansion). We use ICP monitoring for "borderline" cases such as subtle disc changes or in the presence of concerning radiological or clinical features.

Methods: Since 2015, we have routinely carried out simultaneous sleep studies (SS) for children undergoing ICP monitoring to assess the physiological relationship between these phenomena. We report data from all children with craniosynostosis who underwent combined SS/ICP monitoring in this period.

Results: 27 consecutive patients were identified (19 males), with a mean age of 6 years (range 1-18 years). Diagnoses were Apert (8), sagittal (6), multisuture non-syndromic (5), Crouzon (5), Muenke (2) and Pfeiffer (1). Indications for undertaking ICP/SS were: ophthalmic examination findings (13), clinical symptoms e.g. headache (11) and deteriorating visual electrophysiology (7). 10 patients were found to have normal ICP, of which 8 had normal SS and 2 had mild OSA. 14 had rICP, of which 12 had normal SS and 2 had moderately severe OSA. 2 children had low ICP, both with normal SS, and 1 child had borderline ICP, and mild OSA. 10 patients had undergone previous airway interventions. In the high ICP group, physiological rises in pCO_2 caused significant ICP spikes. All children with rICP went on to have vault expansion, the others were managed symptomatically.

Conclusions: There appears to be a relationship between OSA and raised pressure in a proportion of children with craniosynostosis. Understanding this relationship may aid monitoring and decision making in these patients.

OP06

Improvement of sleep apnea after FrontoFacial MonoBloc Advancement (FFMBA): in 108 faciocraniosynostoses

Eric Arnaud, Samer Haber, Giovanna Paternoster, Syril James, Hossein Khonsari, Philippe Meyer (Paris, France)

Background: Long-term effectiveness of Frontofacial Monobloc Advancement (FFMBA) on sleep apnea was analyzed.

Methods: This is a monocentric prospective cohort study on 108 kids with FCS treated by FFMBA with 4 internal distractors and with a 5 months consolidation time. The primary outcome was achieving a normal Apnea Hypopnea Index (AHI) (less than 5/hour) on polysomnography (PSG) without additional surgery. Age at operation was 47.7±36.6 months. Mean follow-up was 56.5±48.6 months.

Results: 69 children (63.9%) achieved at least 50% improvement in AHI. Among them 39 (36.1%) normalized their AHI without additional surgery in 28.3±28.3 months while 16 others (14.8%) did so after additional surgery (ENT). 10 patients (9.3%) eventually relapsed in 22.0±14.0 months. Previous turbinectomy and tonsillectomy were associated with a better response rate, a faster response, a longer period before relapse, and less need for additional surgery ($p<0.05$). FGFR2 mutation, and previous posterior or lateral decompression or Rénier H technique were all associated with less relapse, while previous fronto-orbital or facial advancement were associated with more relapse. Higher AHI at baseline was associated with secondary facial advancement (21 children, 19.4%). Age at operation was not significantly associated with treatment response and relapse. However, young age was associated with previous tracheotomy, higher AHI at baseline, Pfeiffer syndrome, use of transfacial Kirschner wires with external traction, and absence of previous turbinectomy and tonsillectomy, corresponding to greater severity.

Conclusion: FFMBA is an effective procedure to correct or minimize OSA in FCS. Turbinectomy, tonsillectomy and cranial vault expansion are indicated before FFMBA. Previous FOA or Lefort 3 before FFMBA are associated with earlier relapse.

OP07

Pattern of closure of the metopic suture: an anatomical study on 460 normal children

Matthieu Vinchon (Lille, France)

Background: The metopic suture, unlike other sutures of the calvaria, closes during early infancy. The persistence of an open metopic suture is referred to as metopism. However, the timing and pattern of closure of the metopic suture is poorly documented. The following questions remain unanswered: at which ages does the closure of the metopic suture start and end? How do we define metopism? And what are the consequences of metopism?

Material and Methods: Starting in 2012, we collected prospectively 3D-CT scanners of children admitted for head trauma, selecting patients with no previous medical history, and excluding children with chronic modification of the calvaria like in subdural hematoma or hydrocephalus. On the basis of 3D reconstructions, we defined the metopic suture as unfused, partially fused or totally fused.

Results: We studied 460 children, 351 of these being 2 years or younger. The median age of onset and completion of closure of the metopic suture was 4 months and 9 months respectively. The metopic suture remained totally unclosed in 4.7% of children older than one year, a proportion which remained stable in children above 5 years. We conclude that metopism can be asserted if the metopic suture is open at one year.

From the 3D reconstruction, we found that temporal bulging above a pterional narrowing was a hallmark of metopism. In addition, metopism was commonly found in association with cranium bifidum.

Conclusions: 3D-CT of normal children victim of head injury represent an abundant material for the study of anatomy and development.

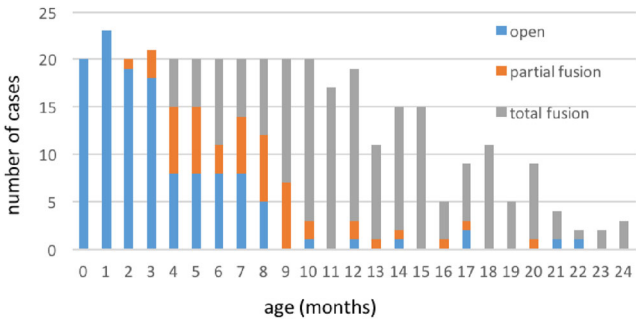

Figure 1: *Pattern of metopic fusion*

OP08

Characterization of the calvarial suture skeletogenic stem cell niche in non-syndromic craniosynostosis: towards cell-specific targeting in the design of advanced therapies

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*Equal contribution

Introduction: Nonsyndromic craniosynostosis (NCS) is a congenital defect due to the premature fusion of skull sutures. The suture mesenchyme houses a skull-specific stem cell niche, which could implicate in premature suture ossification. To test this hypothesis, we characterized the stem cell niche of open and fused sutures of patients with sagittal NCS.
Methods: The expression of lineage-specific markers (THY1, skeletal stemness-marker; TEK, haemopoietic/angiogenic marker; ENPEP and ITGAV, bone marrow stem cells differentiation markers; GLI1, a putative calvarial stemness-marker; AXIN2, mesenchymal cell fate determinant) was analyzed by qPCR and immunofluorescence, in suture tissues and in calvarial stem cells (CMSC). Mesenchymal stem cells isolated from alternative tissue sources served as controls.
Results: Both NCS suture tissues and CMSC isolated thereof expressed reduced levels of *TEK* and *ENPEP* compared with controls. *AXIN2* levels were higher in open suture-derived CMSC than in fused suture-cells and in controls. Upon *in vitro* osteogenic induction, *THY1* and *GLI1* expression decreased, whereas *AXIN2* levels increased, in both open- and fused- suture CMSC. Immunofluorescence showed that AXIN2 is mainly expressed at the sub-endosteal site, while THY1 is primarily expressed in proliferating cells within the trabeculae.
Conclusion: All CMSC expressed the same marker genes, indicating that explant culture allowed selecting comparable cell populations from both sutures and synostoses. Possibly, the tissue microenvironment plays a key role in determining a different CMSC fate leading to enhanced osteogenic differentiation and premature suture closure in NSC. $THY1^+/GLI1^+$ cells represent reasonably the main stem cell population in the calvarial niche, as the expression of these markers decreased upon *in vitro* osteogenic differentiation. Targeting suture stem cells to modulate their osteogenic potential *in vivo* may represent a future strategy to test aimed at implementing advanced therapies with reduced invasiveness and complications.
Acknowledgement: Study supported through funds from: NIH/NIDCR, Federazione Gene and Università Cattolica del Sacro Cuore.

OP09

The 3D evolution of the normal cranium during the first 2 years of live

Jene W. Meulstee, Guido A. de Jong, Wilfred A. Borstlap, Thomas Maal, Hans Delye (Nijmegen, the Netherlands)

Introduction: During the first years of a newborn's life, the cranium grows very rapidly. Insight in this development is essential for monitoring cranial development of new-borns, identify cranial abnormalities and for the follow up of craniosynostosis surgery. This study presents a three-dimensional (3D) evaluation of the normal cranial shape by the use of 3D stereophotographs.
Methods: A total of 150 3D stereophotographs of new-borns without abnormalities were used to evaluate cranial evolution. All 3D photos were acquired between 3 and 24 months of age with a fixed interval of 3 months. This dataset was used to calculate the cranial length, cranial width, cranial index, cranial circumference and volume. Distance maps of the complete 3D morphology were produced to create a 3D perception of the cranial evolution.
Results: Circumference of the head measured on 3D photographs increased from 412 to 502mm (22% increase). Cranial index showed a small decrease, from 78 to 77 (2% decrease). Volume increased from 982 to 1715 ml (75% increase). Evaluation of the 3D distance maps revealed a more prominent growth of the anterior part of the skull between 3 and 12 months. The posterior part of the skull developed more rapidly between 12 and 15 months conform the rest of the skull and an almost uniform growth was seen between 15 and 24 months.

Conclusion: This 3D analysis of the normal cranial shape provides a valuable insight of the evolution of the cranium during the first 24 months of live. This study presents data which can be used for monitoring cranial development and for the follow-up of craniosynostosis surgery.

OP10

Identifying ophthalmic abnormalities and optimizing pediatric ophthalmology screening in non-syndromic craniosynostosis

Aabgina Shafi, Nikolaos Vryonis, Dimitri Shastin, Paul Chumas, Janice Hoole, Vernon Long (Leeds, United Kingdom)

Introduction: The purpose of this study was to compare the ophthalmic abnormalities (viz., refractive error, strabismus, proptosis, papilloedema, or optic disc atrophy) between the children with different type of simple suture and multiple suture non-syndromic craniosynostosis and identify ideal follow-up times for screening.
Methods: Retrospective review of medical records of consecutive children who underwent craniosynostosis surgery at Leeds Teaching Hospitals NHS Trust between January 2009 and December 2013.
Results: A total of 90 children with non-syndromic craniosynostosis were identified for this study. Mean age of craniofacial surgery was of 17.7 months, with a tendency for later age of surgery identified in complex craniosynostosis. Mean logMAR best corrected visual acuity was 0.21. A significant refractive error was found in 24.4% of patients, astigmatism being the most common. Coronal synostosis had the highest incidence of refractive error. Strabismus was present in 14.4% of patients, coronal and complex craniosynostosis having the highest prevalence. Proptosis and papilloedema were only present in complex craniosynostosis. There was no measurable change in refractive error and strabismus pre- and post-craniofacial reconstructive surgery.
Conclusion: This study adds to our understanding of ocular features of non-syndromic craniosynostosis and will help in prognostication and counseling of these patients. Coronal and complex craniosynostosis have the highest incidence of ophthalmic co-morbidity, with sagittal and metopic having the least. If we may infer from this study that sagittal suture craniosynostosis do not develop any clinically significant refractive error or strabismus, it follows that regular screening of this subset of patients is not mandated. This would reduce a significant number of patients to be screened at regular intervals. Not only would this make our screening service more efficient and cost effective, patients would also benefit from reduction in unnecessary appointments. The authors recommend that ophthalmic screening in sagittal suture non-syndromic synostosis is not warranted routinely.

Abstract session 5: Craniofacial III

OP11

Orbital morphology in Crouzon syndrome: a 3D assessment before and after fronto-facial monobloc advancement with internal distraction

Roman Hossein Khonsari, Johan Nysjö, Ronak Sandy, Ingela Nyström, Giovanna Paternoster, Eric Arnaud (Paris, France; Uppsala, Sweden; Aalborg, Denmark)

Aim: Oculo-orbital disproportion is a major concern in Crouzon syndrome (CS), leading to insufficient eyelid closure and eventually functional visual impairment. Fronto-facial monobloc osteotomy aims to correct craniofacial growth deficiencies including oculo-orbital disproportion.
Material and Methods: Twenty-four patients with CS and 48 controls were included. Four time-points were considered: pre-op., early post-op. (within one month after the procedure), 6 months after surgery and 12 months after surgery. Orbits were segmented using a semi-automatic mesh-based method. Mean models were generated using a registration-based technique and were compared visually and quantitatively using color-coded distance maps, Maximum Absolute Distance (MAD), Hausdorff Distance (HD) and Dice Similarly Coefficient (DSC). Orbital symmetry was assessed by mirroring the left side on the right side and computing the volume overlap. We tested MAD≤1.5 mm, HD≤1.5 mm, and DSC≥0.9 between CS and control groups for each time point and used the same criteria to assess symmetry.
Results: Shape normalization was obtained qualitatively in all groups after fronto-facial surgery. Volumes were significantly more similar to normal in the 5-12 years age group after surgery and stayed stable. Volumes were significantly larger than controls in the 0-5 age group before surgery and remained stable. Quantitative assessment based on MAD, HD and DSC showed that normalization wa s obtained after 1 year of follow-up in the 5-12 years group but not in the 0-5 years group, where the 3D shape of the orbits remained different from controls despite significant normalization. There was no tendency for relapse in both age groups.
Conclusion: Fronto-facial surgery significantly normalizes orbital shape and volume in Crouzon syndrome. The 3D assessment of the orbital inner mould is an indicator of the multifactorial effets of monobloc advancement and a method to monitor its stability.

Abstract session 7: Technology in Hydrocephalus

OP12

Upward movement of cerebrospinal fluid in vivo and its relation to the brain venous system

Steffi Dreha-Kulaczewski, Arun Joseph, Klaus-Dietmar Merboldt, Hans-Christoph Ludwig, Jutta Gärtner, Jens Frahm (Goettingen, Germany)

Introduction: CSF dynamics and its impact in the pathogenesis of various forms of hydrocephalus are still poorly understood. Forced inspiration has recently been identified as main driving force of CSF flow using real-time magnetic resonance imaging (MRI). Exploiting technical advances quantitative measurements of flowdirection, velocities and volumes were currently studied.

Methods: Real-time flow MRI was performed at 3 Tesla (Magnetom Prisma, Siemens Healthcare). It combines high spatial ($1.2x1.2x5mm^3$) and temporal (135 ms) resolution and requires acquisition of two high-speed images with differential velocity encodings, perpendicular to the imaging section. Quantitative analyses were conducted using CAIPI prototype software (Fraunhofer MEVIS, Bremen). In 12 healthy subjects CSF flow was measured in four region-of-interests (ROI): aqueduct, at C3, Th2 and Th5. For analysis of venous flow, a ROI was placed in a prominent epidural vein at C3 as part of the posterior venous drainage.

Results: Consistent and prompt upward CSF movement towards the brain in response to forced inspiration was seen in all subjects at the aqueduct, in 11/12 subjects at Th2 and in 4/12 subjects at Th5. The mean CSF flow volume of all subjects during deep inspiration was positive in all three ROIs substantiating its upward directionality. Cardiac-related CSF flow remained minor and variable depending on anatomic conditions. Concomitant analyses of CSF dynamics and epidural venous blood flow at C3 revealed an upward flux of CSF and an enhanced downward flow of venous blood during inspiration.

Conclusion: Our analyses demonstrated CSF and venous flow to be closely communicating cerebral fluid systems where inspiration-induced downward flow of venous blood due to reduced intrathoracic pressure is counterbalanced by an upward movement of CSF. The results extend our understanding of human CSF flux and opens important clinical implications, namely concepts for drug delivery, new classifications and therapeutic options for various forms of hydrocephalus.

OP13

The relationship between intracranial pressure and age – Chasing age related reference values

Sarah Hornshoej Pedersen, Alexander Lilja-Cyron, Marianne Juhler (Copenhagen, Denmark)

Introduction: In neurosurgery, one aim of treatment is normalization of intracranial pressure (ICP). No true reference values for ICP in humans exist and current values are estimates based on measurements in adults who underwent treatment in order to correct ICP.Further, no studies compare ICP measurements in children to measurements in adults and it seems silently assumed that ICP are the same, regardless of the physiological differences. We report ICP values in a "pseudo-normal" group of children and adults.

This study aims:

• To clarify if day and night ICP differs between children and adults

• To examine if age affects ICP

Methods: We analysed data from all non-shunted patients undergoing invasive elective diagnostic 24 hours ICP monitoring from February 2008 to November 2014. Thirty-five patients (14 children, 21 adults) met the inclusion criteria with no subsequent suspicion of increased ICP and therefore no need for pressure-relieving treatment. Follow-up time was 3-9 years.

Data were separated into day and night sequences and the mean value was determined.

Results: 91% of the patients had higher ICP during night-time. Day-time ICP was respectively 2.8±2.2 mmHg in children and 1.9±4.2 mmHg in adults ($p= 0.39$). The difference between day-time and night-time ICP was similar in children ($\Delta ICP=5.8\pm4.0$ mmHg, $p<0.0001$) and adults ($\Delta ICP=6.1\pm3.3$ mmHg, $p<0.0001$).

ICP could be described as a decreasing function of age, with an ICP decrement of 0.69 mmHg per decade ($p=0.015$).

Conclusion: In this study of patients with no ICP related disease, both children and adults showed a nocturnal ICP increase. There were similar differences between day and night ICP in the two groups. However, across all ages, ICP seemed to decrease with age. This could contribute to the understanding of normal human physiology and impact therapeutic interventions, e.g. shunt valve selection or resistance in external ventricular drainage.

OP14

Infant external hydrocephalus is a cerebro-venous disease related to outflow impairment

Laura Sainz Villalba, Laura Laffitte, Martin U. Schuhmann (Madrid, Spain; Tübingen, Germany)

Introduction: External hydrocephalus (eHC) is defined as macrocephaly, enlarged subarachnoid spaces and no or moderate ventricular enlargement. Observed in infants below 1-year eHC subsides spontaneously in the first 2-3 years, however macrocephaly remains. Newer studies point to an impaired reabsorption of CSF due to venous hypertension as the underlying cause of eHC. This study evaluates the

correlation between venous system abnormalities and the volume of the subarachnoid spaces in eHC.
Methods: Infants newly diagnosed with external hydrocephalus received a contrast enhanced MR phlebography with 3 D sinus reconstruction. Clinical presentation and radiological findings were recorded, and venous features were graded according to the number of affected segments and type of abnormality. Ventricular width was assessed with Evans index and frontooccipital horn ratio (FOHR). Extraaxial CSF volume was planimetrically determined using BrainLab 2.0 software.
Results: 13 patients with a mean age of 9 months were studied. Mean Evans index and mean FOHR were 0.27 and0.35, respectively. Subarachnoidal extra-axial CSF volume had an average of 167.25 cm^{3}·. 13/13 patients presented with an abnormal sinus anatomy and stenosis in phlebography. The most frequently affected segment was the sigmoid sinus (in 61.5%). 9/13 children were managed conservatively. The number of affected venous segments showed a highly significant positive correlation to the subarachnoid volume, indicating that a more severe venous outflow impairment leads to ta larger extraaxial CSF volume (ANOVA p=0.0295). No correlation between ventricular width and extraaxial volume was found.
Conclusion: Venous outflow impairment seems to play a major role in the development of eHC. Thus, the name hydrocephalus is misleading since it is not a primary CSF born problem but a venous system disease. Further studies and larger cohorts have to confirm that eHC has the same pathophysiology just at an earlier time point than pseudotumor cerebri.

OP15

Use of EOS® low-dose biplanar X-ray for shunt series in children with hydrocephalus: a preliminary study

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Introduction: Shunt series (SS) are a common diagnostic tool used to verify shunt integrity. SS include X-ray films of the skull, chest, and abdomen, and are often performed either when a shunted patient presents with suspected shunt malfunction, or as a screening test to identify shunt disconnections. Over recent years, EOS low-dose biplanar X-ray have been used for various indications and are associated with significantly reduced radiation doses compared to ordinary X-rays. This is the first publication on the use of EOS as a SS technique.
Methods: Over a period of 4 months, EOS were performed at our center for various orthopedic indications, mostly for evaluation of scoliosis. Nine children were identified as having a VPS and served as the study group. We retrospectively reviewed the shunt visibility and integrity in both EOS scans, as well as regular SS or plain spinal X-rays.
Results: Nine patients with a VPS that underwent EOS imaging of their spine were identified. 3 had bilateral shunts, 8 had either SS or regular spinal X-rays. In all patients, the shunt integrity was easily demonstrated on the EOS images. 2 patients had an identified shunt disconnection confirmed on the EOS images. No shunt related information was missed on the EOS compared to the other x-ray images.
Conclusion: These preliminary results suggest that EOS may be used as an alternative technology to demonstrate shunt integrity instead of regular X-ray SS. Favorable shunt visibility without the need for multiple radiation exposures and image processing (such as stitching), results in a significantly shorter examination time with significant less radiation.

OP16

The ShuntScope – A technique for catheter placement in complex cases of hydrocephalus in Pediatric Neurosurgery

Stefan Linsler, Sebastian Antes, Sebastian Senger, Joachim Oertel (Homburg, Germany)

Objective: The long-term preserving of ventriculoperitoneal shunt function depends on the correct placement of the ventricle catheter. Nearly 4% of ventricular catheters are misplaced. There are already many tools to optimize the insertion, such as neuronavigation or stereotactic techniques.
Methods: We used the new semirigid ShuntScope (Karl Storz, Germany) for the catheter placement in cases of slit ventricles, pseudotumor cerebri, cystic lesions and in cases of aqueductoplasty. The study was carried on 33 patients (19 males, 14 females), mean age 40 (2 months to 17 years). The ShuntScope was used diagnostic and therapeutic. The small outer diameter of 1mm allows to pass through the catheter under directly endoscopic view with a very good image resolution of 10,000 Pixels.
Results: The main value of this new technique was the ability to place the catheter tip accurately within distorted, cystic or small ventricles. Even controlled and guided placements in the fourth ventricle were possible in cases of aqueductoplasty. The semirigid endoscope allowed a safe intraoperative correction of the catheter placement due to a cover sheet function of the ventricular catheter. The postoperative neuroradiological imaging revealed the catheter tip placement analogous to the intraoperative findings and video recording analysis. No bleeding complications or infections occurred.
Conclusion: The ShuntScope is very useful for safe catheter placement in complex cases of hydrocephalus. With this new technique misplacements of shunt catheters are completely avoidable. The ShuntScope technique should be implemented in Pediatric Neurosurgery.

OP17

Computerized Shunt-Infusion-Study (SIS): radiation free, minimal invasive quantitative assessment of hydrocephalus shunt function

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Objective: Hydrocephalus shunt malfunction can – especially in children – occur insidiously without symptoms of raised intracranial pressure (ICP) or without changes in ventricular size. It therefore imposes a diagnostic challenge. Imaging-based shunt-tests are, if at all, qualitative and imply radiation exposure. Computerized shunt-infusion-studies enable a quantitative shunt function assessment. We report on feasibility and results of this technique.

Methods: Shunt-Infusion study (SIS) is performed with two butterfly needles being inserted into the pre-chamber, one for computerized ICP recording, and one for mock CSF infusion. After baseline ICP recording, infusion is started at 1.5 ml/min until a new ICP plateau (ICPpl) is reached. Dedicated software (ICM+) containing the shunt's resistance characteristics, calculates baseline and plateau ICP, ICP-amplitudes, elastance, outflow resistance and critical shunt pressure (CSP = maximum plateau pressure at normal shunt resistance).

Results: 39 SIS were performed in 35 children. Functional shunts were defined by ICPpl >1mmHg below CSP, non-functional as >1mmHg above CSP and borderline in between. 17 (43,6%) of shunts were obstructed, 3 (7,7%) borderline and 19 (48,7%) functional. Baseline ICP in obstructed shunts was significantly above shunt working pressure (n=0.003). CSF outflow resistance and ICP plateau were significantly elevated (n=0.001) in obstructed shunts, cut-off thresholds were 5.9 mmHg/ml/min and 8.5 mmHg, respectively. All obstructed shunts were revised. In 9/17 the ventricular size decreased postoperatively, with 8/17 also showing an improvement on clinical tests or according to parent's reports. In any case an enlargement of the external CSF spaces or a normalised head growth was observed.

Conclusion: SIS is a feasible, elegant and radiation free technique for quantitative shunt assessment to rule out or prove shunt malfunction. Dedicated software containing shunt hydrodynamic characteristics is necessary and small children will need short-term sedation. SIS should become routine in pediatric neurosurgery units.

Abstract session 8: Vascular

OP18

The use of direct ICP and brain tissue oxygen monitoring in the perioperative management of patients with Moyamoya disease

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Introduction: Moyamoya disease is characterised by progressive stenosis of the internal carotid arterial circulation. There is a high risk (10-20%) of ischaemic events during revascularisation surgery. Impaired cerebral autoregulation has been proposed as one of the factors for this. We examined the cerebral oxygenation and indices of autoregulation in the pathological hemisphere during the perioperative period.

Methods: Indirect revascularisation using pial synangiosis was performed in six cerebral hemispheres, in four patients. Patient ages ranged from 8 to 18 years. All patients were Suzuki stage 3. General anaesthesia was maintained with desflurane (1.0 minimum alveolar concentration) and remifentanil (0.05-0.2 mcg/kg/min). Preoperative systolic blood pressure was maintained intraoperatively using an intravenous infusion of noradrenaline. Nimodipine (1.5 mg/kg/h IV) was given intra- and postoperatively. Blood pressure was measured using a radial arterial line, zeroed to the external auditory meatus. Direct ICP and brain tissue oxygen was monitored using a Raumedic PTO catheter sited frontally on the operated hemisphere at the beginning of the procedure and maintained for 48 hours postoperatively.

Results: There were no complications with the use of the PTO catheter. No peri-procedural episodes of clinical ischaemia were recorded.

4 of the 6 hemispheres could be considered ischaemic with brain tissue oxygen below 25mmHg.

Auto-regulatory status was examined across all patients using different methodologies including pressure reactivity index (PRx), low-frequency autoregulation index (LAx), and optimum CPP (CPPOpt). These findings will be presented.

Conclusions: We believe this to be the first reported use of direct cerebral oxygenation monitoring in the perioperative period for patients with Moyamoya disease undergoing indirect revascularisation.

In this patient cohort the measurement of ICP and direct brain tissue oxygenation with the Raumedic PTO catheter is safe.

"Normal" brain oxygenation was noted in 2 of the 6 hemispheres. With angiographic follow-up awaited, could this be a predictor of unsuccessful revascularisation?

OP19

The analysis of the embryonic dural sinuses in infants

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Introduction: The primitive tentorial sinus, occipital sinus, and falcine sinus have been usually believed to obtain its adult

pattern or to regress between the fetal stage and adult. Since the anatomy of these three primitive dural sinuses has been seldom studied in infant population and it has been still unknown when these dural sinuses reach its adult condition. Using computed tomography digital subtraction venography (CT-DSV), we analyzed the anatomy of these embryonic dural sinuses in infants.

Materials: We included 13 infants who underwent CT-DSV prior to their neurosurgery from 2015 to 2017. We also included the 35 cases with unruptured cerebral aneurysm as a normal adult control. Three embryonic dural sinuses; primitive tentorial sinus, occipital sinus, and falcine sinus were analyzed both in the infants and the adults using CT-DSV images. We also analyzed the drainage patterns of the superficial middle cerebral vein (SMCV) in the infants and the adults, which is determined by the anastomosis between the primitive tentorial sinus and the cavernous sinus.

Results: 15.4 %, 46.2%, and none of the infants had the primitive tentorial sinus, the occipital sinus, and the falcine sinus, while 10.0%, 8.6%, and 2.9% of the adults had each embryonic dural sinus. The difference in the variation of SMCV draining pattern between the infants and the adults is insignificant. The incidence of occipital sinus in infants was significantly higher than in adults.

Conclusion: Presumably, the connection between the primitive tentorial sinus and the cavernous sinus is established before birth. The occipital sinus is formed at embryonic stage and mostly regresses after infancy. The falcine sinus is usually obliterated prenatally. The knowledge obtained from the current study is fundamentally important for a pediatric interventional neuroradiologist as well as a pediatric neurosurgeon.

OP20

Association of developmental venous anomalies and formation of “de novo” brain cavernous malformations in pediatric patients

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Objective: Vascular brain malformations in children include developmental venous anomalies (DVA), cavernous malformations (CM), arterio-venous malformations (AVM), and capillary teleangiectasia. They differ by location, clinical presentation, management, and treatment. DVAs, as the most frequent group, may be presented in association with CM. In this work we analyze clinical significance of DVAs, especially in coexistence with CMs.

Methods: Among 109 pediatric patients (0-18 years) with diagnosed vascular brain malformation at Children's Hospital Zagreb-Klaiceva in the period from 2005 to 2017, we analyzed clinical presentation, diagnostic procedure, treatment and results of follow up for patients with DVA and CM, with special emphasis on their coexistence.

Results: Among all pediatric patients with diagnosed vascular brain malformations (n=109), 67 patients had DVA (52 supratentorial, 14 infratentorial, 1 supra+infratentorial), 13 AVM (12 supratentorial, 1 pontine), 27 CM (24 supratentorial, 1 brainstem, 2 cerebellar), and 2 pontine capillary teleangiectasia. In patients with DVA (n=67), 3 of them had associated CM in the close proximity. First of them, a12-year-old boy presented with clinically aggressive infratentorial deep cerebellar CM that was successfully operated after rebleeding on the 11th day following first hemorrhage. The second patient had incidental finding and has been followed. In a third patient, a 14-year old boy with stable epilepsy, “de novo” formation of right temporal CM was detected during the MR-follow-up of the DVA located in the right temporal lobe. Three of 67 patients with DVA (4,5%) were associated with CM, and 1 of 67 patients with DVA (1,5%) in our series developed “de novo” CM during the follow-up period.

Conclusion: Pediatric patients with DVA have to be followed by MRI examinations, in order to detect possible “de novo” formation of brain CM at the site of DVA.

OP21

Our eleven years' experience in Radiosurgery treatment of AVMs in the pediatric population

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Aim: Cerebral arteriovenous malformations (AVMs) are relatively rare cerebrovascular malformations characterized by abnormal connections between arteries and veins. The main feature of this type of vascular malformation is a direct blood flow from an artery into a vein, bypassing normal brain tissue. The annual incidence of AVMs is approximately 1.3 per 100,000 in the general population. Approximately 12 to 18% of all AVMs occur in the pediatric population. The main complication associated with AVMs is hemorrhage, especially common in the pediatric population. There are several types of cerebral AVM treatment: surgical excision, embolization, and radiosurgery. The aim of this study is to present our elven years' experience in the radiosurgical treatment of cerebral AVMs presented in the pediatric population.

Methods: This is a retrospective study that presents a single-center experience in radiosurgical treatment of cerebral AVMs in the pediatric population in the period from Februrary 2006 to April 2017. The diagnosis was made by a combination of clinical examination and magnetic resonance imaging (MRI). In a number of cases the diagnosis was also confirmed by digital subtraction angiography (DSA). All patients underwent radiosurgical treatment of intracranial AVM, including a regular follow up by a neurosurgeon. AVM obliteration was confirmed by MRI or by MRI and DSA.

Results: In the period from Februrary 2006 to April 2017, 28 patients with intracranial AVM, under 18 years of age, were treated with radiosurgery at our center. All patients who did not have a follow up of more than two years were excluded from this survey. Sixty-three percent of the patients were male and the average age was 14 years. The majority of intracranial AVMs were located supratentorially and in the left thalamus. The most common symptoms were headache, paresis and visual problems. In two thirds of all patients the AVM had ruptured before radiosurgical treatment. Only 9.1% of all patients were treated surgically, and 27.3% of them underwent embolization. The average AVM volume was 3.5 cubic centimeters, and the average prescription dose was 22.4 Gray. Obliteration was accomplished in 68.2% of all patients and the average obliteration time was 38 months. Regular annual follow-up was conducted until neuroradiological evidence of obliteration was obtained. All patients had postoperative MRI and in 77.3% obliteration was confirmed by DSA. There were no candidates for re-treatment.
Conclusion: Radiosurgery is a noninvasive, safe, and successful type of treatment in pediatric patients with intracranial AVMs, especially in those in whom the location of the AVM is unsuitable for surgical treatment and embolization.

OP22

Strategy of treatment and outcome of pediatric patients with supratentorial arteriovenous malformations – Own experience

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Introduction and Aim: Despite the rapid evolution of therapeutic modalities, the optimal treatment of arteriovenous malformations (AVMs) in children remains controversial.
The aim of the study was to analyze the outcome and discuss the optimal treatment strategy for pediatric supratentorial AVMs.
Methods and Results: We identified and retrospectively reviewed 49 consecutive patients younger than 18 years (median age 12,2 years) treated for supratentorial AVMs with various combinations of techniques in the Department of Pediatric Neurosurgery in Katowice between 2007-2017.
Thirty-seven (75.5%) patients presented with hemorrhage, 13 (26.5%) with seizures and one (2%) was diagnosed incidentally. Spetzler-Martin score distribution was grade 1 in 8 (17%) cases, grade 2 in 17 (37%), grade 3 in 14(30%), grade 4 in 9 (22%) and grade 5 in 1 (2%) case. Supplementary Lawton-Young score was also evaluated to decide about optimal treatment strategy. Nineteen (39%) patients were graded as grade 1, 25 (51%) as grade 2 and 5 (10%) as grade 3.
Twenty-one (43%) patients underwent surgery as primary treatment. The complete excision of AVM was achieved in 10 (48%) of surgically treated cases. The endovascular procedure was primary therapy in 19 (39%) cases. Total obliteration was obtained in 5 (26%), and partial in 10 (53%) of these patients. Radiosurgery was chosen as primary modality only in 6 (12%) cases. Complete obliteration, proved by angiography performed 5 years after treatment was obtained in 3 cases. Eight patients are still under observation. Multimodal treatment was used in 14 (28.5%) patients. Embolization was followed by surgery in 3 cases, AVM remnant after surgery was embolised in 2 and treated by radiotherapy in 3 cases. Five patients were treated by embolization and radiotherapy and one by all modalities. 3 patients were only observed.
Conclusion: Microsurgical resection remains the gold standard for the treatment of pediatric AVMs, but cases with grade 3 AVMs need multimodal treatment.

OP23

Multimodality management of pediatric brain arteriovenous malformations by an integrated neurovascular team: experience from 33 cases

Mario Valencia, Tiffany Li Teng Kok, Claire Toolis, Sanjay Bhate, Vijeya Ganesan, Adam Rennie, Fergus Robertson, Gregory James (London, United Kingdom)

Background: Brain arteriovenous malformations (bAVMs) in children present a particular challenge. Since the senior neurosurgical author joined our institution's combined pediatric neurovascular team (consisting of interventional neuroradiologists, paediatric neurologists and a specialist nurse) in 2015, we have offered microsurgical resection (MR), endovascular treatment (ET) and stereotactic radiosurgery (SRS), individually and in combination, as options for management of this condition. We review our initial experience.
Methods: We reviewed all children referred for management of bAVM to senior author (2015-present) and undertook database and chart review using a proforma.
Results: 33 consecutive patients were identified (17 male), mean age at presentation 10 years (range 2-15). 27 presented with haemorrhage, 3 incidental, 2 neurological symptoms and 1 with seizures. Of the 27 bleeds, 15 were managed non-operatively, 7 with EVD, 4 with craniotomy and 1 with decompressive craniectomy. 31 bAVMs were supratentorial (28 lobar, 1 chiasmatic, 1 thalamic and 1 corpus callosum) and 2 infratentorial (1 brainstem, 1 cerebellar). Spetzler Martin grades were I (4 patients), II (7), III (14), IV (6) and V (2). 4 bAVMs were managed conservatively, 3 had MR alone, 1 combined MR/ET, 3 ET alone, 3 combined ET/SRS and 19 SRS alone. 7 have angiographic cure (all MR and ET cases), in all SRS cases results are awaited. Outcome (mRS) was 0 (17 patients), 1 (8), 2 (4), 3 (2) and 4 (2). There was no mortality. There were 3 treatment-related complications: transient visual disturbance (2 patients, 1 MR, 1 ET); seizures (1 SRS patient) and hemianopia (1 ET/SRS patient).

Conclusion: A neurovascular team which can offer all 3 options for paediatric bAVM allows tailored treatment with acceptable outcomes. Our series has a preponderance of SRS which likely reflects our status as a tertiary referral centre for this treatment modality. Working in a multidisciplinary team allows a balanced discussion of bAVM cases.

Abstract session 9: New technologies

OP24

Robot-assisted stereotactic brainstem biopsy in children: prospective cohort study

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*Equal contribution

Introduction: Tumours located within the brainstem comprise approximately a tenth of all paediatric brain tumours. Surgical biopsy of these tumours is technically challenging and has historically been associated with considerable risk. To this end, robot-assisted surgery theoretically allows for increased accuracy and precision. In this study we report our experience using the Neuromate robot (Renishaw, Gloucestershire, UK) to perform robot-assisted stereotactic biopsy in children with tumours located within the brainstem.
Methods: An uncontrolled prospective cohort study was performed (Phase II) according to the IDEAL model for safe surgical innovation. The study was conducted at Great Ormond Street Hospital, which acts as regional referral centre for paediatric tumours in North London. All cases were recorded on a prospectively maintained database. The database was searched over a two-year period between the 1stDecember 2015 and the 31stNovember 2017 to identify all children with brainstem tumours that underwent robot-assisted stereotactic brain biopsy. When accessible, the post-operative MRI scans and pre-operative plans were compared to assess the Target Point Localisation Error (TPLE). Adverse events were recorded prospectively according whether they resulted in increased hospital stay, caused neurological injury, or lead to death.
Results: In all, 11 consecutive children were identified with brain tumours located within the brainstem and related structures. In 10/11 cases specimens were diagnostic; in the remaining case a further biopsy was successful. The most frequent pathology was DIPG (7/15). Seven patients underwent an early post-operative volumetric MRI; the calculated median TPLE was 2.7mm (range 0.5-4.2 mm). There were no surgical complications noted.
Conclusions: Robot-assisted stereotactic biopsy in children appears to be feasible and safe. Comparative studies are warranted to further assess the technique.

OP25

First clinical applications of a high-definition 3D exoscope in Pediatric Neurosurgery

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Objective: Ideal visualization tools in micro-neurosurgery should provide magnification, illumination, wide fields of view, ergonomics and unobstructed access to the surgical field. The operative microscope was the predominant innovation in modern neurosurgery. Recently, a high-definition 3D exoscope was developed. We describe our first experience in pediatric neurosurgery.
Methods: The VITOM® 3D exoscope (Karl Storz GmbH, Tuttlingen, Germany) was used during a trial phase in pediatric micro-neurosurgical operations, in direct comparison to an OPMI® PENTERO® operative microscope (Carl Zeiss AG, Jena, Germany). Experiences were evaluated with five-level Likert items regarding ease of preparation, image definition, magnification, illumination, field of view, ergonomics, accessibility of the surgical field and general user friendliness.
Results: Three operations were performed: supratentorial open biopsy in supine, infratentorial brain tumor resection in park bench and myelomeningocele closure in prone position. While preparation and image definition were rated equal for microscope and exoscope, the microscope's field of view, illumination and user friendliness were considered superior, whereas advantages of the exoscope were seen in ergonomics and accessibility of the surgical field. No complications attributed to visualization mode occurred.
Conclusion: In our experience the VITOM® 3D exoscope is an innovative visualization tool with advantages over the microscope in ergonomics and accessibility of the surgical field. However, improvements were deemed necessary with regard to field of view, illumination and user friendliness. While the debate of a "perfect" visualization modality is influenced by personal preference, this novel visualization device has the potential to become a valuable tool in the neurosurgeon's armamentarium.

OP26

Intraoperative navigated ultrasound in Pediatric Neurosurgery: an essential tool with ductile applications

Paolo Frassanito, Gianpiero Tamburrini, Luca Massimi, Federico Bianchi, Elif Başaran Gündoğdu, Massimo Caldarelli (Rome, Italy; Bursa, Turkey)

Background: Technologic improvements have refined and expanded the scope and application of intraoperative US technology (IOUS). Strategies to maximize its efficacy and overcome the various limitations have evolved, in particular the fusion with preoperative MRI to allow neuronavigation. On

these grounds, the role of navigated IOUS (n-IOUS) deserves attention to answer the question "tool or toy?".

Methods: Cranial procedures performed with the aid of n-IOUS (Esaote) in the last 2 years in our institution were collected.

Results: We performed 20 tumor surgical procedures (group I) and 15 endoscopic procedures (group II). In the group I, n-IOUS allowed to locate the lesion accurately in all the cases. Main advantages are the possibility to correct the brain shift and the possibility to place a catheter inside deep-seated lesions with real time navigated imaging. Moreover, n-IOUS allowed to evaluate the extent of resection reliably. Indeed, the surgical plan was changed in about 30% of cases, as n-IOUS showed a residual tumor that was left behind. Concordance of n-IOUS to postoperative MRI was total. In the group II, n-IOUS allowed reliable neuronavigation with real-time IOUS in the cases of multiloculated hydrocephalus with open fontanelle. The use of n-IOUS increased the operative time by 30 minutes in average, mainly due to the registration procedure while the brain shift correction required in average 2-3 minutes. No complication related to the use of n-IOUS was recorded.

Conclusions: The n-IOUS is definitely an essential tool with ductile applications that may improve the neurosurgical practice. Compared to other intraoperative imaging modalities, advantages claimed by n-IOUS are lower cost, higher readiness, with reduction of intraoperative time, real-time imaging, and repeatability. Hybrid system coupled with neuronavigation allows to overcome the limits of IOUS and neuronavigation, since facilitates the interpretation of IOUS on one hand and allows to correct the brain shift on the other.

OP27

Non-invasive assessment of intracranial pressure in pediatric patients using transorbital ultrasound measurement of the Optic Nerve Sheath Diameter (ONSD)

Susanne R. Kerscher, Felix Neunhoeffer, Konstantin Hockel, Martin U. Schuhmann (Tübingen, Germany)

Introduction: Ultrasound-based measurement of the optic nerve sheath diameter (ONSD) has been proven to be a reliable method to assess increased ICP in children of any age group. This study aims to investigate 1) the relationship between ONSD and invasively measured ICP and 2) if ONSD values can indicate a clinical situation with potentially increased ICP in pediatric neurosurgical patients.

Methods: 140 patients aged newborn to 18 years (median 6 years) were investigated. Diagnoses included hydrocephalus (52%), tumor (13%), craniosynostosis (14%) and other intracranial pathologies (21%). Dependent on diagnosis, the investigation was performed in children awake or sedated, pre- and post-operatively, or longitudinally over a period of time, respectively. In 46 patients the mean binocular ONSD was compared to invasively measured ICP.

Results: 46 patients underwent ONSD in parallel to invasive measurement of ICP.The correlation between ONSD and ICP was strong with $r= 0.62$, $p< 0.01$.

77 patients with undoubted symptoms of increased ICP showed increased ONSD values before surgery (mean 5.89 ± 0.82mm) which decreased after surgery (mean 5.14 ± 0.8 mm) ($p<0.001$). Of 89 patients with mild to moderate indices of raised ICP, 66 showed normal ONSD values (mean 4.78 ± 0.38 mm) and were observed only. None needed an intervention in follow-up. In the remaining 23 patients higher ONSD values were found (mean 5.4 ± 0.29 mm). In those invasive ICP measurement was recommended. The ONSD cut-off value with the highest diagnostic value for detecting clinically relevant situations of increased ICP was 5.3 mm (sensitivity of 83%, specificity 94%, OR 47.5, AUROC 0.901).

Conclusion: Transorbital ultrasound measurement of ONSD is a reliable first-line screening tool before further diagnostic steps are initiated. Unnecessary CT imaging as well as delay of treatment can be avoided after a first line orientation using ONSD with regard to ICP.

OP28

Increased total retinal thickness on OCT: a precursor for intracranial hypertension?

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Introduction: One of the challenges in the treatment of patients with craniosynostosis is the detection of Intracranial Hypertension (ICH). An objective and non-invasive tool to assess ICH in craniosynostosis is needed. Optical Coherence Tomography (OCT) is a promising non-invasive screening method that accurately detects changes in the retina. The aim of this study was to evaluate Total Retinal Thickness (TRT) measured on OCT in scaphocephaly patients and to investigate a possible association with skull growth arrest.

Methods: We performed a prospective cohort study at the Dutch Craniofacial Centre in Rotterdam. We included 67 patients with non-syndromic scaphocephaly and 66 control subjects without known optic nerve disease aged 3-13 years. OCT and fundoscopy were obtained in all patients. In scaphocephaly patients, occipitofrontal head circumference was measured as part of the regular treatment protocol. Skull growth arrest was defined as downward deflection or lack of change in occipitofrontal head circumference trajectory. TRT of scaphocephaly patients was evaluated and compared with the normative references derived from the healthy controls.

Results: When compared to the normative references, 8 patients showed increased TRT. Of these patients, 2 had

papilledema on fundoscopy and 5 hadskull growth arrest. An increased TRT was associated with papilledema on fundoscopy and skull growth arrest ($p<0.01$).

Conclusion: An increase in TRT is associated with papilledema and skull growth arrest. This study determines a cluster of patients with increased TRT and symptoms of ICH but with normal fundoscopy. In these patients, increased TRT might be a precursor of ICH compared to papilledema on fundoscopy.

OP29

A multidisciplinary and innovative separation of craniopagus twins

Jesse Taylor, Peter Madsen, Benjamin Kennedy, Phillip Storm, Gregory Heuer (Philadelphia PA, USA)

Introduction: The craniopagus variant of conjoined twins is a rare congenital anomaly. Twins with total fusion where significant vascularity is shared, especially the superior sagittal sinus, have a high morbidity and mortality rate with separation despite advances in surgical technique and perioperative critical care. We report a case of craniopagus twins for whom computer-aided design and modeling (CAD/CAM) surgical planning, novel cranial distractor devices, and intra-operative navigation were used to aid in the planning and completion of a successful separation surgery.

Methods: The craniopagus twins where diagnosed on prenatal ultrasound. Further postnatal imaging demonstrated Total Angular craniopagus twins by the Goodrich classification, with faces in the same direction and minimal fusion of brain parenchyma. The separation was a planned staged approach, first utilizing distraction-contraction to convert the twins to a vertical configuration with less shared surface area. Thereafter, we performed CAD/CAM planned tissue expansion and surgical navigation during the actual separation.

Results: A novel CAD/CAM external distraction device was employed after linear strip craniectomy at 3 months of age. At 4 months of age an external constriction device aided further remodeling to minimize their shared surface area and provide a more favorable anatomy for separation, changing the angle between the twins and improving access to the combined dural sinuses. At 7 months, tissue expanders were placed and the twins were successfully separated at 10 months of age. They were discharged home at 16 months of age, both interactive and with no significant focal neurologic deficits.

Conclusion: We report a successful, multidisciplinary separation of craniopagus twins using CAD/CAM surgical planning, novel cranial distractor devices, and intra-operative navigation. These surgical innovations allowed for early, safe separation which harnessed the regenerative potential of the infant head and neck.

Abstract session 10: Epilepsy

OP30

Dysembryoplastic neuroepithelial tumors – Is the lesionectomy enough for seizures control?

Adrian Iliescu, Catalin Gheorghe Pascal, Iulia Elisabeta Balalau Vapor, Radu Eugen Rizea, Irina Petruta Tudose, Alexandru Tascu (Bucharest, Romania)

Objective: Slow-growing dysembryoplastic neuroepithelial tumors (DNET) are one of the organic causes of seizures in children. Lesionectomy has a good chance to reduce or even cure the seizures. The authors present a series of 21 pediatric cases who underwent surgery for DNET.

Methods: The review included 21 patients operated for DNET between 2007 and 2015. The mean age was 9.8 years old (range 4.5 to 15.1 years). All patients were extensively tested for the focal source of the seizures. Thirteen (61.9%) of the lesions were located in the frontal lobe and 8 (38.0%) were located in the temporal lobe. All children have been submitted to lesionectomyusing neuronavigation. The mean follow-up time was 6 years (range 2.6 to 9.3 years). All the patients had a thoroughly psychological assessment.

Results: Postoperatively, 11 patients (52.3%) were seizures free (Engel Class I A), 6 patients (28.5%) were free of seizures for more than 2 years (Engel Class I C), 3 patients (14.2%) had worthwhile seizure reduction (Engel Class III A) and one patient (4.7%) got worse after surgery (Engel Class IV C). To the four patients included in Engel Class III A and Engel Class IV C which had there lesions in the temporal lobe a second surgery was offered with more extensive resection. Three patients improved and they could be included in Engel Class II B. One patient didn't benefit of surgery.

Conclusions: Lesionectomy is certainly useful in carefully selected cases, but in the presence of diffuse lesions and/or associated anomalies (cortical dysplasia) a more extensive surgery should be offered to the patients.

OP31

Language outcome after left hemispherotomy in children: a case series

Helio Rubens Machado, Joceli R. Silva, Marcelo Volpon, Ursula Thomé, Americo C. Sakamoto, Sara Rosset, Geisa Angelis, Ana Paula A. Hamad (Ribeirao Preto, Brazil)

Introduction: Language deficits are always a concern in epilepsy surgery when operating in the dominant hemisphere. The exact timing for localization and lateralization of language areas is difficult to be established pre-operatively. We studied a series of children with refractory epilepsy treated with left hemispherotomy at our Institution. Our goal was to evaluate the impact of surgery on language outcome.

Subjects and Methods: From 1994 to 2012, 61 children underwent left hemispherotomy. We selected only cases operated on between the ages of 6 and 18 years (33 cases). Only 15 cases had a complete pre and post-operative neuropsychological evaluation for language. Etiology in these cases included Rasmussen encephalitis (6 cases), vascular insults (6 cases) and Cortical Dysplasia (3 cases). Normal milestones were achieved in 10 cases before surgery and 12 children were left-handed. All patients were submitted to a comprehensive clinical and radiological workup prior to the surgical procedure and were operated on using the same technique. Neuropsychological and language assessments were performed pre and postoperatively at variable time frames (from 6 months to 8 years after surgery, mean 3.5 years).
Results: Post-operatively, 80% of the cases (12 children) were seizure free and only 3 children had residual seizures. Language remained unchanged in comparison with the preoperative status in 14 (93.3%) children. 1 child experienced improvement in his language skills.
Conclusions: In children, language is a very complex phenomenon depending on several mechanisms of brain maturation before it can be definitely localized and lateralized. Atypical representation is common and relocation is possible either intra or interhemispheric or even contralateral or bilateral. Children should be thoroughly studied and surgery should not be a priori denied as prognosis is good, especially when surgical treatment is indicated early.

OP32

Peri-insular anterior quadrantotomy: a new concept for frontal lobe epilepsy

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Introduction: Refractory frontal lobe epilepsy is classically treated with frontal lobectomy.
Epilepsy surgery greatly evolved during the last decades in favor of disconnective techniques and we would like to describe a new technique of selective disconnection of the frontal lobe, to perform in cases of refractory epilepsy due to epileptogenic foci involving one frontal lobe (anterior to the motor cortex), in the presence of preserved motor function.
Methods: The representative case of a 9-year-old boy presenting with refractory epilepsy due to epileptogenic foci localized to the right frontal lobe is reported.
The surgical techniques and the clinical outcome are detailed.
Results: Our case index showed a focal cortical dysplasia of the right frontal lobe at cerebral MRI with an optimal clinical, electrophysiological and radiological correlation.
A peri-insular anterior quadrant disconnection (quadrantotomy) was performed. The postoperative period was uneventful, and the patient was in Engels's seizure outcome class I at 3 years of follow-up. A significant cognitive gain was observed during follow up.
This disconnective procedure may be divided into four steps: the supra-insular window, the anterior callosotomy, the intra-frontal and the fronto-basal disconnection. The functional neuro-anatomy was detailed for each step of the surgery.
The cortical and subcortical electrophysiological mapping is an important step in the identification of eloquent cortices and intact neural pathways to guide this disconnective procedure.
Conclusion: The peri-insular anterior quadrantotomy may represent an efficient technique to treat refractory epilepsy when epileptogenic foci are localized to one frontal lobe while preserving residual motor functions. Complications associated with large brain resections may thus be avoided in this pediatric population.

OP33

Surgical strategies for the treatment of epilepsy related to Sturge-Weber syndrome in pediatric patients

Marcelo Volpon Santos, Ursula Thome Costa, Ana Paula Andrade Hamad, Americo Ceiki Sakamoto, Helio Rubens Machado (Ribeirão Preto, Brazil)

Introduction: Sturge-Weber syndrome (SWS), a neurocutaneous disorder characterized by facial and leptomeningeal angiomas, is frequently associated with epilepsy in children (75-90% of cases), which can be localized or widespread within a whole cerebral hemisphere. Since medical therapy is frequently insufficient for seizure control, many patients require surgical treatment that must be tailored according to the clinical and radiological features of each patient to achieve good results. We describe herein the surgical strategies adopted in our pediatric case series of SWS related epilepsy.
Methods: From 1997 to 1996, 11 children (2 boys and 10 girls; mean age: 9 years) diagnosed with SWS underwent 12 operations for epilepsy treatment at our institution. Focal resections were performed in 4 (33,3%), whereas posterior quadrantic disconnections (PQD) were done in 4 (33,3%) and hemispherotomies in 4 (33,3%).
Results: 6 patients (50%) were seizure free and 5 had significant improvement (Engel Class III) at their last assessment. Mean follow-up was 10,3 years (range 2-20 years). There were two reoperations: one girl was initially treated with a focal frontal resection but required a posterior quadrantic disconnection 9 months after the first operation, and became seizure free, and another patient had a redo hemispherotomy

but his post-op seizure status remained unchanged. Only 2 patients (25%) in the PQD or hemispherotomy subgroups did not achieve seizure freedom, whilst 2 of the 4 patients who underwent focal resections still experienced seizures after surgery (50%).

Conclusions: Refractory epilepsy associated with SWS is common and can be satisfactorily controlled with tailored surgery. Extended resections / disconnections provide better seizure outcomes; therefore, surgical strategies should be individualized according to a thorough preoperative workup.

OP34

Prediction of epilepsy status and treatment response in Rett syndrome: a combined support vector machine – Neural network method

Conor Keogh, Giorgio Pini, Daniela Tropea (Dublin, Ireland; Camaiore, Italy)

Introduction: Rett Syndrome is a neurodevelopmental disorder caused by genetic alterations in synaptic regulation and is frequently associated with treatment-resistant epilepsy. Vagus nerve stimulation has demonstrated significant promise in reducing seizure burden in these patients, though identification of surgical candidates remains challenging. The ability to predict epilepsy status and treatment responsiveness therefore offers a valuable method of risk stratification, identifying those in need of greater monitoring and intervention.

Methods: We collected EEG data from a large (n=42) cohort of patients with this rare disorder. Patients were divided based on epilepsy status (no epilepsy: n=18, epilepsy: n=24). The epilepsy group was subdivided based on response to medical treatment (treatment responsive: n=16, treatment resistant: n=8).

Measures of spectral power distribution and inter-electrode coherence were compared between groups to characterise the electrophysiological features associated with epilepsy and treatment resistance. Machine learning classifiers were trained and validated using a 10-fold cross validation approach to assess whether these features predicted epilepsy status and treatment response in a new patient.

Results: We identified asymmetry of power distribution as a marker of epilepsy. A support vector machine trained on the identified asymmetry measures predicted epilepsy status of test data with 69% accuracy (PPV 66.7%, NPV 72.7%).

Treatment responsiveness was associated with differences in the first principal component of inter-electrode coherences, indicating that abnormalities of network-level measures were associated with treatment resistance. A neural network classifier trained using inter-electrode coherence measures predicted treatment resistance with 87.5% accuracy (PPV 77.8%, NPV 93.3%).

Conclusion: We demonstrate the potential predictive value of machine learning approaches using EEG features for epilepsy status and treatment resistance. This provides a potentially clinically valuable methodology for stratification of patients and prediction of those likely to require surgical intervention for epilepsy control following failure of medical therapy.

