

# 44th Annual Meeting of International Society for Pediatric Neurosurgery, Kobe, Japan, Oct 23-27, 2016

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## PLATFORM PRESENTATIONS

Monday, 24 October 2016  
09:00 – 09:32

### Platform presentations 1: Fetal Diagnosis and Management

#### PF-001

**Special topic: Fetal diagnosis and management**

**Zika virus fetal encephalitis outbreak: what is the role of the pediatric neurosurgeon in the response strategy?**

Carlos Eduardo Jucá<sup>1</sup>, André Luiz Pessoa<sup>3</sup>, Erlane Ribeiro<sup>2</sup>, Rafaela Menezes<sup>4</sup>, Thayse Lopes<sup>4</sup>, Vivian Mota<sup>1</sup>, Renata Jucá<sup>5</sup>

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**OBJECTIVE:**Zika virus (ZV) fetal encephalitis is an emergent disease provoking an outbreak in Brazil. It is considered a global emergency by the World Health Organization. Aim of this study is discuss the role of the pediatric neurosurgeon in the response strategy of this new threatening neurological disease.

**MATERIAL-METHODS:**Report of epidemiological, clinical and radiological features of ZV encephalitis outbreak in Brazil. Report of illustrative cases imposing differential diagnosis with pediatric neurosurgical diseases. Establishment of the place of the neurosurgeon in the outbreak task force.

**RESULTS:**From october 2015 to march 2016, 6776 suspected cases of zika virus associated encephalitis (zvae) were notified, 944 confirmed and the others under investigation, with 208 perinatal deaths. Clinical presentation is linked to severe microcephaly. Computer tomography (CT) show brain atrophy and spread calcifications. Association of zika infection and microcephaly was confirmed by polymerase chain reaction in 130 cases. Our pediatric neurosurgery unit received two cases mimicking neurosurgical diseases. Case 1: a 28 years old pregnant woman at the 26th week was referred due to fetal ultrasound suggesting hydrocephalus. Magnetic resonance showed ventricular enlargement as result of brain atrophy. At birth, HC was 31cm, morphology and CT presented the features of zvae. Case 2: A 22 days old male newborn was referred due to suspicion of craniostenosis. The anterior fontanell was closed. HC was 30cm. Clinical aspect and CT

scan were compatible with zvae. Both mothers reported symptoms of ZV fever in the first months of pregnancy. Assesment and follow up by the neurorehabilitation team demonstrated severe neurological disability with upper motor neuron syndrome.

**CONCLUSIONS:**Zvae is a threatening and rapidly spreading outbreak with high potential for mortality and morbidity. The pediatric neurosurgeon must take part in the management task force helping to establish differential diagnosis and to rapidly separate true cases from those related to neurosurgical diseases.

**Keywords:** Zika virus, encephalitis, microcephaly, hidrocephalus.

#### PF-002

**Special topic: Fetal diagnosis and management**

**First 50 fetal in-utero microsurgical myelomeningocele repairs: critical comparison of neurosurgical and maternal outcomes to the MOMS trial**

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**OBJECTIVE:**The Management of Myelomeningocele Study (MOMS) trial results showed improved hydrocephalus and motor outcomes in prenatal repair group compared to the postnatal group. We report our initial 50 case series of fetal in-utero myelomeningocele (MMC) repair experience and outcomes at the Saint Louis Fetal Care Institute. The learning curve was also observed and assessed.

**MATERIAL-METHODS:**We strictly followed the maternal/fetal inclusion and exclusion criteria used by the MOMS trial. A multidisciplinary fetal MMC repair team performed 50 repairs between May 2011 and March 2016. We prospectively followed maternal, fetal and neonatal data. The data was compiled to evaluate the learning curve to meet the MOMS results held as standard of care.

**RESULTS:**All MMC defects underwent successful in-utero repair. Average GA at time of MMC repair was 24+3/7 weeks. Average GA at time of delivery was 34+4/7 weeks, with 50% of pregnancies reached 37 weeks. Operative time of microsurgical MMC repair after hysterotomy averaged 43 minutes. Two perinatal mortalities 2/50(4%) were due to complications of prematurity. CSF diversion rate of all surviving children via VP shunt or ETV was 20/48(41.6%), MOMS trial was 40%. ETV success rate was 8/17(47%) with an average follow up 20 months. When statistically compared to MOMS trial, maternal outcomes were either equivalent or improved for all categories except pulmonary edema, chorioamnionitis and preterm labor. Fetal outcomes were also improved or equivalent. Surgical skin to skin time significantly increased from the first 10 cases to the next 37 cases (p<.001) which then led to significantly increase in days between repair and delivery (p<0.05) and increase in infant birth weight (p<0.05). Ten cases appear to be the threshold for our center to consistency in equivalent results.

**CONCLUSIONS:**Fetal,maternal and neurosurgical outcome equivalency was shown between Saint Louis series and the MOMS trial. Ten cases appear to be the point at which this equivalency is reached. ETV deserves a closer look and may show promising results in the setting of improved hindbrain herniation.

**Keywords:** fetal in-utero surgery, myelomeningocele, spina bifida, MOMS trial

### PF-003

#### Special topic: Fetal diagnosis and management

##### Lessons learned after 180 fetal surgeries for myelomeningocele

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**OBJECTIVE:**To analyze clinically and radiologically 180 MMC fetuses intrauterine operated and compared with 180 patients who underwent surgery after birth by the same surgeon and the same medical service.

**MATERIAL-METHODS:**We analyzed clinically and radiologically 160 MMC fetuses intrauterine operated and compared with 160 patients who underwent surgery after birth by the same surgeon and the same medical service. The evolutionary segment ranged from six months to four years. The following aspects were studied: 1. cephalic perimeter; 2. the presence and Chiari degree; 3. posterior fossa volume; 4. clivus-supraocciput angle variation; 6. valve placement; 7. relationship between functional and anatomic level of the lesion.

**RESULTS:**The operative mortality in fetal surgery was higher than the post-natal, 4.3% and 1.25% respectively. All fetuses who died occurred in the first cases operated, and in the last three years, we had no more mortality. We did not have maternal mortality. In 84.3% of postnatal surgical cases require shunts while in pre-natal only 4.3% needed. The intrauterine operated patients there was a trend macrocephaly associated with non-hypertensive ventriculomegaly. The severity of the Chiari type 2 was lower in intrauterine surgical cases. The volume of the posterior fossa was doubled in patients operated in the fetal period, and the clivus-supraocciput angle was higher in this group of patients. Motor level was better in fetal group than in the postnatal.

**CONCLUSIONS:**Fetal surgery for myelomeningocele correction promotes an increase in the volume of the posterior fossa, turning an obstructive

hydrocephalus in a communicating or non-obstructive hydrocephalus with decreasing degrees of Chiari type 2 and improves the motor level of these patients. A learning curve is critical in reducing fetal mortality and a trained multidisciplinary team is important for obtaining good results.

**Keywords:** fetal myelomeningocele, intrauterine repair, open surgery, hydrocephalus, Chiari 2, fetal surgery

### PF-004

#### Special topic: Fetal diagnosis and management

##### First year neurologic outcome after fetal surgery

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**OBJECTIVE:**Since the results of the Management of Myelomeningocele Study were published, maternal-fetal surgery for the in utero treatment of spina bifida has become accepted as a standard of care alternative. There are two kinds of approach for intrauterine myelomeningocele repair, which are fetoscopic approach and open surgery. Our aim is to present first year neurological evaluation and urodynamic results of four patients who were undergone percutaneous fetoscopic patch closure for myelomeningocele.

**MATERIAL-METHODS:**Four patients were evaluated according to neurological examination, severity of hydrocephalus, urodynamic study results and MRI findings at the end of their first year of life.

**RESULTS:**There was no need for shunting in any of four patients. Over active detrusor and detrusor sphincter dyssynergia were occurred in all patients but clean transient catheterization was needed in only one patient according to urodynamic study parameters. In one patient, clonus was occurred in the sixth month of life and tethered cord releasing operation was performed.

**CONCLUSIONS:**Fetal surgery is associated with spinal segmental neuroprotection and decrease of Chiari malformation and shunt depended hydrocephalus. Further research is needed to better understand the pathophysiology of MMC, the ideal timing and technique of repair, and the long-term impact of in-utero intervention.

**Keywords:** First Year, Fetoscopic Surgery, Neurologic Outcome

Monday, 24 October 2016

11:00 – 12:20

## Platform presentations 2: Spine and Craniofacial

### PF-005

#### Other

##### New classification of spinal lipomas based on embryonic stage

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**OBJECTIVE:**New classification of spinal lipomas based on embryonic stage is proposed.

Spinal lipomas have been regarded as a representational pathology of spina bifida and caused by failed primary neurulation. However, some showed no spina bifida and it is not unusual to involve distal to conus medullares which is formed by secondary neurulation. Most of currently applied classifications use MRI sagittal view.

**MATERIAL-METHODS:**In the new classification, lipomas were classified based on the embryonic stage as follows:

Type 1: pure primary neurulation failure

Type 2: stretching from primary to secondary neurulation failure

Type 3: secondary neurulation failure (early phase)

Type 4: secondary neurulation failure (late phase)

**RESULTS:**Typically, type 1 lipomas correspond with dorsal and part of transitional lipomas with spina bifida. Type 2 lipomas involves more complex form of lipoma such as chaotic type lipomas and lipomyelomeningoceles. Caudal type and filum lipomas correspond with type 3 and 4 irrespectively. Anorectal, urogenital and sacral anomalies which develop at the same time with secondary neurulation associated with type 2 to 4 lipomas, more on type 3 and 4. From surgical view point, type 1 is a good candidate for radical resection of the lipoma. Fusion line is not clear in type 2 with possible inclusion of nerve root inside the ventrally extended lipoma, thus surgery is tough and only partial resection can be possible. Type 3 lipomas fuse with conus at the caudal end and untethering is not complicated. True “spina bifida” can be absent in this group. Surgery for type 4 filum lipoma is straightforward.

**CONCLUSIONS:**In conclusion, this new classification based on embryonic stage seems to provide more practical information in terms of the clinical feature and surgical difficulty.

**Keywords:** spinal lipoma, classification, primary neurulation, secondary neurulation

## PF-006

### Spine

#### Histopathology of the filum terminale in children with and without tethered cord syndrome with attention to the elastic tissue within the filum

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**OBJECTIVE:**To compare histologically transected fila from pediatric patients with tethered cord syndrome (TCS), with and without a low conus with controls, focusing on collagenous and elastic tissue.

**MATERIAL-METHODS:**30 fila from patients with TCS, including 5 where minimal cautery was used prior to filum section, were compared with fila from 27 pediatric cadavers without TCS (controls). Sections of fila were stained with H&E, Masson trichrome and Verhoeff von Gieson elastic stains and 5 also with Gordon and Sweet’s reticulin stain.

**RESULTS:**Fila from controls showed loose fibrovascular connective tissue (FCT) with thin and evenly dispersed elastic fibers (EF). Reticulin fibers (RF) were seen in blood vessel walls and nerve twigs. Fat was identified microscopically in 2 fila. All fila from patients with TCS had dense FCT. The EF were in normal numbers in 13, and focally or diffusely decreased in 12. All 25 patients where the fila were cauterized during resection had thick and coiled EF. Coiling was not seen when minimal cautery was applied. RF were seen in blood vessel walls and nerve twigs. Fat was identified in 19 patients. Findings were similar, whether the conus termination was normal or low.

**CONCLUSIONS:**The fila of all patients with TCS, whether or not the conus was low, showed abnormal FCT. EF numbers were decreased in 48% of patients, however there were thick EF in all patients. Coiling of EF, initially thought to be an abnormality in patients, is probably caused by cautery.

**Keywords:** Occult tethered cord, fibrous connective tissue, reticulin, elastic tissue, cautery

## PF-007

### Spine

#### More than traction? New insights into the pathophysiology of “tethered cord syndrome”

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**OBJECTIVE:**Fixation of intrathecal nervous structures in the spinal canal, caused by various forms of spinal dysraphism or previous intradural surgery, may lead to neurologic signs of “tethered cord syndrome” (TCS). Traction and resulting hypoxia of affected neural structures are considered main driving forces for developing clinical relevant TCS. The aim of this study was to identify further specific mediators of molecular cascades that may contribute to TCS.

**MATERIAL-METHODS:**Ethical approval was obtained. Specimens from 16 TCS-surgeries were investigated; pathology included previous intradural surgery for dysraphism and thickened filum. Clinical characteristics were obtained retrospectively and included neurological status, bowel/bladder-dysfunction, contractures/spacities of lower extremities and back pain. Normal adult spinal cords (sc) (n=4) served as controls. Sections were immunostained with neuroglial, neural crest, mesenchymal and epithelial markers (GFAP, neurofilament (NF 200kD), NeuN, synaptophysin, CNPase, Vimentin). Immunohistochemistry and real-time RT-PCR for inflammatory cytokines (interleukin-1beta (IL-1b) and its receptor IL-1R1, tumor necrosis factor-alpha (TNF-a) and its receptor (TNF-R1), and hypoxia-induced cytokines (erythropoietin (EPO) and its receptor (EPOR)), plus hypoxia inducible factors (HIF-1a/-2a) were performed. Data were analyzed qualitatively and semi-quantitatively. Cellular expression patterns were confirmed by multiple-fluorescence-labeling.

**RESULTS:**All specimen exhibited significant gliosis with strong GFAP- and Vimentin-immunolabeling. IL-1b and TNF-a plus their receptors became detectable in cellular composites of intrathecal nervous structures on significantly elevated level (p<0,05 and p<0,001 vs. control). These cytokines were co-stained with NeuN (TNFa/TNF-R1) and GFAP (IL-1b/IL-1R1). EpoR/Epo was significantly elevated in 85%/92% cases, and co-stained with NeuN and HIF-1a/-2a, which were also induced in TCS specimen.

**CONCLUSIONS:**In addition to hypoxic stress, pro-inflammatory and hypoxia-induced cytokines probably influence symptomatology and outcome of TCS. As specific molecular composites of the underlying pathophysiology they provide potential new targets for experimental therapeutic approaches.

**Keywords:** tethered cord syndrome, spinal dysraphism, neuropathology

## PF-008

### Spine

#### The management of craniocervical junction abnormalities in skeletal dysplasia: experience of a single institute

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**OBJECTIVE:**Craniocervical junction abnormalities are quite common in skeletal dysplasia, which could result in severe neurological complication, and should be appropriately managed by pediatric neurosurgeon. In this study, we discuss the feasible management for this condition based on our clinical experience.

**MATERIAL-METHODS:** Retrospective analyses were performed on 15 cases of craniocervical junction abnormalities in skeletal dysplasia, including nine cases of achondroplasia, two cases of metatropic dysplasia, and one case of congenital spondyloepiphyseal dysplasia, osteogenesis imperfecta, and mucopolysaccharidosis I.

**RESULTS:** Among 15 cases, nine were treated surgically. Eight cases underwent decompression of most stenotic part. Foramen magnum decompression was completed on six cases of achondroplasia who afterwards gained ambulatory ability except one. Upper cervical decompression was done on two cases of metatropic dysplasia who maintained upper extremity function as suits their own condition. One case of osteogenesis imperfecta underwent cerebrospinal fluid diversion only, and her cervical lesion was conservatively managed without any neurological compromise. One case of achondroplasia who presented foramen magnum stenosis and syringomyelia will be operated soon. One case of mucopolysaccharidosis I presented with tetraparesis after falling and severe upper cervical stenosis was detected on MRI, however, he was conservatively managed because of legal guardian issues. The rest of the five cases were carefully monitored based on static and dynamic evaluation of the lesions.

**CONCLUSIONS:** Our results suggest that decompressive surgery for neurocompression is a considerable option even in skeletal dysplastic popularity. However careful monitoring is recommended for latent stenotic lesions if no instability is demonstrated.

**Keywords:** craniocervical junction abnormality, skeletal dysplasia, neurocompression

#### PF-009

##### Other

**A lack of efficacy of an intradural somatic-to-autonomic nerve anastomosis for bladder control in children with spinal dysraphism: results of a prospective, randomized, double-blind study**

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**OBJECTIVE:** Xiao et al. and other investigators have reported high success rates (70–85%) after an intradural somatic-to-autonomic (e.g., L-5 to S3–4) nerve transfer procedure in children with spinal dysraphism. These authors have suggested this “skin-CNS-bladder” reflex arc allows spontaneous, controlled voiding in children with neurogenic bladder dysfunction. The study reported here was designed to more rigorously investigate the effectiveness of the procedure.

**MATERIAL-METHODS:** Children with neurogenic bladder dysfunction related to myelomeningocele or lipomyelomeningocele, who were scheduled for spinal cord detethering (DT), were enrolled in the study. Patients were randomized between two groups at the time of surgery: one group underwent only spinal cord detethering (DT) and the other underwent the Xiao procedure (XP) in addition to spinal cord detethering (DT+X). Patients, families, and study investigators, all of whom were blinded to the assigned study group, analyzed the patients' neurologic and urologic outcome at regular intervals during the 3-year follow-up.

**RESULTS:** Twenty patients were enrolled in the study: 10 underwent only DT and the other 10 underwent DT+X. Patients who underwent the XP were no more likely to be able to void, to control their urination, to achieve continence, or to have a demonstrable urodynamic bladder contractions in response to cutaneous stimulation than patients who only underwent spinal cord DT. Performance of the XP during DT was more likely to lead to improvements in quality of life, bladder capacity and bladder over-activity, all of which may be attributable to the effect of sacral rhizotomy instead of bladder reinnervation.

**CONCLUSIONS:** The performance of the Xiao Procedure did not result in an improvement in neurologic or urologic outcome. The results of this RCT are in agreement with recently published, similarly disappointing results in spinal cord injury. This study raises doubts about the clinical applicability of this procedure in humans before confirmatory animal studies are completed.

**Keywords:** tethered spinal cord, spinal dysraphism, Xiao procedure, nerve transfer, voiding, incontinence

#### PF-010

##### Other

**Craniovertebral instability: a description of a new syndrome**

Harold ReKate, Timothy White, Orseola Arapi  
The Chiari Institute

**OBJECTIVE:** Patients with reducible distortion of the anterior brainstem by the odontoid process may present with symptomatic Chiari I malformation. The most common underlying cause of this condition is joint hypermobility in the context of inheritable abnormalities of connective tissue (Ehlers Danlos syndrome type 3 and Marfan's syndrome).

Here, we posit a new syndrome of interest to neurosurgeons occurring in patients with EDS that consists of cranio-cervical instability, dysautonomia and chronic fatigue. Recognition of these overlapping symptoms in patients who fail to improve following Chiari decompression or with characteristic clinical and radiographic features prior to intervention may benefit from occipito-cervical stabilization.

**MATERIAL-METHODS:** A retrospective chart review was conducted that included patients seen by the principal investigator between January 2012 to March 2016. All included patients met criteria for EDS 3, radiographic evidence of cranio-cervical instability (CCI), dysautonomia, and symptoms of chronic fatigue.

**RESULTS:** A total of 26 patients were identified. The average Beighton Score was 6/9. All patients had a previous diagnosis of POTS, 100% of patient complained of chronic fatigue, 77% of patients had a previous diagnosis of IBS, and 69% of patients complained of a confusional state as well as memory problems. All cases had radiographic evidence of instability at the CVJ.

**CONCLUSIONS:** We propose that the changes occurring with the CCJ in patients with EDS leads to anterior brainstem compression and is associated with both dysautonomia as well as chronic fatigue. We are currently pursuing this relationship with a prospective outcomes study to assess cranio-cervical fusion for correction of the CCI and its impact on patient symptomatology. Patients undergoing decompression for Chiari I malformation that also have evidence of dysautonomia should be followed postoperatively for progression of cranio-cervical instability.

**Keywords:** hindbrain herniation, Postural orthostatic tachycardia syndrome, headache

#### PF-011

##### Spine

**Significance of increased initial pB-C2 distance on radiographic outcomes in Chiari type 1 malformation**

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**OBJECTIVE:** The presence of ventral brainstem compression (VBSC) in the setting of Chiari malformation type 1 (CM1) is well-known, but management

remains controversial. One measure of VBSC is the pB-C2 measurement proposed by Grabb and Oakes, who hypothesized that a pB-C2 of greater than 9mm requires occipital cervical fusion. In our institution we do not routinely instrument these patients, so we evaluated the outcome of patients with initial pB-C2 greater than 9mm undergoing craniocervical decompression without fusion.

**MATERIAL-METHODS:**All patients who underwent initial craniocervical decompression for CM1 by the senior authors between 2003 and 2006 were included. The pB-C2 was measured on the pre-operative and last post-operative MRI. The change in mean pre- and post-operative pB-C2 of patients with a pB-C2 greater than or less than 9mm were then compared. Patients were excluded if they did not have appropriate imaging available.

**RESULTS:**A total of 54 patients met the inclusion criteria, of which 8 had an initial pB-C2 greater than 9mm. Median follow up was 36 months. Average initial pB-C2 was 6.1mm (5.5mm in <9mm group vs 9.4mm in >9mm group). Average pB-C2 at last follow up was also 6.1mm (5.8mm vs 8mm). The change in pB-C2 in the <9mm group was +0.3mm (95% CI -0.1mm to +0.6mm) versus -1.2mm (95% CI -2.6mm to +0.3mm) in the >9mm group. There was no significant difference in syrinx resolution between the groups.

**CONCLUSIONS:**The question of whether to perform a fusion on patients with VBSC in CM1 remains controversial. In our series, we demonstrate that patients with an initial pB-C2 >9mm did not have any significant progression in their pB-C2, and in most cases had an improvement in the severity of their VBSC. This suggests that patients with more severe VBSC can be safely managed initially with decompression alone, although further study is warranted.

**Keywords:** Chiari malformation, occipital-cervical fusion, ventral brainstem compression, pB-C2, craniocervical decompression

#### PF-012

##### Spine

#### Growth and alignment following subaxial cervical spine instrumentation and fusion in pediatric patients: preliminary results from a multi-institutional study of the Pediatric Craniocervical Society

Richard Anderson<sup>1</sup>, Hannah Goldstein<sup>1</sup>, Doug Brockmeyer<sup>2</sup>, David Pincus<sup>3</sup>, Gerald Tuite<sup>4</sup>, Andrew Jea<sup>5</sup>, Todd Hankinson<sup>6</sup>, Nick Wetjen<sup>7</sup>, Cormac Maher<sup>8</sup>, Sean Lew<sup>9</sup>, Joshua Payhs<sup>11</sup>, Jonathan Martin, And Others On Behalf Of The Pediatric Craniocervical Society<sup>10</sup>

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**OBJECTIVE:**Literature has suggested that the cervical spine reaches biomechanical maturity by approximately 9 years of age. However, more recent work has shown that there is continued anatomic growth up to 18 years. The

purpose of this study was to determine the effects of rigid instrumentation and fusion on the growing subaxial spine.

**MATERIAL-METHODS:**A multi-institutional retrospective chart review was conducted for children < 16 years of age who underwent instrumentation and fusion of the subaxial spine at more than 20 participating centers. All patients had > 1 year of follow-up with radiographically confirmed fusion. Preoperative, immediate postoperative, and most recent follow-up radiographs and/or CT scans were evaluated to assess changes in spinal growth and alignment.

**RESULTS:**In this preliminary analysis of 40 patients, 55% were male with an average age of 11.8 years. Etiologies consisted primarily of trauma (55%), congenital (20%), or neoplasia (20%). The leading presenting symptom was pain. On average, 3 levels were fused, most often using a posterior approach. All patients experienced improvement or resolution of symptoms at an average of 3 years follow-up. In children < 10 years, there was an average of 2.1mm of vertical subaxial cervical spine growth/year in the instrumented population, compared to a normal growth rate of 2.2–2.6mm/year. In children > 10, there was an average of 2.6mm/year of growth in the instrumented population, compared to the normal average growth rate of 1.5–3.1mm/year. Adjacent level unintended fusion or progressive deformity was rare.

**CONCLUSIONS:**Most children undergoing subaxial cervical spine fusion with rigid instrumentation have some continued growth within the fused levels. This appears to be independent of age at the time of fusion, specific levels, or number of levels fused. Adjacent segment problems are uncommon in the early postoperative years, but continued close long-term follow-up is needed.

**Keywords:** spine, instrumentation, fusion, growth,

#### PF-013

##### Spine

#### Safety and efficacy of posterior hemivertebrectomy in congenital scoliosis in children

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**OBJECTIVE:**To study the efficacy and safety of Posterior Hemivertebrectomy and short segment pedicle/ screw fixation as a one stage procedure in Congenital Scoliosis.

**MATERIAL-METHODS:**20 patients with congenital hemi vertebra and progressive scoliosis were treated with this procedure under intra-operative neuromonitoring. Posterior exposure and Transpedicular Fixation of the adjoining vertebrae was done. The hemivertebra was approached from the convex side and transpedicular decancellation followed by excision was done. Controlled compression at the convexity was done for curve correction.

**RESULTS:** There were 9 males and 11 female patients with mean age of 7.53 (1.4 – 24) yrs at presentation and mean follow up was 24 months (6 – 55 months). Average number of hemivertebra removed was 1.4 (1- 3) and mean pre op and post op coronal cob angle was 53.29 degrees and 27.24 degrees respectively. Mean pre op and post op sagittal cobb angle was 38.18 degrees and 17.43 degrees respectively Mean coronal and sagittal cob correction percentage achieved was 48% and 46.9% respectively. Mean loss of coronal and sagittal correction at final follow up was 6.43 % and 5.54 % respectively. There was superficial wound healing problem in one patient, which was managed with antibiotics and dressings. There were no cases of implant failure or post op neurological deterioration except in one patient which improved partially at 3 months.

**CONCLUSIONS:**Posterior Hemivertebrectomy is a less invasive and effective procedure for congenital scoliosis, and one which can be safely performed in children with deformity provided it is done with neurophysiological monitoring.

**Keywords:** hemivertebrectomy, congenital scoliosis

## PF-014

## Spine

**Intraoperative neurophysiological monitoring in brainstem and complex spinal surgery: optional or mandatory?**

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**OBJECTIVE:** Resection of brainstem and spinal tumours and surgical correction of craniocervical instability are particularly challenging in children. We reviewed the role of intraoperative neurophysiological monitoring (IONM) in preventing neurological sequelae.

**MATERIAL-METHODS:** Twenty procedures in 19 children aged 4–15 (mean 7.5) years were monitored using combined somatosensory evoked potentials (SSEPs) and transcranial motor evoked potentials (TcMEPs), under total intravenous anaesthesia (TIVA), with brain-stem auditory evoked potentials (BAEPs) and free-running EMG (frEMG) where indicated.

Evoked potentials (EPs) were recorded at pre-operative awake baseline, post-anaesthetic induction, after prone positioning, and continuously (SSEPs) or intermittently (TcMEPs) throughout surgery.

**RESULTS:** Nine underwent excision of brainstem/spinal lesions, eight occipito-cervical fusion for instability/rotatory subluxation, three detethering/lumbar laminoplasty. Pre-operative lower limb SSEPs were absent in one child with progressive paraparesis and delayed in another with lower limb hyper-reflexia; two had torticollis.

Significant IONM changes occurred in 14 procedures (70%). Changes in TcMEP amplitude or frEMG with screw insertion in six cranio-cervical fusions resolved after appropriate response (pause in surgery, increasing blood pressure, removing minimal pressure or screw adjustment). The seventh was monitored without recordable lower limb baseline responses, with no adverse sequelae at cervical level, but sustained ischaemic injury to the upper thoracic cord undetected by exclusively upper limb IONM.

Amplitude drop or loss of unilateral TcMEPs occurred during late tumour removal in 4 cases where SSEPs remained stable. TcMEPs returned incompletely in one anterior T5 tumour with associated post-operative transient ipsilateral leg weakness. During a brainstem tumour resection, persistent SSEP amplitude drop with elevated BP lead to restriction of further excision. In two cord detetherings, MEPs/frEMG changed transiently without sequelae.

**CONCLUSIONS:** Brainstem/spinal surgery in our practice was made safer using combined IONM. It detected a higher number of brainstem/cord events than anaesthetic vital monitoring, guiding surgery to avoid permanent sequelae and should be considered mandatory.

**Keywords:** craniocervical instability, occipitocervical fixation, intraoperative neuromonitoring, high cervical spinal tumours, pre-operative somatosensory evoked potentials

Monday, 24 October 2016  
14:15 – 15:20

## Platform Presentations 3: Craniofacial | Epilepsy & Functional

## PF-015

## Epilepsy and functional

**Epilepsy surgery in pediatric intractable epilepsy with destructive encephalopathy**

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**OBJECTIVE:** The aim of the current study is to review the clinical features, surgery outcomes and parental satisfaction of children with destructive encephalopathy who underwent epilepsy surgery due to medically intractable seizures.

**MATERIAL-METHODS:** 48 patients who underwent epilepsy surgery from October 2003 to August 2011 at Severance Children's Hospital have been reviewed. The survey was conducted for functional outcomes and parental satisfaction at least 1 year after the surgery.

**RESULTS:** Epileptic encephalopathy including Lennox-Gastaut syndrome and infantile spasms was more prevalent than symptomatic focal epilepsy. Hypoxic ischemic injury accounted for most of the underlying etiology of the destructive encephalopathy, followed by central nervous system infection and head trauma. 27 patients (56.3%) underwent resective surgery and 21 patients (43.7%) underwent palliative surgery. 16 patients (33.3%) achieved seizure free and 27 parents (87.5%) reported satisfaction with the outcome of their children's epilepsy surgery. In addition, 14 parents (77.8%) whose children were not seizure free reported satisfaction with their children's improvement in cognitive and behavior issues.

**CONCLUSIONS:** Epilepsy surgery in destructive encephalopathy was effective for controlling seizures. Parents reported satisfaction not only with the surgical outcomes, but also with improvement of cognitive and behavior issues.