Abstract session 11: Functional diagnostics

OP35

Bilateral Deep Brain Stimulation (DBS) of Gpi for secondary dystonia. A series of fourteen patients with different etiologies

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Introduction: The efficacy of bilateral DBS of Gpi is encouraging but not yet validated given the small series of patients. Aim of this study is reporting the outcome of fourteen patients submitted to bilateral DBS of Gpi for secondary dystonia with different etiology.

Materials and Methods: Since 2009 to 2017 fourteen patients (mean age 13,5 years-old; range 4,5-30 yrs) with secondary generalized and multifocal dystonia received bilateral stereotactic electrode implantation of Gpi nucleus for DBS. Main etiology was ICP (9/14) followed by metabolic disease (9/14); one patient developed secondary dystonia twenty years after cranial surgery for craniopharyngioma. Three patients were admitted in ICU because of status dystonicus due to metabolic disease. Average history of disease was 7 years (range 1 mos-18 years).

Results: Monopolar and high-frequency bilateral stimulationof Gpi was started one month after surgery and gradually programmed according to the suggested parameters (3.5 V, 80 usec, 130 Hz). Mean folow-up was 40 months (3-74 months). No side effects were reported during the stimulation. Surgical outcomes were evaluated according to the Burke-Fahn-Marsden Rating Score (BFMDRS). At last follow-up control, BFMDRS improved in about 35% of case, especially in metabolic disease (40%) as compared to ICP (30,1%). Besides objective improvement documented by BFMDRS, all patients but one referred a subjective improvement and a less perception of disability. Two out of three patients with status dystonicus recovered after DBS and left ICU; one died because of multiorgan failure despite surgery.

Conclusions: This series provides new evidence about the efficacy of bilateral DBS-Gpi as a treatment of secondary dystonia. Surgical outcomes should be evaluated not only

with BFMDRS but especially according to patients' perceived quality of life after surgery. Status dystonicus may be successfully treated too by bilateral DBS of Gpi.

OP36

Deep brain stimulation in pediatric patients: institutional experience

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Introduction: DBS is initially used for treatment of essential tremor and Parkinson's disease in adults. In 1996, a child with severe life-threatening dystonia was offered DBS to the internal globus pallidus (Gpi) with lasting efficacy at 20 years. Since that time, increasing number of children benefited from DBS.
Methods: We retrospectively evaluated our database of patients who underwent DBS from 2011 to 2017. All patients ≤17 years of age at the time of implantation of DBS were included in this series. Subjective Benefit Rating Scale (SBRS) is used to evaluate clinical outcome.
Results: Between May 2014 and October 2017, 11 children underwent DBS procedure in our institution. 6 of them were female and 5 of them were male. Mean age at surgery was 11,8 ± 4,06 years (range 5-17 years). In our series, 4 patients had primary dystonia (36,3%), 2 patients had secondary dystonia (18,1%), 2 patients had juvenile parkinsonism (18,1%), 2 patients had Tourette Syndrome (18,1%) and 1 patient had cerebral palsy (9%). Two juvenile parkinsonism patients underwent bilateral STN DBS while other 9 patients underwent bilateral Gpi DBS. SBRS scores were 1,75±0,5 for patients with primary dystonia, 3±0 for patients with juvenile parkinsonism, 2,5±0,7 for patients with Tourette Syndrome, 2,5±0,7 for patients with secondary dystonia and 1 for patient with cerebral palsy.
None of the patients experienced any intracerebral hemorrhage or other serious adverse neurological effect related to the DBS. Wound complications occurred in two patients.
Conclusion: There are many references in literature that support DBS as a treatment option for pediatric patients with medically refractory neurological disorders. DBS has replaced ablative procedures as a treatment of choice not only for adult patients, but also for pediatric patients. Wound related complications still remain the most common problem in pediatric patients. Development of smaller and more flexible hardware will improve quality of children's life and minimize wound related complications in the future.

OP37

Lower limb goniometry as a reliable tool for measuring outcome after spasticity treatment with selective dorsal rhizotomy

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Introduction: Selective Dorsal Rhizotomy (SDR) is an undisputable treatment option for spasticity caused by cerebral palsy (CP). Outcome after SDR is usually assessed by subjective scales and quality of life scores, but also by goniometry, which is the measurement of the total motion of a specific joint. It provides a quantitative analysis of SDR efficacy, and is commonly used on rehabilitation follow-up. This study aimed to evaluate goniometric parameters of patients with CP that have undergone SDR in our service in order to assess post-operative outcome.
Methods: The authors reviewed the medical charts of pediatric patients with CP associated spasticity treated by SDR between June 2010 and July 2017. Demographic data, number of spastic limbs, and pre-operative GMFCS levels were collected, along with goniometry of hip abduction, bilateral popliteal, and foot dorsiflexion angles prior to surgery and 6 months post-operatively.
Results: 35 patients were included (19 males and 16 females; mean age 8,7 years). 13 (37,1%) had lower limb bilateral paresis, 10 (28,5%) triparesis and 14 (40%) tetraparesis. Preoperative GMFCS distribution was: 1 level II, 7 level III, 16 level IV and 10 level V. Improvement of the hip abduction angle was seen in 84,6%, bilateral popliteal angle in in 69,2%, and foot dorsiflexion angle in 61,5%.
Conclusions: Goniometry is a reliable method for quantitative evaluation of the benefits of SDR, demonstrating its efficacy and correlating with other scores as a reliable outcome assessment tool. It is simple and therefore might be performed by all clinical members of the rehabilitation team involved in care of spastic patients.

OP38

Progress in motor function in children with cerebral palsy who undergo single-level selective dorsal rhizotomy: a prospective cohort study

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Background: Selectivedorsal rhizotomy (SDR) is an established treatment for spasticity in children with spastic cerebral palsy. Spasticity is permanently reduced immediately after SDR.In well-selected children, SDR leads to an improvement in motor function and independence. This study aims to define the improvement in motor function over the first two years after SDR.
Methods: Prospectively collected data from the Great Ormond Street Hospital SDR Database (2013-2017) were

analysed. All patients underwent single-level SDR with intraoperative neurophysiological monitoring. The gross motor function measure (GMFM-88) was evaluated preoperatively, at 6, 12 and 24 months after surgery. The patients were stratified by their initial gross motor function classification system (GMFCS). We investigated GMFM domains individually, using the paired t test, as well as in a multivariate linear mixed-effects model (LMEM) encompassing multiple domains, with change in the GMFM score over time as the primary outcome.

Results: 91 children were included; mean age at surgery was 6.2 years. In GMFCS II children (n = 28) the GMFM-88 improved from 87.3% at baseline to 90.8%($p<0.001$), 93.6%($p<0.001$), and 91.1%($p=0.06$) at 6, 12 and 24 months respectively, with maximal improvement in the E domain (walking, running, jumping) (66.0%(baseline) to 81.3%(24 months), $p<0.0001$). GMFCS level III (n=46) children improved from 64.5% at baseline to 69.0%($p<0.0001$), 71.6%($p<0.0001$) and 72.5%($p<0.0001$) at 6, 12 and 24 months respectively, with maximal improvement in the D (standing) domain (25.6(baseline) to 45.5 (24 months), $p<0.0001$). GMFCS level IV (n=17) children showed improvement from 41.6% at baseline to 46.9% ($p=0.06$), 48.3%($p<0.001$), and 47.0% ($p=0.01$) at 6, 12 and 24 months respectively, with maximal improvement in the C (crawling, kneeling) domain (39.1%(baseline) to 53.5%(12 months), $p=0.003$)

Conclusions: In our LMEMage or gender did not correlate with outcome. This series confirms significant time-dependent improvement in GMFM-88 in children with spastic diplegia who undergo single-level SDR.

OP39

Obstetric brachial palsy – Endoscopic neurolysis and decompression

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Objective: To evaluate the possibility of surgical treatment of plexopathy with endoscopic assistance.

Methods: From 2015 to 2017, 17 patients (age range, 1 months-2 years) with brachial palsy underwent endoscopic neurolysis and decompression. The technique consisted of using complex endoscopic equipment and instruments through axillaris along the neuro-vascular bundle to the primary trunks. Elaborate electrophysiological monitoring was used. We have assessed the effectiveness of surgical treatment used 3 criteria. There are flexion of the forearm, total abduction and using the arm in games. We used British Medical Research Council grading system (BMRC) for the evaluation of the movement and special scale (0-3 points) for evaluation of using the arm in games. Follow-up was 12±0,7 months.

Results: Flexion of the forearm before operation was 1,35 ± 0,36, after operation 3,41± 0,30.

	BEFORE OPERATION	AFTER, 12 MONTHS
M0	**8**	**0**
M1	**1**	**2**
M2	**4**	**1**
M3	**2**	**5**
M4	**2**	**6**
M5	**0**	**3**

Table 1

Total abduction before operation was 0,59 ± 0,27, after operation 3,18 ± 0,34.

	BEFORE OPERATION	AFTER, 12 MONTHS
M0	**13**	**1**
M1	**0**	**0**
M2	**2**	**5**
M3	**2**	**4**
M4	**0**	**3**
M5	**0**	**5**

Table 2

Using the arm in games before operation was 0,47 ± 0,19, after operation 2,06 ± 0,25.

	BEFORE OPERATION	AFTER, 12 MONTHS
NOT USING	**12**	**2**
PARTIALLY USING, BUT DOESN'T KEEP TOY	**2**	**2**
PARTIALLY USING, KEEPS TOY	**3**	**6**
FULL	**0**	**7**

Table 3

Conclusion: Endoscopic neurolysis and decompression brachial plexus is effective treatment obstetric brachial palsy.

Abstract session 12: Dysraphism I

OP40

Does intraoperative measurement of BCR (Blubocavernosus reflex) and sphincter MEP correlate with urological outcome in complex lipoma surgery?

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Introduction: Sphincter MEP and BCR have been incorporated into spinal cord untethering surgery for complex lipomas. The purpose of this study was to assess whether intraoperative electrophysiological findings correlate with urological outcome.
Methods: Between 2015 and 2017, 31 children with complex lipomas underwent total or near total lipoma resection. Indications for surgery were prophylactic (15), abnormal urology profile (9), motor dysfunction (4) and 2 with motor dysfunction and urological concerns. Sphincter MEP and BCR were monitored in all cases. A grading score for BCR was developed to allow intra and inter patient comparison. All children in this study had urological evaluations at least 3 months following surgery.
Results: 19 children had good baseline and end of procedure BCR, the majority of these children ended up dry (11) or where still in nappies (4) at the time point of the follow up. Only 4 children with good BCR commenced or continued CIC post operatively. 12 children had poor BCR at the end of the procedure (4 had been good at baseline but deteriorated at the end of surgery). In this group only 1 attained continence, 9 children commenced or continued CIC.
Conclusions: Urinary continence correlates with good BCR at the end of surgery. If the BCR is poor at baseline or is lost during surgery there is an increased likelihood for CIC. These results suggest that BCR might provide an objective assessment of urological prognosis in initial evaluation of children with complex dysraphism.

OP41

Distal spinal cord and neural placode stimulation in newborns with myelomeningocele

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Introduction: Monopolar stimulation of the distal spinal cord and neural placode was previously reported in lumbosacral myelomeningocele and it was suggested that monopolar stimulation of the lateral spinal cord does not cause any contraction of functional lower extremity muscles.
Patients and Methods: We planned an electrophysiological study in 10 newborns with lumbosacral myelomeningoceles. During the surgery, bipolar stimulation was performed from the distal spinal cord and neural placode respectively and we tried to record muscles responses from the lower extremities.
Results: We received responses from the bipolar stimulation of the neural placode just close to the exit of the roots in all patients. But we could receive responses with distal spinal cord stimulation in only 3 patients and the amplitude of the responses was very low. We could not obtain responses in 7 patients during surgery.
Conclusion: This result showed that the primitive neural placode has low electrophysiological capacity to transfer the stimulations to the lower extremity muscles. This may be secondary to the low myelination of the primitive neural placode.

OP42

Pro-inflammatory, pro-apoptotic and neuroprotective mechanisms in spinal dysraphism: important factors to be considered in therapeutically approaches

Friederike Knerlich-Lukoschus, Gesa Cohrs (Sankt Augustin, Germany; Kiel, Germany)

Objective: Despite the complexity of spinal dysraphism (SD) and its comorbidities, the primary therapy is limited to surgical reconstruction and, in case myelomeningocele (mmc), coverage of the spinal defect and treatment of associated comorbidities. Tertiary molecular cascades which are induced by the "second hit" to the placodes, after initial impaired neurulation ("first hit"), offer targets for adjuvant therapies and potentially influence the pathology of associated comorbidities (like mmc-associated hydrocephalus). Different cytokines and their potential involvement of tertiary lesion cascades in open and closed spinal dysraphism will be presented.
Methods: Human material collected at post-natal repair surgeries for mmc and spinal lipomyelomenincoceles (LMMC), plus material from untethering surgeries (TC) were investigated for expression of inflammatory cellular changes (expression of vimentin, GFAP, BLBP, CD11b, CD68 etc.). Normal fetal and adult sc specimens served as controls. Pro-inflammatory cytokines like TNFalpha, IL-1beta, and its receptors were analyzed by immunohistochemistry (IHC), real-time RT-PCR, double-labeling. Associated pro-apoptotic and hypoxic mediators were analyzed similarly. Further, erythropoietin and its receptor were investigated as neuroprotective cytokine counterpart.
Results: In all SD and TC specimens, IL-1b/IL1-R1 and TNFa/TNF-R1 were expressed on higher mRNA and IHC level compared to normal sc controls. In MMC and TC mRNA level were slightly higher compared to LMMC. Cytokines were co-expressed with GFAP, inflammatory cells and neuronal elements. In parallel, pro-apoptotoic mediators like cleaved caspase-3 and morphological signs for apotptoic cell death (i.e. TUNEL labeled cells) became apparent in neuroepithelial tissues. As cellular component, inflammatory cells and activated astroglial cells became apparent in both dysraphic- and TC-groups. Also, hints for a hypoxic component were found by induction of HIF1/2a. As endogenous neuroprotective cytokine effector/receptor-pair, eryothorpoietin and especially EPO-R was found on higher expression levels especially in mmc specimens.
Conclusions: Molecular tertiary lesion cascades, specifically induction of pro-inflammatory cytokines along with the expression of associated apoptotic and hypoxic mediators, are detectible in neuroepithelia of open as well as of closed SD, and consecutive TC-derived specimens. At least inflammation is therefore a potentially important factor that might determine

the outcome, clinical course and comorbidities of SD. The hypothesis of inflammatory processes as unifying concept of SD has to be verified further by investigating the expression time-course during the pre-natal fetal course – preliminary data on these findings will be presented in a consecutive contribution of our group.

OP43

Water-tightness increases reversal of hindbrain herniation and improves neuromotor function in the lamb model for spina bifida repair

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Aim: Experimental animal and clinical research have demonstrated that prenatal anatomical 2 to 3-layer fetal repair of open spina bifida (SB) improves neonatal outcome. The reason why only 36 to 86% of patients have complete reversal of hindbrain herniation (HH) and 44 to 69% have improvement of leg neuromotor function, and others not, remains uncertain. One potential reason is incomplete closure, leading to persistent leakage of cerebrospinal fluid (CSF) and direct damage to the spinal cord. We aimed to experimentally test this hypothesis and determined the consequences of watertightness of the repair in the SB lamb model.

Methods: 75-day lambs (term=145d) underwent standardized induction of lumbar SB consisting of a 4.2x4.2cm circular skin resection, and a laminectomy, durectomy and myelotomy over 5 vertebral levels or more (L1-6). They were assigned to 3 groups: unrepaired SB (SB group) or standardized 2-layer repair at 100d using Duragen® patch and skin closure, which was either watertight or was not. An intra-operative fluorescein test was performed after skin closure completion to determine whether the closure was watertight (watertight group) or not (non-watertight group). Lambs were delivered at term (mean=143d). Survival after the repair until term C-section was the safety outcome measure. At 1-2d after birth, the primary efficacy outcome variable (HH on Magnetic Resonance Imaging [MRI]) and secondary efficacy outcomes (skin closure, CSF leakage [blotting paper test], hindlimb neuromotor function [joint score & motor grade], ventriculomegaly on MRI [parietal diameter >2mm on axial plane], Motor/Sensory Evoked Potential [MEP/SEP] measurements) was recorded and assessed by 2 independent assessors blinded to the treatment groups. Statistical analysis compared the repaired to the unrepaired SB fetuses.

Results: 38 fetuses were included, and SB surgically induced (n=10 SB, n=9 watertight and n=19 non-watertight). Survival was comparable between the 3 groups, i.e. 70% (7/10), 78% (7/9) and 64% (7/11) respectively. Watertight repair induced complete (100%) reversal of HH, better neuromotor function and presence of bilateral hindlimb MEPs (table 1). Despite 86% of HH reversal, non-watertight repair was associated with poor hindlimb neuromotor function and low presence of bilateral hindlimb MEPs. All lambs that underwent repair had at birth complete skin closure and a lower rate of clinically obvious CSF leakage than SB lambs. They also had ventriculomegaly and absence of SEPs on hindlimbs (Table 1).

Groups	**Unrepaired SB**	**Watertight SB repair**		**Non-watertight SB repair**	
Number	10	9	*p value*	19	*p value*
Primary safety outcome					
Survival at birth	70% (7/10)	78% (7/9)	*p=0.708*	64% (7/11)	*p=0.763*
Primary efficacy outcome					
Reversal of HH on MRI	14% (1/7)	100% (7/7) *	*p=0.002*	86% (6/7) *	*p=0.01*
Secondary efficacy outcomes					
CSF lumbar leakage	100% (7/7)	14% (1/7) *	*p=0.002*	43% (3/7) *	*p=0.023*
HL neurologic examination	N=7	N=6		N=7	
Mean HL joint movement score (max=12)	3.2±2.19	9.3±4.14 *	*p=0.032*	7.25±4.24	*P=0.055*
Mean locomotor grade (max=15)	1.4±0.72	4.2±2.22 *	*p=0.017*	2.86 ±1.69	*p=0.240*
Mean parietal ventricular diameter on MRI **	4.1 ±1.59	3.6 ±1.53	*p=0.1148*	4.0±1.60	*p=0.7190*
Presence of MEP in both HL	40% (2/5)	100% (3/3)		50% (2/4)	
Latency	22,70 ±20,44	38,72 ±19,69	*p=0.274*	23,30 ±18,29	*p=0.998*
Area-Under-the-Curve	3,52 ±,52	11,84 ±10,31 *	*p=0.035*	5,06 ±3,94	*p=0.850*
Peak-to-Peak amplitude	0,08 ±0,07	0,24 ±0,17 *	*p=0.044*	0,14 ±0,12	*p=0.519*
Presence of SEP in both HL	0% (0/4)	0% (0/4)	*p=1.0000*	0% (0/4)	*p=1.0000*

Table 1: *Comparison of outcomes between repaired (watertight or non-watertight) and unrepaired SB lambs (superiority study). All data showed equal variance. For normally distributed continuous variables, one-way ANOVA combined with post Hoc Tukey's multiple comparisons was used. Kruskal-Wallis test with its post Hoc test was used to compare categorical variables and non-normally distributed continuous variables.*

*A p-value <0.05 was considered significant and is marked with an *.*

** Ventriculomegaly is defined as a mean ventricular diameter of >2mm on axial slides. Abbreviations: SB, spina bifida; CSF, cerebrospinal fluid; MRI, magnetic resonance imaging; HL, hindlimb; MEP, motor evoked potentials; SEP, somatosensory evoked potentials.

Conclusion: In the SB fetal lamb model, watertight 2-layer fetal SB repair is safe in terms of survival. Such repair effectively and completely reverses HH and improves hindlimb neuromotor function. Non-watertight repair does not yield as much neuroprotection.

OP44

Percutaneous fetoscopic patch coverage of spina bifida aperta creates new commodity of patients

Thomas Fortmann, Angela Brentrup (Münster, Germany)

Introduction: In 2003 the pediatrician Thomas Kohl started the percutaneous fetoscopic patch coverage of spina bifida aperta in Germany. Due to this new kind of surgery a new commodity of patients developed. These patients have different problems than postnatally operated spina bifida patients.

Methods: We report of our experiences in Muenster of eight patients who present themselves with different kind of problems.

Results: Wound healing is a big issue for these patients. Two patients underwent postpartal surgery due to dural tears, one twice. Two patients had local infections resulting in prolonged wound healing. One patient developed a secondary meningomyelocele and needed open surgery. The anatomy of the celes is not respected as in postpartal surgery. This means that in case of revision surgery, it is difficult to separate adequate layers of tissue for covering the defects. Two patients already suffered from tethered cord. Two patients have a remaining anchor, which makes MRI imaging difficult/impossible. Chiari malformations are not prevented by fetoscopic coverage – one patient even needed decompression. Six patients still developed a hydrocephalus and needed a shunt supply.

Conclusion: From our experience in Muenster a new commodity of patients developed after the beginning of fetoscopic coverage of spina bifida aperta, which needs special attention due to a difficult wound healing, a different postsurgical anatomy and so far uncommon problems for spina bifida patients.

OP45

Functional outcome after microsurgical detethering in children and young adults

Aurelia Peraud, Nicole Terpolilli (Munich, Germany)

Objective: Tethered Cord Syndrome (TCS) comprises a combination of neurological, musculoskeletal and urological disturbances caused by abnormal tension of the spinal cord. Microsurgical detethering is the therapy of choice in symptomatic TCS, but ideal timing is still unclear. The aim of the current study was to analyze the outcome after surgical detethering with regard to age, preoperative symptomology, and neurological outcome.

Methods: Clinical records of all patients under the age of 20 years microsurgically treated at our department for tethered cord syndrome and presented to our outpatient clinic at least once for follow-up between 01/2003 and 10/2016 were retrospectively analyzed.

Results: 59 patients (27 male, 32 female) diagnosed with symptomatic tethered cord syndrome were included in the current study. Patients were postoperatively followed up for 2.6 years on average (0.2-9.7 years). Most common initial symptoms included bladder disturbances (89.8%), muscle weakness (83.1%), foot deformities (45.8%), and back pain (27.1%). Mean age at operation was 6.1 years (10 days-20.0 years). All patients underwent microsurgical detethering under intraoperative electrophysiological monitoring. There was no postoperative worsening of symptoms. Bladder function improved in 67.3% of cases presenting with this symptom; recovery of urinary function tended to be more complete when patients were operated before the age of 4 years. Pain abated in 62.5% of patients. Motor function significantly improved in 67.3% of patients. Of note, while of the 17 patients that were too young to stand and walk at the time of operation (below the age of 12 months) 11 were ambulatory at the time of last check-up, only one of 9 patients with detethering after the age of 1 year regained the ability to ambulate.

Conclusion: Our results indicate that neurological outcome, especially recovery of urinary function, after microsurgical dethetering, is more favourable when surgery is performed early in life and when symptoms are mild. Further and prospective analyses are needed in order to validate these findings.

Abstract session 14: Hydrocephalus I

OP46

Premature infants less than 1500g birth weight: incidence of intraventricular hemorrhage and hydrocephalus. A single institute analysis

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Objective: With changed treatment concepts in neonatology the mortality and occurrence of intraventricular hemorrhage (IVH) in premature babies could be lowered. The incidence of IVH in very low birthweight infants (VLBW <1500g) varies in the literature between 20% (Murphy 2002) and 61% (Sajjadian 2009). The aim of our study was to analyze the incidence of IVH, hydrocephalus (HC) and shunt dependency in our institution.
Methods: We retrospectively analyzed all preterm babies equal or less than 1500g, which were born in the years 2011 to 2015 in the children's hospital. In addition to birth weight the occurrence of cerebral hemorrhages and/or ventricular dilatation were investigated as well as the frequency of shunt dependency.
Results: 170 VLBW infants (89 males, 81 females) were included. The lowest birth weight was 360g and on average 1119g. In 153 cases a cesarean section was required, 53 times it was a multiple pregnancy. The overall mortality was 3%. Brain sonography was done routineously and showed an IVH in 35 cases (20.6%). Twelve children developed unilateral hemorrhage, in 23 cases the bleeding occurred bilaterally. A moderate to severe IVH (grade 3-4) were observed in 7% and a mild IVH (grade 1-2) in 13.5%. A reservoir was required in 10 cases (5.9%). All these children (8 males, 2 females) developed shunt dependency. A nonadjustable valve with integrated gravitational unit was implanted in 90% and once an adjustable valve without gravitational unit. In 50% initially antibiotic impregnated catheter system (AIS) was used. The follow up time ranged between two (homeland-related further treatment) and 60 months. No mortality and no intraoperative complications occurred in this high risk group. One child did not require revision in the follow up period, one revision became necessary in 4 cases. The remaining had 2 to 3 revision surgeries. Seizures were observed in 2 children, all children are developmentally retarded in varying degrees.
Conclusions: In the high risk VLBW group the mortality rate was low with 3%. Compared to the literature, the rate of hemorrhage is in the lower range at 20.6%. Only 5.9% became shunt-dependent during the course. Unfortunately, all hydrocephalic children are suffering from retardation.

OP47

The management of post-hemorrhagic hydrocephalus in preterm infants: retrospective analysis of 57 cases treated with Ommaya reservoir, endoscopic third ventriculostomy and ventriculoperitoneal shunt

Paola Ragazzi, Valentina Pennacchietti, Pierpaolo Gaglini, Mario Cacciacarne, Christian Carlino, Paolo Galletto, Luciana Costa, Rossana Bagna, Paola Peretta (Turin, Italy)

Introduction: Endoscopic third ventriculostomy (ETV) is an established treatment for obstructive hydrocephalus, but its role in post-hemorrhagic hydrocephalus (PHH) is controversial. Here we describe the safety and efficacy of ETV as an option in the management of PHH in preterm infants.
Methods: We retrospectively analyzed 57 preterm patients (2002-2017) affected by PHH and treated with a ventricular catheter connected to an Ommaya reservoir and intermittent CSF aspiration. In patients with long-lasting hydrocephalus we performed a ventriculoperitoneal (VP) shunt or an ETV.
Results: Mean gestational age of the population was28.5 weeks. Three patients had grade II, 18 III and 36 IV intraventricular hemorrhage. Mean age at reservoir implant was 19.8 days, mean interval between aspiration 2.5 days. Complications occurred in 10 infants, leading in one case to reservoir replacement. Four patients presented meningitis requiring external ventricular drainage. The Ommaya reservoir was punctured on average 11.2 times per patient. Four patients died for complications unrelated to reservoir, one was lost at follow-up. Thirteen children resolved hydrocephalus with Ommaya tapping, whereas 39 required a permanent treatment: 18 a VP shunt, 21 an ETV. Three out of the 18 shunted patients had malfunction: they underwent ETV and shunt removal after 6.1-20.5 months; one had a new VP shunt 9.9 months after ETV. Among the 21 patients with ETV, 11 underwent a VP implant because of progressive hydrocephalus. Two had a redo-ETV with shunt removal, but only one remained shunt free. At the end of the follow-up 26 children were shunt free (50%).
Conclusion: Ommaya reservoir, VP shunt and ETV significantly reduced shunt dependency in a traditionally shunt dependent population. ETV is a feasible and effective treatment alternative to shunt implant. ETV with VP shunt removal is an alternative for shunt revision. A careful study of the imaging supports the selection of ETV candidates.

OP48

CSF upward flow during neuroendoscopy of obstructive hydrocephalus: What is the implication for ETV success?

Hans Christoph Ludwig, Hans Christoph Bock, Steffi Dreha-Kulaczewski (Göttingen, Germany)

Objective: Contemporary hydrocephalus (HC) concepts are based on animal experiments done by Dandy, Key and Retzius. According to these classifications the choroid plexus was established as the site of CSF production and the arachnoid villi as the structures of its absorption. Nowadays gated flow MRI has supported the hypothesis of bulk flow and piston like movements out of the ventricles towards arachnoid absorption. Many publications support mixing and diffusion characteristics without directed flow of cerebral and spinal CSF. Recent studies applying real time-MRI with very high temporal resolution identified respiration dependent CSF

movements in contrast to our current concepts. In light of these new findings, we have reanalysed our neuroendoscopic third ventriculostomy (ETV) procedures focusing on upward CSF movements. Furthermore, we measured Fronto-Occipital-Horn-Ratios (FOR), assessed flow void signals in pre- and postoperative clinical MRIs and related the results to ETV outcome.

Material and Methods: Video analyses of intraoperatively documented trapping mechanisms during neuroendoscopy and an evaluation of MRI flow void signals in 46 pediatric patients with obstructive HC have been conducted. Mechanisms of trapping were investigated for underlying breathing induced dynamics. ETV outcome was estimated after 5 y (mean) follow up.

Results: Trapping mechanisms could be identified in 85 % of the MRIs. Intraoperative CSF dynamics was in 64 % related to breathing as revealed by video documentation. Mean FOR in successful ETV declined in 40.4 % from 0.67 to 0.59 (mean) and without success from 0.67 to 0.62.

Conclusion: 85% of MRIs revealed a CSF trapping component with opposite flow directed from spinal canal into the ventricles or cystic compartments. This observation is in agreement with recent *in vivo* real time-MRI findings. The implication of any cranial directed CSF flow might explain some of the ETV results in childhood.

OP49

Management of External Ventricular Drains (EVD) – Systematic review of measures to reduce infection rates

Christopher Munoz-Bendix, Ann-Kristin Schmitz, Hans-Jakob Steiger, Thomas Beez (Düsseldorf, Germany)

Background: EVD infection rates are as high as 30%. Attempts have been made to reduce infection rates by improving implantation technique, handling protocol and ventricular catheters. We systematically review the impact of such attempts on EVD infection rate.

Methods: A systematic review of PubMed database was performed to identify controlled studies analyzing measures to reduce EVD infection. Full-text articles were reviewed according to Cochrane Guidelines. Data was analyzed with Review Manager Version 5.3 (Cochrane Collaboration, 2014) and odds ratios (OR) with 95% confidence intervals (CI) were calculated.

Results: Sixteen studies with a total of 3,613 events were eligible for inclusion in this pooled analysis: 3 studies compared standard with silver-impregnated catheters, 7 studies compared standard and antibiotic-impregnated catheters, 3 studies compared standard care with special "infection control bundles" and 3 studies compared standard with long tunnelling. Antibiotic-impregnated catheters significantly reduced the infection rate with OR 0.17 (CI 0.11-0.28). "Infection control bundles" also showed a significant effect with OR 0.10 (CI 0.05-0.21). In contrast, silver-impregnated catheters and long tunnelling only showed a tendency towards infection reduction with OR 0.65 (CI 0.16-2.71) and 0.32 (CI 0.07-1.38), respectively. The crude overall infection rates for cohorts in the individual studies ranged from 3% to 27%.

Conclusions: Antibiotic-impregnated catheters and improved implantation and handling techniques, summarized in "infection control bundles", both significantly reduce EVD infection rates. When performing this systematic review it became evident that 1) infection rates varied widely, partially due to different definitions of "EVD infection" and that 2) some interventions might have an impact on infection rates, but were never analyzed separately in a controlled fashion (e.g. experience of the surgeon, frequency of CSF sampling, frequency and type of wound dressing). Based on systematic literature review a gold standard can be defined, but several questions remain open regarding the "perfect" EVD.

Abstract session 16: Neuro-Oncology I

OP50

Recurrent posterior fossa ependymoma in children: the role of surgery

Matthieu Vinchon, Camille Di Palma, Pierre Leblond (Lille, France; Caen, France)

Background and Purpose: Posterior fossa ependymoma (PFE) in children is considered a surgical disease because chemotherapy has little impact and radiotherapy is limited in young children. However, even when resection is apparently total, the recurrence rate is high. The role and impact of reoperation for PFE has been little studied.

Material and Methods: In order to study the results of reoperation, we reviewed our experience with PFE in children during the last two decades. This is a monocentric, retrospective study.

Results: Between 1997 and 2017, we operated 41 children for PFE. Resection was apparently total in 36 (88%), and postoperative irradiation was given in 28 (68%). After a mean follow-up of 68 months, 19 patients (46%) showed tumor progression; among these, 14 were reoperated; the other 6 were deemed not suitable for surgery on account of metastasis, inaccessible tumor, or sequellae of previous surgery. Overall, 9 patients (22%) died of tumor progression.

The event-free survival (EFS) and overall survival (OS) after first surgery were 45% and 77% at five years respectively. The OS at five years after tumor progression was 69% in the reoperated group, VS 0 in the non-reoperated group.

Conclusion: PFE in children has a high rate of tumor recurrence. In our study, the OS rate after reoperation is comparable to the OS rate after the first operation. Reoperation with total second resection appears to offer the best chances of cure. Reoperation for recurrent PFE

should be proposed as curative means whenever possible, including cases with single metastasis

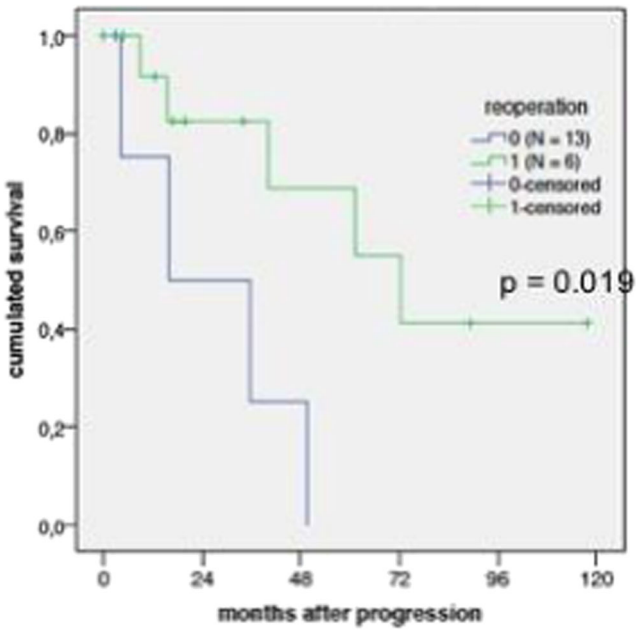

Figure 1: *OS after tumor progression*

OP51

Repeat surgery after sub- or near-total resections of pediatric brain tumors

Ann Kristin Schmitz, Christopher Munoz-Bendix, Sevgi Sarikaya-Seiwert, Hans-Jakob Steiger, Thomas Beez (Düsseldorf, Germany; Bonn, Germany)

Objective: Neurosurgical resection is commonly the first step in the treatment of brain tumors in children. Several studies have established gross total resection as an important predictor of outcome. However, postoperative tumor residuals can occur for intentional or unintentional reasons. Decision-making regarding a repeat surgery can be challenging, as risk and benefit are difficult to estimate. We thus analyzed feasibility, safety and benefit of repeat surgery after sub- or near-total resections.

Methods: In this retrospective analysis, we enrolled pediatric brain tumor patients who received subtotal (>1,5 cm^2 remnant) or near-total (<1,5cm^2 remnant) neurosurgical resections of WHO grade II, III or IV brain tumors between 2007 and 2017. Operations were performed in a standardized setting with neuronavigation or ultrasound-guidance and intraoperative neurophysiological monitoring (IOM). Volumetric measurements of tumor residuals were performed on 72h-postoperative contrast-enhanced MRI scans. Neurological status and perioperative morbidity were recorded. Additional clinical data was obtained from patient charts.

Results: We identified 39 patients (12 female; mean age: 8±5 years). Histological entities included ependymoma WHOII/III (n=8), glioblastoma WHO IV (n=9), astrocytoma WHO II/III (n=6), medulloblastoma WHO IV (n=11), ATRT WHO IV (n=3), PNET WHO IV (n=2). We observed 14 near-total and 25 subtotal resections. Eleven patients received early repeat surgery. Postoperative MRI scans revealed a tumor volume reduction in all cases (seven near-total and four gross total resections). The extent of repeat resection was limited by visual or neurophysiological identification of eloquent structures. No new perioperative morbidity or surgery-associated mortality was recorded.

Conclusions: Repeat surgery with IOM after incomplete initial resection did not increase surgical morbidity or mortality and thus appears to be feasible and safe. However, its role regarding oncological outcome has to be further investigated. Additional factors (such as further delay of adjuvant therapy) might be relevant in the risk-benefit assessment during decision-making for repeat surgery.

OP52

Posterior Fossa Syndrome (PFS). A possible etiology

Barbara Spacca, Flavio Giordano, Elena Arcovio, Francesca Livi, Lorenzo Genitori (Florence, Italy)

Introduction: Posterior Fossa Syndrome is a complication after posterior fossa surgery difficult to foresee. It is associated to tumor resection but can result after any cerebellar damage. Signs and symptoms are various (cognitive, neurological, behavior) but it is typically represented by troubles in speech. Evolution can be different, and recover can be partial. Still open is the debate over the reason why patients develop PFS in 8-30% after surgery.

Methods: Between 2005 and 2016, 193 patients were operated for PF tumors. Inclusion criteria were: age between 2 and 18 years; first surgery; no previous language or movement disorders, normal level of consciousness on admission.

Patients were divided into two groups: who developed a PFS and who did not. Retrospectively, we assessed surgical, clinical and radiological details. MRI scans were reviewed for brainstem infiltration at Peri Acqueduttal Grey (PAG) and Superior Cerebellar Pedunculus (SCP). Statistical analysis was performed (Chi-square).

Results: Seventeen patients, younger than sixteen years, developed PFS within 72 hours after surgery. In this group: transvermian route was the most common (70.6% vs 28.4%); gross total removal was obtained in 58.8% (65.3% in the whole cohort); histology was medulloblastoma in 52.9% (26.7% in the others) and LGG in 35.3% (45.5% in the others). Eleven recovered after a mean of 9.4 months; six had persistent neurological and behavioral disorders.

Brainstem infiltration was evident in 95 patients (49.2%):15 of them (15.7%) developed PFS. Chi-square revealed a

positive association between brainstem infiltration ($p<0.001$) and trans vermian surgery ($p<0.015$) with PFS.
Conclusions: Brainstem infiltration and transvermian surgery were associated with a higher risk of PFS with a statistically significant association. A hypothesis may be the damaged induced by the infiltration itself and/or by the surgery to the PAG, garbling its central linking role between brainstem and forebrain and its activity in modulation of the verbal response to pain and stress.

OP53

Long-term outcome after treatment for pediatric medulloblastoma and supratentorial PNET – What life do the survivors get as adults?

Radek Frič, Bernt J. Due-Tønnessen, Einar Stensvold, Tryggve Lundar (Oslo, Norway)

Aims: Medulloblastoma (MB) and supratentorial primitive neuroectodermal tumor (CNS-PNET) are the most common malignant pediatric brain tumors. In this presentation, we reflect the outcome of treatment of the cohort of children with MB/CNS-PNET treated at Oslo University Hospital during the last four decades.
Material and Methods: The background for the presentation were the previously reviewed data from all patients < 20 years of age diagnosed and treated for MB/CNS-PNET at our institution between 1974 and 2013. In this presentation, we specifically focus on long-term survivors.
Results: The cohort consists of 158 patients (median age at presentation 7.1 years, 41% girls). Gross total resection (GTR) was achieved in 118 patients (75%). 126 patients received radiotherapy as a part of the primary treatment whilst 24 patients did not due to their low age. The overall 5-year survival rate for MB/CNS-PNET was 54% (MB 57%, CNS-PNET 41%). 5-year overall survival for patients with GTR versus those with non-GTR differed significantly (64% versus 22%). At the time of analysis, 63 patients were alive and disease-free, one was alive with disease, and 96 patients were deceased: 84 of these (88%) due to progression of their tumor and 12 patients due to supposed late effects of the treatment. We noticed 11 cases of secondary malignancies, three of them with fatal outcome. Parameters of quality of life from long-term survivors are also presented, indicating decreasing quality of life over time.
Conclusions: Survival data from this historical series of children treated for MB/CNS-PNET are comparable to data from other population-based studies. Importantly though, we noticed very late reccurences and secondary malignancies even in long-term survivors. Our data from the analysis spanning 40 years of treatment stress the need for life-long follow-up after treatment of MB/CNS-PNET in the childhood.

OP54

Prone versus sitting position in pediatric low-grade posterior fossa tumors

Valentina Baro, Luca Denaro, Domenico d'Avella (Padova, Italy)

Background: The right intraoperative positioning of the patient is a fundamental element in all surgical procedures, to obtain a good surgical field and ergonomic gestures for the surgeon, and to guarantee a comfortable/safe position for the patient. Throughout history two positions were adopted to access posterior cranial fossa: prone and sitting. Both positions highlighted advantages and disadvantages, in fact, there are several reviews and case-reports on postoperative complications after posterior fossa surgery. However, at present, comparisons between these positions are rare in literature and the lack of homogeneity in procedures and diagnosis impedes a bias-free comparison of postoperative outcome and there is only one report in children so far. Pediatric patients represent a population that could mainly benefit from a low-rate of postoperative complications.
Aim: The aim of this study is examining the intraoperative and postoperative complications, and the follow-up of pediatric patients affected by low-grade tumors of posterior cranial fossa, in particular, pilocytic astrocytoma (grade I WHO), and comparing their occurrence in patients operated in the prone position with those in the sitting position.
Materials and Methods: We retrospectively assessed 30 pediatric patients affected by pilocytic astrocytoma of the posterior cranial fossa, from the beginning of the symptomatology, during the hospitalization, to the postoperative course, extending our analysis with clinical and neuroradiological follow-up. Two centers were involved in the analysis because of the standardized use of only one position, prone or sitting, for approaching these lesions. 16 patients underwent posterior fossa craniotomy in the prone position (from 1999 to 2015) and 14 in prone position (from 2011 to 2017). Medical records were used to get the preoperative, intraoperative and postoperative data, while follow-up data and the actual health status were obtained from medical reports of the clinical and neuroradiological examinations, and through telephone interviews with questionnaire administrations.
Results: Preoperative data didn't show statistically significant differences between the two groups. Intraoperative and postoperative complications were similar among the two groups, and small differences could not be ascribed to patient positioning (p-value > .05).
Conclusions: These data suggest that Concorde position and sitting position can be considered safe in suboccipital craniotomies, and anesthesiological complications assume nowadays a second-rate importance. Neither prone nor sitting position is complication-free in the postoperative course, but there aren't statistically significant differences which tend to prefer one

position. Further studies are needed to show if there are possible differences between prone and sitting position for other frequent pediatric tumors such as medulloblastomas and ependymomas.

OP55

Pineal tumors in children: the French Register and the Lyon experience

Carmine Mottolese (Lyon, France)

Introduction: Pineal tumors account for 0,5-1% of all CNS tumors. 50% of all pineal tumors are benign tumors that need a surgical removal.

We report the experience of the Lyon and of the French Register of the pineal tumors focusing on pediatric cases.

Material: 812 patients belong to the French Register of the pineal tumors: 284 represent pediatric patients.

Pineal parenchymal tumors (PPT) represent 36% of cases, germinal tumors 25%, glial tumors 14%, papillary tumors 7%, 15 % pineal cysts and other13%. In the series of Lyon PPT represent 20% and germinal tumors 26%. 190 patients were treated for hydrocephalus: 116 by a shunt and 46 patients by endoscopic ventriculostomy: for 38 patients no data were found.

94 patients had a biopsy: 24 by endoscopy, 53 in stereotactic conditions and 17 by direct approach.

138 patients underwent surgical resection by suboccipital-transtentorial either supracerebellar infratentorial approach.

Results: Analysis of the results shows a tight relationship with histology with better survival for benign lesions. The overall rate of survival was of 84%:

pinealocytomas 100%; PTT grade II 83%; PTT grade II – III 83%.

In patients with pinealoblastoma, survival does not exceed 25% with a particularly poor outcome for children younger than 3 years.

Survival for germinomas was of 88% while for mixed secreting germ line tumors the overall survival was of only 15%.

For pineal gliomas the survival was 85%.

Conclusion: Pineal tumors in children represent a difficult challenge still to-day.

Pynelocytomas are rare tumors in children.

The decisional tree depends on radiological and markers studies and need a multidisciplinary discussion. Benign lesions are treated with a low rate of sequels while malignant lesions as pinealoblastomas have still a bad prognosis.

OP56

Awake brain surgery in children

Laura-Nanna Lohkamp, Pierre-Aurelien Beuriat, Michel Desmurget, Alexandru Szathmari, Cecile Faure-Conter, Didier Frappaz, Federico Di Rocco, Carmine Mottolese (Lyon, France)

Introduction: Awake brain surgery has limited indications in children due to age and neuropsychological aspects interfering with feasibility and psychological outcome. Few case series have been reported in children focusing on the differences compared to adults. Neuropsychological testing and monitoring may have an important impact on ameliorating eligibility of children undergoing awake procedures. This study reports our criteria for practicability, neuropsychological testing and diversified outcome aspects of awake brain surgery in children.

Methods: Retrospective review and prospective outcome analysis of all children who underwent awake brain surgery in Lyon between 2005 and 2017.

Results: Among 16 children considered for awake brain surgery 14 were accounted eligible after neuropsychological evaluation and underwent asleep-awake-asleep brain surgery. The cohort included 4 males and 10 females. The median age at surgery was 13.8 years, (range: 9.9 to 17.6 years). The indications were related to CNS-tumors in eloquent areas, mostly of glial origin. Intra-operative testing included cortical electromagnetic stimulation during speech or motor activity. A complete tumor removal was achieved in 10 patients. Transient neurological deficits were postoperatively observed in 2 patients, whereas severe psychological troubles occurred in 1 child aged 10 years old. 2 patients showed persistent attention deficits, which were most probably related to the tumor location itself, specific cortical and subcortical regions respectively. Two patients died during follow-up due to tumor progression. The mean duration of follow up was 21,7 months (range: 3,4 to 46,8 months).

Conclusion: Awake brain surgery was shown to be beneficial in older children in terms of efficient tumor resection beside simultaneous preservation of neurological functions. Neuropsychological testing after surgery is essential to determine the individual psychological outcome, which can be altered in a minority of patients despite a careful preoperative evaluation. Preemptive preparation and neuropsychological sustain may contribute to realize awake brain surgery in even younger children.

Abstract session 17: Neuro-Oncology II

OP57

Proteomic analysis of pediatric glioblastoma

Luca Massimi, Claudia Martelli, Alessia Nesticò, Gianpiero Tamburrini, Paolo Frassanito, Massimo Caldarelli, Massimo Castagnola, Claudia Desiderio (Rome, Italy)

Introduction: To date, the proteomic analysis has been mainly focused on the adult glioblastoma (GBM), only a very few studies concerning the pediatric variant. These studies are based on the enzimatic pre-digestion of the proteins, while here we present the result of a study realized through the top-down approach.

Patients and Methods: The samples of the last 6 pediatric patients (4 males, 2 females; mean age at surgery: 8 years) consecutively operated on for GBM (WHO IV) at our Institution were analyzed. The steps consisted in: 1) Extraction of the protein acid soluble fraction from tissue samples; 2) Analysis by HPLC-ESI-LTQ-Orbitrap Elite; 3) Identification and characterization of the proteins; 4) Data interpretation.
Results: The proteome of the pediatric GBM was characterized by a predominant presence of hemoglobin alpha-chain, beta-thymosin 4 peptide, ubiquitin proteoforms, alpha-defensins, protein S100A6 and Heat Shock 10 kDa protein. More in detail: 1) The entire form of ubiquitin was absent in favor of the truncated forms, particularly of des-RGG form. Ubiquitin is involved in the tumorigenesis; 2) High levels of the des-AGES form of the thymosin beta-4 were detected. This peptide is the most abundant among the beta-thymosin family and seems to be involved in the tumor invasion; 3) The defensins would not characterize the GBM but could indicate the inflammatory reaction against the tumor, which varies according to the patient; 4) Different chains of hemoglobin have been described in GBM. Our data encourage a deeper investigation on its role of in the GBM cells; 5) S100A6 and Heat Shock proteins could be involved in the tumor-related oxidative stress.
Conclusions: Compared with other pediatric brain tumors that we investigated by a proteomic analysis (medulloblastoma and pilocytic astrocytoma), GBM is characterized by the absence of entire ubiquitin and by the presence of truncated forms of this protein and truncated forms of thymosin beta-4. These proteins may be considered as new markers of GBM and new hints for investigating possible therapeutic targets.

OP58

Are pediatric glioblastomas different from adults?

Heidi Baechli, Ahmed El Damaty, Olaf Witt, Felix Sahm, Andreas Unterberg, Stefan Pfister, Till Milde (Heidelberg, Germany)

Objective: Pediatric glioblastomas (PGBM) accounts for 8-12% of all primary CNS tumors among children with an incidence between 0.85 per 100.000. The classification of brain tumors has previously been based on histology but has now been fundamentally changed by the addition molecular data. The histological appearance of these tumors is malignant, but molecularly there are distinct entities with divergent biology and different prognosis. We here report 5 cases that were initially diagnosed as pGBM, whose diagnosis changed significantly after molecular analysis.
Methods: Five children were included, aged between 7 and 14 years at diagnosis. The neuropathology of case 1 was a mesencephalic GBM (central view: anaplastic astrocytoma III), case 2 a GBM of the right cerebellar peduncle, case 3 a left frontal GBM with giant cells and case 4 a right parieto-occipital anaplastic astrocytoma III. Case 5 was a (insert location) GBM IV. Case 1, 2 and 5 were incompletely resected, case 3 and 4 were completely resected. All cases were treated by chemo- and /radiotherapy.
Results: Diagnoses and/or prognoses changed significantly after molecular profiling: case 1, 2and 4 were reclassified as low grade gliomas, case 3 as a pleomorphic malignant glioma with germline alteration and case 5 unchanged a GBM IV. DNA sequencing revealed important molecular alterations in 3 cases: a TP53 mutation (tumor and germline) in the pleomorphic malignant glioma, a BRAF V 600E mutation, a pilocytic astrocytoma, and an IDH1 mutation. Follow up revealed stable disease or complete remission, respectively, over 7 years in case 1, 2 and 4, a complete remission over 2,5 years in case 3, and a stable disease over 3 years in case 5.
Conclusion: PGBMs are distinct from adult GBMs and molecular analysis can identify miss-classified low grade tumors, underlying cancer predisposition syndromes, and molecular subgroups with better prognosis.

OP59

Role of repressive histone modification marks and histone Lysine Methyltransferases SETDB1 and EZH2 in pediatric brain tumors

Alexia Klonou, Antonios N. Gargalionis, Penelope Korkolopoulou, Marios Themistocleous, Athanasios G. Papavassiliou, Spyros Sgouros, Christina Piperi (Athens, Greece)

Introduction: Chromatin remodeling alterations such as histone methylation induced by their respective histone methyltransferases are considered to have a prognostic value in gliomagenesis and in pediatric gliomas onset. The aim of the study was to investigate the differential expression of repressive histone marks H3K9me3, H4K20me, active histone mark H3K4me3 and linker histone H1x in pediatric gliomas. In addition, expression of lysine N-methyltransferases SETDB1 and the Enhancer of zeste homolog 2 (EZH2) was evaluated.
Methods: Archival human glioma tissues and normal brain samples were provided by the Neurosurgery Departments of "Mitera" and "Agia Sophia" Pediatric Hospitals and the study was approved by the University of Athens Medical School Ethics Committee.
Protein expression was evaluated immunohistochemically as H-score (intensity multiplied with cell percentage, 0-300) in 20 pediatric tumor samples (18 astrocytomas grade II-III, 2 glioblastomas; age 3-14 years old; 11 males, 9 females) and in 5 samples of normal brain tissue.
Results: Increased nuclear staining of H3K9me3 and H4K20me3 repressive marks was observed in astrocytomas (median H-score 298 and 295 respectively). Moderate nuclear staining was obtained for H3K4me3 and SETDB1 (median H-

score 190 and 120 respectively), whereas EZH2 and H1x presented no significant nuclear expression. A positive association of SETDB1 with H3K9me3 expression was observed (p=0.043). SETDB1 staining was significantly elevated in male compared to female children (p=0.007) and tended to be higher in glioblastomas compared to astrocytomas II-III (p=0.071).

Conclusion: These findings indicate the possible involvement of H3K9me3, H4K20me3 and H3K4me3 histone marks in the pathogenesis of pediatric gliomas. The histone methyltransferase SETDB1 was found to play a significant role in modulating gene expression, possibly by inducing H3K9me3 epigenetic mark. Future studies validating these chromatin remodeling changes in larger cohorts are needed along with elucidation of the underlying molecular mechanisms for potential therapeutic targeting.

OP60

The design and preliminary findings of the German CyberKnife registry for pediatric patients with CNS lesions

Laura-Nanna Lohkamp, Arne Grün, Peter Vajkoczy, Volker Budach, Markus Kufeld (Berlin, Germany)

Introduction: Although CyberKnife radiosurgery is a well-established method in adults with CNS lesions scientific evidence in pediatric patients remains scarce for all entities. In addition, the incidence of pediatric CNS lesions amenable to radiosurgery is low, thus clinical trials for evaluating this method and its indications in children are mandatory. The purpose of this study is to evaluate the efficacy, safety and outcome of pediatric CNS lesions treated, using the CyberKnife radiosurgery system.

Material and Methods: The radiosurgery registry for pediatric patients (≤ 18 years) with CNS lesions is designed as an interdisciplinary multicenter observational study, intending to include patients on a retrospective and prospective basis. Epidemiologic, clinical and imaging data will be collected and the follow-up will be monitored electronically throughout a five-year period. Primary endpoint is clinical outcome for benign lesions at five years of follow-up and for malign lesions local tumor control at 1- and 2-years of follow-up. Secondary endpoints are radiation toxicity, side effects and neurocognitive development.

Results: Patient enrollment was initiated at 11 treatment centers throughout Germany. Since 2005 95 treatment interventions have been carried out, 29% of them with palliative and 71% with curative intention. 38 of the treated lesions were benign CNS tumors, followed by 27 vascular lesions and 9 ependymomas. Other treatment indications were CNS metastases, craniopharyngeomas and germline tumors. Treatment interventions required anesthetic support in half of the patients and 11 of 95 patients were treated repetitively.

Conclusion: The multicenter CyberKnife registry for pediatric patients with CNS lesions is an efficient tool to collect comprehensive treatment and follow up data of all children who underwent CyberKnife radiosurgery in Germany. Their retrospective and prospective analysis might clarify indications, outcome and treatment efficacy of CyberKnife radiosurgery in this patient group. The registry will serve as a basis for future clinical trials.

OP61

Early treatment of complex located pediatric low-grade gliomas using Iodine-125 brachytherapy alone or in combination with microsurgery

Mathias Kunz, Silke Nachbichler, Lorenz Ertl, Gunter Fesl, Rupert Egensperger, Maximilian Niyazi, Irene Schmid, Jörg-Christian, Aurelia Peraud, Friedrich-Wilhelm Kreth (Munich, Germany)

Introduction: To analyze efficacy, functional outcome, and treatment toxicity of low-dose rate I-125 brachytherapy (SBT) alone or in combination with best safe resection (in case of larger tumor volumes) as first-line treatment for pediatric low-grade gliomas (PLGGs) not suitable for complete resection.

Methods: Consecutively treated (2000-2014) complex located circumscribed WHO grade I/II PLGGs were included. For small tumors (≤4cm in diameter) SBT alone was performed; for larger tumors best safe resection and subsequent SBT was chosen. Temporary Iodine-125 seeds were used (median reference dose: 54Gy). Treatment response was estimated with the modified MacDonald criteria. Analysis of functional outcome included ophthalmological, endocrinological and neurological evaluation. Survival was analyzed with the Kaplan-Meier method. Prognostic factors were obtained from proportional hazards models. Toxicity was categorized according to the Common Terminology Criteria for Adverse Events.