**Keywords:** Intractable epilepsy, Destructive encephalopathy, Epilepsy surgery

## PF-016

## Epilepsy and functional

**Callosotomy for drop events and peri-insular hemispherotomy following neonatal strokes are highly effective single stage operations in pediatric epilepsy surgery**

Jeffrey P Blount<sup>1</sup>, Brandon G Rocque<sup>1</sup>, Curtis J Rozzelle<sup>1</sup>, Hyunmi Kim<sup>2</sup>, Monisha Goyal<sup>2</sup>, Rani Singh<sup>2</sup>, Pongkiat Kankirawatana<sup>2</sup>

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**OBJECTIVE:** Despite increasing evidence that surgery is highly effective and safe for children with medically resistant epilepsy (MRE) there are still limited numbers of programs offering epilepsy surgery for children. This is particularly true in developing countries and is largely due to prohibitive costs of invasive monitoring. As such there is a demand for clear demonstration of effective single stage operations which can offer high effectiveness and safety and have candidacy determined by clinical, imaging and EEG means.

**MATERIAL-METHODS:** A 10 year experience at an academic referral center was reviewed. Cohorts of single stage procedures were identified and studied for effectiveness and safety.

**RESULTS:** Between January 2004 and January 2014 over 700 Epilepsy Surgery procedures were performed at our center. These included 41 hemispherectomies (PIH) and 50 corpus callosotomies. Among PIH procedures there were 28 performed in cases of MRE arising from a prior cerebral infarct. 4 children required a new shunt to manage hydrocephalus but there was no other long term unexpected morbidity. The vast majority (25/28) were

rendered seizure free and there was no mortality in this part of the hemispherectomy cohort. Of the 50 patients undergoing corpus callosotomy 38 suffered primarily drop seizures. This sub-group of the larger cohort showed excellent response to corpus callosotomy with all patients showing an immediate elimination in drop events that persisted for varying lengths of time.

**CONCLUSIONS:**This large retrospective survey identifies two distinct cohorts of single stage procedures that showed high effectiveness and high safety. Suitable candidates for these procedures can be identified with clinical, CT or MRI imaging and basic EEG. As such these may represent suitable and optimal initial procedures for Pediatric Surgical Epilepsy programs in environments of constrained resources.

**Keywords:** hemispherectomy, corpus callosotomy, epilepsy surgery, developing countries

## PF-017

### Epilepsy and functional

#### Epilepsy associated glioneuronal tumours in children: is it time for a more aggressive management strategy?

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**OBJECTIVE:**Glioneuronal tumours (GNT) are composed of a mixture of neuronal and glial elements and are a recognized cause of pediatric epilepsy. In cases presenting to our institution with seizures, standard management has comprised an initial period of radiological surveillance and anti-epileptic medication with surgery reserved for those with refractory seizures or radiological progression. We aim to describe the natural history of patients presenting in this manner and report the surgical results in order to inform management strategy. **MATERIAL-METHODS:**The electronic clinical document database at Great Ormond Street Hospital for Children was interrogated to find patients presenting with seizures and either a presumed radiological diagnosis of GNT or a confirmed histological diagnosis in those who had undergone neurosurgical resection.

**RESULTS:**141 patients treated between 1995 and 2015 were identified (91 male, 60 female). Median age at seizure onset was 3 years. 120 patients (79.5%) underwent surgery with the indication being refractory epilepsy in 111 (92.5%), and tumor progression in 9 (7.5%). Median duration from seizure onset to surgery was 4.13 years. Histopathological review identified dysembryoplastic neuroepithelial tumor in 77 (64%) and ganglioglioma in 43 (36%). 5 patients (4.4%) suffered surgical adverse events (2 episodes of temporary blurring of vision, 1 empyema, 1 subdural hematoma requiring evacuation, and 1 subgaleal collection. No permanent neurological deficits were recorded. 74 patients (80%) were seizure free (Engel Class I) at last follow up (median 2 years post surgery). Radiological complete resection was associated with higher rates of seizure freedom ( $p=0.026$ ).

**CONCLUSIONS:**The vast majority of children presenting with epilepsy and presumed glioneuronal tumors eventually undergo surgery. Neurosurgical resection is safe and highly effective at controlling seizures. Earlier surgical intervention may be associated with improved cognitive and seizure outcomes. Where considered safe, surgical resection should be offered to this patient group before current criteria for medical refractoriness are met.

**Keywords:** Pediatric epilepsy surgery, glioneuronal tumor

## PF-018

### Epilepsy and functional

#### Predicting successful antiepileptic drug withdrawal in children after epilepsy surgery

Michal Tichy<sup>1</sup>, Josef Dvorak<sup>1</sup>, Alena Jahodova<sup>2</sup>, Martin Kudr<sup>2</sup>, Vladimír Komarek<sup>2</sup>, Pavel Krsek<sup>2</sup>

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<sup>2</sup>Department of Pediatric Neurology, Charles University, 2nd Faculty of Medicine, Motol University Hospital, Prague, Czech Republic

**OBJECTIVE:**To assess variables influencing successful antiepileptic drug (AED) discontinuation after pediatric excisional epilepsy surgery.

**MATERIAL-METHODS:**Data of 103 children who underwent excisional epilepsy surgery at Motol Epilepsy Centre between 2002 and 2011 were retrospectively analyzed. All patients were followed up for more than two postoperative years. We statistically compared subjects who became seizure-free and drug-free for at least 1 year with children experiencing seizure recurrence during tapering-down medications.

**RESULTS:**AEDs were postsurgically preserved in 27, reduced in 52 and completely withdrawn in 24 children. Seizure recurrence experienced 23/27(85%) subjects with preserved AEDs, 13/52(25%) patients with reduced AEDs and 4/24(16,6%) children after AED discontinuation. Seizure-free drug-free patients ( $n=20$ ) significantly differed from subjects experiencing seizures during or after AED discontinuation ( $n=17$ ): age at seizure onset (7.7 vs. 3.3 years), seizure frequency (30% vs. 76,5% patients with daily seizures), mental disability (20% vs. 53% mentally-disabled subjects), need of long-term intracranial EEG study (10% vs. 35% cases), localization of resections (95% vs. 29% temporal lobe surgeries), extent of surgeries (0% vs. 41% of multilobar resections), incidence of postoperative EEG spikes (present in 25% vs. 65% of cases) as well as etiology (prevailing hippocampal sclerosis and benign tumors vs. cortical dysplasia).

**CONCLUSIONS:**Successful AED withdrawal was predicted by later seizure onset, less frequent seizures, normal intelligence, temporal lesion other than cortical dysplasia, need of less extensive resection without long-term intracranial EEG study. Assessing completeness of resections at the time of surgery does not reliably predict patient's chances to become postoperatively seizure-free and drug-free.

**Keywords:** Epilepsy surgery, seizure -free, drug- free, discontinuation of medication

## PF-019

### Neuro-oncology

#### Initial experience with endoscopic ultrasonic aspirator in pure neuroendoscopic removal of intraventricular tumors

Giuseppe Cinalli, Imperato Alessia, Spennato Pietro, Di Martino Giuliana, Nicosia Giancarlo, Ruggiero Claudio, Mironi Giuseppe

Department of Pediatric Neurosurgery, Santobono-Pausilipon Children's Hospital

**OBJECTIVE:**Neuroendoscopic excision of intraventricular tumors is difficult and time consuming because of lack of an effective decompression system that can be used through the working channel of the endoscope. The authors report the utilisation of endoscopic ultrasonic aspirator in the resection of intraventricular tumours.

**MATERIAL-METHODS:**Nine pediatric patients (6 M, 3 F) aged between 1 and 15 years old were operated by a purely endoscopic approach using an endoscopic ultrasonic aspirator. Two patients presented intraventricular metastases by high grade tumors (medulloblastoma, AT/RT), 2 Subependymal giant

cell astrocytomas, 2 patients low grade intra/paraventricular tumors and 3 patients presented suprasellar tumors (2 craniopharyngiomas and 1 optic pathway glioma). Hydrocephalus was present in 3 cases. In all cases endoscopic trajectory and ventricular access were guided by neuronavigation. All the patients, but one, were operated through a precoronal approach. In one case the patient was operated through a posterior parietal approach. Endoscopic technique was based on endoscopic visualization of the tumor, ventricular washing to dilate the ventricles and to control bleeding, biopsy of the tumor and aspiration of the tumor with the ultrasonic aspiration. Bleeding was controlled with irrigation, monopolar coagulation and thulium LASER.

**RESULTS:**In 6 cases the resection was total or near total (more than 90% of the lesion). In three cases the resection was partial. Histological evaluation of the collected material (withdrawn with biopsy forceps and aspirated with ultrasonic aspirator) was diagnostic in all cases. The length of operations ranged between 60 and 120 minutes. There were not complications. All the procedures were performed by an expert endoscopic surgeon.

**CONCLUSIONS:**Ultrasonic aspiration is a highly effective tumor decompression system that can be effectively used in a purely endoscopic approach to intraventricular lesions.

**Keywords:** Neuroendoscopy, tuberous sclerosis, intraventricular tumor, medulloblastoma, biopsy, ultrasonic aspirator

#### PF-020

##### Craniofacial

**Preoperative complete cognitive evaluation in children operated on for single suture craniosynostoses: early recognition and rehabilitation of selective impaired functions contribute to an improvement of long term neurodevelopmental outcome**

Gianpiero Tamburrini<sup>1</sup>, Daniela Chieffo<sup>2</sup>, Federica Moriconi<sup>2</sup>, Irene Bernardini<sup>2</sup>, Luca Massimi<sup>1</sup>, Paolo Frassanito<sup>1</sup>, Massimo Caldarelli<sup>1</sup>

<sup>1</sup>Pediatric Neurosurgery, Institute of Neurosurgery, Catholic University Medical School, Rome, Italy

<sup>2</sup>Pediatric Neurology Unit, Catholic University Medical School, Rome, Italy

**OBJECTIVE:**The goal of our study was to provide the opportunity to reliably assess longitudinally neurocognitive functions in children operated on for monosutural craniosynostosis and to evaluate the possibility of an early neurorehabilitation programme to prevent possible neurocognitive disorders.

**MATERIAL-METHODS:**Formal neurocognitive and neurolinguistic investigations were carried out. Neurocognitive investigations consisted of Griffith Mental Developmental Scale (GMDS), the Wechsler preschool children Intelligence Scale-III (WIPPSI III), Various aspects of neuropsychological function, i.e. visual attention (LEITER Scale), and verbal memory, were investigated by means of the Nepsy II. Neurolinguistic evaluation was performed by phonological and lexical examination (naming and comprehension-TFL). The Beery test was used to evaluate visualperceptual skills.

**RESULTS:**Between January 2008 and December 2015 223 children affected by single suture craniosynostoses have been evaluated and treated for a single suture craniosynostosis at the Catholic University Medical School, in Rome, Italy. 78 of them have completed a selective neuro cognitive evaluation in four longitudinal time periods

At 6 months Global IQ was in the normal range in 68 children (87%) and borderline in 10 (13%); selective neurocognitive evaluation showed visual attention deficits and motor hypercinesia in 23/30 (77%) children affected by scaphocephaly, 18/30 (60%) children affected by anterior plagiocephaly and 7/18 (39%) children affected by trigonocephaly (Group A). Moreover 18/30 children with scaphocephaly (60%), 12/30 children affected by anterior plagiocephaly (40%) and 10/18 children affected by trigonocephaly (55%) showed expressive language disorders (Group B). A selective rehabilitation programme was started.

At a mean follow-up of 6.4 yrs, global IQ impairment improved in all but 4 children. Attention and language deficit showed a progressive and longitudinal improvement. Reading and writing impairments persisted in a minority of them. **CONCLUSIONS:**In conclusion a longitudinal preoperative and postoperative neurodevelopmental evaluation programme might improve the long term cognitive outcome of children affected by single suture craniosynostoses.

**Keywords:** Single suture craniosynostosis, Neurocognitive function, Rehabilitation

#### PF-021

##### Craniofacial

**Intraoperative administration of ε-aminocaproic acid is associated with reduced blood loss in children undergoing craniofacial reconstructive surgery: a retrospective analysis**

Srijaya Reddy<sup>1</sup>, Taylor Mann<sup>2</sup>, Heather Gordish Dressman<sup>2</sup>, John Myseros<sup>1</sup>, Suresh Magge<sup>1</sup>, Chima Oluigbo<sup>1</sup>, Richard Levy<sup>2</sup>, Robert Keating<sup>1</sup>

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<sup>2</sup>Department of Anesthesia, Children's National Medical Center, Washington, DC USA

**OBJECTIVE:**Historically, surgical correction of craniosynostosis has resulted in significant blood loss often requiring transfusion. While ε-aminocaproic acid (EACA) has previously been shown to reduce intraop hemorrhage in children undergoing craniofacial surgery, previous efforts have involved variable dosing of EACA. The aim of this study was to determine if using targeted dosing of EACA was associated with decreased hemorrhage in children undergoing craniofacial surgery

**MATERIAL-METHODS:**Children undergoing calvarial vault reconstruction surgery at Children's National Medical Center, Washington DC (07/13 -12/15) were retrospectively evaluated. Patients receiving EACA intraoperatively (100 mg/kg loading dose, 40 mg/kg/hr continuous infusion) were compared to those not receiving EACA. Demographic data and perioperative variables were assessed. Primary outcome measures:intraoperative calculated blood loss, RBC transfusion volume, blood donor exposures, and postoperative surgical drain output; secondary outcome measures:operative time, number of sutures repaired, ICU/hospital LOS, intraoperative crystalloid/colloid administration, perioperative hematocrit and coagulation times. Normality of all outcomes was determined, means were compared using student's t-test ( $p < 0.05$  significant).

**RESULTS:**44 patients (4m-8yrs) were included in the study; 18 received EACA and 26 did not. Both the EACA and non-EACA groups were analyzed for confounding characteristics and surgical variables and no significant differences between the two groups were found. The EACA group had significantly lower calculated blood loss ( $37 \pm 18$  vs.  $63 \pm 45$  mL/kg,  $P = 0.004$ ) reduced RBC transfusion requirements ( $14 \pm 12$  vs.  $30 \pm 24$  mL/kg,  $P < 0.001$ ) than non-EACA group. There was no significant difference in postoperative surgical drain output nor morbidity between the two groups

**CONCLUSIONS:**EACA, administered at the recommended dosing regimen, was associated with significantly reduced intraoperative hemorrhage in children undergoing craniofacial reconstructive surgery. Thus, EACA should be considered as an integral component of any blood conservation strategy in young children undergoing craniofacial surgery. A prospective, randomized controlled trial would be valuable in search of the Holy Grail of transfusion-less craniofacial surgery.

**Keywords:** craniofacial surgery, amicar, reduced bleeding

## PF-022

## Craniofacial

**Neuroradiological management and surgical decision making for separation of craniopagus twins**

James Tait Goodrich<sup>1</sup>, Douglas Cochrane<sup>2</sup>, Oren Tepper<sup>3</sup>, Ahmed Al Ferayan<sup>4</sup>, Richard Hayward<sup>5</sup>, Riyadh Alokai<sup>6</sup>, Jack Farinhas<sup>7</sup>

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<sup>7</sup>Department of Radiology (Neuroradiology), Montefiore Medical Center, Bronx, New York USA

**OBJECTIVE:**We will review a series of seven craniopagus twins and discuss the uniqueness of the various vascular connections and then point out which cases have a more likely chance of reduced morbidity and mortality with a staged surgical separation.

**MATERIAL-METHODS:**Our centers have been involved in the surgical management and diagnostic treatment of 12 sets of twins. An additional six sets of twins did not have surgery due to death, religious or for ethical reasons. This extensive experience allows now a more accurate evaluation for those children that have suitable vascularity and an anatomical interface for separation. The anatomical conjoining, degree of vascular sharing and the angulations can have a prognostic significance on the outcome. Recent addition of sophisticated 3-D modeling/3-D printing has also had an important impact on surgical planning. We will review the unique characteristics of several craniopagus cases along with findings that can significantly influence the decision to operate.

**RESULTS:**In our various cases we have been able to accomplish successful separations with no mortality and lower risk of morbidity based on previous series. Each craniopagus twin set has a unique degree of conjoining and the preoperative assessment needs to focus on the vascular connections and the brain interface. With careful and thorough radiological studies these interfaces and anatomical connections can be assessed and the decision then made if a surgical correction can be undertaken with reduced morbidity and mortality.

**CONCLUSIONS:**With the staged surgical separation techniques the incidence of mortality has been significantly reduced along with morbidity. With our reviewed of now eighteen cases of craniopagus twins it has become clear that there still remain a subpopulation of craniopagus twins that are too complex in their vascular connections and conjoined brains and as a result the risk of a separation exceeds the benefits for the children.

**Keywords:** conjoined twins, craniofacial surgery, Siamese twins, craniopagus twins, conjoined twins

Monday, 24 October 2016  
16:00 – 16:30

## Special Symposium 1: Epilepsy & Functional: Selective Dorsal Rhizotomy

## PF-023

## Other

### Outcomes of selective dorsal rhizotomy for children with spastic cerebral palsy using multi-level laminoplasty versus single-level laminectomy

Sui To Wong, Jason Mk Ho, Kwong Yui Yam

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**OBJECTIVE:**Selective dorsal rhizotomy (SDR) can be done with multi-level laminoplasty (MLL) or single-level laminectomy (SLL). At our institute, we changed the operative approach from MLL to SLL in 2006, thus created 2 groups of SDR patients. We reviewed our prospectively collected data to study the outcomes of these 2 groups of patients.

**MATERIAL-METHODS:**From 2003 to 2010, fifty-five children with spastic cerebral palsy underwent SDR at our hospital. MLL was used in 33 patients between June 2003 and October 2006 (Group MLL); SLL was used in 22 patients between November 2006 and August 2010 (Group SLL). All patients went through the same pre-operative and post-operative physiotherapy protocols. Post-operative complications were documented. All patients were evaluated pre-operatively and post-operatively in multiple clinical domains, using Gross Motor Function Classification System (GMFCS), modified Ashworth scale (mAS), range of movement lower limb joints (ROM), gait analysis (gait pattern, walking speed, oxygen consumption, stride length), hip and spine x-rays, and urodynamic studies. Statistical analysis was performed with SPSS version 22.

**RESULTS:**Mean age at operation was 8.4 in Group MLL, 7.0 in Group SLL. Preoperative distributions on GMFCS and mAS in both groups were similar. The median length of follow-up was 11 years in Group MLL, 7.5 years in Group SLL. Muscle tone was improved in the majority in both groups: 88% in Group MLL, 94% in Group SLL. No statistical significant difference was found between the 2 groups in post-SDR mAS, ROM, gait analysis, and rates of hip subluxation and spinal deformity. In patients with urinary symptoms before SDR, 60% resolved in Group MLL, and 50% resolved in Group SLL ( $p=0.691$ ).

**CONCLUSIONS:**Both SDR techniques can alleviate problems of spastic cerebral palsy with same efficacy and low complication rates.

**Keywords:** selective dorsal rhizotomy

## PF-024

## Epilepsy and functional

### Long term impact of selective dorsal rhizotomy on the quality of life of ambulant children with diplegic cerebral palsy

Marina Pitsika<sup>1</sup>, Mary Cramp<sup>2</sup>, Jenny Smith<sup>1</sup>, Balazs Markia<sup>1</sup>, Guy Atherton<sup>3</sup>, Anna Clarke<sup>3</sup>, Ian K Pople<sup>1</sup>, Richard J Edwards<sup>1</sup>

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<sup>3</sup>Department of Paediatric Orthopaedics, Bristol Royal Hospital for Children, Bristol, United Kingdom

**OBJECTIVE:**Selective dorsal rhizotomy (SDR) is known to improve functional outcome in diplegic cerebral palsy (CP), but its impact on quality of life has not been established. The aim of this study was to evaluate the impact of SDR on the quality of life of children with diplegic CP

**MATERIAL-METHODS:**A single-centre prospective study of children (female  $n=24$  (35%), male  $n=43$  (65%), mean age 6.5 years (range 3-14)) with diplegic CP that underwent SDR between 2011 and 2015. Children were assessed pre-operatively, and at 6, 12 and 24- months post-operatively. Demographic data were recorded, quality of life was assessed using the CP-QoL questionnaire for caregivers and a statistical analysis of the outcome scores (Modified Ashworth Score for spasticity-MAS, Gross Motor Function Classification System level-GMFCS, Gross Motor Function Measure-GMFM-66) was performed

**RESULTS:**67 caregivers completed a 12-month follow up and of these 37 completed a 24-month follow-up. GMFSC was level 2 in  $n=17$  and level

3 in n=50, mean GMFM-66 pre-surgery was 57.1 (range 44.8–81.9) and improved to 61.4 and 62.9 at 12 and 24 months respectively ( $p<0.0001$ ). There was statistically significant improvement in 5 out of the 7 domains of quality of life (Participation, Access to social services, Pain & Physical Health, Function, Emotional Well-Being- $p<0.01$ ) and non-significant improvements in the remaining two domains (Family Health and Social Well-being/acceptance), at 12 months that remained significant at 2 years follow-up. There was significant correlation with improvements in pain and spasticity scores ( $p=0.04$ ), as well as change in pain and participation ( $p=0.02$ ) and emotional well-being ( $p=0.01$ ). Pre-operative GMFCS and the improvement in function (GMFM) did not correlate with improvements in CP-QoL.

**CONCLUSIONS:** In ambulant children with diplegic CP, SDR can offer a significant and lasting improvement in quality of life, primarily due to the impact SDR has on reducing lower limb spasticity and improving of pain

**Keywords:** Selective Dorsal Rhizotomy, Spastic Diplegia

## PF-025

### Epilepsy and functional

#### Selective dorsal rhizotomy in patients with severe spasticity with GMFCS 4-5: results and lessons learnt

Liana Adani Beni<sup>1</sup>, Eli Ashkenazi<sup>2</sup>, Hila Ben Pazi<sup>3</sup>, Ron Lamdan<sup>4</sup>

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<sup>4</sup>Hadassah Medical Center, Jerusalem, Israel

**OBJECTIVE:** Selective Dorsal Rhizotomy (SDR) is a well-accepted treatment in children with cerebral palsy (CP) suffering from Spastic Diplegia but less for patients with quadriplegia and limited ambulation. SDR is not commonly used in children with GMFCS 4-5. We report our experience with SDR in CP patients with severe spasticity and motor impairment.

**MATERIAL-METHODS:** In a prospective series of spastic 23 patients with GMFCS 4 or worse who underwent SDR, 7 were classified as GMFCS 5 (ages 3-17y). All underwent SDR after the option of ITB was thoroughly discussed with the parents. In 22/23 ITB was offered as a valid and even preferable option to the parents, but SDR was chosen mainly because of fear from infection and preference to avoid a foreign body. One patient could not receive ITB because of sensitivity and anaphylactic reaction. The patients were variable regarding communication skills, and their clinical profiles.

**RESULTS:** Indications for surgery were pain, discomfort, difficulty in care giving due to extreme spasticity, difficulty in sitting and need for frequent adaptation of satisfactory wheelchair. All suffered pain, either related to motion or in rest and had abnormal hips on X rays. Half had scoliosis, and one was previously operated for scoliosis. In 90% of the patients there was significant improvement in pain, but all reported facilitation in transfers, in handling and in ability to perform physiotherapy better to preoperative period. Improved sitting was reported in the patients with significant scoliosis. There was no mortality, one complication of wound infection and meningitis in 1 patient, who underwent superficial revision of the wound and recovered well. There were 2 intraoperative anesthetic complications of bronchospasm during induction that resolved completely.

**CONCLUSIONS:** We conclude that SDR can be safely performed in patients with GMFCS 4-5 and that in most patients the goals are effectively achieved with minimal risk.

**Keywords:** CEREBRAL PALSY, SDR, Rhizotomy, GMFCS, QUADRIPARESIS, PAIN

## PF-026

### Epilepsy and functional

#### Selective dorsal rhizotomy for hereditary spastic paraparesis in children

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**OBJECTIVE:** The aim of this study was to determine the outcomes for children who underwent selective dorsal rhizotomy (SDR) for the treatment of spasticity related to spinal pathology.

**MATERIAL-METHODS:** We performed a retrospective review of all cases of SDR at our institution over the last 30 years and identified patients in whom spasticity was attributed to spinal rather than cerebral pathology. We gathered demographic information and recorded functional status and spasticity scores pre-operatively and over long-term follow-up.

**RESULTS:** We identified 4 patients who underwent SDR for spinal-related spasticity. All four had hereditary spastic paraparesis (HSP). All patients had reduced spasticity in the lower limbs after SDR, which was maintained over long-term follow-up. 2 patients had a more severe and progressive subtype of HSP, and both these patients exhibited functional decline over time despite improvement in tone.

**CONCLUSIONS:** Our findings suggest SDR is a reasonable option to consider for relief of spinal-related spasticity in uncomplicated hereditary spastic paraparesis. However, SDR for the treatment of complicated HSP seems to carry more risks and have a less predictable outcome. Overall, SDR is probably best reserved for pathologies that are relatively stable in their disease course.

**Keywords:** Hereditary spastic paraparesis, Selective dorsal rhizotomy, Spasticity

Monday, 24 October 2016

16:30 – 17:10

## Platform Presentations 5: Epilepsy & Functional

### PF-027

#### Epilepsy and functional

#### Effects of focal brain cooling in patients with intractable epilepsy: changes in intraoperative electrocorticogram and neurotransmitter concentrations

Sadahiro Nomura, Takao Inoue, Hirochika Imoto, Yuichi Maruta, Eiichi Suehiro, Yuya Hirayama, Michiyasu Suzuki

Department of Neurosurgery, Yamaguchi University School of Medicine

**OBJECTIVE:** Focal excision is indicated for patients with medically intractable epilepsy, but unable to be performed in eloquent brain regions. We developed a focal brain cooling treatment and successfully terminated epileptic discharge in animal models of epilepsy. In the present study, we applied this method to epilepsy patients during surgery and assessed its effectiveness using electrocorticogram (ECoG) monitoring and measured neurotransmitter concentrations.

**MATERIAL-METHODS:** Six patients (6–18 years old) with intractable epilepsy were included in this study. Epileptic focus was determined by video-ECoG monitoring. Prior to focal excision, cooling at 15°C for 30 min was performed within the area to be resected. Temperature and ECoG were continuously monitored. An extracellular fluid specimen was obtained and the concentration of

excitatory (glutamate, Glu) and inhibitory neurotransmitters (gamma-aminobutyric acid, GABA) was measured. The study protocol was approved by the institutional review board of Yamaguchi University Hospital.

**RESULTS:**The starting temperature of the brain's surface was 35°C and immediately dropped to 15°C after focal cooling. Epileptic discharges were alleviated, and the ECoG power spectra extracted from the fast beta band (14–24 Hz) was reduced from  $5.4 \times 10^{-9} \text{ V}^2$  to  $0.78 \times 10^{-9} \text{ V}^2$  after cooling. This reduction continued until the postcooling period. The concentration of Glu was 46.0  $\mu\text{mol/L}$  before cooling and decreased to 33.1  $\mu\text{mol/L}$  (72.0%) and 25.3  $\mu\text{mol/L}$  (54.9%) during the cooling and the postcooling periods, respectively. The concentration of GABA also decreased from 408 nmol/L to 253 nmol/L (62.7%) and 304 nmol/L (89.0%) during the cooling and postcooling periods, respectively.

**CONCLUSIONS:**Focal brain cooling inhibited abnormal neuronal excitation and neurotransmitter release from the presynaptic vesicles. The decrease in extracellular Glu specifically contributes to the inactivation of seizure propagation. The development of this focal brain cooling method, together with the development of implantable devices, would help patients with unresectable foci.

**Keywords:** Intractable epilepsy, Focal brain cooling, Neurotransmitters

## PF-028

### Epilepsy and functional

#### Altered regional homogeneity in epileptic patients with infantile spasm: a resting-state fMRI study

Qian Chen

Department of Neurosurgery, Shenzhen Children's Hospital, Shenzhen, China

**OBJECTIVE:**To investigate regional homogeneity (ReHo) changes in Infantile spasm (IS) patients.

**MATERIAL-METHODS:**Resting-state fMRI was performed on 11 patients with IS, along with 35 age- and sex-matched healthy subjects.

**RESULTS:**Group comparisons between the two groups demonstrate that the pattern of regional synchronization in IS patients is changed. Decreased ReHo values were found in default mode network, bilateral motor-related areas and left occipital gyrus of the patient group. Increased ReHo was found in regions of cingulum, cerebellum, supplementary motor area and brain deep nucleus, such as hippocampus, caudate, thalamus and insula. The significant differences might indicate that epileptic action have some injurious effects on the motor, executive and cognitive related regions. In addition, ReHo values of left precuneus and right superior frontal gyrus were associated with the epilepsy duration in the IS group. The correlation results indicate that the involvement of these regions may be related to the seizure generation.

**CONCLUSIONS:**IS may have an injurious effect on the brain activation. The findings may shed new light on the understanding the neural mechanism of IS epilepsy. IS syndrome is an age-related epileptic encephalopathy that occurs in children.

**Keywords:** epilepsy, resting-state fMRI, regional homogeneity, Infantile spasm, thalamus, DMN

## PF-029

### Epilepsy and functional

#### The role of intraoperative MRI in influencing the outcomes of resective epilepsy surgery: an initial experience

Santosh Mohan Rao Kanangi<sup>1</sup>, Sonia Tejada<sup>1</sup>, Andrea McLaren<sup>1</sup>, Ben Cooper<sup>1</sup>, Kay Hall<sup>1</sup>, Shivram Avula<sup>2</sup>, Sasha C Burn<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, Alder Hey Children's NHS foundation Trust, Liverpool L12 2AP, UK.

<sup>2</sup>Department of Radiology, Alder Hey Children's Foundation Trust, Liverpool, UK.

**OBJECTIVE:**The role of image guidance (in our case intra-operative MRI-ioMRI) in improving epilepsy surgery outcomes is still unclear. We present our surgical data analyses to shed some light on this question.

**MATERIAL-METHODS:**This retrospective study, included 65 patients aged between 2-18 years, operated at Alder Hey for epilepsy (excluding VNS) between 2009-2015. Follow-up was between 13 months-6 years. Only patients wherein a period of more than 12 months had elapsed post-surgery were included, lest factors like anti-epileptic drug adjustments & rehabilitation confound results.

The operated patients had multiple pathologies. Cortical dysplasia-8, neuronal heterotopia & cortical dysplasia-1, MTS & focal cortical dysplasia (FCD)-2, tuber & FCD-1, isolated tubers-5, oligodendroglioma (G2)-13, hemimegalencephaly-1, angiocentric glioma-1, ganglioglioma-1, fibrillary astrocytoma-2, DNET-1, SEGCA-1, cavernoma-2, hypothalamic hamartomas-7, MTS-6, FCD 1a-3, 1c-2, IIa-3, IIb-3, IIIa-2, IIIb-1, III (NOS)-1, recurrent oligodendroglioma (G3)-1, gliosis-2, hippocampal sclerosis-2, encephalitis-1.

The patients were asked 7 questions about seizure frequency, alertness, social interaction, memory, mood, sleep patterns and the caregiver's quality of life comprising an Impact on the Quality of Life Questionnaire. The responses were graded from 0-5 in ascending order of improvement.

**RESULTS:**Actions following an ioMRI were categorized as: Closure-44, further resection & closure-13, further resection, repeat ioMRI & closure-6. 1 patient had a closure and an immediate post-op MRI while another had no ioMRI. The outcomes of the aforementioned subsets were compared. ioMRI aided in the completeness of excision in tumours & cavernomas. In cases where the primary goal was disconnection, ioMRI use showed better outcome scores.

Patient response rate was 72%. Scores of 4 or 5 were good outcomes. The most significant improvements were in seizure frequency (74.4%), alertness (56.8%), interaction with the environment (50%) and caregiver's quality of life (62.9%).

**CONCLUSIONS:**ioMRI provides a clear and immediate picture of the anatomical completeness of the procedure for documentation enabling the surgeon to speak to the family with confidence at the end of the procedure, which greatly enhances parental satisfaction. Preliminary data suggests a trend towards better long term outcomes in epilepsy surgery with ioMRI. More studies are needed for validation.

**Keywords:** Epilepsy surgery, intra-operative MRI (ioMRI), Quality of life outcome

## PF-030

### Epilepsy and functional

#### Comparison in outcomes between corpus callosotomy and vagus nerve stimulation for medically refractory epilepsy in children

Takamichi Yamamoto, Tomohiro Yamazoe, Hirokazu Nakatogawa, Ayataka Fujimoto, Daiki Uchida, Naoto Kuroda, Chikanori Inenaga, Tokutaro Tanaka  
Department of Neurosurgery, Seirei Hamamatsu General Hospital, Hamamatsu, Japan

**OBJECTIVE:**Vagus nerve stimulation (VNS) therapy has become gradually popular as a palliative treatment option for intractable epilepsy in Japan. Corpus callosotomy (CC) is palliative as an intracranial epilepsy surgery. However, we do not have much information whether VNS or CC is better for each patient. The object of this study was to assess the efficacy of VNS and CC in seizure reduction for pediatric patients who underwent these procedures aged 10 years old and younger.

**MATERIAL-METHODS:**Twenty-three patients who underwent VNS implantation from 2011 through 2015 were retrospectively reviewed. Outcomes of VNS was evaluated using the McHugh (MH) Outcome Classification. In the same way, 11 patients who underwent CC in the same period of time were reviewed.

**RESULTS:**In patients treated by VNS, a more than 80% reduction in seizure frequency (MH Class 1) was obtained in 8 patients (38%), and a 50-79% seizure reduction (MH Class 2) in 6 patients (29%). Four out of 9 patients (44%) with VNS followed up for more than 2 years achieved MH Class 1,

although 4 out of 12 patients (33%) less than 2 years after the implantation showed MH Class 1. In patients who underwent CC, a more than 50% reduction was achieved in 5 patients (45%). Especially in epileptic falls, 4 out of 5 patient (80%) who underwent CC obtained seizure freedom. Improvement in their behavior was also seen in 6 patients (55%) several months after CC.

**CONCLUSIONS:** VNS and CC were effective treatment options as palliation for pediatric patients with medically refractory epilepsy. Although CC is an invasive procedure, CC has potentials in obliterating epileptic falls and improvement of mental development. On the other hand, VNS is less invasive and can be effective in about 50% of patients, although VNS needs time for programming and its output to be increased.

**Keywords:** medically refractory epilepsy, corpus callosotomy, vagus nerve stimulation, palliation, children

## PF-031

### Neuro-oncology

#### Surgical management of hypothalamic hamartomas with epilepsy: a 5-year experience with 92 cases

Chunde Li, Shiqi Luo

Department of Neurosurgery, Beijing Tiantan Hospital, Capital University of Medical Sciences, Beijing, People's Republic of China

**OBJECTIVE:** Hypothalamic hamartomas (HHs) require surgical treatment in patients presenting with refractory epilepsy.

**MATERIAL-METHODS:** The authors report on a single-center series of 92 patients (55 males, 37 females) who underwent surgery between January 2011 and December 2015. They experienced several types of seizure (gelastic, tonic, partial, atonic, generalized tonic clonic).

**RESULTS:** 55 were males and 37 were females; The average age was 6.8 years (range: 1.5 to 34 years), pterional approach were used in six patients (Type II) and total resection were achieved in five patients. Transcallosal interforniceal approach were used in 86 cases (Type II: two cases; Type III: 47 cases; Type IV: 37 cases) and subtotal resection were achieved in Type II: 50%; Type III: 47%; Type IV: 97%. One patient (Type III) died in the five day after operation. The average Follow-up time were 31 months (range from 4 to 63 months). After surgery, 62.5% (5/8) of patients with Type II HHs, 67.4% (31/46) with Type III, and 97% (36/37) with Type IV were free of epileptic seizures. One patient (Type III) developed hydrocephalus three months after HHs resection and were cured by V-P shunt operation.

**CONCLUSIONS:** In the present series, 97% of patients affected by Type IV HHs became seizure free; of those with Type III HHs at presentation, 67% recovered. Data from our series demonstrate that transcallosal interforniceal approach can cured more HHs patients than before.

**Keywords:** transcallosal interforniceal approach, Hypothalamic hamartoma, Intractable epilepsy, Surgery

## PF-032

### Craniofacial

#### Decompressive cranioplasty for mild metopic suture synostosis with clinical symptoms can improve function of the frontal lobe: a prospective dual institutional analysis including a natural history period

Kazuaki Shimoji<sup>1</sup>, Osamu Akiyama<sup>1</sup>, Takaoki Kimura<sup>1</sup>, Takeyoshi Shimoji<sup>2</sup>, Masakazu Miyajima<sup>1</sup>, Hajime Arai<sup>1</sup>

<sup>1</sup>Department of Neurosurgery, Juntendo University Tokyo, Japan

<sup>2</sup>Department of Neurosurgery, Okinawa prefectural southern medical center / children's medical center, Okinawa, Japan

**OBJECTIVE:** Generally, Increased intracranial pressure (ICP) and clinical symptoms are not considered as a symptom of single suture synostosis. On the other hand, there are reports that there are raised ICP and developmental delay in some degree in this pathology. To prove the efficacy of decompressive cranioplasty for mild metopic suture synostosis with clinical symptoms, we conducted a dual institutional prospective study including a natural history phase.

**MATERIAL-METHODS:** The inclusion criteria were children from 2-4 years old who have a bony ridge in the forehead with symptoms such as language delay and autistic tendency. 5 neurophysiological tests including Developmental Quotient (DQ) and Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS) a moderate test based on Childhood Autism Rating Scale (CARS), National Rehabilitation Center Sign-Significance (NRC S-S) test (A test for language understanding and expression) have been conducted at 4 time points (3 months pre-op, pre-op, 3 and 6 months post-op). The term from 3 months before surgery and just before surgery was considered as a natural history. The primary end point was configured as improvement of DQ in 6 months after surgery and the secondary end point was configured as the improvement of other tests.

**RESULTS:** 27 cases underwent surgery at the age of 3.2±0.9 years old. Compared to the natural history, improvement was seen in cognitive-adaptive scale and total scale in DQ also in PARS and expression of language in NRC S-S test both in 3 and 6 months post-op terms, statistically significantly.

**CONCLUSIONS:** The area decompressed is including the Broca area and the so-called mirror- neuron system which is now suspected to be related to autism. Although the natural history term was relatively short, the improved items of the tests are related to these area which is encouraging that this surgery may lead to an improvement of the function of the frontal lobe.

**Keywords:** single suture synostosis, intracranial pressure, language delay, autism

## PF-033

### Craniofacial

#### The surgical selection of distraction osteogenesis for craniosynostosis in our facility

Hirokatsu Osawa, Daimon Shiraishi, Mihoko Kato

Department of Neurosurgery, Aichi Children's Health and Medical Center, Obu, Japan

**OBJECTIVE:** The surgical selection for craniosynostosis is still a difficult issue because each procedure has the benefits and drawbacks. In many papers, conventional remodeling and distraction osteogenesis have been compared with, however, few papers about a new Multi-directional Calvarial Distraction Osteogenesis (MCDO) which has different concept from conventional distraction method has been discussed in terms of surgical indications. In our facility, we choose the type of distraction methods including MCDO according to symptoms in each case.

**MATERIAL-METHODS:** In order to investigate surgical selection and outcome for distraction osteogenesis including MCDO, we evaluated 58 cases with surgical cranial remodeling between 2010 and 2015 using CT scan, MRI images and chart review retrospectively.

**RESULTS:** This study includes 14 cases for conventional remodeling, 33 cases for conventional distraction method, and 10 cases for MCDO. The conventional distraction method would be applied to almost all types of fused sutures, on the other hand, MCDO tends to be used for sagittal or pan synostosis without plagiocephaly. Of the MCDO cases, the median age was older and the average surgical duration was 110 minutes longer than the conventional distraction method. On the basis of CT scan data, the change of postoperative cephalic index was 1.5% in the MCDO and 7.0% in the conventional distraction.

**CONCLUSIONS:**MCDO would be an effective procedure because the procedure has characteristics to maintain and enlarge skull shape in a balanced manner. In this study, we also discuss surgical indications and outcome in each procedure.

**Keywords:** Distraction Osteogenesis, Craniosynostosis, Surgical selection

#### PF-034

##### Craniofacial

##### Correction of nasal angulation in children with unilateral coronal synostosis

Hamilton Matushita<sup>1</sup>, Nivaldo Alonso<sup>2</sup>, Daniel Dante Cardeal<sup>1</sup>, Fernanda Gonçalves Andrade<sup>1</sup>, Manoel Jacobsen Teixeira<sup>1</sup>

<sup>1</sup>Department of Neurosurgery of São Paulo University

<sup>2</sup>Department of Plastic Surgery of São Paulo University

**OBJECTIVE:**Nasal bone remodeling or repositioning performed simultaneously with frontal-orbital advancement in children with unilateral coronal synostosis is still a controversial issue, mainly related to the necessity of correction. The aim of this study is to contribute in elucidating this problem. **MATERIAL-METHODS:**We report on 23 consecutive cases out of 94 children with unilateral coronal synostosis, in whom primary correction of the nasal deviation was performed. The technique comprised a remodeling of the internal e superior quadrant of the margins of the affected orbit with vertical osteotomy of the nasal bone. We measured the angular deviation of the nasal deviation in clinical photographs and preoperative 3D-CT Scans (Measure Software – Softonic – Windows).

The t test was used to compare the preoperative and postoperative of each patient. Significance was adopted at  $p < 0,05$  (Minitab 17 Software).

**RESULTS:**The mean age at surgery was 10,3 months (range 5 – 36 months). Gender distribution female/male was 14/9 = 1.5 The mean angular deviation of the nose preoperatively calculated on photography was 7,95 degrees (range - 5,8 to 9,9 degrees) and on pre-op CT Scan was 12,21 degrees (range - 5,4 to 16,3 degrees). All patients demonstrate post-operatively reduction of nasal angulation, mean deviation was 1,7 degrees (range - zero to 2,4 degrees) at the postoperative evaluation. The reduction was statistically significant ( $p < 0,05$ ). In all patients the aesthetic results were excellent. No patients required touch-up procedures.

**CONCLUSIONS:**This technique of correction of nasal angulation required a limited dissection of the soft nasal tissue and can be applied to any form or intensity of nasal angulation in children with unilateral coronal synostosis

**Keywords:** unicoronal synostotic plagiocephaly, craniosynostosis, nasal deviation, asymmetry.

Tuesday, 25 October 2016

08:00 – 09:00

## Platform presentations 6: Education | Trauma and Cerebrovascular

#### PF-035

##### Trauma

##### Clinical decision rule to obtain CT scan for infants with minor head injury: comparison with PECARN and application of quantification methods type II

Tadashi Miyagawa, Mariko Yabuki, Yoshiyuki Watanabe, Koichi Tamaki, Horohide Karasudani, Akira Yamaura

Department of Pediatric Neurosurgery, Matsudo City Hospital, Matsudo, JAPAN

**OBJECTIVE:**Of the currently published clinical decision rules(CDR) for the management of minor head injury (mHI) in children, the PECARN rule appears to be the best to identify children at very low risk of clinically important traumatic brain injuries (ciTBI). However, even a well-validated rule may not be the best for other settings because of population, clinical settings and practitioners. The aim of this study was to assess applicability of the PECARN rule for infants with mHI in Japan and to apply quantification methods type II to make a new simple CDR to use.

**MATERIAL-METHODS:**This retrospective cohort study was performed according to the PECARN. From 2005 to 2014, 1091 infants (younger than 2 years) with mHI were enrolled in this study. Data were compared to the results in the PECARN **RESULTS:**Based on PECARN definition, ciTBI in this study showed 2.6%, higher than that in PECARN. When applying the prediction tree for ciTBI in children younger than 2 years, the risk of ciTBI for those with none of six predictors was 0.7% which also showed higher compared to PECARN. The prediction rule had a negative predictive value of 99.3% and a sensitivity of 85.7%. Also when applying the suggested CT algorithm for children younger than 2 years, 10.9% of infants were classified as CT recommended, the risk of ciTBI for this group was 15.1%, which showed much higher than that in PECARN. Quantification methods type II was applied for making a new simple CDR. The prediction rule had a negative predictive value of 99.6% and a sensitivity of 99.2%.

**CONCLUSIONS:**The PECARN rule would successfully be applied in Japan. A new CDR produced with quantification methods type II would be better to identify children at very low risk of ciTBI.

**Keywords:** clinical decision rule, clinically important traumatic brain injury, CT, PECARN, quantification methods type II

#### PF-036

##### Trauma

##### Resource utilization in the transfer of pediatric patients with head injuries

Clay M Elswick<sup>1</sup>, Lori A Gurien<sup>2</sup>, Deidre L Wyrick<sup>2</sup>, Mallikarjuna R Rettiganti<sup>2</sup>, Marie E Saylor<sup>2</sup>, Ambre L Pownall<sup>1</sup>, Diaa Bahgat<sup>1</sup>, Eylem Ocal<sup>1</sup>, Robert T Maxson<sup>2</sup>, Gregory W Albert<sup>1</sup>

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**OBJECTIVE:**No guidelines exist for interfacility transfer of head-injured children to pediatric level I trauma centers. Previous studies have suggested that helicopter emergency medical services (HEMS) may be overutilized in pediatric trauma. The purpose of our study is to evaluate for reasonable interfacility transport of head-injured children by HEMS to an ACS verified level I pediatric trauma center and to determine criteria that would predict need for HEMS transport.

**MATERIAL-METHODS:**All trauma patients transferred by HEMS from an outside facility to our trauma center and underwent neurosurgical consultation were identified from 2008-2012. Patients without head injuries or found to have severe injury of another body region were excluded. Reasonable HEMS transport was defined as patients who died from their head injuries or required neurosurgical intervention (surgery or bedside procedure). Factors associated with reasonable HEMS transport were identified using a logistic regression model which included demographic variables, Glasgow Coma Scale (GCS), intubation, and CT findings. **RESULTS:**373 patients met inclusion criteria. 257 (69%) were classified as non-reasonable HEMS transport. In the reasonable HEMS transport group (N=116), 26 patients died (22%). Within the non-reasonable HEMS transport group, 61% (N=157) were admitted to the ICU, 2% (N=5) to the intermediate unit, and 37% (N=94) to the floor. One patient was discharged home from the trauma bay. 30% (N=78) of the non-reasonable HEMS transfers were discharged home in under 24 hours. Factors associated with reasonable air transport included decreased GCS (3-8) and the following CT FINDINGS: mass effect/midline shift/

herniation, edema/loss of gray-white matter/hypoxic-ischemic appearance, epidural hematoma, and open and depressed skull fracture.

**CONCLUSIONS:** HEMS is overutilized in pediatric trauma patients with head injuries. Patients that lack the characteristics defined as predictive of death or need for neurosurgical intervention may not need to be transferred by air, but instead by ground, resulting in better allocation of costly resources.

**Keywords:** Helicopter transport, closed head injury

## PF-037

### Trauma

#### Therapeutic strategy for severe traumatic brain injury in children and indication of decompressive craniectomy: a single center experience

Takashi Araki<sup>1</sup>, Hiroyuki Yokota<sup>1</sup>, Akira Fuse<sup>1</sup>, Shoji Yokobori<sup>1</sup>, Hidetaka Onda<sup>1</sup>, Kentaro Kuwamoto<sup>1</sup>, Takahiro Kanaya<sup>1</sup>, Akio Morita<sup>2</sup>

<sup>1</sup>Department of Emergency and Critical Care Medicine

<sup>2</sup>Department of Neurosurgery

**OBJECTIVE:** To describe the usual practice for various intracranial pressure (ICP)-targetted medical therapies and indication of decompressive craniectomy (DC) for intracranial hypertension with severe traumatic brain injury (TBI) in children.

**MATERIAL-METHODS:** A retrospective chart review of data was performed using a database to pediatric patients (<16 years old) with TBI on the Nippon Medical School Hospital between January 2002 and December 2009. Medical charts were reviewed for documented performance of specific elements derived from Guidelines for the Management of Severe Head Injury, 2nd Edition, the Japan Society of Neurotraumatology

**RESULTS:** RESULTS: 122 pediatric TBI patients studied and fifty patients were severe TBI (GCS<8). Patients were categorized into two group, with medically treated (n=39: group M) and surgically treated (n=11: group S). ICP monitoring was used in 78% (39/50) of children with severe TBI, 79.5% (31/39) of them showed ICP decrease with ICP-targetted medical therapies, such as hyperosmolar diuretics (17/39:57%), mild hyperventilation (10/39:30%), the use of sedatives (9/39: 30%). DC was performed in 38% (19/50) of children and the ICP sensor insertion (OR 9.17, 95%CI 1.07-78.50), the intracranial hemorrhagic lesion (OR 47.33, 95%CI 6/16-363.44) were found to have strong correlation with DC. The group S had a pupillary abnormality (8 vs 5, p<0.01), had a higher the Injury Severity Scale score (32.8±10.2 vs 9.4 ±8.6, p<0.01), hypotension (6 vs 2, p<0.01) compared to the group M. At the most recent follow-up examination, favorable outcome were seen on 76.5% (29/39) children of group M and 78.9% (15/19) children of group S.

**CONCLUSIONS:** Reducing ICP is crucial factor in patients' survival with severe TBI in children. DC might be beneficial in the management of refractory intracranial hypertension. Long-term follow-up is important to determine neurological sequelae associated with TBI. Children make significant functional gains during inpatient rehabilitation and better prognosis can be expected with multidisciplinary management.

**Keywords:** Therapeutic strategy, traumatic brain injury, decompressive craniectomy

## PF-038

### Vascular

#### Pediatric spinal vascular malformation: considerations of classification, diagnosis and treatment

Gao Zeng, Hong Qi Zhang, Xing Long Zhi, Jian Xin Du, Feng Ling

Department of neurosurgery, XuanWu hospital, Capital medical university, Beijing, China

**OBJECTIVE:** Spinal vascular malformations are rare diseases in pediatric age. Despite of great advances with technological and materials in recent years, there continue to pose a challenge to treatment these complex disorders. The objective of this work was the retrospective study for these challenging diseases according to the single center of 13 years experiences.

**MATERIAL-METHODS:** 138 pediatric spinal vascular malformation patients (≤14-year-old) hospitalized in 2002-2015 in Xuanwu hospital capital medical university were reviewed, and the characteristics of the classification, epidemiology, clinical symptoms, diagnoses, and therapies were analyzed.

**RESULTS:** Pediatric spinal vascular malformations occupied only 7% of all cases during the same period. The highest incidence was seen during 8-14 years old. Spinal cord arteriovenous malformation (SCAVM) was the most common (51.4%) in pediatric patients, while Type II and Type III perimedullary arteriovenous fistula (PMAVF) (26.8%) was the second, followed by Cobb's syndrome (7.9%). No dual arteriovenous fistulae were found. 70% pediatric patients presented with hemorrhage. Embolization therapy can cure near half of pediatric spinal vascular malformation patients.

**CONCLUSIONS:** There is much difference in many aspects between the pediatric and adult population in spinal vascular malformations. Clearly classification is the base of making correct treatment strategy. Embolization is the effective method for the children in order to keep the spinal column stable. For the complicated cases, partial embolization aimed at aneurysms and the suspected part to be the chief mechanism of the neurological deterioration is recommended. Combined surgery is an effective technique for some SCAVM, dissect the AVM nidus strictly around the membrane boundary between the nidus and normal tissue.

**Keywords:** Pediatric, spinal vascular malformation, Spinal cord arteriovenous malformation, perimedullary arteriovenous fistula, Cobb's syndrome

## PF-039

### Vascular

#### Posterior cerebral artery stenosis and posterior revascularization surgery in pediatric patients with moyamoya disease

Tomomi Kimiwada<sup>1</sup>, Toshiaki Hayashi<sup>2</sup>, Reizo Shirane<sup>1</sup>, Teiji Tominaga<sup>3</sup>

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<sup>3</sup>Department of Neurosurgery, Tohoku University Graduate School of Medicine, Sendai, Japan

**OBJECTIVE:** To investigate the presence of posterior cerebral artery (PCA) stenosis before and after anterior circulation revascularization surgery and clinical characteristics in pediatric patients with moyamoya disease (MMD).

**MATERIAL-METHODS:** 62 pediatric patients treated in our hospital, under 16 years old with MMD, were analyzed.

**RESULTS:** Twenty-three (37%) of pediatric MMD patients present with PCA stenosis at the time of initial diagnosis. A strong correlation between the presence of infarction and PCA stenosis before anterior revascularization was found (p<0.001). In addition, progressive PCA stenosis was found in 12 (19.4%) patients following anterior revascularization. Presence of infarction and younger age at the time of initial diagnosis were risk factors for progressive PCA stenosis after anterior revascularization (p<0.001 and p=0.002, respectively). We performed posterior circulation revascularization surgery including occipital artery – posterior cerebral artery bypass for nine of 12 patients with progressive PCA stenosis. All of the patients showed symptomatic and radiologic improvement.

**CONCLUSIONS:** PCA stenosis is an important clinical factor related to poor prognosis in pediatric MMD. We should be aware of the possibility of progressive PCA stenosis during the postoperative follow-up period and consider posterior circulation revascularization surgery.

**Keywords:** moyamoya disease, posterior cerebral artery stenosis, posterior revascularization

#### PF-040

##### Vascular

#### **Pediatric brain cavernous malformations and rebleeding: infratentorial versus supratentorial clinical presentation and surgical treatment**

Miroslav Gjurašin<sup>1</sup>, Vlatka Mejaški Bošnjak<sup>2</sup>, Vlasta Djuranović<sup>2</sup>, Ana Tripalo Batoš<sup>3</sup>, Goran Roić<sup>3</sup>, Pavle Miklič<sup>1</sup>, Jadranka Sekelj Fureš<sup>2</sup>, Ljiljana Popović<sup>4</sup>

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**OBJECTIVE:** Vascular brain malformations in children include cavernous malformations (CM), arterio-venous malformations (AVM), developmental venous anomalies (DVA) and capillary teleangiectasia. They differ by location, clinical presentation, management, and treatment. In this work we analyze the treatment of pediatric patients with CM operated at our institution, with special emphasis on supratentorial vs. infratentorial features.

**MATERIAL-METHODS:** Among 72 pediatric patients (0-18 years) with diagnosed vascular brain malformation at Children's Hospital Zagreb Klaiceva in the period from 2005 to 2015., we analyzed clinical presentation, diagnostic procedure, treatment and results for operated patients with CM.

**RESULTS:** Among all pediatric patients with diagnosed vascular brain malformations (n=72), 47 patients had venous angioma (35 supratentorial, 11 infratentorial, 1 multiple), 12 arterio-venous malformation (11 supratentorial, 1 pontine), 11 cavernoma (10 supratentorial, 1 cerebellar), and 2 pontine capillary teleangiectasia.

In the supratentorial CM group, 4 patients presented with seizures, 3 with headache, one with speech disorders, one with disoriented behavior, and one with leg weakness.

A 12-year-old boy with infratentorial deep cerebellar CM presented with vomiting, nystagmus, ataxia and vertigo; this patient had associated cerebellar DVA with typical sign of caput medusae where distal radicles covered posterior and upper aspect of the CM located next to the IV ventricle. Preoperatively, on the 11th day after first hemorrhage, he suffered significant rebleeding with CM enlargement and clinical deterioration, but without intraventricular blood penetration. Ten of 11 CM patients were operated and recovered, among them the patient with infratentorial CM recovered with partial hearing impairment.

**CONCLUSIONS:** While all supratentorial CM showed rather stable clinical condition, one infratentorial CM presented in our series had aggressive behavior with early preoperative rebleeding, significant enlargement of the CM volume, and neurological deterioration. Early diagnosis and timely treatment significantly contribute to functional recovery and optimal clinical result, especially for infratentorial CM.

**Keywords:** pediatric cavernous malformation, infratentorial, rebleeding

Wednesday, 26 October 2016

09:15 – 10:35

## Platform presentations 7: Neuro-Oncology

#### PF-042

##### Special topic: Molecular biology

#### **Intraoperative neurophysiology in pediatric supratentorial surgery: experience with 59 cases**

Akiva Korn, Haggai Benvenisti, Mona Jubran, Yifat Bitan Talmor, Perla Ekstein, Shlomi Constantini, Jonathan Roth  
Dana Children's Hospital, Tel-Aviv Medical Center, Tel-Aviv, Israel

**OBJECTIVE:** The utilization of intraoperative neurophysiology (ION) to map and assess various functions in supratentorial surgery is well documented and commonplace in the adult setting. It is less described in the pediatric setting due to the relatively low incidence of relevant pathologies, as well as relative difficulty to implement these methods for physiological reasons. The applicability has yet to be established as feasible or beneficial, and optimizations of current techniques are not known.

**MATERIAL-METHODS:** Retrospective analysis was undertaken in all pediatric supratentorial surgery utilizing ION of the motor system over a period of 9 years. Intraoperative findings were correlated with short and long term postoperative clinical outcome. The monitoring impact on surgical course was evaluated on a per case basis.

**RESULTS:** Data of 59 patients were analyzed (avg age  $94.1 \pm 58.6$  months). Deep lesions consisted of 18.5% of the total group, superficial lesions 54.2%, lesions with both deep and superficial components 22.0% and ventricular 5.1%. Monopolar mapping of the motor cortex was significantly more successful than bipolar mapping (84.6% vs 28.1% of trials respectively), the youngest age mappable was 3 vs 154 months for either method respectively. Successful mapping of the primary sensory cortex correlated with increasing age, and was found at  $107.4 \pm 59.5$  mo., all older than 6 mo. 23 patients had full clinical and ION motor correlations. The predictability of a postoperative motor deficit inversely correlated with the scrtMEP value.

**CONCLUSIONS:** The relative difficulty of motor mapping and monitoring correlates with the timeline of maturation of this systems. ION is applicable in the pediatric population; however with certain limitations depending mainly on age. Yet when successful, ION has a positive impact on surgical decision making, ultimately providing an added element of safety to these patients.

**Keywords:** intraoperative neurophysiological monitoring, tumor resection, motor function, supratentorial surgery, epilepsy

#### PF-043

##### Neuro-oncology

#### **Neoadjuvant chemotherapy for infantile brain tumors in infants and young children**

Hideki Ogiwara

National Center For Child Health And Development

**OBJECTIVE:** Complete removal of infantile brain tumors is often difficult due to large size and high vascularity, while degree of resection is related to their prognosis in most cases. Neoadjuvant chemotherapy may facilitate resection by reducing the vascularity of the tumor.

**MATERIAL-METHODS:** Retrospective review of infants who underwent tumor removal after neoadjuvant chemotherapy was performed.

**RESULTS:**Nine patients underwent surgical resection after neoadjuvant chemotherapy. The mean age was 18 months. Five patients underwent partial resection, and 4 underwent biopsy as an initial surgery. The histopathological diagnoses were ependymoma in 2 patients, anaplastic ependymoma in 1, primitive neuroectodermal tumor (PNET) in 2, choroid plexus carcinoma in 1, atypical teratoid/rhabdoid tumor (AT/RT) in 1, glioblastoma in 1, and embryonal tumor with abundant neuropil and true rosettes (ETANTR) in 1. After 2 to 4 courses of multiagent chemotherapy, the second-look surgery was performed. The tumor volume was reduced to varying degrees in 5 patients (56%) after chemotherapy. Intraoperatively, the vascularity of the tumor was considerably reduced and the tumor was more circumscribed in all cases. Gross total resection was achieved in 8 patients (89%) and near-total resection in 1 (11%). Histopathological examination demonstrated fibrotic tissue circumscribing the tumor in 6 cases (67%, 6/9). The average blood loss was 20% of the estimated blood volume, and 3 (33%) required a blood transfusion. There was no surgical mortality. One patient had transient dysphasia postoperatively. The mean follow-up period was 28 months. At the last follow-up, 2 patients (22%) died (due to tumor progression in 1 and sepsis in 1), and 4 patients (44%) had no recurrence of the tumor.