Results: Fifty-eight patients were included treated either with SBT alone (n=39) or with SBT plus microsurgery (n=19). Five-year progression free survival was 87%. Two patients had died due to tumor progression. Among survivors, improvement/ stabilization/deterioration of functional deficits was seen in 20/14/5 patients, respectively. Complete/partial response had beneficial impact on functional scores (p=0.02). The 5-year estimated risk to receive adjuvant radiotherapy/chemotherapy was 5.2%. The overall early (delayed) toxicity rate was 8.6% (10.3%), respectively. No permanent morbidity occurred.

Conclusion: In complex located PLGGs, early SBT alone or combined with best safe resection preserves/improves functional scores and results in tumor control rates usually achieved with complete resection. Long-term analysis is necessary for confirmation of these results.

Abstract session 20: Neuro-Oncology V

OP62

Optic pathway gliomas of the pediatric age: impact of neurosurgery on quality of life

Alessia Imperato, Giuseppe Cinalli, Giulia Meccariello (Pozzilli, Italy; Naples, Italy)

Introduction: Optic pathway gliomas represent 1 to 5 % of paediatric CNS tumours. They can be sporadic or syndromic, often associated with NF1. Clinical manifestation comprises visual deficit, hydrocephalus, endocrinological disturbances. Despite benign histology, surgical radicality is impaired by optic pathway and/or hypothalamic infiltration. Treatment is multidisciplinary and comprises surgery, chemo- and radiotherapy.
Methods: We revised 58 patients with OPG followed at Neurosurgical Department of AORN Santobono-Pausilipon. Data were collected concerning pre- and post-operative endocrinological and visual function. Progression of disease and tumour volume were measured on MRI. When possible ophthalmologic data were completed with optic pathway tractography. Tumours were classified according to the modified Dodge classification (Taylor 2008).
Results: Median age at diagnosis was 6 years (6 months to 18 years). 36 patients had diagnosis of NF1 (31/36 were not candidate to surgery). 5 NF1 and 22 non-syndromic patients were operated: 6 patients achieved disease control with surgery, 21 patients had adjuvant chemo- and radiotherapy. At diagnosis 23 patients had visual disturbances and 6 endocrinological deficits. At the end of the follow up we evaluated:
-Visual outcome: improved in 10 patients, stable deficit in 6, worsened in 7 (in 2 cases due to tumour recurrence). Patients with no pre-operative deficit (4) had unremarkable visual outcome.
-Endocrinological outcome: 9 cases of new deficits.
-Disease control: 1 patient was cured, 23 patients reached partial control (subtotal removal in 2 cases and partial removal in 21); 1 is in progression, 1 died before adjuvant chemotherapy and 1 patient died after stabilization of the disease (domestic accident).
Conclusions: Surgery has gained increasing importance in long-term management of OPG. Our data suggest surgical indication for patients with disease progression. Adjuvant therapy should be reserved to patients with post-operative symptomatic residuals. Surgery has proven effective in treating cystic tumours, intracranial hypertension and in stabilizing or even improving visual outcome.

OP63

Edema of the optic tracts in tumors of the sellar region: clinical and visual implications in the pediatric population

Laurent Riffaud, Maia Proisy, Celine Chappe, Aurore Bussat (Rennes, France)

Objective: Tumoral-related edema of the optic tracts (EOT) corresponds to a preferential posterior distribution of peritumoral edema along the white matter tract of the visual system. To date, the consequence of EOT has never been evaluated specifically in the pediatric population. In this study, the authors have attempted to identify clinical and radiological features associated with the development of EOT and the specific influence of this edema on visual function.
Methods: A retrospective review was carried out of data collection from patients under 18 years operated on for a tumor of the sellar region at our institution between January 2005 and January 2016. Data was collected on patient characteristics, ophthalmological evaluations and neuroimaging findings. To evaluate and compare visual function impairment, ophthalmological data was converted into a global visual function score which took into account visual acuity and visual field evaluations and laterality deficiencies. Visual acuity score was defined according to the International Classification of Diseases. Visual field deficiencies were converted into a score 0 to 2. Two opposing groups were then distinguished according to the presence or absence of EOT. We compared visual acuity, visual fields and global scores between the groups before and after treatment.
Results: Twenty-six patients were included in the study: 17 craniopharyngiomas, 3 pilocytic astrocytomas, 2 gangliogliomas, 2 germ cell tumors, 1 macroprolactinoma and 1 Rathke cleft's cyst. There were 11 children in the group with edema and 15 children in the group without edema. None of the following criteria were statistically different between the 2 groups: age, gender, clinical symptoms at presentation (endocrine deficiency, intracranial hypertension signs), incidence of hydrocephalus, compression of optic tracts and mass effect on optic chiasm, tumor size and localization, intratumoral cysts, treatment, type of tumor, recurrence.
Median global visual function score and visual acuity score were not statistically different between the groups either at presentation or at final evaluation. Visual field score was lower (more deficiency) in the group with edema than in the group without edema ($p < 0.05$): 89% of the patients with edema had severe or mild visual field impairment versus only 40% in the group of patients without edema. At final examination following treatment, visual field scores were no longer different between the 2 groups. Although not significant, the number of patients with optic disc pallor was higher in the group without edema both at diagnosis and at final examination.
Conclusions: Our study confirms that edema of the optic tracts in a context of sellar region tumor in children is not necessarily associated with a less favorable visual prognosis

OP64

Thalamopeduncular tumors in children – Treatment strategy and outcome

Marek Mandera, Mikołaj Zimny (Katowice, Poland)

Introduction: Thalamopeduncular tumors although rare remain significant therapeutic challenge. The aim of the study was to analyze the results of treatment and discuss the preoperative planning on the basis of MR and diffusion tensor (DTI) imaging.
Methods and Results: We identified and analyzed retrospectively 9 pediatric cases of thalamopeduncular tumors treated surgically in the Department of Pediatric Neurosurgery in Katowice between 2007-2016. The mean age of patients was 7 years (ranged from 3 to 16 years). The mean follow-up was 55.6 months (ranged from 6 to 112 months). Clinically all patients presented hemiparesis at admission. Preoperative MRI was performed in all patients and DTI in 7 cases. Corticospinal tract (CST) was distorted anterolaterally in 4 cases, laterally in 2 and medially in one case. The choice of surgical approach was based on MR details of the tumor and CST distortion. Four patients were operated on by transcortical, via middle temporal gyrus approach, 3 cases by anterior interhemispheric, transcallosal, transforaminal approach and in 2 cases transylvian, transinsular approach was used. Near-total resection (>90%) was achieved in 5 and subtotal in remaining 4 patients. Pathological examination revealed pilocytic astrocytoma in 8 cases and pilomyxoid astrocytoma in 1 case. Two patients were operated twice due to progression of the tumor remnant. Postoperatively 4 patients presented mild progression of deficit, however the deficits improved after 2-14 days in 3 of them. Hemiparesis was on the preoperative level in three cases after surgery. Adjuvant chemotherapy or radiotherapy was used in none of patients.
Conclusions: Preoperative DTI allows to visualize the distortion of CST by the tumor and enables planning of the optimal surgical approach. As significant majority of thalamopeduncular tumors corresponds histologically to pilocytic astrocytoma total surgical removal should be attempted in most of them.

OP65
Surgical treatment of thalamic tumors in children
Alessia Imperato, Giuseppe Cinalli (Pozzilli, Italy; Naples, Italy)

Introduction: Improvement in microsurgical techniques and neuroimaging has allowed safer resection of thalamic tumors, once considered inoperable.
Methods: We reviewed medical records, imaging studies, operative and pathology reports of pediatric thalamic tumors treated at the Neurosurgical Department of AORN Santobono-Pausilipon.Neuronavigation (integrated with DTI studies) and intra-operative monitoring were used. The extent of tumor resection was assessed on MRI (within 24 hours): partial (< 90% STR), near-total (> 90% NTR), or total (no residual tumor GTR).
Results: Since 2002, 27 children with thalamic tumors were treated: 9 unilateral tumors, 16 thalamo-peduncular tumors and 2 patients with bilateral tumor (who underwent endoscopic biopsy and implantation of VP shunt). 25 patients received tumor resection. Different surgical approaches were chosen according to tumor location and displacement of the cortico-spinal tract. In 12 cases, multiple procedures were performed: in 7 cases, as part of a planned multistage resection, in the remaining 5 to manage recurrence or regrowth of residue. GTR was reached in 15 patients (63.2%), STR in 4 cases (15.7%) and PR in 6 patients (21.1%). Eighteen patients harbored low grade tumors. In this group, the mean follow-up was 45 months (range 4-132): one patient is dead, 12 patients are alive with no evidence of disease, 4 patients are alive with stable disease, 1 is lost to follow up. All patients are independent in their daily lives. The outcome of high grade tumors (9 patients) was very poor: 2 patients died immediately after surgery, 6 patients died for progressive disease, 1 patient is alive with residual disease.
Conclusion: Our institutional review seems to offer further evidence in favor of resection of thalamic tumors. In low-grade gliomas, radical resection can be curative without complementary treatment. Recurrences or residual regrowth can be safely managed surgically. In high grade tumors, the role of surgical resection remains matter of debate.

Flash Presentations

Abstract session 3: Craniofacial II

FP01
Chari malformation management in Bilambdoid and sagittal synostosis
Giovanna Paternoster, Syril James, Federico Di Rocco, Michel Zerah, Eric Arnaud (Paris, France)

Introduction: The ideal surgical treatment of Bilambdoid and sagittal synostosis-BLSS is not yet well defined. Several aspects of this specific group of complex synostosis need to be considered and could influence the surgical approach: Chiari malformation (CM), syringomyelia, sleep apnea syndrome (SOAS), morphological phenotype. In particular, the tonsillar herniation seems to be responsible of an increased re-operation rate.
Patients and Methods: Thirty-one non-syndromic patients were retrospectively analysed in our Department, from 1972 and 2014. We reviewed the clinical findings, radiological evolution and their surgical management.
Results: Four out of 31 patients were not operated on: 2/4 presented CM, asymptomatic and stable at long follow-up; no syringomyelia; no papilledema.

Twenty-seven cases required surgery: 8/27 patients (29,6%) were re-operated and 4 of them (14,8%) needed more than one procedure.
Nineteen operated patients presented CM at initial diagnosis, in 1 case associated with syringomyelia and in 3 cases with SOAS: in 13 patients CM remained radiologically stable, in 1 improved after surgery and in 5 of them worsened (in 1 case in association with syringomyelia that appeared 4 years later).
In all of these patients a cranial volte remodelling was performed and in 4/19 was necessary a redox for aesthetic reasons.
In 15 patients a posterior decompression (with distractors in 5 cases) was realised, in 4 of them associated to biparietal remodelling (BPR) and FM decompression: even if age at surgery was less than 12 months, no one showed clinical or radiological deterioration or required further surgery.
In 4/14 was associated to BPR: 1 patient operated at 3 months of age developed a syringomyelia and required FM decompression and further a 4V stent in a third surgical step; in 1 case a new frontal orbital advancement was necessary 4 years later for esthetical cause.
In the group of 7/14 patients submitted to a simple posterior expansion, in 2 cases a FM decompression became necessary for a CM aggravation with SOAS apparition at 3y FU and 2/6 required also a frontal orbital remodelling. One of 4 patients submitted to simple BPR required a further posterior expansion for morphological motive and CM aggravation.
Conclusion: The management of CM in BLSS is essential to reduce the number of surgical procedure and to improve patient outcome and evolution. Morphological evaluation (phenotype), age at diagnosis and at surgery, should be consider in the definition of the surgical plan.

FP02

Establishing standards of care in craniosynostosis: results from a survey of ERN CRANIO member institutions

Tiffany Berrington, Matthias Schulz, Hans Delye, Gregory James (London, United Kingdom; Berlin, Germany; Nijmegen, the Netherlands)

Introduction: European Reference Networks (ERNs) are networks involving hospitals with particular expertise in rare conditions. ERN-CRANIO focusses on rare disorders of the skull and face, including craniosynostosis. It consists of 29 member-institutions in 11 countries.
Methods: We undertook an electronic survey to understand current practice in craniosynostosis across ERN-CRANIO. 19/29 (66%) units replied. For single suture cases, 87.5% of units had a specific management protocol. 53.3% of units saw 51-100 new cases/year, 26.7% 21-50 and 20% <20. Units followed up children until 10 yrs/age or less (18.8%), age 11-15 (31.3%), age 16-20 (37.5%) and 12.5% of units never discharged single suture cases.
Results: A variety of surgical techniques are used. For sagittal synostosis, endoscopic + helmet (35.7%), spring assisted cranioplasty (21.4%), total calvarial remodelling (14.3%), open suturectomy + barrel stave (14.3%), modified Pi (7.1%) or parental choice (7.1%). 78.6% preferred surgery at 3-5 months. For metopic and unicoronal synostosis, 64.3% used fronto-orbital remodelling (bandeau), 14.3% non-bandeau frontal-orbital remodelling, 7.1% endoscopic + helmet, 7.1% dynamic cranioplasty and 7.1% parental choice; with 57.1% recommending surgery at 6-12 months. For multisutural syndromic craniosynostosis, 87.5% of centres have a management protocol. 31.3% of centres see 6-10 new cases a year, 25% 11-20, 25% <5, and 12.5% >20. Most centres (80%) either never discharge syndromic patients or follow until adulthood. 50% of centres transition all such patients to an adult service. 78.6% of units perform prophylactic vault expansion on syndromic cases, whilst 21.4% wait for clinical indication. 30.8% of units prefer to operate 3-5 months, whilst 69.2% prefer to wait for 6-12 months. Again, a wide variety of techniques are used across the ERN for initial surgery in syndromic children, most commonly frontal advancement (30.8%).
Conclusion: This study provides a useful snapshot of current standards of care in craniosynostosis across the high volume centres in the ERN.

FP03

Neurocognitive implications of sagittal craniosynostosis: role of the surgical treatment and genetic background

Daniela Pia Rosaria Chieffo, Wanda Lattanzi, Valentina Arcangeli, Camilla Zanetti, Paolo Frassanito, Luca Massimi, Massimo Caldarelli, Gianpiero Tamburrini (Rome, Italy)

Objective: To make a comprehensive neurocognitive, neuroradiological and genetic analysis of children affected by sagittal craniosynostosis, to estimate possible predictive factors of neurocognitive impairment.
Methods: All children diagnosed with isolated sagittal craniosynostosis from January 2014 to January 2017 were included in this study. Before surgery (T0) all children underwent a comprehensive neurocognitive evaluation and CT/MR scan. *SMAD6* and *BMP2* genes were sequenced in all patients to assess the presence of mutations/risk alleles. Gene expression profiles were analyzed in suture tissues collected during surgery. Selective neurocognitive function evaluation was planned at 6 months (T1), 12 months (T2) and 3 years (T3) after the surgical treatment.
Results: At T0 24% patients (20/82) had borderline/lower scores (</=85) in one or more scales; 15 of them showed a specific drop in a single scale, the coordinate o-manual scale being the most frequently compromised. The remaining 5 children showed a more severe profile, with borderline/lower scores in two or more scales.

CT/MR allowed excluding morphological abnormalities of both cortical and white matter structures in all cases. Increased subarachnoid spaces around the cerebral hemispheres was documented in 60 cases (73.1%).
Gene profiling allowed evidencing a specific signature of overexpressed genes.
The neurocognitive evaluation at T1 allowed evidencing borderline/lower scores (</=85) in one or more scale in 8 out of 56 tested children (14.2%), 5 of which showing drops in a single scale. Language function more frequently compromised. The remaining three children (5.3%) showed a more severe profile, with borderline/lower scores in two or more scales.
Conclusions: The persisting drop of neurocognitive functions after surgery in selected children, could be related to neuroradiological and genetic findings. The improvement observed over time in a consistent proportion of cases might be related to the improvement of an initial immaturity of the sensory-motor system.

FP04

Cognitive assessment of children with Crouzon and Pfeiffer syndromes

Giovanna Paternoster, Xavier Dahiez, Leslie Tabuteau, Karine Andre, Michel Zerah, Eric Arnaud (Paris, France)

Aim: Syndromic faciocraniosynostoses are generally believed to be associated with severe cognitive impairement, due to FGFR2-related brain malformations and prolonged episodes of intra-cranial hypertension. Here we intended to assess several specific cognitive tasks in Crouzon syndrome (CS) and Pfeiffer syndrome (PS) patients in order to provide further details on the superior brain functions in these children.
Methods: We included 8 patients with CS (age 8.62±1.25) and 6 patients with PS (mean age 8.18±2.29); 13/14 patients benefited from fronto-facial monobloc advancement (FFMBA) at an early age (2.67±2.52). Specific cognitive tests for executive functions (WISC-IV, WNV, Animal Stroop Test, TEA-Ch, Tower of London Test, Corsi block-tapping test), attentional functions (TEA-Ch) and social cognition (NEPSY-II) were conducted after an initial assessment using Raven's Progressive Matrices (PM-47). A BRIEF questionnaire was filled by the parents of the children before every assessment.
Results: Patients with CS and PS both have satisfactory cognitive abilities, based on the parents reports (BRIEF) and the patients' own assessments. Patients with PS do not have poorer test results than patients with CS: the two main affected functions were divided attention in CS and PS, and cognitive inhibition only in CS.
Conclusion: CS and PS are not associated with poor cognitive prognosis. These two FGFR2-related faciocraniosynostoses are often grouped into one spectrum. Our results tend to show that their cognitive patterns are distinctive, and could indicate that specific rehabilitation programs should be designed for CS and PS in order to focus on their predominantly affected cognitive functions.

FP05

Does early surgery for sagittal synostosis result in better cognitive outcomes?

Maggie Bellew, Rachel Mandela, Paul Chumas (Leeds, United Kingdom)

Introduction: To ascertain whether age at time of surgery has an impact on later developmental attainment for children with Sagittal Synostosis (SS).
Methods: The developmental outcome data from patients who had surgery for SS and who attended for their routine pre-op, 6 months post-op, and five years of age developmental assessments (n=50) and 10-year IQ assessment (n=49) were examined, comparing whether they had surgery at <6 months, 7-12 months or >12 months.
Results: A significant effect for age at time of surgery was identified at 6 months post-op and 10 years, but not at 5 years, in terms of GQ, FSIQ and PIQ.It showed that surgery is optimally undertaken at 6 months or less, with those receiving surgery at 12 months or later performing the least well.
Conclusions: The results of this study suggest that early surgery for sagittal synostosis may improve cognitive outcomes. This also has potential implications for the type of surgery offered, if surgery is to be undertaken before six months of age.

FP06

Single suture craniosynostosis: preoperative and postoperative evaluation of neurocognitive development

Daniela Pia Rosaria Chieffo, Federica Moriconi, Paolo Frassanito, Luca Massimi, Valentina Arcangeli, Massimo Caldarelli, Gianpiero Tamburrini (Rome, Italy)

Objective: A comprehensive neurocognitive evaluation of children with single suture craniosynostosis before and after surgery to define risk factors which might predict neurocognitive development.
Methods: All children with single suture craniosynostosis operated on from January2014 to January 2017 were enrolled in the study. A comprehensive neurocognitive evaluation was performed before surgery (T0) and 6 months after surgery (T1).
Results: 133 patients (mean age: 6 months) were enrolled at T0 (scaphocephaly: 82 (59%), trigonocephaly 35 (25%), anterior plagiocephaly 16 (11%).
100 patients performed a screening 6 months after surgery (T1) (mean age: 14 months), 56 of them affected by scaphocephaly, 29 by trigonocephaly, 15 by anterior plagiocephaly.

At T0, 27.1% of the patients (36/133) had a drop in one or more scales: 24.3% of the patients with scaphocephaly, 37.5% of the children anterior plagiocephaly, 28.5% of the children with trigonocephaly.
20% of the patients evaluated at T1had a persisting drop in one or more scale: 14.3% of the patients with scaphocephaly, 33.3% of the children with anterior plagiocephaly and 3.4% of the children with trigonocephaly.
The most compromised scale at T0 was the one of hand-eye coordination (13 cases), more frequent in patients with scaphocephaly (9/13).
In patients with global impairment (13 cases), the most compromised groups were patients with scaphocephaly (5/13) and trigonocephaly (5/13).
At 6 months the most frequently compromised scale was the scale of language (4 cases), more frequently in children with scaphocephaly (3/4 cases). Children with anterior plagiocephaly had more frequently a persistence of eye-coordination defects.
Among patients with globally impaired profile (11 cases) the most frequently compromised were children with trigonocephaly (6/11) and scaphocephaly (3/11).
Conclusions: The most frequent persisting specific drop at 6 months concerned language in children with scaphocephaly. If global neurocognitive delay was persisting 6 months after surgery it was more frequent in patients with scaphocephaly and trigonocephaly.

FP07

Neurodevelopmental aspects of non-syndromic craniosynostosis

Dawid Larysz, Dowgierd Krzysztof, Agnieszka Rożek, Patrycja Larysz, Marek Mandera (Gliwice, Poland; Olsztyn, Poland; Katowice, Poland)

Introduction: Non-syndromic craniosynostosis could be associated with developmental problems including improper speech acquisition, dyspraxia, learning disabilities and behavioral problems. Many hypotheses were proposed to explain aforementioned phenomena, concerning associated intracerebral anomalies, abnormal shape of brain in malformed skull and local intracranial hypertension with local hypoperfusion. Aim of the study was prospective analysis of frequency and type of neurodevelopmental problems in children with non-syndromic craniosynostosis and attempt to find factors that could influence their development.
Methods: Studied group consisted of 300 children with non-syndromic craniosynostosis. Inclusion criteria consisted of: age before one year, confirmation of CSO in CT examination, absence of associated brain malformations and absence of other skull and other congenital anomalies. Developmental examinations were performed by team consisted of pediatric neuropsychologist, speech therapist, neurosurgeon and occupational therapist. We used Polish version of MFED and DSR tests, neuropsychological and speech evaluation, before surgery, than each 3 months until 2 years, and then each 6 months. Mean follow-up was 6 years (ranged from 3 to 10 years).
Results: Proper development was found in unoperated children in 57,1% of children before 6 months of age and 50,0% before 1 year of age. 6 months after the surgery proper development was found in 78,57% of children, in 80,0% (12 months after the surgery) and in 86,67%. The most frequent were speech acquisition problems and dyspraxia. Unoperated children with sagittal synostosis have more frequent developmental problems before 6 months of age than with metopic or unicoronal synostosis. In most children we found problems with polish speech pronunciation, and slight deficits in muscle strength and symmetry.
Conclusion: Developmental problems affect more than 45% of children with non-syndromic craniosynostosis preoperatively. The most frequent abnormalities are speech acquisition delay and motor development abnormalities. The etiology of developmental problems seems to be multifactorial. Abnormal morphology of the speech apparatus and deviations of the center of gravity of the head could play important role in developmental deficits.

Abstract session 4: Trauma & Miscellaneous

FP08

Can artificial intelligence predict the necessity of CT scan for infants with minor head injury?

Tadashi Miyagawa, Mariko Yabuki, Yoshiyuki Watanabe, Koichi Tamaki, Hirohide Karasudani, Akira Yamaura (Matsudo, Japan)

Objectives: For the management of minor head injury (mHI) in children, we previously published data regarding applicability of the PECARN rule for infants with mHI in Japan and proposed quantification methods type II as a new simple clinical decision rule. These methods could be satisfactory in clinical settings, however a more high-quality method has been expected. The purpose of this study was to assess the feasibility of artificial intelligence (AI) to predict the necessity of CT scan for infants with mHI.
Methods: From 2005 to 2014, 1091 infants (younger than 2 years) with mHI were enrolled in this retrospective study. Clinical data were analyzed using decision tree analysis (DTA) in AI to predict the necessity of CT scan for infants with mHI.
Results: Machine learning with DTA was able to create a learning algorithm for AI to predict the necessity of CT scan for infants with mHI. Variables which were included in this DTA were scalp hematoma, alternation of consciousness, number of vomiting, Glasgow Coma Scale, age, gender,

LOC duration, and not acting normally per parent. The machine learning with DTA had an accuracy of 96.1%, a negative precision value of 97.0%, a positive recall of 98.8%, and a negative F-measure of 98.1%. Those values showed this learning algorithm for AI could be used to predict the necessity of CT scan for infants with mHI.
Conclusions: The learning algorithm for AI created in this study would be better to identify infants who need CT scan after mHI. This study indicated AI would be able to predict the necessity of CT scan for infants with mHI.

FP09

MRI utility in C-spine clearance following pediatric trauma

Michael Handler, Krista Greenan, David Mirksy, Graber Sarah, Noah Hubbell, Nicholas Stence (Aurora CO, USA)

Introduction: Numerous protocols have been introduced to clear the cervical spine (C-spine) in pediatric trauma patients, including use of MRI for C-spine clearance in patients who cannot participate in a neurological examination. In the adult literature, patients with negative CT C- spine imaging have a very low incidence of an undetected spine injury. In children, who may theoretically have a higher risk of traumatic ligamentous injury, the risk of a C-spine injury with a negative CT scan is unknown.
Methods: All trauma patients treated at a Level I Pediatric Trauma Center from 1995 to 2015 were retrospectively reviewed for the details of their injury, their C-spine clearance and imaging. Patients with both CT and MRI of the C-spine were included Imaging studies were interpreted by two pediatric neuro-radiologists and validated for concurrence. Imaging characteristics were tabulated independently, and findings were compared between CT and MRI.
Results: 223 patients the met inclusion criteria. 159 (71%) had a normal CT. No patient with a normal CT had an unstable injury demonstrated on MRI. Patients with normal CTs who had MRI changes had increased T2 signal in the interspinous ligaments and soft tissues, but no significant ligamentous disruption or disc herniation requiring intervention. 37 patients (17%) had instability demonstrated on both studies and required intervention.
Conclusions: Pediatric patients who have undergone a CT of the cervical spine that is normal will not benefit with the addition of a cervical MRI. Routine use of MRI for C-spine clearance will not improve patient safety by detecting occult significant injury even in patients who cannot be cleared clinically.

FP10

The predictive validity of the Glasgow Coma Scale vs "Glasgow Outcome Scale Extended Pediatric" and "Kings Outcome Scale for Children with Head Injury"

Andrew Croall, Shona Forsyth, Emer Campbell, Meharpal Sangra, Anthony Amato-Watkins, Lorraine Todd, Roddy O'Kane (Glasgow, United Kingdom)

Introduction: A universally accepted method of measuring traumatic brain injury (TBI) outcome in paediatric populations has yet to be established. Two outcome scales, "Kings Outcome Scale for Children with Head Injury" (KOSCHI) and the "Glasgow Outcome Scale extended Pediatric" (GOSeP) have been tailored to children and young adults. We sought to investigate their utility on paediatric patients admitted with head injury at our institution.
Methods: A retrospective case note review of 120 patients with TBI admitted to our institution was performed between March 2012 and March 2017. Demographic details and initial Glasgow Coma Scale ((GCS) (<6 hours post-injury)) was recorded. Where possible a KOSCHI and GOSeP was calculated from outpatient clinic follow-up letters.
A telephone interview was conducted with patient carers to determine KOSCHI and GOSeP. The predictive validity of each scale was determined via analysis of correlation of GCS severities to outcome scale categories.
Results: GCS was available in 82 patients: 60 minor (13-15), 9 moderate (9-12), and 14 severe (<9) TBI's. There were 54 males and 28 females. Age at injury ranged from 1-month to 188-months-old (Median age ±1SD)(62.5±58).
A KOSCHI and GOSeP score could be determined in 47 patients from case notes recorded >3-months post-TBI. Overall, KOSCHI and GOSeP correlation to GCS scores was determined, as were subgrouping into age groups (<2, 2-5, and >5-years-old). Overall and subgroups correlation coefficients showed minimal to no correlation.
41 telephone interviews of patients >6 months post-TBI were successfully performed. KOSCHI (r_s=0.119 (p=0.229)), GOSeP (r_s=-0.109 (p=0.250)), and age subgroups of both scales showed poor levels of correlation.
Conclusion: Neither overall or subgrouping by age of KOSCHI or GOSeP Outcome Scores provided any meaningful correlation to GCS Scores recorded within 6-hours post-TBI. Thus, GCS could not appropriately predict outcomes for either scales across the age ranges of pediatric TBI.

FP11

Pediatric cerebral abscesses: a single UK center experience

Samuel Hall, Shirley Yadu, Benjamin Gaastra, Nijaguna Mathad, Ryan Waters, Aabir Chakraborty, Vassilios Tsitouras (Southampton, United Kingdom)

Introduction: Cerebral abscesses are relatively less common in the pediatric population compared to adults. The mainstay of treatment is surgery with prolonged administration of antibiotics, although significant variations regarding the optimal

management still exist. The aim of this study was to investigate the epidemiology, treatment and outcomes of our centre's pediatric cerebral abscess workload.

Methods: Patients were identified from hospital records (2003-2017) using ICD10 code G06.0 and the results screened for age and pathology. Only patients <18 years old with a cerebral abscess were eligible. Case notes were reviewed for abscess location, microbiology results, types of surgical intervention, and outcome using the Glasgow Outcome Score at 3 months.

Results: Twenty-two patients were identified (mean age 7.0 ±5.4years, male n=12) over the 15-year period. The most common location was the frontal lobe (n=10). Twelve patients (55%) had an identifiable source including; ENT infections n=2, dental work n=2, meningitis n=2, pulmonary aspergilloma n=1 and iatrogenic following ICP bolt insertion n=1. Streptococcus was the most common organism (n=14) with the remainder comprising a mixture of Gram+ve or Gram–ve organisms. All patients underwent surgery with 18 patients having a burr hole aspiration of which 9 required at least one repeat aspiration (total 30 aspirations; 1.66/patient) and the other 4 patients having a craniotomy and excision. Neither age, location nor organism were associated with a higher rate of re-aspiration. Of the 19 patients with available follow-up data, 18 returned to GOS 5 and one patient was GOS 4.

Conclusion: Pediatric cerebral abscesses are an uncommon diagnosis however most have a preceding infection and Streptococcus is commonly identified as the causative organism so antimicrobial therapy should be chosen accordingly. All of our patients underwent surgical intervention and received intravenous antibiotics with a favourable outcome. Parents should be counselled to expect more than one aspiration.

FP12

Brain abscess in children: predisposing factors, management and outcomes

Vianney Gilard, Kevin Beccaria, Syril James, Thomas Blauwblomme, Giovanna Paternoster, Timothée De Saint Denis, Marie Bourgeois, Stéphane Blanot, Stéphanie Puget, Dominic Thompson, Michel Zerah, Martin Tisdall (Rouen, France; Paris, France; London, United Kingdom)

Introduction: Brain abscess is a rare but life-threatening infection in children. Presenting symptoms may be non-specific and can lead to delayed diagnosis. The aim of our study was to better characterize clinical presentation, management and outcome in infants and children with brain abscesses.

Methods: The authors conducted a retrospective multicenter study over a 25 years period (1992-2017). During this period, 116 children and 28 infants (age< 1 year) with brain abscess were treated in 2 national reference centers. The following data were assessed: clinical presentation, predisposing factors, radiological data, microbiology results , management and outcome.

Results: Mean age at diagnosis was 102.3±54 months in children and 2.5±2.4 months in infants. Predisposing factors were different between age groups with a predominance of meningitis in infants in 64% of cases versus 3% in children ($p<0.01$). Concerning clinical presentation, fever and meningism were more frequent in infants ($p<0.01$). On cerebral imaging, brain abscesses were hemispheric in 81% and deeply located in 11%. 115 patients were treated with aspiration, 11 with excision and in 18 patients; aspiration was combined with instillation of topical antibiotics. Re-operation was required in 29 children versus 1 infant ($p<0.01$). In our cohort, the mortality rate was 4%. At 3 months follow-up, the outcome was favorable in 86% of children versus in 68% of infants ($p=0.03$).

Conclusion: We report outcomes of children with brain abscesses in a large cohort from two level 1 pediatric centers. There seems to be a dichotomy between children and infants in terms of predisposing factors and outcome. Despite an improvement in its management, brain abscess in pediatric population remains a serious condition. Predisposing factors must be sought and treated to prevent from recurrence.

FP13

Pediatric Neurosurgery malpractice claims in Germany

Thomas Beez, Beate Weber, Hans-Jakob Steiger, Sebastian A. Ahmadi (Düsseldorf, Germany)

Objective: Limited data on malpractice claims is available for pediatric neurosurgery. Aim of this study was to analyze malpractice claims faced by pediatric neurosurgeons in a large German Medical Council coverage area, representing 60,000 physicians and 10 million inhabitants.

Methods: We analyzed malpractice claims in neurosurgical patients aged 18 years and younger that were completed by the arbitration board of the North Rhine Medical Council from 2011 to 2015, in relation to overall number of neurosurgical procedures. Claims were categorized into cranial, spinal and neuro-interventional. Arbitration decisions were categorized into treatment-associated damage *with* medical error, treatment-associated damage *without* error or disease-related outcome. Severity was graded from negligible (grade 1) to death (grade 6).

Results: From a total of 8,381 malpractice claims, seven concerned Pediatric Neurosurgery. Median age was 12 years (range 1-17). Claims were related to cranial (N = 5), spinal (N = 1) and neuro-interventional (N=1) procedures. Surgical cases comprised three ventriculoperitoneal shunt (VPS) operations, two cranioplasties and one spinal fusion after trauma. During the study period, 4,500 VPS operations, 1,484 cranioplasties and 84 spinal fusion operations were performed in children in the coverage area, accounting for malpractice

claim rates of 0.07%, 0.13% and 1.19%, respectively. Damage was treatment-associated with medical error in one and treatment-associated without error in six cases; thus, all claims occurred in cases with complications. Severity was grade 2 (transient minor) in three, grade 3 (transient major) in one, and grade 5 (permanent major) in three cases.
Conclusions: Median malpractice claim rate was 0.13% of pediatric neurosurgery cases. While treatment-associated damage was confirmed in all cases, erroneous treatment was found in only one. The most common scenario leading to a malpractice claim was a CSF diversion procedure with complicated course, likely reflecting the high volume of hydrocephalus in pediatric neurosurgery.

Abstract session 5: Craniofacial III

FP14
Cephalic measurements on 3D photogrammetry after spring-assisted correction for scaphocephaly

Maik Tenhagen, Katya A.L. Mauff, Irene M. Mathijssen, Marie-Lise C. van Veelen (Rotterdam, the Netherlands)

Introduction: Scaphocephaly, caused by the premature fusion of the sagittal suture, characteristically presents itself with an elongated head, bitemporal narrowing, frontal bossing and a prominent occiput. Spring-assisted correction is a relatively new surgical technique to correct scaphocephaly. Advances in 3D photographic technology could provide help in analyzing postoperative outcome following spring-assisted correction. This study aimed to examine the utility of 3D photogrammetry for evaluating surgical results by performing cephalometric measurements on a large cohort of scaphocephaly patients.
Methods: The 3D photogrammetric images of 81 patients (72 male, 9 female) were collected between March 2013 and August 2016.Volume, cephalic index and head circumference were derived from the 3D scans. Volume changes were analyzed over time. The head circumference and cephalic index acquired from 3D scans were compared to age-matched measurements performed manually and on x-ray respectively. The intra- and interrater reliability for calculating the volume were determined. Statistical analysis was performed with the use of models correcting for repeated measurements.
Results: Mean 3D follow-up was 1.2 years (range 0.25-6.9 years). A curve fitted to the observed data shows a mean volume approximately 2SD above the corrected average volume in the normal male population. Due to limited data, no statements could be made for the female population. Excellent correlation was achieved in the comparison of head circumference ($r = 0.947$) and cephalic index ($r = 0.835$) on 3D scans as opposed to those measurements performed manually and on x-ray respectively.
Conclusion: This research provides further insight into the volume development up until 4 years after spring-assisted correction. Postoperative volume was overall larger than in the normal population. Performing cephalometric measurements on 3D photogrammetry proved to be a good alternative for measurements performed manually or on x-ray.

FP15
Radiation-free 3D head shape and volume evaluation after endoscopically assisted strip craniectomy followed by helmet therapy for scaphocephaly

Guido de Jong, Jene Meulstee, Erik van Lindert, Wilfred Borstlap, Thomas Maal, Hans Delye (Nijmegen, the Netherlands)

Objective: Post-operative follow-up in craniosynostosis is still mainly done using radiation techniques. Sequential radiation-free follow-up techniques (e.g. 3D stereophotogrammetry) are hindered by the lack of consistent markers like bony landmarks often restricting evaluation to subjective comparison. However, using the computed cranial focal point (CCFP), it is possible to perform correct sequential image superposition and objective evaluation. We used this technique for mean volume and 3D shape change evaluation of the head based on 3D Photos after endoscopically assisted scaphocephaly surgery with helmet therapy.
Methods: We performed a mean head 3D shape and volume evaluation on age grouped 3D Photos (n=176) of children that underwent endoscopically assisted scaphocephaly surgery with helmet therapy. We used CT scans (n=96) of age grouped children as reference for comparison. We performed a mean 3D shape evolution analysis and calculated both the volume and cephalic index (CI) over time.
Results: The mean volume followed the reference group with deviations at the time of pre-surgery. The mean CI was initially 69.5% and increased to 77.0% at around 9 months from where it gradually declined to between 72-73% from hereon. The 3D head shape showed the highest amount of growth in the parietal area especially in the first few months after surgery. There were minor differences in both the total volume and posterior to total volume ratios between the craniosynostosis group and healthy references.
Conclusion: Using a novel technique we were able to objectively evaluate 3D head shape, volume and CI using stereophotogrammetry after endoscopically assisted scaphocephaly correction. The most prominent 3D shape change was around the surgical site and the CI showed initial increase post-surgery with some decrease over time. Volumes showed minimal differences compared to the reference group.

FP16
Use of Oral Device in stabilization of facial advancement after early remove of Osteodistraction device for postoperative sequelae

Giulio Gasparini, Michela Perina, Camillo Azzuni, Luca Massimi, Gianpiero Tamburrini, Sandro Pelo (Rome, Italy)

Introduction: The risk of relapse in children undergoing craniofacial advancement is very high; it is more serious in patients treated with osteodistraction because of the absence of rigid internal fixation systems. Our standard therapeutic protocol includes a stabilization period of 120 days after the last activation of the osteodistraction device.
When the removal of the distraction device is needed before the fixed term, it is often impossible to reposition a new fixation system. This complication might dramatically increase the relapse rate of the malformation because the regrowth bone is immature.
The alternative we propose in these cases is to use an oral device designed to help the splanchnocranium not to lose its advanced position.
Technical Note: We called the oral device "Maxillary Advancement Contention (MAC)". It is a functional activator with dental retention. It has an Adam's hook on last erupted on both side and an occlusal plane in acrylic resin to prevent eruption and mesialization of the lower primary teeth. A vestibular arch is inserted on its frontal side in order to stop the advancement of lower incisors. The device is implanted immediately after the removal of the distraction device and is left in place for at least 3 months.
Discussion: In children with faciocraniostenosis functional complications might force to anticipate the time of craniofacial advancement. The early age of surgery in these children increases the risk of early mobilization of the osteodistraction device. The MAC system might help, with a minimally invasive procedure, to maintain the obtained advancement allowing the stabilization of the regrowth bone.

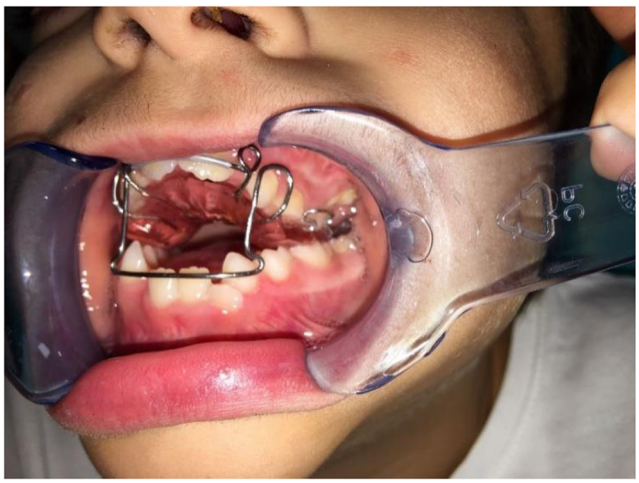

Figure 1: *Oral Device*

FP17

Three-dimensional preoperative virtual planning and template use for surgical correction of unicoronal and metopic craniosynostosis

Mariana Alamar, Joaquin Andermatte, Santiago Candela, Gemma Garcia, Antonio Guillén, Patricia Puerta, Jordi Muchart, Enrique Ferrer (Barcelona, Spain; San Sebastian, Spain)

Introduction: Surgical correction of craniosynostosis of the anterior cranial vault deformities relies most of the time on the surgeon's vision to estimate the shape of a normal forehead and orbital rim. Computer-aided design/computer aided manufacture (CAD/CAM) techniques can be used to create a preoperative planification to guide the osteotomies and the placement of the bone segmentsto achieve a more precise correction. The aim of the study was to compare two groups of patients, one with virtual surgical planning and the other one with the standard technique.
Methods: We compare 5 patients (3 coronal and 2 metopic craniosynostosis) who underwent unilateral and bilateral fronto-orbital advancement using CAD/CAM preoperative surgical planning and 10 patients (6 coronal and 4 metopic) intervened by conventional technique to measure surgical outcomes. Anthropometric measures were taken in CT scans before surgery and 6 months after surgery. Other variables such as blood loss and operative time were taken.
Results: Patients treated with CAD/CAM preoperative surgical planning showed greater correction than the group treated with the standard technique. Both in the immediate postoperative period and in 6-month follow up. There is also a reduction in surgical time and blood loss.
Conclusion: The use of virtual surgery and prefabricated cutting guides allows for a more precise and rapid reconstruction. It can improve the safety and the aesthetical results.

Abstract session 6: Craniofacial IV

FP18

Ultrasound examination of the cranial suture as a method for diagnosing craniosynostosis in children

Albert Sufianov, Olga Sadykova, Iurii Iakimov, Galina Sufianova, Rinat Sufianov (Tiumen, Russia)

Introduction: Computed tomography with 3D skull reconstruction, which is recognized as the "gold standard" in the diagnosis of craniosynostosis, has a significant drawback associated with radiation exposure, which can be significant for an intensively growing brain at the age of 6-12 months. The use of ultrasound in the visualization of the seams of the skull is a possible research method that

replaces computed tomography. The purpose of the work is to illustrate the possibility of applying an ultrasound examination method for visualization of the cranial sutures in the diagnosis of craniosynostosis.

Materials and Methods: An analysis of the ultrasound of the skull sutures in 45 children (31 boys and 14 girls) at an average age of 4.5±1.2 months was performed (from 1.5 to 12 months), which were examined and treated at the Federal Center of Neurosurgery, Tiumen, Russia. Ultrasound examination of the skull sutures was performed using the Toshiba Aplio 500 expert class device (TUS-A500) using a linear sensor for scanning (PLT-1005BT) with a frequency of 10 MHz and a 58 mm aperture with a water kit (Toshiba Water Bag Kit) and a scanning depth of 2 cm (Figure 1). The use of an aquatic environment made it possible to strengthen the acoustic window and improve the visualization of the seam. To assess the condition of the cranial sutures, we developed a simple ultrasound scheme, according to which the study was performed. Scanning of the cranial suture was carried out all along its length in a plane perpendicular to the corresponding seam in the sequence from the coronary suture on the right and left to the lambdoid suture on the right and left to the sagittal suture and to the metopic suture.

Results and Discussion: According to the data of ultrasound examination of the suture of the skull, 14 children had positional posterior and anterior plagiocephaly (5 girls and 9 boys), 12 children - trigocephalus, 18 cases - scaphocephaly and 1 child combined craniosynostosis - combination of defeat of the metopic and right side of the coronary suture of the skull.

Diagnostic visualization allows you to confirm the craniosynostosis of a particular suture and to evaluate the deformity of the skull characteristic for each of them. Ultrasonographic signs of craniosynostosis: 1) absence of hypoechogenic continuous linear space; 2) loss of dentition and unevenness of the internal suture margin; 3) asymmetric fontanels; 4) loss of slope of the edge; 5) the presence of a bone protrusion or roller; 6) the presence of a "transition zone" in the incomplete form (subtotal) of craniosynostosis.

Important for a neurosurgeon for planning treatment is the accuracy of localization of the site of premature penetration of the suture, bone thickness, deformation of the osseous crest, connection with venous sinuses, emissary veins. The use in the narrative of the ultrasound protocol of this classification, in our opinion, greatly facilitates preoperative surgical planning and dynamic observation in the postoperative period.

Conclusion: The ultrasound of the cranial sutures can be considered as an alternative CT of the skull method of diagnosing craniosynostosis, which can also be used as a screening test for suspected craniosynostosis in a child.

Figure 1: *Probe*

FP19

Prevalence and severity of positional plagiocephaly in children and adolescents

Valeria Blé, Alexandru Szathmari, Pierre Aurelien Beuriat, Federico Di Rocco, Laura Nanna Lohkamp, Pierre Antherieu, Carmine Mottolese (Lyon, France)

Introduction: Though Positional Posterior Plagiocephaly (PPP) is common in infants since the pediatric recommendations of "Back to Sleep" and several aspects of its natural history remain unclear. The aim of this study is to understand the actual prevalence and severity of PPP in unselected pediatric population.

Methods: Head CT scan performed for head trauma from September 2016 to September 2017 were retrospectively analyzed in 165 children ranging from 0 to 18 years of age (101 boys).

Cranial Vault Asymmetry Index (CVAI) was calculated at the level of the superior orbital rim and 3,5% was considered as threshold of asymmetry. The results were analyzed according to different age: Group I: 1month to 12 months of age (37 patients), group II: 2 to 4 years (32), group III: 5 to 8 years (36), group IV: 9 to 12 (27), and group V: 13 to 18 years (33) and severity of asymmetry: mild group (CVAI range: 3.5-7%), moderate group (CVAI range:7-12%) and severe group (CVAI>12%).

Results: The total prevalence of PPP was of 25%. The prevalence in Group I was estimated to be 40,5%, 15,6% in Group II, 30,5% in Group III, 18,5% in Group IV and 12% Group V.

The degree of deformation varied from 3.5% and 15.09%, most children have a mild asymmetry. The degree of the asymmetry varied according to the age groups but moderate asymmetry could be found at all ages even in Group IV and V. One child (Group II) presented a severe asymmetry.
Conclusion: This study analyses the PPP in an unselected unbiased pediatric population, shows that PPP has still a high prevalence in adolescents. It confirms that the prevalence of deformational plagiocephaly is more common than usually reported and that PPP persists at a late age.

FP20
Examining the use of resorbable plates in cranial vault reconstruction in 182 pediatric patients: a retrospective cohort study
Frank Reilly, John Caird, Dylan Murray (Dublin, Ireland)

Background: Resorbable fixationis preferable over non resorbable plates and screws in paediatric cranial vault procedures particularly in the management of craniosynostosis (CS). Few studies estimated the prevalence of complications nor characterising patient and surgical factors associated with complication incidence. Our objective was to determine the prevalence of complications associated with the use of Inion resorbable plates in paediatric patients undergoing cranial vault reconstruction and examine patient and surgical factor which may influence complication incidence.
Methods: A retrospective study of 182 patients who underwent cranial vault reconstruction using resorbable plate fixation under a single operating Surgeon from 2008-2016 at a the National Paediatric Craniofacial Centre (NPCC) at the Children's University Hospital in Temple Street (CUH). Complications were identified from a prospectively maintained database and medical note review, and categorised on the basis of whether the plate contributed to the development of the complication. A number of key patient characteristics and surgical variables were also recorded. and examined for statistical associations with complications.
Results: Twenty-two patients (12.1%) experienced a complication of which 8 cases were fully attributable to the inserted plate and a further 5 cases where the plates partially contributed to the complication. Males were less likely to develop a complication however in stratified analyses excluding patients older than 7 years and those with an underlying genetic syndrome (which may represent clinically different subpopulations), increasing age and lower weight but not gender were associated with complication incidence.
Conclusion: Our complication rate using Inion plates has an equivalent safety profile to other fixation devices reported in the literature. Gender was the only factor significantly associated with complication development, however in restricted samples excluding firstly older patients and secondly those with an underlying genetic syndrome, increasing age and lower weight were more important factors. No surgical factors were identified to be associated with complication incidence such as the number of plates inserted – however volume of transfused red blood cells may possibly be associated complications, requiring further study in larger samples.

FP21
Anterior skull base and pericranial flaps ossification after frontofacial monobloc advancement
Giovanna Paternoster, Anne Morice, Anne Ostertag, Syril James, Martine Cihen-Solal, Hossein Khonsari, Eric Arnaud (Paris, France)

Introduction: Frontofacial monobloc advancement surgery (FFS) creates retrofrontal dead spaces and a communication between the anterior cranial fossa and nasal cavities. To tackle with this issue, we usually perform trans-orbital double pericranial pedicled flaps (PF). The aim of our study was to evaluate the post-operative ossification of the anterior skull base and of the PF following FFS, and to identify factors influencing this ossification.
Methods: Measurements of the skull base only (SB) and of the ossified PF together with the SB (SB-OPF) were performed on CT-scan at nasofrontal (NF) and (2) nasoethmoïdofrontal junctions (NEF). The total thickness of the skull vault was measured and plotted using the Wall Thickness Analysis module of VGStudio Max 3.0 (Volume Graphics, Heidelberg, Germany) and a qualitative defect score (DS) for the SB was computed.
Results: Twenty-two patients operated at 3.1 years ±1.6 were included: 14 with Crouzon syndrome (CS), 5 with Pfeiffer syndrome (PS) and 3 with Apert syndrome (AS). On immediate post-distraction CT-scans (median post-operative period of 23.4 days ±10.6), PF and distraction gaps were not ossified. SB was thinner for patients (all syndromes alike) than in controls at the NEF point only ($p=0.02$). One- and five-year post-operative CT-scans were performed after median post-operative periods of 1 year ±0.17 and 5 years ±1.1, respectively. The distraction gap was completely ossified in the midline anterior SB in all patients at these two timepoints. SB-OPF was thicker in patients at these two timepoints ($p<0.005$ and 0.02). Patients with PS had significantly thicker SB and OPF thicknesses ($p=0.01$ and 0.03) and lower DS than patients with CS or AS ($P=0.03$) at one-year post-operatively.
Conclusion: Our results show ossification of PF and total reossification of the anterior SB midline in all patients. We thus recommend performing pericranial pedicled flaps in FFS in order to potentialize reossification of anterior SB.

FP22
Fronto-orbital advancement and reconstruction using reverse frontal bone graft without using the removed orbital bar as part of the reconstruction

Asim Sheikh, Moritz Schramm, Paula Carter, John Russell, Mark Liddington, Paul Chumas (Leeds, United Kingdom)

Aims: FOAR is the standard treatment for correction of metopic and coronal synostosis. There are many techniques described to achieve this, but no standardized technique is applicable for all FOARs. We have attempted to standardize our FOAR by using a technique of reverse frontal bone graft for FOAR and using the removed orbital bar for bone dust for on-lay. New orbital rims are created in the inferior surface of the reversed frontal bone and the length of the reversed frontal bone is used to create width to deal with the temporal "thinning".
Methods: Since April 2014, 13 patients have been operated using this technique. A standard frontal bone graft and orbital bar are marked out and removed. The frontal graft is then reversed, and supra orbital margins are created on the "new" inferior surface. The reversed frontal graft is then placed in the desired location (neutral for metopic and advanced for coronal synostosis be that uni or bilateral) and secured to the nasion by absorbable sutures, and laterally, by absorbable plate and screws. The orbital bar is then drilled down to make bone dust which is used to fill gaps and for on-lay in the temporal region and on exposed dura.
Results: Over the years, our technique for FOAR (Marchac template and then reversed frontal graft) has evolved but has always utilized the orbital bar as part of the reconstruction. This evolution is a reflection of no technique successfully managing the various competing aims and the differences encountered between cases and diagnoses. The method described here allows for a more standard technique and the 13 patients who have undergone this approach to date have had good cosmetic results with no specific complications.

FP23

Analysis of craniosynostosis cases treated with posterior cranial vault distraction and frontal orbital advancement/remodeling

Daiki Senda, Kazuaki Shimoji, Doruk Orgun, Masakazu Miyajima, Hiroshi Mizuno, Yuzo Komuro (Tokyo, Japan)

Introduction: Craniosynostosis patients with shortened occipitofrontal diameter are mainly treated with posterior cranial vault distraction osteogenesis (PVDO) in our institution. If further intracranial volume (ICV) expansion is needed, additional treatment with frontal orbital advancement (FOA) is done. On the other hand, frontal orbital remodeling (FOR) is done for better aesthetic results. In this study, we have investigated post-treatment ICVchanges in patients with craniosynostosis with the aforementioned methods.
Methods: Patients who underwent FOA or FOR in addition to PVDO at Juntendo University Hospital between 2011 and 2017 were reviewed. Patient characteristics, length of distraction, ICV, and pre- and postoperative CT scan findings were evaluated. ICV was measured using Mimics$^{\circ R}$ on 3DCT. Morphological changes over time were also assessed.
Results: Nine patients (5 male, 4 female) aged from 5 months to 6 years 8 months (mean 26.6 months) at the time of PVDO were reviewed. For PVDO, the distraction length was 18 to 38mm, the ICV change was 100 to 281 mL, and the enlargement ratio of ICV was 109% to 134%. 2 patients were further operated with FOA for ICV expansion while the remaining 7 with FOR for aesthetic improvement.
With FOA, ICV change was 73 to 120 mL while enlargement ratio of ICV was 108% to 114%. With FOR, ICV change was 27 to 77 mL while enlargement ratio of ICV was 104% to 107%.
Conclusion: In patients with severe deformity craniosynostosis, PVDO is performed first in order to expand the ICV and decompress the posterior cranial fossa. Furthermore, FOA is performed if any extra increase in ICV is needed. This approach seems to enable larger expansions of ICV compared with other conventional methods. Furthermore, we think that FOR should be reserved for patients in whom adequate ICV levels are achieved with PVDO yet frontal reshaping is necessary.

FP24

Endoscopic endonasal skull base surgery for 40 pediatric cases: Ankara University experience

Gokmen Kahilogullari, Cem Meco, Suha Beton, Murat Zaimoglu, Hazan Basak, Agahan Unlu (Ankara, Turkey; Salzburg, Austria)

Objective: To discuss the endoscopic endonasal approach in pediatric age group with its advantages and disadvantages.
Methods: Retrospective analysis of 40 pediatric patients that are operated via endonasal endoscopic approach for various indications except angiofibroma in Ankara University Medical Faculty between years 2010 to 2016 December.
Results: 25 patients (62.5%) were male and 15 patients were female (37.5%). The main age was 10.4 (2-18). Twelve of the cases were craniopharyngioma (30%), 8 pituitary adenoma (20%), 4 traumatic CSF rhinorrhea (10%), 4 meningocele (10%), 2 germinoma (5%), 2 malign tumors (5%), 2 pituitary inflammation (5%), 1 odontoidectomy (2%), 1 fibrous dysplasia (2%), 1 hemangiopericytoma (2%), 1 neurocytoma (2%), 1 dermod cyst (2%), 1 fibrous dysplasia (2%) and 1 capillary hemangioma (2%). Among 27 patients with pathological results; total or gross total excision was achieved in 23 (85%), subtotal resection was achieved in 2 (7%). In 2 cases of malign tumors (7%) only biopsy was made. Either traumatic or due to tumor resection, all dura lesions along the skull base was reached with endonasal endoscopic approach and watertight sealing was achieved. No patient had a postoperative CSF leak or meningitis. 9 patients had transient diabetes insipidus, 1

patient had temporary loss of lateral gaze and the case after odontoidectomy had pneumocephalus one week after surgery following a sneeze attack. One patient was died because of endocrinological failure and hypo-hypernatremia imbalance.
Conclusion: Endonasal endoscopic approach is an effective method for surgery in managing various pathologies of the pediatric age group. Due to its less invasive nature, it protects the developing bony structures of the face and the skull, while achieving satisfactory outcomes. Nevertheless narrow transnasal corridor as well as inadequate sphenoid sinus pneumatization could be the main handicaps of this approach in pediatric patients.