**CONCLUSIONS:**Neoadjuvant chemotherapy for infantile brain tumors was effective in reduction of tumor vascularity and clarification of the tumor-brain interface, which significantly facilitated the maximal tumor resection.

**Keywords:** neoadjuvant chemotherapy, infantile brain tumors, second-look surgery

#### PF-045

##### Neuro-oncology

##### Transcallosal interforaminal approach to pineal region tumors in 704 children: operative technique and results

Wei Liu, Zhenyu Ma, Shiqi Luo, Chunde Li, Jian Gong, Yongji Tian  
Department of Neurosurgery, TianTan Hospital, Capital Medical University, Beijing, China

**OBJECTIVE:**Tumors of the pineal region are histopathologically heterogeneous in children and surgical treatment remains challenging. This study was to describe the techniques and clinical results in children who were treated and histologically verified using a transcallosal interforaminal microsurgical approach.

**MATERIAL-METHODS:**From 1998 to 2015, 704 children (564 boys and 140 girls, 1 to 15 years), with pineal region tumors were underwent tumor resection in our ward. According to the histological subtypes and tumor marker in blood, all patients were treated using this approach followed with or without radiation and chemotherapy.

**RESULTS:**Five hundred ninety-one patients had germ cell tumors(234, mature teratomas; 303, immature teratomas or mixed germ cell tumors; 23, germinomas; 20, choriocarcinomas; 7, yolk sac tumors; 4, embryonal carcinoma); 45, pineoblastomas; 15, pineocytomas; 10, cavernous hemangiomas; 24, astrocytomas; 2, glioblastomas; and 17, dermoid cysts. From 1998 to 2005, 215(86%) tumors were totally removed, 30 subtotally removed, and 5 partly removed. From 2006 to 2015, 428(94.3%) were totally removed and 26 subtotally removed. There were no surgical mortality and no subsequent instances of disconnection syndrome. Short-term memory deficits appeared in 143 patients who had giant tumor and resolved within 6 months in most. There were 8 patients with preoperative unconsciousness that resolved within 4–20 days after surgery. Parinaud's syndrome was observed in 19 patients after surgery; 27 appeared preoperatively who resolved within 4 months. A total of 245 patients who were followed during 9 months and 17 years were evaluated. The progression-free survival was achieved in all mature teratomas and 40 immature teratomas or mixed germ cell tumors. Nine choriocarcinomas, 4 yolk sac tumors, 7 pineoblastomas and 1 glioblastomas were dead following neoplasm recurrence.

**CONCLUSIONS:**This approach is useful and safe for removing tumors in and around pineal region without major complications. Compared with other approaches, complete resection can be achieved in the more children.

**Keywords:** third ventricle, transcallosal surgery, pineal region tumors, follow-up, childre

#### PF-046

##### Neuro-oncology

##### Dose-reduced radiotherapy with platinum-based chemotherapy has improved quality of life in long-term survivors with germinoma

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<sup>2</sup>Department of Neurosurgery, Hiroshima University Hospital, Hiroshima, Japan

**OBJECTIVE:**To evaluate the efficacy of dose-reduced radiotherapy with platinum-based chemotherapy to improve quality of life in germinoma patients (pts).

**MATERIAL-METHODS:**We analyzed 45 pts with histologically verified germinoma. They were treated with 3–8 cycles of platinum-based chemotherapy concomitantly with 24-Gy radiation during 1st to 3rd cycle of chemotherapy. The median age was 14.5 years (9–39 years) including 38 male and 7 female. The lesions involved neurohypophysis in 23 and never in other 22. The median follow-up periods were 185 months (68–235 months).

**RESULTS:**All tumors disappeared completely. Tumor recurred in 6 pts, respectively 25, 25, 83, 105 and 156 months after initial therapy. All achieved second complete remission by chemo-radiotherapy. Chronic myelocytic leukemia and parotid carcinoma were observed as second malignancy and well controlled with additional therapy. Overall survival rate is 100%. Relapse-free and event-free rate in 10-year were 92.5% and 85.7%, respectively. The anterior pituitary function in 18 of 22 pts with neurohypophyseal lesion was always preserved and occasionally improved resulting one year after treatment. Clinically, 6 males recovered from ED, 2 females maintained regular menstrual cycle and 31 had regular occupation.

**CONCLUSIONS:**This treatment provided germinoma patients with satisfactory outcome and quality of life.

**Keywords:** germinoma, long-term survivor, chemotherapy, dose-reduced radiotherapy, second tumor, quality of life

#### PF-046

##### Neuro-oncology

##### Analysis about hypersensitivity of intracranial germinomas to low-dose radiation: relationship between volumetric changes and diagnostic radiation dose

Naoki Kagawa<sup>1</sup>, Ryuichi Hirayama<sup>1</sup>, Yasunori Fujimoto<sup>1</sup>, Yasuyoshi Chiba<sup>2</sup>, Chisato Yokota<sup>1</sup>, Shogo Fukuya<sup>1</sup>, Daisuke Eino<sup>1</sup>, Koji Takano<sup>1</sup>, Manabu Kinoshita<sup>3</sup>, Naoya Hashimoto<sup>4</sup>, Toshiki Yoshimine<sup>1</sup>

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**OBJECTIVE:**Spontaneous regressions in intracranial germinomas have been reported in some cases, but the natural history of them has not been well

known. To answer a part of that question, we retrospectively measured the tumor volume before and after chemo-radiotherapy and analyzed factors that influence shrinkage of tumor volume.

**MATERIAL-METHODS:**Thirty-one cases with primary intracranial germinomas and HCG-producing germinomas were treated in our hospital from 1994 to 2014. In twelve of them, plural MRI scans were done before the first course of chemotherapy regimen. Their age ranged from 8 to 26 years. Three cases were bifocal type. Biopsies were performed in all regions. Tumor volume of fifteen lesions was analyzed by volumetric assessment based on MRI. Ratio of volumetric change between the first MRI on admission and the scan immediately before chemotherapy was defined as shrinking rate (%). Period between disease onset and the first chemotherapy was 20 to 47 days. Diagnostic radiation dose was calculated in each case.

**RESULTS:**Initial tumor volume ranged from 0.962 to 72.356 cubic centimeters (mean: 8.55). Diagnostic radiation dose: 52.2 to 910.1 mGy. Shrinking rate ranged from -57.8 to 85.4% (mean: 24.0). In only 7 regions, shrinking rate was within 30%. Both shrinking rate and modified shrinking rate corrected by the number of days was significant positively influenced by diagnostic radiation dose ( $p=0.02$ ,  $p=0.009$ ). Shrinking rate was negatively influenced by initial volume ( $p<0.05$ ). But, it had no correlation with age, sex, histopathological parameters.

**CONCLUSIONS:**This study shows that the volumes of intracranial germinomas are changing dynamically before chemoradiotherapy in many cases and also their shrinkage is mainly influenced by low-dose radiation at diagnosis.

**Keywords:** intracranial germ cell tumor, radiosensitivity, volumetric change, diagnostic radiation dose

#### PF-048

##### Neuro-oncology

##### Vascularization of optic gliomas: primitive invertebrate-like channels-clinical and therapeutic implications

David Houston Harter<sup>1</sup>, Matija Snudr<sup>2</sup>, Pamela Wu<sup>3</sup>, Guoan Zhang<sup>2</sup>, Matthias Karajannis<sup>2</sup>, Jeffery H Wisoff<sup>2</sup>, Benjamin Cohen<sup>4</sup>, Tara S Jennings<sup>2</sup>, Seema Shroff<sup>2</sup>, Valerio Ortenzi<sup>2</sup>, Ranjan Jain<sup>4</sup>, David Zagzag<sup>2</sup>

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<sup>3</sup>Department of Biochemistry and Molecular Pharmacology, New York University Medical School, New York, NY

<sup>4</sup>Department of Radiology, New York University Medical School and Langone Medical center, New York, NY

**OBJECTIVE:**Optic gliomas are characterized as pilocytic astrocytoma (PA) or pilomyxoid astrocytoma (PMXA). Prominent chondroid myxoid matrix is typical of PMXA but not PA. We investigated the composition of myxoid matrix and its role in vascularization of optic gliomas.

**MATERIAL-METHODS:**We reviewed clinicopathological data of 120 patients with optic glioma diagnosed at NYU Langone Medical Center from 1996 to 2014. We analyzed microvascular density (MVD), perfusion, hypoxia and proliferation by immunohistochemistry and ultrastructural features by electron microscopy. Liquid chromatography–mass spectrometry (LC-MS) was performed to identify components of the myxoid matrix in PMXA.

**RESULTS:**PMXA showed significantly lower MVD by CD34 (8.1 vs 14.5,  $p$ -value  $< 0.002$ ) and Erg (7 vs. 13.6,  $p$ -value 0.003) than PA, however GLUT-1 showed equal distribution. Electron microscopy showed that PMXA contains both regular blood vessels with endothelial lining and channels completely lacking endothelia and smooth muscle. LC-MS stratified optic gliomas into three distinct groups. We identified 5389 proteins of which 188 were differentially expressed in the three groups ( $p < 0.05$ , Benjamini-Hochberg adjustment). Between PA and PMXA, most of differentially expressed proteins (146/

188) displayed a positive fold change (increasing in PMXA relative to PA), and a minority (42/188) showed a negative fold change. Abundant extracellular matrix proteins were a chondroitin sulfate proteoglycan versican (VCAN 3.7-fold increase  $Q=0.000463$ ) and its paralog vertebrate Hyaluronan and Proteoglycan Link Protein 1 (HAPLN1, 22-fold increase from the PA to the PMXA group  $Q=4.60 \times 10^{-7}$ ).

**CONCLUSIONS:**Optic gliomas develop endothelium-independent channels evocative invertebrate blood supply. The myxoid matrix is composed of VCAN and linking paralog HAPLN1. Targeting the myxoid matrix may provide novel avenues for therapy of optic gliomas and PMA.

**Keywords:** Optic glioma, Pilomyxoid astrocytoma, Pilocytic astrocytoma

#### PF-049

##### Neuro-oncology

##### Surgical treatment of 88 optic pathway gliomas

Eveline Teresa Hidalgo<sup>1</sup>, Svetlana Kvint<sup>1</sup>, Cordelia Orillac<sup>1</sup>, Emily North<sup>1</sup>, Cheddi Thomas<sup>2</sup>, Yosef Dastagirzada<sup>1</sup>, Matija Snudr<sup>2</sup>, Jeffrey H. Wisoff<sup>1</sup>

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<sup>2</sup>Department of Pathology, NYU Langone Medical Center, New York, USA

**OBJECTIVE:**Pediatric optic pathway gliomas (OPGs) are often considered benign, but can have detrimental effects on the quality of life, impair vision and are a potentially lethal disease. The aim of this study is to report the characteristics and outcomes of surgically treated OPGs and to identify candidates for different treatment strategies.

**MATERIAL-METHODS:**Retrospective chart review of consecutive pediatric patients with surgically treated OPGs by a single surgeon at our institution from 1985-2015. Three treatment pathways were defined: surgery without planned adjuvant therapy (1), surgery with planned adjuvant therapy (2) and patients with prior treatment (3).

**RESULTS:**88 patients – 49 male and 39 female – were included in analysis. 8 patients had NF1. Pathology revealed pilocytic astrocytoma (85.2%), pilomyxoid astrocytoma (8%), and pilocytic/pilomyxoid astrocytoma (5.7%). Radiologic location of the tumor was: hypothalamic (87%), and involvement of only chiasm and/or tract in (13%). Median age at diagnosis was 4 years, median age at surgery was 6 years, and median time from diagnosis to surgery was 1 year.

At the time of the study:

Pathway 1: 37 patients; median PFS  $84 \pm 33.4$ ; median OS of 118 months (range: 24 – 337 months); OS rate 68%.

Pathway 2: 9 patients; median PFS  $45 \pm 10.9$  months; median OS of 127 months (range: 23 – 368 months); OS rate 67%.

Pathway 3: 42 patients; median PFS  $74 \pm 13.3$  months; median OS of 69 months (range: 7 – 356) months; OS rate 76%.

**CONCLUSIONS:**The role of surgery in the treatment of pediatric OPGs depends on patient characteristics and tumor biology. With the adequate therapeutic strategy, long-term PFS and OS can be achieved.

**Keywords:** Optic Pathway Gliomas Hypothalamic Glioma Surgical Treatment

#### PF-048

##### Neuro-oncology

##### Childhood craniopharyngioma: results of a prospective study of risk-adapted strategies to spare hypothalamus structures

Stephanie Puget<sup>1</sup>, Kevin Beccaria<sup>1</sup>, Thomas Blauwblomme<sup>1</sup>, Michel Zerah<sup>1</sup>, Michel Polack<sup>2</sup>, Delphine Zenaty<sup>4</sup>, Christian Sainte Rose<sup>1</sup>, Claire Alapetite<sup>3</sup>

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<sup>4</sup>Departement of Pediatric Endocrinology, Robert Debre, Paris

**OBJECTIVE:**The surgical management of pediatric craniopharyngiomas remains one of the more controversial topics in pediatric neurosurgery. There has been a growing worldwide advocacy for less extensive resection and for the utilization of multimodality therapy to limit morbidity.

**MATERIAL-METHODS:**Since 2002, we developed and applied prospectively a risk-adapted strategies designed to preserve hypothalamic structures (Puget et al, JNS 2007). We report our results for hypothalamic disturbances, endocrinological deficits and tumor control

**RESULTS:**The authors reviewed data obtained in a prospective cohort of 111 children who underwent resection between 2002 and 2014. Our Preoperative Hypothalamus-Involvement grading stratified patients as grade 0 (16%), grade 1 (32%) and grade 2 (52%), correlated to preoperative hyperphagia and obesity. The Postoperative Hypothalamic-Damage grading found 41.4% of grade 0 (intact hypothalamus), 57% of grade 1 (residu on the hypothalamus) and 1.6% of grade 2 (hypothalamus damage), also correlated to clinical hypothalamus dysfunction. A complete resection (CR) was achieved in 54 patients with endonasal approach in 28. The remaining 57 children received radiotherapy on the residu (protons for 52 patients). One patient died in the postoperative course due to a vessel injury. The post operative morbidity comprised 1 hemiparesis, 1 epilepsy and 1 memory deficiency. We observe no morbid hyperphagia, even if some patients gained weight, or major behavioral troubles. After a median FU of 6 years, we observed 10 relapses after CR and 4 after incomplete resection. Compared to our historical cohort with the aim of CR, we observed less endocrinological deficits and 4 patients had no hormonal substitution. We observed no complications due to RT except a basal meningioma which was removed.

**CONCLUSIONS:**Hypothalamus-sparing surgery decreases the occurrence of severe obesity and hypothalamus dysfunction without increasing the local recurrence rate. Our risk-adapted grading system should be evaluated in multicenters to validate its reproducibility and accuracy.

**Keywords:** craniopharyngioma, hypothalamus, risk-adapted strategy, obesity

## PF-051

### Neuro-oncology

#### Quality of life and sexual function in adulthood two decades after gross-total resection of childhood craniopharyngioma

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Division of Pediatric Neurosurgery, Department of Neurosurgery, NYU Langone Medical Center

**OBJECTIVE:**Gross-total resection of craniopharyngioma is associated with complications that potentially affect quality of life. This study was designed to investigate the impact of gross-total resection on the long-term quality of life and sexual functioning in adulthood.

**MATERIAL-METHODS:**55 consecutive pediatric patients treated with primary gross-total resection for craniopharyngioma were included in this retrospective fixed cohort study. A multidimensional questionnaire-based quality of life instrument, the SF36v1 and the Medical Outcomes Study family and sexual functioning scale, was chosen to analyze follow-up data. Additionally, patients were asked to fill out a Questionnaire about medication, visual impairment, education and family life.

**RESULTS:**Of 43 mailed questionnaires 23 were returned (response rate 55%); 17 patients were lost to follow up, 3 individuals were reluctant to participate, 7 patients died and 5 patients were minors at the time of study. The median length of follow up was 19 years (range 10-31). BMI was underweight in 1 patient (4%), normal in 3 patients (13%), overweight in 8 patients (35%), obesity in 4 patients (17%) and severe obesity in 7 patients (31%). 18 out of 19 patients reported about sexual functioning, of whom 53% reported at least 'a little of a problem' in one or more areas of sexual functioning.

The mean SF 36v total score was 51.9 for PCS (physical health) and 48.5 for MCS (mental health), no significant difference between the patient cohort and the normal population.

No significant correlation between BMI and PCS, but significant correlation between BMI and MCS.

44% Patients reported to have excellent or very good health in general.

**CONCLUSIONS:**In a cohort of adults who underwent gross-total resection for craniopharyngioma in childhood, quality of life scores according to the SF 36 instrument were not different from those in a normal sample population. Sexual dysfunction is slightly more prevalent in this population.

**Keywords:** Craniopharyngioma, Long-term Outcome, Quality of Life, Sexual Function

## PF-052

### Neuro-oncology

#### Pediatric chordomas: results of a multicentric series of 38 children and proposal for a histopathological prognostic grading system and new therapeutic strategies

Kévin Beccaria<sup>1</sup>, Arnault Tauziède Espariat<sup>2</sup>, Marc Polivka<sup>3</sup>, Christian Sainte Rose<sup>1</sup>, Michel Zerah<sup>1</sup>, Pascale Varlet<sup>2</sup>, Stéphanie Puget<sup>1</sup>

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**OBJECTIVE:**Pediatric chordomas are rare slow-growing malignant neoplasms and few data are available on their optimal therapeutic strategy and outcome. This study aimed at evaluating their management and identifying prognostic factors.

**MATERIAL-METHODS:**This multicentric retrospective study included 38 children from 4 French centers, having a chordoma diagnosed between 1966 and 2012. Clinical, radiological, histopathological data and the type of treatment and outcome were analyzed.

**RESULTS:**The median age was 12.2 years-old with a male predominance (24 M/14 F). Most of chordomas were classical forms (45.5%), especially located in the clivus and cranio-cervical junction (84.2%). The overall survival (OS) was 66.6% and 58.6%, and the progression free survival (PFS) was 55.7% and 52% at 5 and 10 years respectively. A histopathological/immunohistochemical grading system recently proposed for adults based on histopathological subtype, mitoses, apoptosis, prominent nucleoli, necrosis, Ki67 and p53 labelling index was applied for this pediatric series. Importantly, in multivariate analysis it significantly correlated to outcome (PFS and OS,  $p=0.022$  and  $p=0.001$  respectively) and the loss of BAF47 immunoeexpression appeared as a significant independent prognostic factor (PFS,  $p=0.039$ ). A tendency for a better outcome (OS and PFS) was observed in case of complete removal compared to incomplete removal, especially when it was obtained before radiotherapy ( $p=0.16$  and  $p=0.37$  for OS and PFS before radiotherapy,  $p=0.27$  and  $p=0.43$  for OS and PFS after radiotherapy, respectively).

**CONCLUSIONS:**We identified clinical, histopathological and immunohistochemical parameters correlated to pediatric chordomas prognosis. A new grading system combined with the quality of surgical resection could help in

stratifying patients to postpone radiotherapy in case of low risk. Targeted therapy (EGFR, VEGF and STAT3 pathways) and re-irradiation may be discussed as potential therapeutic strategies.

**Keywords:** chordomas, histopathological grading, prognostic factors, management

Wednesday, 26 October 2016

10:50 – 11:55

## Platform presentations 8: Neuro-oncology

### PF-053

#### Neuro-oncology

##### The NOPHO-European study on cerebellar mutism syndrome

Morten Wibroe<sup>1, 2</sup>, Shivaram Avula<sup>3</sup>, Johan Cappelen<sup>4</sup>, Charlotte Castor<sup>5</sup>, Niels Clausen<sup>6</sup>, Irene Devenney<sup>7</sup>, Greg Fellows<sup>8</sup>, Pernilla Grillner<sup>9</sup>, Ramneek Gupta<sup>10</sup>, Bengt Gustavsson<sup>11</sup>, Mats Heyman<sup>12</sup>, Stefan Holm<sup>9</sup>, Atte Karpainen<sup>13</sup>, Rosita Kiudeliene<sup>14</sup>, Camilla Klausen<sup>15</sup>, Päivi Lähteenmäki<sup>16</sup>, Stephen Lewis<sup>17</sup>, Tuula Lönnqvist<sup>18</sup>, Conor Mallucci<sup>19</sup>, René Mathiasen<sup>1</sup>, Mattias Mattson<sup>20</sup>, Pelle Nilsson<sup>21</sup>, Kristiina Nordfors<sup>22</sup>, Per Nyman<sup>7</sup>, Karsten Nysom<sup>1</sup>, Karin Persson<sup>23</sup>, Jouni Pesola<sup>24</sup>, Barry Pizer<sup>25</sup>, Olof Rask<sup>26</sup>, Magnus Sabel<sup>27</sup>, Kjeld Schmiegelow<sup>1</sup>, Astrid Sehested<sup>1</sup>, Ingrid Tonning-Olsson<sup>5, 28</sup>, Ingrid Kristin Torsvik<sup>29</sup>, Kirsten van Baarsen<sup>30</sup>, David Walker<sup>31, 32</sup>, Mia Westerholm-Ormio<sup>18</sup>, Barbara Zetterqvist<sup>33</sup>, Marianne Juhler<sup>2</sup>

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20. Department of Pediatrics, University Hospital of Umeå, Umeå, Sweden
21. Department of Neurosurgery, Uppsala University Hospital, Uppsala, Sweden
22. Department of Pediatrics, Tampere University Hospital, Tampere, Finland

23. BarnReHab Skåne, Lund, Sweden
24. Department of Pediatrics, Kuopio University Hospital, Kuopio, Finland
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26. Department of Pediatrics, Skåne University Hospital, Malmö, Sweden
27. Department of Pediatrics, Queen Silvia Children's Hospital, Sahlgrenska University hospital, Göteborg, Sweden
28. Department of Psychology, Lund University, Lund, Sweden
29. Department of Pediatrics, Haukeland University Hospital, Bergen, Norway
30. Department of Neurosurgery, Radboud University Medical Centre, Nijmegen, The Netherlands
31. Children Brain Tumour Research Centre, Nottingham, UK
32. HeadSmart Be Brain Tumour Aware, Nottingham, UK
33. Department of Clinical Intervention and Technique, Karolinska University Hospital, Stockholm, Sweden

**OBJECTIVE:**The cerebellar mutism syndrome (CMS) is one of the most disabling late effects after neurosurgery for a posterior fossa tumour in childhood. The reported incidences vary substantially in previous studies. The pathophysiology is unknown, but damage to cerebello-thalamo-cerebral circuits is likely. The study focuses on the risk factors for development and severity of CMS including surgery (approaches, techniques and tissue and vascular damage, re-operation) and host genome variants.

**MATERIAL-METHODS:**Multicentre study developed as a NOPHO collaborative study coordinated from Rigshospitalet, Copenhagen with online data registration and database management at Karolinska, Stockholm and quarterly online participant meetings. Registration includes clinical data and speech samples collected preoperatively and at four defined postoperative points for the subsequent 12 months. Therapy, including neurosurgery, is by local standards. A blood sample for genetic analysis is collected from all patients. Imaging is collected and reviewed centrally.

**RESULTS:**The study aims to recruit 500 children. It opened in five Nordic and Baltic countries during 2014/2015; in the Netherlands in February 2016 and will open in the UK during 2016. Two German centres will join in 2017. The target accrual of 500 patients will be reached by the end of 2018. As of March 2016, 70 patients have been included from 12 centres. Mutism has occurred in 7 cases.

**CONCLUSIONS:**The study will be the largest prospective international study on CMS to date, and the first one to 1) systematically register surgery, use of steroids, standardized speech samples and 2) to investigate the influence of host genome.

**Keywords:** Cancer, Brain tumour, Pediatric, Neurosurgery, Cerebellar Mutism Syndrome, Genetics

### PF-054

#### Neuro-oncology

##### Survival in pediatric medulloblastoma: a population-based observational study to improve prognostication

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**OBJECTIVE:** Medulloblastoma continues to be the most common form of deadly brain malignancy of childhood. The purpose of this study was to utilize a big-data approach to investigate the changing trends in incidence, prevalence, demographics and survival in a contemporary cohort of pediatric medulloblastoma.

**MATERIAL-METHODS:** Patients diagnosed with medulloblastomas (1973–2012) in the Surveillance, Epidemiology, and End Results Program database were included in the study. The primary outcome, overall survival, was ascertained using a time-to-event (TTE) measure. Variables included age at diagnosis, gender, race, state, year of birth, radiation therapy, extent of surgery, duration of follow-up or overall survival, and cause of death.

**RESULTS:** There were 1735 patients with a median (IQR) age at diagnosis of 7 (4–11) years, predominantly white children (n=1438, 83%) above that expected from the US population presuming no race predilection ( $\chi^2$  statistic = 21.6,  $p < 0.001$ ). The incidence and prevalence of pediatric medulloblastoma has remained stable over the past 4 decades. There is a critical time point at 1990 when the overall survival has drastically improved. In a contemporary cohort, the percentage of participants alive was 86%, 70% and 63% at 1, 5, and 10 years, respectively. Multivariate Cox-Regression model demonstrated Radiation (HR: 0.37; 95% CI: 0.30–0.46,  $p < 0.001$ ) and Surgery (HR: 0.42; 95% CI: 0.30–0.58,  $p < 0.001$ ) independently predict long-term outcome. The probability of mortality from a neurological cause is less than 5% in patients who are alive 8 years following diagnosis.

**CONCLUSIONS:** Overall survival has improved over the last 4 decades, with a critical improvement around 1990. The majority of patients survive, especially those alive 8 years following initial diagnosis. The likelihood of mortality secondary to medulloblastoma disease is rare following this time period. Big-data approaches to clinical data allows us to discover significant associations with extreme precision ultimately enhancing our understanding of disease behaviour.

**Keywords:** Medulloblastoma, Epidemiology, Survival, Time to Event, Population study, Prognosis

## PF-055

### Neuro-oncology

#### Repositioning disulfiram as a radio-sensitizer against atypical teratoid/rhabdoid tumor (AT/RT)

Seung Ki Kim<sup>1</sup>, Young Eun Lee<sup>1</sup>, Seung Ah Choi<sup>1</sup>, Pil Ae Kwak<sup>1</sup>, Kyu Chang Wang<sup>1</sup>, Ji Hoon Phi<sup>1</sup>, Ji Yeoun Lee<sup>1</sup>, Sangjoon Chong<sup>1</sup>, Kyeong Min Joo<sup>2</sup>, Kook Hee Yang<sup>3</sup>

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**OBJECTIVE:** Atypical teratoid/rhabdoid tumors (AT/RT) are one of the most malignant pediatric brain tumor with an average survival of less than 1 year. We reported the therapeutic effects of disulfiram (DSF), an irreversible inhibitor of aldehyde dehydrogenase (ALDH), against brain tumor initiating cells (BTIC) from AT/RT. The goal of this study is to evaluate anti-cancer synergistic effect of DSF and radiation

therapy (RT) combination against AT/RT and to confirm the potent radio-sensitizing activity of DSF on AT/RT cells.

**MATERIAL-METHODS:** The responses to combination of DSF and RT were determined by in vitro assay of clonogenic formation, DNA damage, autophagy and apoptosis, as well as protein expression using AT/RT cell lines and primary cultured cells. The therapeutic effect for short-term and long-term was confirmed using AT/RT orthotopic mouse model.

**RESULTS:** Combination of DSF and RT is effective in decreasing clonogenicity of AT/RT cells in vitro. A sensitizer enhancement ratio (SER0.5) of DSF was 1.21 ~ 1.58 for AT/RT cells. Combined treatment increased the protein expression of  $\gamma$ -H2AX and LC3B, whereas decreased the Survivin and BCL2. The combination treatment produced abundant  $\gamma$ -H2AX foci and strongly promoted the autophagic cell death in all AT/RT cells. In AT/RT mouse model, the combination treatment significantly reduced the tumor growth (\* $P < 0.05$ ) and prolonged the survival rate (\*\* $P < 0.001$ ) compared to single treatment.

**CONCLUSIONS:** Our results demonstrated that the combination therapy with DSF and RT has a synergistic therapeutic effect on AT/RT, suggesting a potential clinical application for AT/RT patients.

**Keywords:** Atypical teratoid/rhabdoid tumor, disulfiram, aldehyde dehydrogenase, radiation therapy

## PF-056

### Neuro-oncology

#### Expression of FAT1 and prognosis of children with medulloblastoma

Hao Li

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**OBJECTIVE:** Determine the value of FAT1 in predicting the prognosis of children with medulloblastoma.

**MATERIAL-METHODS:** The mutation patterns of FAT1 were determined by whole exome sequencing of medulloblastoma tissues from children. The level of FAT1 expression in these tumors was determined by immunohistochemistry. The role of FAT1 in medulloblastoma was studied by RNAi while the specific inhibitor of Wnt/beta-catenin pathway was employed to analyze the mechanism of FAT1. Kaplan-Meier analysis was used to assess the value of FAT1 in predicting the prognosis of children with medulloblastoma.

**RESULTS:** Whole exome sequencing of 26 medulloblastoma tissues indicated mutations in more than one disease-related gene. Eight tissues had missense mutations in FAT1 that transformed it into an oncogene. Fluorescence quantitative PCR and immunohistochemical assays showed that expression of FAT1 in tumors was significantly lower than in normal brain tissue, and that down-regulation of FAT1 expression was associated with enhanced proliferation of medulloblastoma cells. Moreover, the expression of four genes including DKK, LEF1, beta-catenin, and cyclin D1 was significantly upregulated in the shFAT1-Daoy cells. But when the shFAT1-Daoy cells were treated with LGK-974, specific inhibitor of beta-catenin, expression of beta-catenin decreased greatly. In all, FAT1 probably function as a suppressor in medulloblastoma cells Daoy through Wnt/beta-catenin pathway. The immunohistochemistry results indicated that FAT1 expression was low in 26 tumors and high in 14 tumors. The overall survival of patients with high FAT1 expression (36 months) was significantly longer than that of patients with low FAT1 expression (12 months).