Abstract session 13: Dysraphism II & Spine

FP25

Comparison of percutaneous minimally invasive fetoscopic surgery and open fetal surgery: single center experience

Huseyin Canaz, Ibrahim Alatas, Ayten Saracoglu, Ali Gedikbasi (Istanbul, Turkey)

Myelomeningocele (MMC), one of the most common congenital malformations, can result in severe lifelong disabilities, including paraplegia, hydrocephalus, Chiari II malformation, incontinence, sexual dysfunction, skeletal deformations and mental impairment. MMC was the first nonlethal anomaly to be treated by fetal surgery. As a result of developments in 3 decades, fetal surgery took its place in guidelines. There are still two different techniques for fetal MMC repair. Fetoscopic technique is used in Germany, Spain, Brasil and Poland. Open fetal surgery is used in USA, Switzerland, Poland, Belgium, Brazil and Turkey. These two techniques also contain some variations in different countries.
We performed percutaneous minimally invasive fetoscopic surgery in 4 cases in 2015 and open fetal surgery in 3 cases in 2017 in Istanbul Bilim University Florence Nightingale Spina Bifida Center. At this point we are the only center who have experience about both techniques. Although short term results are similar in both techniques, there are some differences in surgical management, anesthesiology, postoperative and postnatal care.
Our aim is to compare both techniques and share our experience.

FP26

Open spina bifida in Ireland: neurosurgical management over 9 years

Maria Nunez Sayar, John Caird, Jane Leonard, Darach Crimmins (Dublin, Ireland)

Introduction: Children's University Hospital Temple Street is the National Paediatric Neurosurgery Centre (NPNC) and has managed all new patients born with open myelomeningocele (MMC).
Methods: Prospective collection of clinical data and imaging on all children born 2008-2016 with MMC referred to NPNC.
Results: A total of 185 newborns with MMC were referred.
85% of MMCs were diagnosed antenatally. Over 90% were born at term, with normal birthweight and normal Apgars. There is a larger number of higher lesions compared with similar international cohorts.
All bar 4(2%) of the lesions were closed surgically, 50% within 48 hours of delivery. Later closure is associated with a higher rate of shunt infection and meningitis. 78% of children required a VP shunt. Shunt rates are lowest in sacral lesions (50%). There is a high rate of shunt revision (57% of patients) and infection (decreasing in latter years). Associated Chiari 2 features are universal in these patients, but symptoms are rare. Mobility is lesion-level dependent; with all sacral-level lesion patients that are alive after 3 being independently mobile as opposed to only 3% of those with thoracic lesions. Cognitive function is generally good. 95% of children alive at 3 years are normal or have only mild intellectual difficulties. Seven (4% of children alive at follow up) have bladder continence.
Ten patients have died (5%) mostly due to brainstem related issues.
Conclusion: Myelomeningocele remains a prevalent condition in Ireland. It is associated with need for multiple surgeries and hospital stays. Shunt rates are high with ongoing issues with malfunction and infection. Half of children are wheelchair users after 3 years of age, but cognitive function is good. Virtually all children are incontinent. Mortality rates are low with modern aggressive management and less than similar historical cohorts.

FP27

Post-natal management of myelomeningocele: long term outcome with a multidisciplinary team experience

Alexandru Szathmari, Pierre-Aurélien Beuriat, Isabelle Poirot, Frederic Hameury, Christophe Rousselle, Isabelle Sabatier, Federico Di Rocco, Carmine Mottolese (Bron, France)

Introduction: Myelomeningocele is a complex anomaly. Multidisciplinary management is mandatory. We report the long-term outcome of a single centre experience with a systematic multidisciplinary management.
Materials and Methods: We included all patient (46 patients) who were seen in our out-patient consultation from October 2015 to June 2016 operated from a MMC. All patient had a cranio spinal MRI at birth before the surgical closure of malformation and during follow-up. Clinical outcome was assessed prospectively.
Results: Mean follow up time was 8,1 years. The level of the vertebral malformation was sacral or lower lumbar (≤ L4) in 27 cases (58,7%); higher lumbar (between L1 and L3) in 5

cases (10,9%) and thoracic in 14 cases (30,4%). Antenatal diagnosis was done in 33 cases (72%). Only 78,3% of the patient had a Chiari II malformation at birth before the closure of the malformation. 76% of the patients were operated in the first 24h of life. 60,8% had a surgery for hydrocephalus. Seven had a retetherd cord syndrome and were re-operated. 80% had orthopaedic problems. 57% of the patient were able to walk. 96% were involved in a school program: normal in 70% and adapted in 30%. Vesico-sphincterian problems with intermittent catheterization were found in 60% of the patient. Only 35% of the patient had a normal bowel management.
Conclusions: This study confirms the progress made on the post-natal management of MMC patients. The multidisciplinary management is nowadays mandatory and have permitted, with the improvement of the neurosurgical techniques, to improve clinical outcomes. Our series showed that 57% the patient can walk and that 96% are able to attend a school program. The bladder and bowel management remains a key point for quality of life of this patient.

FP28

Prognosis of surgical treatment of the tethered cord syndrome in children

Kirill Sysoev, William Khachatryan (St. Petersburg, Russia)

Purpose: The aim of this study was to identify the factors relevant to the prognosis of the outcome of the surgical treatment of the tethered cord syndrome (TCS).
Methods: The results of surgical treatment performed on 58 children with TCS were analyzed, with follow-up periods ranging from 6 months to 5 years. The data of preoperative clinical and instrumental examinations, as well as those of intraoperative electrophysiological diagnostics and morphometry, were compared with the dynamics of the TCS clinical presentation.
Results: The recovery rate was significantly higher in children with filum terminale abnormality (p=0.014), as well as Grade I tethering (p=0.0037), and when the spinal cord tracts at the level of intervention were intact (p=0.018). Complete untethering (p=0.04) and a low threshold value of amperage in direct stimulation (<1 mA) (p=0.016) were identified as factors for a favourable outcome. Worsening of neurological symptoms was more frequent in children operated over the age of 10 (p=0.03), when the TCS was manifested exclusively through the pelvic dysfunction (p=0.00004), if the F-wave block is less than 30% (p=0.0045) and the stimulation threshold during root mapping ranged from 1 to 5 mA (p=0.01).
Conclusion: The operation is recommended when structural changes are minimal. In case of severe structural changes, if the spinal cord tracts are intact, the indications for operation are determined by the risk of irreversible structural changes due to the natural course of the disease, although the risks are substantially higher.

FP29

Tethered cord syndrome in young children – Functional outcome after microsurgical detethering

Aurelia Peraud, Nicole Terpolilli (Munich, Germany)

Objective: Tethered Cord Syndrome (TCS) is characterized by neurological, musculoskeletal and urological symptoms that are caused by abnormal tension of the spinal cord. Microsurgical detethering is the therapy of choice in symptomatic TCS, but ideal timing of surgery especially in young kids is still unclear. The aim of our study was to analyse the outcome after surgical detethering with regard to age and neurological outcome.
Methods: Clinical records of all patients under the age of 18 years undergoing surgery for TCS and regularly followed-up in our outpatient clinic from 01/2003-10/2017 were retrospectively analysed.
Results: 62 patients were included and followed up for 3.0 years after surgery. Most common initial symptoms included bladder disturbances (87%), muscle weakness (82%), foot deformities (45.8%), bowel dysfunction (30.6%), and back pain (27.1%). Mean age at operation was 5.7 years. 39 patients (62.9%) had undergone repair of open spinal dysraphism within 4 months post-birth. All patients underwent microsurgical detethering under intraoperative electrophysiological monitoring. Bladder function improved in 64.8% of cases presenting with this symptom; recovery of urinary function tended to be more complete when patients were operated before the age of 4 years. Bowel dysfunction only rarely improved (15.8%). Pain abated in 62.5% of patients. Motor function significantly improved in 74.5% of patients. Of note, while 12/17 patients that were too young to stand and walk at the time of operation were ambulatory at last check-up, only one of 9 patients detethered after the age of 1 year regained the ability to ambulate. Four patients (6.5%, all operated after 2.5 years of age) developed symptomatic re-tehtering.
Conclusion: Neurological outcome, especially recovery of motor function after microsurgical dethetering, seems to be more favourable when surgery is performed early in life and when symptoms are mild. Further and prospective analyses are needed in order to validate these findings.

FP30

The different faces of limited dorsal myeloschizis: before and after 2010, before and after birth

Honorine Maes, Sven Bamps, Katrien Jansen, Frank Van Calenbergh (Leuven, Belgium)

Introduction: Limited dorsal myeloschizis (LDM) has long been confused with meningomyelocoele (MMC). Pang introduced in 2010 LDM as a separate identity, characterized by a focal limited spina bifida with a fibroneural stalk connecting the spinal cord to a normal overlaying skin, caused by an

incomplete disjunction of the neural tube in the embryo. Children with LDM mostly have a milder neurological deficit compared to MMC, have less Chiari-II malformation and less hydrocephalus.
Methods: Five children have been operated in our centre between 1999 and the present for LDM. One child was referred from abroad for fetal surgery for suspected MMC and was only diagnosed on the maternal-fetal MRI. The repair was done in another country but confirmed the diagnosis of LDM. We analyzed diagnostic features, operative technique and clinical outcome.
Results: All patients had a lumbar saccular LDM. It was ruptured at birth in three neonates. They all underwent an intervention to untether the cord of the overlaying skin and reconstruct the spinal canal. In all children we intraoperatively saw a stalk connecting the skin with the spinal cord. Three children had a VP shunt for hydrocephalus. Only one child had Chiari II malformation diagnosed on postnatal MRI. Two children were deemed grade 0, three were grade 1 and one was grade 3 according to the grading of Pang.
Conclusion: LDM is a neural tube defect, recognised by a stalk tethering a focal defect of the spinal cord to the skin. We confirm that the outcome in these children is better than in the usual MMC patient. It is therefore in 2017 imperative to make this diagnosis during fetal life, in order to avoid unnecessary fetal surgery but also termination of pregnancy after counselling as if it was a very severe case of MMC.

FP31
Limited dorsal myeloschisis: the Kolkata experience
Kaushik Sil, Sandip Chatterjee (Kolkata, India)

Aims: Limited Dorsal Myeloschisis (LDM) is an ill recognized form of spinal dysraphism. Here, due to incomplete dysjunction between cutaneous and neuro ectoderm, a fibroneural stalk extends from the dorsal neural tube to the skin. There is a CSF collection around the tube leading to a saccular variety and there is an atretic variety with a skin pit or dimple. Confusion occurs between this and meningomyelocele in saccular variety and with other closed spinal dysraphisms in atretic variety – so the operating surgeon needs to be aware of this distinct entity.
Materials: We present our experience of 24 cases which presented at our centre. 4 of them were recurrent. There were 7 in cervical location, 2 in dorsolumbar region and 15 in lumbosacral region. Most were of saccular variety. There was associated myelocystocele in 2 cases and complex spinal dysraphism in 8 cases. Presentation was mostly in early childhood with tethered cord syndrome, with age range varying from birth to 17years. One of the patients presented at the age of 60 years! None of the recurrent cases were diagnosed as LDM in the first surgery. All patients were diagnosed clinicoradiologically and underwent surgery. At surgery, the stalk was isolated from the cord and transected. Histopathology showed glioneural tissue with mesodermal elements. All children had good neurological recovery.
Conclusions: LDM is a distinct entity and should be kept in mind during surgery of closed spinal dysraphisms. Focal closed spinal defect with fibroneural stalk should alert the surgeon about this possibility.

FP32
Lumbosacral lypoma – Removing technic by the traction bi-coagulation dissection
Albert Sufianov, Marat Gizatullin, Iurii Yakimov, Alexandr Skripnicov, Olga Klimenko (Tyumen, Russia)

Objective: Total and safe resection of lumbosacral lipomas is main important for success surgery. Since 2015, we have performed 31 total/near-total resections of lipomas by the traction bi-coagulation dissection.
Methods: From 2015 to 2017, 31 patients (age range, 2 months-52 years) with dorsal, transitional, caudal lipomas, lypomielomeningocele underwent total or near-total lipoma resection. Ninety percent of the patients were children younger than 18 years and 10% were adults. The technique consisted of using traction bi-coagulation dissection (strip technique) between the fat and the neural plate along a white fibrous plane. Elaborate electrophysiological monitoring was used.
Results: Three postoperative observations concern us. 31 patients, 18 (59%) had no residual fat on postoperative magnetic resonance imaging; 10 patients (35%) had less than 20 mm3 of residual fat; and 3 patients (6%), had more than 20 mm of fat. We used Spina Bifida Neurological Scale (SBNS) for test patients before and after surgery. 21 (68%) had the same level, 7 (23%) had improving their condition, and 3 (9%) had deterioration.
Conclusion: Strip technique resection of lumbosacral lipoma is effective treatment.

FP33
Intraoperative neuromonitoring in surgery for split cord malformations
Ozkan Tehli, Yusuf Izci (Ankara, Turkey)

Introduction: Split cord malformations (SCM) are rare in children. They may cause neurological and urological deficits. Surgery is the main treatment method of SCM. Electrophysiological monitoring is currently in use during the surgery of SCM.
Patients and Methods: We operated 20 children with SCM in the last 2 years. Intraoperative neuromonitoring was used in all patients and motor evoked potentials (MEP), free-run EMG and neurostimulation were

preferred for neuromonitoring. MEP was used before the surgery and after surgery to check the functionality of the spinal cord. Free-run EMG was used during the resection of the splitting lesion and cutting the filum terminale. Neurostimulation was used for during the untethering the spinal cord and determination of the filum terminale.
Results: No neurological deterioration has been occured after surgery. All children were discharged without any complication.
Conclusion: Intraoperative neuromonitoring is very important for the preservation of the functional neural structures and for a safe and effective surgery in patients with SCM.

FP34

Complex combined neurosurgical and orthopedic spinal procedures in pediatric patients: a case-based study

Maria Lucia Licci, Daniel Studer, Carol Hasler, Axel Terrier, Raphael Guzman, Jehuda Soleman (Basel, Switzerland)

Introduction: Correction of spinal deformity in pediatric patients with associated neurological abnormalities, often related to syndromic or genetic diseases, requires complex multidisciplinary treatments and procedures. Many patients undergo combined neurosurgical and orthopedic spinal surgeries either as a single-stage treatment or by interval staging of the deformity correction and the neurosurgical procedure. Our aim is to present our single-center experience in patients who underwent complex combined neurosurgical and orthopedic spinal procedures within the last 5 years.
Methods: A retrospective review of all pediatric patients (<18 years old) undergoing a surgical procedure involving a pediatric neurosurgeon and an orthopedic spine surgeon over a 5-year period was performed. Medical charts of these patients were searched for baseline characteristics, underlying disease or syndrome, pathology indicating surgery, surgical procedure, results of intra-operative spinal cord monitoring and postoperative complications.
Results: Out of all neurosurgical patients treated between the year 2013 and the year 2017, we included 16 patients undergoing a total of 18 combined neurosurgical and orthopedic spinal procedures. 62.5% (n=10) were females and 32.5% (n=6) males, with a mean age of 12.5 years. 2 patients suffered from a syndromic disease, 3 patients suffered a genetic disease (e.g. NF), 4 patients presented with spastic cerebral palsy, while in 7 patients no underlying disease was apparent. In 5 patients the reason for surgery was neoplastic. 9 patients underwent stabilization procedure due to scoliosis combined with a neurosurgical procedure (e.g. untethering, revision of a baclofen pump, spinal canal decompression, etc). Intraoperative neuromonitoring was used in 88% (n=16) of the cases. Surgical morbidity rate was 11.1% and mortality rate was 0%.
Conclusion: In pediatric patients suffering from syndromic or genetic diseases the treatment can be often very challenging. Good collaboration between highly specialized pediatric neurosurgical and pediatric orthopedic spinal teams and correct timing of staged procedures is needed to achieve good outcome in these complex cases.

FP35

Use of percutaneous osteosynthesis in pediatric thoracolumbar fractures

Timothée de Saint-Denis, Dialla Thomas, Syril James, Lotfi Miladi, Michel Zerah, Christophe Glorion (Paris, France)

Child spine fractures represent a minimal part of both spine fractures and pediatric fractures. The potential of growth offers a chance of remodeling but can lead to an evolutive deformity. Percutaneous osteosynthesis has already been settled as a gold standard treatment for some adult spine fractures. Minimal invasive fixation can reduce muscle aggression and definitive arthrodesis.
We started using percutaneous treatment for non-neurologic chance or burst fractures two years ago. 8 patients between 9 and 16 years old have been treated. Reduction and consolidation permitted an early material removal at six months. No complication was observed. With a Follow up between 6 months and 2 years, no pseudoarthrodesis was observed.
This local experience confirms percutaneous osteosynthesis as validated treatment choice for child thoracolombar fractures. Use of this technique could be extended as a temporary internal bracing for some spine deformity.

Abstract session 15: Hydrocephalus II

FP36

The correlation between Third Ventricle Diameter (TVD) and ventricular indices at the time of diagnosis and during its evolution in pediatric hydrocephalus

Louise L. Schweizer, Martin U. Schuhmann, Susanne R. Kerscher (Tübingen, Germany)

Objective: Measurement of ventricular width is important in pediatric patients with hydrocephalus. At the present it is assessed using cranial MRI or CT, coming along with risks of anesthesia in small children or radiation. As the third ventricle is accessible via the temporal bone window using ultrasound, measurement of its diameter could be a reliable and radiation-free alternative to assess ventricular changes. A necessary precondition is, that changes of third ventricle diameter (TVD) correlate to changes of the whole ventricular system. In this study TVD was compared to standard ventricular indices to investigate its correlation.
Methods: 394 MRT/CT images from 117 pediatric patients (median age 6.5 years) with hydrocephalus were measured at

the time of diagnosis and during acute and chronic changes. Measurements included axial TVD and three standard measures of the lateral ventricles (Evans Index, FOHR and Cella Media Index (CMI)). TVD and lateral ventricle measurements were compared using Spearman's and Pearson correlation coefficient at the time of diagnosis, therapy and acute shunt dysfunction. Furthermore, a correlation within subjects was calculated in 8 patients.

Results: The best linear correlation was found between TVD and CMI after initial therapy (r=0.7), at acute shunt dysfunction (R=0.702) and after shunt revision (r=0.566). After shunt revision changes of TVD correlated well to changes in Evans (r=0.609), FOHR (r=0.74), $p<0.01$. The correlation within subjects were outstanding Evans Index (r=0.988), FOHR Index (=0.99) and CMI (0.99).

Conclusion: TVD showed a significant correlationwith all lateral ventricle indices at the time of diagnosis, therapy, shunt dysfunction and during chronic changes. It is therefore an excellent mirror of ventricular changes in pediatric hydrocephalus. Thus, TVD measurements by ultrasound can potentially substitute standard imaging to assess changes in ventricular width.

FP37

Assessment of intracranial pressure and ventricular size in Pediatric Neurosurgery using combined ultrasound measurement of the Optic Nerve Sheath Diameter (ONSD) and Third Ventricle Diameter (TVD)

Susanne R. Kerscher, Marcel Kullmann, Annette Weichselbaum, Andrea Bevot, Martin U. Schuhmann (Tübingen, Germany)

Introduction: In neurosurgery a large number of pathologies is associated with increased intracranial pressure (ICP). ICP increase can be caused by hydrocephalus, idiopathic intracranial hypertension (IIH) or brain edema. Non-invasive diagnostics to assess ICP and differentiate between hydrocephalus and other entities are needed in pediatric and adult patients. This study aims to investigate the combined use of ultrasound ONSD and TVD to assess ICP and ventricular size in pediatric patients.

Methods: This prospective study includes 87 patients (median 7 years) diagnosed with hydrocephalus (53%), IIH (22%), tumor (8%) and other intracranial pathologies (17%). Binocular ONSD was measured transorbitally using a 12MHz linear probe. TVD was quantified with a transtemporal approach using a phased-array 1-4MHz transducer placed at the temporal window. All measurements were done with patient in supine position.

Results: 33 patients presented increased ONSD (mean 5.9 ±0.6mm) and TVD (mean 7.86±5.86mm) and underwent ICP decreasing therapy. In 14 patients ONSD (5.9±0.6mm) was enlarged while TVD (3.97±2.39mm) was only marginally increased. In these cases further diagnostic procedures were recommended. 40 patients presented with normal ONSD (4.98±0.6mm) and normal to marginally increased TVD (4.25±3.58mm). Watch-and-wait was performed and none needed an intervention up to now. In 21 patients TVD was measured before/after hydrocephalus therapy and in 7 without intervention. The relative difference was higher in patients with therapy (31 ± 19.7%) than without (4.8± 4.2%), $p<0.001$. A change of > 7.65% is highly sensitive and specific for true and relevant ventricle enlargement (sensitivity 90.5%, specificity 87.5%, AUROC 0.939, OR 57).

Conclusion: Transorbital ONSD is a reliable method to assess ICP and can be combined with transtemporal TVD for identification of possible ICP increases. Changes of TVD over time can be used for diagnosis of clinically relevant ventricular enlargements in pediatric patients.

FP38

Effect of ventricular volumes on post-hemorrhagic developmental outcome

Marcus Lo, Roy Eagleson, Jessica Kishimoto, Sandrine de Ribaupierre (London ON, Canada)

Introduction: Intraventricular hemorrhage affects preterm infants, and even though its incidence has gone down, the best management of posthemorrhagic hydrocephalus is still controversial. 2D Ultrasound is traditionally used for following cases of ventricular enlargement, however, 3D US enables a more accurate reflection of volumetric changes and allows the clinician to decide whether an intervention is needed with posthemorrhagic ventriculomegaly.

Methods: We have studied 57 preterm infants with IVH and looked at their outcome during the following two years by recording scores such as the AIMS, INFANIB, and Bayley-III assessment. Overall, half of the cohort had a low grade IVH, and the other half had a higher grade. We also looked at the effect of highest ventricular volume (assessed with 3D ultrasound, and followed over the length of the intensive care stay after birth, usually 1-3 months), the need of an intervention, and correlated it with the outcomes.

Results: As expected, the lower grades did better in all assessment scores, but interestingly, the grade III tended to be worse than the grade IV. In our population however, it seemed that the surviving grade III had more other prematurity issues than the surviving grade IV, and we could see an effect of ventricular volume on the outcome.

Discussion: 3D US enables clinicians to follow ventricular volume in IVH patients better than 2D US, as the shape of the ventricle is irregular and there tends to be a posterior enlargement which can be monitored more effectively on the 3D US than the 2D images. We have shown in the past that 3D US was therefore valuable,

but we can now show the effect of volume on neuropsychological assessments as early as two years of age.
Conclusion: Following accurate ventricular volume can help in the management of posthermorrhagic hydrocephalus and might affect outcome.

FP39

Patterns of ventricular dilatation in post hemorrhagic hydrocephalus predicting timing and usefulness for Ventriculo-Peritoneal Shunt (VPS) placement in premature infants

Federico Di Rocco, Coline Peron, Laura-Nanna Lohkamp, Franck Plaisant, Olivier Claris, Pierre-Aurelien Beuriat, Alexandru Szathmari, Carmine Mottolese (Lyon, France)

Introduction: Intraventricular hemorrhage in prematures may result in a post-hemorrhagic hydrocephalus. The pattern of evolution of the ventricular dilatation after the hemorrhage and its impact on treatment decision and outcome remain unclear. The aim of the study was to evaluate such patterns for incorporating them in clinical decision-making.
Material and Methods: Retrospective observational single-center study including all premature infants, who presented with severe intraventricular hemorrhage, realized between 2009 and 2017 in Lyon. Ventricular dilatation kinetics have been analyzed with respect to different treatment modalities and risk factors for VPS placement. Outcome at age two was analysed.
Results: Seventy-five patients with severe intra-ventricular hemorrhage were observed. Fifty of them died. Among the twenty-five surviving patients twenty-two presented with hydrocephalus. Eight patients did not require any treatment, 7 patients underwent a temporary treatment via lumbar or ventricular punctures and 7 patients required finally a VPS. The mean interval between the initial puncture and VPS placement was 24 days. Risk factors for VPS requirement were: arterial hypotension with catecholamine dependency, bilateral hemorrhage, clinical and radiological signs of ventricular dilatation. Ventricular dilatation was regressive after 17 days after lumbar or ventricular puncture in patients that received a temporary treatment. The development at two years of age was more affected in infants that underwent VPS placement, showing a delay in psychomotor and speech development. Cerebral palsy was shown to occur more frequently after bilateral hemorrhage, but was not associated to VPS placement.
Conclusion: Premature infants with intra-ventricular hemorrhage and post hemorrhagic hydrocephalus who underwent VPS placement are at higher risk for neurologic deficits that the other treatment groups. Bilateral hemorrhage was highly related to catecholamine treatment and to requirement of VPS placement after 17 days of temporary treatment. An early VPS placement may have a preventive effect on persistent neurologic deficits related to intracranial hypertension.

FP40

Ventriculoperitoneal shunt valve pressure adjustments in hydrocephalic infants below the age of six months according to head circumferences and transfontanelle ultrasonography measurements

Adriano Cattani, Franziska Schwarzer, Mario Schwarzer, Andrea Spyrantis, Gerhard Marquardt, Susanne Schubert-Bast, Marco B. Bartels, Volker Seifert, Thomas M. Freiman (Frankfurt, Germany)

Objective: Ventriculoperitonal shunt (VP-Shunt) is the standard hydrocephalus treatment in newborns. However, the right time of VP-Shunt insertion with or without additional components as larger sized adjustable valve pressure system or anti-siphon device in developing children who are still unable to walk is still matter of intensive debates. Therefore, we observed hydrocephalic infants below the age of six months who were implanted with a VP-shunt and adjustable valve pressure as well an anti-siphon system and performed the needed valve pressure adjustments according to their head circumferences (HC) measurements and ventricular size in order to avoid macro- or microcephaly and consequently disturbance during brain development.
Method: We included 31 hydrocephalic children aged from one day up to 6 months of life, inclusive low-weight preterm infants, in a 3 years period between 2014 and 2017 admitted in our institution. All children were implanted with the proGAV VP-Shunt with an adjustable valve pressure system with range between zero and 20 cm H_2O together with a pumping-chamber and a gravity dependent valve with fixed pressure at 20 cm H_2O. Prospective head circumferences and transfontanelle ultrasonographic ventricular size measurements were performed with follow-up amounted between 10 days and 2 months. At VP-Shunt implantation the standard valve pressure was initially set to 5 cm H_2O. Valve pressure lower adjustments were done, if the HC exceeded the 90^{th} percentile and higher adjustments if the HC was lower than the 10^{th} percentile. Outcomes of children with post intraventricular haemorrhage hydrocephalus (pIVHH; n=13), and mature children with congenital or acquired hydrocephalus (non-intraventricular haemorrhage hydrocephalus; nIVHH; n=18) were analysed separately.
Results: Most children (n=23, 74%) needed valve pressure adjustments to maintain a normal head growth. In particular most of the children with pIVHH were preterm infants and needed more valve pressure adjustments (mean: ~3 versus 1.5 per children), had higher rate of shunt infections (~61% versus 39%) and revisions (mean: 4.5 versus ~2 per children) comparing to nIVHH. Overall, 90% of all implanted children had HC percentile improved according to HC measurements deceleration or at least stabilised according to HC stagnation (45% improvement; 45% stabilisation and 10% degradation of HC) at the first follow up after VP-Shunt insertion.

Conclusion: These results indicate that using a permanent proGAV valves presents the advantage of adjusting pressure as needed to efficiently manage hydrocephalus in children regardless aetiologies. However more precautions should be taken, particularly in the group of post-IVH children, to better set the valve pressure at insertion and to decrease complication risk. In addition, the risk of asymptomatic complications (e.g. subdural haematoma, hygrom) is present in all hydrocephalic children, and ultrasonography should be performed systematically during the follow up period.

FP41

Anatomical changes in the floor of the third ventricle in patients with hydrocephalus due to pure aqueduct of sylvius stenosis before and after endoscopic third ventriculostomy

Tuncer Turhan, Elif Bolat, Kadri Emre Çalışkan (Izmir, Turkey)

Introduction: Some radiological changes in the floor of the third ventricle has been described in brain magnetic resonance images (BMRI) of the patients with obstructive type hydrocephalus. Frequently, the floor of the third ventricle (tFTV) where is called tuber cinerum bulges downward into the interpeduncular cistern in cases of aqueduct stenosis.
This is thought to be due to a pressure difference between the intracisternal area and the ventricular system. Therefore there is a general acceptance that endoscopic third ventriculostomy (E3V) operation is more successful in this patients. This study was designed to investigate the validity of this hypothesis.
Methods: Patients who underwent E3V operation between 2010-2017 in our clinic were retrospectively reviewed. Patients with complete occlusion in the aqueduct region who had not undergone any previous operation or intracranial disease were selected. The radiological and clinical data of these patients were collected and evaluated statistically by SPSS 17 program.
Results: 83 (46 were female and 37 were male) patients with aqueduct stenosis were accepted to this study. The mean age was 19.1 years (min 11 days, maximum 71 years). After E3V operation, the mean follow-up period was 20.84 months. When preoperative brain MRI examinations of these patients were examined, it was observed that 38 patients had bulging to the interpeduncular cistern at tFTV. After the E3V operation, the anatomy of tFTV improved in 23 patients. There was no change in postoperative brain MRI of other 15 patients.
E3V success rate in patients with bulging on tFTV preoperatively was calculated as 77.7%.In patients without bulging in this area in preoperative MRI, E3V success rate was 81.5%. However, there is no statistical difference between these two groups (chi-square p=0.441).
38 patients with bulging on tFTV in preoperative MRI were evaluated separately.Success rate in patients with no changes in tFTV in brain MRI after the operation was calculated as 66.6% and success rate is 91.3% in patients with bulging improvement. However, there is no statistically significant difference between these two groups (chi-square p=0.07).
Conclusion: In patients with aqueduct stenosis, changes in tFTV support the presence of hydrocephalus, but not a stand-alone indicator of E3V success.

FP42

Hydrocephalus following resection of posterior fossa tumors in children: a proposed abbreviation of the Preoperative Prediction Rule

Kevin Owusu-Agyemang, Emer Campbell, Anthony Amato-Watkins (Glasgow, United Kingdom)

Introduction: Approximately one-third of children with posterior fossa tumors require permanent CSF diversion. The Canadian-Preoperative-Prediction-Rule-for-Hydrocephalus (CPPRH) and subsequent modification mCPPRH were devised as evidence-based tools to rationalise the increasing use of prophylactic endoscopic third ventriculostomy (ETV) in managing the risk of this condition. The authors sought to critically appraise the mCPPRH.
Methods: A retrospective record review of 65 consecutive children (age <16) with posterior fossa tumours treated at the Royal Hospital for Children in Glasgow between 2012 and 2017. Univariate and multivariate analyses of patient, tumourand operative characteristics. Primary outcome was ventricular shunting at 6-months. The mCPPRH and Endoscopic-Third-Ventriculostomy-Success-Score (ETVSS) were assessed, with an abbreviated, composite predictive score applied.
Results: Fifty patients were evaluated. Of the variables identified as significant predictors of post-resection hydrocephalus in the mCPPRH; age < 2-years and moderate/severe hydrocephalus were found to be significant predictors for our cohort. No significant univariate associations with the outcome was found for the operative variables: pre-resection external ventricular drain, craniectomy and ETV.
A high mCPPRH (scores ≥ 5) remained a significant predictor of post-resection hydrocephalus with observed likelihood ratio (LR) of (4.391, p=0.036). The need for ventricular shunting in our series was predicted by a low ETVSS (<70). A high abbreviated mCPPRH comprised of age <2-years (score 3), moderate/severe hydrocephalus (score 2), transependymal oedema (score 1), with adjustment for ETVSS <70 (score 1) was a significant predictor (LR 8.639, p=0.003) of permanent CSF diversion with a similar area under the ROC curve of 0.649 to the mCPPRH.
Conclusion: The incidence of ventricular shunting post-tumour resection was predicted by the abbreviated score. This simplified model relies on more objective variables and by identifying patients at high risk of developing post-

resection hydrocephalus will help target research efforts at this group, as well as prepare patients and their relatives for expected outcomes.

FP43

Ventriculo-pleural shunts in children

Marina Brigui, Dominic Thompson, Gregory James, Kristian Aquilina (London, United Kingdom)

Background: Ventriculoperitoneal shunts are the most common shunts used in paediatric neurosurgical practice. When the peritoneum is unsuitable for cerebrospinal fluid (CSF) drainage, the atrium or the pleural cavity are used instead. We have previously reported on the relatively high morbidity and short longevity of ventriculoatrial shunts[1]. In this study, the indications, complications and survival of pleural shunts are reviewed.

Materials and Methods: Retrospective analysis of all children who have undergone shunt surgery in our institution over the last 25 years.

Results: From 1993 to 2017, 32 children between 1 and 15 years old (mean 9.4) have undergone 36 ventriculopleural shunt insertions, representing 0.65% of the 5508 shunt procedures performed over this period. There were no infants in this series. These children had from 1 to 33 previous shunt surgeries (mean 7.6). Post-operatively, 18 shunts (50%) needed revision, after an average of 5.8 years. Ventriculopleural shunt survival was 69% at 6 months, 64% at 1 year, 22% at 5 years and 14% at 10 years. There were 2 cases of infection, one CSF leakage along the tract, one wound breakdown and the rest had shunt blockage. One child suffered a respiratory arrest unrelated to shunt insertion. Mean duration of follow-up was 12 years (range 0.3 to 25 years).

Conclusion: Ventriculopleural shunts can be safely used as an alternative to ventriculoperitoneal shunts in children over 1 year when the peritoneum is not available for CSF drainage. Their longevity and complication profile is better than that for VA shunts in our series.

Reference:

1 Ultrasound guided placement of the distal catheter in paediatric ventriculoatrial shunts – an appraisal of efficacy and complications. Clark DJ, Chakraborty A, Roebuck DJ, Thompson DNP. Childs Nerv Syst (2016) 32:1219-1225.

FP44

Progressive programmable valves failure in children with normal CSF – A technical problem of valve design?

Stephanie Anetsberger, Ahmed El Damaty, Andreas Unterberg, Heidi Bächli (Heidelberg, Germany)

Objective: Hydrocephalus is one of the most common diseases encountered in the field of Pediatric Neurosurgery. This condition is often treated with implantation of a ventriculoperitoneal shunt. Shunt system design has evolved to minimize failure rates and simplify management. We implement the programmable Miethke valves, which have been shown to reduce the problem of CSF overdrainage in children. The proGAV 2.0 is the latest, technically improved programmable valve system currently available. This system utilizes a differential pressure unit combined with a gravitational unit that could also be adjustable to prevent overdrainage. We frequently observed failures of these programmable valves in the last 10 months, possible reasons are discussed.

Methods: From March 2017 until November 2017, we explanted 16 defect valves in children with shunt failure, those children were between the ages of 10 months and 18 years. CSF was collected from each case intraoperatively and examined. The explanted valves were sent to the manufacturer for analysis. The valves removed had been implanted between 2006 and 2017. Hydrocephalus in these children had developed secondary to: intraventricular haemorrhage (31%), tumor (23%), aqueductal stenosis (23%), MMC (15%) and Dandy Walker syndrome (8%). Indication for surgical revision was either a symptomatic over- (54%) or underdrainage (46%) in patients in whom valves were no longer adjustable.

Results: 62% of the children had slit ventricles. The malfunctioning valves were proGAV (25%), proGAV 2.0 (37,5%) and proSA (37,5%). All patients had normal CSF including protein levels. Despite normal CSF protein, in all explanted valves, sedimentation of detritus was found when were opened and examined by the manufacturer.

Conclusion: To our understanding possible reasons for the valve failure in patients with normal CSF protein, could be the narrow space inside the valve which possibly leads in combination with a narrow valve outlet to sedimentation of detritus, partially also as a consequence of slit ventricles. Sedimentation likely blocks the rotor in the valve, thereby hindering programmability. This may play a significant role in causing the over- or underdrainage observed in these patients.

FP45

Robot-assisted neuroendoscopic intracranial membrane resection in premature infants with post-hemorrhagic hydrocephalus and complex cyst formation

Andrea Spyrantis, Adriano Cattani, Volker Seifert, Susanne Schubert-Bast, Thomas Freiman (Frankfurt am Main, Germany)

Introduction: Complex intraventricular cyst formations are observed after intraventricular hemorrhage in premature infants, in particular after cerebrospinal fluid (CSF) diversion-associated infections. The resection of multiple cyst membranes is crucial for creating one common CSF space. Since membranes have a complex structure, navigated procedures are

required. We report our experience with robot guided minimally invasive endoscopic intervention and microsurgical operations.
Methods: Two children were operated using a microsurgical approach guided by neuronavigation, two were operated endoscopically with the endoscope guided by the robotic surgery assistant (ROSA). All four infants were premature, had intraventricular hemorrhage, hydrocephalus and developed intracerebral cysts. Due to the soft scull, two of four children were not fixed in the Mayfield clamp, but fixed by tapes. For the robot guided endoscopic approach, referencing with preoperative magnetic resonance imaging was accomplished by facial laser scan.
Results: The children had between six and 23 CSF catheter procedures before and a minimum of 6 cysts. Diameters of microsurgical craniotomies were 3 cm or larger. Due to the thickness and vascularization of the membranes, pure endoscopic operations were not possible and microsurgical instruments were used. The robot reached all planned endocopic trajectories to resect membranes. This was not achieved in microsurgical cases, despite neuronavigation. Membrane resection was followed by external ventricular drainage for seven to 14 days and thereafter permanent shunt implantation.
Conclusion: Due to thick cyst walls robot-guided endoscopic treatment could not be carried out as intraventricular endoscopy but as endoscopic assisted microsurgery. However, the anatomy of the intracerebral cysts was better visible than with microscope, only.

FP46

Wound dehiscence after CSF shunt surgery: options for surgical management

Alexandra Honekamp, Julia Nassrin Masomi-Bornwasser, Martin Glaser, Wolfgang Wagner (Meppen, Germany; Mainz, Germany)

Introduction: In cases of bacterial CSF shunt infection, explantation of the complete system, temporary external CSF derivation and re-insertion of a new shunt after recovery from infection is the classical management. However, in cases of wound dehiscence over the shunt without signs of CSF or systemical infection, local skin revision without shunt explantation system may be sufficient.
Methods: Between 2002 and 2017 in Mainz, 32 patients (23 male, 9 female) underwent 34 operations for local skin revision after shunt surgery without explantation of the system. Age at shunt surgery was 1 day - 16 years (median 195 days). Time interval between shunt surgery and wound revision was 5 - 298 (median 21) days. Skin dehiscence was directly over or adjacent to the shunt material in all cases (frontal, retroaurikular, occipital, clavicular, abdominal or temporoparietal). Follow-up was 1.6 - 181 (median 29) months.
Results: There was no subsequent shunt infection after 27 revision surgeries. In 2 of the remaining 7 cases, infection occurred more than 6 months later; in another case it happened after a primarily abdominal infection, in the beginning without meningitis. That is, the relationship between wound revision and subsequent shunt infection was probable in 4 out of 34 operations, none in 2 cases, and questionable in 1 case. Median age at revision surgery of patients with infection was 154 days, without infection 195 days. Age at surgery, site of wound dehiscence or liquorrhoea did not correlate with infection risk.
Conclusion: In cases of a wound dehision after shunt surgery without signs of local or systemic infection, it is warranted to perform as a first step a local skin revision without shunt explantation, with the chance of avoiding additional operations or longer hospital stays. When local or systemic infection is present, the explantation of the shunt system is inevitable.

FP47

Cerebrospinal fluid shunt-associated infections in pediatric patients: A retrospective cohort study over a 10-year period

Luise Martin, Nora Renz, Tobias Finger, Andrej Trampuz, Ulrich-Wilhelm Thomale (Berlin, Germany)

Background: Cerebrospinal fluid (CSF) shunt-associated infections in children are challenging due to quick progression of inflammation with potential irreversible brain damage. The aim of this study is to describe clinical features, microbiological findings and treatment of pediatric patients with shunt infections.
Materials/Methods: Consecutive patients aged <18 years with first diagnosed episode of shunt infection from January 2007 until August 2017 were included. We retrospectively collected data by chart review. Shunt-associated infection was defined by the Center for Diseases Control (CDC) definition criteria.
Results: We included 48 children with a median patient age of 2,5 years (range, 1 month to 17 years), 19 were female (40%). Infections involved 47 ventriculoperitoneal and 1 ventriculoatrial shunts. Main etiology of hydrocephalus was posthemorrhagic (n=22). The median time between shunt implantation (or last revision surgery) and manifestation of infection was 47 days (range, 1 day to 7.8 years). Most infections occurred within one year after surgery (n=33, 69%), among them 16 within the first postoperative month. Fever (>38°) was present in 33 (75%), local inflammation in 8 children (17%). Serum CRP was elevated (>8 mg/l) in 38 patients (79%). In CSF, leukocyte count was $>5x10^6$ cells/l in 39 patients (90%). The causing pathogen, cultured from CSF, was detected in 27 infections (59%); 19 infections (41%) remained culture-negative. The most

common pathogens were coagulase-negative staphylococci (n=13), *Staphylococcus aureus* (n=3), gram-negative bacilli (n=6) and enterococci (n=3). All patients received intravenous antibiotic treatment (median:18 days (range, 5-37 days). Surgical procedure was performed in 46 patients; predominantly 2-stage shunt replacement with temporary external ventricular drainage (n=31,67%).

Conclusions: CSF shunt-associated infection is a serious complication in children, usually manifesting within the first months after shunt implantation. CSF leukocyte count was normal in 10%. Almost half of infections remained culture-negative, potentially due to early antimicrobial therapy preceding diagnostic sampling.

FP48

Does the CSF protein concentration influence shunt survival in pediatric patients?

Emer Campbell, Anthony Amato-Watkins, Roddy O'Kane, Meharpal Sangra (Glasgow, United Kingdom)

Introduction: Currently CSF shunt insertion is one of the mainstays of treatment of hydrocephalus; and whilst this continues, shunt failure will continue to be a cause of significant morbidity in children with hydrocephalus.

We sought to determine the impact of the CSF protein concentration at the time of insertion on the survival of the CSF shunt; if the CSF protein is elevated, is the clinical course of the shunt affected?

Methods: Prospective surveillance of all CSF shunt insertions performed in our unit from 1st January 2012 to 30th June 2016 in patients aged < 16yrs at the time of operation. Normal CSF protein concentration defined ≤0.50g/L; high CSF protein >0.50g/L.

Survival assessed by Kaplan Meier analysis and Chi-square analysis of 30-day, 1 year and overall lifespan of shunt.

Results: 162 CSF shunt insertions were performed in 120 patients. CSF protein concentration prior to insertion of the shunt was known in 111 procedures. CSF protein recorded as normal in 62 cases and high in 49 cases.

No difference was detected in either overall CSF shunt survival or one-year shunt survival for high CSF protein concentrations compared to normal CSF concentrations.

>A significant difference was detected in 30-day survival; shunts inserted into CSF with a high protein concentration were more likely to fail within 30 days than shunts inserted into CSF with a normal protein concentration, (Normal CSF failure rate 7/62, High CSF, failure rate: 16/49, p=0.006).

Conclusion: Patients with elevated CSF protein concentrations are more likely to suffer an early shunt failure (within 30 days of insertion); however, if their shunt survives to beyond 30 days, the risk of failure is not different to patients with normal CSF protein concentrations.

Abstract session 18: Neuro-Oncology III

FP49

Improved risk-stratification for posterior fossa ependymoma of childhood considering clinical, histological and genetic features

Stephanie Jünger, Martin Mynarek, E. Dörner, A. zur Mühlen, K. von Hoff, Stefan Rutkowski, A. von Bueren, Torsten Pietsch (Bonn, Germany; Hamburg, Germany)

Introduction: We aimed to identify independent outcome predictors in children with posterior fossa ependymomas.

Methods: Tumor samples of 134 patients aged 0.2 -15.9 years treated according to HIT protocols were analyzed for histological features including mitotic activity, necrosis and vascular proliferation and genomic alterations by SNP and molecular inversion probe analysis. Survival analysis was performed by Kaplan-Meier method with log rank test and Cox regression.

Results: Residual tumor, chromosome 1q gain and structural genomic alterations were identified as predictors of significantly shorter event-free (EFS) and overall survival (OS). Furthermore, vascular proliferation, necrosis and high mitotic activity were predictive for shorter OS. Multivariate Cox regression revealed residual tumor, chromosome 1q gain, vascular proliferation and high mitotic activity as independent predictors of OS; for EFS, chromosome 1q gain, residual tumor and mitotic activity were predictive. We found evidence that tumors of different age groups are distinct concerning the parameter chromosome 1q gain, which was absent in tumors of children younger than 18 months of age at the time of diagnosis.

Conclusion: The independent outcome predictors residual tumor, chromosome 1q gain and mitotic activity can be integrated in an improved risk-stratification model for posterior fossa ependymoma of childhood., which outperforms current stratification procedures.

FP50

Incidental pediatric brain tumors: an international survey

Jonathan Roth, Jehuda Soleman, Robert F. Keating, Dimitrios Paraskevopoulos, Shlomi Constantini (Tel-Aviv, Israel; Basel, Switzerland; Washington DC, USA; London, United Kingdom)

Introduction: Incidental pediatric brain tumors (IPBT) are increasingly being diagnosed. Currently, there is no consensus regarding the need and timing of treatment of such lesions. In the current study, we exemplify treatment trends among pediatric neurosurgeons and oncologists in regard to a sample of 6 incidental lesions, as well as trends for approval of growth-hormone replacement therapy (GHRT).

Methods: A questionnaire presenting 6 different cases was spread via the following platforms to all their members by and email:
• ISPN
• ESPN
• Listserv - reference: http://www.neurosurgeryresearch.net
• SIOP
• ISPNO 2016 congress participants
The survey was open for 3 months. Collected data included basic information concerning the responder (surgeon / oncologist, experience, continent of practice), as well as responses to multiple questions regarding treatment of the child (follow / biopsy / resection), permission to supply GHRT, and free text for comments.
Results: 143 responses were eligible for analysis (92 neurosurgeons, 51 oncologists, from a total of 6 continents). Results for each case will be presented separately. However, across all 6 cases, a stable lesion over time lead to a shift in treatment recommendation to a more conservative one, while lesions with a fast change were treated more aggressively. Neither profession, nor experience had a consistent impact on the recommendations. GHRT were more often approved once lesions were stable over time compared to baseline.
Conclusions: Treatment recommendations are not absolute, and are influenced by many factors. However, stable lesions lead to a shift in recommendation towards a "follow" approach, while growing lesions shifted recommendations toward a "biopsy" or "resection" approach. This stresses the need for better understanding the natural course of incidental brain tumors in children, as well as evaluating the potential risk for malignant.

FP51

The most influential papers in the neurosurgical management of pediatric central nervous system tumors

Charlotte Burford, Yasmine Cherfi, Bassel Zebian (London, United Kingdom)

Introduction: Citation number can be used as a measure of the influence an academic publication has. Papers in paediatric neurosurgery with more than 50 citations and a citation rate of more than 5 per year have been suggested to be high impact. This study aims to determine and categorise the most highly cited papers related to the neurosurgical management of paediatric brain tumours between 2005-2010.
Methods: A previous study (Wilcox et al., 2013) identified the three journals which published the most influential papers in paediatric neurosurgery: Child's Nervous System, Journal of Neurosurgery: Paediatrics and Paediatric Neurosurgery. All papers published by these journals between 2005-2010 were collected from Web of Science. Papers related to the neurosurgical management of paediatric central nervous system neoplasms were identified and the 50 most cited were determined. Papers not relating to surgical management were excluded.
Results: 54% of the most highly cited papers were published in *Child's Nervous System*, 26% in *Journal of Neurosurgery: Paediatrics* and 20% in *Paediatric Neurosurgery*. Citation number ranged from 27-93 (mean = 43) and citations per year ranged from 2.31-8.25. 24% had more than 50 citations, 8 of which had a citation rate of more than 5 per year. 42% of the 50 most highly cited and 70% of the top 10 most cited papers were related to the surgical management of craniopharyngiomas. 10% were related to surgical management of posterior fossa tumours. 24% of papers were review articles and 72% were experiential series. There were no randomised controlled trials.
Conclusion: Between 2005-2010 the majority of highly influential papers related to the surgical management of paediatric tumours were regarding craniopharyngiomas. 8 studies could be considered high impact within the field of paediatric neurosurgery.

FP52

A web-based application for complication registration in neuro-oncological trials

Lars Van der Loo, Erik Van Lindert (Nijmegen, the Netherlands)

Introduction: Accurate registration of intra- and postoperative complications is indispensable in providing best care, assuring quality and improving outcomes of paediatric neurosurgery. Usually, the treating surgeon or other staff members of the department register complications in a standardized manner for all their patients. However, complication registries can differ greatly between various centres. The aim of this study therefore, was to develop a uniform complication registration database for all patients treated by physicians involved with the Brain Tumour Group of the European Society for Paediatric Oncology (SIOPE-BTG).
Methods: We developed a web-based application for the registration of complications for all patients included in SIOPE-BTG-affiliated clinical trials. Complication definitions were formulated using the Canadian Paediatric Neurosurgery Study Group's definitions of complications in paediatric neurosurgery. Definitions for intraoperative complications were obtained from research by the Radboud University Medical Centre in the Netherlands. Complications were classified using the Ibañez Classification. The web-based application outputs digital files which can be filed in patient notes or electronic patient records, as well as in trial databases. A small pilot study is being performed in our hospital. Complications are recorded at 30 days post-operatively and limited to patients with brain tumours.
Results: A Wordpress-driven web-based application was successfully developed. This application was developed in strict adherence to Dutch Law regarding safety of information. A small pilot study is being performed;

preliminary results yield high physician satisfaction about the ease of use of the web-based application and the highly usable output format.
Conclusions: A web-based complication registration system offers clinicians a user-friendly and accurate tool for easy registration of complications. This SIOPE-BTG approved application combines multiple consensus definitions on intra- and post-operative complications and provides standardized data to assess complication rates in clinical trials. Further implementation of this tool in the SIOPE-BTG is planned.

FP53

Intraoperative MRI use in pediatric brain tumor surgery

William White, Ashok Raghavan, Hesham Zaki, John McMullan, Saurabh Sinha, Patricia De Lacy, Shungu Ushewokunze (Sheffield, United Kingdom)

Introduction: Intraoperative MRI (IoMRI) is a valuable tool in paediatric neurosurgery to maximize brain tumour resection rates and minimize morbidity. The availability of this technology worldwide is limited due to high costs. An alternative is to perform an immediate MRI under the same general anaesthetic. We have compared the use of immediate post operative MRI scans to our recent experience of using an intraoperative MRI scanner.
Methods: A retrospective review was performed that included 109 patients. Patients that underwent an IoMRI (Group A) were compared to those that underwent an immediate post-op MRI (Group B). In Group A 23 patients underwent 23 operations, between December 2015 and September 2017. In Group B 87 patients underwent 110 operations between January 2006 and December 2014.
Results: The mean total anaesthetic time was 554 minutes (n=22, SD=106) in Group A and 500 minutes (n=107, SD=137) in Group B (p=0.087). The transfer time to MRI was 28 minutes (n=21, SD=10.2) in Group A and 38 minutes (n=106, SD=18.4) in Group B (p<0.001). Further resection was performed after MRI in 4 (17.4%) patients in Group A and 3 (2.7%) in Group B. The mean total anaesthetic time when further resection was undertaken after MRI scan was 610 minutes in Group A and 680 minutes in Group B.
In the cases where the surgical intent was for a complete resection, this was achieved in 65% of cases: in Group A 55% (n=19) and in Group B 68% (n=81) (p=0.209).
Conclusions: IoMRI allows for quicker transfer to MRI compared to immediate postoperative MRI. When further resections are carried out IoMRI reduces the duration of surgery. In a resource limited setting the use of immediate MRI is a feasible alternative to IoMRI in paediatric brain tumour surgery.

FP54

Retrospective single-center study of the prevalence of hydrocephalus in pediatric brain-tumor patients and its perioperative treatment paradigm

Andreas Schaumann, Charlotte Hammar, Matthias Schulz, Ulrich-Wilhelm Thomale (Berlin, Germany)

Objective: The necessity of hydrocephalus treatment in pediatric brain tumor patients suffering from different tumor entities is yet heterogeneously approached. We retrospectively reviewed our perioperative hydrocephalus treatment paradigm in pediatric brain tumor patients during a 5-year period.
Methods: Our brain tumor data base was evaluated regarding the prevalence of hydrocephalus and its perioperative treatment strategies. From May 2010 to April 2015 we performed 341 tumor related operations such as total, subtotal, partial removal or biopsy in 206 pediatric patients (108 male (53,2%, mean age at tumor-intervention: 7,58 years), 98 female (46,8%, mean age at tumor intervention: 7,39 years), who offered sufficient follow-up for this study. Hydrocephalus related operations such as external ventricular drain (EVD), endoscopic third-ventriculo-cisternostomy (ETV) or ventriculo-perioneal (VP) shunt were evaluated in this patient cohort.
Results: Out of 206 patients 101 (49%) patients needed a CSF diverting procedure.
74 (35,9%) patients received the intervention before tumor surgery (20 (27%) ETV, 51 (68%) EVD and 13 (17,5%) VP-Shunt). In the latter group 10 (9,9%) patients had a preexisting shunting device ad referal. In addition, postoperatively 23 (11,1%) patients received a CSF relieve by ETV (n=5; 21,7%) or shunt (n=18; 78,3%). The ratio for a temporary csf-diversion (EVD) related to diagnosis were 35,6% pilocytic astrozytoma, 33,3% anapl. Ependymoma, 31,2% medulloblastoma and 40% ATRT. 56 patients received a permanent CSF diverting procedure by either ETV (n=25) or VP shunt (n=29) implantation with a ratio related to diagnosis of 13,6% pilocytic astrozytoma, 33,3% anapl. Ependymoma, 37,5% medulloblastoma and 33,3% ATRT.
Conclusion: Based on the practical routine from a five-year period treating pediatric brain tumor patients we conclude a relevant rate of 49% for temporary CSF diversion with only 14% (n=29) of patients required a VP-shunt.

FP55

Spinal cord tumors in children: 17 years of experience in Lyon

Alexandru Szathmari, Pierre-Aurelien Beuriat, Alexandre Vasiljevic, Didier Frappaz, Cecile Faure Conter, Line Claude, Valeria Blé, Laura-Nanna Lockhamp, Pierre Antherieu, Federico Di Rocco, Carmine Mottolese (Lyon, France)

Introduction: Spinal cord tumors (SCT) are rare in children. We reviewed all spinal cord tumors operated in Lyon in the past 17 years.
Methods: All patients were extracted from our data base. Medical files were then retrospectively review.

Results: From January 2000 to December 2017, 23 spinal cord tumors were operated. The mean age at diagnosis was 9,6-year-old. The mean follow-up time was 4,9 years.
Symptoms were back pain in 7 cases, back pain and lower limb motor deficit in 5 cases, lower limb motor deficit only in 4 cases and others in 6 (scoliosis, sacral malformation…).
The interval between the beginning of the symptoms and the diagnosis was between 4 to 12 weeks.
The localization of the tumor was cervical in 1case, cervicothoracic in 2, thoraco-lumbar in 6, thoracic in 6, Lumbar in 4, Lumbo-sacral in 2 and sacral in 1.
The quality of the surgical resection was accessed on a post-operative MRI realized within 48hours after the surgery: 4 cases of Near Total Removal, 13 cases of Growth Total Removal and 6 cases of Sub Total Removal.
Two patients presented a post-operative complication.
Histological diagnosis consisted of 4 myxopapillary ependymomas, 5 anaplastic ependymomas, 4 pilocytic astrocytomas, 4 metastases of supra tentorial tumors, 1 neuroblastoma, 1 unclassified malignant tumor, 1 embryonary rhabdomyosarcoma, 2 PNET and 1 gangliocytoma.
Thirteen patients received no complementary treatment, 5 chemotherapies only, 3 radiations only and 2 chemotherapy and radiations.
At last follow up, 12 patients were cured, 7 were in complete remission, 1 progressed and 3 died (2 because of disease progression).
Conclusions: SCT are very rare in children. Management required surgery and sometimes complementary treatment. The prognosis is good for benign tumors with a low morbidity.

FP56
Evaluation of tumor biopsies performed via intraventricular endoscopic approach

Elif Bolat, Taylan Özgür Kiliç, Tuncer Turhan (Izmir, Turkey)

Introduction: Endoscopic devices used especially in last 20 years have brought significant advances in neurosurgery. With the introduction of neurosurgical endoscopic procedures, accessing to challenging regions became possible with excellent point of view.
It is obvious that in order to establish worldwide standardization about efficacy and safety of endoscopic surgical procedures, neurosurgeons need more information about preoperative patient characteristics and postoperative success and complication results.
In this study we presented the outcomes of our 12-year experience of intraventricular endoscopic tumor biopsies.
Methods: In our descriptive study, between January 2005 and December 2017, demographics, mass localization, pathology results, presence of hydrocephalus, postoperative complications and mortality rates of the patients who underwent intraventricular endoscopic biopsy procedure in Ege University Department of Neurosurgery were retrospectively analyzed.
Results: Totally 65 patients underwent intraventricular endoscopic biopsy procedure. Of the patients, 32 were male (49.2%) and 33 were female (50.7%). Mean age of the patients was 237.9±238.6 (1.0-840.0) months.
The most common mass locations were pineal region (33.8%), third ventricul (15.4%) and suprasellar region (13.8%). Pathology results revealed colloid cyst in 10 (15%), pilocytic astrocytoma in 8 (12.3%) and low-grade glioma in 6 (9.2%) patients.
Fifty patients (76.9%) had hydrocephalus prior to the procedure and 48 (96%) of them underwent endoscopic third ventriculostomy and 2 of them (4%) underwent aquaduct stent placement. Short-term success rate of the procedure about hydrocephalus was 100%.
Fifty-six patients were completed the procedure without any complications, however the most commonly seen complication was intraventricular hemorrhage (13.8%) after endoscopic biopsy procedure. Overall mortality rate was 3.1% and the most common cause of mortality was peroperative intraventricular hemorrhage.
Conclusion: Intraventricular endoscopic tumor biopsy is a safe and effective technique in patients with various tumor locations.

FP57
Tumor progression pattern of recurrent pilocytic astrocytoma

Christos Chamilos, Panayiotis Kokkalis, Spyros Sgouros (Athens, Greece)

Introduction: Pilocytic astrocytomas are common brain tumors in children mostly occurring in the posterior fossa. Optimal management is gross – total resection and follow-up. The event-free survival is considered excellent.
Methods: We present a retrospective review of pediatric patients with pilocytic astrocytomas between 2007 and 2017 in a single institution. Clinical radiological and tumor characteristics were reviewed.
Results: There have been 23 cases (11male) with mean age at presentation 7.16 years and mean follow-up 3.5 years. There were 5 patients that had 2 operations and 1 had 3 operations. Two of the cases had a very early recurrence (under 1 year) and three cases had a late recurrence (more than 2 years). Three of the recurrences had a very small residue (<10mm) at post-op MRI. There are also 3 non-operated cases with a small residue that has not progressed.
Conclusions: In our series there is not a certain recognized pattern of tumor recurrence. The early recurrences didn't present certain pathologic markers that could explain the case (ki-67 or BRAF). On the other hand there were no significant correlation between small remnants at post-op scans and

recurrences. In our series even though a gross total removal was achieved and the survival rate is 100%, a significant rate of patients (21%) presented a recurrence.

Abstract session 19: Neuro-Oncology IV

FP58

Place of surgery in the treatment of SubEpedymal Giant-cell Astrocytoma (SEGA) in children. About a series over 15 years

Alexandru Szathmari, Pierre-Aurelien Beuriat, Federico Di Rocco, Isabelle Sabatier, Christophe Rousselle, Alexis Arzimanoglou, Carmine Mottolese (Lyon, France)

Introduction: Subependymal giant cell astrocytoma (SEGA) is a specific form of brain tumors often associated with Tuberous Sclerosis Complex (TSC). The purpose of this study is that of analyzing the therapeutic options for pediatric SEGA through our series.

Patients and Methods: Among 13 patients operated between 2001 and 2016 from an SEGA at the East Hospital Group of Lyon, 8 were children. These are 2 boys and 6 girls aged 2 to 14 (average 7). The average duration of follow-up was 6 years.

Results: Of the 8 patients, 7 had a known history of epilepsy and 5 are known to have TSC. In 5 patients, intracranial hypertension was related to an acute hydrocephalus. In these cases, the intracranial hypertension was severe with neurological and ophthalmological disorders (papilledema and visual acuity). The location of the tumor was in 7 cases in the right lateral ventricle, in 1 in the left ventricle and in 1 case there was an extension in the 3rd ventricle. All patients were operated on by a direct approach with complete excision in 6 cases. In 3 cases, a ventriculoperitoneal shunt was necessary before the excision. An anti-comitial treatment could be stopped in a patient. Treatment with mTOR inhibitor was not used.

Conclusions: Our analysis confirms the importance of systematic monitoring of SEGA by brain imaging to detect their evolution and prevent the appearance of signs of intracranial hypertension. Complete removal is possible, sometimes in two stages. Neo-adjuvant treatment with mTOR inhibitor was not used in our series due to diagnostic circumstances and due to the long-term concerns of this treatment in children.

FP59

Neurosurgical implications of neurofibromatosis type 1 in a single-institutional pediatric series

Veronica Saletti, Silvia Esposito, Luisa Chiapparini, Paolo Ferroli, Francesco Di Meco, Laura Valentini (Milan, Italy)

Introduction: Neurofibromatosis 1 (NF1) is an autosomal dominant, multi-system, tumor suppressor disorder with variable complications some of which may require the attention of a neurosurgeon.

Method: With the aim to explore the role of neurosurgery in a cohort of pediatric NF1 patients, we retrospectively reviewed records of 306 NF1 children at the IRCCS Foundation Carlo Besta Neurological Institute of Milan from 2009 to 2017.

Results: 18 of the 306 (5.9%) NF1 children underwent neurosurgical procedures; 7 of 18 patients (30.9%) required multiple surgeries for more than one lesion.

7 of 18 (38.9%) children, median age 7 years 6 months, underwent neurosurgery for brain tumors. 65 of the 306 (21.2%) NF1 children developed 68 brain tumors: 44 optic pathway gliomas (OPG) and 24 non-optic intracranial tumors. Neurosurgery was required in 1 of 44 (2.3%) OPG, in 4 of 8 (50%) hemispheric and cerebellar low-grade gliomas, in 3 high-grade gliomas and in 1 medulloblastoma.

7 of 18 (38.9%) children with paraspinal tumors, median age 12 years 6 months, required neurosurgery; histological diagnosis was MPNST in one patient and neurofibromas in the others. In addition 6 children (2,0%) required shunt insertion for hydrocephalus, and 2 children required neurosurgical procedures for moya-moya syndrome in one (0.3%) and cerebral cavernous malformation in the other.

Conclusion: Despite the high incidence of brain tumors in NF1 children, the role of neurosurgery appears to be limited. The majority of brain tumors in NF1 are represented by low-grade lesions with slow and indolent growth; however, there is a small but important risk that some of the extra-optic lesions may become high grade over time.

The role of neurosurgery appears prevalent at the spinal level in relation to the symptomatic paraspinal neurofibromas, particularly in adolescence.

Hydrocephalus and cerebrovascular anomalies are uncommon, but possible complications that can seriously complicate the course of the disease.

FP60

Neurofibromatosis type 1: a multidisciplinary approach from the pediatric neurosurgeon's point of view

Dalila Forte, Amets Sagarribay, Ana Cordeiro, Carolina Pinheiro, Sónia Gomes, Marta Amorim, Rita Silva, Telma Francisco, Miguel Correia, Mário Matos (Lisbon, Portugal)

Introduction: Neurofibromatosis type 1 (NF1) is a common genetic disorder caused by a mutation in the NF1 gene on chromosome 17 and is associated with multisistemic manifestations including susceptibility for benign and malignant tumors of the central and peripheral nervous system. NF1 patients are assessed regularly by an organised multidisciplinary team in our institution.

Purpose: To analyse a NF 1 pediatric population followed by a multidisciplinary team, considering particularly neurological manifestations and neurosurgical care.

Methods: Review of all NF 1 cases currently followed by the Neurocutaneous Diseases multidisciplinary team in Hospital

Dona Estefânia. All cases included until November 2017 in the team's database were reviewed. Results were compared with other series previous published in the literature.

Results: A total of 73 patients were included, with a median age of 11±5,6 years, male:female ratio being 1:1. Genetic testing was performed in 54 (74%) patients and molecular confirmation obtained in 45 (61,6%) patients. Family history was absent in 56,1% of our population.

NIH criteria were formally satisfied in 89% of the patients. Nearly all (72) patients presented six or more café-au-lait macules, 55% presented axillary or inguinal freckling, 38% presented two or more neurofibromas or one or more plexiform neurofibromas, 27% presented Lisch nodules and 7% presented sphenoid wing or long-bone dysplasia.

Regarding neuroimaging findings, 68% of the patients displayed Unidentified Bright Objects (UBOs) on cranial MRI, 11 (15%) presented scoliosis, 7 (9,5%) presented some degree of ventriculomegaly or ventricular dysmorphy. In one patient, choroid plexus hypertrophy was present. Four cases of vascular lesions were recorded, three cases of arterial stenosis and one developmental venous anomaly (AVD). Chiari type 1 malformation was present in four patients, two requiring surgery. Optic gliomas were documented in 21 (28,7%) patients, in 16 (21,9%) bilaterally. Plexiform neurofibromas were noted in 18 (24,6%) patients, involving predominantly the cervical and dorsal regions but also cranial nerves. Brain-stem lesions were found in four patients.

Neurological examination was altered in 13 (17,8%) patients. Developmental delay was present in 20 (27,4%), behaviour problems in 22 (30%) and learning disabilities in 32 (43,8%) patients.

Neurosurgical treatment was performed in 14 (19%) patients. The most frequent procedure was laminectomy for removal of neurofibromas, mainly cervical (6 patients), supratentorial craniotomy (3 patients) and infratentorial craniotomy (1 patient). Pathology revealed 2 pilocytic astrocytomas, 1 DNET and 1 Gangioglioma. Suboccipital craniectomy and C1 laminectomy was performed to treat a Chiari type 1 malformation. Two endoscopic third-ventriculostomies were performed and one ventriculo-peritoneal shunt was placed.

Conclusion: Neurological manifestations are common in NF1. Neurosurgical intervention is required in a small group of patients. Pediatric Neurosurgeons should be part of a multidisciplinary team in the long-term management of these patients, and contribute both in imaging surveillance and surgical intervention.

FP61

Surgical management of vestibular schwannomas in pediatric patients affected by neurofibromatosis type 2

Mario Giordano, Massimo Gallieni, A. Samii, Concezio Di Rocco, M. Samii (Hannover, Germany; Magdeburg, Germany)

Objective: Vestibular schwannomas (VS) affect young patients with Neurofibromatosis type 2 (NF2) and cause very serious problems for hearing, facial expression, and brainstem function. Our objective is to present our clinical experience with pediatric NF2 patients and to analyse the surgical indication, its timing and the factors influencing the surgical outcome.

Methods: We performed 11 surgical procedures for VS in 9 patients affected by NF2. The mean age at operation was 13.6 years (range 12-16 years). Eight tumors were located on the right and 3 on the left side. In two patients VSs on both sides were treated. One case had been previously treated in another department and had complete preoperative facial and hearing deficit. Regarding the remaining 10 surgeries: five had preoperatively serviceable hearing, 2 not-serviceable hearing and 3 anacusia on the operated tumor side; all the treated case had intact facial nerve function. All cases were treated using a retrosigmoid craniotomy in semisitting position. The patients' clinical symptoms and signs at the moment of the operation, the functional hearing outcomes, and follow-up are analysed.

Results: Total tumor removal was achieved in 10 surgical procedures. In 1 case**,** deliberately subtotal removal was performed for brainstem decompression and hearing preservation in the only hearing ear. Ten of the operated tumors (91%) were T4 VS or larger. After tumor resection, anatomic facial nerve preservation was achieved in all the 10 cases firstly treated in our Institution. Using the Gardner-Robertson scale only 2 patients had postoperative worsening of the hearing from serviceable to not-serviceable.

Conclusions: The goal of the surgical treatment in subjects with NF2 should be complete tumor excision. Actually, when operated on in specialized centers the total tumor removal can be obtained even in huge lesions with a great possibility of facial function preservation. In our experience, hearing can be preserved if still present before the operation, in a high proportion of cases though the risk of anacusia should be taken into consideration and discussed with the patient and the family.

FP62

Pediatric brain tumors of the first three years of life

Federico Bianchi, Paolo Frassanito, Luca Massimi, Massimo Caldarelli, Gianpiero Tamburrini (Rome, Italy)

Objective: The aim of this study was to review our institutional experience concerning children affected by brain tumors of the first three years of life and to compare it with literature data in order to remark the peculiarities of these tumors compared with those occurring during later children growth.

Methods: We reviewed all clinical charts of children aged less than 3 years, treated at our Institution for a brain tumor.

Results: 206 patients fulfilled the selection criteria (116M/89F, median age: 12 months). The most frequent clinical symptoms at diagnosis were raised ICP (59 cases), seizures (16 cases) and focal neurological deficits (56 cases).

There were 104 supratentorial cerebral tumors, 98 infratentorial ones and 3 meningeal tumors. Supratentorial tumors were histologically benign in 66% of the cases. 43 of them were located in the midline with a related mortality rate of 39%; 26 tumors were intraventricular and were related with a 19% mortality rate, whereas 35 were hemispheric and were related with a mortality rate of 37%. Infratentorial tumors were mostly malignant, 40 of them located at the level of the midline (37,5% mortality rate), 19 being intraventricular (mortality rate: 42%) and 38 hemispheric (39,5% rate of mortality).
Conclusion: We confirm a slight prevalence of supratentorial tumors in this age group. Supratentorial tumors were mostly benign, but interesting more frequently the midline or the ventricular system, with a consequent more relevant long term neurological impairment if compared with hemispheric tumors, more frequent in older children. Histologically malignant tumors were uncommon in the supratentorial space; their survival rate was better if compared with older children with a comparable histology. Infratentorial tumors were mostly malignant with an even more aggressive behavior compared with those occurring in older children.

FP63

Brain tumors of the first years of life: seven years of IOP/ GRAACC/ Federal University of Sao Paolo experience

Patricia Alessandra Dastoli, Jardel Mendonca Nicácio, Marcos Devanir da Silva Costa, Andrea Maria Cappellano, Frederico Adolfo Benevides Silva, Nasjla Saba Silva, Sergio Cavalheiro (Sao Paolo, Brazil)

Introduction: Among children with brain tumors, the babies have the worst prognosis and the most difficult treatment. Most of the brain tumors of the first year of life correspond to voluminous, malignant and highly vascularized masses. In spite of a high operative mortality, surgery still appears to be the more effective therapy. The postoperative chemotherapy isgiven in order to permit a delay in the delivery of radiation to developing brain or to enable more aggressive surgeries in better clinical conditions.
Objective: To analyze the several treatment strategies and outcomes ofbrain tumors in patients up to one year of age in ten years of experience in a single institution.
Methods: The authors retrospectively evaluated 63 patients under one year of age with brain tumors treated between 2010 and 2017, at IOP/GRAACC/UNIFESP. Data regarding initial clinical presentation, treatment modalities and outcomes were collected.
Results: From 63 patients under one year of age treated with brain tumors, 61 were eligible for evaluation and 2 were excluded for loss of follow-up. 30 were girls and 31 boys. The mean age at treatment was 6 months (range: 1 day-12 months). 29 babies presented with signs and symptoms of intracranial hypertension, 11 babies with epileptic seizures, 8 of them initially showed impairment of low cranial nerves. The other patients had other less frequent signs and symptoms. 45 tumors were located in the supratentorial compartment (23 cortical tumors, 8 suprasellars tumors, 8 tumors in the lateral ventricules, 4 optical pathways tumors and 2 pineal tumors) and 16 were infratentorially (10 cerebellar vermis, 6 brain stem tumors). Nine patients were diagnosed with tuberous sclerosis, 2 patients with neurofibromatosis type 1(NF1), 2 diagnosed with Li Fraumeni syndrome and 1 with Gorlin syndrome. The patients with tuberous sclerosis and NF1 were treated clinically. The most common histological types were :11 rhabdoid teratoid, 9 low grade astrocytoma, 5 choroid plexus carcinoma and 5 glioblastoma. Surgery is the treatment of choice. Malignant tumors which could not be completely resected in only one surgery required others surgeries alternated with cycles of chemotherapy. Ten patients underwent more than one surgery (2 to 6 resections) 8 deaths occurred. The mean follow-up was 3y10m (range: 2 days- 7y9m).
Conclusions: Treatment strategies for patients with tumors in the first year of life is still controversial. Gross total resection is the goal of surgical treatment, but sometimes this is impossible in the first approach. To decrease the high intraoperative mortality, these patients can undergo as many surgeries as necessary for total tumor resection. The malignant brain tumors were treated with postoperative chemotherapy.

FP64

Oncolytic virus for DIPG: a clinical trial

Sonia Tejada, Ricardo Diez-Valle, Marisol Gonzalez-Huarriz, Pablo Dominguez, Ana Patiño, Marta Alonso (Pamplona, Spain)

Introduction: Diffuse Intrinsic Pontine Gliomas (DIPG) are tumors with no effective treatment, where radiotherapy remains as the standard of care. Targeted therapies and specially immunotherapy could be the alternative to treat these infiltrative tumors, killing tumor cells and avoiding neurological deficits. Oncolytic adenoviruses are a type of immunotherapy with the capacity to wake up the patient´s own immune system. Adenovirus DNX-2401 is a conditionally replicating virus that has proven safe in glioma clinical trials in our institution and others.
Methods: A phase I clinical trial with DNX-2401 for patients with newly diagnosed DIPG to assess the MTD is taking place in our hospital (N=12). Tumor biopsy is performed through the cerebellar peduncle, followed by virus injection. The virus is injected using a new cannula for brain infusion, MEMS cannula (Alcyone Lifesciences) that prevents the reflux. Virus will be injected starting with 10^{10} pv. The trial is uncontrolled, unicentric with a 3 + 3 design.
Tumor sample from the biopsy is dedicated to genetic studies and for the generation of cell lines.
Results: All the clinical data from the trial available until May 2018 will be presented during the congress, to date just

one patient has been enrolled. Immediately after surgery an intraoperative MRI verified the location of the infusion and the absence of complications. The patient presented gait and swallowing impairment in the next 48 hours, with clinical improvement during the next week. Radiotherapy was initiated 3 weeks after virus injection. Biological studies are currently undergoing.

Conclusions: The infusion with the DNX-2401 of the first DIPG patient has shown to be safe. The patient was home one week after the biopsy and viral infusion. The information from this clinical could be very valuable to provide relevant safety data that allows for a radically different clinical trial for DIPG.

FP65

Diagnosis and treatment of pediatric brainstem tumors (a 30-year institutional review)

Liudmyla Verbova, Andriy Shaversky, Yury Orlov, Andriy Vashchenko, Tetiana Malysheva, Ivan Protcenko, Leonid Marushchnko, Andriy Svist, Mykola Plavsky, Volodymyr Mykchaliyuk, Ruslan Gavrish, Kristina Robak (Kyiv, Ukraine)

Background: Brainstem tumors comprise 10-25 % of all pediatric intracranial tumors.

Material and Methods: Between 1986 and 2015, 268 patients with the brainstem tumors were treated at the Institute: 134 male and 134 female. The age of patients was from 1 month to 16 years (average age – 9 years). According to the localization we used the classification: - midbrain tumors – 77 (≈29%), - pons – 56 (≈21%), - medulla – 25 (≈9 %), -pons + medulla – 91 (≈34%), -medulla + cervical spine – 19 (≈7%). CT and MRT identified growth pattern, exophytic component, cyst formation. The tumors were verified in 94 patients (≈35%): astrocytomas – 69 (≈73,4%), ependymomas – 17 (≈18%), glioblastomas – 3 (≈3,2%), PNET – 3 (≈3,2%), gemangioblastoma – 2 (≈2,2%). The symptom duration was from 1 week till 2 years. Clinical features were dominated by increased ICP syndrome in 188 patients (≈70%).

184 patients underwent next procedures: total resection – 3, subtotal resection – 17, partial removal – 35, cyst puncture and biopsy – 6, decompressive operation – 5, shunting operation – 118.

Results: 33 patients died at the clinic in different time; 14 – after tumor removal, 10 – after shunting operation, 9 – without surgery. The main reasons of vital complications: poor patients condition, giant tumors size, vascular disorders in brainstem and hemorrhage in the tumors. Follow –up data from 6 months to 16 years (average, 3 years) is available for 105 patients (≈45%).

Conclusions: Focal and cystic tumors and also the tumors with exophytic component require the surgical treatment. The midbrain tumors have slow growing nature and that's why these lesions don't require surgery till its progression. Malignant tumors need adjuvant therapy. Patient's condition, severity of neurologic deficit, tumor size influence on the treatment results.

FP66

Chiasmato-hypothalamic gliomas in children: there is still a place for an extended surgery?

Nicolas Pianton, Alexandru Szathmari, Pierre Aurelien Beuriat, Federico Di Rocco, Alexandre Vassilievic, Cecile Conter, Didier Frappaz, Linne Claude, Carmine Mottolese (Lyon, France)

Introduction: Chiasmato hypothalamic gliomas represent 5%-7% of all CNS system tumors and 65% occur in children less than 5 years old.

Generally, they are benign, chemiosensible and radio sensible. Their natural history is variable and the relationships between the disease and neurofibromatosis I are not completely clear. We report a series of patients focusing on the evolution of the disease related to different strategies.

Material: 19 patients have been treated between 2000 and 2017: 57% boys and 43% girls.

66% of patients presented visual problems: nystagmus 24%, oculomotor palsy 16% and reduced visual acuity in 26 %. 28 % presented intracranial hypertensive signs and 4% a precocious puberty.

18% of patients had a biopsy before chemotherapy; 25% of patients had a partial removal and 57 % an extended removal. 47 % of tumor were pylocytic astrocytomas, 28% diffuse astrocytomas and 25 % gangliocytomas.

48% of children were treated with chemotherapy and 13 % with radiotherapy.

Results: 45% of patients had a good quality of life with a good schooling, 19% mild sequels with an adapted schooling, 13% presented a visual deterioration and 24% of patients were bedridden. The analysis of visual troubles shoved: 58 % of patients presented an improvement of vision after surgery, 34 % a stabilization while 8% a loss of vision.

Conclusion: The aim of therapy in children with HCGLGG is to control tumor progression preserving function. A multidisciplinary discussion is necessary before clinical decision making. Chemotherapy represent the first line of treatment, radiotherapy is discussed in function of the evolution of the disease while surgery is reserved for progressive disease avoiding visual and endocrinological sequels for a good quality of life.

Poster Presentations

Poster track 1.1: Craniofacial I

PP001

Use of distraction osteogenesis for unicoronal craniosynostosis

Gregory Heuer, Ian Hoppe, Lawrence Lin, Rosaline Zhang, Scott Bartlett, Jesse Taylor (Philadelphia PA, USA)

Introduction: Patients with unicoronal craniosynostosis (UCS) are traditionally treated with frontoorbital advancement (FOA). While effective, FOA can associated with significant blood loss and a tendency for postoperative ocular dismotility. Distraction osteogenesis (DO) has been successfully applied to other craniofacial abnormalties. We describe the use of DO in the treatment of UCS, comparing the outcomes to FOA.
Method: Patients undergoing DO for UCS at our institution (11 patients) were compared to the most recent 11 patients undergoing FOA.Patient age, operative time, blood loss, blood replacement, length of stay (LOS), complications and the development of new onset strabismus following surgery were documented.
Results: A chi-square analysis and student's t test were utilized for analysis. Mean follow-up time was 23.2 months in the DO group and 31 months in the FOA group. Patients undergoing DO compared to FOA were significantly younger (5.3 and 9.1 months, $p < 0.01$), experienced significantly less operative time (119 and 192 minutes, $p < 0.01$), significantly less blood loss (146 and 373 ml, $p < 0.05$), and significantly less blood replacement (220 and 431 ml, $p < 0.05$). The length of stay for the initial procedure was less in the DO group compared to FOA, but did not reach significance (3.1 and 4.5 days, $p = 0.10$). The mean distance distracted was 36mm in the DO group. One patient in the DO group experienced a new-onset strabismus postoperatively compared with 5 in the FOA group.
Conclusions: DO for the treatment of UCS provides a favorable risk profile compared with traditional FOA.The trend towards a decreased incidence of postoperative strabismus suggests that gradual movement of the orbit is favorable with regards to the accommodation of ocular muscles to the new orbital position.

PP002

Computer assisted virtual planning and surgical templates manufacturing in craniosynostosis surgery

Juan Vicente Darriba Alles, Ángela Moreno Gutiérrez, Gemma Arenas de Frutos, Santiago Ochandiano Caicoya, Roberto García Leal (Madrid, Spain)

Introduction: Achieving the best aesthetic outcome using frontoorbital remodeling to treat craniosynostosis involving coronal and metopic sutures is complex and challenging. Computer assisted virtual planning and individualized surgical templates manufacturing is a useful tool for optimizing surgical results. We describe our results using this technique since 2016.
Methods: A virtual working model was generated using a 3D cranial CT scan obtained preoperatively. Frontoorbital remodeling virtual planning was carried out by simulating the osteotomies in the orbital bandeau and the frontal bones to achieve the desired configuration. This process was performed through a close collaboration between medical and engineering teams. Then the cutting guides and negative and positive stereolitography (SLI) templates were manufactured and used in the remodeling during surgical procedure. Bone fragments were fixed with resorbable material (poli- D, L - lactic mesh, plates and pins) using ultrasound technique.
Results: Six patients underwent surgery for correction of trigonocephaly (4 patients) and anterior plagiocephaly (2 patients). The medium age at the time of surgery was 9 months. Both intra- and postoperative results were satisfactory. No surgery complications occurred to date. The use of surgical individualized preformed SLA templates allows a simple, accurate and expeditious handling during surgery procedure as well as excellent result in craniofacial remodeling.
Conclusion: Virtual planning and computed assisted SLA templates modeling have been shown to be interesting and advantageous tools for the successful treatment of craniofacial deformities that may be considered.

PP003

TCF 12-associated craniosynostosis: clinical series of 17 patients

Marina Brigui, Federica Ruggiero, Louise Wilson, Justine O'Hara, David Dunaway, Owase Jeelani, Gregory James (London, United Kingdom)

Introduction: TCF12 mutation is a recently described genetic aetiology of syndromic craniosynostosis. We review the clinical characteristics of this syndrome in a series of children from our institution.
Methods: We identified all children referred to our Craniofacial Service in which a confirmed genetic diagnosis of TCF12 mutation was found and undertook a proforma-based review of clinical records, radiological images and photographs.
Results: 17 consecutive patients aged 1-20 years were identified, including 9 boys and 8 girls. All 17 patients had craniosynostosis. 14 children had bicoronal synostosis and 3 unicoronal. A severe degree of brachyturricephaly was noted in the majority of bicoronal cases. 16 of 17 children underwent surgical correction. The average age at first surgery was 20 months. 6 children needed more than one operation (most commonly fronto-orbital advancement remodelling following initial spring-assisted posterior vault expansion). In these 6 this was an expected 2 stage surgical plan, due to the severity of the brachyturricephaly. 1 child had hearing loss and abnormal limbs, and 1 child had malformed toes. Developmental delay was found in 3 patients. No cases of intracranial hypertension were identified.
Conclusion: This is the largest UK clinical series to date and adds significantly to global experience. Despite the severity of the clinical phenotype in terms of skull deformity, ophthalmic evidence of intracranial hypertension was not seen in our TCF12 cohort. Developmental delay in this group does not

appear to correspond with intracranial hypertension. Many children required both posterior and anterior expansion to correct the severe brachyturricephaly that appears to be characteristic of the syndrome.

PP004

Frontal widening and remodeling for scaphocephalic children older than one

Eric Arnaud, X.L. Jing, Giovanna Paternoster, Syril James, X. Liu, Michel Zerah (Paris, France)

Introduction: Surgical correction for scaphocephaly is recommended before 6 months of age to provide more complete skull remodeling with a better mental outcome. However, in late presenting patients, alternative techniques may be used to address transverse frontal narrowing and anterior bulge.

Patients and Methods: Among a larger group of operated patients, eighteen children aged between 15 months and 6 years were retrospectively evaluated because of sufficient imaging. Mean age at surgery was 28 months and follow up was 23 months (Minimum 1 year and maximum 4 years). The forehead was split in two halves and a 1 cm wide strip of bone was fixed in the middle. A bilateral advancement was provided by sagittal split in both lateral aspects of upper orbits. The morphology of the forehead was assessed by two measurements on the imaging before surgery and immediately after in most patients, and 1 year after in 5 patients only because of the limitation of irradiation.

The anterior bulging was assessed by the fronto-nasal angle (FNA), corresponding to the angle between the lines tangential to the nasal bone and to the most prominent part of the forehead taking the nasion as the summit.

Transverse diameters of the forehead were assessed by ratios of the most transverse width over transverse distance between fronto-zygomatic junctions.

Statistical analysis was carried out using Student paired and unpaired t-tests.

Results: In all patients but one a transverse widening and a posterior tilting of the forehead were obtained ($p<0.05$). In one patient, the midline sagittal strip of the forehead consolidated with a midline ridge. The transverse dimension of forehead increased and remained stable at time of follow-up.

Discussion: It is recommended to address sagittal synostosis correction before 6 months of age. However, in patients operated late after 12 months of age, some forehead remodeling needs to be performed in order achieve better esthetic results.

PP005

Early Preemptive Transfusion Infusion (EPTI) approach for pediatric complex cranial vault reconstructive surgery: a single center observational study

Mirella Seveso, Dario Caldiroli, Marika Furlanetto, Laura Valentini (Milan, Italy)

Background: Significant blood loss during intra-operative vault surgical remodelling represents the principal issues in pediatric patients harbouring craniosynostosis. Adequate evaluation of blood loss is difficult, and literature only suggest context-sensitive approach to transfusion according to patient characteristics and clinical settings. A pilot study[1] conducted by our Institution defined and demonstrated that EPTI approach may improve the well–being outcomes (re-feeding time and LOS) in infants undergoing open cranioplasty[2]. In the present study we retrospectively analyzed our outcomes on a larger series of infants after protocol standardization. We also addressed the pre-emptive ERCM (estimated red cell mass) as the constant variable in our transfusional program.

Methods: A working database comprising clinical data, electronic charts, neuroradiological images, intraoperative notes, laboratory exams and infusional-transfusional requirements of 92 patients operated from 2010 to 2015 was created. The pre-emptive ERCM was calculated based on Kearney formula and the difference between total transfused RCM and pre-emptive was analysed.

Results: The median re-feeding time was 4 hours, only one patient required ICU admission; the median LOS was 6 days and only 19.6% (N=18) received postoperative transfusions. Median total intraoperative crystalloid infusion was 104 ml/Kg (range 50-200 ml/Kg). Colloid use was not a routine practice (N=12 patients). Median intraoperative fresh frozen plasma trasfusion was 9,5 ml/Kg (range 0-60 ml/Kg). Median total transfused RCM (Red Cell Mass) was 187 ml (range 68 – -377 ml). Only in three patients the total transfused RCM was inferior to calculated pre-emptive ERCM. In 33 patients (36%) total transfused RCM was at least twice pre-emptive ERCM.

Conclusion: Our data confirm that EPTI strategy improves well-being outcomes and recovery. Further prospective studies in a larger series and different settings are needed to validate the early standardized approach.

References:

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PP006

Results of experience with endoscopic craniectomy for craniosynostosis treatment

Ricardo Gomez-Espinosa, Jesus Gomez-Plascencia (Guadalajara, Mexico)

Introduction: Results of craniosynostosis treatment have been notoriously improved after the introduction of endoscopic craniectomy techniques, resulting in shorter hospital stay,

diminished blood loss, less operative time with less exposition to anesthesia, all of which result in a reduced global risk for the patients and lesser economic burden.

The present report compares the results between the first endoscopic craniectomy for craniosynostosis, just after the author's training in neuroendoscopy, and the last one, performed in his daily practice.

Methods: Results regarding hospital stay, surgical time and anesthesia time and blood transfusion of the first early endoscopic craniectomy performed by the autor just upon his return from training periods at two european neurosurgical centers of excellence were compared to those of the last one performed after his return to his daily practice; both patients presented with scaphocephaly and both underwent endoscopic midline craniectomy through incisions 2.5 cm in endoscopic craniectomy length, 1 cm posterior to coronal and 1cm anterior to lambdoid sutures.

Results: In both cases with identical pathologies there were no surgical nor anesthetic complications. The first case had longer surgical and anesthetic times and required blood transfusion, and presented two 1 and 3 mm respectively, dural tears which were repaired without further complications. The second case did not require blood transfusion and did not have any dural tears.

Conclusions: Results confirm that endoscopic craniectomy is a safe technique for early correction of craniosynostosis; the difference found in surgical and anesthetic times as well as in the transfusion required can be interpreted as due to more experience with the procedure. As any surgical technique endoscopic craniectomy must have a specific training to avoid serious complications.

Poster track 1.2: Craniofacial II

PP007

Robinow syndrome in a newborn presenting with hydrocephalus and craniosynostosis

Koichiro Sakamoto, Sandro von Däniken, Bedirhan Boztepe, Kazuaki Shimoji, Takaoki Kimura, Masakazu Miyajima, Hajime Arai (Urayasu, Japan; Bern, Switzerland; Vienna, Austria)

Background: Robinow syndrome is a rare entity with a characteristic appearance, such as hypertelorism, short stature, mesomelic shortening of the limbs, hypoplastic genitalia, and rib and vertebral anomalies. We had treated a case of Robinow syndrome who developed hydrocephalus and craniosynostosis which is not usually associated.

Case: The ventricle enlargement was detected during pregnancy in this baby girl. She did not develop hydrocephalus just after birth. Her facial appearance was fetus-like, so the pediatricians had suspected Robinow syndrome. During follow up, her head circumference enlarged rapidly when she was 3 months old. Her conscious level was not disturbed but she had a tight fontanel and sunset phenomenon was recognized. Hydrocephalus was detected with images so that she underwent ventriculo-peritoneal shunting (VPS). Her head enlargement seized. 6 months after surgery, her parents noticed the brachycephalic shape of her head. A CT and MRI scan were conducted and her bilateral coronal, bilateral lambdoid and the sagittal suture were fused in addition with a tonsillar herniation. Since the sutures were not remaining, we diagnosed that this was a primary pansynostosis rather than secondary craniosynostosis due to VPS. Posterior cranial vault distraction with foramen magnum decompression (FMD) was conducted. The distraction was performed by 1mm per day up to 30mm. After a consolidation period of 2 months, the distractors were removed. This distraction resulted in a 15.4% increase (+196cc) of the intracranial space with an improvement of the chronic tonsillar herniation.

To confirm the diagnosis, a genetic testing was conducted. The analysis showed ROR2 Exon3 (c233 c>t p. Thr 78 Met), which is found in the recessive type of Robinow syndrome.

Conclusion: We report this case as the first to our best knowledge of Robinow Disease presenting with hydrocephalus and craniosynostosis. Posterior cranial vault distraction with FMD is a useful way to treat this condition.

PP008

Brain anomalies in orofaciaodigital type 1 syndrome: combined clinical and fundamental approach

Pernelle Pulh, Brunella Franco, Paul T. Sharpe, Roman Hossein Khonsari (Paris, France; Naples, Italy; London, United Kingdom)

Aim and Scope: Brain anomalies in orofaciaodigital type 1 (OFD1) syndrome are not well described and their origin is not well understood. OFD1 is a cilia-related protein and its mutation interferes with the Sonic Hedgehod (SHH) signaling pathway.

Material and Methods: Here we combined clinical MRI data from a family with OFD1 syndrome and expression patterns of SHH-related genes (*Gli1*, *Gli2*, *Axin2*, *Ptch1*, *Shh*) in the brain of mice with an *OFD1* knock-out mutation at various stages of development (E12, E14, E16, E18).

Results and Discussion: The morphology of the brain in OFD1 corresponds to the previously described pattern of brain malformations in cilia-related craniofacial syndromes. The specific disturbance of ventralizing SHH-related genes helps to understand the origin of these anomalies.

PP009

Macroglossia after craniofacial and posterior fossa surgery in children

Roman Hossein Khonsari, Jebrane Bouaoud, Thierry Briac, Syril James, Ian Henessy, Antonio Vecchione, Eric Arnaud (Paris, France; Dublin, Ireland)

Aim: Acute tongue swelling after craniofacial and posterior fossa surgery has been rarely described and its management is challenging. Here we report 3 cases of acute macroglossia occurring in children after neurosurgical and craniofacial procedures and discuss the treatment options in this condition.
Methods: We report 3 cases of acute macroglossia occurring in children after neurosurgical procedures and review the literature on the subject.
Results: A 15-year-old boy with Hurler syndrome and a 12-year-old girl with Down syndrome and Chiari malformation underwent neurosurgical procedures in prone position for respectively foramen magnum decompression and posterior vault distraction. Both children had previous moderate macroglossia due to their congenital conditions. A 20 months-old boy was operated on for a posterior fossa tumor in sitting position and similarly developed acute post-operative macroglossia. In all three cases, acute macroglossia caused extreme difficulties in airway management. One patient required partial glossectomy due to extensive tongue necrosis and another required urgent mandibular distraction in order to provide space for the tongue.
Conclusions: Factors usually incriminated in acute macroglossia after prolonged prone or sitting position during surgery are lymphatic, venous and arterial obstructions and neurogenic edema. Here in two of the cases, the previous congenital moderate macroglossia may have favored the onset of the tongue swelling. Based on these three cases and an extensive review of the literature, we propose guidelines in order to prevent this adverse event; based on simple peri-operative measures and post-operative management.

PP010

Craniosynostosis in children with hypophosphatasia

Vadim Ivanov, Alexander Kim, Natalia Kolbina (St. Petersburg, Russia)

Introduction: Hypophosphatasia is a rare metabolic disease with incidence approximately 1:100000. Clinical picture can vary widely, including cases with complex craniosynostosis. Described occurrence of craniosynostosis in children with hypophosphatasia is about 30-40%. A distinctive feature of Craniosynostosis in Hypophosphatasia is the involvement of some cranial sutures in the formation of Pancraniosynostosis.
Materials and Methods: Since 2016 in Almazov National Medical Research Centre were treated 10 patients with hypophosphatase. Craniosynostis was detected in 2 patients: a 3-year-old-child with pancranostenosis and a child of 5 years with a saggital suture synostis. Both children received a substitution therapy asphotasealfa. Tactic of surgical treatment was determined according to the Neuroimaging Methods of examination, cereblal's oximetry data, clinical expertise.
Results: An analysis of the treatment literature facts indicate a remitting character of the formation of craniostenosis in children with hypophosphatase. The method of the choice in such case cases can may be a distraction methods of correction of craniosynostosis. The combination of a surgical correction with a drug therapy also can affect the frequency of restonosis formation and the skull defects persistence after performed correction.
Conclusions: The planning of the surgical treatment of craniostenoses in children with hypophosphatosis should take into account the particularities of the bone tissue regeneration in this pathology, considering the medication therapy. Expediently performing screening of the hypophosphatase for children with combined forms of craniosynostosis in the absence of a phenotypically obvious syndromal form of craniosynostosis.

PP011

The impact of surgery timing for craniosynostosis on cognitive outcomes: a systematic review

Rachel Mandela, Maggie Bellew, Paul Chumas, Hannah Nash (Leeds, United Kingdom)

Introduction: Research has established that craniosynostosis is associated with higher incidence of learning difficulties and that patients with this condition have a lower distribution of scores on cognitive tests than norms. This systematic review seeks to address the question of whether there is an optimal age for surgery in terms of neurodevelopmental outcomes.
Methods: Databases were searched between October–November 2016 and searches repeated on the 11th July 2017.Search terms included *children, craniosynostosis, cognitive* and *developmental*.
Studies were selected according to PICOS criteria: *Participants* - were children diagnosed with non-syndromic craniosynostosis, who were no older than five years of age at timing of surgery. *Intervention* - studies focused on corrective surgery for craniosynostosis. *Comparison* - studies categorised and compared groups of children who underwent surgery at different ages. Studies (such as those employing a correlational design) which did not compare different age-at-surgery groups were excluded. *Outcome* - Studies which conducted tests of cognitive and neurodevelopmental outcomes in children after surgery were included. Quality was assessed using the Quality Assessment Tool for Studies with Diverse Design.
Results: Ten studies met the criteria. Five studies found a beneficial effect of earlier surgery and five did not. The average quality score was slightly higher in the studies that did not find an effect, but the two highest quality studies did find an effect.
Conclusions: The results of the review were inconclusive. However, although only half the studies found a beneficial effect for earlier surgery, no studies found a beneficial effect of later surgery.

PP012

Skull X-rays in the diagnosis of abnormal skull shapes

Dylan Murray, John Caird, Hugh O'Sullivan (Dublin, Ireland)

Introduction: Differentiation between plagiocephaly and craniosynostosis continues to provide a challenge for the clinician. Historically, initial investigations would have included a skull x-ray. However, it is widely accepted that this investigation is difficult to interpret and its value in diagnosis of craniosynostosis is debated. Aside from the burden this places on radiology departments, it also exposes children to mutagenic ionising radiation.
Aim and Objectives: Examine the correlation between reported skull x-rays and clinical diagnosis of craniosynostosis.
Methods: Retrospective chart review was conducted as a 2-part study in children who were referred with a skull x-ray.
Part A: Children referred to the NPCC with a suspected skull abnormality between 1st January 2015 and 30th May 2017
Part B: Children who underwent surgery for a confirmed craniosynostosis between 1st January 2011 - 25th October 2017.
Results and Findings: Part A: 300 children were referred with 59 skull x-rays. This represented 20% of all patients referred during the time period. Of these 44 (73%) were found to be a match with 15 (27%) not matching the final clinical diagnosis.
Part B: 274 children underwent surgery for a confirmed craniosynostosis between 1st January 2011 - 25th October 2017. 63 pts had skull x-ray on referral- this represents 23% of all operated children in the time period. Of these 41 (63%) were found to be a match with 17 (29%) not matching the final clinical diagnosis. 5 (8%) were inconclusive.
Conclusions: Part A: 25% of children referred to the NPCC and 92% of children who underwent surgery at the NPCC the x-ray did not contribute to their management.
Clinicians should check with the NPCC with respect to the protocol for x-rays where craniosynostosis is suspected as in most cases the diagnosis is clinically obvious to the craniofacial surgeon.

Poster track 1.3: Vascular

PP013

Vascular diseases in Pediatric Neurosurgery

Martin Scholz, Friedhelm Brassel, Thorsten Rosenbaum, Ameer Alyeldien, Lutz Schreiber (Duisburg, Germany)

Objective: The vascular diseases are rare in paediatric neurosurgery, nevertheless they are very important diseases.
The vascular pathologies as angiomas, Vena-Galeni-Malformations and cerebral ischemia need a fast and interdisciplinary management by neuro-radiologists, neurosurgeons and pediatrics.
Methods: A case series of 4 special cases of paediatric vascular diseases, which are managed in our department in the last 3 years, will be presented. A bleeding angioma, cerebellar ischemia in Kawasaki-Syndrome, a nerve-conflict of an AICA-Loop inside the porus acusticus internus and a spontaneous bleeding of an unknown Vena-Galeni-Malformation in a 14 years-old boy will be presented.
Results: All patients need a special individualized and interdisciplinary teamwork and management to reach the optimal outcomes.
Conclusions: The interdisciplinary teamwork offers the patients a fast and optimal treatment and of course the best outcomes.
An interdepartmental synergy and cooperation is very important to create a team that facilitate and optimize the management of the paediatric vascular diseases on a standard manner.

PP014

Aggressive Moyamoya disease in an infant

Amets Sagarribay, Dalila Forte, Gonçalo Januário, Miguel Correia, Rita Silva, Isabel Fragata, Carla Conceição, Mário Matos (Lisbon, Portugal)

Introduction: Moyamoya is a rare disease in Western countries, mainly in Europe, with an incidence that ranges from 0.5 in East Asia to 0.08 per 100.000 population. Characterized by progressive bilateral distal ICA stenosis and the formation of abnormal vascular network, it can be divided in two incidence peaks: at 5 to 15 and at 30 to 40 years of age, been females twice affected. Presentation in children is typically related to ischemic events, 40% as TIA and 40% as stroke. There are few reports of its presentation at infant stage as a result of fast aggressive evolution.
Material and Methods: The authors describe a case of a six months white male with aggressive moyamoya disease diagnosed after stroke and review the literature.
Results: A six months white male was admitted to our emergency department after seizure and new onset of hemipharesis. There were no previous history of either infection disease or fever. There were no haematological problems, neurocutaneous stigmata or familial history, Down´s syndrome, congenital heart disease or perinatal adverse events. CT scan, MRI/MRA, diffusion-perfusion MRI and cerebral angiography revealed bilateral carotid stenosis, moyamoya vessels, bilateral cerebral hypoperfusion and ischemic changes at distal cortical áreas. The authors initiated antiplatelet therapy and performed a bilateral multiple burr hole indirect revascularization procedure. The postoperative period went uneventful. Six months after surgery images showed significant bilateral increase in cerebral perfusion with multiple ECA tributaries entering cranial volt at burr holes site. From neurological point of view he has a normal development and had a progressive recovery from hemipharesis. The authors show dynamic angiography and diffusion-perfusion MRI images pre- and postoperative, and surgical procedure.

Discussion: There are few reports of moyamoya disease diagnosed at first year of life. From the author´s point of view the best surgical technique in this scenario is multiple burr holes indirect revascularization because it enables an easy technical procedure, bilateral approach, multiple anastomosis points, optimal revascularization and low complication rate.

PP015

The management of subdural hematomas in newborns: a case series

Peter Spazzapan, Tomaž Velnar (Ljubljana, Slovenia)

Introduction: Subdural haematomas may be frequently encountered in newborns and young children. They may result as a consequence of various pathology, such as metabolic diseases, trauma, child abuse, coagulopaties, prematurity, forced delivery or vascular pathology. In case of neurological symptoms and brain compression, a surgical intervention is necessary. We describe the symptoms, surgical outcome and prognosis of children with subdural haemorrhage of various aetiologies.
Methods: From 2015 to 2017, children treated at our centre due to subdural haematomas of were evaluated. A retrospective analysis of clinical data was performed. The clinical condition, neurological condition, mode of treatment and outcome were taken into account.
Results: In this time period, seven children were treated at our department. The median age was 1 year. Six children were operated on under emergency conditions due to brain oedema and herniation. In one, the haematoma resolved spontaneously. Five children had symptoms of elevated intracranial pressure within the first 24 hours of life, in one the neurological deterioration occurred after 24 hours. Each patient had a cranial ultrasound followed by MR that confirmed the bleeding. The result of bleeding was in two respective cases birth trauma and child abuse, in four prematurity and in one coagulopathy. With a mean follow-up of 6 months, one patient died and one had mild developmental delay. In other, a normal development was observed.
Conclusion: In neonates and small children with subdural bleeding, surgery can be safely performed in those who have clinical deterioration and radiological signs of brain oedema or compression. As a result of a good long-term neurological outcome, aggressive surgical and supportive treatment should be carried out.

PP016

Vein of Galen aneurysmal malformation. Report of 3 cases

Peter Spazzapan, Tomaž Velnar, Zoran Milošević (Ljubljana, Slovenia)

Introduction: The vein of Galen aneurysmal malformation (VGAM) is a rare intracranial arteriovenous malformation (AVM), that can cause severe morbidity and mortality in early postnatal period and during childhood. Arterial blood from the choroidal arteries drains directly into the median vein of the prosencephalon, which is the precursor of the vein of Galen.
Methods: We retrospectively analayzed three cases of VGAM diagnosed and treated in our Institution between 2010 and 2015. We analyzed the angioarchitecture of the vascular lesion, the symptoms, the treatment modality and the outcomes of the three patients.
Results: All three cases of VGAM diagnosed and treated in the early neonatal period by neuroradiological embolization. One child had a hydrocephalus, which resolved after embolization and ETV. All three patients had an excellent outcome at a mean follow-up of 5 years.
Conclusions: Patients with VGAM present different age-related clinical manifestations. In the neonatal period the predominant symptoms are related to a high cardiac output and to consequent cardiac failure. Management of VGAM poses many therapeutic challanges. Neuroradiological intervention has shown to control cardiac failure and to occlude the AVM with much less risks compared to neurosurgical intervention. Hydrocephalus, which may be associated to VGAM is caused mainly by venous hypertension. With an early and correct diagnosis and treatment these children can develop normally and have a normal neurological outcome.

PP017

Spontaneous giant basilar tip aneurysm in a child presenting as obstructive hydrocephalus

Dalila Forte, Luís Cardoso, Gonçalo Januário, Ricardo Nogueira, João Reis, Miguel Correia, Amets Sagarribay, Mário Matos (Lisbon, Portugal)

Introduction: Pediatric intracranial aneurysms are rare. Giant aneurysms (≥25mm) are found more frequently in children than in adults, and favour particularly the vertebrobasilar system.
Case Description: A previously healthy 12-year-old boy presented with a 2-month history of progressively worsening headache and intermittent vomiting. Cranial Computed tomography (CT) revealed a circumferential, hyperdense lesion in the third ventricle and associated obstructive hydrocephalus. Brain magnetic resonance imaging (MRI) revealed an aneurysm of the basilar artery compressing the midbrain and the sylvian aqueduct with hyperactive flux. No signs of subarachnoid haemorrhage (SAH) were present. Neurological deterioration occurred with installation of anisocoria and left hemiparesis after 48h without CT scan changes. Cerebral angiography was performed revealing an aneurysm (25,7 x 33,3mm) at the bifurcation of the basilar artery, oriented superiorly and posteriorly. Since it was impossible to identify the origin of the right posterior cerebral artery, partial occlusion of the aneurysm was

obtained with coiling. Ventriculoperitoneal (VP) shunting was carried out afterwards using a programmable pressure valve. Control CT scan after endovascular treatment and VP shunt placement showed no signs of SAH and reduction of ventricular size. The patient showed progressive clinical improvement but presented with transient drowsiness and left hemiparesis. Control MRI after 2 months showed adequate control of the hydrocephalus with no signs of hyperdrainage and persistence of residual aneurysmal flow without enlargement of the sac. Bilateral infarct of the cingulate gyrus and corpus callosum area was apparent, probably due to decompression-related vascular lesion after shunting, thus explaining the transient clinical worsening. A multidisciplinary rehabilitation program was started with progressive improvement and definitive endovascular treatment will be scheduled.

Conclusion: Basilar artery aneurysms with associated hydrocephalus have complex underlying physiological mechanisms. Multimodal, multi-staged treatment including endovascular and neurosurgical is appropriate. Lifelong follow-up is essential due to high recurrence and "de novo" aneurysm formation rates in pediatric population.

PP018

RBM3 in neuroprotection and neurogenesis after experimental ischemic stroke

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Introduction: Moderate cerebral hypothermia (32-33°C) is a potent therapeutic tool to decrease brain damage after e.g. hypoxic-ischemic encephalopathy. The RNA-binding protein RBM3 has been identified as a mediator of neuroprotection afforded by hypothermia in-vitro and in-vivo. In the adult brain, RBM3 expression is present in neurogenic niches but it is unclear whether RBM3 can regulate neural stem/progenitor cells (NSPCs) proliferation and promote neurogenesis after ischemic stroke.

Methods: C57BL/6J RBM3 wild-type (WT) and knock-out (KO) mice were used in this study. For in-vitro experiments, NSPCs were isolated from 2-3-month-old adult mice and oxygen glucose deprivation-reoxygenation (OGD/R) was performed. For ischemic stroke in-vivo, the right common carotid artery (RCCA) of adult mice was ligated, animals were exposed to 8% oxygen for 20 min at 37°C and injected serially with BrdU. Seven days after stroke the infarction size was analyzed and the numbers of proliferating NSPCs and newborn neuroblasts were counted. Putative interactors of RBM3 were analyzed in NSPCs and further characterized in-vivo to identify downstream effectors of RBM3.

Results: Without OGD/R stress, proliferation and neurosphere formation did not differ between RBM3 WT and KO NSPCs. In contrast, after OGD/R stress the absence of RBM3 resulted in less BrdU+ cells. In the stroke model there was little difference of infarction volume between RBM3 WT and KO mice. In WT mice the number of proliferating NSPC and newborn neuroblasts in ipsilateral subgranular zone (SGZ) was significantly larger than in contralateral SGZ. In RBM3 KO mice the induction of neurogenesis was absent. No difference was found in neurogenesis in subventricular zone upon RBM3 expression. On the molecular level we found evidence that RBM3 interacts with the insulin growth factor pathway, which is involved in NSPCs maintenance and neurogenesis.

Conclusion: Our data suggest that RBM3 is involved in promoting neurogenesis upon hypoxic-ischemic brain injury.

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Poster track 1.4: Trauma

PP019

Landmines from the neurosurgical perspectives in children

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Introduction: Landmines are the most dangerous and insidious war weapons. Although they are mostly used in war times, these weapons are also used in terrorist action, especially hand-made landmines. Military personnel are the usual targets of the landmines, but children also also affected especially during the in-city conflicts.

Patients and Methods: 35 patients were transferred to our hospital with the head or spine injuries secondary to landmine explosions.

Results: 22 patients were injured with stone and mud, but 13 patients were injured with metallic fragments. 2 of 35 patients were children. They had head injuries with metallic fragments after the land mines explosions. Both of them were with their mothers during the explosions. They underwent removal of the fragments and discharged without any deficit.

Conclusion: Children were also the victims of landmine explosions. Their treatment is the same with adults. But close follow-up and psychological support are needed for these children.

PP020

Early resorption of acute subdural hematoma in a child

Ozkan Tehli, Yusuf Izci (Ankara, Turkey)

Background: Acute traumatic subdural hematomas are usually lethal and require emergent surgical treatment. But cranial anatomy and physiology of children is different than the adults.

Case Report: A 2-years old male patient presented with the history of head trauma. He had a transient memory loss, but he

was neurologically intact at admission. Computed tomography revealed an acute subdural hematoma at the left frontoparietal region. The thickness of the hematoma was between 1 and 2 cm, and there was no midline shift. As the child had no neurological deficit and no shift, we observed the child for 3 days. At the end of follow-up, the computed tomography showed spontaneous resorption of the subdural hematoma and the child discharged without any neurological deficit.
Discussion: Acute subdural hematoma might be recovered spontaneously. Cerebrospinal fluid may have a role on the cleaning of subdural hematoma.