**CONCLUSIONS:** High expression of FAT1 in the medulloblastoma tissues of children correlated with longer overall survival. And Wnt signaling pathway play an important role in the FAT1 function.

**Keywords:** children medulloblastoma whole-exome sequencing FAT1 prognosis

## PF-057

## Neuro-oncology

**High survival rate in pediatric medulloblastoma with risk-adapted combined chemotherapy and reduced craniospinal radiotherapy**

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**OBJECTIVE:**Although multimodal treatment including surgical resection, radiotherapy, and chemotherapy has led to an improvement of long-term outcomes in pediatric patients with medulloblastoma, treatment-related toxicity often has a major impact on long-term quality of survival. To improve long-term survival and to reduce treatment-related toxicity, we developed novel adjuvant protocol including combined chemotherapy with reduced-dose radiotherapy and risk-adapted protocols.

**MATERIAL-METHODS:**Seventeen patients with medulloblastoma (10 with standard-risk disease and 7 with high-risk disease) were treated at Okayama University Hospital between 1997 and 2015. After surgical resection, standard-risk patients were treated with one cycle of ICE (IFO, CBDCA, VP-16)/VCEC (VCR, CPA, VP-16, CDDP) followed by craniospinal irradiation and 2 cycles of ICE/VCEC. Treatment of high-risk patients consisted of 2 to 3 cycles of ICE/VCEC followed by craniospinal irradiation and 2 cycles of high dose ICE or TEC (Thiotepa, VP-16, CBDCA). Irradiation of the entire central nervous system (24 Gy for the whole brain and spine) was carried out, and a boost irradiation dose of 30 Gy was given to the extended local area to the patients older than 3 years old.

**RESULTS:**With a median follow-up of 102 months (range 8–211), 16 complete responses and one partial response were observed. Five-year survival rate of standard-risk and high-risk group were 90% and 86%, respectively. No patient died of treatment-related toxicities. One patient with standard-risk disease developed dissemination after completing chemoradiotherapy and died 24 months after surgery. One patient with high-risk disease died of AML 5 years after initial diagnosis. Treatment related-toxicity include 7 hypopituitarism and 6 hearing disturbance. All of the survivors had no neurocognitive deficit.

**CONCLUSIONS:**Postoperative combined chemotherapy and reduced radiotherapy with risk-adapted protocols may improve survival rates and decrease the treatment-related toxicity in pediatric patients with medulloblastoma.

**Keywords:** Medulloblastoma, chemotherapy, radiation,

## PF-058

## Neuro-oncology

**Analysis of prognostic factors and survival in pediatric pineoblastomas: a population-based study using the SEER database**

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**OBJECTIVE:**Pineoblastomas are rare, malignant, pineal region lesions that account for <0.1% of intracranial tumors and retain a poor prognosis. We investigated the clinicopathological factors associated with survival using a

nationwide cohort of patients registered in the Surveillance, Epidemiology, and End Results (SEER) cancer database.

**MATERIAL-METHODS:**Pediatric patients (age<19 years) diagnosed with pineoblastoma between 1973 and 2010 were identified using the SEER registry. Patient, tumor, and treatment characteristics, including age at diagnosis, sex, race, extent of surgical resection, and radiation were evaluated. Kaplan-Meier survival analysis and Cox proportional hazard models (CPHM) were used to examine the effect of variables on outcome.

**RESULTS:**119 patients (61 male and 58 female) were identified with a median age at diagnosis of 6 years. The median overall survival (mOS) was 73 months. Surgery was performed in 97 patients (81.5%) with 36 patients (30.2%) undergoing biopsy and 25 patients (21%) undergoing gross total resection (GTR). 85 patients (71.4%) received either adjuvant or radiotherapy alone. Univariate CPHM identified a markedly worse prognosis for children aged<5 years compared with older patients (hazard ratio, 6.64; 95% CI, 2.91–15.14; p<0.0001; mOS 16 months). Radiotherapy was an important predictor of overall survival (hazard ratio, 0.37; 95% CI, 0.214–0.645; p=0.0005; mOS 202 months). Sex or extent of resection did not reach this level of significance. Multivariate CPHM confirmed older age and radiotherapy as independent prognostic variables. In our experience, 4 patients (median age 6; range 1–10 years) between 2000 and 2013 were diagnosed with pineoblastoma. GTR and radiotherapy were performed in 75% of patients with similar outcomes seen in the SEER analyses.

**CONCLUSIONS:**Older age at diagnosis and radiotherapy were associated with improved survival in pediatric pineoblastoma patients. Surgery alone did not confer a survival benefit. Multicenter, international tumor databases have the potential to impact our understanding of rare brain tumors.

**Keywords:** pineoblastoma, SEER, prognosis

## PF-059

## Special topic: Molecular biology

**Molecular classification of medulloblastoma in Japan**

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<sup>9</sup>Department of Neuro-Oncology/Neurosurgery, International Medical Center, Saitama Medical University, Hidaka, Japan

**OBJECTIVE:**Recent intensive genomic and molecular biological analyses of medulloblastomas have revealed that they are at least classified in to four core subgroups: WNT, SHH, Group 3, and Group 4 based on difference in cytogenetics, mutational spectra, and gene expression signatures, as well as in clinical phenotypes and outcomes. This four-subgroup system of

medulloblastomas will become not only a novel prognostic marker but also lead to improved diagnosis and risk stratification systems in combination with metastatic, cytogenetics, and/or mutational statuses. To deal with this progress, we have formed the Japanese Pediatric Molecular Neuro-oncology Group (JPMNG) and initiated a clinical research project to establish a nationwide network of a molecular diagnosis system for pediatric brain tumors in Japan.

**MATERIAL-METHODS:** Fresh, fresh-frozen and/or formalin-fixed paraffin-embedded archived tissue specimens are collected. Optimal diagnostic methods are being set up to reliably and reproducibly classify them into molecular subgroups according to the consensus criteria. These include gene expression analysis using the NanoString nCounter system, immunohistochemistry, and DNA sequencing.

**RESULTS:** We have so far collected a total of 303 cases including 169 medulloblastomas and 134 ependymomas. Analysis using 149 medulloblastomas indicated that proportions of four core subgroups were WNT (12.1%), SHH (28.9%), Group 3 (15.4%) and Group 4 (43.6%), respectively. CTNNB1 mutations were found in 88.9% of WNT-MB, and TP53 mutations were identified in 17.6% of WNT-MB and 24.3% of SHH-MB. Mutation of TERT promoter was also found in 5 SHH tumors.

**CONCLUSIONS:** We expect that the JPMNG project will improve the molecular diagnosis of pediatric brain tumors, leading to more appropriate treatments and better clinical outcomes in Japan.

**Keywords:** medulloblastomas, molecular diagnosis, molecular subgroup, prognostic marker

## PF-060

### Neuro-oncology

#### Incomplete tumor resection and second surgery in pediatric patients with ependymoma treated within the HIT2000 trial

Gertrud Pascalias Kammler<sup>1</sup>, Kara Krajewski<sup>1</sup>, Manfred Westphal<sup>1</sup>, Katja Von Hoff<sup>2</sup>, Carsten Friedrich<sup>2</sup>, Stefan Rutkowski<sup>2</sup>, Jürgen Krauss<sup>4</sup>, Monika Warmuth Metz<sup>5</sup>, Rolf Dieter Kortmann<sup>3</sup>

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**OBJECTIVE:** Gross total resection is key in the treatment of pediatric ependymoma. The purpose was to evaluate the assessment of residual tumor and evaluate the impact on survival associated with second surgeries taking histological grading into account.

**MATERIAL-METHODS:** Patients 0-21 years diagnosed with non-metastatic intracranial ependymoma and treated within the prospective, multicenter HIT2000 trial were included. Pre- and postoperative imaging underwent prospective central neuroradiological review. Histological grading also underwent central review. Evaluation for second surgery was recommended whenever feasible independent of adjuvant therapy.

**RESULTS:** Of 291 patients included in the trial, central review of postoperative imaging was sufficient for further classification for 206 patients. 137 patients were classified as R0 postoperatively and 69 as R+ according to central neuroradiological review. Histological grade II Ependymoma was found in 19/137 R0 patients and 10/69 R+ patients; 118/137 R0 and 59/69 R+ patients had grade III Ependymoma after initial surgery. 24/69 R+ patients underwent second surgery before relapse, 11 before and 13 after onset of adjuvant therapy; 12/24 were

then R0 after a maximum of 3 procedures. 3-year OS for primary GTR was 92%±2% vs. secondary GTR at 89%±11% vs. persisting R+ at 69%±6% (p=0.001). 5-year OS for WHO °III tumors was 80 ±5% for primary GTR vs. 53±25% for secondary GTR vs 55±8% for persisting R+ patients; 5-year OS for WHO °II tumors was 88±12% for primary GTR vs. 35±26.4% for persisting R+ (n=1 patient with secondary GTR); (p =0.01). Residual tumor measured 10.7cm<sup>3</sup> ± 14.2 (corrected mean); range: 0.07-49 cm<sup>3</sup>. There is a strong tendency for best overall survival in patients with <1cm<sup>3</sup> residual tumor, possibly due to more intensive treatment regimens in this subgroup.

**CONCLUSIONS:** Gross total resection remains an important prognostic factor. Secondary GTR may confer a survival advantage, regardless of histological grading.

**Keywords:** ependymoma, surgical resection, overall survival

Wednesday, 26 October 2016

15:00 – 15:20

## Platform presentations 9: Neuro-oncology

### PF-061

#### Neuro-oncology

##### Role of surgery in pediatric intrinsic brain stem lesions: CCHE-57357 experience

Mohamed Ahmed El Beltagy<sup>1</sup>, Mohamed Sa'ad Zaghoul<sup>2</sup>, Madiha Awad<sup>3</sup>, Amal Refaat<sup>4</sup>, Hala Taha<sup>5</sup>, Amal Mosa'ab Abdelaziz<sup>6</sup>

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**OBJECTIVE:** Intrinsic brain stem lesions (BSLs) are among the most aggressive lesions that can affect the pediatric central nervous system (CNS). They commonly affect children and young adults under the age of twenty years. Unlike other CNS regions, surgery is not the first line of management in BSLs due to the critical location and associated surgical morbidities. The role of surgery in affecting or even changing the line of management in those critical lesions is questioned.

**MATERIAL-METHODS:** Retrospective evaluation of BSLs managed in Children's Cancer Hospital of Egypt (CCHE-57357) between July 2007 and July 2015. Two hundred and eighty five patients were radiologically diagnosed as intrinsic BSLs. MRI was the diagnostic imaging modality of choice in all cases. 37 patients (12.9%) out of the 285 BSLs showed atypical radiographic patterns and were assigned to Frameless stereotactic-guided biopsy.

**RESULTS:** Fourteen patients (37.8%) out of the thirty seven showed high grade glial tumor and received radiotherapy with mean follow up period of 6 months. While, twenty one patients (56.8%) showed low grade glial tumor with mean follow up period of 15 months; of these, thirteen patients received radiotherapy, five patients received chemotherapy and three were clinically

followed up. Two patients out of the thirty seven had non-malignant BSLs. The first patient underwent near total resection after initial biopsy proved tuberculous granuloma while the other patient had an epidermoid cyst. Table (1) The five years overall survival for BSLs is 22.3% whereas the event free survival is 18.7%.

Table (1) Management of Brain Stem Lesions

**CONCLUSIONS:** Surgery has a beneficial role in focal intrinsic brain stem lesions since different pathologies may be encountered as inflammatory and low grade pathologies which may decide further treatment lines.

**Keywords:** brain stem lesions, glioma, neurosurgery, radiotherapy, stereotactic biopsy

## PF-062

### Neuro-oncology

#### Robot-assisted multi-catheter chronic intermittent convection enhanced drug delivery for the treatment of paediatric diffuse intrinsic pontine glioma: surgical case series of 10 patients

William GB Singleton<sup>1</sup>, Neil U Barua<sup>1</sup>, James A Morgan<sup>4</sup>, Owen Lewis<sup>2</sup>, Dave Johnson<sup>2</sup>, Max Woolley<sup>2</sup>, Richard J Edwards<sup>4</sup>, Stephen P Lewis<sup>3</sup>, Steven S Gill<sup>4</sup>

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**OBJECTIVE:** Treatment failure in diffuse intrinsic pontine glioma (DIPG) is in part due to poor CNS penetration of systemic chemotherapy at non-toxic doses. Convection enhanced delivery (CED) of chemotherapeutics to the paediatric brainstem through surgically placed micro-catheters overcomes the limitations of systemic treatment. We describe our surgical solution that facilitates accurate and reproducible drug delivery to the pons without the need for repeated surgical intervention.

**MATERIAL-METHODS:** 10 children (ages 4-12 years) have been treated in a compassionate treatment program of CED for radiological or biopsy proven DIPG using a multi-catheter intermittent infusion regime. All children underwent prone robot-assisted stereotactic 3T MRI & CT angiography guided implantation of 4 recessed step catheters using bilateral trans-frontal and trans-cerebellar trajectories. Infusion of drug was performed with the patients awake on the ward post operatively via an implanted transcutaneous port and repeated every 4-8 weeks. Drug distribution has been modelled using intra-infusion volume FLAIR and T2 3T MRI.

**RESULTS:** Of the 10 patients treated, there has been no surgical complication or post-operative morbidity. The surgical procedure is well tolerated, and all patients have been quick to recover. Surgical time is on average 5 hours (range 4-8). Infusion of drug has commenced as early as 3 hours after catheter and port implantation. Infusion volumes have ranged from 2.5-12 mls per single infusion. Transient neurological deficits have been seen during infusion, which have resolved prior to a repeat cycle of therapy.

**CONCLUSIONS:** Therapeutic volumes of drug to cover the paediatric pons can be achieved using a multi-catheter technique utilising a transcutaneous port that facilitates repeated drug delivery in a ward based setting. The procedure is well tolerated by children, and provides a stable platform for the investigation of direct administration of chemotherapy for the treatment of DIPG.

**Keywords:** Convection enhanced delivery, DIPG, Robotic Surgery, Brainstem, Stereotaxy.

Thursday, 27 October 2016

10:50 – 12:50

## Platform presentations 10: Hydrocephalus

### PF-064

#### Hydrocephalus

#### Viewing on hydrocephalus from bottom to top: what can we learn from real-time MRI?

Hans Christoph Ludwig<sup>1</sup>, Hans Christoph Bock<sup>1</sup>, Steffi Dreha Kulaczewski<sup>2</sup>

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<sup>2</sup>Department of Pediatric Neurology, University Medical Center Göttingen

**OBJECTIVE:** Contemporary hydrocephalus (HC) concepts are still derived mainly from the work of Dandy (1913) as well as Key and Retzius (1875). Based on few animal studies, they determined that 80% of CSF was produced in the choroid plexus and absorbed at the arachnoid villi. Cardiac gated flow MRI has further supported the idea of bulk flow and piston like CSF dynamics out of the ventricles towards the site of arachnoid absorption. However, these concepts are in contrast to the pathophysiology of many CSF related diseases such as normal pressure hydrocephalus, pseudotumor cerebri, Chiari malformation, arachnoid cysts, and spontaneous intracranial hypotension. Furthermore endoscopically gained visualisation of trapping mechanisms in various types of occlusive HC does not support the common notion of craniocaudal directed CSF movements. Recent studies applying Real-Time MRI (Dreha-Kulaczewski et al, 2015) have opened new insights into CSF dynamics induced by forced breathing and may revise our current understanding of disturbances of CSF flow.

**MATERIAL-METHODS:** Video analysis of intraoperatively documented trapping mechanisms during microsurgery or neuroendoscopy (N=260) in different types of occlusive HC have been conducted to reveal breathing related CSF movement and dynamics. The mechanisms of trapping were investigated for underlying mechanical fluid forces and flow characteristics. These data were related to recent concepts of CSF movements and underlying driving forces.

**RESULTS:** Trapping mechanisms could be identified in nearly all types of occlusive HC. Intraoperative documented CSF flow dynamic was mainly related to ventilated breathing and followed a spinal to cranial directed movement.

**CONCLUSIONS:** Most types of occlusive HC revealed trapping of CSF moving upwards from spinal canal to the ventricles or into cystic compartments. These observations are supported by results of recent Real-Time MRI studies and might change our common concept of the underlying pathophysiology of different subtypes of hydrocephalus.

**Keywords:** Hydrocephalus, Trapping, Real Time MRI, CSF

### PF-065

#### Hydrocephalus

#### Using MRI to establish patency between adjacent CSF compartments

J. Gordon McComb<sup>1</sup>, Edward Frederick Melamed<sup>1</sup>, Skorn Ponrartana<sup>2</sup>, Eisha Christian<sup>1</sup>, Stefan Bluml<sup>2</sup>, Mathew Borzage<sup>2</sup>

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**OBJECTIVE:** The ability to establish patency of cerebral spinal fluid (CSF) flow between adjacent central nervous system (CNS) compartments is of importance in the diagnosis and treatment of patients with CSF flow obstruction in various areas. The method described was originally derived from arterial spin labeling (ASL) of blood flowing in vessels. This technique, referred to as

time-spatial labeling inversion pulse (time-SLIP), was further modified to non-invasively visualize CSF pulsatile and turbulent flow between two regions of interest.

**MATERIAL-METHODS:**The presence of CSF flow was examined at the foramen Magnum (FMag), the aqueduct of Sylvius (AS), the foramen of Magendie (FMgd), the floor of the third ventricle (3rd V), and at the foramen of Monro (FM). The studies were compared with clinical information and classified as true positive, true negative, false positive, and false negative based on expectation of patency.

**RESULTS:**A total of 83 studies were done on 51 patients. Among positive readings of the FMag, AS, 3rd V, and FM, 49/49, 40/40, 15/15, and 9/9 were true, while 3/5, 20/30, 2/5, and 0/2 negative readings were true, respectively. The sensitivity and specificity percentages of the technique at the FMag, AS, 3rd V, and FM were, respectively, 96, 100; 80, 100; 83, 100, and 82, undefined. Negative likelihood ratios for these sites were, respectively, 0.04, 0.20, 0.17, and undefined.

**CONCLUSIONS:**Establishing qualitative patency between adjacent CSF compartments using MRI is possible with a modified ASL technique. The freely selectable tag allows CSF flow to be visualized in any direction or location. This technique can be used post-endoscopic third ventriculostomy to evaluate the patency of the fenestration in addition to evaluating pre- and postoperative craniocervical decompression changes to CSF flow. This technique has excellent (above 80%) sensitivity, specificity, and negative likelihood ratio at all sites studied.

**Keywords:** MRI, Cerebrospinal Fluid, CSF Flow, Hydrocephalus, time SLIP, arterial spin labeling

## PF-066

### Hydrocephalus

#### Prediction of shunt failure facilitated by rapid (<40 sec) and accurate volumetric analysis

Tushar Jha<sup>1</sup>, Zakia Barnawi<sup>3</sup>, Mark Quigley<sup>2</sup>, Tiffani Defreitas<sup>2</sup>, John Myseros<sup>2</sup>, Suresh Magge<sup>2</sup>, Chima Oluigbo<sup>2</sup>, Robert Keating<sup>2</sup>

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**OBJECTIVE:**Determination of shunt failure is fraught with numerous difficulties, particularly when there is lack of meaningful change in the size of the ventricles(5% of patients). Currently, obtaining accurate volumetrics of CSF spaces offers the best chance for an objective assessment of shunt malfunction. Historically this process has been time consuming and required considerable expertise, effectively limiting it's usefulness as a routine tool. We set out to develop a rapid and accurate means of measuring volumetrics of CSF spaces, to facilitate timely and accurate diagnosis of shunt malfunctions.

**MATERIAL-METHODS:**CT scans were analyzed retrospectively in 26 shunt patients at CNMC (Washington,DC) employing manual and automated methods to measure ventricular volume. Manual measurements were produced using OsiriX software whereas automated measurements were produced using proprietary software. Variables assessed included time taken for analysis and accuracy of measured volume between multiple users. CSF volume assessment was also correlated with patients who eventually underwent shunt revision to assess whether there was any predictive value with respect to shunt failure.

**RESULTS:**Using an improved proprietary algorithm, average time of automated analysis of ventricular volume was 38.3 vs 2244 secs for the manual

technique. Correlation between three independent examiners was >99%. The standard deviation between both techniques was +/- 1.43%. 4/26 patients had unchanged CT's at the time of shunt evaluation and 3/4 of these patients demonstrated meaningful increase(8-10%) in their ventricular volume despite being read unchanged by radiology.

**CONCLUSIONS:**Ventricular volumes of CT scans in pediatric patients with hydrocephalus can be measured with efficacy and accuracy with the novel automated software presented here. The application of this technology is especially valuable in patients who present with symptoms of potential shunt failure without any recognizable increase in radiographic ventricle size. In such cases, this software may provide timely information that may prevent delay in treatment, and potential medical misadventure

**Keywords:** shunt failure, hydrocephalus, automated volumetrics

## PF-067

### Hydrocephalus

#### Differences in intracranial pressure seen in children and adults could be caused by age differences

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**OBJECTIVE:**In neurosurgery, one aim of treatment is normalization of intracranial pressure (ICP). No studies compare ICP measurements in children to measurements in adults and it seems silently assumed that reference values are the same, regardless of the physiological differences.

This study AIMS:

- To clarify if day and night ICP differs between children and adults
- To examine if age affects ICP

**MATERIAL-METHODS:**We analysed data from all non-shunted patients with suspicion of hydrocephalus or idiopathic intracranial hypertension (IIH), undergoing invasive elective diagnostic 24 hours ICP monitoring from February 2008 to November 2014. 130 patients were included. In this abstract we compare younger children (n=41, mean age=6.6 years, range 1;12) with adults (n=71, mean age=50.1 years, range 18;85).

We separated data into day and night sequences and determined the mean value for both sequences.

**RESULTS:**We found a nocturnal ICP increase (p<0.0001) in 95% of the patients. The mean intrapersonal difference in  $\Delta$ ICP (for day and night pressure) was nearly identical in children (6.6 mmHg) and adults (6.3 mmHg) p=0.8. However, there were noteworthy differences between the groups;

- Children had both higher mean ICPday (7.0 versus 2.5 mmHg, p<0.0001) and mean ICPnight (12.3 versus 8.9 mmHg, p=0.001) than adults.
- In adults both ICPday and ICPnight decreased with age (decrement of 0.1 mmHg per year). In comparison, children presented almost ten times the age related decrement.

**CONCLUSIONS:**Both groups showed a nocturnal ICP increase. Measured ICP values were however, higher in children than in adults both day and night. ICP could furthermore be described as a linearly decreasing function of age, with a remarkably steeper slope from age 1 to 12.

This could indicate that ICP reference values for children should be different than those for adults.

The results are based on a pseudo normal study population and contribute to an estimation of normal human physiology.

**Keywords:** intracranial pressure, pediatric reference values, normal human physiology, hydrocephalus

**PF-068****Other****Cerebrospinal fluid leakage in childhood and adolescence**

Koichi Takahashi  
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**OBJECTIVE:**Cerebrospinal fluid (CSF) leakage has increasingly been recognized, but few investigations have examined younger patients. This study investigated 129 patients with CSF leakage of onset in childhood or adolescence. The causes, outcomes, and clinical features of these patients were analyzed.

**MATERIAL-METHODS:**The 129 patients (65 boys, 64 girls) had an onset earlier than age 15. Diagnosis of possible CSF leakage was made based on clinical symptoms and findings of magnetic resonance imaging (MRI) and radioisotope (RI) cisternography. Indications for an epidural blood patch (EBP) were presence of parathecal activity (direct evidence) or decreases in RI residual activity without clear detection of parathecal activity (30% at 24 h after RI injection) (indirect evidence). Statistical analysis of the outcomes between patients with direct evidence and patients with indirect evidence on RI cisternography were performed.

**RESULTS:**All patients experienced one or more symptoms such as intractable headache, nausea, vertigo, fatigue, neck pain and sleep disturbance. In RI cisternographic findings, 68 patients (52.7%) had direct findings and 61 patients (47.3%) had indirect findings. Regarding outcomes, 93.0% of patients showed good or moderate recovery after treatment with an epidural blood patch (EBP). There were no significant differences in the outcomes of direct versus indirect evidence on RI cisternography.

**CONCLUSIONS:**CSF leakage is not yet well recognized, particularly in child and adolescent patients. This study indicates that EBP is also effective for younger patients. If patients experience various intractable symptoms such as headache, nausea, vertigo, and so on, CSF leakage should be considered in the differential diagnosis.

**Keywords:** cerebrospinal fluid leakage, intracranial hypotension syndrome, cerebrospinal fluid hypovolemia, pediatrics, epidural blood patch

**PF-069****Other****Altered cerebrospinal fluid dynamics in neurofibromatosis 1: severe arachnoid thickening, which can also be caused by asymmetrical CSF pressure through sphenoid defect, may cause abnormal CSF dynamics**

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**OBJECTIVE:**The object of the study was to understand abnormal dynamic of CSF in patient with neurofibromatosis 1, which may cause temporal lobe herniation and bulging of temporal fossa.

**MATERIAL-METHODS:**Four Patients, three females and one boy, with neurofibromatosis type 1 (NF1) presented with similar craniofacial deformity, which consisted of pulsatile exophthalmos, an enlarged bony orbit, orbital neurofibroma, dysplasia of the sphenoid wing with the presence of a herniation of the temporal lobe into the orbit, and a bulging temporal fossa.

IICP signs were found in all 4 patients. And abnormally thickened arachnoid membrane, which could not be fenestrated for combination with the basal cistern, was seen in one patient during the operation.

**RESULTS:**Protruding temporal lobe, which was one of the main symptoms in NF 1 patients, could be stopped by control of IICP via programmable ventriculoperitoneal shunt (VPS) implantation or extra ventricle drainage in three patients. The dense fibrosis of arachnoid membrane and consequent altered hemispheric cerebrospinal fluid dynamics may IICP in NF patient. Such IICP may consequently cause temporal lobe herniation and the orbitotemporal deformity.

**CONCLUSIONS:**There is a tendency to severe fibrosis of arachnoid membrane in patients with NF, which might cause abnormal CSF dynamic. This mechanism may not present with general findings of hydrocephalus, which often can delay diagnosis. Coexisting sphenoid dysplasia may aggravate abnormal CSF dynamic. In such cases, before considering any surgical reconstruction, the control of ICP via shunting should be primarily considered.

**Keywords:** Neurofibromatosis, sphenoid dysplasia, orbito-temporal deformity, high intracranial pressure

**PF-070****Hydrocephalus****Endoscopic third ventriculostomy in infants younger than 6 months old: rate of success and prognostic factors**

Carlos Bennett<sup>1</sup>, Andres Horlacher<sup>2</sup>, Manuel Gonzalez<sup>2</sup>, Julio Meza<sup>2</sup>, Nicole Loyola<sup>2</sup>, Rodrigo Riveros<sup>2</sup>, Cox Pablo<sup>2</sup>

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**OBJECTIVE:**To determine the success rate in infants less than 6 months olds operated with endoscopic third ventriculostomy for hydrocephalus, and to identify risk factors for failure of the procedure or predictive factors for success.

**MATERIAL-METHODS:**We analyzed a retrospective cohort of 26 infants less than 6 months old patients operated with endoscopic third ventriculostomy (ETV) at a single institution. Success rate was defined as no need of a shunt device during follow-up. Variables evaluated included: age, etiology of the hydrocephalus, morphology of the hydrocephalus, presence of previous shunt, dilatation of frontal subarachnoid space, bulging of the floor of the third ventricle and experience of the neurosurgeon.

In addition we performed a systematic review of the literature and meta-analysis of the subset of infants less than 6 months old operated with ETV across different studies.

**RESULTS:**There were 17 patients with aqueductal stenosis, 7 patients with posthemorrhagic hydrocephalus, 2 patients with Dandy Walker malformation. 52% of patients were free of shunt at the end of follow up. Etiology was not a prognostic factor, but triventricular morphology was significant in predicting freedom of shunt. Infants with more than 1 cm of frontal subarachnoid space had worse results (72% of failure). In our systematic review and meta-analysis, we found 235 patients across different studies, and overall success rate was 56,17%. The experience of the surgeon was a significant prognostic factor. There were contradictory reports regarding etiology.

**CONCLUSIONS:**Contrary to some reports, we found in our series that age of the patient does imply a higher risk of ETV failure, independent of etiology. The experience of the surgeon, morphology of the hydrocephalus and the size of the subarachnoid space should be taken into account in decision making process. The ETV success score does not seem to predict well the possibility of failure in this particular age group.

**Keywords:** endoscopic third ventriculostomy, young infants

## PF-071

## Hydrocephalus

**Effective timing for myelomeningocele repair and simultaneous ventriculoperitoneal shunting**

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**OBJECTIVE:**The prenatal diagnosis of myelomeningocele (MMC) has increased. In MMC with ventriculomegaly, the optimum timing for ventriculoperitoneal (V-P) shunting is still controversial. We retrospectively evaluated cases in which MMC repair and V-P shunting were performed simultaneously on the day of birth, and compared the outcomes with cases in which the surgery was performed on a different postnatal day.

**MATERIAL-METHODS:**We reviewed 41 cases that were diagnosed prenatally with MMC with hydrocephalus and were treated at our institutions since 2000. The cases were divided according to whether the newborns underwent simultaneous MMC repair and V-P shunting on the day of birth (group A) or on a different postnatal day (group B). The timing of surgery, central spinal fluid (CSF) leakage from the surgical site, length of hospitalization, and rates of shunt occlusion, shunt infection, and symptomatic type II Chiari malformation were compared between the two groups.

**RESULTS:**In group A (19 cases), MMC repair and V-P shunting were performed immediately after caesarean section. In group B (22 cases), MMC repair and V-P shunting were performed one day after the caesarean section in three cases, and on a later day in 19 cases. CSF leakage from the surgical wound was not observed in group A (0%) but was present in 5 cases in group B (22.7%). In groups A and B, shunt occlusion occurred in 42.1% and 45.5% of the cases, respectively; shunt infection occurred in 0% and 31.8%, and symptomatic type II Chiari malformation requiring foramen magnum decompression occurred in 10.5% and 18.1%. The average length of hospitalization was 49 days and 105, respectively.