PP021
Facing traumatic brain injuries in children and young adults
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Aim: Aim of this study was to highlight the etiological pattern and the distribution of Traumatic Brain Injuries in Children and Young Adults (<18 years).
Methods and Results: From January 2014 to January 2016 (24 months), more than 400 young patients presenting to Volos General Hospital Emergency Department with Traumatic Brain Injuries were included in this study.
All the patients underwent clinical and radiological evaluation. Injuries were caused mainly by road traffic accidents (200-50%), followed by falls-domestic accidents (100-25%) and other types (assault, sport injuries, other) (100-25%).
Only 20% (40) of the road traffic accidents were wearing safety equipment. Motor bike and bicycle accidents in male patients are the most common causes of Traumatic Brain Injuries (63,63%).
Discussion: The appropriate pediatric neurosurgical care (3 consultant neurosurgeons) and the neurosurgical evaluation are very important in order to minimize the serious consequences of the Traumatic Brain Injuries. Safety equipment and safety driving can minimize serious traumatic consequences.

PP022
Bilateral split decompressive craniotomy in the management of cerebral edema caused by acute necrotizing encephalitis in a child with known ANA IgA vasculitis
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Introduction: Acute necrotising encephalitis is a severe form of influenza related encephalopathy and is most commonly due to influenza A infection. It causes death or very severe neurological disability in approximately 70% of patients. Patients may develop altered mental status which rapidly progresses to coma within 24-72 hours of fever onset and may present with or without seizures. Here we present the case of a child who was diagnosed with acute necrotising encephalopathy on the background of a previous diagnosis of ANA positive IgA vasculitis.
Methods: A 13-year-old male presented to our unit acutely with a deterioration in consciousness (GCS = 8). This followed a 3-day history of fever, lethargy, two episodes of vomiting and some confusion. He was intubated and ventilated on arrival. Initial brain imaging showed cerebral oedema and was suggestive of encephalitis with no evidence of abscess or empyema. Virology confirmed Influenza A H3N2 infection. He was initially managed on dexamethasone, IVIG and Tamiflu. An ICP bolt was inserted following a seizure like episode. Intracranial pressure continued to rise, despite medical management with mannitol, hypertonic saline, hyperventilation and sedation. He underwent a bilateral split decompressive craniotomy.
Results: Post-operatively he developed a DVT requiring insertion of an IVC filter due to risk of intracranial haemorrhage. He made a slow neurological recovery post-operatively and was discharged to a neuro-rehabilitation unit 3 months after his initial presentation, having undergone extensive inpatient rehabilitation. At last review, 7 months post-operatively, he was making good progress and has been discharged from rehabilitation services.
Conclusion: Acute necrotising encephalitis is a life-threatening condition. Decompressive craniotomy can be used successfully to manage refractory intracranial hypertension with a good functional recovery post-operatively.

PP023
Decompressive craniectomy in case of Herpes Encephalitis
Alya Hasan, Lamya Alsarraf (Shuweikh, Kuwait)

Introduction: Herpes simplex encephalitis is treated medically and uncommonly requires acute interventional surgery. We will present a case that required decompressive craniectomy post brain edema related to encephalitis and focal status epilepticus.
Methods: A case review of a serologically proven herpes encephalitis case of a 2-year female that required surgical intervention.
Results: This 2.5-year-old female presented with abnormal behavior, speech arrest and staring. She was started on anti-epiletics. A MRI brain revealed a small area of hemorrhagic focus in the right anterior inferior temporal gyrus area with surrounding perifocal edema. The child became unresponsive no abnormal movements and developed a fever. An EEG revealed focal status epilepticus originating from the right side. A follow up MRI revealed cortical gryral effacement right temporal, cingulate gyri; subfrontal areas right and left; along with uncal herniation and a significant midline shift right to left. The imaging findings were attributed to her encephalitis and focal status epilepticus. Clinically the child deteriorated,

developing anisocoria with signs of Cushing triad. An urgent decompressive craniectomy was performed as a lifesaving procedure. Serologically evidence of HSV 1 was obtained and temporal lobe biopsy was of lymphocytic meningoencephiltis. The child survived however at 1 year follow up she had severe cognitive disabilities and was bed bound.
Conclusion: Surgical intervention in these cases can be lifesaving, however long-term disabilities relating to the underlying encephalitis affect significantly the outcome.

PP024

Cranioplasty with a polymethylmethacrylate customized cranial implant in pediatric age: a single-center experience

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Introduction: Cranioplasty is a surgical procedure used to repair skull defects. Optimal strategy of reconstruction used to achieve the best result remains a challenge, especially in pediatric patients in whom there is a continuing skull growth that makes the choice of material more difficult.
The native bone flap is universally accepted as the preferred option in pediatric patients; In case of unavilability the authors' choice of prosthetic material is a customized polymethylmethacrylate (PMMA) implant.
Methods: We present the results of our clinical series of 12 custom-made PMMA implants in pediatric patients who had undergone cranioplasty at our center between 2006 and 2013. All patients or parents were administered a questionnaire to assess the aesthetic result.
Results: The age at craniectomy was from 5 months to 12.5 years and the mean age at cranioplasty was 84.33 months. The mean extension of the plastic was 56.83 cm2 and the mean time from craniectomy to cranioplasty was 9.25 months. The mean follow-up duration was 55.7 months. We did not observe any major complications; only 3 patients had minor and moderate complications (prosthesis dislocation, granuloma formation, fluid collection).
Conclusions: PMMA resulted in a low complication rate and the custom-made technique was associated with an excellent grade of patient and parent satisfaction on long-term follow up.

Poster track 1.5: Hydrocephalus I

PP025

Chronic overdrainage syndrome: pathophysiological insights based on ICP analysis. A case-based review

Laura Sainz Villalba, Konstantin Hockel, Martin U. Schuhmann (Madrid, Spain; Tübingen, Germany)

Introduction: Chronic overdrainage affects shunted patients producing a variety of symptoms that may be misdiagnosed. Early described as a triad of intermittent type headache, small "slit" ventricles and position related symptoms, a new concept has evolved beyond ventricular catheter obstruction. A cascade of pathophysiological steps regarding CSF dynamics and venous dynamics occurs with an overdraining shunt system. There is mounting evidence that changes in this cerebrospinal venous system are a key factor to the development of chronic overdrainage syndrome (COS).
Methods: We review the possible pathophysiological changes introduce by overdrainage to the CSF and venous compartment. On the basis of ICP recordings from a29 year old woman with a shunt since the postnatal period suffering from most severe intermittent headaches, despite unchanged ventricular size and an open shunt we explain thepathophysiology of COS.
Results: Massively increased ICPs and a continuous B wave "storm" during the headache attacks were recorded with compromised intracranial compliance despite an open shunt. When mobilized to upright position, her ICPs were dropping to - 17 mmHg. By treating the patient with an adjustable gravity unit the symptomatology gradually improved and finally disappeared.
Conclusion: Symptomatology can only be explained by sudden venous occlusion. Abnormal venous distensibility follows as a result of chronic overdrainage and prones to collapse with less transmural pressure gradients. The collapsed venous system will increase ICP, collapsing veins even further leading to compensatory reserve exhaustion and impaired autoregulatory phenomena.

PP026

Symptoms, signs and clinico-radiological patterns of presentation in shunt overdrainage syndrome. Analysis of a series of 100 patients

Bienvenido Ros Lopez, Sara Iglesias Moroño, Miguel Segura Fernandez-Nogueras, Antonio Selfa Rodriguez, Miguel Angel Arraez Sanchez (Malaga, Spain)

Introduction: Important problems still exist in shunt overdrainage syndrome (SOS) concerning the definition of the entity and its classification. The purpose of this paper is to describe our experience about the clinico-radiological patterns of presentation in SOS.
Methods: Between 2003 and 2015, 100 patients with a diagnosis of SOS have been included. The inclusion criteria consisted of the appearance of any symptoms/signs that improved with measures to reduce the amount of drainage through the shunt. Following data were recorded: gender, perinatal history, age at first shunting, etiology, type of shunt, age at diagnosis, symptoms/signs at presentation, radiological findings and valve refilling characteristics.
Results: The series consists of 56 male and 44 female patients. Mean age at first shunting was 17.90 months. The most

frequent etiology was posthemorraghic (33 cases) and most patients (61 cases) were first implanted with fixed pressure valves. The mean age at diagnosis of SOS was 73.98 months. 67 patients debuted with headache as the main symptom. 31 patients debuted with atypical pictures, such as hypotonia-irritability (20 patients), seizures (5 patients), "dorsal midbrain syndrome" (1 patient), "pseudotumor cerebri-like" picture (2 patients), late deterioration in spina bifida (2 patients), orthostatic intolerance syndrome (1 patient). 2 patients were included in a pre-symptomatic state with no symptoms but marked cranial dysmorphia. According to the characteristics of the symptoms and signs, radiology and pump refilling, we grouped the patients in different patterns: pre-symptomatic (PRES) pattern (2 cases), pattern A (18 cases) with orthostatic symptoms, pattern B (41 cases) with intermittent symptoms, pattern C (27 cases) with chronic symptoms, pattern D (5 cases) with "pseudotumor cerebri like" picture,pattern I (6 cases) with ventricular isolation and pattern F (1 case) corresponding to repeated proximal shunt failure.
Conclusions: The identification of clinico-radiological patterns in SOS may help in the early diagnosis of this condition, especially in cases of atypical presentation.

PP027
Comparison of Third Ventricle Diameter (TVD) using transtemporal UltraSound (US) measurement and Magnetic Resonance Imaging (MRI) in Pediatric Neurosurgery
Susanne R. Kerscher, Martin U. Schuhmann (Tübingen, Germany)

Introduction: We previously proved that changes of TVD in MRI scans are a reliable mirror of changes of the lateral ventricles. Since the third ventricle is easily accessible in more than 90% of children and adults using ultrasound via the transtemporal bone window, US assessment of TVD would be most helpful in clinical routine care of patients with hydrocephalus. This study aims to investigate whether there is a good correlation between US and MRI based TVD and if both methods measure the same value.
Methods: This prospective study includes 52 patients aged newborn to 18 years. Diagnoses encompassed hydrocephalus (50%), tumor (13.5%) and various other pathologies (36.5%). TVD was measured by transtemporal window using a phased-array 1-4 MHz transducer with patient in supine position. Mean TVD was created out of 3 measurements. This was compared to TVD on T1 weighted axial MRI of the brain which were simultaneously (usually within 24 hours) aquired.
Results: Overall mean values for TVD were 6.44±5.27 and 6.29±4.94 mm for ultrasound and MRI, respectively. There was a strong positive correlation between TVD measured on MRI compared to ultrasound ($r=0.982$, $p<0.01$). Bland and Altman analysis of the two methods showed a mean bias of 0.15 mm with limits of agreement of -1.37 and 1.67 mm. 98% of the values were within the limits of agreement.
Conclusion: US and MRI based measurements of TVD are well comparable, but US TVD is in mean about 0.15 mm larger than MRI TVD, most likely due to a more angulated assessment. When considering this offset, ultrasound TVD can be used for follow-up evaluations of ventricular width in neurosurgery instead of MRI scans.

PP028
Predictors of QoL in pediatric hydrocephalus using HOQ – Spanish version
Sara Iglesias, Bienvenido Ros, Miguel Segura, Andrea Delgado, Francisca Rius, Miguel Angel Arraez (Malaga, Spain)

Introduction: The aim of the study was to assess whether the HOQ-Spanish version could be useful to analyse predictors of functional outcome of the shunt.
Methods: Parents of paediatric patients with hydrocephalus attending our neurosurgery outpatient clinic between March 2015 and April 2016 were invited to enrol in the study and complete the HOQ-Spanish version. Age at diagnosis and at the time of the study, clinical data, shunt complications and socioeconomical factors were recorded. Independent variables related to the HOQ scores were studied.
Results: A total of 132 patients participated in the study. The mean overall HOQ score was 0.68 (on a scale from 0 (worse) to 1.0 (best)). Factors related to a worse quality of life were: seizures, motor or visual impairment, shunt infection, need for shunt revision, number of shunt revisions, symptomatic overdrainage and older age at the time of the study.
Conclusion: The HOQ-Spanish version is useful to achieve factors related to QoL in pediatric hydrocephalus. Some of them, as shunt complications, could be influenced by our practice.

PP029
Ventriculo-sinustransversostomy as an alternative treatment for hydrocephalus in children
Mikhail Nikolaenko, Konstantin Samochernich, William Khachatryan (St. Petersburg, Russia)

Introduction: CSF-shunting is still one of the main methods of treatment shunto-dependent form of hydrocephalus in children. Classical methods of liquor-shunting operations are ventriculo-peritoneostomy, less often ventriculo-atriostomy. In cases where contraindications to the implementation of these interventions are identified, ventriculo-sinus operations may be an alternative treatment for hydrocephalus.
Methods and Results: We, in the period from 2015 to 2017, carried out and improved the methodology of conducting 38 ventriculo-sinustransversostomy. According to the results,

which established that the development of hyperdrainage states was not observed, but in 13 cases, hypodrainage states were registered, which led to the need for shunt revisions. When analyzing the data it was found that the hypodrainage state developed against a background of high pressure in the transverse sinus of the dura mater, which prevents the correct operation of the shunting system. Taking into account that in the system "upper sagittal sinus - jugular vein - right atrium" there is a decrease in venous pressure, and when positioning the venous catheter of the shunting system into the right atrium, a negative pressure is recorded at the time of diastole.
During the operation, after puncture of the transverse sinus and implantation of the venous catheter, the pressure in different sections of the sinus is measured, in the case of recording a venous pressure less than 60 mm of water column, the operation is terminated with the position of the catheter in the transverse sinus. But when the venous pressure is recorded more than 60 mm of water column, the catheter is displaced along the blood stream towards the right atrium with a stage-by-stage monitoring of venous pressure in different parts of the venous system, when the venous pressure is recorded less than 60 mm of water column, the catheter is left in this position, which is optimal for correct operation of the liquor-shunting operation and allows to achieve control over hydrocephalus.
Conclusion: Ventriculo-sinustransversostomy can be an alternative method of treatment of hydrocephalus in children in those cases when classical operations are not possible. This kind of CSF-shuntingcan become universal, since hyperdrainage conditions are not recorded on our material, and the positioning of the intermediate and distal part of the shunt into the venous channel not only repeats the physiological mechanism of liquor resorption, but also prevents the development of complications of the CSF-shunting, which improves the parameters of hydrocephalus treatment in children.

PP030

Comparison of quality metrics for pediatric shunt surgery

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Objective: Despite an increasing interest in objective quality metrics for medical care, no common outcome measures exist for shunt surgery. In the present study we calculated published metrics in a single data set and compared results of a small pediatric neurosurgery unit to published benchmarks.
Methods: A retrospective analysis of shunt surgery at our unit in 2015 was performed. Demographic, clinical and radiological variables were extracted from medical notes. Surgical Activity Rate (SAR), Revision Quotient (RQ), 30-Day Shunt Malfunction Rate, 90-Day Global Shunt Revision Rate and Preventable Shunt Revision Rate (PSRR) were calculated.
Results: Sixty shunt operations in 39 children were performed at our institution in 2015, comprising 18 (39%) new shunt insertions and 42 (70%) revisions. The median age of this cohort was 18 months (range 0.03-204 months). The etiologies were post-hemorrhagic (n=16, 41%), congenital (n=11, 28%), tumor-associated (n=8, 21%) hydrocephalus, as well as other causes in 4 patients (10%). The majority of cases were ventriculoperitoneal shunts (n=55, 92%) inserted through an occipital burr hole (n=41, 68%). Thirteen shunt failures occurred within 90 days after index surgery, predominantly due to proximal failure (n=6, 46%). The SAR was 0.127, the RQ was 2.333, the 30-Day Shunt Malfunction Rate was 0.166, the 90-Day Global Shunt Revision Rate was 21.7% and the PSRR was 38.5%.
Conclusions: Quality metrics obtained for our dataset correlate with published values and, analyzed in conjunction, offer measurement of quality that can be compared across studies using common metrics and can be considered patient-oriented, easily measurable and potentially modifiable. When judging such results it has to be considered that shunt revisions are not exclusively linked to suboptimal medical care, but partially intrinsic to the disease and procedure and thus to a certain degree beyond a neurosurgeon's control.

Poster track 1.6: Arachnoid cysts

PP031

Management and outcomes of antenatally diagnosed intracranial arachnoid cysts: a single UK center experience

Samuel Hall, Alex Smedley, Nijaguna Mathad, Ryan Waters, Aabir Chakraborty, Vassilios Tsitouras (Southampton, United Kingdom)

Introduction: Arachnoid cysts are common developmental abnormalities encountered in pediatric neurosurgery. However, only a small minority are diagnosed intra-uterine. The aim of this study is to evaluate the management and outcomes of antenatally diagnosed arachnoid cysts in our institution.
Methods: A retrospective case note review was conducted of all patients with an intracranial arachnoid cyst who presented between 2005-2016 at a single neurosurgical centre in the United Kingdom. All patients with an arachnoid cyst diagnosed on antenatal imaging were eligible for inclusion.
Results: Nine patients (5 male, 4 female) were identified and regarded eligible for inclusion out of 739 arachnoid cysts diagnosed by our centre over this time period. The locations of the cysts were: posterior fossa n=3, quadrigeminal cistern n=2 and one each of convexity, interhemispheric, intraventricular and suprasellar. Six patients were surgically treated (5 with endoscopic fenestrations and 1 with cysto-peritoneal shunt) which occurred on an average of 81 days after birth. Associated hydrocephalus was found in 2 patients (22%) who were managed with endoscopic third ventriculostomy at the time of fenestration and both later required CSF

shunting. Two patients, including one of the patients with hydrocephalus, required re-fenestration for enlarging cyst volume. Five of the surgical patients and all three conservatively managed patients, had normal development. The remaining patient had spina bifida and associated developmental delay. The average clinical follow-up was 35 months.
Conclusion: Antenatal diagnosis remains an uncommon presentation of arachnoid cysts despite the widespread application of antenatal ultrasonographic screening. Once diagnosed, a higher proportion will require surgery than cysts diagnosed later in life. Neuroendoscopic management is a safe and effective option as a first line treatment choice. Reasonably early intervention is also safe and the overall neurodevelopmental outcome is favourable.

PP032

A single center's experience of the operative management strategies of intracranial arachnoid cysts

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Introduction: Considerable debate continues surrounding the various operative management strategies for arachnoid cysts. We report our institutional experiences on the operative management and post-operative outcomes for intracranial arachnoid cysts.
Methods: This was a single centre, retrospective review of all newly operated intra-cranial arachnoid cyst between 2012-2016. Patients were categorised according to treatment into direct (craniotomy + cyst fenestration/excision, neuroendoscopic fenestration and CP shunt insertion) and indirect (VP shunt insertion, ETV) groups. Case note review was carried out to examine for subsequent interventions and clinical follow up.
Results: 23 newly operated cases were identified under the study time period. 65% were male (n=15) and 35% were female (n=8). Mean age at first neurosurgical intervention was 3.8 years (range 0.01-15.9). Mean length of follow up was 3.6 years (range 0.8-5.4 years). The arachnoid cysts were topographically classified into; retrocerebellar (26%, n=6), quadrageminal (22%, n=5), sylvian fissure (13%, n=3) intraventricular (13%, n=3), pre-pontine (9%, n=2), suprasellar (9%, n=2), interhemispheric (4%, n=1) and hemispheric (4%, n=1).
19 patients underwent direct treatments; 9 endoscopic fenestration, 8 craniotomy and 2 CP-shunt. 5 of the endoscopic fenestration patients did not require further intervention, 2 underwent subsequent open fenestration and 2 VP-shunts. 4 patients who underwent craniotomy did not require subsequent intervention, 4 patients went on to require VP-shunt (n=2), CP-shunt (n=1) and subdural peritoneal shunt (n=1) insertion. Both patients treated via CP-shunt required further intervention.
4 patients underwent indirect treatments; 3 ETV and 1 VP-shunt. All patients who underwent indirect treatment required further intervention.
Conclusions: Our results highlight more favourable outcomes for direct treatment utilising endoscopy or craniotomy. All patient undergoing indirect treatment or direct, cysto-peritoneal shunting required subsequent surgical intervention. Given the limitations in our small sample size, these results should primarily be interpreted as a further contribution to the ongoing debate surrounding the complex management of these patients.

PP033

Successful endoscopic fenestration of arachnoid cysts in symptomatic children: how does the cyst volume change?

Marina Pitsika, Spyros Sgouros (Newcastle Upon Tyne, United Kingdom; Athens, Greece)

Introduction: Endoscopic fenestration remains a first line treatment option for symptomatic arachnoid cysts. After fenestration the cyst does not collapse, but seems to reach an equilibrium. The aim of this study was to evaluate the change in the cyst volume following successful fenestration.
Methods: DICOM data analysis was performed, using semi-automatic segmentation (seed growing technique, manual correction) for serial measurements of the cyst volumes on 4 children (1 female, 3 males) that had symptomatic arachnoid cysts (middle fossa n=2, choroidal fissure n=1, posterior fossa n=1), were treated with endoscopic fenestration between 2009 and 2014 and experienced symptom resolution post operatively. Average follow up was 20.5 months (range 3-48).
Results: Significant cyst volume reduction was seen in all four patients. Patient 1 cyst volume pre-operatively was 336 cm^3 and decreased by approximately 40cm^3 per month for the first three months (to 70% of the original size), while it remained stable after that. In patient 2 original cyst volume was 12.64 cm^3 and reduced by 11.3 cm^3 in 3 months. In patient 3 pre-operative cyst volume was 104.68 cm^3 and reduced with a rate of 16cm^3 per month for two months (70% of original size) and remained stable after that and similarly patient 4 from a pre-operative volume of 124.81 cm^3 reduced by approximately 11cm^3 per months for the first seven months (to 43% of the original size) reaching a plateau after that. There has been no late increase in volume.
Conclusions: Significant reduction in arachnoid cyst size is seen in patients who responded clinically to endoscopic fenestration. The cyst volume seems to decrease gradually in the first 3 months and reaches a plateau by 7 months. Complete resolution of symptoms in the presence of residual volume may indicate that cyst size below a threshold may not directly correlate with clinical status.

PP034

Management of intracranial arachnoid cysts – Clinical and radiological outcome

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Introduction: Management of intracranial arachnoid cysts (AC) remains topic of debate and different treatment approaches exist. Principal options are either establishing a permanent communication with intracranial CSF compartments by endoscopic or microsurgical techniques or extracranial diversion of the cyst content by a shunt.
Methods: A retrospective survey of all patient files identified all patients who were operatively treated at the Division of Paediatric Neurosurgery at Charité Universitätsmedizin Berlin from 2007 to 2017. In order to evaluate efficacy of performed treatment 1) all available pre- and postoperative radiological image sets were reviewed to assess volume changes, 2) development of preoperative symptoms after the operation was assessed and 3) complications and reoperation rates are reported.
Results: During the observation period 94 children with intracranial AC of different locations were operatively treated. There were 35 patients with Sylvian fissure arachnoid cysts undergoing 39 operations, 15 patients with suprasellar AC and 18 operative procedures, 14 patients with intra/paraventricular AC and 15 operative procedures, 14 patients with posterior fossa AC and 18 operative procedures, 6 patients with cysts of midline septum or quadrigeminal cistern and 6 procedures, 5 patients with interhemispheric AC and 5 procedures, 3 patients with temporo-mesial cysts and 4 procedures and 2 patients with convexity AC undergoing 2 operative procedures. Mainly endoscopic techniques – occasionally augmented with stent implantation – were used to achieve communication with regular intracranial CSF circulation, followed by microsurgical techniques and rarely extracranial CSF diversion via a shunt. For most AC locations, a significant decrease of cyst volume after the respective operative procedure could be demonstrated. Associated clinical symptoms are reported and were dependent on cyst location and generally improved after surgical treatment. Few patients needed re-operation due to either insufficiently decreased AC size, persisting symptoms or surgical complications. This was mostly dependent on cyst location as well.
Conclusion: The presented series of intracranial AC proves the efficacy of establishing a communication between the AC and regular CSF circulation to achieve clinical and radiological improvement. CSF diversion to an extracranial compartment was only exceptionally necessary.

PP035

A diagnostic conundrum: temporal arachnoid cyst and limbic encephalitis

Zarela Krause Molle, Hans-Jakob Steiger, Michael Karenfort, Thomas Beez (Düsseldorf, Germany)

Objective: Arachnoid cysts (AC) have been associated with seizure activity as well as behavioural and psychiatric symptoms. However, in case of atypical presentation the diagnostic challenge of identifying the AC as causative pathology or as mere bystander can arise.
Methods: We present a case with two concomitant intracranial pathologies and highlight the diagnostic challenges and therapeutic considerations.
Results: A 15-year-old boy presented with dizziness, diffuse dysesthesia and loss of the sense of reality. MRI revealed a large right temporal AC with midline shift and significant compression of the right cerebral hemisphere, classified as Galassi type III. Electroencephalography (EEG) did not demonstrate epileptiform activity. Microsurgical fenestration was performed without complications. After initial improvement, the patient presented with episodes of acute anxiety, disorientation, memory loss and gustatory and olfactory sensations. EEG now showed scattered sharp waves. Treatment with levetiracetam was commenced. MRI revealed decrease in arachnoid cyst size and improvement in right hemispheric compression. However, fluid attenuated inversion recovery (FLAIR) sequences now demonstrated increased signal intensity of right hippocampus. In retrospect, subtle limbic signal alteration could be detected also on preoperative MRI, although findings were masked by severe compression due to the untreated AC. Limbic encephalitis was confirmed by pathological glutamate decarboxylase (GAD) auto-antibody serum titre of 1:40,000.
Conclusions: This is a case of an atypical clinical presentation initially attributed to a Galassi type III AC. As symptoms persisted despite successful fenestration, further diagnostic workup revealed a concomitant rare diagnosis of limbic encephalitis. In retrospect, the patient's symptoms appear to be related to autoimmune disease rather than to the large AC. This diagnostic conundrum highlights the complexity of differentiating causal and incidental findings in patients presenting with atypical clinical features.

PP036

Neuroendoscopy in children – 13-year experience

Frantisek Horn, Dana Kuniakova, Martin Smrek, Michal Petrík, Martin Lindák, Dominika Stevkova, Miroslava Sarkanyova (Bratislava, Slovak Republic)

Introduction: Authors present their experiences with neuroendoscopy from the first operation in 2004 in children with hydrocephalus since 2017. Retrospective study is focused on the diagnosis of hydrocephalus and the age at the time of surgery. Long term follow of the patients is routine in their conditions.
Method: Flexiendoscope was used in 127 children and 144 procedures. Approaches: Endoscopic third ventriculostomy (ETV), aqueductoplasty (AP), cystostomy (CP), septetstomy (SP) and chorioid plexus coagulation (CPC). According to the age, patients were diveded to two groups: first - A -from newborn til 6month and second - B - from 0,5 -19 years. Type of

hydrocephalus (H): Congenital, Posthemorrhagic (PHH), Postinfectious (PI), Meningomyelocele (MMC), Tumor (T) and others.

Results: In group A 33% was success rate within the first year after the procedure, in older patients, group B, the success rate was 66,7%. Neuroendoscopy, as the most successful procedure was in children with congenital H. Most failure type was in children withPHH. 10 newborns were treated with neuroendoscopy and one is still VPS free. In other nine paitents we used combined procedure neuroendoscopy and after 3,5 months (1-9) we need to put the VPS. In 4 patients with MMC the procedure was successful only in 3 of them. ETV was the most frequent procedure, then AP, CP, SP. CPC and ETV we used only in one patient with Dandy type malformation, after three months we have to put VPS.

Discussion: Treatment with CSF shunting or with neuroendoscopy does not appear to be associated of any substantial difference in quality of life outcome. In our previous study, Dubravova, 2014, in which we compared the results of neuroendoscopy versus VPS, the overall complication rate for endoscopic interventions was significantly lower compared to shunts (p=0.017). Bowes at al., 2017 published, that intraventricular endoscopy is a safe procedure in pediatric patients of all ages, although it might be associated with increased shunt rates after endoscopic surgery, especially ETV, in young infants. Our limited experiences are the same.

Conclusions: It looks, that neuroendoscopy it is still acceptable procedure in children with hydrocephalus and with the combination of other techniques (VPS) it gives the possibility for normal life in children with hydrocephalus, also in younger babies in the first 6 months of life.

Poster track 2.1: Epilepsy & Functional diagnostics

PP037

The Mozart effect in children with epilepsy: a review of the literature

Vaitsa Giannouli, Nikolaos Syrmos (Thessaloniki, Greece)

Introduction: Mozart's sonata for two pianos in D major (K448) has been widely used in research regarding neuropsychological assessment, and it has been shown to decrease interictal EEG discharges and recurrence of clinical seizures not only in adults, but also in young patients. The objective of this study is to examine if Mozart's music has an effect on brain activity in epileptic infants/children.

Methods: A literature search of the electronic databases PubMed-MEDLINE, EMBASE and Google Scholar was performed to identify studies published before September 2017. Five original studies presenting raw experimental and clinical data were included.

Results: All studies indicated that recurrence of first unprovoked seizure, seizure frequencies, and epileptiform discharges were significantly reduced after listening to Mozart's K.448 in children with epilepsy. Thus, there is preliminary evidence that listening to Mozart's music, short-term and long-term, may contribute to decreases in epileptiform discharge and seizure frequency in children with epilepsy, even in refractory type, a finding that can be incorporated in clinical practice.

Conclusions: Although further analyses are required, the results support a beneficial influence of a specific excerpt of Mozart's music in the heterogeneous group of pediatric epilepsies. Future research should further investigate if the cognitive and neuropsychological impairments of pediatric epilepsy of varying types can also be influenced by exposure to music.

PP038

Predictors of the surgical treatment of temporal epilepsy in children and their importance in the personalization of surgery

Konstantin Abramov, Roza Khachatryan, Magomed Mamatkhanov, Konstantin Lebedev, William Khachatryan (St. Petersburg, Russia)

Purpose of the Study: Study of predictors affecting the outcome of surgical treatment of children with symptomatic temporal epilepsy.

Materials and Methods: In 80 patients aged 2-17 years with symptomatic temporal drug-resistant epilepsy, operated in the Russian neurosurgery institute after A.L. Polenov from 2011 to 2016, were studied the factors affecting the outcome of surgical treatment in the near and distant periods.

Results: In this population, the children were dominated by boys (51.25%). More than 50% of patients were 8-13 years old, the average age was $M = 10.5 \pm 0.45$ years (median = 11 years). The manifestation of the disease in the majority (55%) of the patients was before the age of 1 year (median = 3 years), the average age was $M = 4.54 \pm 0.46$ years, the most frequent age of onset was 1 year (mode = 1 year). Structural changes in the group of children with drug-resistant temporal epilepsy have been established in the overwhelming majority (93.8%) of cases. In particular, neoplastic lesions of different histocellular structure, hippocampal sclerosis in 15%, cortical malformation of 11.25%, vascular malformations in 5%, arachnoid cysts and brain atrophy in 26.25% are established in 22.5%. Seizure outcome was extremely satisfactory in 94% of patients, including the termination of seizures (like Engel I) in the nearest postoperative period (up to 12 months) in 72.5% of cases, and in the distant (from 1 to 5 years years) in 54.6% of cases. Favorable prognostic factors of the effectiveness of surgical treatment in the long-term period were: male sex ($p < 0.01$), age at the onset of the disease more than 7 years ($p < 0.01$), duration of the disease less than 3 years ($p < 0.05$) less than 3 anticonvulsants ($p < 0.01$), no history of febrile seizures, epileptic reactions ($p < 0.05$), absence of comorbid conditions ($p < 0.05$), serial seizures and statuses ($p < 0.01$), absence of early postoperative seizures ($p < 0.01$). Left-sided resections (p

<0.05), carrying out of lesionectomy (p <0,05) were more effective, removal of neoplastic lesions, vascular malformations, cortical tubers (epileptogenic foci) (p <0,01). The treatment of temporal epilepsy in the long-term period with the consequences of neuroinfection, against cicatrical and gliotic and atrophic brain changes, with arachnoid cysts (p <0.01), and also in the detection of epileptic leukoencephalopathy (p <0.05) proved unfavorable in the prognostic plan. Subpial multifocal resections (p <0.05) were less effective for the treatment of temporal epilepsy.

Conclusion: The rational tactics of treatment of temporal drug-resistant epilepsy in children should take into account the child's developmental features, the presence and nature of intercurrent pathology, clinical examination and introscopy data, and also take into account the alleged etiologic factor, the features of the pathological process and the prognosis of the proposed operation.

PP039

Pediatric refractory epilepsy with multiple focal cortical dysplasia lesions close to eloquent area: a case illustration of epilepsy surgery

Emily Kit-Ying Chan, Ho Wan Leung, Lai Wah Fung, Wai Sang Poon, Claire Ka-Yee Lau, Xian Lun Zhu, Aubery Ka-Pui Li (Hong Kong)

Introduction: Focal Cortical Dysplasia (FCD) is a disorder of neuronal migration with its prevalence in epilepsy ganging 5-25%. Multiple FCD related refractory epilepsy could be common especially in the paediatric population. The surgical planning is challenging in terms of localization of culprit lesion and function preservation in surgery involving eloquent areas by means of function mapping. We would like to describe a case of refractory epilepsy with multiple FCD lesions and the surgical treatment process.

Case Report: Yu is a 9-year-old boy. He was born full term with an unremarkable perinatal course. He remained well with normal development and good academic results until his first tonic-clonic seizure episode at 7 years old. Over 2 years, he has progressed to suffer from daily seizures up to 6 times per day despite multiple medications. He also experienced increasing difficulty to manage schoolwork.

He was normal in neurological examination. MRI brain T2/FLAIR and Double Inversion Recovery sequences showed hyper-intense lesions in the gray-white junction of bilateral frontal, right parietal lobe and right posterior parietal lobe suggestive multiple FCD [Figure 1]. Video EEG of habitual seizure showed onset from the right parietal lobe lesions in close proximity to the primary motor-sensory cortex [Figure 2]. Functional MRI (motor task) and DTI demonstrated the motor and sensory fibre tracks just escaped from these two FCD lesions [Figure 3].

Intracranial depth electrodes targeting at the two FCD lesions and a subdural grid were implanted. Extra-operative mapping and video EEG confirmed that clinical seizure onset is from the parietal lesion and subclinical seizure onset from the frontal lesion. Motor and sensory function in close proximity to the lesion were mapped out [figure 4]. Both lesions were then excised in a 2nd stage operation under general anesthesia with MEP/SSEP monitoring and neuro-navigation. Intra-operative ECoG demonstrated significant reduction of epileptic spikes after excision. Post-operatively, there was no neurological deficits and he remained seizure free so far [Figure 5, post-op image].

Conclusion: Dedicated structural and functional MRI, intracranial electrode for extra-operative VEEG plus function mapping, lesion excision under GA with intra-operative physiological monitoring (IOM) contributed to the successful epilepsy surgery.

PP040

Selective dorsal rhizotomy in hemi-paretic/-plegic children with spasticity: case report and review of the literature

Michal Petrík, Dana Kuniaková, Martin Smrek, František Horn (Bratislava, Slovak Republic)

Introduction: Disturbances in upper motor neuron system lead to spasticity, which is main cause of impaired motor function in children with spastic cerebral palsy. Selective dorsal rhizotomy is well-developed surgical method, which originated in 1980s with introduced and well-reviewed electrophysiological intraoperative criteria. Modified Ashworth scale is widely used to evaluate patients preoperatively and after selective dorsal rhizotomy. We report on one case of unilateral selective dorsal rhizotomy in child with spastic hemiparesis. Literature has been reviewed for cerebral palsy patients diagnosed with hemiplegia or hemiparesis who underwent selective dorsal rhizotomy procedure unilaterally or bilaterally in both pediatric and adult population.

Methods: Selective dorsal rhizotomy was done at 7 years of age on the child with right-sided spastic hemiparesis. Intraoperative neuromonitoring was used during surgery. Baclofen test was performed before the surgery and relaxation under general anesthesia was used to evaluate contractures and range of movement on affected side. We used modified Ashworth scale to evaluate muscle tone. Video was documented throughout the course of diagnostic and therapeutic process.

Results: Decreased tone was measured with modified Ashworth scale after the surgery. Quality of gait was assessed with visual gait analysis scale. Gait parameters were compared before and after surgery. Beneficial effects on both spasticity and gait led to the gain in functional status and quality of life of the child.

Conclusions: Selective dorsal rhizotomy can be safely used as a method in selected cases of hemiparesis/hemiplegia that are not responding to medication treatment. Precise diagnostic evaluation and selection of cases is recommended. In surgical therapy we recommend to approach selectively on affected side as both patient numbers in literature and knowledge on precise pathophysiological mechanisms are limited.

PP041

Survival of telemetric intracranial pressure sensors

Nicolas Nørager, Alexander Lilja-Cyron, Sara Duus, Marianne Juhler (Copenhagen, Denmark)

Introduction: The monitoring of intracranial pressure (ICP) is a cornerstone diagnostic tool in pediatric neurosurgery. Relatively recently, telemetric ICP monitoring devices, Raumedic Neurovent-P-tel (NPT), have become available, allowing for sampling of ICP curves without repeated surgical insertions of a cabled ICP sensor, thereby lowering the treatment risks considerably. The Raumedic Neurovent-P-tel is clinically approved for 90 days, but previous studies have detected seemingly useful ICP beyond this period. The objective of our study was to investigate the survival time and clinical applicability of the Raumedic Neurovent-P-tel.
Method: We retrospectively identified all patients in our clinic that had undergone implantation of NPT. Information regarding implantation date, explantation date and date of measurement error was collected. The Kaplan-Meier method was then used to analyze survival. Explanted NPT was tested for zero drift.
Results: The Kaplan Meier analysis was based on 106 included NPT. Of these, 60 became non-functioning during the observation period either due to inability to find signal (20), misleadingly low ICP (13) or explantation (27). In total, 46 sensors were censored. A median survival time of 224 days (95% confidence interval, 145 to 303) was found. 85% of implanted NPT had a survival time beyond the clinically approved 90 days. Nine NPTs were explanted before the clinically approved implantation period of 90 days due to defect probe (4), skin damage or infection (4) or unknown reason (1). Median zero drift was 3 mmHg in explanted sensors.
Conclusion: Many NPTs were useful far beyond the present standard for clinical applicability. Our main worry is mistreatment on the basis of wrong ICP curve. In this regard, we found the main problem to be low ICP values. Therefore, it seems that at least findings of high pressure can still be used beyond 90 days.
Funding: Lundbeck Fonden

PP042

Neuromonitoring for Pediatric Surgery

Miroslava Sárkányová, Michal Petrík, František Horn (Bratislava, Slovak Republic)

Introduction: Neurophysiologic monitoring is a name for set of methods that are used for evaluation of functional state of neural systems in real-time. The using of neuromonitoring brings many benefits within diagnostics, when even the smallest changes can be captured in the neurological image. The neurophysiologic methods are very beneficial during many operations because the risk of post-operational neurological deficits can be significantly reduced. With their help, the occurrence of iatrogenic complications during the operations is minimized. The neuromonitoring methods are useful for determination of prognosis and tracking of treatment results too. The goal of our research is to analyze, extend and improve efficiency of the usage of neurophysiologic monitoring in our clinic of pediatric surgery in Bratislava.
Methods: The neuromonitoring methods that are used in our clinic are electromyography and evoked potentials. Within the evoked potentials, we are using methods for monitoring of motoric evoked potentials and somatosensoric evoked potentials. These methods are actively used in our clinic since 2013.
Results: The analyzed dataset contains 23 patients that conducted surgical operations between years 2013 and 2016. The research deals with three types of operations, where the neuromonitoring methods play critical role for the operation outcomes. During operations of thyroid, the electromyography was used two times and the evoked potentials were applied eleven times. Within operations of selective dorsal rhinotomy for children suffering from spasticity, the electromyography was used nine times. For operations of two children with neural tube defects, the electromyography was used in both cases and the evoked potentials were not applicable because of too low age of the patients. None of the neuromonitoring records were completely without any artifacts.
Conclusion: Usage of neuromonitoring methods during surgical operations minimizes the risk of permanent damaging of affected nerves and thus the safety of surgical interventions is significantly increased.

Poster track 2.2: Dysraphism I

PP043

Myelomeningocele operated with electrophysiological neuromonitoring, child's case

F. Bouaré, D. Mpando, M. Laghmari, M. Lmejjati, H. Ghannane, N. Kissani, S. Ait Benali (Marrakech, Morocco)

Introduction: Myelomeningocele is a complex form of spina bifida. It is characterized by herniation of the spinal cord, raciness and meninges through a vertebral bone defect. His treatment is surgical. The aim of this work is to specify the importance of surgery and the role of peroperative electrophysiological neuromonitoring in the treatment of myelomeningocele.

Materials and Method: This is a reported case of the service.
Observation and Result: KG, 7 months old, had a well-defined lumbar lesion, skin-covered, renal on palpation, which had been evolving since birth (Fig. 1). The neurological examination found a monoparesie of the lower right limb. Lumbar MRI (Figs 2 and 3) found spina bifida with lumbar myelomeningocele. The treatment was surgical with electrophysiological neuromonitoring peroperatively. During the operation, the roots were stimulated (Fig. 4) and activity signals (Fig. 5) were detected with neuromonitoring. Parts of the myelomeningocele that gave activity signals were spared and re-integrated. We made a tight closure of the hard mother and a closing of the skin.
Discussion and Conclusion: The surgery allows the reconstruction of the anatomy, to avoid aggravation of the clinical picture. The electrophysiological neuromonitoring helps to limit the damage of the surgery and to preserve the functional roots.

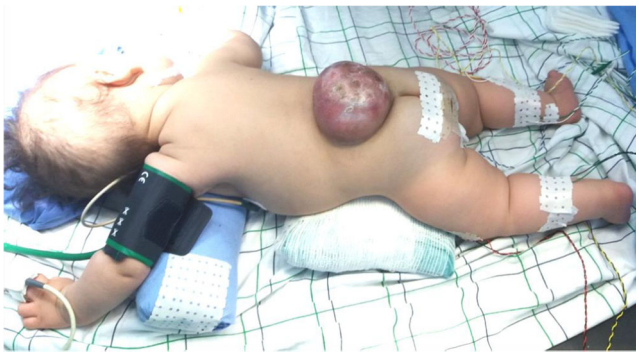

Figure 1: *Lumbar myelomeningocele*

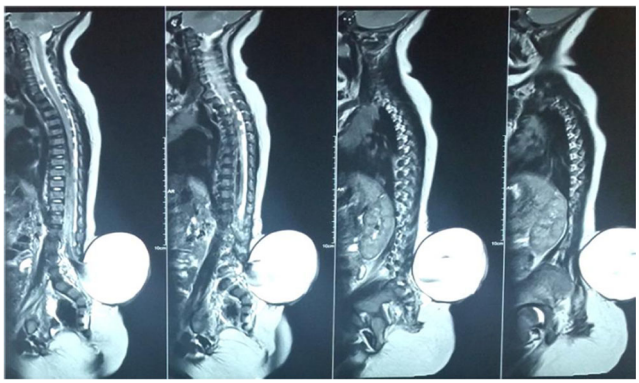

Figure 2: *Medullary MRI, sagittal section, T2 sequence showing lumbar myelomeningocele*

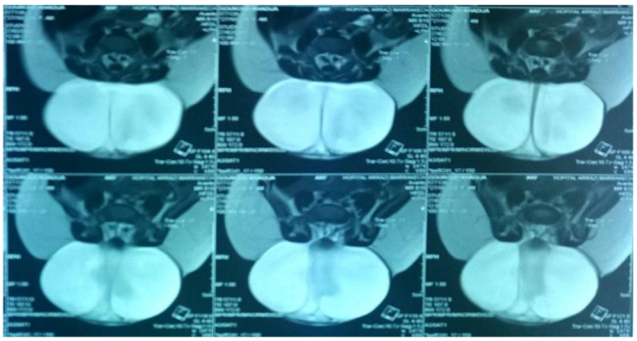

Figure 3: *Medullary MRI, axiale section, T2 sequence showing lumbar myelomeningocele*

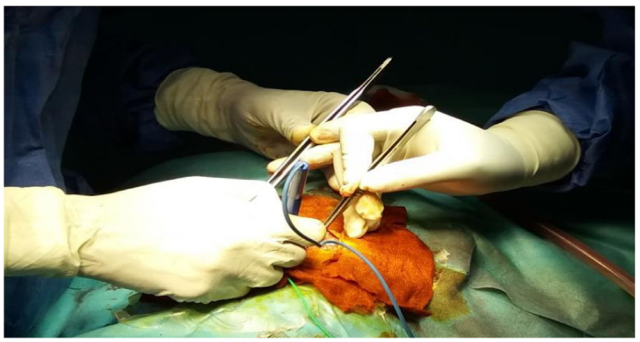

Figure 4: *Nerve roots stimulation*

Figure 5: *Detection of activity signals*

PP044
Correlation of SEP results with clinical and radiological assessments of myelomeningocele patients
Gokhan Canaz, Huseyin Canaz, Ibrahim Alatas, Zeliha Matur (Istanbul, Turkey)

Introduction: In the post-op follow-ups of patients who had undergone myelomeningocele repair, magnetic resonance imaging with the clinical evaluation is not sufficient to the

evaluation of lower extremity sensory and motor functions. Electrophysiological deteriorations of medulla spinalis in lesion levels and any progression or changes in electrophysiological basis are important to predict problems before they become obvious in clinic.

Material and Method: Posterior tibial nerve somatosensory evoked potentials of 36 patients, who had operated for myelomeningocele closure had recorded and neurological examinations were done in the same session. Fusion defect levels were Results were correlated with clinical functional lesion levels, levels of fusion defect and ambulation levels.

Results: 36 patients, aged 3-16 years old (mean:8,22) were evaluated. Posterior tibial nerve somatosensory evoked potentials revealed significant correlation with ambulation levels (r:0,471, $p<0.01$), levels of fusion defect levels (r-,366, $p<0.05$) and clinical functional lesion levels (r-,556, $p<0.01$).

Conclusion: As a non-invasive and sensitive diagnostic tool, somatosensory evoked potentials should be an indispensable method in follow-ups of spina bifida patients.

PP045

Prognostic significance in congenital heart disease accompanied with neural tube defect and Jarcho-Levin syndrome

Ibrahim Alatas, Ayhan Cevik, Berivan Sezen, Huseyin Canaz (Istanbul, Turkey)

Introduction and Aim: Jarcho-Levin syndrome is a genetic disorder with catastrophic vertebra anomaly, characterized by spondylocostal dysostosis and spondylothoracic dysostosis, which is rare associated with multiple vertebrae and rib anomalies, congenital heart diseases and neural tube defects.

Neural tube defects and congenital heart diseases most common multifactorial inheritance which play roles in pathogenesis in this syndrome. neural tube defects and congenital heart disease associated with Jarcho-Levin syndrome play an important role in the morbidity and mortality of patients.

In this study, we aimed to determine the prognostic factor by determining the percentadge and to observe of accompanying cardiac anomalies and the percentadge of non-cardiac anomalies by examining the cases with retrospective neural tube defect and Jarcho-Levin syndrome in two separate groups.

Method: The patients who were diagnosed with neural tube defect after delivery at the Medicine Faculty of Bilim University Florence Nightingale Hospital between October 2014 and September 2017 were evaluated retrospectively by forming two separate groups according to isolated neural tube defect and Jarko-Levin syndrome. Term healthy babies who consultated for cardiac murmur and electrocardiographic findings that are appropriate for their age, choosed for as a control group. Patient and control groups were evaluated for age, sex, body weight, accompanying anomalies. Standard echocardiographic evaluation procedured by the same pediatric cardiologist for all patients and healthy control group.

Result: 42 (%63.6) patients with isolated neural tube defect to study, 24(%36.3) patients with Jarko-Levin syndrome diagnosis and 30 healthy babies were included. Age, gender and accompanying anomalies were not statistically significant between the isolated neural tube defect and Jarko-Levin syndrome groups. 7(%16.6) cases of isolated neural tube defect and 1(%4.1) case of Jarko-Levin syndrome were diagnosed as congenital heart disease. 7 of the patients with isolated neural tube defect and 13 patients with Jarko-Levin Syndrome had patent foramen ovale (PFO).

Conclusion: Jarko-Levin syndrome increase in the frequency of PFO detected in diagnosed cases may have clinical importance in the management, especially patients with right-left shunt (ventriculoatrial shunt or surgical procedure).

PP046

Growing curves in spina bifida

Ibrahim Alatas, Vuslat Ozer, Berivan Sezen (Istanbul, Turkey)

Objective: The purpose of this study is to evaluate an alternative growth development curve to the age-matched growth chart which is considered as a health indicator of the child and that the World Health Organization has been recommended for healthy children by regularly checking children with spina bifida.

Methods: Our study was conducted on 295 female and 265 male patients enrolled in the Şişli Florence Nightingale Hospital's Spina Bifida Research Center. At determined control intervalsthe height, weight and head circumference of the patients were measured by the supervised and trained healthcare workers. The LMS method and the SPSS program were used for the analysis of the data and the calculation of the percentile.

Results: It has been observed weights of the children with spina bifida disease, couldn't reach WHO's data. It occured that these children had a lot of operations. For this reason, when the patients examined, instabilities were seen in the growth and development graphs. These instabilities were seen not only in the post-operative period but also because they were very influential, such as bladder and gastrointestinal system disorders, which were the consequences of spina bifida. When the children with spina bifida were examined, it was observed that the height's average in the 25-75th percentile at birth, but in the following months decreased to 50 percentile in males and 25 percentile in females. This retardation also depends on the ability of the spinal cord to vary according to the grade of defect present in the cord. Hydrocephalus, involve major differences due on head circumference measurements of children in spina bifida. When every patient examined individually it has clearly shown us that these differences depend on the operations.

Conclusion: It is shown that the percentages of height and weight compared to the data of the world health organization and the head circumference of children with spina bifida concentrate on 90 and above percentiles. It is shown that progressed hydrocephalus in children can be so misleading even in the weight measurements. In order to support the growth and development of children with spina bifida, it is necessary to interpret the deformities secondary to the disease and to obtain a meaningful result.

PP047

Spina bifida with additional anomalies

Ibrahim Alatas, Revna Cetiner, Vuslat Ozer, Berivan Sezen (Istanbul, Turkey)

Introduction: Spina Bifida is a congenital neural tube defect which is an incomplete closure in the spinal column. It is seen as an additional anomalies and defects secondary to spinal cord openness and deterioration of drainage of the cerebrospinal fluid. It is difficult to predict all the additional anomalies that may occur in this unexplained etiology. Therefore, it is important to extend the spectrum. It's known hydrocephalus, tethered spinal cord syndrome, Chiari malformation, lower extremity deformities, organ anomalies, also deformities which orthopedics and pediatric surgery (interested) involved, are frequently seen with spina bifida. In this study 553 patients were examined for additional secondary anomalies developing in this disease.
Methods: 553 patients (303 females, 250 males) between 0-19 years of age were examined in the spina bifida clinic. Secondary syndrome and deformities related to spina bifida were examined. Anomalies and pediatric surgery, orthopedic operations were recorded.
Results: Hydrocephalus was the most frequent additional anomaly seen in 60% of the patients (332 patients). 52% of patients had been follow up for tethered cord syndrome. 121 patients had lower extremity deformities and 22 of them were pes planus. There were short legs, shortness Achilles tendon, pes equinovarus and AFO use.
4 of the patients had a heart puncture, 3 had a single kidney, 3 had a horseshoe kidney, 3 had dextrocardia, 4 had a kidney localization anomaly, 1 had one arm, and 1 had atopic dermatitis. In pediatric surgery most common operations were inguinal hernias (39) and undescended testis (13). Also there were coanal atresia and intestinal perfusion disorders. 26 patients had been dislocated hip surgery and they followed-up. 11 patients had scoliosis operation. 16 patients had feet, achilles tendon or hip deformity loosening operations.
Conclusion: Spina bifida is a disease unknown etiology and frequently accompanied by additional anomalies. these defects are multisystemic and various. Frequency of association between spina bifida and additional anomalies also prior knowledge of anomalies are crucial for the management of the disease.

PP048

Jarko-Levin syndrome, spina bifida and chest deformities

Ibrahim Alatas (Istanbul, Turkey)

Introduction: Jarcho-Levin syndrome (JLS) is a genetic disorder that occurs with various anomalies in the ribs and vertebrae. It is known that spina bifida and JLS can be seen at the same time. Chest deformities are common in JLS, but in spina bifida the main problem is on vertebra. Etiologies are not certainly known in both diseases. In this study, a detailed investigation was carried out in a large group of JLS and spina bifida patients to inquire about the relationship between rib and vertebral anomalies and also these 2 diseases if they both initiate each other.
Method: 107 patients with JLS who came to the spina bifida clinic participated in the study. Many parameters such as vertebral anomalies, angulation direction and progression, chest deformities, rib anomalies, lateral bar infusion, dextrocardia were examined in patients. Rib anomaly and angulation of scoliosis searched on the patients whether the same direction. For this data, patients' chest X-ray was used. Rib anomalies are classified as decomposition, deficiency, fusion or combination of these variations. SPSS software was used for data.
Results: 110 patients participated in the research, including 69 girls and 38 boys. 7 patients' Chest X-Ray could not be evaluated. There were 73 patients with scoliosis. Pectus carinatus deformities were detected in 45 patients (3 patient's likely). Patients rib anomalies; 10 of them with fusion, 6 with deficiency, 1 with disruption, 8 with disruption and deficiency, 13 with disruption and fusion, 24 with deficiency and fusion and 29 with disruption, deficiency and fusion. Rib anomaly and angulation of scoliosis searched whether in the same direction in 29 patients. In 20 patients of were in same, 6 with opposite and 3 with both side anomalies. 3 patients had dekstrocardia, but also 2 other patients had got inappropriatechest X-ray. One of the patient probably had dextrocardia, but the other was not.
Conclusion: Chest deformities are common in JLS and vertebra anomalies common in Spina Bifida diseases. It is possible to have an idea about the origin of the developments by examining in detail the deformities of the patients who have these diseases together. It is also considerably that the normally uncommon chest deformities (pectus carinatus) are frequent in these patients. This data could contribute to the literature.

PP049

Appropriate skinfold measurements for spina bifida

Ibrahim Alatas (Istanbul, Turkey)

Introduction: Spina bifida is a neural tube defect which is seen in 1-2 out of 1000 people in the world and frequently with deformities in vertebra, thorax and locomotor system. Skinfold measurements are important, especially when evaluating pediatric patients, are also substantial in these patients. Ambulatory problems are seen in most of the patients who has

accompanying deformities. Patients use crutches and wheelchairs so often. Physical disability caused by common additional deformities, and disproportionate use of ancillary equipment disrupt the fat distribution, commonly used triceps, biceps, and subscapular areas, and thus the skinfold measurement. In this study, the suprailiac area, which is not affected by these events, was found suitable for patients with spina bifida.
Method: 493 patients between 1 month to 19 years of age were examined. Patient's vertebral anomalies were evaluated. Patient's biceps, triceps, scapular and suprailiac skinfold measurements were taken with anthropometric measurements. anthropometric measurements were evaluated in percentile curves of Turkish children. Skinfold measurements were evaluated in WHO's percentile curves. LMS method and SPSS program used on statistical study.
Result: 263 female and 230 male patients participated our research. Patient's vertebral anomalies were evaluated. Of the 263 female patients 43 with scoliosis, 20 with kyphosis, 16 with kyphoscoliosis, 3 with spondylopathy, 1 with spondylopathy and kyphosis and 1 with Klippel-Feil syndrome. Of the 230 male patients, 39 with scoliosis, 22 with kyphosis, 11 with kyphoscoliosis, and 1 with a hemivertebra and a butterfly vertebra. The skinfold results of the patients were evaluated in the percentile chart.
Conclusion: Skinfold measurements are important in monitoring growth and development of children. The fat distribution of the upper-lower body is in particular disproportionate due to the movement problems of the children with spina bifida who has deformity and using ancillary equipment. As a result, standardized biceps, triceps and subscapular skinfold measurements give false measurements in children with spina bifida. Our evaluations and our observations which we had on our patient that the result of the skinfold measurement that will give the correct result if we use the suprailiac area where unaffected of the motion and deformities. It is also necessary to establish specific measurement curves for these patients to follow and treat correctly.

Poster track 2.3: Dysraphism II

PP050
Psychiatric features of parents of children with spina bifida

Ibrahim Alatas, Filiz Izci, Revna Cetiner, Serkan Zincir, Huseyin Canaz (Istanbul, Turkey)

Objective: Spina Bifida (SB) is a closure defect of the neural tube. Affecting multiple systems of the body, this disease also affects families psychologically. In this study, our aim was to investigate levels of psychiatric symptoms, depression, anxiety, despair and coping with stress in parents of children with Spina Bifida.
Method: From the follow-up patients' families of our hospital's neurosurgery unit, a total number of 80parents were included in this study. Sociodemographic data form, The Structured Clinical Interview -Clinical Version (SCID-I / CV) for DSM-IV Axis Diagnosis, Beck Anxiety Inventory (BAI), Beck Depression Inventory (BDI), Symptom Checklist (SCL-90-R), Beck Hopelessness Scale (BHS) and Coping with Stress Scale were performed.
Results: The mean age of parents of children with Spina Bifida diagnosis was 34.44±7.00. Psychiatric symptoms and inventory scores are displayed on the table.

	Mean	Standard Deviation
Scl-90	0,86	0,63
Beck Depression Inventory	13,00	10,32
Beck Anxiety Inventory	12,93	11,71
Beck Hopelessness Scale	5,30	3,74
Coping with Stress	52,94	8,53
Pessimism	8,29	3,80
Somatization	0,92	0,77
Anxiety	0,78	0,69
Obsession	1,20	0,76
Depression	0,98	0,76
Interpersonal Sensitivity	1,16	0,80
Psychotic	0,57	0,52
Paranoid	0,95	0,69
Anger	0,97	0,85
Phobic	0,52	0,65
Other	0,96	0,77
General Symptom Index	0,89	0,61

Table 1: *Clinic Inventory Scores of Cases*

Conclusion: It was determined that psychiatric symptoms such as anxiety, depression, difficulty in coping with stress can be seen among parents of children with SB.
This suggests that parents of patients with diseases like SB should get the needed psychiatric help and supportive care during the course of treatment.

PP051
Establishing a retinoic induced MMC model in rats for investigating molecular lesion cascades during the post-neurulation fetal and perinatal development

Gesa Cohrs, Jan-Philip Sürie, Christian Vokuhl, Janka Held-Feindt, Michael Synowitz, Friederike Knerlich-Lukoschus (Kiel, Germany; Sankt Augustin, Germany)

Objective: Our previous findings regarding post-natal human mmc tissues imply that specific molecular lesioning cascades are induced by secondary damaging of the exposed placodes after initial impaired neurulation ("first/second-hit"-theory). Assessment of the relevance of such mechanisms necessitates investigations of their onset and time-course during fetal development under standardized conditions. We therefore established the retinoic-induced mmc model in rats (according

to Danzer et al 2005, Exp. Neurol.). Preliminary investigations of pro-inflammatory cytokine expression on E16, E18, and E22 were performed.

Methods: First, the optimal retinoic acid (RA, Sigma Aldrich) dose was determined by gavage-feeding time-dated Sprague-Dawley rats with 50, 60, 70 mg/kg RA at E10 (maternal n=15, newborn=163). Control animals received olive oil alone (maternal n=5, newborn n=67). Newborns were delivered by C-section at E16, E18, E22. Each newborn underwent detailed inspection. The neural axis was analyzed by H&E and immunostaining for neuroglial (Neuronal nuclei (NeuN, NF200kD, GFAP, Iba1), neural crest (Nestin, BLBP), inflammatory cell markers and Interleukin-1beta/-receptor (IL-1b/ IL-1R1), TNFalpha/TNF-Receptor (TNFa/TNF-R). Immunoreactivities were analyzed qualitatively and semiquantitatively (ImageJ 1.43u, NIH, USA).

Results: In the 50mg-RA group isolated MMC appeared in 4.3%, craniofacial anomalies in 7.2%, anencephaly in 1.4% of cases. In the 60mg-RA group MMC appeared in 97.8%, anencephaly in 20%, craniofacial anomalies in 4.4%. Caudal regression was seen in 24.4%. In the 70mg group MMC appeared in 62.1%, anencephaly in 24.1%, craniofacial abnormalities in 37.9%. Placode neuroepithelia exhibited typical cellular profiles with hints for inflammatory cell marker expression and significant astrogliosis. Preliminary analyzes confirmed induction of investigated cytokines during fetal development.

Conclusion: Application of 60mg/kg of RA on E10 reliably induced open spinal dysraphic defects with hints for specific inflammatory post-lesional cascades in respective placodes. This model enables investigations of fetal tertiary lesion cascades of open dysraphism, thus providing new-insights and targets for innovative approaches.

PP052

Use of homologous cryopreserved amniotic membrane in the repair of myelomeningocele: technical note

Elisabetta Marton, Enrico Giordan, Giorgio Gioffré, Giuseppe Canova, Adolfo Paolin, Grazia Marina Mazzucco, Pierluigi Longatti (Treviso, Italy)

Objective: Standard surgical management of myelomeningocele (MMC)aims at protecting the exposed neural tissue by reconstruction of the overlying tissue layers. Dural reconstruction often requires a synthetic dural substitute. Unfortunately, all the available materials seem to fail in preventing adhesive scars and re-tethering. We describe the use of banked homologous amniotic membrane (AM) as a dural substitute for MMC repair in newborns.

Methods: AM was obtained from donors undergoing cesarean section. Under sterile conditions, AM was carefully detached from the chorion, rinsed in saline solution, and cryopreserved in liquid nitrogen. Three newborns affected by open MMC were operated within 24 hours from delivery, whilst two cases were operated one and two weeks after, respectively. The AM patch was used in different positions and settings. The amnion was sutured both on the placode and on the dura in two cases, whereas in the other three patients it was placed and sutured on the dural layer only.

Results: No adverse events occurred. Surgical wounds healed without complications and MRI scans taken at 3 and 6 months after surgery showed a satisfying de-tethering of the spinal cord with no signs of new adhesions.

Conclusions: We suggest a standardized technique which consists in the interposition of AM between the closed placode-dura and the myocutaneous flap as a protective tissue for the treatment of MMC in newborns. We postulate that the use of an AM patch can combine the mechanical properties of a dural substitute with the anti-adhesive properties of the amnion, with a synergistic effect. Such features are most important in preventing tethering and facilitating de-tethering. The use of a homologous banked AM patch provides an opportunity to have a great amount of quality sterile material with viable anti-inflammatory and anti-adhesive properties.

PP053

Prenatal diagnostics of encephaloceles and myelomeningoceles and their further neurosurgical treatment

Leonid Marushchenko, Lydmila Verbova, Ivan Protsenko, Pavlo Plavsky, Volodymir Mikhalyuk, Andriy Svyst, Myroslava Marushchenko (Kyiv, Ukraine)

Introduction: Prenatal diagnostics of neural tube defects allows to increase the effectiveness of surgical treatment of neonates with CNS dysraphia in cases when the parents decide to preserve pregnancy. The purpose of the study is to assess the possibilities of diagnostics of encephalocele and myelomeningocele of the fetus for further surgical treatment.

Methods: In current study the analysis of treatment of 53 children with neural tube defects for 2008-2017 years was performed. The median gestational age of diagnostics of CNS malformations according to the ultrasound data was 23,7±4,4 weeks. MRI of the fetus was performed in 27(50,9%) cases. Encephalocele was diagnosed in 5(9,4%) causes, myelomeningocele – in 48(90,5%). Fronto-ethmoidal encephaloceles were detected in 2(3,8%) patients, occipital – in 3(5,6%); myelomeningoceles of cervical spine – in 4(7,5%), thoracic – in 6(11,3%), lumbosacral spine – in 38(71,7%). Associated hydrocephalus was determined in 42(79,2%) patients, Chiari II malformation in 41(77,3%). Postnatal follow-up examination in all cases confirmed prenatal diagnosis.

Results: Cesarean delivery was in 44(83%) cases in order to prevent damage of hernial sac. Children with hernias complicated with the liquorrhea and with a threat of rupture of the hernial sac were transferred to Department of Pediatric

Neurosurgery. 26(49,0%) newborns with ruptured myelomeningocele and with a threat of rupture were operated during first 24-48 hours after birth. In 27(50,9%) children with preserved hernial sac reconstructive operations were performed within 1-4 months of life.
Conclusion: Ultrasound and MRI prenatal diagnostics of neural tube defects allows to determine the type of CNS malformation, combined congenital pathology as important prognostic signs of the further child's development. Prenatal diagnostics makes it possible to plan the surgical treatment of neural tube defects in newborns immediately after birth and to transfer these operations from urgent to planned emergency, thereby increasing the effectiveness of surgical treatment of this category of patients.

PP054

Dizygotic twins of different gender with concordant myelomeningocele: a case report and review of the literature

Cécile Balmer, Sarah Stricker, Raphael Guzman, Jehuda Soleman (Basel, Switzerland)

Introduction: Myelomeningocele (MMC) is a common subtype of congenital neural tube defects (NTD). Twins show higher rates of NTD, whereas the concordance of spina bifida in dizygotic twins is extremely rare.
Methods: We present a case of dizygotic unlike-sex twins presenting with concordant MMC, live-born and surgically treated within 48h of life. For further analysis of a possible underlying genetic cause a genetic analysis of the twins is being performed.
Results: Dizygotic twins of different gender were born at term to a 35-year-old woman after an uneventful pregnancy. Due to infertility caused by endometriosis, the twins were conceived through intracytoplasmic sperm injection without preimplantation diagnostics. There was no family history of congenital malformations or consanguinity. Folic acid was supplemented peri-conceptually and throughout the whole pregnancy, while mild gestational diabetes was treated with diet only. At 18 weeks of gestation both twins were diagnosed with a lumbosacral MMC by ultrasound. The known MMC was accompanied by ventriculomegaly and Arnold-Chiari Malformation type II. Both twins underwent successful surgical repair of the MMC 48 hours after birth, while due to progressing hydrocephalus both underwent the insertion of a ventriculo-peritoneal shunt one week later. The further postoperative course was uneventful. The genetic analysis is still ongoing and will be presented.
Conclusion: Concordance of MMC in twins is a rare finding. Only 18 pairs of twins, of which 8 were dizygotic twins, have been described so far. Results of genetic analysis in twins with MMC are lacking in all reports so far. We will present a review of the literature, the results of our genetic analysis and discuss possible implications of these genetic results.

PP055

Myelomeningocele: 10 years of follow up in Slovenia

Peter Spazzapan, Tomaž Velnar (Ljubljana, Slovenia)

Introduction: Myelomeningocele (MMC) is a congenital malformation that results from a failure of neurulation. Beyond the exposed spinal cord, virtually all patients with MMC have an associated Arnold Chiari malformation and most of them require a shunt. Some of them present progressive neurological deficits caused by tethered spinal cord and by hydrosyringomyelia.
Methods: We present a retrospective analysis of all children treated in Slovenia for MMC in the last 10 years (2007-2017). All neurosurgical aspects were evaluated: the need for shunt placement and shunt revision, the need for cranio-cervical decompression, for spinal cord detethering and for surgical treatment of hydrosyringomyelia.
Results: 19 children were treated in Slovenia for MMC from 2007 to 2017. They were all followed by a multidisciplinary team. The mean follow-up was of 5 years. All MMC were repaired within 48 hours of life. 12/19 needed a shunt and 11/12 needed a shunt revision during the follow-up. 3/19 children needed a cranio-cervical decompression for a symptomatic Arnold-Chiari malformation, 2 children needed a spinal cord detethering procedure for progressive neurological deficits. No patient had a direct surgical procedure of the hydrosyringomyelia. Different stages of ambulatory problems were present among 19 children: 10 presented complete paraplegia, 5 presented normal ambulatory capacities, 4 were able to walk with orthoses. Only 1/19 children had normal urinary function, 1/19 had urinary incontinence, 17/19 had a neurogenic bladder. 1/19 child had an incontinent bowel, all the others had different levels of chronic constipation. An orthopaedic follow up was arranged for all the patients.
Conclusions: Children with MMC must be followed throughout childhood by a multidiciplinary team. The results of our MMC series is not differing much from those of larger series. Among all neurosurgical procedures shunt revision was the most frequent. Shunt function should be first to be assessed whenever a child with MMC presents with clinical deterioration.

PP056

Spina bifida: routine or urgent surgery?

Ivan Protsenko, Leonid Marushchenko, Pavlo Plavsky, Volodymyr Mykhalyuk, Andriy Svyst, Lyudmyla Verbova (Kyiv, Ukraine)

Introduction: Nowadays congenital malformations of neonates still remain to be challenging medical, social, and ethical issues, and spina bifida is among the most common of them.
Objective: To improve the treatment of children with spina bifida.

Materials and Methods: 183 children with spina bifida have been treated along three years in our clinic. 54 patients (29.5%) had the liquorrhea at the admission and 65 (35.5%) children were under its threat. These 119 (65%) patients underwent urgent surgical treatment, while the others − the routine one.
Results: The basic principles of spinal herniation surgery are the removal of hernia sac, restoration of dura mater and soft tissues integrity around hernia, and the release of a spinal cord fixation. Urgent and routine surgery approaches for this spina bifida is almost the same, though the routine surgery has more benefits such as enough lead time for the detailed preoperative examination and thorough planning of the surgical treatment. The algorithm of diagnostics and therapy of children with the spina bifida depends on the integrity of the hernia sac, liquorrhea presence, and the severity of concomitant hydrocephalus. In case of spina bifida combined with the progressing hydrocephalus both dysraphia and shunting surgery were performed. Ifsimultaneous operation was associated with the extreme risk, liquor shunting has been performed first, and after a few days hernia has been removed (or vice versa).
Conclusions: The algorithm of diagnostics and therapy depends on the spina bifida character and requires individual approach in every single case.

Poster track 2.4: CVJ & Spine

PP057

A presumed atlanto-axial osteomyelitis with secondary cervical subluxation in a neonate: a case report

Andrew Croall, Kevin Agyemang, Anthony Amato-Watkins, Emer Campbell (Glasgow, United Kingdom)

Introduction: The management of osteomyelitis of the upper cervical spine has been extensively discussed in the neurosurgical literature in the last three decades. We present the youngest patient, a neonate, with suspected upper cervical osteomyelitis and discuss their non-operative management.
Methods: A 4-week-old neonate, born at term, presented with an out-of-hospital cardiac arrest. Initial investigations identified sepsis, but no obvious source. Following the development of secondary central respiratory failure and quadriparesis due to spinal cord injury, a delayed diagnosis of atlanto-axial subluxation was made. A retropharyngeal abscess was identified and following rupture; microbiological cultures grew *Staphylococcus aureus*. The differential diagnosis included Grisel Syndrome (inflammatory induced ligamentous laxity with secondary subluxation).
The patient was treated non-operatively, with both intravenous and oral courses of antibiotics for a period of 3-months. The cervical instability was controlled via a bespoke cervical orthosis for the duration of hospital admission. Respiratory and neurological function improved.
We describe the difficulties encountered in the MRI interpretation of possible infection in the upper cervical spine of neonates and describe the challenges in managing instability of the upper cervical spine in a neonate.
Conclusion: Conservative management of probable osteomyelitis with antibiotics and immobilisation has thus far proven successful in a neonate with a high cervical cord injury.

PP058

The use of translaminar C2 screws in occiptocervical fixation for pediatric patients with C1-C2 instability due to muccopolysaccharidosis: a case report

Marcos Dalsin, Alvaro Ernani Georg, Elisa Bonotto Pietta, Ápio Cláudio Martins Antunes, Jorge Wladimir Junqueira Bizzi (Porto Alegre, Brazil)

Introduction: Muccopolysaccharidosis (MPS) is a metabolic disease caused by the absence or malfunction of lysosomal enzymes needed to breakdown glycosaminoglycans. In MPS type IV (Morquio), these molecules accumulate inside cells and connective tissue causing progressive damage, including skeletal dysplasia and, frequently, odontoid hypoplasia with subsequent spine instability. In pediatric patients, cervical spine arthordesis is a challenge due to the smaller anatomical structures and more delicate bone tissue.
Objective: To describe the use of translaminar C2 screws as an alternative in occiptocervical fixation in a pediatric patient with MPS with craniocervical junction instability.
Case Report: A 4-year-old female with a recent diagnosis of MPS type IV was brought to the emergency unit with sudden tetraparesis (strength grade 1) after falling to the floor. Magnetic resonance imaging (MRI) showed invagination of the posterior arch of C1 through the foramen magnum and odontoid hypoplasia causing spinal cord compression. A suboccipital craniectomy with posterior C1 arch removal was performed to decompress the spinal cord. The instability was treated with posterior fixation using an occipital plate connected to two translaminar C2 screws, and the arthrodesis was performed with autologus bone from the 5th costal arch. The patient developed progressive neurological improvement (strength grade 4).
Discussion: Arthrodesis of upper cervical spine in small children is difficult and even more challenging in MPS patients due to abnormal bone development. In general, posterior wirings fixations are safe and technically simple procedures, with, however, high failure fusion rates. Transarticular C1–C2 screws and C2 pedicle screw provide high fusion rates. However, are associated with the risk of vertebral artery injury and are technically challenging due to smaller bone size, immature bone formation or variable anatomy. The use of translaminar C2 screws has been demonstrated to be an effective and safe technique

for internal fixation in pediatric patients with atlantoaxial instability from different etiologies, including MPS.

PP059

Currarino syndrome: case report and review of the literature

Dominika Števková, Frantisek Horn, Miroslava Fuňáková, Dana Kuniaková, Martin Smrek (Bratislava, Slovak Republic)

Introduction: Currarino syndrome is a hereditary complex of malformations characterized by 3 basic features called Currarino trias – anorectal malformation, sacral defect and presacral mass. Incomplete form missing one or two of the triad´s elements is also known and the clinical manifestation is extremely variable even in one family.
Methods: We present a case of a boy born in December 2008 with meningomyelocele in lumbosacral region covered with full skin, anal atresia with perineal fistula and partial sacral bone agenesis. Canalis sacralis was open in dorsal part and intradural accumulation of fibrolipomatous tissue was connected to subcutaneous lipoma spread diffusely in gluteal region. The tethered-cord was confirmed on MRI.
Results: The surgical resection of lipoma was realized in September 2009 because of development of the tethered-cord syndrome. Thanks to sufficiency of fistula and good clinical state of patient the perineal anoplasty and creation of neoanus was realized only in March 2017.
Conclusion: Therapy of Currarino syndrome depends on the clinical manifestation. The aim was to report our experience and the surgical management of children with Currarino syndrome at Paediatric Surgery Department of University Children Hospital in Bratislava. We also summarize information about Currarino syndrome, its manifestation, inheritance, diagnostic methods, therapeutic options and prognosis. Five electronic databases were used to explore the literature, including PubMed, Scopus, Medline, ScienceDirect and Embase.

PP060

Lumbosacral lipomas: do lipids play a role in disease progression?

Victoria Jones, Dominic Thompson (London, United Kingdom)

Introduction: Lumbosacral lipomas are a rare congenital pathology with variable natural history but the potential to cause significant neuro-urological disability. Although near-total resection of the lipoma tissue is the current gold standard treatment, there are at present, no good markers to guide timing of surgery. We have compared lipid abundance between symptomatic and asymptomatic lumbosacral lipoma patients to determine if lipid abundance is associated with disease progression.
Methods: CSF, plasma and urine samples were collected from patients undergoing near-total resection of a lumbosacral lipoma. Four patients were ostensibly asymptomatic and 13 patients had proven symptoms based on neurological and urodynamic assessment. Lipids were extracted, separated with high performance liquid chromatography, ionised with electrospray ionisation and analysed with Orbitrap mass spectrometry to generate a lipidomics profile. Further identification of lipids was achieved through comparison and analysis of retention time and fragmentation patterns of known standards using triple quadrupole mass spectrometry.
Results: Over 4000 lipids were detected in CSF samples with 528 showing a significant difference in abundance between symptomatic and asymptomatic patients ($p<0.05$). Of note, a large number of these lipids were more abundant in asymptomatic patients than symptomatic patients. This distribution was seen in both plasma and urine samples. Targeted lipid assays revealed a difference in abundance of both phosphatidylethanolamines and phosphatidylcholines.
Conclusion: There are significant differences in the abundances of different lipid subtypes between children whose lumbosacral lipomas are symptomatic and those that remain asymptomatic. This could provide the basis for the future development of a biomarker to predict disease progression. The increase in abundance of many different lipids in asymptomatic patients suggests that some lipids may play a protective role, potentially limiting disease progression.
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PP061

Achondroplasia and Langerhans cell histiocytosis of the spine. An unusual association

Pasquale Gallo, Lesley Simpson (Edinburgh, United Kingdom)

Objective: Achondroplasia, the most common skeletal dysplasia, is caused by a mutation of fibroblast growth factor receptor-3. Langerhans cell histiocytosis (LCH), a myeloid-derived dendritic cell disorder,is a rare condition, estimated to be between two and ten cases per 1 million children aged 15 years or younger.
LCH causes destructive lesions in a child's spine. Few large, long-term studies have evaluated the clinical and radiographic presentation, natural history and outcomes of modern treatment approaches after the diagnosis of this disease in children. Nevertheless no description exists in the medical literature, to our knowledge, of LCH affecting the spine of a patient with achondroplasia.
Case Report: A 7-year-old achondroplastic boy, known to our Department for previous foramen magnum decompression performed when he was 5-year-old, was admitted with a 3 weeks history of mid-thoracic back pain following a fall. Over the 7 days prior to admission this pain had become severe. His left leg had also started giving way and he was

using a walking stick to mobilize. A MRI spine on admission showed abnormalities consistent with a pathological fracture of the T1 vertebrae as well as soft tissue extending posteriorly from this and compressing the spinal cord. He was initially managed with bed rest and analgesia. A CT guided needle biopsy was performed which revealed a LCH. He had also a whole body MRI that showed a focal abnormality of the right acetabulum compatible with a further LCH deposit.

Following these results his case was discussed in a multidisciplinary meeting. Neurosurgical treatment was not advised and despite a multifocal bone LCH is often treated with pulses of Prednisone and Vinblastine we elected to treat more conservatively up front with anti-inflammatory Indomethacin. We felt that, in this young man with significant spinal deformity (exaggerated lumbosacral lordosis and thoracolumbar kyphosis, as well as scoliosis), it was best avoid steroids for the increased risk of osteopenia and further vertebral fragility.

He was discharged from the Hospital with a Somi-brace, extensive physiotherapy and occupational therapy input.

Six months after the start of the Indomethacin treatment the pain was completely resolved and the brace was discontinued. Serial follow-up spine MRIs showed a progressive resolution of the pathological fracture of the T1 vertebrae as well as the soft tissue extending posteriorly from this with complete resolution of the spinal cord compression. The child gradually returned to his previous daily life and school activities.

Patient has been on indomethacin for one year and tolerating it very well, with Omeprazole for gastric protection with no relapse of the disease.

Conclusions: This exceptional case highlights that in patients with achondroplasia, even with significant spinal deformity, the natural history of spine LCH is such that aggressive surgical management can be avoided. Indomethacin, extensive physiotherapy, occupational therapy input and follow-up are necessary to monitor recovery and spinal balance.

Poster track 2.5: Neonatal hydrocephalus

PP062

Neonatal ventriculoperitoneal shunts continue to have a higher failure rate than those of older children

Maya Kommer, Michael Canty, Anthony Amato-Watkins, Emer Campbell, Meharpal Sangra, Roddy O'Kane (Glasgow, United Kingdom)

Introduction: Many factors, including age, determine failure rates of ventriculoperitoneal (VP) shunt insertion. The goal of this study was to determine whether neonatal ventriculoperitoneal shunts continue to have higher failure rates than those of older children.

Methods: We identified all paediatric patients that had undergone a primary ventriculoperitoneal shunt insertion between January 2012 and December 2015. Data was collected retrospectively from our operative database and patient records.

Results: 22 neonates received primary ventriculoperitoneal shunts in the study period. Five were premature. The most common aetiology of hydrocephalus was dysraphism followed by intraventricular haemorrhage. Three patients had a ventriculosubgaleal shunt inserted prior to definitive VP shunt insertion, two had an external ventricular drain (EVD). 59% (13 patients) had their shunt revised at an average of 255 days post insertion (range 10-1235 days). Reasons for removal were blockage (five), migration/malposition (four), overdrainage (two), infection (one), unknown (one). The one-year revision rate was 45.5%.

39 patients in the non-neonate group had a primary ventriculoperitoneal shunt at an average age of 4.8 years (range 100 days to 15.9 years). The most common aetiology of hydrocephalus was congenital followed by tumour. Three had had an EVD prior to VP shunt insertion. 41% (16 patients) had their shunt revised at an average of 289 days post insertion (range 7-1042 days). Reasons for revision were blockage (four), infection (four), headache (three) malposition/migration (two), disconnection (two) and one subdural haematoma. The one-year revision rate was 28.2%.

Conclusion: Neonatal shunts are much more likely to fail than those of older children. We should aim to identify strategies to reduce this by either delaying insertion where possible or using techniques such as image guidance to reduce rates of malposition.

PP063

Subgaleal shunting for post-hemorrhagic hydrocephalus in preterm infants – A series of 17 cases

Alexandre Canheu, Marcio Francisco Lehmann, Felipe Inacio Ferreira Da Silva, Bruno Loof Amorim, Francisco Spessato Pesente, Sergio Murilo Georgeto (Londrina, Brazil)

Introduction: Posthemorrhagic hydrocephalus comprises the most common complication in those premature neonates whose suffered germinative matrix hemorrhage, and its treatment is still matter of controversies. These infants usually weight less than 1500g and a temporary CSF diversion device is needed. This paper depicts the initial 17 cases of ventriculosubgaleal shunting (VSGS) in prematures who suffered grades III and IV periventricular hemorrhage and hipertensive hydrocephalus.

Methods: We analyzed a series of 17 preterm infants who underwent VSGSfor posthemorrhagic hydrocephalus between July 2015 and November 2017. The median gestation age was 28 weeks (range 23-32). The median weight was 890 grams (range 625-1615). The mean span of time harboring de VSGS was 59,3 days (range 39-78).

Results: The cases were studied for: 1) revision of the system: 4 subjects (23,5%) had 2 system revisions each due to

obstruction; 2) shunt related complications:5infants (29%) had positive CSF culture and the VSG converted to EVD and further VP shunt. (2 of these showed previous CSF fistula through the wound); 4) convertion to VP shunt: 9 children (52,3%) showed persistent hydrocephalus at further investigation, and underwent to VP shunt after stable clinical condition and weight higher than 2000g. One death occurred in a very low birth weight subject who developed severe thrombocytopenia and pulmonary hemorrhage.
Conclusions: VSGS is a good alternative method to treat post-hemorrhagic hydrocephalus, specially in preterm infant whose need a temporary shunt device. The low rate of severe complications and encouraging results about persistent hydrocephalus should guide further investigation and larger cases series.

PP064
Subgaleal shunting for post-hemorrhagic hydrocephalus – Technical note
Alexandre Casagrande Canheu, Marcio Francisco Lehmann, Felipe Inacio Ferreira Da Silva, Bruno Loof Amorim, Francisco Spessato Pesente, Sergio Murilo Georgeto (Londrina, Brazil)

Introduction: Subgaleal shunting (SGS) for posthemorrhgic hydrocephalus (PHH), is considered as an option to control the intracranial hypertension of those preterms affected. This paper suggests a standard technique of valveless shunt.
Methods: A right precoronal 2 cm "S" shape incision is made and subgaleal (SG) space is entered. A blunt dissection creates a pocket, as big as one can do, within the SG space giving enough room to house de CSF drained from the ventricle. A burr hole is made at the Koch's point followed by standard puncture of the frontal horn. The ideal catheter is 10-14 cm, one piece, with inline reservoir and distal end catheter with no second piece or ligatures, and no valve as well; thus, the hemorrhagic CSF can flow directly to the SG pocket as needed. The better closure should be done in two layer sutures and we prefer non-absorbable stitches at the skin, decreasing dehiscence of the wound stretched by the CSF pocket.
Discussion: The "S" shape incision gives more elasticity to accommodate the reservoir beneath the thin skin as well allows the more generous dissection of the SG space; the aesthetics is improved too. We always implant valveless devices to ensure the ongoing drainage of the CSF and lessering the obstructions. A ventricular catheter with inline reservoir and distal end is preferable rather than a free catheter since the first can be punctured anytime when the outflow is getting obstructed; and better than Omaya reservoir, in our opinion, since the last is bigger in a higher profile and do not collect CSF at the SG space.
Conclusions: SGS is a good alternative method to treat PHH, and this technique variation proposed herein can maintain the CSF shunt for a longer time span lowering complications

PP065
The concentration of Doublecortin in the cerebrospinal fluid from human infants is developmentally regulated
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Introduction: Doublecortin (DCX) is a microtubule-associated protein specifically expressed in neuronal precursor cells and migrating neurons, and mutations in the human DCX gene cause lissencephaly or subcortical band heterotopia. DCX is a neurodevelopmentally regulated protein, and its concentration in the rodent and human brain, as well as in the cerebrospinal fluid (CSF-DCX) from rodents, declines sharply during early postnatal stages. Our group recently showed that a hypoxic-ischemic (HI) brain injury in the rat neonate significantly disrupted the physiological downregulation of CSF-DCX, leading to a significant increase in CSF-DCX for up to a week post HI. This increase correlated positively with the severity of HI-induced brain infarct, and also, but to a lesser extent, with HI-induced neurogenesis. In the current study, we address the clinical relevance of DCX in the CSF by measuring DCX concentration in the CSF from human infants, and examining the potential relationship between CSF-DCX and clinical data.
Methods: CSF was collected from pediatric patients requiring neurosurgical treatment. CSF was centrifuged at 2000 rpm at 4°C, and supernatant stored at -80°C until analysis. DCX concentration was measured with a sandwich immunoassay using electrochemiluminesence (Mesoscale platform) with a detection limit of 5 pg/ml. Clinical data was retrospectively collected from individual patient charts. Chronological age was adjusted for premature birth.
Results: A total of 61 CSF samples from 39 patients (20 females) were collected between February 2013 and June 2016. Out of these 61 samples, 34 were repeated collection from 13 patients. The patients' age ranged from 0.28 years before due date to 18 years (median 0.64 years; interquartile range [IQR] 0.07, 5.56 years). Indication for neurosurgical treatment were congenital hydrocephalus (8 patients), baclofen pump implantation (8), infection (5), neonatal intraventricular hemorrhage (4), symptomatic arachnoid cysts (4), tumors (4), shunt dysfunction (2) and other indications (4). DCX could be quantified in 29 samples from 16 patients. All 14 patients who were less than 4 months old had detectable DCX levels ranging from 13 to 21'568 pg/ml (median 612 pg/ml; IQR 129, 3'495 pg/ml) with DCX levels decreasing with age. Indication for neurosurgical treatment among those patients were congenital hydrocephalus (5 patients), neonatal intraventricular hemorrhage (4), infection (3), and symptomatic arachnoid cysts (2). Only two patients older than 4 months had detectable DCX in the CSF: a 3-year-old female with an ependymoma (DCX

level in the CSF 93.5 pg/ml) and a 5-year-old male with a grade III astrocytoma (331 pg/ml).
Conclusions: Our preliminary results show an age-dependent downregulation of DCX protein in the CSF of humans until 4 months of age, similar to that observed in rodent neonates/juveniles. This time window for the detection of DCX in human CSF strikingly coincides with the recently described extensive migration of DCX positive cells to the frontal lobe in the human brain that persists for 5 months after birth. To further understand the clinical significance of DCX in the CSF, the human CSF samples will also be evaluated for markers of neuronal damage (S100beta and NSE).

PP066
Shunt valve use in neonatal hydrocephalus: a single center experience
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Introduction: The management of hydrocephalus in neonates presents a unique set of challenges compared to older children. Shunt survival is the desired goal of treatment. Complications such as infection and over drainage add a significant burden to this patient population. We sought to determine whether there is any difference in the use of a flow-controlled valve over a differential pressure valve with an anti-siphon device in this patient population.
Methods: A retrospective case series of primary ventriculopertioneal shunt procedures in the first four weeks of life from August 2011 to August 2016 at Sheffield Children's Hospital was performed. Eighteen patients were included in this study, four were born prematurely and fourteen at full term.Seven neonates had a flow regulated (Orbis-Sigma) valve implanted and eleven had an adjustable valve (Miethke ProGav).
Results: The one-year shunt survival was 18% (2/11) for the adjustable valve group and 43% (3/7) for the flow regulated valve group. Shunt failure was due to distal blockage (3), infection (3), proximal catheter disconnection (1), proximal blockage (1), unspecified blockage (1) in the adjustable valve group.In the flow regulated valve group shunt failure was due to infection (1), proximal blockage (1), pseudomeningocele (1) and unspecified shunt dysfunction (1).
Evidence of significant over drainage was seen in four patients. This included three patients primarily treated with adjustable valves. These patients had their valve settings increased to 8, 8 and 10 respectively. One patient primarily treated with a flow regulated valve also developed over drainage.
Conclusion: One-year shunt survival rates are poor in neonates regardless of shunt valve selection. Our case series demonstrates that flow regulated valves may potentially be associated with a lower incidence of over drainage in neonates.Symptomatic overdrainage can be managed by increasing shunt settings in patients with adjustable valves.

PP067
Borehole-Reservoir in patients with hydrocephalus and a weight under 2000 gram
Lutz Schreiber, Francisco Brevis, Ameer Alyeldien, Martin Scholz (Duisburg, Germany)

Object: Hydrocephalus in newborns and early born children is a well-known disease which can be treated with lumber puncture or ventricular tapping through the fontanelle.
As a repetitive procedure, the ventricular tapping is not preferred as it is a brain traumatising procedure. The Borehole-Reservoir is a good alternative which allows aspiration of the CSF for a couple of weeks.
Method: In the last two years, we have been managing all the hydrocephalus cases, in which the patient weights less than 2000gram, with a Borehole reservoir (Mini-Rickham). We have been doing one to two punctures per day to reduce the intracranial pressure. The surgery is ultrasound-navigated. We have been using a 3.5 to 4.5 cm long ventricle catheter, depending on the intraoperative ultrasound.
Once the weight of 2000gram is reached, a shunt will be connected to the borehole reservoir.
Results: No infection was documented in all cases with a borehole-reservoir.
A daily puncture of the reservoir has been done up to 6 weeks in case of very early born children.
In one case an emergency surgery had to be done with the age of 11 months as the child grew and thereupon the ventricle catheter has been gone too short.
As a standard, all the ventricular catheters have been replaced with a longer one (longer than 4.5 cm) during the first year of life.
Follow-up ultrasound examinations were being done every 3 months during the first year of life.
Conclusion: Implanting a borehole reservoir is a good alternative technique to manage the hydrocephalus cases before implanting a shunt in very early born babies, having the advantage of reducing the trauma of the brain.
It is important to check the length of the ventricular catheter in standardized regular follow-up sessions to plan the re-surgery to replace the ventricular catheter with a longer one.
The follow-up sessions should be ultrasound assisted.
The infection's rate is very low.

Poster track 2.6: ETV

PP068
Endoscopic third ventriculostomy in the management of hydrocephalus and outcome analysis in 51 children
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Introduction: Endoscopic third ventriculostomy (ETV) is a safe method of choice in treatment of hydrocephalus. Age and etiology are important factors that determine success rates of endoscopic third ventriculostomy (ETV). In our study, we retrospectively analyzed single surgeon experience the data of with 51 children who underwent ETV treatment for different etiologies and different age groups of the hydrocephalus between 2001 and 2016.
Methods: Between 2001 and 2016, 51 children patients with obstructive hydrocephalus underwent ETV. The patients fell into three groups. These groups were <6 months of age, 6-24 months of age, >24 months of age. All ETVs were performed by the same neurosurgeon.
Results: Overall success rate of ETV was in 80% for all etiology and ages. In patients for all etiology, the success rates of <6 months of age, 6-24 months of age, and >24 months were 56.2%, 88.9% and 94.1%, respectively. In aqueductal stenosis, the highest success rate was obtained. Success rates of post-hemorrhagic, post-infectious and spina bifida related hydrocephalus were 60%, 50%, and 14.3%, respectively. While success rate at the first ETV attempt was 85.3%, in previous V-P shunt performed patients, it was 76.9%.
Conclusion: Based on our experience, ETV could be the first method of choice for hydrocephalus even in children younger than 6 months of age. In our experience, factors indicating potential failure of ETV were young age and etiology like hemorrhage, infection other than isolated aqueductal stenosis. ETV is the method of choice in obstructive hydrocephalus even in patients with former shunting. Fast formation of new arachnoid membranes and lower pressure gradient in infants than in older children can play a role in ETV failure.

PP069

The factors of efficacy of endoscopic third ventriculostomy in children under one year with obstructive hydrocephalus

Vasiliy Danilin, German Letyagin, Sergey Kim (Novosibirsk, Russia)

Introduction: Endoscopic surgery for obstructive hydrocephalus in children is an alternative to shunts. At present, more and more the effectiveness of endoscopic third ventriculostomy (ETV) in infants is discussed among neurosurgeons. In order to increase the effectiveness of ETV, many surgeons asking questions about what factors may influence on the efficiency of the operation in the younger age group?
Objective: To study the factors affecting the efficiency of ETV.
Material and Methods: In our clinic for 2012-2016 performed 88 endoscopic interventions for children the age under of one year. 43 children (that's 48.9% of the total number of operations) required in the subsequent execution of shunt, one child made repeated ETV. The median time to onset of clinical manifestations of closing the stoma was 3.9 months (116 days).
Results: The most effective operation was in congenital of obstructive hydrocephalus (64.5% effective operations), as well as the age over of 6 months (63.6% effective operations). In posthemorrhagic hydrocephalus efficiency was 42.6%. In intraventricular hemorrhage (IVH) 4 degree an efficiency operation the lowest was 25%. The overall effectiveness of ETV in children under one year in our study was 51.1%.
Conclusion: With congenital occlusive hydrocephalus, the efficacy of ETV is maximal and reaches 64.5%. Despite the lower efficiency ETV in children under one year at a posthemorrhagic hydrocephalus, this type of operation is a very good alternative to shunts.

PP070

Repeated endoscopic third-ventriculostomy: our case series and review of the literature

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Introduction: Endoscopic third ventriculostomy (ETV) is a safe and effective treatment for triventricular hydrocephalus. Failure rates in children range from 10% to 40%. In case of closure of the stoma, a shunt must be implanted or a repeated ETV can be performed. This choice may be treacherous and no protocol have been outlined in literature yet.
Materials and Methods: Our retrospective analysis includes 151 children who had undergone ETV from January 2006 to August 2017 in our Center. In 29 patients (mean age 5,9 years) we performed a repeated ETV. Etiologies of hydrocephalus included: 11 tumors, 7 intraventricular hemorrhages, 4 aqueductal stenosis in NF1, 4 primary acqueductal stenosis, one lumbosacral myelomeningocele, one vein of Galen malformation, one Blake's pouch cyst. The interval between first and second ETV ranged from 3 days to 8 years. The median follow-up was 64,5 months.
Results: In 22 patients we twice performed an ETV, while in 7 cases we proceeded with a third attempt. Among the 22 patients, 5 needed a ventriculo-peritoneal shunt, while in the third ETV group 2 patients required a shunt because of closure of the stoma. We directly performed a shunt implant, when a malresorptive nature of the hydrocephalus was established.
Our overall ETV failure rate at first attempt was 19,3%, while failure after repeated attempts was 27,6%. In 51,7% of the cases the ETV failed within 6 months after surgery, in 37,9% within the first 3 months. A repetition of the ETV was successful in 72,4 % of the patients.
Conclusions: According to our data and to the current literature, a redo-ETV is a valid and relatively safe treatment after the failure of a previous ETV. According to our experience, even a third attempt with an ETV can be effective, if the surgeon has ruled out the malresorptive mechanism of the hydrocephalus.

PP071

Endoscopic third ventriculostomy in hydrocephalus management in children – Comparing with adults

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Introduction: Neuroendoscopy has become a routine procedure in neurosurgery and has multiple indications including ETV, CPC, septostomy, cyst fenestration, biopsy, drainage, or resection of intraventricular tumors, colloid cysts, hematoma evacuation, pituitary surgery, craniosynostosis surgery, spine surgery, and as an adjuvant in microsurgery, allowing the operating surgeon to "look around the corner.

Aim of Study: To review the indications and results of ETV in hydrocephalus managment in children, comparing with adults.

Methods: We report on patients operated for hydrocephalus at the Department of Neurosurgery at the University Hospital Center "Mother Teresa" Tirana, between 2011 and 2016.

Results: During the study period we treated 156 hydrocephalus patients in 212 interventions. The interventions were categorised in 115 shunt interventions, 42 combined interventions of which 25 ETV, and 55 endoscopy interventions of which 36 ETV. 61 interventions (30% of procedures / 33% of patients) were ETV of which 28 ETV were performed on pediatric patients.

The age range from 3 months to 69 years-old (median 32 years-old). The follow-up period was 5 to 71 months (mean 49 months).

In children it was most often performed for extrinsic aqueductal stenosis (8), intrinsic aqueductal stenosis (7) and shunt malfunction (5). In adults it was most often performed for extrinsic aqueductal stenosis (13), shunt malfunction (7), Chiari I (5).

No mortality occured and morbidity was observed in 1 (2,5%) case (transient left occulomotor palsy). ETV failure was observed in 12 (19,7%) cases for a success rate of 80%.

Conclusions: ETV is an established method for the treatment of non-communicating hydrocephalus in children and adults. Carefully selected patients of non-communicating HCP and of HCP previously shunted benefit from ETV. Success rate is 80%. NPH patients (iNPH or after aSAH) often fail.

PP072

Deliberate endoscopic third ventriculostomy for hydrocephalus in infants. Towards a reduction in the morbidity of ulterior shunts?

Mathieu Helleringer, Olivier Klein (Nancy, France)

Background: The treatment of hydrocephalus by endoscopic third ventriculostomy (ETV) has a high risk of failures in infants less than 1 year old whereas the ventriculoperitoneal shunt can lead to a significant morbidity in the same age group. We wonder if ETV first could reduce the morbidity of subsequent shunt placement. The aim of this study is to check if deliberate ETV lead t reduction morbidity of ulterior hydrocephalus treatment in infants under 1 year.

Patients and Methods: 51 children less than 1 year treated for hydrocephalus from January 2011 to April 2016 were included in this retrospective study: 16 with ETV and 34 with shunt. Children with history of shunting before ETV procedure were excluded. CSF flow MRI was performed postoperatively to assess the ETV functioning except for two children. ETV success was defined as no need of shunting over a one-year period. Mean follow up was 33 months.

Results: Mean ages at the first treatment were 113 and 166 days for shunt and ETV groups respectively (p=0.8).41% of ETV (6/16 patients) were considered as success at first treatment and 50% (8/16) after second ETV. The same result was observed in patients with shunt devices 47% (16/34 patients). Among the ETV group, five patients were treated five patients for arachnoid cyst with hydrocephalus and four of them were considered as success. Complications were observed in 19 patients with shunt devices (56%) and in 2 patients with ETV (12,5%) (p=0.004). Only 3 patients with shunt after ETV had complications (37%).

Conclusion: As a high rate of complications was observed in infants less than 1 year old with shunt devices, we suggested to perform ETV in this population as the first intention although its transient functioning.

PP073

An optimized method of minimally invasive endoscopic third ventriculocisternostomy for children with occlusive hydrocephalus

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Introduction: In this article, we present an optimized minimally invasive method of ETV for children with occlusive hydrocephalus.

Methods: The study comprises sixty-four consecutive pediatric cases (34 boys and 30 girls aged from 1 month to 5 years) of occlusive hydrocephalus from various etiologies, which were treated with an improved technique of ETV. Mean clinical follow up period after ETV was 24.2±3.8 months.

Results: Application of the new technique made it possible to significantly reduce the length of the soft tissue incision for access, and the use of upgraded instruments allowed to perform a twist drill hole in the skull to less than half a usual size. Access to the brain and lateral ventricle was performed by blunt trephination of the dura without the need for significant corticectomy or coagulation, and yielded minimal damage to the brain, which is very important in patients of young age. Continued endoscopic control during the approach down to

the lateral ventricle increases safety and decreases risk of injury and can be performed in cases of pathologies affecting the anatomical relationships of the lateral and third ventricle. Mortality in our cohort was 0% and there were no post operative neurological, endocrinological or infectious complications. Patency rates of the first ETV performed was 78%, with the remaining patients requiring additional surgical procedures for complicated settings.
Conclusion: This new technique of minimally invasive ETV placement in pediatric patients is an effective and safe method to treat occlusive hydrocephalus and can be recommended for extensive clinical use.

Poster track 3.1: Oncology: General I

PP074
Treatment strategy for the expanding cyst of pediatric pilocytic astrocytomas
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Introduction: Although pilocytic astrocytoma (PA) shows a good prognosis after the total removal, but it cannot be completely removed depending on the location. If the residual tumor shows with cysts, it may increase with tumor growth, or only cyst may be enlarged. A treatment strategy for cystic expansion of pediatric PA was examined.
Methods: Thirteen cases of intracranial PA cases (average: 9.4 years old) who were performed the initial operation in 2000-2015 were investigated.
Results: Among them, nine cases with cystic components were found. Among the 9 cases, 4 cases didn't show postoperative cyst expansion (2 cases without recurrence after total removal, 2 cases after partial removal without postoperative treatment), 5 cases (4 cases of partial removal, 1 case of biopsy + cyst fenestration) showed cystic expansion of the remaining lesion. In the 5 cases, locations were 3 cases of optic pathway / hypothalamus, 1 case of cerebellum, and 1 case of brain stem. One case was totally removed by craniotomy, 1 case was performed cyst fenestration + partial removal (partial cauterization), 1 case was confirmed spontaneous regression by follow up observation. Two patients underwent chemotherapy (CDDP + VCR), among them, a case showed regression, but a case gradually increased and finally needed a fenestration. Ki-67 index of these expanded cases showed 2.1% on average, and that was lower than other groups. BRAF V600E was wild type in the two cases examined.
Conclusion: Cystic expansion may not be associated with spontaneous regression or increased tumor. Controlling cysts by chemotherapy including Bevacizumab is controversial. If it is asymptomatic or minor symptoms, spontaneous regression may be expected, but if symptomatic or expansion progression is rapid, surgical treatment is inevitable. In that cases, consider partial extraction as well as fenestration, depending on cyst contents, Ommaya reservoir is also considered.

PP075
Management of the immature teratoma in neonates: case report
Claudiu Matei, Monica Dragomir, Stefan Ioan Florian (Sibiu, Romania; Bucharest, Romania; Cluj-Napoca, Romania)

Introduction: Immature teratoma is a rare tumor classified as germ cell tumor. Immature teratomas are well-defined tumors, including hair, bone, cartilage, teeth, dermal cells; occasional intratumoral haemorrhages may occur. Microscopically immature teratoma contains elements from all three germ layers: ectoderm, mesoderm and endoderm. Diagnosis is based on imaging, serology by tumor markers and pathological appearance. Management of newborns with immature teratoma includes surgery and chemotherapy; radiotherapy is not part of a therapeutic armamentarium of children less than 5 years of age.
Methods: The aim of this paper is to highlight the main diagnostic and therapeutic features of the immature teratoma.
Case Presentation: A one-month-old girl patient was admitted to the Neurosurgical Department of Polisano Hospital in December 2014.The imagistic studies revealed a giant intracranial tumor located on the right brain hemisphere, with an apparent origin in the pineal region.
Results: The treatment was multimodal, consisted of three therapeutic sequences: surgery-chemotherapy-surgery. The follow up imagistic exam at two years show no tumor recurrence and tumor markers are in normal range. The little girl has a normal psychomotor development with a very good quality of life.
Conclusions: Immature teratoma is a very rare tumor, but unfortunately with is a lack of strong evidence data regarding the diagnostic and management. In our case, the immediate prognosis was favorable due to a prompt management consisting of three therapeutic sessions: surgery - chemotherapy - surgery. The medical literature attests a perioperative mortality of 17%, a normal psychomotor development in 46% of cases, a survival rate at 5 years of 70% and 76% of these patients have a satisfactory quality of life.

PP076
Infantile myofibromatosis of the skull base: two case reports
Gloria Tresserras, Giuseppe Mirone, Ursula Ferrara, Giuseppe Cinalli (Naples, Italy)

Introduction: Infantile Myofibromatosis is the most common fibrous tumor of infancy and early childhood, showing aggressive behavior in some cases. There are two types

of presentation: solitary or multi-centric lesions involving skin, subcutaneous tissue, muscle, bone and viscera.
Methods: We will present two cases of Infantile Myofibrotamosis of the skull base. We retrospectively analyzed the location, clinic manifestations, treatment and the follow-up of the cases. Our results are compared with those published in the literature and correlate with our management.
Results: One of them are located at anterior fossa extending along the ethmoid bone; the second was located in posterior fossa, extending up to supratentorial compartment. Surgical therapy is recommended in cases of mass effect or progressive increasing size. Partial removal of the occipital mass was achieved in one case, whereas the remaining lesion gradually reduced in size at follow-up. In the other case, complete tumor removal was obtained in ethmoidal mass, with no recurrences.
Conclusion: Infantile Myofibromatosis is a fibrous mesenchymal tumor. Intracranial involvement is rare and only four cases of the skull base have been published. The prognosis is generally good if the tumor does not involve visceral organs and complete spontaneous regression can be seen. The first choice for management is biopsy and close observation. When the tumor grows and shows signs of mass effect, resection should be considered. No consistent results of chemotherapy have been published.

PP077

Desmoplastic infantile ganglioglioma/astrocytoma – A retrospective series of 9 cases

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Introduction: Desmoplastic gangliogliomas/astrocytomas (DIG/DIA) are rare tumors predominantly arising in the pediatric population. This series aimed at evaluating their characteristics and management and determining potential mutations leading to their development.
Methods: This retrospective study included 9 pediatric patients treated between 1996 and 2014 for a DIG/DIA at Necker – Enfants Malades Hospital, Paris, France. Clinical, radiological, histopathological data and the type of treatment and outcome were analyzed. A whole genome sequencing (WES) was attempted in 8 cases.
Results: The median age was 9 months (1-38 months) with 5 boys and 4 girls. Symptoms leading to diagnosis were motor deficiency (5/9), intracranial hypertension (4/9), seizure (3/9) or macrocrania (3/9), and were progressing from some days to 6 months. Tumors involved predominantly the temporal (7/9) and parietal (4/9) lobes, with 5/9 multilobar tumors. In MRI, a contrast enhancement after gadolinium injection was observed in all cases and a cystic component was present in 7/9 tumors. No metastasis was detected at diagnosis. In histopathological exam, 4/9 cases presented with high mitotic activity and necrosis. All patients of the series were operated on in first intention. Three patients received chemotherapy for progressive disease. After a mean follow-up of 75 months, 4 patients were in complete remission, 3 patients had a stable disease, one patient had died and one had been lost to follow-up. No recurrent mutation was detected in WES in this series.
Conclusion: Desmoplastic gangliogliomas/astrocytomas (DIG/DIA) are rare tumors with a generally good outcome. Management is based on surgery, which has to be as complete as possible. Chemotherapy is a second intention treatment. No recurrent mutation has been detected that could be targeted by therapy.

PP078

The role of neurosurgery in pediatric versus adult Langerhans cell histiocytosis

Hosai Sadat, Hans-Jakob Steiger, Sevgi Sarikaya-Seiwert, Kerim Beseoglu, Thomas Beez (Düsseldorf, Germany; Bonn, Germany)

Objective: Langerhans cell histiocytosis (LCH) is a rare proliferative disorder of dendritic cells. Since pediatric disease is more common, guidelines were established in pediatric oncology. As there is no comprehensive analysis of differences between adults and children, recommendations in adults are partially derived from pediatric literature. Here, we analyzed the role of neurosurgery in LCH, with focus on children versus adults.
Methods: LCH patients treated between 2000-2017 were stratified for age (≤17 years versus >17 years) and demographic, medical and radiological variables retrieved.
Results: Twenty patients (10 female, 10 male) were stratified into pediatric (N = 12, mean age 7 years, range 1-17 years) or adult (N = 8, mean age 25 years, range 18-40 years). Presenting symptoms in children versus adults were localized pain in 50% versus 42% and palpable swelling in 25% versus 33%, respectively. Imaging revealed a skull lesion in 75% and 88%, with dural invasion in 33% and 38%. Two children presented with spinal and one adult with suprasellar lesion. Multi-organ involvement was found in two children and one adult. Whole-body MRI was performed in 92% of children, but only 38% of adults. Neurosurgical intervention was resection in 75% of children and 88% of adults and biopsy in the remaining, followed by chemotherapy in 50% of children and 25% of adults. At mean follow-up of 6 years (range 1-17 years), complete remission was achieved in 92% and 88%, respectively. No disease-related mortality occurred.
Conclusion: LCH mainly affects bone and rarely presents with dural or intradural involvement. Multi-organ disease occurs at comparable rates. Whole-body imaging was performed in almost all children, but only in one-third of adults. This might reflect stringent adherence to pediatric oncology guidelines, or better clinical guidance on pediatric LCH. Our study

highlights potential areas for research, which would be best performed in multi-center approach.

PP079

Use of tubular retractors in the minimally invasive complete resection of an intraventricular meningioma in a child

John Hanrahan, Prajwal Ghimire, Anna Oviedova, Wisam Al-Faiadh, Bassel Zebian (London, United Kingdom)

Introduction: Meningiomas are amongst the commonest primary brain tumours, accounting for approximately 30% of primary intracranial tumours in adults. Intraventricular meningiomas account for less than 5% of all meningiomas. They are a significant surgical challenge. Accessing these lesions usually requires the creation of a corticotomy and a corridor through brain parenchyma coupled with significant retraction increasing the risk of morbidity especially in large lesions which require a larger corticotomy and corridor.
Case Report: We report a case of a three-year old female undergoing complete macroscopic resection of an intraventricular meningioma using a monoportal endoscopic approach followed by the use of tubular retractors coupled with the microscope. She had presented with a four-month history of worsening ataxic gait and increasing falls, and a two-week history of early-morning vomiting. She had been previously fit and well. Brain MRI revealed a supratentorial lesion 7.5 x 6 x 4cm in size located in the left lateral ventricle. Complete resection was achieved through a staged minimally invasive approach, confirmed by post-operative MRI imaging. The tumour was classified as a WHO grade 1 meningioma. At six months follow-up, she was clinically well with no residual or recurrence.
Conclusion: Our case highlights the challenges posed by large intraventricular meningiomas. We have demonstrated that a minimally invasive approach facilitated by tubular retractors is safe and can achieve complete resection, although a staged procedure was required.