**CONCLUSIONS:**Performing simultaneous MMC repair and V-P shunting immediately after delivery reduced the risk of CSF leakage, shunt infection, and symptomatic type II Chiari malformation, and shortened the hospital stay.

**Keywords:** myelomeningocele, hydrocephalus, simultaneous operation

## PF-072

## Hydrocephalus

**Rate of shunt revision as a function of age in a population of myelomeningocele patients with shunted hydrocephalus**

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**OBJECTIVE:**It is generally accepted that cerebrospinal fluid shunts fail most frequently in the first years of life. The purpose of this study is to describe the risk of shunt failure for a given patient age in a well defined cohort of patients with shunted hydrocephalus secondary to myelomeningocele.

**MATERIAL-METHODS:**We analyzed data from our institutional spina bifida (SB) research database including all pediatric patients with myelomeningocele and shunted hydrocephalus. For our entire population, we determined the number of shunt revisions in each year of life. We then calculated the number of patients at risk for shunt revision during each year of life, thus enabling us to calculate the rate of shunt revision per patient in each year of life. In this way, we are able to evaluate the timing of all shunt revision operations for our entire clinic population and evaluate the likelihood of having a shunt revision during each year of life.

**RESULTS:**A total of 655 patients are enrolled in our SB research database. 408 of these have a diagnosis of myelomeningocele. 333 patients have had at least one shunt revision and thus are included in this study. The mean age is 14.2 + 7.2 years (median 14, range 0-34 yrs). There were 94 shunt revisions in the first year of life. This represents a rate of 0.28 revisions per patient year. Rate of shunt revision per patient year decreases with increasing age, with the exception of a increase in frequency in the early teen years.

**CONCLUSIONS:**These data substantiate the idea that shunt revision surgeries in myelomeningocele patients become less likely as a patient ages. We are able to estimate the risk of shunt failure for a given myelomeningocele patient as a function of age. This will be useful for counseling patients and families.

**Keywords:** Hydrocephalus, myelomeningocele, shunt, aging, shunt revision

## PF-073

## Hydrocephalus

**Incidence of ventriculoperitoneal shunt failure in children less than one year of age: observational review of prospectively collected data**

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**OBJECTIVE:INTRODUCTION:** Ventriculoperitoneal (VP) shunt failure is more common in patients less than one-year-old. This study presents the results obtained over one year of data collection as a single center experience.

**MATERIAL-METHODS:METHODODOLOGY:** All patients performed hydrocephalus (HC) related surgery in our institute were gathered starting from April 2015 till March 2016. Collected data in the manual and electronic-based data bank were studied prospectively aiming firstly to detect the incidence of shunt failure in this age group and secondly to analyze the different patterns of failure.

**RESULTS:RESULTS:** In the period starting from April 2015 till end of March 2016, among 458 consecutive patients collected, 203 were less than one year of age at the time of surgery and underwent VP shunt as the primary treatment for HC, males represented 129 cases while females represented 74 cases of patients, the mean age was approximately five months.180 cases had communicating and 23 cases had non communicating HC. 46 cases required revisions with an overall incidence of 22.8%. Seven cases required replacement of the shunt system. An alternative to simple peritoneal implantation was performed in 22 cases; among these four were inserted in the right atrium; and 12 in the gall bladder. In two cases laparoscopic revision was performed. The mean number of revisions was 3.68 with minimum of one revision per case and the maximum number of 25 revisions. Nine cases were complicated by exposure, 19 were complicated by infection. Ten patients died during perioperative period and one case was lost in follow up thus excluded from the study.

**CONCLUSIONS/CONCLUSION:** Age at the first VP shunt insertion remains to be a major factor in predicting the incidence of shunt failure. Close monitoring and early detection of shunt failure represent the keystone for management of such common complication in HC.

**Keywords:** Hydrocephalus, shunt failure, shunt infection

#### PF-074

##### Hydrocephalus

#### A classification proposal for “ventriculostoma reclosure pattern” based on experience in 50 pediatric cases with repeat ETV procedure

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**OBJECTIVE:**The reclosure of the ventriculostoma is one of the major causes of failed ETV procedure in children. The present study investigates the types and features of ventriculostoma closure patterns and suggests a classification for ventriculostoma reclosure subtypes based on the observations of 50 repeat-ETV procedures in pediatric cases.

**MATERIAL-METHODS:**The data of 64 pediatric triventricular hydrocephalus cases which have undergone repeat ETV for the failure of initial ETV procedure due to ventriculostoma closure were retrospectively analyzed. Besides clinical signs and symptoms, verification of the ventriculostoma failure was confirmed with cine phase-contrast magnetic resonance imaging, which is regarded as an accurate method of detecting postoperative fenestration patency. Patient records on clinical features and video recordings of the endoscopic procedures were reviewed in detail. The cases with a history of intraventricular hemorrhage, CSF-infection or CSF-shunt surgery were not included. The cases with incompletely penetrated membranes during the initial ETV procedure were also excluded from the study.

**RESULTS:**In 50 cases which have met the above criteria, ventriculostoma reclosure patterns were classified according to the features of the reclosed region. Three major types of ventriculostoma closure were described: Type-1: Closure of ventriculostoma with gliosis or scar tissue that results in a non-translucent/opaque third ventricle floor; Type-2: Narrowing/closure of ventriculostoma by translucent membranes; Type-3: Patent ventriculostoma but CSF flow is blocked by newly formed membranes in the basal cisterns.

**CONCLUSIONS:**This classification may be useful in further studies which will intend to investigate success rate of the repeat-ETV procedure in pediatric cases in detail.

**Keywords:** classification, ETV, reclosure, ventriculostoma

#### PF-075

##### Hydrocephalus

#### The role of endoscopic third ventriculostomy in previously shunted children admitted with shunt malfunction

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**OBJECTIVE:**Endoscopic third ventriculostomy (ETV) is a routine and safe procedure for therapy of obstructive hydrocephalus. The role of endoscopic treatment in the management of shunt malfunction was rarely evaluated. The aim of our study is to evaluate the success rate of ETV with/without choroid plexus coagulation (CPC) in pediatric patients formerly treated by ventriculoperitoneal (V-P) shunt implantation.

**MATERIAL-METHODS:**We retrospectively reviewed the records of 30 consecutive patients with shunt malfunction/ infection who were treated at our institution with ETV/CPC.

**RESULTS:**30 patients with shunt failure, 20 males and 10 females, were enrolled with the mean age of 52.96 +/- 49.83 months (2 – 168) at the time of endoscopic treatment. There were 14 cases of shunt malfunction and 16 cases of shunt infection in this series. During postoperative assessments, 12 patients (40%) required new ventricular shunt implantation within a mean period of 24.4 +/- 22.7 days (10 – 95) from endoscopic procedure. 17 patients (56.7%) did well without need to any further intervention during a follow-up period of 6 – 48 months (18.05 +/- 10.22). One patient expired about 1 month after procedure.

**CONCLUSIONS:**The use of ETV with/without CPC in patients with shunt failure resulted in shunt independence in 56.7% of cases. Endoscopic procedures can be an alternative method of treatment in patients with former V-P shunt implantation.

**Keywords:** Shunt, Hydrocephalus, ETV, malfunction

#### PF-076

##### Other

#### Triclosan-coated sutures in preventing surgical site infection in children: a randomized controlled series

Willy Serlo<sup>1</sup>, Marjo Renko<sup>2</sup>, Niko Paalanne<sup>1</sup>, Terhi Tapaiaainen<sup>1</sup>, Matti Hinkanen<sup>1</sup>, Tytti Pokka<sup>1</sup>, Sohvi Kinnula<sup>1</sup>, Juha Jaakko Sinikumpu<sup>1</sup>, Matti Uhari<sup>1</sup>

<sup>1</sup>Oulu University Hospital

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**OBJECTIVE:**Surgical site infections (SSIs) remain a pervasive problem in all surgery. Triclosan coated sutures have been suggested to decrease the occurrence of SSIs in adult patients but the evidence of the efficacy is scanty especially in children.

**MATERIAL-METHODS:**We designed a randomized, double-blind, placebo-controlled trial among 1615 children younger than 18 years attending surgery. Participants were recruited between September 2010 and December 2014 among consecutive patients coming to operation theatre for surgery with a plan to use resorbable sutures. Children were randomly allocated to receive either triclosan-coated or non-coated resorbable sutures. The primary outcome was the occurrence of superficial or deep SSI according to the Centers for Disease Control and Prevention criteria within 30 days after the operation

**RESULTS:**In the intention-to-treat-analysis any SSI occurred in 2.8 % (22/791) of the patients allocated to triclosan-coated sutures and in 5.8 % (46/797) of the patients having non-coated sutures (proportion difference (D) 3.0 %, 95 % confidence interval (CI) from 1.0 to 5.1 %, P=0.003). To prevent one SSI, triclosan-coated sutures should be used in 33 children (CI 20 to 98). Deep SSIs were found in 0.4 % (3/791) of the patients in the triclosan group and in 2.0 % (16/797) in the control group (D 1.6 %, CI 0.6 to 2.9 %, P=0.004, RR 0.21, CI 0.06 to 0.67).

**CONCLUSIONS:**Triclosan-coated sutures were effective in preventing SSIs in children by 52 % and deep SSIs by 75 %. The effect was seen even among children with a low risk for SSIs.

**Keywords:** infection prevention, coated sutures

#### PF-078

##### Other

#### The role of mixed reality simulation in neurosurgical education

Giselle Coelho Caselato<sup>1</sup>, Benjamin Warf<sup>2</sup>, Nelci Zanon<sup>1</sup>

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<sup>2</sup>Department of Pediatric Neurosurgery - Boston Children Hospital / Harvard Medical School, Boston, United States of America

**OBJECTIVE:**The Neurosurgical education is a long, laborious process, that requires many years of supervised, hands-on training. The development of surgical simulation platforms is therefore essential to reducing the risk of potentially serious intraoperative errors arising from inexperience. The main goal is to propose a new tool for neurosurgical education, associating virtual and realistic simulation (mixed reality), for craniostyosis correction (scaphocephaly type) and neuroendoscopic training.

**MATERIAL-METHODS:**Tridimensional videos were developed by 3DS Max program. The simulated approaches were: the Renier's H technique for craniostyosis correction, choroid plexus cauterization and endoscopic thirdventriculostomy. The physical simulators were made with a synthetic thermo-retractile and thermo-sensible rubber which, when combined with different polymers, produces more than 30 different formulas. These formulas present textures, consistencies and mechanical resistance similar to many human tissues.

**RESULTS:**The virtual and physical simulators for craniostyosis and neuroendoscopy were approved by an expert surgery team. The proportion of the answers was estimated by the confidence intervals.

**CONCLUSIONS:**The experts conclude that this virtual simulation provides a highly effective way of working with 3D data and it significantly enhances teaching of surgical anatomy and operative strategies in neurosurgical field. A mixed simulation provided the desired results of both physical and virtual simulators in achieving the psychomotor and cognitive apprenticeship. The combination of these tools may potentially improve and abbreviate the non experienced learning curve, in a safe manner.

**Keywords:** simulation; physical; virtual; craniostyosis; hydrocephalus; education

**PF-079**

**Other**

### Publication rate of oral presentations at the International Society for Pediatric Neurosurgery (ISPN) annual meetings

Christopher Bonfield

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**OBJECTIVE:**The International Society for Pediatric Neurosurgery (ISPN) meeting serves as an important forum to present, discuss, and disseminate current academic research. One ultimate aim of abstracts presented at these meetings is to be published in full in a peer-reviewed journal. The purpose of this study was to investigate the publication rates of oral presentations at the ISPN annual meeting in peer-reviewed journals.

**MATERIAL-METHODS:**All oral presentations from the 2009, 2010, and 2011 ISPN annual meetings were reviewed. Author and title information were then used to search PubMed to identify those abstracts that had been published in peer-reviewed journals. The title of the journal, year of publication after the meeting, and the country of origin of the authors were also recorded.

**RESULTS:**In total, 163 of 401 oral presented abstracts (40.6%) were found to have been published. The vast majority (95.1%) were published within 4 years of presentation at the meeting. *Child's Nervous System* (29.4%), *Journal of Neurosurgery: Pediatrics* (14.1%), and *Neurosurgery* (8.6%) were the three most common journals for published abstracts. The United States of America, the Republic of Korea, and India produced the most published articles.

**CONCLUSIONS:**Oral presentations at the annual ISPN meetings have an overall publication rate of 40% in peer-reviewed journals, which is comparable to other national and international neurosurgery meetings. This illustrates the high quality of papers presented at the meeting and supports the ISPN as being a strong platform for excellent current research in neurosurgery.

**Keywords:** International Society for Pediatric Neurosurgery, annual meeting, publication rate, oral presentation, abstract

## FLASH PRESENTATIONS

Monday, 24 October 2016

09:32 – 10:00

### Flash session 1: Fetal diagnosis and management

**FL-001**

**Special topic: Fetal diagnosis and management**

**Fetal closed spinal malformations: detection and outcome**

Liana Adani Beni<sup>1</sup>, Moshe Brinsein<sup>2</sup>, Ygal Wolman<sup>3</sup>, Yuval Oz<sup>4</sup>

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<sup>4</sup>Ziv Medical Center, Zafed, Israel

**OBJECTIVE:**Fetal ultrasound has improved tremendously in the last two decades. Still, fetal diagnosis of tethered cord (TCS) or Closed Spinal Dysraphism (CSD) is not common. Moreover, fetal diagnosis of TCS may lead, in some countries to a large number of terminations of pregnancies, and even though it is a diagnostic challenge there is an important question of what would be the implication of improved diagnosis of TCS on the pregnancies.

The aim of this report was to summarize our experience together with the ultrasonographers, in a pilot that was conducted in order to see whether it is possible to better detect fetal Closed Spinal Dysraphism (CSD) / TCS and whether detection of such cases will impact the outcome of pregnancies.

**MATERIAL-METHODS:**With collaboration of several centers of OBG and ultrasonographers, US detection of level of conus was attempted. In cases of fetal diagnosis of TCS, parents were referred to senior author for fetal counseling.

**RESULTS:**We present our experience in fetal counseling for Fetal CSD diagnosed between 20-36 weeks of gestation, the implications of such diagnosis on the pregnancies reported, and the outcome of the babies that were born. The challenge of fetal counseling and the medicolegal aspects together with the psychological impact on the future parents are also presented.

**CONCLUSIONS:**The attempt to diagnose level of conus medullaris led to diagnosis of TCS in some cases. Fetal MRI was indicated for most pregnancies, in order to characterize better the congenital spinal malformation and to exclude additional malformations of the CNS. Attempt to define the level of conus may improve detection of fetal TCS/CSD however whether it is still to be determined if fetal diagnosis is of benefit in such cases.

**Keywords:** Fetal, Prenatal, Closed spinal dysthaphism, outcome, TCS

**FL-002**

**Special topic: Fetal diagnosis and management**

**Three-dimensional modeling of fetal myelomeningocele**

Michael H Handler<sup>1</sup>, Maggie M Hodges<sup>2</sup>, Jennifer L Wagner<sup>3</sup>, David M Mirsky<sup>4</sup>, Ahmed Marwan<sup>5</sup>, Robin Shandas<sup>5</sup>, Timothy M Crombleholm<sup>5</sup>, Kenneth W Liechty<sup>5</sup>

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<sup>5</sup>Department of Surgery, Children's Hospital Colorado and University of Colorado, Aurora, CO, USA

**OBJECTIVE:**Open fetal surgery to close myelomeningoceles (MMC) has become widely accepted since the successful completion of the MOMS trial. The occurrence of premature delivery and fetal well-being are directly related to the length in time of the fetal repair. When primary closure is precluded, a biologic patch may be incorporated, lengthening the procedure. We postulate that preoperative modeling of the MMC defect utilizing three-dimensional (3-D) imaging and 3-D printing technology will improve operative times, by predicting the need for a patch and providing a template to create one preoperatively.

**MATERIAL-METHODS:**We identified 9 myelomeningoceles that were candidates for fetal repair. Standard 3-D imaging sequences were acquired in addition to the routine magnetic resonance Imaging (MRI) protocol. DICOM images were converted into stereolithography files. During the process, 3-D surfaces were converted to patterns allowing 3-D printing using acrylonitrile butadiene styrene. These models were then used to predict the need for patch reconstruction in fetal MMC closure, plan the operative repair, and create a sterile template to prepare the patch early intraoperatively.

**RESULTS:**3-D modeling accurately predicted the size of the MMC defect. In the 6 patients where an Alloderm patch was required, preoperative modeling closely projected the ideal patch size with minimal modification. Early intraoperative sizing of the patch allowed completion of fetal repair within the goal of 30 minutes in all but one case.

**CONCLUSIONS:**Preoperative 3-D modeling can accurately demonstrate MMC anatomy and predict the need for a patch. Early intraoperative creation of an appropriately sized patch using the 3-D generated template contributes to reduced operative time, potentially contributing to a decrease in premature delivery and intraoperative complications. 3-D modeling may have implications for progress in fetoscopic intervention and the development of advanced biomaterials.

**Keywords:** myelomeningocele, fetal surgery, patching, modeling, surgical technique

## FL-003

### Spine

#### A new method for evaluating myelomeningocele lesion level in fetuses

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**OBJECTIVE:**Effective prenatal counseling for a fetus with myelomeningocele (MMC) requires a prediction of postnatal motor function. However, determining the precise level of MMC in the fetus is difficult. We retrospectively assessed MMC lesion levels on fetal MRIs to determine whether postnatal motor function can be predicted prenatally.

**MATERIAL-METHODS:**We examined fetal MRIs for 18 cases of prenatally diagnosed MMC that were treated at our institute from 2012 to 2016. We excluded two cases with complicated chromosomal aberration or osteogenesis imperfecta, and included the remaining 16 cases. We examined prenatal MRIs obtained with half-Fourier acquisition single-shot turbo spin-echo (HASTE), and estimated the level of the MMC lesion by setting the level of the right hilus renalis as the level of the L1 vertebral body (hilus renalis method) or by setting the last bright disc space of the lumbosacral spine as L5-S1 (disk method). The lesion level

was assigned as the highest vertebral level involved in a sagittal view of the spine. All infants underwent MMC repair on the day of birth. An orthopedic surgeon evaluated the motor functions at 3 months of age. A radiologist determined the anatomical MMC level from postnatal imaging.

**RESULTS:**We could estimate the lesion level from fetal MRIs for all 16 cases using the hilus renalis method, but for only 11 cases using the disc method. Our estimated level from fetal MRIs matched the postnatal anatomical level in 83% of the cases when using the hilus renalis method, and in 50% when using the disc method. The estimated level matched postnatal motor function in 11 cases (68%) when using the hilus renalis method, and in 6 cases (37%) when using the disc method.

**CONCLUSIONS:**The hilus renalis method may allow us to predict postnatal motor function from the MRI of a fetus with MMC.

**Keywords:** prenatal MRI, lesion level, motor function, myelomeningocele, hilus renalis

## FL-004

### Special topic: Fetal diagnosis and management

#### Fetal ventriculomegaly: external validity of a fetal MRI-based model to predict the need for postnatal CSF diversion

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**OBJECTIVE:**Not all children diagnosed in-utero with fetal ventriculomegaly (FV) require cerebrospinal fluid (CSF) diversion after birth. Using image analysis and machine learning techniques, we developed fetal MRI-based models to predict the need for post-natal shunting in FV patients. To assess its external validity, we set out to apply this approach to a dataset from another institution.

**MATERIAL-METHODS:**FV patients undergoing fetal MRI at a minimum gestational age (GA) of 20 weeks comprised the cohort on which the models were trained. Separate models were derived based on four imaging features: atrial diameter (AD); fronto-occipital horn ratio (FOHR); CSF volume normalized to brain size obtained using computer-aided segmentation; and a voxel-to-voxel comparison using aligned images. The accuracy of the models on the training cohort was assessed via leave-one-out cross validation. The trained models were then applied to a replication cohort consisting of subjects with fetal MRIs obtained at another institution. Accuracy and receiver operator characteristic analyses were performed.

**RESULTS:**Fifty FV patients (25 shunted, per HCRN guidelines) undergoing fetal MRI at a median gestational age of 26 weeks were used to train the models. In the training cohort, median time to post-natal shunting was 6 days, and median follow-up time among non-shunted patients was 29 months. In

this cohort, the models correctly classified subjects as needing a shunt with 80% maximum accuracy. The replication cohort consisted of 26 patients, of which 4 underwent post-natal shunting. When applied to the replication cohort, models based on AD and CSF volume had the highest predictive value with an accuracy approaching 100%.

**CONCLUSIONS:** Our findings support the generalizability of a fetal MRI-based model for predicting the need for post-natal CSF diversion and highlight AD and CSF-segmented volume as features that remain predictive across institutions. Future studies will focus on assessing the models prospectively.

**Keywords:** fetal, ventriculomegaly, hydrocephalus, shunt, machine learning

## FL-005

### Special topic: Fetal diagnosis and management

#### Prenatal closure of myelomeningoceles: does it reduce the need for VP shunts?

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**OBJECTIVE:** We undertook a study to determine the number of children who meet current neuroanatomical criteria for offering prenatal closure of myelomeningoceles (MM) at our fetal health center (FHC).

**MATERIAL-METHODS:** Children born with spina bifida who had been evaluated prenatally at our FHC were analyzed in regard to prenatal vs. postnatal closure, prenatal ventricular width, postnatal neurological level, and whether a VP shunt was placed.

**RESULTS:** 10 fetuses were identified prenatally to harbor MM and were evaluated and delivered at the FHC. One had a prenatal closure elsewhere and now has a shunt. 8 of the remaining group met criteria for prenatal closure. Of these 8, six (75%) have VP shunts; the other two have ventriculomegaly. 4 other fetuses had prenatal closure elsewhere; only 2 meet current inclusion criteria. Of these, three have a VP shunt (75%).

**CONCLUSIONS:** there is no discernable difference in the rate of VP shunts in our two groups (75%), pre-natal vs. post-natal closure. This rate is higher than that reported in multicenter studies which show a clear reduction in VP shunt placement in the prenatally closed group. Our anomalous outcome may reflect the smaller number of children in this study

**Keywords:** Myelomeningocele, prenatal surgery, VP shunt, hydrocephalus

Monday, 24 October 2016

16:30 – 18:00

## Flash Session 2: Craniofacial

### FL-017

#### Craniofacial

##### Craniopagus surgery as an emergency

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**OBJECTIVE:** Conjoined twin is a rare embryonic anomaly and craniopagus is much more uncommon. There are reports of surgical separation which have been done in stable general condition of twins including good weight and

medical situation at the presence of extensive preoperative radiologic and laboratory investigations. In spite of thorough evaluation and preparation surgical outcome is not very well.

**MATERIAL-METHODS:** Here we report a case of craniopagus twin of partial type. One of the twins died of respiratory failure probably subsequent to aspiration at age of 28 days of life. Twins had history of prematurity and birth at gestational age of 7 months who were urgently admitted from another center due to death of one of them. Surgical separation considered as immediately after their admission in our center and performing a rapid radiologic evaluation. The laboratory results for the surviving twin indicated normal coagulation functions and surgery was performed to prevent coagulopathy and sepsis. The twins were attached to each other at the frontoparietal vertex. They had brain MRI when were admitted in another center. We performed emergent brain CT with CT angiography to assess the bony, vascular, and nervous system interconnections.

**RESULTS:** Separation was successfully performed with saving the alive one. The skin, dura matter of dead neonate were used for reconstruction of another one.

**CONCLUSIONS:** A multi-disciplinary approach is essential for management of conjoined twins. Emergency separations bring dismal results due to bad emergent condition of patients. Delayed separation provides more favorable results due to well-planned strategies, team preparedness, and better patient conditions. In case of death of one twin emergency surgery is indicated. The advantage and disadvantage of this kind of surgery are discussed.

**Keywords:** Craniopagus, death of one twin, surgery

## FL-018

### Special topic: Neuro-imaging

#### Frequency and age distribution of accessory cranial sutures in the parietal and occipital bones

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**OBJECTIVE:** Various accessory cranial sutures are present in the parietal and occipital bones in pediatric population due to their complex ossification patterns. The parietal and occipital bones ossify from two and six ossification centers, respectively, and accessory cranial sutures can be identified at the boundary of these ossification centers. The purpose of this study is to clarify the frequency and age distribution of accessory cranial sutures.

**MATERIAL-METHODS:** Among the patients with mild head trauma less than 15 years old from April 2012 to March 2014, 515 patients (mean 3.8 years old) who were performed CT scan were enrolled in this study. Presence of accessory cranial sutures in the parietal and occipital bones were retrospectively evaluated on 3D images, and frequency in each age group were analyzed.

**RESULTS:** In the parietal bone, intraparietal sutures were present in 4.3% of all the patients. In the occipital bone, mendosal sutures, superior longitudinal fissure, midline occipital fissure, and innominate sutures were identified in 24.3%, 5.6%, 1.2%, and 40.4%, respectively. These accessory cranial sutures were frequently observed in younger age group.

**CONCLUSIONS:** A diagnosis of normal accessory cranial suture should be considered when encountering extra lines in the parietal and occipital bones in young patients. Accurate knowledge of accessory cranial sutures are essential for precise differential diagnosis between fracture lines and suture lines.

**Keywords:** accessory cranial suture, CT scan

## FL-019

## Craniofacial

**Craniosynostosis versus early fontanel closure: can misconceptions influence referral and surgery timing?**Carlos Eduardo Jucá<sup>1</sup>, Gabriela Oliveira<sup>2</sup>, Livia Freitas<sup>3</sup><sup>1</sup>Universidade de Fortaleza (UNIFOR), Fortaleza, Brazil; Centro Universitário Christus - Unichristus, Fortaleza, Brazil, Hospital Infantil Albert Sabin, Fortaleza, Brazil; Instituto Hippocampus, Fortaleza, Brazil.<sup>2</sup>Centro Universitário Christus - Unichristus, Fortaleza, Brazil.<sup>3</sup>Universidade de Fortaleza (UNIFOR), Fortaleza, Brazil.

**OBJECTIVE:** Timely diagnosis of craniosynostosis is mandatory to appropriate management. Patients and families depend on referral from general pediatricians and practitioners to have access to a pediatric neurosurgeon assessment. Aim of this study is to evaluate current pediatrician's conceptions about craniosynostosis and fontanel closure (FC) and compare to objective clinical findings.

**MATERIAL-METHODS:** A questionnaire about craniosynostosis and FC was proposed to 50 pediatricians and 100 mothers of non neurosurgical patients in a pediatric hospital in Fortaleza, Brazil. A two years service casuistic of craniosynostosis was reviewed to compare the data with clinical findings.

**RESULTS:** Among general population, 87% of responders said there is association between early FC and craniosynostosis, but only 2,5% could define craniosynostosis. 50% of pediatricians said FC should happen between 13-18 months, 54% believe in a link of FC time and craniosynostosis. When facing early FC, 60% of pediatricians consider direct referral to a neurosurgeon, while 25% prefer surveilling head circumference and neurological development. 98% of pediatricians claimed have previous knowledge about craniosynostosis, but 46% pointed out a link between suture fusion and FC. Asked about clinical presentation, 39% said craniofacial deformities are the main manifestations of early FC, mental retardation in second place. In the last two years, 17 children had a craniosynostosis surgery in our service, 10 scaphocephalies, 6 anterior plagiocephalies and 1 trigonocephaly. All of them were diagnosed based on cranial deformities, not early FC, 47% of them had an open fontanel at diagnosis. Two scaphocephalic patients arrived older than one year, referral being retarded due to existence of an open fontanel.

**CONCLUSIONS:** Diagnostic confusion involving craniosynostosis and FC are the cause of inappropriate referral and may lead to useless neurosurgical assessments or retarded surgery timing. Pediatric neurosurgeons should have a role in the education of practitioners and the general population to avoid these errors.

**Keywords:** Craniosynostosis, fontanel, cranial sutures.

## FL-021

## Craniofacial

**Is the previous concept true that intracranial pressure is low in microcephalic children? Results of intracranial pressure monitoring in 24 microcephalic children with developmental delay**Soo Han Yoon<sup>1</sup>, Dong Ha Park<sup>2</sup><sup>1</sup>Department of Neurosurgery, Ajou University School of Medicine, Suwon, Korea 443-721<sup>2</sup>Department of Plastic and Reconstructive Surgery, Ajou University School of Medicine, Suwon, Korea 443-721

**OBJECTIVE:** After craniectomy had begun more than 100 years ago both for children with microcephaly and with craniosynostosis, surgery for children with microcephaly was discontinued in the early 1900's because of high mortality and no effectiveness, while surgery for children with craniosynostosis has continued and involved many techniques. However, development of the

new direct gradual cranial expansion surgical technique has been applied to the treatment of children with post-shunt microcephaly and slit ventricle syndrome, such that we should study the intracranial pressure (ICP) in children with microcephaly to evaluate the possibility of new surgical techniques for children with microcephaly.

**MATERIAL-METHODS:** We analyzed age, sex, mean ICP, head size and developmental score in 24 microcephalic children with developmental delay who received the Camino cortical intracranial pressure monitoring during 2015.

**RESULTS:** Twenty four microcephalic children with developmental delay consisted of 9 boys and 15 girls, and the mean age was  $4.9 \pm 2.0$  years. ICP changes according to age was  $y = -1.8156x + 27.721$ . There were no statistical differences of developmental scores and head sizes between children with high ICP and with low ICP. The mean ICP was  $18.7 \pm 8.6$  mmHg and in 9 children it was less than 15mmHg, 5 were between 15 and 20mmHg, and 10 were more than 20mmHg.

**CONCLUSIONS:** We suggest that a significant portion of microcephalic children with developmental delay have high ICP that cannot be expected from head sizes or development scores and that has decreasing tendency with age.