Poster track 3.2: Oncology: General II

PP080

The Dutch Pediatric Neurosurgery Network, the first results

Leonie van den Abbeele, Erik van Lindert (Nijmegen, the Netherlands)

Introduction: Pediatric neurosurgery in the Netherlands is performed in the seven academic hospitals by 14 neurosurgeons, all but one part-time pediatric neurosurgeon. About 1000 patients are being treated per year and 1500 surgeries performed. There is significant centralization of some subspecialties (epilepsy, plexus, craniosynostosis) between the hospitals and ongoing further centralization of other subspecialties (oncology, spinal dysgraphia) are threatening the discipline of pediatric neurosurgery.
Method: We initiated a Dutch Pediatric Neurosurgery Network in 2015 in which the Dutch pediatric neurosurgeons join in a virtual pediatric neurosurgical center. The network makes it possible to have online consultations with other pediatric neurosurgeons in other hospitals, and to share protocols and images as well as collaborate in clinical studies. In the Radboud University hospital we started in 2016 with a digital outpatient clinic in which teleconsultation in part replaces outpatient clinic visits. Neurosurgical patients and their parents have access to our network. They can find reliable information on their diseases and ask questions to the pediatric neurosurgical team.
Results: After one year the Radboud university hospital has about 80 patients in their virtual outpatient clinic. Parents are enthusiastic about the possibility to have a quick consultation on their concerns. Asking a question online feels like less time claiming. We also use our digital network for post operative checkups for minor surgery and save patients travel and waiting time, while parents do not have to take a leave from work. The DPNN is bringing the digital outpatients clinic also to the other academic hospitals. The 14 neurosurgeons collaborate in the Dutch Pediatric Neurosurgery Study Group for initiating and performing nationwide clinical studies and in October 2017 the first collective article on repeat third ventriculostomy was published.
Conclusion: All Dutch pediatric neurosurgeons working together in all aspects of pediatric neurosurgery and research and joining forces with parents and patients will have to secure and improve the future of Dutch Pediatric Neurosurgery.

PP081

The NOPHO-European study on the cerebellar mutism syndrome: study update

Jonathan Grønbæk (Copenhagen, Denmark)

Objective: The cerebellar mutism syndrome (CMS) is one of the most disabling adverse effects after neurosurgery for a posterior fossa tumor in childhood. The reported incidences vary substantially in previous studies. The pathophysiology is unknown, but damage to the cerebello-thalamo-cerebral circuits is likely. The aim of this study is to investigate risk factors for development and severity of CMS including surgery (approaches, techniques and tissue and vascular damage, re-operation) and host genome variants.
Materials and Methods: This multicenter study was originally developed as a NOPHO collaborative study coordinated from Rigshospitalet, Copenhagen with target accrual of 500 patients. During the last three years patients have been included in this study in 17 centers from eight European countries. Data are collected prospectively in

standardized clinical research forms (CRF) and registered online in a database developed for the study. Automated reminder-emails on missing or incomplete CRFs are sent out to the participating clinicians. Online participant meetings are held quarterly. Registration includes clinical data and speech samples collected preoperatively and at four defined postoperative points for the subsequent 12 months. Therapy is by local standards. A blood sample for genetic analysis is collected from all patients. Imaging is collected and reviewed centrally.

Results: The study opened in five Nordic and Baltic countries during 2014/2015; in the Netherlands in February 2016 and in the United Kingdom and Hungary in 2017. Centers from the United Kingdom and Germany are expected to join in 2018. As of November 2017, 182 patients have been included with an overall inclusion rate of 92% and CRF completion rate of 82%. Mutism has occurred in 19 cases (10%) and reduced speech is seen in 17 cases (9%). The target of 500 patients is estimated to be reached in spring 2021 by current trajectories.

Conclusion: The NOPHO-European study of the cerebellar mutism syndrome is running as planned with a high inclusion and data completion rate. Impaired speech is seen in 19% of patients of which muteness is accounting for 10%.

The study is estimated to reach 500 included patients in 2021 and thus the largest prospective international study on CMS to date. It is the first to 1) systematically register surgery, use of steroids, standardized speech samples and 2) to investigate the influence of host genome.

European neurosurgical centers are encouraged to join this study.

PP082

Surgery of the small tectal tumors in children

Taisiya B. Bazarkhandaeva, Williyam A. Khachatryan (St. Petersburg, Russia)

Introduction: It's believed that in the management of the small tectal tumors of the brain the wait-and-see tactic is more reasonable.

Methods: The analysis of the results of the examination and treatment of 12 children with tectal tumors up to 5см^3 was carried out, admitted over the period of 2002-2014. The age of the children ranged from 6 to 17 years. The average fallow-up period was 4.5 years. The diagnostics included clinical and laboratory examinations, neuroophthalmological, otoneurological examinations, evoked potentials of the brainstem, MRI, MR-tractography, PET with labeled methionine. The tumor volume was calculated by the formula $V = 4\pi [a \times b \times c]/3$.

Results: The main clinical presentation before surgery were hypertension-hydrocephalic and quadrigeminal syndromes. Tumor resection was performed in 6 children, CSF-shunting operation – in 4, radiosurgical treatment – in 1, and in 1 case the child was under observation. Histological examination revealed glioma of grade I-II in all patients. The position on the operating table in all cases – sitting position using supracerebellar subtentorial approach. To remove the intrinsic tumors we used median intercolliculiar wound corridor. After the operation, the patients' condition stabilized, regressed hypertension-hydrocephalic syndrome, and no new focal neurological symptoms were noted. It should be noted that regression of hypertensive syndrome was achieved after the operation; however, focal neurological deficit, caused by a neoplastic process, was persistent.

Conclusion: In view of the persistency of the preoperative neurological deficits and the risk of their progression, as well as the absence of the severe complication after operation the surgery for these patients can be the method of choice of the treatment.

PP083

Securing hemostasis in pediatric low-grade posterior fossa tumors: the value of thrombin-gelatin hemostatic matrix

Valentina Baro, Luca Denaro, Domenico d'Avella (Padova, Italy)

Background: Effective hemostasis is of utmost importance in neurosurgical practice. Pediatric low-grade posterior fossa lesions notoriously reach considerable dimensions in a complex anatomical region at the time of presentation, therefore achieving a valid hemostasis can be troublesome and time consuming. Gelatin thrombin hemostatic matrix is a useful tool widely used in neurosurgery.

Objective: We report our experience concerning the use of thrombin gelatin hemostatic matrix to strengthen final hemostasis after posterior fossa low-grade tumor surgery in children. To our knowledge this is the first report of the use of hemostatic matrix in pediatric neurosurgery.

Materials and Methods: Between 2012 and 2016, 23 consecutive patients underwent posterior fossa surgery for low-grade gliomas in our Department. The mean age at surgery was 6.35 years, 12 patients were female and 11 male. Histology was Pilocytic Astrocytoma in 15 cases, Ependymoma in 7 cases and Gangliocytoma in one case. At the end of surgery an effective hemostasis was achieved using standard methods and, in order to strengthened final hemostasis, we use a thrombin-gelatin hemostatic matrix.

Results: The post-operative MRI performed in-between 24-48 hours after surgery did not show any bleeding or ischemic or other complications imputable to the matrix. During radiological and clinical follow-up none of the patients showed any delayed complication related to the matrix.

Conclusions: In our practice we found thrombin-gelatin hemostatic matrix sealant a valid and safe tool for strengthening hemostasis in pediatric low-grade posterior fossa surgery resulting in reduced damage to the surrounding structures and preventing post-operative hemorrhage.

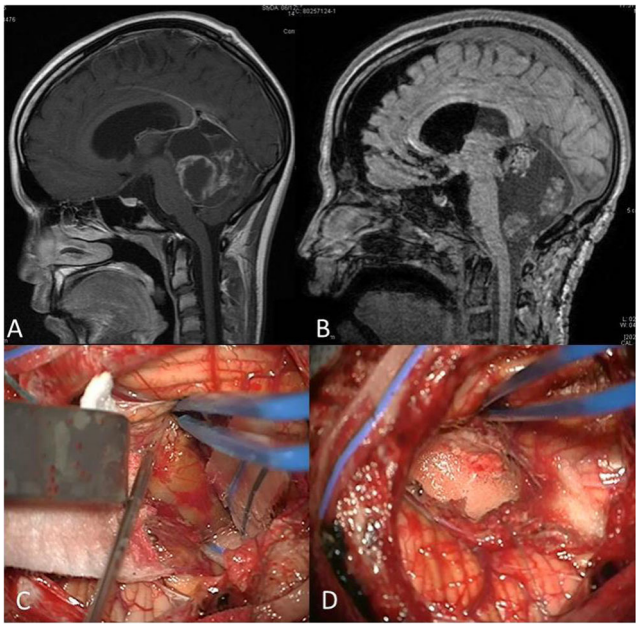

Figure 1: *A: Preoperative gadolinium-enhanced MRI shows a lesion within the fourth ventricle. B: Postoperative T1 weighted MRI shows the gross total resection without hemorragic complication. Histology revealed a Gangliocytoma WHO grade I. C: Intraoperative picture showing tumor exposal below the cerebellar hemispheres. D: Intraoperative picture showing the surgical field filled by thrombin-gelatin hemostatic matrix sealant.*

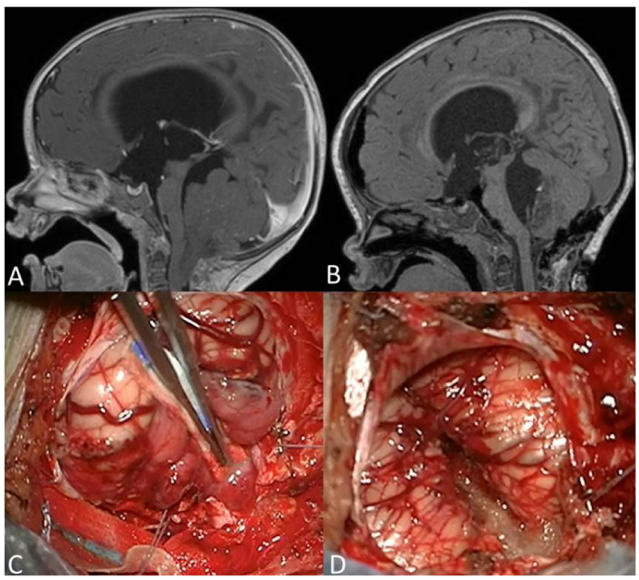

Figure 2: *A: Preoperative gadolinium-enhanced MRI shows a lesion within the fourth ventricle extending across the foramen magnum. B: Postoperative T1 weighted MRI confirmed the gross total resection without hemorragic complication. Histology revealed a Ependymoma WHO grade II. C: intraoperative picture showing the lesions protruding below cerebellar hemispheres crossing the foramen magnum. D: intraoperative picture showing the the surgical field filled by thrombin-gelatin hemostatic matrix sealant.*

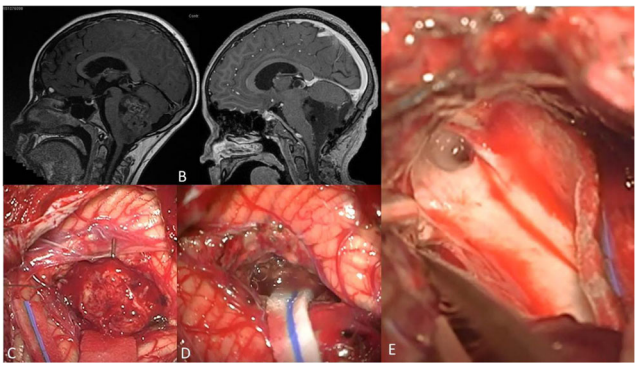

Figure 3: *A: Preoperative gadolinium-enhanced MRI shows a lesion within the fourth ventricle. B: postoperative gadolinium-enhanced MRI shows the gross total removal without hemorragic complication. Histology revealed a Pilocityc Astrocytoma WHO grade I. C: intraoperative picture showing the lesion below the cerebellar hemispheres. D: intraoperative picture showing the surgical field after the irrigation and suction of the matrix. E: intraoperative picture showing the floor of the fourth ventricles after tumor removal.*

PP084

Half/partial facial colliculus syndrome

Amets Sagarribay, Gonçalo Januário, Dalila Forte, Miguel Correia, Carla Conceição, José Pedro Vieira, Mário Matos (Lisbon, Portugal)

Introduction: Facial colliculus syndrome comprises ipsilateral lower motor neuron pattern of facial nerve palsy, ipsilateral VI nerve palsy and, sometimes, conjugate gaze palsy due to an associated contralateral medial rectus palsy (internuclear ophtalmoplegia). This syndrome may be seen in relationship with tumors, demyelination, infection or vascular diseases that compromise facial colliculus at the dorsal pons in the floor of the fourth ventricle. Facial nerve palsy alone related to IV ventricle lesions may be defined as "half/partial facial colliculus syndrome" as the authors show in relationship with fourth ventricle epiermoid tumor.

Material and Methods: The authors describe the case of a fourteen years old girl with intermittent peripheral facial nerve palsy related to fourth ventricle epidermoid tumor and review literature.

Results: A fourteen years old girl was admitted at our institution due to left peripheral facial nerve palsy (House-Brackmann IV/VI) and headache. CT scan and MRI showed quadrigeminal cistern lipoma and IV

ventricle epidermoid tumor. She recovered from facial palsy some days after admission and also from headaches and was discharged. Some weeks after was admitted again with facial nerve palsy and headache so the authors decided to remove the tumor. At surgery there was an intimate relationship between the upper part of the epidermoid tumour and left facial colliculus. The postoperative period was uneventful and there were complete recovery and no more facial palsy episodes. The authors show preoperative images, operative videos in both microscopic and endoscopic views.

Discussion: The authors consider peripheral facial nerve palsy alone as part of facial colliculus syndrome ("half/partial facial colliculus syndrome") associated to some lesions of the fourth ventricle.

Poster track 3.3: Oncology: Spine & Craniofacial

PP085

Intramedullary epidermoid cyst revealing undiagnosed adult diastematomyelia

Alessia Imperato, Sergio Paolini (Pozzilli, Italy)

Introduction: Epidermoid cysts represent less than 1% of all intraspinal tumours. They are slow-growing lesions that occur as a result of inclusion of ectodermal tissue during the closure of the primitive neural tube. Usual location is intradural and extramedullary. We report the case of a woman with intramedullary epidermoid cyst discovered at the level of a previously unknown diastematomyelia.

Case Report: A 53-year-old woman, with a clinical history of mammary cancer treated 2 years before, referred to our Institution complaining lumbar pain with extension to the lateral surface of the right leg. Neurologic examination revealed urinary incontinence and gait instability. Spinal MRI showed a dysraphic lumbar spine with a low lying and bifid cord: at L3 the right hemicord presented a round mass, hyperintense on T1-weighted images, hypointense on T2, with inhomogeneous and intense contrast enhancement.

We performed surgery under neurophysiologic monitoring: after L3 laminectomy and dura mater incision, the right hemicord was found swollen and covered by a small lipoma, tightly adherent to the cord. Posterior myelotomy revealed a cystic mass with a greenish dense content and a solid core was gently separated from neural tissue. A diagnosis of intramedullary epidermoid cyst was made. The postoperative course was uneventful, and the patient's neurologic status improved, with neither urinary nor motor deficit.

Discussion: Diastematomyelia is rare congenital anomaly. Its embryology has been clearly explained by *Pang et al*. The association of diastematomyelia with epidermoid cyst can be expected on the basis of the unified theory of split cord malformations, but only few reports of have been described. Moreover, intramedullary location of epidermoid tumors is very rare. The present case provides further support to the unified theory of embryogenesis for double spinal cord malformations.

PP086

Pilocytic astrocytoma of the conus medullaris: a pediatric case

Charlotte Burford, Wisam Al-Faiadh, Jose P. Lavrador, Bassel Zebian (London, United Kingdom)

Background: Approximately 5% of all paediatric spinal tumours are intramedullary and of these, approximately 10% are found in the conus medullaris. In adults, a histological diagnosis of myxopapillary ependymoma is most common. In the current literature, only three adult cases of pilocytic astrocytoma of the conus medullaris (PACM) are reported. Here we report the first paediatric case of a PACM in 7-year-old female.

Methods: A 7-year-old girl presented to her local hospital with a 6-week history of hip and groin pain, worsening when lying down. Prior to this presentation she was otherwise fit and well. Imaging at the local unit demonstrated a lesion of the conus, which was part cystic, part solid in nature. She was subsequently referred to our unit for further management.

Results: The patient underwent a T11-L2 laminotomy and linear midline durotomy. An expanded conus was seen and a left paramedian myelotomy was made, under ultrasound-guidance, allowing drainage of the cyst. Debulking of the solid component of the tumour was then performed with intraoperative neuromonitoring. Neuropathological examination was consistent with a diagnosis of pilocytic astrocytoma. A dose of 20mg/kg of 5-ALA had been administered pre-operatively but no intraoperative fluorescence was seen. Post-operatively she was neurologically intact and her symptoms resolved.

Conclusion: Pilocytic astrocytoma of the conus meduallaris is a rare lesion, which can present in both adult and paediatric patients. An attempt at debulking is indicated in symptomatic patients with large lesions and can be safely achieved with both ultrasound guidance and intraoperative neuromonitoring after careful examination of high resolution MRI.

PP087

Surgical long-term outcome in a series of intramedullary tumors

Laura Grazia Valentini, Marika Furlanetto, Luisa Chiapparini, Micol Babini, Bianca Pollo, Roberto Cordella, Veronica Saletti (Milan, Italy)

Introduction: Intramedullary tumors are quite rare in the pediatric population, representing less than 1% of CNS lesions

in this age group. First surgical series are reported in the eighties and the role of IntraOperative neurophysiolocial Monitoring (IOM) is crucial to obtain their safe removal.

Patients and Methods: In a 25 years period (1992-2017) 28 children were operated on for Intamedullary Tumors at our Institution; 22 males and 6 females; mean age at surgery was9,8 years, median 11 y, with a range from 1 to 16 years. Two patients were "infants" (< 1 year). Most frequent presenting symptom was back pain (35%), especially along the night, followed by segmentary strength loss; in 3 cases rapidly progressive scoliosis was present before surgery. The diagnosis was performed in every case by whole spine c.e. MRI.All children were operated by a laminotomic approach, except 4, in which just a biopsy was performed due to the massive medullary edema; in these cases, a decompressive duroplasty was associated to a laminectomy.IOM was performed since the end of the ninenties.

Results: Hystopatological exams revealed14 low grade gliomas, 6 high grade (III/IV) gliomas, 2 ependimomas; 2 presented hemangioblastomas (1 with multiple localizations); in the remaining 4 cases a malformative intramedullary tumor was documented, associated with some mild stigmata of dysraphysms just in one child; the malformative lesions displayed a pattern of growth slowly ascending in the centromedullary canal. The diagnosis was delayed in all cases, with a long-time span between first symptom and diagnosis. Brief, long and very long-term outcome was strictly related with the histology, but also to preoperative deterioration level and to type of surgery.

Conclusion: The results will be discussed in detail, focusing on the role of IOM improvements, reoperations, adjunctive treatment and spine stabilization.

PP088

Surgically seeded tumor after dumbbell shape craniopharyngioma surgery

Amets Sagarribay, Dalila Forte, Gonçalo Januário, Marcos Veiga, Miguel Correia, Carla Conceição, Manuela Mafra, Mário Matos (Lisbon, Portugal)

Introduction: Craniopharyngioma metastasis is a rare condition with few reports in literature. Some of previously reported cases divides craniopharyngioma metastasis as primary or secondary to surgical procedures. Dumbbell shaped are also rare subtypes of craniopharyngioma and, in many cases, with such configuration that need to be planned in a staged manner in order to safely removal.

Material and Methods: The authors describe the case of an eight years old boy previously treated to remove a dumbbell shaped craniopharyngioma that showed a new lesion out from its original tumor site but correlated with previous surgical corridor. They describe the case and review literature.

Results: An eight years old boy with a rare dumbbell shaped craniopharyngioma was surgically treated in a staged manner due to tumor configuration as it grew from sella turcica to fill the III ventricle coming between ACoA and chiasm through lamina terminalis. The authors performed a three steps surgery: an anterior transcallosal, right supraorbital/ subfrontal and endoscopic transnasal transsphenoidal approaches, with a gross total removal. Few months after MRI studies showed no tumor relapse but a new lesion at the right lesser sphenoid wing. Differential diagnosis were brain abscess, granuloma and seeded craniopharyngioma. As this lesion grew with time the authors decided to re-operate, performed a supraorbital/ subfrontal approach to remove it and confirm a seeded craniopharyngioma with no relation with previous tumor site. After 4 years of follow up no relapse or new lesion could be seen in repeated MRI studies. The authors show pre- and postoperative MRI, surgical planning, surgical videos and histological images.

Discussion: There are few reports of craniopharyngioma metastasis or "seeded tumor". As the authors show it might not be related with malignant behavior but as cellular seeding during surgical approaches that may keep in mind when imaging follow-up studies show new lesions in a previous surgical corridor.

PP089

Pediatric craniopharyngioma early recurrence after gross total removal and treatment with γ-knife. Report of 2 cases

Christos Chamilos, Panayiotis Kokkalis, Spyros Sgouros (Athens, Greece)

Objective: The optimal management of recurrent pediatric craniopharyngioma remains controversial. We report 2 children with craniopharyngioma which recurred very early after initial excision and were treated successfully with γ-knife.

Patients: Patient 1: a 7-year old girl presented with loss of visual acuity, weight loss and vomiting. She had big solid 3rd ventricular tumor. She underwent gross total resection with 2-step surgery (transcallosal and pterional). A small remnant on the right optic foramen grew 2mm in 4 months. She underwent γ-knife irradiation to the remnant. Follow-up scan at 12 months showed no further growth. Patient 2: a 5-year old boy presented with headaches, vomiting and papilledema. He had a suprasellar lesion with s large cystic component. A gross total resection was achieved via pterional approach. At the 3-month follow-up scan a cystic recurrence was shown for which he underwent a second operation and γ-knife adjuvant irradiation at the sellar region. Follow-up scan at 21 months showed no further recurrence. Both patients experienced no significant post irradiation side effects.

Conclusion: Recurrence of craniopharyngioma may occur even after apparent radical excision. Prompt management of residual or recurring disease by GKS, repeat surgery, or a combination of both is usually successful in controlling

further tumor growth. GKS is a relatively safe modality for the treatment of recurrent or residual craniopharyngiomas, and it is associated with improved tumor control and reduced in-field recurrence rates.

PP090

Craniopharyngiomas: surgical results

Liudmyla Verbova, Mykhailo Shamaev, Andryi Shaversky, Tetiana Malysheva, Dmytro Tsyurupa (Kyiv, Ukraine)

Background: Craniopharyngiomas account for 6-10% of all intracranial tumors in children and 80% of hypothalamo-hypophyseal tumors. The own clinical material is investigated.
Material and Methods: Between 1993-2016 ears 157 patients were treated at the Institute: adults - 62 (average age - 33), children - 95 (average age - 8); female: male = 1:1.
Tumor localization: endo-suprasellar - 14 children/6 adults, suprasellar - extraventricular - 39/24, extra-intraventricular - 40/27, intraventricular - 2/5. 157 patients underwent next procedures: total resection - 26/12, subtotal resection - 27/21, partial resection - 31/19, reservoir Ommaya - 6/5, shunting operation- 6/7.
Results: Mortality after surgery - 14 children (3 patients with suprasellar-extraventricular tumors; 10 patients - extra-intraventricular tumors, 1 patient - intraventricular tumor); 13 adults (4 patients with suprasellar-extraventricular tumors; 9 patients - extra-intraventricular tumors).
The median follow-up for children was 5 years, for adults - 3 years: 31 patients had recurrences,8 patients died of the disease.
Conclusion: Craniopharyngiomas are hypothalamo-hypophyseal tumors. The striving for radical surgery is not always justifiable, especially in adults because of hypothalamus damage. The treatment choice must be individualized according the patient condition, cyst component, hydrocephalus, direction of tumor growth.

Poster track 3.4: Oncology: Ventricles & CSF

PP091

CSF disturbances after transcallosal resection

Christian Dorfer, Gregor Kasprian, Julia Vendl, Thomas Czech (Vienna, Austria)

Objective: To evaluate the frequency and potential predicting factors for CSF disturbances in a consecutive series of patients operated through a transcallosal approach.
Methods: We retrospectively reviewed the medical charts of all patients operated at our institution via a transcallosal approach between January 2002 and December 2016. The following radiological parameters were assessed on pre- and multiple postoperative MR/CT images: ventricular size, as well as subdural space and interhemispheric fissure width. Statistical analyses were performed to identify clinical and radiological predicting factors for shunt dependent hydrocephalus within three months after surgery.
Results: We identified 74 patients (female n=40, male n=34; median age 17.6yrs, range from 6 mos - 76 yrs). An EVD was placed preoperatively in 18 patients (24.3%). Histology revealed a colloid cyst in 15 (20.3%), pilocytic astrocytoma in 14 (18.9%), diffuse astrocytoma WHO II in 5 (6.8%), diffuse astrocytoma WHO III+IV in 7 (9.5%), subependymal giant cell astrocytoma in 5 (6.8%), craniopharyngioma in 5 (6.8%) and other in 23 (31.0%). After a median time of 24 days (range from 10 days to 3 mos) since transcallosal resection implantation of a VP-shunt was necessary in 9 patients (12.2%) and a subduro-peritoneal shunt in 5 (6.6%). The mean age at shunt implantation was 14.3 yrs. The necessity for postoperative shunt implantation was significantly associated with younger age.
Conclusion: The incidence of early shunt dependent hydrocephalus after transcallosal resection is high (18.8%), especially in younger children. The reason for shunt dependency is multifactorial.

PP092

Efficacy of ETV in the treatment of hydrocephalus after removal of posterior fossa tumor in children

Marina Pitsika, Panayiotis Kokkalis, Georgios Papaevangelou, Christos Chamilos, Spyros Sgouros (Athens, Greece)

Objective: To analyze the efficacy of Endoscopic Third Ventriculostomy (ETV) in the management of hydrocephalus after posterior fossa tumour surgery in children.
Materials and Methods: A retrospective review was performed of 55 posterior fossa tumour resections performed in 44 patients during 2008-2017 (11 operations were for recurrences). All patients had tumour excision as the first operation, regardless of ventricular size. Management of hydrocephalus was performed postoperatively in the presence of symptoms of intracranial hypertension in the presence of large ventricles, regardless of pseudomeningocele. Another 12 patients who had 28 operations in the same period were excluded as they had a shunt insertion elsewhere first, before tumour resection.
Results: Hydrocephalus was present before tumour resection in 29 patients (53%). After tumour excision, 15 patients in total (27%) required permanent treatment for hydrocephalus. Of these, 4 patients did not have hydrocephalus at presentation, but developed after tumour resection (2 midline and 2 had laterally placed tumours) and 3 patients were shunted late after tumour excision (2, 5 and 10 months respectively). In 10 patients ETV was performed as first procedure; 8 of those required subsequent ventriculo-peritoneal shunt. Hence the success rate of ETV was 20%. All patients that ETV failed had pseudomeningocele

postoperatively. Of those who had ETV that failed, 2 patients had wound CSF leak postoperatively.
Conclusion: ETV when employed in the management of hydrocephalus after removal of a posterior fossa tumour in children has a poor success rate. Presence of pseudomeningocele is a negative predisposing factor for ETV success.

PP093

Neuroendoscopic approach to midline lesions causing hydrocephalus in pediatric population: a consecutive series

Afonso Antonio Montalvo, Angela Moreno Gutiérrez, Juan Vicente Darriba Alles, Oscar Lucas Gil de Sagredo del Corral, Marc Valera Melé, Vicente Casitas Hernando, Roberto García Leal (Madrid, Spain)

Introduction: Endoscopic third ventriculostomy (ETV) is nowadays an effective treatment for obstructive hydrocephalus and allows, in some cases, to diagnose the cause by an endoscopic tumor biopsy (ETB) with a single-trajectory approach. However, little has been reported about this technique when used in pediatric population with midline lesions causing obstructive hydrocephalus. We report our experience with this technique performed in children.
Methods: Since January 2000, we reviewed 10 consecutive cases of hydrocephalus secondary to midline lesions. Medical records, neuroimaging studies, operative notes and histological findings were analyzed.
Results: There were 9 female and 1 male patients with a mean age of 10,4 years (range 6-14 years). Headache, vomits and diplopia were the main symptoms. At the last follow up two patients had undergone repeated ETV and one of them finally required a shunt placement, giving an overall success rate of 90% (9 of 10 cases) with a mean follow-up of 48 months (range 2 -179 months). ETB was successfully performed in 6 patients (60%) resulting in 3 low grade tectal gliomas, 2 pineal tumors and 1 ependymal cyst. In the rest four cases radiological diagnosis was low grade tectal glioma and no further attempt was made to obtain tissue for diagnosis. There was no major morbidity and mortality related to the procedure.
Conclusions: Neuroendoscopic approach is a safe and effective procedure for treating hydrocephalus associated with midline lesions in children and can provide histological diagnosis at the same time by a single-trajectory approach.

PP094

Ventricular endoscopy in treatment of obstructive hydrocephalus and tumor verification of the 3rd ventricle region

Volodymyr Mykhalyuk, Ludmyla Verbova, Pavlo Plavsky, Andrij Svyst, Artur Mumliev, Nikolai Guk, Dmytro Tsyurupa (Kyiv, Ukraine)

Background: Tumors in the III-rd ventricular region have a common anatomical location, yet they can represent a diverse spectrum of histopathology, radiological characteristics and prognosis. These tumors are accompanied by hydrocephalus in majority of cases.
Methods: Between of 2013-2017 years twelve patients aged 4 months - 17 years (7,9 ± 3,5), with tumors in the III-rd ventricle were treated in our institution, five (41,6%) boys and seven girls (58,4%). All patients were operated by precoronal approach through lateral ventricle. Surgical procedures included: Endoscopic cysts aspiration/Ommaya reservoir implantation – 6; ETV – 6; Endoscopic septostomy – 3; Endoscopic biopsy – 9. Endoscopic intervention was done as primary in nine cases and secondary in three cases - (one case, after the removal of pilocytic astrocytoma and two cases after VP shunt). The goals of endoscopic intervention were: reduction of hydrocephalus and if possible histological verification of the tumor.
Results: In all cases, at least one goal been achieved. Regression of hydrocephalus achieved in 11 cases (91,6%). Endoscopic biopsy followed by histological verification were achieved in 7 cases (58,3%). After endoscopic procedures two patients required second operation (tumor resection):one patient with craniopharyngioma and compression of chiasm by solid part of the tumor and one patient with a failed biopsy - the tumor expansion in the posterior fossa (in the following verified as germinoma). Cases of pilocytic astrocytoma (2) and craniopharyngioma (1)– were followed up. Two patients (germinomas) was done RT, CRT after surgical treatment required.
Conclusion: Endoscopy is a useful technique for achieving shunt-independent control of obstructive hydrocephalus as well as simultaneously obtaining biopsy of III-rd ventricular tumors. A proper histological verification of these tumors significantly helps in effective management of these lesions as well as in making prognosis.

PP095

A rare case of intraventricular ganglioglioma in a 13 years old adolescent

Izzet Durmusalioglu, Gokhan Canaz, Burak Gunduz, Erhan Emel, Evsen Apaydin, M. Bilge Bilgic (Istanbul, Turkey)

Introduction: Gangliogliomas (GG) are rare, low grade primary central nervous system (CNS) tumours accounting for 0.5-1.7% of all neuroepithelial tumours. Their usual presentation is medically refractory epilepsy during the first 3 decades

of life. Gangliogliomas appears most commonly in the temporal lobe, followed by the frontal, parietal, and occipital lobes, and, less frequently, in the thalamus, cerebellum, brainstem, and spinal cord. We report a case of ganglioglioma in the left lateral ventricle, which is a rare location for this type of tumour.

Case Report: 13 years old female patient admitted our emergency room with 2-3 minutes length generalised tonic-clonic seizure. No seizure anamnesis was present. At the evaluation after the postictal period, neurological examination was totally normal. Computer tomography showed a giant 8x6.5 cm cystic mass. In the MRI, 8x6.5cm cystic mass, originating from left lateral ventricle was revealed (Fig. 1). There was a 2x1.5cm solid part with peripheric enhancement. As preliminary diagnosis choroid plexus papilloma and ependymoma were considered and the operation was planned.

Using left parieto-occipital transsulcal approach, near-total resection was performed. Per-operatively, solid component was seen as adhered to ventricle wall. Samples were sent to Istanbul University, Istanbul Faculty of Medicine, Department of Pathology for examination.

Postop no complication was seen. Patients neurological examination was normal. Postop MRI revealed that near total resection was achieved (Fig. 2). Anti-epileptic medication was composed, and the patient was discharged postoperative 5th day.

Pathological examination revealed Who grade I ganglioglioma (Fig. 3 and 4).

Discussion: Both parenchymal and intraventricular GG have a predilection for males and tend to occur mostly in children and young adults; however, the peak age of occurrence is slightly higher in intraventricular GG. Our patient was a 13 years old girl. The most common presenting symptom of patients with gangliogliomas is seizures, with an incidence of 75-100% according to the literature. GG are most of the time described as partially cystic mass with a solid part.

Currently, total resection is suggested as the most widely recognized approach for treatment. The range of resection is regarded as patient's clinical presentation, tumour localization and tumour size. Survival rates were reported 100-75% with total resection and 75-25% with subtotal resection in literature. We are believing these variations between numbers are highly related to tumour localization and size, in patient basis. Adjuvant therapy is controversial after subtotal resection.

In the latest edition of the WHO classification of tumours of the central nervous system published in 2016 GG were assigned to WHO Grade I.

Conclusion: GG are rarely taken place in intraventricular localization. These tumours should be considered in the differential diagnosis of intraventricular lesions, especially in young patients. Complete surgical resection is the widely accepted treatment of choice.

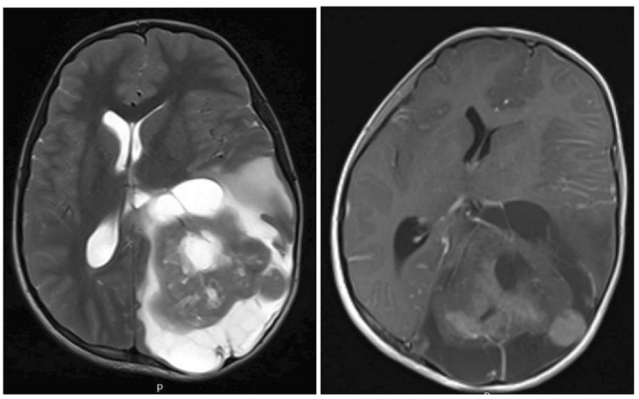

Figure 1

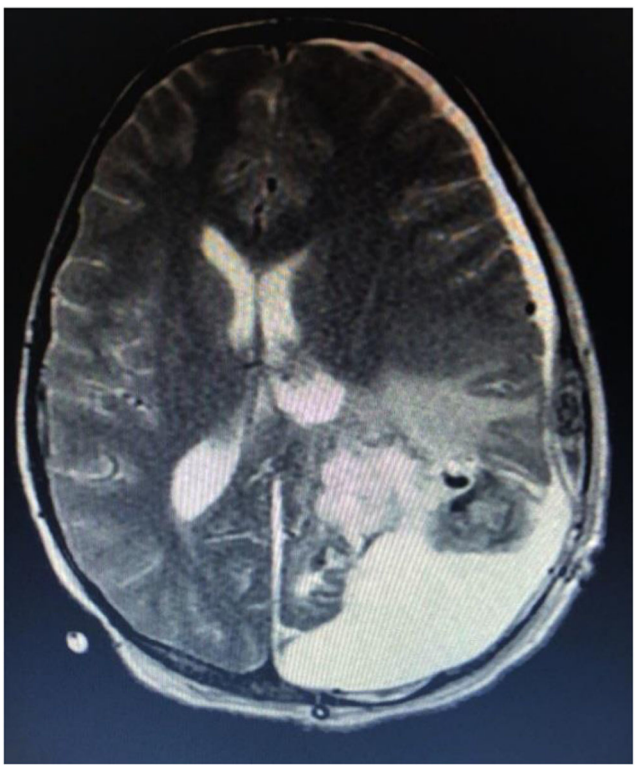

Figure 2

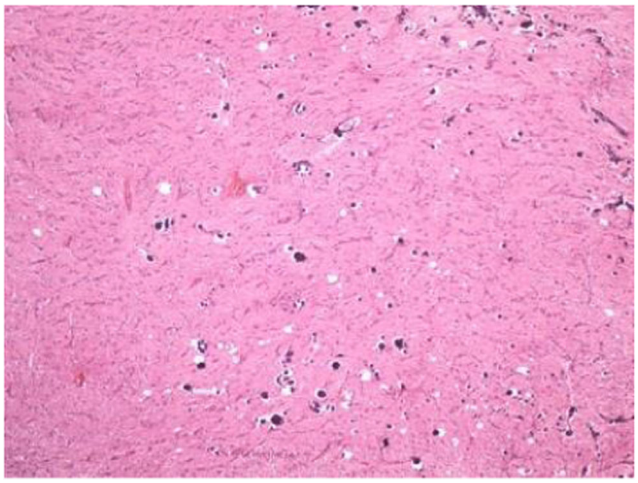

Figure 3: *Low grade neuroepithelial tumor with extensive microcalcifications (X40, H&E)*

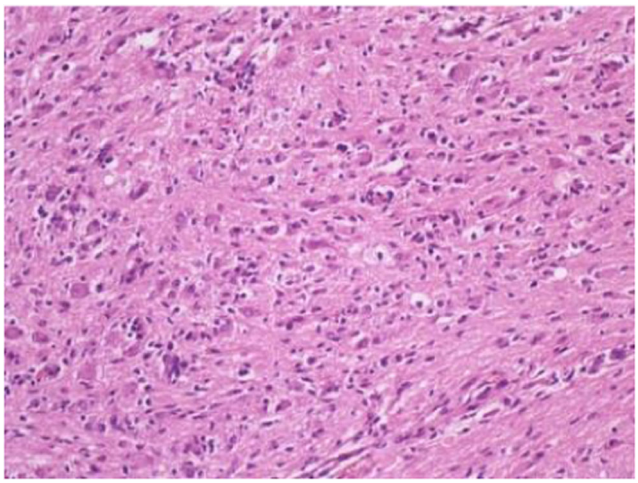

Figure 4: *The dysplastic neuronal component was prominent (x200, H&E)*

PP096

Water tight dural closure in pediatric craniotomies: is it really needed?

Jonathan Roth, Haggai Benvenisti, Shlomi Constantini (Tel-Aviv, Israel)

Introduction: Dural closure (DC) following intracranial procedures is considered crucial to reduce postoperative complications such as pseudomeningoceles (PM), CSF leak, hydrocephalus, infections and more. However, water tight dural closure (WTDC) is often difficult to achieve, and may be time consuming. We describe our experience with various DC techniques, mostly a minimal, non-WTDC, and focus on postoperative complications.

Methods: Data was retrospectively collected. Redo and craniectomy cases were excluded. Collected data included demographics, surgical etiology, sizes of resected pathology and craniotomy, region of pathology, ventricular opening, usage of drains and shunts, DC technique, and complications (focusing on PM, leaks, infections, and hydrocephalus).

Results: 163 cases, aged 3-222 months (90±56) were included. Main surgical indications included tumor resection (114), and epilepsy resections (27). 122 cases were supratentorial. The ventricular system was opened in 69 cases. In 30 cases the dura was left open, in 68 slight dural approximation was done, in 47 dura was closed but not in a WT fashion, and in 18 a WTDC was done. Fibrin glue was used in 22 cases. In 156 cases, a dural substitute was placed over the dural opening (Duragen (131), and Gelfoam (80)). One patient (0.6%) had a CSF leak. At 3 months, 18% had a radiological PM, but only 8.6% were noticed clinically. At 1 year, 9% had a radiological PM, but only 3% were noticed clinically. Overall, 3% needed a tap of a PM, and 16 patients (10%) underwent additional CSF diversion procedures. There were no postoperative infections. Only an infratentorial location was significantly associated with PM.

Conclusions: Non-WTDC following cranial surgery in children was associated with a low rate of clinical significant PM, infections, leaks, and hydrocephalus. Non-WTDC is fast, and spares the need to harvest additional tissues such as galea or muscle, thus minimizing the surgical incision as possible.

Poster track 3.5: Miscellaneous

PP097

Idiopathic intracranial hypertension in children: about a case

Cherkaoui Mandour, Cherif Abad El Asri, Miloudi Gazzaz, Brahim El Mostarchid (Rabat, Morocco)

Idiopathic intracranial hypertension (IIH) has been well described in adults but it is a rare neurological disorder in children. It is characterized by raised intracranial pressure (ICP) in the absence of brain parenchymal lesion, vascular malformations, hydrocephalus, or central nervous system infection.

The diagnosis is usually confirmed by high opening pressure of cerebrospinal fluid (CSF) with exclusion of secondary causes of intracranial hypertension. If not treated properly, it may lead to severe visual dysfunction. We review through a case of a 05-year-old girl the pathogenesis, clinical presentation, diagnostic criteria and management of IIH in children.

PP098

Intra-orbital hydatid cyst of the child: an observation

F. Bouaré, D. Mpando, M. Laghmari, M. Lmejjati, H. Ghannane, S. Ait Benali (Marrakech, Morocco)

Introduction: The hydatid cyst of the orbit is a parasitosis due to the development of the tænia echinoccocus granulosus larvae in the orbital cavity. It is an infection that accounts for 1 to 2% of all hydatidoses. Areas of predilection exist despite prophylactic measures in Africa, Latin America and Asia. The aim of this work is to specify the pathophysiology of the intraorbitallocation of the hydatid cyst and the medico-surgical therapeutic modalities in the literature through a reported case.

Materials and Method: This is a reported case of the service.

Observation – Result: It was a 7-year-old child, who has no particular history, who presented a right exophthalmia, little red, little painful, non-pulsatile progressive appearance for 3 months with episodic tearing and visual fog. Orbital CT showed a non-enhancement cystic lesion in the supero-external angle of the right eye. Orbital MRI showed the same appearance. The hydatid serology was positive. The treatment was surgical. Operative follow-up was marked by palpebral edema and phimosis which declined.

Discussion – Conclusion: The treatment is medico-surgical with antiparasitic and lateral orbitotomy by the coronal or ciliary approach, or fronto-orbital approach.

PP099

CLN2 disease (neuronal ceroid lipofuscinosis type 2): experience in the real world with cerliponase alfa intracerebroventricular enzyme replacement therapy in a public hospital in Cordoba, Argentina

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Introduction: Neuronal ceroid lipofuscinosis (NCLs) are one of the most common autosomal recessive neurodegenerative diseases in childhood (1:100,000 live births), characterized by the lysosomal deposition of autofluorescent material throughout the body particularly affecting the brain and retina. Currently the NCLs are classified into 14 types according to the affected gene. CLN2 disease is related to deficiency of the lysosomal enzyme tripeptidyl peptidase 1 (TPP1). Characterized by epilepsy, cognitive impairment, progressive motor dysfunction and visual deficit, leading to a vegetative state with premature death.

Objective: We present two patients ages 5 and 2 yo diagnosed with CLN2 disease who were started on intracerebroventricular enzyme replacement therapy with cerliponase alfa, a recombinant form of TPP1.

Description: Two patients with the classical late infantile phenotype of CLN2 disease were selected for intracerebroventricular cerliponase alfa therapy. Prior to infusion, successful Rickham reservoir implantation via neuronavigation guidance occurred. A post-surgical brain CT ensured catheter location. Infusions were started using the protocol developed by the hospital service. Using an antiseptic technique, the reservoir was punctured, 2 ml of CSF were removed and sent for analysis. Subsequently, the catheter was connected to the infusion system (flow 2.5 ml/h and total volume 10 ml). The procedure was well tolerated, no adverse events documented, and the patients were discharged on average 24 hrs. post-infusion with plan to repeat every 14 days for life.

Conclusion: Implantation of Rickham reservoir at the right frontal lobe level can be safely performed using the neuronavigation system in patients with CLN2 disease. Intracerebroventricular cerliponase alfa treatment can be performed in our hospital following our local protocol in accordance with the information provided by the supplier of the drug. These represent the first infusions of cerliponase alfa in Latin America demonstrating potential of cerliponase alfa to treat CLN2 patients in the region.

PP100

Spinal epidural abscess after perinatal pneumothorax in an 8-week old baby – A case report

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Objective: We present a case of a massive spinal epidural abscess following early-onset sepsis and pneumothorax.

Methods: An 8-week-old white female presented to the emergency department with a flaccid paraparesis. Parents reported an undulating symptomatology concerning motor deficit. Furthermore, the child did not tolerate prone position anymore. Fever, persisting for three days, was observed 10 days before admission, which showed spontaneous regression. The girl had been born at 41 weeks of gestation, the early postnatal period was complicated by respiratory distress after birth which led to admission on the pediatric ICU, followed by antibiotic therapy with Ampicillin and Tobramycin for 5 days. Additionally, a pneumothorax on the right side was diagnosed, which was treated conservatively.

Magnetic resonance imaging after admission revealed a spinal epidural abscess extending from T1-T9 with maximum expansion from T3-T6, compressing the spinal cord, further osteomyelitis and expansion in dorsal soft tissue was noted. There was a remarkable enhancement along the dorsal soft tissue with connection to the right lung with adjacent atelectasis.

Results: Diagnosis of a spinal abscess with spinal cord compression was made, and the patient underwent urgent neurosurgical intervention. Laminoplasty T3-T5 was performed for

abscess drainage and spinal cord decompression. In the postoperative course the girl recovered quickly with full recovery of the motor deficits. Staphylococcus aureus was grown from pus drained in theatre. The patient was started on a 5-week course of Cefazolin and Clindamycin. Postoperative MRI showed sufficient spinal cord decompression. Baseline immunology tests showed no pathologies. Neurological examination at the time of hospital discharge was entirely normal.
Conclusion: Spinal epidural abscess is rare in newborns and has to be considered as differential diagnosis in children with progressive neurological deficits and fever. Whereas osteomyelitis is a well-recognized complication, spinal abscess with spinal cord compression has rarely been described.

PP101
Multiple brain abscess in a child: two-stages surgical treatment
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Introduction: Brain abscess is a rare and very dangerous condition for children. It may be secondary to sinusitis or otitis media but sometimes it is difficult to find any source for abscess. It is usually solitaire, but it may be also multiple.
Case Report: We reported 4-month old male child who was presented with increased head circumference since birth. The magnetic resonance imaging revealed multiple intracranial abscesses located in the left hemisphere causing midline shift and significant edema. There was no history of infection since birth. The patient underwent 2-stages surgical treatment and all abscesses were evacuated with small burrholes and irrigated with saline solution. The abscess culture revealed methicilline-sensitive staphylococcus aureus and appropriate antibiotherapy was performed. The child was completely improved after the treatment.
Conclusion: Children with multiple brain abscesses should be treated multi-stages if necessary. Appropriate antibiotic regiment is required in order to obtain better clinical outcome.

Poster track 3.6: Hydrocephalus II

PP102
Reducing shunt dependency in post-hemorrhagic ventricular dilatation by improving protocols
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Introduction: Posthemorrhagic ventricular dilatation (PHVD) of infants is usually treated by temporizing measures, e.g. lumbar punctures, ventricular reservoirs, ventriculosubgaleal drains or endoscopic clot-removal, while 50-100% of these patients will eventually receive a permanent ventriculoperitoneal shunt and become shunt dependent. Over a period of 14 years we have tried to reduce shunt dependency and complications of treatment by several different measures. This study is performed to evaluate the result of that undertaking.
Methods: This is a retrospective study of all infants receiving surgical treatment for PHVD between 2004 and 2017. Standard therapy is installing subcutaneous neonatal ventricular reservoirs frontally and repeated reservoir taps (initially twice daily) over a prolonged period of time. In 2008 we changed the location of surgery from the OR to the incubator on the neonatal ICU ward; in 2011 the puncture regime of the reservoir was adapted to reduce the infection rate and in 2013 we converted a procedure previously under general anesthesia into a procedure in local anesthesia.
Results: We have surgically treated 36 infants. Reservoir taps were performed for a period of on average 4 weeks (2-6 weeks) and usually started 2-3 weeks after birth. Overall, 11 infants (31%) became shunt dependent. In the first half of the study period the shunt dependency rate was 63% and in the second half 5% (1 out of 20 patients, p=0,0003). The overall infection rate was 11%, 4 patients (11%) died of comorbidity related to prematurity and 4 new cerebral hematomas were induced by surgery (11%). Two patients required revision of their reservoir because of obstruction, while infections of reservoir were treated conservatively.
Conclusion: A minimalized surgical procedure, a strict puncture regime by specially trained personnel and a prolonged tapping period all contribute to an extreme low rate of shunt dependency without creating arrested hydrocephalus.

PP103
Feasibility of neuroendoscopic lavage prior to shunt implantation in neonates with post-hemorrhagic hydrocephalus
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Objective: Neonates with posthemorrhagic hydrocephalus (PHH) are at high risk for shunt failure, with increased cerebrospinal fluid (CSF) protein being a presumed relevant variable. Neuroendoscopic lavage (NL) has been performed to reduce ventricular blood in premature babies, with positive published results. We have recently adopted this technique and started our learning curve with NL used as adjunct therapy in cases of PHH with indication for permanent CSF diversion, primarily aiming at improving CSF conditions and thus potentially reducing occlusion risk.
Methods: We report results of NL performed at our institution. Clinical, radiological and laboratory data was retrieved retrospectively.
Results: Three neonates born at a mean gestational age of 35 weeks (range 27-40 weeks) were treated with NL at a mean of 20 days after birth (range 7-39 days). All children suffered symptomatic PHH with a mean preoperative frontal to

occipital horn ratio of 0.58 (range 0.42-0.69). External ventricular drain (N=1) and ventricular reservoir (N=1) were inserted prior to NL. Reason for NL was active hydrocephalus with indication for permanent CSF diversion, but persistent mean CSF protein of 317mg/dl (range 130-600mg/dl). NL of both lateral ventricles and third ventricle was performed via a unilateral approach and septostomy. No complications occurred and NL resulted in reduction of mean CSF protein to 60mg/dl (range 15-104mg/dl) and radiological reduction of blood clot volume. VPS insertion was performed directly after NL (N=2) and with latency of 20 days (N=1). No VPS occlusion was observed within 30 days, although one infection occurred.
Conclusion: NL reduces CSF protein and radiologically visible blood clot volume. No complications or early VPS failures were encountered in this series, but certainly larger cohorts are required to assess the value of NL with regard to improving VPS outcome or even avoiding VPS in PHH.

PP104

Endoscopic lavage of massive chronic subdural hematoma – Adoption of a technique from ventricular neuroendoscopy

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Introduction: Subdural fluid collections are frequently encountered in young children after non-accidental injury. In a subset of patients, these collections progress in size and ultimately require permanent drainage, which is commonly achieved with subduro-peritoneal shunts. However, excessive protein and cellular contents in the fluid are risk factors of shunt failure. Here, we describe the adoption of an endoscopic lavage technique established for ventricular endoscopy with the aim of improving fluid condition prior to shunting.
Methods: We present a case of subdural fluid collections secondary to non-accidental injury, where permanent shunting was required, but could not be performed due to excessive protein and cellular levels in the subdural fluid despite conventional burr hole drainage.
Results: A two-month old male infant presented with bulging and tense fontanelle, reduced level of consciousness, bradycardia and significant macrocephaly. Computed tomography demonstrated massive bilateral low attenuation subdural fluid collections, reaching a diameter of 4.5 cm. Emergency burr hole washout and insertion of subdural drains was performed. However, despite prolonged drainage over 10 days, mean protein level remained at 544 mg/dl (range 506-600 mg/dl) and mean erythrocyte count at 6,493/μl (range 3,555-9,356/μl). The child required continuous drainage to avoid clinical deterioration due to raised intracranial pressure, but fluid condition was considered incompatible with permanent subduro-peritoneal shunting. We therefore performed an endoscopic subdural lavage with careful evacuation of residual blood deposits. No complications were encountered. Postoperatively, mean protein level was 292 mg/dl (range 244-320 mg/dl) and mean erythrocyte count was 101/μl (range 29-154/μl).
Conclusion: Endoscopic lavage could be safely performed in a case of massive subdural low attenuation fluid collections, where conventional burr hole drainage failed to improve protein and cellular contents as a prerequisite for successful permanent shunting. We conclude that adoption of this technique can be helpful in selected cases as a salvage procedure.

PP105

The role of Choroid Plexus Cauterization (CPC) before shunt revision

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Introduction: For patients who repeat Ventriculo-peritoneal shunt (VPS) malfunction due to distal side troubles, we should find other distal space for CSF absorption. Usually Ventriculo-arterial shunt (VAS) or Venticulo-pleural shunt (VPLS) are selected as a second choice. We report a case with repeated infectious trouble of VPS and hesitated to select VAS.
Case Report: The patient was a 12-year-old girl. Her gestational age was 26w and her body weight was 958g at birth. She developed hydrocephalus caused by intraventricular hemorrhage. Repeated shunt infectious troubles finally created severe adhesion of the peritoneal cavity which was diagnosed by pediatric surgeons using laparoscope. VAS was thought to be inappropriate because of multiple history of infection. We decided to select VPLS. However, her CSF output volume from external ventricular drainage was over 250ml/day. The volume seemed too much for daily pleural absorption. CPC was done as preparation procedure before VPLS to reduce CSF volume. After CPC, her CSF output volume reduced to 80ml/day. We successfully performed VPLS without any trouble. She did not develop any respiratory failure due to pleural effusion during two years after surgery. VPLS is an alternative CSF diversion for VPS. Pleural effusion becomes a problem especially for infant. However, there is no information about tolerable volume of pleural effusion. Thoracic surgeons usually remove chest drainage tube less than 100ml/day in adults. Therefore, we tried to reduce CSF volume at least 100ml/day by CPC.
Discussion: This case suggests that CPC could provide a good clinical condition for starting VPLS.

PP106

Abdominal cerebrospinal fluid pseudocysts related to ventriculo-peritoneal shunt

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Introduction: Abdominal cerebrospinal fluid (CSF) pseudocysts related to ventriculo-peritoneal shunts are rare. We present a small series of such patients with emphasis on laparoscopic management.

Methods: We present 4 patients (2 boys) aged between 3 and 9 years old, with ventriculo-peritoneal shunt and an abdominal CSF pseudocyst. Factors predisposing to the development of abdominal pseudocysts were analyzed.

Results: All patients presented with abdominal pain and distention and shunt malfunction. A history of prior abdominal surgery other than shunt revision was found in 3 patients and of necrotizing enterocolitis in 2 patients. A history of prior shunt infection was present in 1 patient. The diagnosis of abdominal CSF pseudocyst was made by ultrasound in all cases. Abdominal MR scan was also performed to assess the anatomic details of the cyst. In all cases laparoscopy was utilized to marsipulise a portion of the pseudocyst, remove the shunt catheter tip from the residual pseudocyst, and reposition it in the peritoneal cavity. The patients experienced no complications from the procedure, and there has been no recurrence of the pseudocyst after 6 months of follow-up.

Conclusions: Abdominal CSF pseudocyst is an uncommon complication of V-P shunts. Ultrasound is useful in diagnosis and follow up. Cyst marsipulisation and repositioning of the shunt tip can be safely achieved with minimally invasive laparoscopic techniques obviating the need for open surgery, which may compound abdominal adhesions with time.