**Keywords:** developmental delay, head size, intracranial pressure, microcephaly

## FL-022

## Craniofacial

**Does the addition of barrel staves improve results in endoscopic strip craniectomy for sagittal craniosynostosis?**Suresh N Magge<sup>1</sup>, Benjamin C Wood<sup>2</sup>, Edward S Ahn<sup>3</sup>, Joanna Y Wang<sup>3</sup>, Robert F Keating<sup>1</sup>, Gary F Rogers<sup>2</sup><sup>1</sup>Division of Neurosurgery, Childrens National Health System, Washington, DC, USA<sup>2</sup>Division of Plastic Surgery, Childrens National Health System, Washington, DC, USA<sup>3</sup>Division of Neurosurgery, Johns Hopkins University, Baltimore, MD, USA

**OBJECTIVE:** There exists a wide variation in the endoscopic approaches used for sagittal craniosynostosis. This study compares outcomes of patients undergoing strip craniectomy versus strip craniectomy plus barrel staves for the treatment of sagittal craniosynostosis.

**MATERIAL-METHODS:** An IRB-approved, multi-center, retrospective review was conducted of 73 patients undergoing endoscopic surgery at two institutions. Group A (34 patients) was treated with simple strip craniectomy. Group B (39 patients) was treated with strip craniectomy plus barrel staves. Outcomes were analyzed using imaging data from the same orthotic manufacturer treating patients from both institutions.

**RESULTS:** The mean operative time was significantly shorter in Group A: 71.6 minutes for Group A, and 111 minutes for Group B ( $p < 0.01$ ). The mean anesthetic time was also significantly shorter in Group A: 161.7 minutes for Group A, and 195 minutes for Group B ( $p < 0.01$ ). Mean hospital stay was similar for both (1.2 days for Group A, 1.4 days for Group B,  $p = 0.1$ ). For Group A, preoperative cephalic index (CI) was 72.6, immediate postop CI was 73.8, and final CI was 80.4, with a mean follow-up period for 13.2 months. For Group B, preoperative CI was 71.0, immediate postop CI was 73.6, and final CI was 79.6, with mean follow-up of 19.4 months. There was no statistical difference in change of CI between the two groups (10.5% change in Group A, 12.2% change in Group B,  $p = 0.15$ ).

**CONCLUSIONS:** Both endoscopic strip craniectomy and strip craniectomy with barrel-staving produce good outcomes. But the addition of barrel staves does not seem to improve the results as there is no statistical difference between the two techniques with regard to change in CI. However, the addition of barrel staves adds operative time and anesthesia time, and therefore barrel staves may not be warranted in the endoscopic treatment of sagittal craniosynostosis.

**Keywords:** craniosynostosis, sagittal, endoscopic, strip craniectomy, barrel staves

## FL-023

## Craniofacial

**Management of sagittal craniosynostosis in Mansoura University Hospital: assessment of two surgical techniques**

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<sup>2</sup>Department of Plastic Surgery, Mansoura University, Mansoura, Egypt

**OBJECTIVE:** Isolated sagittal synostosis causing scaphocephaly is the most common form of craniosynostosis, accounting for 40% of cases. Authors compared outcome of two surgical techniques; peninsula-shaped extended linear craniectomy and parasagittal biparietal bone remodeling with extended strip craniectomy.

**MATERIAL-METHODS:** This is a prospective study including all the non-syndromic scaphocephalic cases, younger than 1 year, admitted to Mansoura University Hospital, Pediatric Neurosurgery Unit and treated surgically from 2005 to 2015. The cases were assigned randomly to two groups: Group 1, 26 patients who underwent peninsula-shaped extended linear craniectomy, and Group 2, 26 patients who underwent parasagittal biparietal bone remodeling with extended strip craniectomy. The following variables were analyzed: sex, age, weight, cranial index (CI), surgical time, blood loss, hospital admission time, and postoperative complications.

**RESULTS:** Among 164 children admitted with craniosynostosis, only 52 children had isolated scaphocephaly. The final average age was 5.7 months, the median operative time was found to be 55 minutes in group 1, compared with 130 minutes in group 2 ( $p < 0.0001$ ); while the post-operative hospitalization time was 3 days, compared with 6 days in group 2 ( $p < 0.0001$ ). The median intra and post-operative blood transfusion in group 1 and group 2 was 60 ml and 200 ml ( $p < 0.0001$ ), respectively. There were no complications. The median CI preoperatively was 65 and 66 ( $p < 0.153$ ), while immediate post-operatively was 69, and 74 ( $p < 0.001$ ), and at 1-year post-operatively was 72 and 73 ( $p < 0.147$ ), when we compared groups 1 and 2 respectively.

**CONCLUSIONS:** In term of cranial index (CI), although early outcome of remodeling group proved statistically superior to extended linear craniectomy group, the long term outcome after 1 year showed no statistically significant difference. Additionally, in view of the shorter operative time, shorter hospital stay and less blood loss, we recommend extended linear craniectomy to be more widely applied.

**Keywords:** craniosynostosis, scaphocephaly

## FL-025

## Craniofacial

**Risk for Chiari malformation type I in sagittal synostosis treated by an open technique**

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**OBJECTIVE:** There are still many discussions about indications (cosmetic or functional) and techniques for surgery in sagittal synostosis (SS); also the results are matter for debate, for cognitive outcome and volume correction. Chiari I Malformation (CMI) is a well known association with complex craniosynostosis, reported quite rarely in association with monosutural ones, both untreated and operated. The present study analyses risk for CMI before and after cranioplasty for SS.

**MATERIAL-METHODS:** We retrospectively reviewed 120 pure sagittal synostotic children operated since 2000 by the same open technique (details of cranial vault remodelling will be described); all the patients were submitted preoperative 3DCT scan and to clinical genetic evaluation and genetic exams were prescribed in selected cases to exclude syndromic cases. When aged 5 years or more all patients were submitted to a postoperative MRI with the aim to search for CMI.

**RESULTS:** Age at surgery ranged between 4 and 48 months; at CT scan CMI was suspected in 5 cases, all submitted to MRI, that documented tonsils descent only in two; another case of CMI was documented at MRI in a child 4 years. MRI at 5 years follow up was performed in 90 children: only the child with delayed diagnosis and treatment still had a tonsil herniation.

**CONCLUSIONS:** CT scan is able both to diagnose true sagittal synostosis and to rise the suspect of tonsils herniation. The incidence of CMI in this series of treated SS was 2% preoperatively and null postoperatively, except for the case with delayed diagnosis and treatment.

The present study suggests that early Sagittal Synostosis correction by cranial vault remodelling hereby presented has the functional meaning to prevent the delayed occurrence of CMI. Also the so called “mininvasive” techniques need a longitudinal evaluation not of cosmetic results, but also of functional effect, in terms of capability preventing CMI occurrence.

**Keywords:** Sagittal Synostosis, Chiari Malformation Type I

## FL-026

## Craniofacial

**Outcome 25 years after surgery for scaphocephaly**

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<sup>3</sup>Department of Children and Adolescent, Oulu University Hospital, Oulu, Finland

**OBJECTIVE:** The purpose of the study was to evaluate the late outcome of adult patients treated for scaphocephaly during early childhood.

**MATERIAL-METHODS:** The study population consisted of patients operated for scaphocephaly at Oulu University Hospital between 1978 and 1998. Patients with any associated disease (i.e. hydrocephalus) were excluded. 41 patients fulfilled the criteria and approved to participate in the follow up evaluation. Patients were treated with parasagittal craniotomies with silicone membrane interposition (8 cases), suturectomy with or without dural split (6), and various forms of H-plasty (27). The mean age at the time of operation was 5.1 months. Control persons were age and sex matched and randomly selected from the national register. Both patients and controls answered questions about their education, housing, marital status, employment and health condition. The participants rated their own satisfaction with their appearance on the 10-point Visual Analogue Scale (VAS). From the standardized photographs taken during the follow up visit independent panels consisting of medical professionals and lay persons evaluated the cosmetic outcome using also VAS scale.

**RESULTS:** The patients mean age at follow up was 26.9 years. There was no difference in education, housing, marital status, employment, presence of mental health problems or satisfaction with own appearance between patients and controls. The panels gave a lower score to patients compared to controls (6.0 and 6.7 accordingly). The used operation method did not influence the cosmetic outcome, as judged by the patients themselves or by the independent panels.

**CONCLUSIONS:** Patients treated for scaphocephaly do as well as controls when evaluated by education, employment, mental morbidity and general

health. They are also equally satisfied with their facial appearance as healthy controls. These despite that 30 % of the patients were treated with old-fashioned methods. However, independent evaluators score lower points for patients than controls.

**Keywords:** craniostynostoses, scaphocephaly, outcome, cranioplasty

#### FL-027

##### Craniofacial

#### MCDO (multi-directional cranial distraction osteogenesis) system for craniostynostosis

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**OBJECTIVE:**Distraction osteogenesis for the treatment of craniostynostosis has been accepted by many craniofacial surgeons. This technique has several advantages, but is still in need of improvement. We have developed a completely new type of distraction system called multidirectional cranial distraction osteogenesis (MCDO). The MCDO system consists of a disposable clear plastic frame, anchor pins and extension rods for fixing the frame on the skull, traction pins for pulling the bone pieces up, a distractor fixed on the frame, and plastic flanges for protecting the holes.

**MATERIAL-METHODS:**We retrospectively evaluated 36 children with craniostynostosis treated with the MCDO system in our institute between 2005 and 2014. Median patient age was 12 months. Follow up was 18–125 months (mean, 64 months).

**RESULTS:**The number of osteotomy was 9 to 30 pieces (mean, 14 pieces). The mean blood transfusion was 32 ml/kg. The mean postoperative hospital stay was 19 days. The phase of activation was 8 to 14 days (mean, 10 days) and consolidation period was 14 to 63 days (mean, 43 days). The morphological results of all case were satisfactory. Nine patients had complications, including one with meningitis, four with transient subcutaneous cerebrospinal fluid (CSF) leakage, and four with pin-track infections that caused loosening of the frame. However, all of the patients achieved the scheduled consolidation. **CONCLUSIONS:**The MCDO system allows the contour of any kind of skull deformities as desired under the concept of distraction osteogenesis. This method has the following benefits, such as a high flexibility of reshaping, shorter treatment period and less invasive secondary intervention. It offered numerous advantages over not only the distraction method with unidirectional internal devices but also conventional cranioplasty for the patient with craniostynostosis.

**Keywords:** craniostynostosis distraction osteogenesis multidirectional cranial distraction osteogenesis

#### FL-028

##### Craniofacial

#### Computer-assisted shape descriptors for skull morphology in craniostynostosis

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**OBJECTIVE:**Our aim was to develop a novel method for characterizing common skull deformities with high sensitivity and specificity, based on two-dimensional (2D) shape descriptors in computed tomography (CT) images.

**MATERIAL-METHODS:**Between 2003 and 2014, 44 normal subjects and 39 infants with craniostynostosis (sagittal, 29; bicoronal, 10) enrolled for analysis. Mean age overall was 16 months (range, 1–120 months), with a male:female ratio of 56:29. Two reference planes, sagittal (S-plane: through top of lateral ventricle) and coronal (C-plane: at maximum dimension of fourth ventricle), were utilized to formulate three 2D shape descriptors (cranial index [CI], cranial radius index [CR], and cranial extreme spot index [CES]), which were then applied to S- and C- plane target images of both groups.

**RESULTS:**In infants with sagittal craniostynostosis, CI in S-plane (S-CI) usually was <1.0 (mean, 0.78; range, 0.67–0.95), with CR consistently at 3 and a characteristic CES pattern of two discrete hot spots oriented diagonally. In the bicoronal craniostynostosis subset, CI was >1.0 (mean 1.11; range, 1.04–1.25), with CR at –3 and a CES pattern of four discrete diagonally oriented hot spots. Scatter plots underscored the highly intuitive joint performance of CI and CES in distinguishing normal and deformed states. Altogether, these novel 2D shape descriptors enabled effective discrimination of sagittal and bicoronal skull deformities.

**CONCLUSIONS:**Newly developed 2D shape descriptors for cranial CT imaging enabled recognition of common skull deformities with statistical significance, perhaps providing impetus for automated CT-based diagnosis of craniostynostosis.

**Keywords:** Shape descriptor, Craniostynostosis, Scaphocephaly, severity index, Cranial index, Cranial spectrum

#### FL-029

##### Craniofacial

#### Skull comparison using curvature maps: first step in the development of fully objective, automated surgical planning techniques

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**OBJECTIVE:**3D virtual planning of open cranial vault reconstruction is used to simulate and define an operative plan for craniostynostosis surgery. However, virtual planning techniques are subjective and dependant on the experience and preferences of the surgical team. In order to enable further development of a truly objective automated 3D pre-operative planning technique for open cranial vault reconstructions, we created curvature maps for the comparison of the patient's skull with an age-specific normative skull model. **MATERIAL-METHODS:**Normative skull models were created from sets of cranial CT-scans with no cranial or neural abnormality. Age groups were 0.5, 3, 6, 9 and 12 months. For each scan a triangulated mesh of the skull was created using marching cubes based on a 167 HU threshold. All skulls were oriented similarly within the coordinate system and ray casting was used to obtain sampled 3D metrical data of the inner and outer layer of the skulls. This

data was used to create normalised skull models for each age group. The marching cubes, registration and sampling method were also used on a trigonocephaly, scaphocephaly and anterior plagiocephaly case. Finally, curvature maps were computed using quadric surface fitting.

**RESULTS:** Normalised skull models and corresponding curvature maps have been created for the age-groups of 0.5, 3, 6, 9 and 12 months. A pilot comparison of the curvature maps was made with a trigonocephaly, scaphocephaly and anterior plagiocephaly case.

**CONCLUSIONS:** It is shown that curvature maps allow the comparison of craniosynostosis skulls with age-appropriate normative skulls. This study showed the first step towards an automated objective virtual pre-operative planning technique for open cranial vault reconstructions. Further research will focus on the development of an automated technique that determines the optimal osteotomy lines based on this comparison.

**Keywords:** Craniosynostosis, normative skull models, curvature map, objective automated surgical planning

## FL-030

### Craniofacial

#### Radiation-free 3D head shape and volume evaluation after endoscopically assisted trigonocephaly surgery

Guido de Jong<sup>1</sup>, Manon Tolhuisen<sup>1</sup>, Jene Meulstee<sup>2</sup>, Erik Van Lindert<sup>1</sup>, Wilfred Borstlap<sup>2</sup>, Thomas Maal<sup>2</sup>, Hans Delye<sup>1</sup>

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**OBJECTIVE:** Post-operative follow-up in craniosynostosis is still mainly done using radiation techniques. Sequential radiation-free follow-up techniques (e.g. 3D stereofotogrammetry) 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 calculate the sella turcica position, allowing correct sequential image superposition and objective evaluation. We used this technique for mean volume and shape change evaluation of the head based on 3D Photos after endoscopically assisted trigonocephaly surgery.

**MATERIAL-METHODS:** We performed a mean head shape and volume evaluation on age grouped 3D Photos (n=86) of 21 children that underwent endoscopically assisted trigonocephaly surgery with helmet therapy. We used CT-scans of healthy children (n=56) as reference. The CCFP and nasion were used to orient the 3D Photos. We performed a mean shape evolution analysis and calculated the anterior fossa to total volume ratio (A/T-ratio). The volume- and A/T-ratio pattern were compared with the reference group.

**RESULTS:** The mean anterior fossa volume evolved from 294ml (36.8% A/T-ratio) pre-surgery to 664ml (36.0% A/T-ratio) at 37-48 months post-surgery. Post-surgery the A/T-ratio increases to 38.1% at 5-7 months to decrease to 35.1% at 25-36 months. The reference group showed a near similar volume- and A/T-ratio pattern. In the first 18 months there is a predominant growth around the resected metopic suture. Between 18 and 24 months we observed mostly anterior orbital rim growth. From 24 months till 36-48 months the head grows predominantly at the temporal area. The least outward growth was observed at the temporal bones.

**CONCLUSIONS:** Using a novel technique we were able to objectively evaluate head shape and volume using stereophotogrammetry after endoscopically assisted trigonocephaly correction. The A/T-ratio and volume growth pattern of endoscopically treated patients is near identical to that of the normal reference group.

**Keywords:** 3D Photo, Craniosynostosis, Evaluation, Surgery, Volume, Shape

## FL-031

### Craniofacial

#### Results of craniosynostosis surgery: about 56 cases

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**OBJECTIVE:** During the last decade, the development of different technologies applied to craniofacial surgery has determined a different approach to craniosynostosis compared to the past. For instance, there is no doubt in the fact that resorbable osteosynthesis has improved the stability of the cranial assemblies, osteogenic distraction stresses to find further indications in these treatments and endoscopic approaches make some of these procedures less invasive. We retrospectively reviewed two centers (author) experience with the surgical treatment of craniosynostosis.

**MATERIAL-METHODS:** Fifty-six cases of craniosynostosis were treated in the forenamed centers between august 2009 and March 2016. They were 9 syndromic malformations (5 Apert, 1 Crouzon, 3 clover leaf skull) and 45 nonsyndromic (21 saggittal, 13 coronal, 8 metopic, 3 lambdoid, 5 multiple sutures affected).

**RESULTS:** The mean age of these children at time of surgery was 8.9 months (range, 3 months to 3 years) with a slight male predominance. All our patients underwent neuroradiological evaluation including plain skull radiographs, CT scan of skull and brain with 3-D reconstructions. The surgical treatment consisted in classic suturectomy (bear skin) plus parietal and frontal modelling in sagittal cases, fronto-orbital advancement plus frontal modeling for uni or bilateral coronal cases, parieto-ccipital advancement plus parietal (sunrise) and occipital modeling (barrel stave) in uni or bilateral lambdoid cases, and combination of these depending of the suture(s) that were affected in multiple suture cases. Some post-operative complications were observed (CSF leakage and foreign body reaction to poliglicolic acid suture and plates) and one death due to hypoxia related to transoperative pulmonary failure. None case of recurrence after suturectomy were observed.

**CONCLUSIONS:** Based on our experience, the surgical techniques allows immediate correction of the cranial deformity increasing the vault convexity, also provided the resolution of cerebral compression by increasing intracranial volume, with very low rate of complications.

**Keywords:** Craniosynostosis, Suturectomy, Cranial modelling

## FL-032

### Craniofacial

#### Surgical treatment of non-syndromic craniosynostosis: the Rady Children's Hospital experience

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**OBJECTIVE:** The authors review their 15 year single institution experience in the surgical treatment of non-syndromic craniosynostosis utilizing variations of both open and endoscopic techniques.

**MATERIAL-METHODS:**A retrospective chart review was carried out on all patients undergoing surgical treatment of non-syndromic craniosynostosis at Rady Children's Hospital from 2000-2015. All patients were operated on by a single neurosurgeon (HM) in collaboration with two plastic and reconstructive surgeons (SC, AG) as part of our institution's craniofacial team.

**RESULTS:**514 patients (280 open, 234 endoscopic) underwent initial surgical repair of craniosynostosis (age 1-149 months, median 6.5 months). Diagnoses included 274 (53%) sagittal, 123 (24%) metopic, 82 (16%) unicoronal, 14 (3%) lambdoid, 6 (1%) bicoronal, and 11 (2%) multisutural. Endoscopic patients had a median age of 3.8 months (range 1-12 months) and included 190 (81%) sagittal, 31 (13%) metopic, 10 (4%) unicoronal, and 3 (1%) lambdoid synostoses. Comparing open to endoscopic patients, median age (11.1 to 3.8 months), mean surgery time (140 to 66 minutes), mean EBL (137 to 35cc), mean % weight EBL (16 to 6), OR transfusion rate (84 to 47%), mean amount blood transfused (184 to 62cc), and mean length of stay (3.1 to 1.7 days) all reached statistical significance ( $p < 0.001$ ). Only 27 (5%) patients (19 open and 8 endoscopic) required post op care in ICU setting. 6 open patients (1% of all, 2% of open) had post-op infections, 5 of which required surgical intervention. 23 (4%) patients (17 open and 6 endoscopic) had dural tears noted and repaired primarily during surgery. No patient required reoperation for csf leak/pseudomeningocele/growing skull fracture equivalent. No patient had evidence of symptomatic air embolism, requirement of emergent reoperation, or significant long term morbidity or mortality.

**CONCLUSIONS:**An experienced, multidisciplinary team approach leads to excellence in the treatment of craniosynostosis. A significant subgroup of patients likely benefit from endoscopic, minimally invasive techniques.

**Keywords:** craniosynostosis, non-syndromic, endoscopic, multidisciplinary

#### FL-033

##### Craniofacial

##### Geometric correction of non-syndromic craniosynostosis: aesthetic outcome analysis

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**OBJECTIVE:**To describe a novel technique and to analyze the aesthetic results.

Various surgical techniques have been described in the literature for correction of craniofacial deformities caused by craniosynostosis. The literature still lacks a simple and reproducible technique that can be applied to the wide range of craniosynostosis deformities to achieve predictable and reproducible aesthetic results.

**MATERIAL-METHODS:**A retrospective analysis of patients who underwent craniosynostosis correction in a tertiary academic hospital during the period August 2014 and May 2015 was done. Pre and post-operative photographs were used for subjective assessment of the aesthetic outcome using Whitaker scoring method by three independent evaluators. (Category I - no revision; category II - soft-tissue or lesser bone- contouring revisions desirable; category III - major alternative osteotomies / bone grafting procedures needed; category IV - craniofacial procedures duplicating / exceeding in extent the original surgery needed)

**RESULTS:**Thirteen patients with craniosynostosis - unicoronal (N=7), bicoronal (N=4) and metopic suture synostosis (N=2) - underwent

Fronto-orbital advancement and cranioplasty using the principle of geometric correction method. Based on Pre and Postoperative subjective evaluation using Whitaker Scoring Method, the aesthetic outcome of 56.4% patients was category I and 41% and 2% had category II and III outcome respectively.

**CONCLUSIONS:**The geometric correction method provides good aesthetic outcome in non-syndromic craniosynostosis deformities. This technique is universally reproducible and the same principle can be applied in management of various types craniosynostosis deformities.

**Keywords:** Craniosynostoses, aesthetic outcome

#### FL-034

##### Craniofacial

##### Does early cosmetic evaluation predict the cosmetic outcome 20 years later in patients treated for craniosynostosis during early childhood?

Willy Serlo<sup>1</sup>, Niina Salokorpi<sup>2</sup>, Tuula Savolainen<sup>3</sup>, Juha Jaakko Sinikumpu<sup>1</sup>, Pertti Pirttiniemi<sup>3</sup>

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**OBJECTIVE:**To evaluate how well early postoperative evaluation of the cosmetic outcome after craniosynostosis surgery do correlate with the outcome judged over 20 years later when the patients are adults.

**MATERIAL-METHODS:**The study population consisted of patients operated due to craniosynostosis at Oulu University Hospital between 1976 and 1996, whose cosmetical results were evaluated on a score from 1 to 4 early after primary surgery (mean 33 month, range 1 month to 9.7 years). From 40 patients who were included in the first evaluation, 22 patients approved to come for a new follow up visit during 2015-2016 (mean 20 years later). At the late follow up examination patients were scored for cosmetic outcome by a nurse, a dentist and a paediatric neurosurgeon on the 10-point Visual Analogue Scale (VAS). The participants to rated also their own subjective satisfaction of their cosmetic outcome on the same scale.

**RESULTS:**Those patients who had been 30 years (28-34 years) earlier scored as excellent (1) gained in mean 8.1 points out of 10 (6 patients); those who were early judged as good (2) gained in mean also 8.1 points (14 patients) and those scored as fair (3) gained 6.7 points (2 patients). We found no patients that had been scored as 4 (poor) on the first follow up. Patients own satisfaction with appearance was accordingly 6.1, 7.9 and 8.8.

**CONCLUSIONS:**Early evaluation of the cosmetic outcome (mean 3 years after operation) correlated well with the cosmetic outcome 20 years later. Patients, whose outcome was judged as excellent or good at the early follow up, received higher scores on late follow-up than those who were primarily judged as poor.

**Keywords:** craniosynostosis, cranioplasty, outcome

#### FL-035

##### Craniofacial

##### Long-term results of frontal and bilateral cranial distraction osteogenesis for multi-suture craniosynostosis

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**OBJECTIVE:**The authors retrospectively analyzed effectiveness of frontal and bilateral cranial distraction osteogenesis for oxycephalic multi-suture craniosynostosis, especially in the cases with long-term follow-up.

**MATERIAL-METHODS:**Between October 2000 and May 2011, 15 patients with multi-suture craniosynostosis (Crouzon: 9, Other: 6) underwent cranial distraction osteogenesis, expanding skull both anteriorly and bilaterally. The age at the surgery ranged from one year and 6 months to 9 years (median: 2 years and 7 months). The osteotomies were made in fronto-orbital area including anterior skull base and in biparietal area. After the osteotomies, two or four distraction devices (Keisei Ika Kogyo, Japan) were attached at bicoronal area for frontal expansion. In addition, one or two devices were applied each side at parietal bones for bilateral dilatation. The final cranial expansion ranged from 10 to 28 mm (median: 21.5 mm) anteriorly; from 6 to 25 mm (median: 18.5 mm) bilaterally. The median duration of follow-up was 10 years and 1 month (ranged from 4 years and 10 months to 15 years and 5 months).

**RESULTS:**After the treatment, copper beaten appearances of the skulls disappeared in all patients. In addition, CT scans showed good visualization of cortical sulci in all cases. One patient had CSF leakage around the distraction device and underwent removal of the device. Another patient had dislocation of the distractor just after surgery. The distraction device was applied again. However, neither serious infection nor other complication occurred. No recurrence of cranial stenosis was evident during the follow-up periods.

**CONCLUSIONS:**In the view point of long-term follow-up, frontal and bilateral cranial distraction osteogenesis is useful treatment for oxycephalic multi-suture craniosynostosis.

**Keywords:** craniosynostosis, distraction osteogenesis, Crouzon syndrome, oxycephaly

## FL-035

### Craniofacial

**The long term outcome of the management of active hydrocephalus in children with complex craniosynostoses at diagnosis: is it always possible to avoid further constriction?**

Gianpiero Tamburrini, Fabrizio Pignotti, Paolo Frassanito, Luca Massimi, Massimo Caldarelli

Pediatric Neurosurgery, Institute of Neurosurgery, Catholic University Medical School, Rome, Italy

**OBJECTIVE:**Few data are available in the literature concerning the long-term outcome of the management of children with complex craniosynostoses presenting at diagnosis with active hydrocephalus. The objective of the present study was to evaluate the results of the different available surgical options adopted in the authors Institution and to compare personal results with data from the literature

**MATERIAL-METHODS:**We retrospectively analyzed the data of all patients affected by syndromic CRS with clinical and radiological evidence of active hydrocephalus at diagnosis admitted in our Department from January 2000 to January 2016

**RESULTS:**22 patients with complex CRS and concomitant Hy were identified (F=12, 10=M, mean age: 5,4 mts). Thirteen of these children were affected by Crouzon syndrome (59,1%), 4 (18,2%) by Pfeiffer syndrome, 2 (9,1%) by Apert syndrome, 2 (9,1%) by oxycephaly and 1 (4,5%) by a Mercedes Benz

syndrome. Parieto-occipital expansive cranioplasty was the primary procedure attempted in all cases. At a mean follow-up of 6.5 yrs., the hydrocephalus was controlled only by this procedure in 15/22 children (68,2%). The clinical and radiological persistence of the hydrocephalus led to implant a VP shunt in 5 cases (5/22 cases=22,7%); shunt implantation was complicated by multiple shunt revisions in 1 case and by recurrence of the cranial constriction, requiring a further cranial expansion in the remaining 4 cases.

The last two children (2/22=9.1%) were successfully managed by an endoscopic third ventriculostomy (ETV), aided by a temporary ventriculo-subgaleal shunt in one of them.

**CONCLUSIONS:**Parieto-occipital expansion cranioplasty is the primary procedure to consider for the management of active hydrocephalus in children with complex craniosynostoses. VP shunt should be avoided whenever possible due to the high rate of related complications. ETV can be considered as an alternative in case of the active persistence of the ventricular dilation.

**Keywords:** Complex craniosynostosis, hydrocephalus, expansion cranioplasty

Monday, 24 October 2016

17:10 – 18:00

## Flash Session 3: Epilepsy & Functional

### FL-006

#### Special topic: Neuro-imaging

#### 99mTc -HMPAO SPECT co-registered to MRI in children candidate for epilepsy surgery

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**OBJECTIVE:**In the pre-operative planning of epilepsy surgery SPECT is still the single imaging entity that can depict cerebral blood flow ictally and therefore in special cases with MRI negative patients can ad valuable information of the potential seizure onset zone. Subtraction of interictal from ictal SPECT images with subsequent co-registration to MRI (SISCOM) is an ad on with higher sensitivity especially in extra-temporal regions. The aim of this study was to demonstrate the usefulness of SISCOM in children with medical intractable epilepsy in the diagnostic workup and pre-operative planning of epilepsy surgery.

**MATERIAL-METHODS:**Forty-three children (mean ages 11,5 yrs; range 3-18 yrs, 27 male) were enrolled in the studied from 2009-2015. Ictal injection could be initiated on either clinical seizure (n=35) or at start of EEG changes (n=8). SPECT scanning was performed using a three-headed IRIX SPECT scanner. For the MRI co-registration we used the MRI done as part of the standard epilepsy-surgery program and a post-operative MRI control.

**RESULTS:**In 3 children SISCOM was not completed since the interictal study or the MRI could not be performed. 27 children had a positive SISCOM with focal hyperperfusion. SISCOM was negative or inconclusive in 13 children (3 with fast secondary generalisation, 6 short seizure <30 sec, 1 no seizure). 14

children subsequently underwent surgery of which 4 had a negative SISCOM. SISCOM was concordant to site and lobe of surgery in 80%.

**CONCLUSIONS:**In a little more than one third of the children referred to a SISCOM examination the children go on to surgery even though 29% have a negative SPECT. Negative SPECT scans are often associated with fast generalisation or very short seizure. SISCOM was concordant to site and lobe of surgery in 80%.

**Keywords:** SPECT, SISCOM, Epilepsy Surgery

**FL-007**

### Epilepsy and functional

#### Surgical outcome and neurocognitive development after surgery in infantile intractable epilepsy patients

Ju Seong Kim<sup>1</sup>, Chae Lin Lee<sup>1</sup>, Ju Hyeon Kim<sup>1</sup>, Eun Kyung Park<sup>1</sup>, Kyu Won Shim<sup>1</sup>, Seung Woo Park<sup>2</sup>, Dong Seok Kim<sup>1</sup>

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<sup>2</sup>Department of Neurosurgery, Hospital of Kangwon University, Chuncheon, Korea

**OBJECTIVE:**Infantile intractable epilepsy, such as cortical malformation and infantile spasm could have a significant effect on the long term seizure control and neurocognitive development. However, major neurosurgery in infant patients that are very difficult and dangerous procedures. Nevertheless remove the epileptogenic focus through early surgical treatment can achieve good epilepsy control and appropriate neurocognitive development

**MATERIAL-METHODS:**This study was designed retrospective analysis for Infantile epilepsy patients who underwent epilepsy surgery from 2006 to 2015 in Severance Children's Hospital. We have analyzed the neurocognitive development before and after surgery, as well as epilepsy surgical outcomes and complications.

**RESULTS:**Totally, 109 patients underwent epilepsy surgery. (Corpus callosotomy; 33, Resective surgery; 48, Hemispherotomy; 22, Posterior quadrantectomy; 3, Endoscopic hamartoma disconnection; 1, Resective surgery due to lateralization after callosotomy; 2) 78% of patient achieved seizure free. And among the patient except callosotomy, it showed a successful seizure control rate (>90% reduction) at 98%. In addition, neurocognitive outcomes were could not reach the standard development of the same age groups but it could be confirmed the inner personal development.

**CONCLUSIONS:**Epilepsy surgery in infant is difficult and dangerous procedure, but it could help to achieve the excellent epilepsy control rate and proper development of neurocognitive functions.

**Keywords:** infant, outcome, epilepsy surgery, neurocognitive development

**FL-008**

### Epilepsy and functional

#### Epilepsy surgery in children below five years: an institutional experience

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**OBJECTIVE:**The diagnosis and treatment of epilepsy has evolved with the advent of better investigative techniques changing the approach towards epilepsy.

We present our experience of surgery for refractory epilepsy in children below 5 years of age.

**MATERIAL-METHODS:**Twenty eight children below 5 years, diagnosed to have epilepsy, were included in this study. All these patients were on multiple anti-epileptic drugs for adequate period without any relief from seizures.

The patients underwent thorough evaluation. Fourteen of these children had focal cortical dysplasia. Three had perinatal stroke, two each had tumors and tuberous sclerosis. There were one each with neonatal hypoglycemic brain injury, lobar holoprosencephaly and hemimegalencephaly. Four children had non-specific radiological findings.

Fourteen patients underwent lobar resections, seven underwent hemispheric procedures, one underwent corpus callosotomy, and the rest underwent lesionectomies.

**RESULTS:**The age for onset of seizures was as early as day one, and the average age at surgery was 2.7 years. The youngest was operated at 9 months and the eldest at five years. Thirteen were operated on the right side while another thirteen for the left.

Histopathology revealed various pathologies including focal cortical dysplasia, tumours, gliosis.

Fourteen were completely seizure free while four had a rare occasional episode. Three patients had more than 70% reduction in seizure frequency while another four had a seizure freedom of less than 70%. Two children had the same frequency of seizures as pre-op. The longest follow up has been thirteen years with an average of 4.5 years.

**CONCLUSIONS:**Improved understanding of etiopathology and natural evolution of refractory epilepsy permits early surgery in young children.

Resective procedures and patients with hemispheric pathologies have a better surgical outcome.

**Keywords:** Epilepsy surgery, Children below five years

**FL-009**

### Epilepsy and functional

#### The features of pediatric epileptic surgery

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**OBJECTIVE:**The aim of this study was to reveal the features of pediatric epilepsy and its surgical management.

**MATERIAL-METHODS:**We retrospectively reviewed the data of 108 patients who underwent epilepsy surgery in Osaka University Hospital during last 5 years. Surgical procedures include radical surgery, corpus callosotomy, and implantation of veragus nerve stimulation system. We analyzed accompanying mental retardation, etiology of the epilepsy, location of epileptic focus, and outcome of surgery. The location of epileptic focus was diagnosed with semiology, MRI findings, MEG, scalp-EEG, video-EEG, ECoG, and FDG-PET. The outcome of surgery was evaluated with the ILAE new classification.

**RESULTS:**Of 108 patients, 45 patients were under 16 year-old. About 70% of pediatric patients had mental retardation. This ratio is higher than that of adult patients. The epilepsy focus located in the temporal lobe in 20%, in the extratemporal lobe in 13%, and multilobar, multiple, or unknown were in 67% of patients. The radical surgery was undergone in half of pediatric patients and 67% of adult patients. Surgical outcome was categorized Class 1 in 73% pediatric patients with radical surgery, which was comparable to that of adult patients.

**CONCLUSIONS:**The features of pediatric epilepsy were very different from those of adult epilepsy in many aspects including focus location, accompanied mental retardation. The short term surgical outcome of both groups was, however, almost comparable.

**Keywords:** epileptic surgery, pediatric epilepsy

**FL-011****Epilepsy and functional****Callosotomy in children: complications and perioperative blood loss**

Daniel T Nilsson, Bertil Rydenhag

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**OBJECTIVE:** Corpus callosotomy is indicated for intractable drop attacks. Data on complication rates after this procedure is scarce. We reviewed the complication rates and perioperative blood loss in callosotomy in children operated 1991–2015 and compared complications in children aged 0–7 and 8–18 years.

**MATERIAL-METHODS:** Children aged up to 18 years that had undergone a callosotomy at Sahlgrenska university Hospital, Gothenburg, Sweden 1991–2015 were identified. Postoperative complications and perioperative blood loss were recorded. Complications were defined as minor if symptoms resolved within three months or major if lasting more than three months.

**RESULTS:** Thirty-seven children, 16 girls and 21 boys had a callosotomy. Eight children had an anterior 2/3 callosotomy, 29 had a complete callosotomy. Mean age was 7.8 (range 0.5–17) years, 22 children were age 0–7 and 15 children were age 8–18. Four (11%) patients had a transient left paresis of the leg, one of them also in the arm, all of them in the group 8–18 years. Ten (27%) patients had a marked drowsiness postoperatively, 8 (53%) were in children aged 8–18, lasting 2–14 days and two (9%) were in the younger age group, lasting one and two days respectively. One cerebrospinal fluid leak and one case of air embolus terminating surgery (caused by a malfunctioning infusion pump) before callosotomy was started were found (patient excluded from further analysis). Mean perioperative blood loss (data available in 24 children) was 113 (range 40–400) ml, one patient had a blood transfusion.

**CONCLUSIONS:** Corpus callosotomy is a low-risk procedure, even though we found a higher rate of transient hemiparesis and drowsiness in children over age 7. Mean perioperative blood loss was low, rarely requiring transfusion. No permanent morbidity or mortality was found.

**Keywords:** callosotomy, complication, epilepsy surgery

**FL-012****Epilepsy and functional****Long-term seizure remission and developmental gains after total corpus callosotomy in children with intractable epilepsy**

Masaki Iwasaki<sup>1</sup>, Mitsugu Uematsu<sup>2</sup>, Naomi Hino Fukuyo<sup>2</sup>, Shin Ichiro Osawa<sup>1</sup>, Yoshiteru Shimoda<sup>1</sup>, Kazutaka Jin<sup>3</sup>, Nobukazu Nakasato<sup>3</sup>, Teiji Tominaga<sup>1</sup>

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<sup>3</sup>Department of Epileptology, Tohoku University Graduate School of Medicine, Sendai, Japan

**OBJECTIVE:** This study was aimed to determine what preoperative profiles were associated with seizure remission after corpus callosotomy and whether such seizure outcome was associated with the postoperative developmental outcome.

**MATERIAL-METHODS:** This retrospective study included 26 consecutive patients with childhood onset epilepsy who underwent one-stage total corpus callosotomy at our institution and were followed up for a minimum of 1 year. The age at surgery ranged from 13 months to 32 years (median 6 years). The association between postoperative seizure freedom and preoperative profiles, post-operative developmental gains was examined.

**RESULTS:** Five patients achieved seizure freedom (Engel class I), and 10 patients achieved worthwhile reduction of seizures (class III), whereas the remaining patients had a class IV outcome. All five seizure-free patients had "lack of abnormal magnetic resonance imaging findings", "lack of proven etiology of seizures", and underwent "surgery at age 6 years or younger". These three factors were associated with seizure freedom ( $p < 0.05$ , Fisher exact test). Post-operative gains in developmental quotient were significantly better in patients with seizure freedom than in those without ( $p < 0.05$ , Mann Whitney U test).

**CONCLUSIONS:** Our study replicated the notion that seizure remission can be achieved after total corpus callosotomy in subsets of patients with medically-uncontrolled epilepsy, and suggested that a better developmental outcome can be expected in patients benefiting from seizure freedom.

**Keywords:** Epilepsy surgery, Corpus callosotomy, Development, Seizure outcome, Generalized epilepsy

**FL-013****Epilepsy and functional****Clinical outcome of total corpus callosotomy in a single epilepsy centre**

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**OBJECTIVE:** Corpus callosotomy is good to alleviate drug-resistant generalized epileptic seizures, especially when definitive epilepsy surgery is not indicated. Total corpus callosotomy may have even better seizure outcome without increase incidence of disconnection syndrome. The object of this study was to investigate clinical outcomes of total corpus callosotomy in our institute.

**MATERIAL-METHODS:** Thirteen consecutive patients with infantile or early childhood onset epilepsy underwent 1 or 2 staged total corpus callosotomy for alleviation of seizures. Their age at complete surgery ranged from 2 months to 24 y/o (median 6 years). All patients suffered from generalised seizures. 2 patients had MRI abnormality, including bilateral periventricular nodular heterotopia and band-form heterotopia. The postoperative seizure outcome, cognitive function, and quality of life were evaluated.

**RESULTS:** Post-operative seizure reduction was most effective in atonic fall, followed by generalised tonic seizures. All patients showed improvement in life quality and stable or gained in cognitive function. Post-operative disconnection syndrome was usually transient with apathy, akinetic mutism (10 cases, < 2 weeks), and alien hand syndrome (2 cases, < 6 months).

**CONCLUSIONS:** Complete seizure remission is still rare after total corpus callosotomy in drug-resistant epilepsy. One-stage total corpus callosotomy at a young age patients or older patients with intellectual disability may provide a good and safe choice of palliative epilepsy surgery.

**Keywords:** epilepsy surgery, corpus callosotomy, seizure outcome, cognitive outcome, life quality

**FL-014****Epilepsy and functional****Vagal nerve stimulator revision surgery in children**

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<sup>2</sup>Department of Child Neurology, Maastricht University Medical Center, Maastricht, The Netherlands

**OBJECTIVE:**With a growing number of vagus nerve stimulation (VNS) implants for refractory epilepsy, a growing need for removal or replacement of non-functional or defective VNS systems has emerged. Whereas pulse generators are removed or replaced routinely, leads are considered more challenging because of scarring close to larynx, large vessels, and electrode-nerve complex. We report our surgical technique, intraoperative findings, postoperative complications, and outcome with regard to seizure control.

**MATERIAL-METHODS:**We retrospectively reviewed our patient registry for all VNS surgeries performed between January 2007 and January 2016. All individuals who had their electrode removed or replaced before the age of 18 years were included. Surgical reports, pre- and postoperative files including follow-up from the referring child neurologist were analysed.

**RESULTS:**In 16 children (including 10 boys) the system was completely removed (n=6), completely revised (n=5), or explored resulting in one additional explantation and four new pulse generators without electrode change. Revision indications were low impedance (n=1), high impedance with (n=3) or without (n=1) seizure increase, system failure notice with (n=1) or without (n=1) seizure increase, system non-operational for some time with (n=1) or without (n=3) discomfort, battery near-end-of-life with seizure increase (n=1), battery empty without communication (n=2), increasing spastic hemiparesis (n=1), and need for MR imaging (n=1). Relevant intraoperative findings were defective electrode insulation (n=3), overstretched or broken electrode (n=2), a loose screw in between pulse generator and electrode without actual disconnection (n=1), and an atrophic nerve (n=1). Intended complete electrode removal was always successful (n=7). There were no infections, no major complications, and one small laceration of the jugular vein. Seizure frequency was unaltered or improved.

**CONCLUSIONS:**In our experience, VNS revision surgery in children is feasible and safe. Although initial results are promising, additional follow-up is needed to assess whether lead replacement may affect VNS effectiveness.

**Keywords:** epilepsy, lead, neuromodulation, revision, vagus nerve stimulation

## FL-016

### Epilepsy and functional

#### MRI laser ablation for cavernomas in children: initial experience in 6 cavernomas and technical notes

Zulma Sarah Tovar Spinoza, Kalliopi Petropoulou  
SUNY Upstate Medical University

**OBJECTIVE:**Evaluate the effectiveness of MRI guided laser ablation (LITT) in the treatment of brain cavernous malformations (cavernomas).

**MATERIAL-METHODS:**Three pediatric patients with small (less than 3 centimeters) cavernous malformations and five cavernomas were treated with MRI guided LITT from 2014 until April 2016. We used the Visualase (Medtronic, USA) and Neuroblate (Monteris, USA) systems. We revised the effectiveness of the use of different lasers and the volumetric follow up of the ablated lesions using sequential MRI.

**RESULTS:**One ablation was complicated with post ablation bleed. All the cavernomas demonstrated reduction in volume, one cavernoma required a second ablation due to incomplete first ablation. We found differences on results given the type of laser and the technique of ablating the lesions.

**CONCLUSIONS:**MRI guided LITT is an alternative option for treating small cavernous malformations specially if they are located in deep locations in the brain.

Appropriate surgical technique and use of the application are essential for the success of this procedure.

**Keywords:** Laser ablation, cavernous malformation, MRI, pediatrics.

Tuesday, 25 October 2016

08:50 – 10:15

## Flash Session 4: Trauma and Cerebrovascular

### FL-037

#### Trauma

#### Intracranial hemorrhage in newborns (excluding intraventricular hemorrhage in premature infants)

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**OBJECTIVE:**To analyze common characteristics of intracranial hemorrhage in newborn infants.

**MATERIAL-METHODS:**We retrospectively reviewed charts from cases of intracranial hemorrhage in newborns seen at our department from April 2012 to March 2016. Cases of intraventricular hemorrhage or subependymal hemorrhage in premature infants were excluded.

**RESULTS:**This study included 22 newborns, including seven premature (median gestational age 35.7 weeks) and fifteen mature newborn infants (median gestational age 39.0 weeks). The median birth weight was 2699 g; this included six infant with low birth weight and one with very low birth weight. The most common symptoms at presentation were apnea (32%), seizures (18%), vomiting (18%), asphyxia (18%). Ten infants (45%) had subdural hematoma. Four infants (18%) had spontaneous superficial parenchymal or leptomeningial hemorrhage. Six infants (27%) had intraventricular hemorrhage. Two infants (9%) had intraparenchymal hemorrhage. Eighteen infants (82%) had intracranial hemorrhage due to injuries during birth; 44% of these infants were delivered with mechanical assistance by vacuum or forceps extraction. Two infants (9%) had intracranial hemorrhage due to intracranial hemorrhage in the womb. One infant (5%) had intraventricular hemorrhage due to hypoxic-ischemic encephalopathy. One infant (5%) had a subdural hematoma due to hemophilia. Two infants (18%) required neurosurgical treatment due to the intracranial hemorrhage. Only four infants (18%) had a neurodevelopmental disorder.

**CONCLUSIONS:**Although this series is too small to draw firm conclusions, it is remarkable that the highest rates of intracranial hemorrhage in newborns were subdural hematoma due to birth injuries.

**Keywords:** intracranial hemorrhage, term newborn infants, birth injuries, choroid plexus hemorrhage

### FL-038

#### Trauma

#### Correlation between retinal hemorrhages and acute subdural hematomas of infants

Ishizaki Ryuji, Wataya Takafumi, Tashiro Yuzuru

Department of Neurosurgery, Shizuoka Children's Hospital, Shizuoka, Japan

**OBJECTIVE:**Retinal hemorrhages are often seen in infants with abusive head trauma(AHT), and characteristic findings are widespread multilayered retinal hemorrhage(RH) and acute subdural hematoma(ASDH). We examined characters of retinal hemorrhages of infants associated with ASDH.

**MATERIAL-METHODS:**We performed a retrospective review of 14 cases of under 1-year infants diagnosed traumatic ASDH and RH in our hospital from April, 2007 to December, 2014. We classified them by correlation of laterality of RH and ASDH with the examination of each characteristic.

**RESULTS:**There were four types of correlation between RH and ASDH. Type 1 consists of 6 cases combined with unilateral ASDH and ipsilateral RH. Type 2 includes 4 cases with unilateral ASDH and contralateral RH. Type 3 has 2 cases with unilateral ASDH and bilateral RH. Type 4 involves 2 cases with bilateral ASDH and RH. Characteristics of type 1 were under 6 month old, other image findings such as contusion and brain atrophy, and developmental delay, while these of type 2 were over 6 month old, no other image findings, and normal development. Characteristics of type 3 were over 6 month old, brain swelling, and sequelae of hemiparesis and developmental delay. There were apparent differences between type 1 and type 2, which may become the clue to judge whether AHT or not. Type 3 cases showed signs of high intracranial pressure and bilateral RH, suggesting that the cause of type 3 is different from that of two former types.

**CONCLUSIONS:**These results might suggest that ASDH with ipsilateral RH could be caused by AHT.

**Keywords:** retinal hemorrhage, acute subdural hematoma, infant, abusive head trauma

#### FL-039

##### Trauma

#### Outcome comparison between surgically treated chronic subdural hematoma and subdural effusion in infants

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<sup>2</sup>Department of Neurosurgery, Hospital Kuala Lumpur, Kuala Lumpur, Malaysia

**OBJECTIVE:**Chronic subdural hematoma (CSDH) and subdural effusion (SDE) are two common conditions in infants who may require subdural drainage. CSDH in infants are mostly due to non-accidental injury (NAI). SDE occurs in infant with acute bacterial meningitis. This paper is to compare the outcome between surgically treated CSDH and SDE in infants at Hospital Sungai Buloh, Malaysia.

**MATERIAL-METHODS:**Infants with final diagnoses of either CSDH or SDE who have been treated surgically with subdural drainage between 2008 and 2015 were recruited.

**RESULTS:**CSDHs were found in 21 cases (52.5 percent) while remaining 19 cases were SDEs (47.5 percent). Only one case of CSDH presented with history of recent head injury, while remaining 20 cases were diagnosed with suspected NAI. The mean ages at presentation were 3.76 months (range 3 to 7 months) and 3.37 months (range 1 to 6 months) in both CSDH and SDE groups respectively. The gender was predominantly male with 66.7 percent and 78.9 percent respectively.

The follow-up periods were from minimum four months up to maximum of seven years. The neurological outcomes of both CSDH and SDE groups were almost similar with no significant statistical differences between groups. One infant in the SDE group succumbed due to Salmonella infection. Two infants from each CSDH and SDE groups defaulted follow-up. In both CSDH and SDE groups, 14.3 and 10.5 percent developed cerebral palsy, 19.1 and 26.4 percent developed motor deficits, while 19.0 and 15.8 percent respectively

were diagnosed as epilepsy that require long term anti-epileptic medications and rehabilitation. Only 57.1 and 47.4 percent respectively improved with normal developmental milestones.

**CONCLUSIONS:**The outcomes of both surgically treated infants with CSDHs and SDEs were similarly poor. Almost half of infants with CSDHs which were mostly due to non-accidental injury and half of infants with SDEs suffer from long term neurological sequelae.

**Keywords:** chronic subdural hematoma, subdural effusion, infant, non-accidental injury, acute bacterial meningitis

#### FL-041

##### Trauma

#### Decompressive craniectomy for severe traumatic brain injury in children

Kenichi Usami<sup>1</sup>, Juliette Montmayeur<sup>2</sup>, Kevin Beccaria<sup>1</sup>, Thomas Blauwblomme<sup>1</sup>, Giovanna Paternoster<sup>1</sup>, Philippe Meyer<sup>2</sup>, Marie Bourgeois<sup>1</sup>, Timothé De Saint Denis<sup>1</sup>, Cyril James<sup>1</sup>, Michel Zerah<sup>1</sup>, Christian Sainte Rose<sup>1</sup>, Stephanie Puget<sup>1</sup>

<sup>1</sup>Department of Pediatric Neurosurgery, Necker Hospital, Université Paris Descartes, Sorbonne Paris Cité, Paris, France

<sup>2</sup>Department of Pediatric Neuro-Critical Care and Anesthesiology, Necker Hospital, Université Paris Descartes, Sorbonne Paris Cité, Paris, France

**OBJECTIVE:**Decompressive craniectomy (DC) is performed to decrease the raised intracranial pressure as a second-line treatment for severe traumatic brain injury (sTBI). The definitive indication and the optimal timing of DC for sTBI in pediatric patients are still unclear. Objectives of this study are to clarify the clinical characteristics of pediatric patients who underwent DC for sTBI and to identify prognostic factors.

**MATERIAL-METHODS:**Thirty-three children who underwent DC for sTBI between 1999 and 2015 in a single center were retrospectively reviewed. Patients in whom the worst GCS score was more than 8 before DC were excluded. Median age at injury was 7.7 years (1.2 – 15.0). For analysis, patients were divided into two groups based on Glasgow Outcome Scale (GOS) score, poor outcome (GOS score 1 - 3, n=18) and favorable outcome (GOS score 4 and 5, n=15).

**RESULTS:**We found statistical significant association between the 2 outcome groups. (1) the initial GCS score evaluated by ambulance staffs (5.1±2.2 vs 7.0 ±2.4, p=0.047), and (2) the delay from injury to DC (32.5±47.9 vs 9.5±10.8 hours, p=0.04). Age, clinical symptoms, neuroradiological findings at initial evaluation, and the value of intracranial pressure (ICP) did not affect outcome.

**CONCLUSIONS:**Early DC achieves favorable outcome regardless of the value of ICP. From our results, DC should be performed as soon as possible as the GOS depends on the delay from injury to DC irrespective of the age, type of lesion, or type of injury.

**Keywords:** severe head trauma, surgical indication, optimal timing, outcome, prognostic factor

#### FL-042

##### Trauma

#### Referral pattern and outcome in pediatric cervical spine trauma in a UK pediatric major trauma centre: a two year prospective case series study

Vita Stagno, Balazs Markia, Richard J Edwards

Department of Neurosurgery, Bristol Royal Hospital for Children, Bristol U.K.

**OBJECTIVE:**Pediatric cervical trauma is a rare. There is significant variability between centres in the practice of investigating and treating

pediatric cervical spine trauma. The aim of the study is to audit our practice in a UK Pediatric Major Trauma Centre.

**MATERIAL-METHODS:**A referral database with continuous prospective data collection was used to identify all cervical spine trauma referrals. Data analysis was done based on presenting symptoms, mechanism of injury, diagnostic modalities, findings on imaging, usage of fixation collar and treatment modalities.

**RESULTS:**35 patients were referred to neurosurgery in our regional pediatric major trauma centre with cervical spine trauma over a 2-year period. Average age: 10-years. Leading presenting symptom was torticollis in under-4s and paraesthesia followed by pain alone in older children. Only 4 children had a motor deficit. Midline pain was not present in 5 cases. Sports injury (n=14) was the most common mechanism of injury (rugby (n=9), trampolining (n=3) and horse riding (n=2)); MVC was the cause in only 3 cases. Imaging: CT scan (n=28) and MRI (n=24), both modalities (n=18). 3 patients had isolated ligamentous injuries, 2 of which were unstable, both of which were also associated with CT abnormalities. MRI was used in all cases with neurological signs. Cross-sectional imaging was normal in 21 cases. 22 patients did not require treatment, 7 patients were treated with a hard collar, 2 patients with torticollis required MUA and collar; 2 with HALO fixation and 2 patients needed instrumented fusion. All children recovered without permanent neurological deficit.

**CONCLUSIONS:**Cervical spine injury accounts for only 2.3% of neurosurgical referrals in our busy tertiary unit. There is a significant shift in mechanism of injury, with sports injury now the leading cause in our paediatric population. There is a decrease in SCI caused by MVC. Only 17% of referred patients required surgical intervention.

**Keywords:** Cervical spine; trauma; MRI; ligamentous injury; Sports injury

#### FL-043

##### Trauma

##### Fractures of the skull base in children

Mikle Talabaev, Evgenij Mironec, Kevin Fernando Venegas Hidalgo  
Republican Research and Clinical Center of Neurology and Neurosurgery of Belarus, Department of pediatric neurosurgery

**OBJECTIVE:**to evaluate the frequency and features of the skull base fracture in children.

**MATERIAL-METHODS:**409 pediatric patients of 0 to 16 years were treated in the period from 2013 to 2015 in Republican Research and Clinical Center of Neurology and Neurosurgery, (94.4%) with moderate (TBI II) and (23, 5.6%) with severe (TBI III).

**RESULTS:**71 (17.4%) patients with TBI II and III were diagnosed with fractures of the skull base from which 64 (90.1%) had continued fracture to the skull base and in 7 (9.9%) patients had only base fractures (in 2 patients anterior cranial fossa (ACF) and 5 middle cranial fossa (MCF)). The majority of cases, 33 (46.5%) were consisted of patients aged 3 to 6 years. At the age of 2 years, this trauma was found only in 2 fractures of the frontal bone and the transition to ACF. Fracture of the base of the (ACF) was diagnosed in 38 (53.5%), (MCF) 31 (43.6%) patients, the fracture of both ACF and MCF diagnosed in 2 (2.8%).

In 34 (89.4%) patients with fractures of ACF were seen periorbital hematoma, in 4 (10.5%) were presented with nasal liquororrhea, strabismus associated with damage to the upper rectus muscle in 2 (5.3%). Fractures MCF were presented by otorrhea in 16 (51.6%) patients, hearing loss on the side of lesions in 6 (19.4%), feeling the crunch (Pneumarthrosis) in the area of the temporomandibular joint (TMJ) in 3 (9.7%), peripheral facial nerve paresis in 3 (9.7%). CT examination for fractures ACF revealed: pneumocephalus in 7 (18.4%), epidural hematoma in 3 (7.9%), concussion in 2 (5.3%) patients. Among traumatic injury with ACF present longitudinal fracture of the temporal bone pyramid made up - 77.4% (24), pneumocephalus met in 18 (58.0%), epidural hematoma in 5 (16.1%) patients. In 5 (16.1%) cases revealed no direct radiographic

evidence of fracture MCF base - the presence of air in the TMJ, including 2 patients it was the main fracture CT characteristic.

Epidural hematomas in all patients (8) required surgery. 2 patients ACF with nasal liquororrhea had treated.

**CONCLUSIONS:**1. isolated fractures of the skull base in pediatric patients are seldom; 2. Conservative treatment of liquororrhea in fractures of the skull base is effective in most patients. 3. Pneumarthrosis (TMJ) is rarely described and is not a direct sign of MCF fracture.

**Keywords:** Skull base fractures, temporomandibular pneumoarthrosis in children

#### FL-044

##### Other

##### Neurotrauma care depends on effective team attributes: evidence from a discrete choice experiment

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<sup>1</sup>University of British Columbia, Vancouver, BC

<sup>2</sup>British Columbia Childrens Hospital, Vancouver, BC

**OBJECTIVE:**Trauma hospitals rely on multidisciplinary healthcare teams to successfully manage acute patient admissions. The leader of a neurotrauma team is tasked with ensuring the team functions effectively to produce optimal patient outcomes. Although an effective trauma team leader is often thought to be self-evident - there is little formal literature identifying the leadership characteristics and attributes associated with optimal trauma team performance. The purpose of this study was to elicit the trauma team leader traits and characteristics deemed of greatest utility by members of the trauma team.

**MATERIAL-METHODS:**Semi-structured interviews with trauma team members in British Columbia, Canada were conducted to develop attributes and attribute levels for a discrete choice experiment (DCE). The DCE questionnaire contained 10 choice-sets with varying six attributes. Multinomial logit modeling was used to determine the relative utility of each attribute included in the experiment and segmented models were also developed to evaluate differences in preferences between subgroups.

**RESULTS:**Among the 64 respondents, 74% were female, 36% were physicians, and 36% had been members of the trauma team for more than 10 years. The attributes most strongly preferred by the respondents were collaborative, communication, and decisiveness. The attribute of least utility was experience. The specific leadership qualities that provided the most utility to the trauma team were "actively involves input for team" (0.70; SE: 0.11) and "concise communications, at time closed-loop" (0.52; SE: 0.09). "Hesitant and unclear communication" (-0.88; SE: 0.09) and "often indecisive" (-0.68; SE: 0.10) were deemed most detrimental to the team's function.

**CONCLUSIONS:**The findings of this study reveal the leadership characteristics preferred by trauma team members. Knowing the attributes of a successful team will result in education and refinement of team composition, which will incrementally improve neurotrauma care.

**Keywords:** Trauma, Education

#### FL-045

##### Vascular

##### Outcome of arteriovenous malformation treatment in children

Tristan Van Doornmaal<sup>1</sup>, Rob Lo<sup>2</sup>, Peter Woerdeman<sup>1</sup>, Gerard De Kort<sup>2</sup>, Albert Van Der Zwan<sup>1</sup>, Sen Han<sup>1</sup>

<sup>1</sup>Department of neurosurgery, University Medical Center Utrecht, Utrecht, The Netherlands

<sup>2</sup>Department of neuroradiology, University Medical Center Utrecht, Utrecht, The Netherlands

**OBJECTIVE:**Cerebral arteriovenous malformations (cAVM's) are rare lesions, especially in children. Therefore, treatment outcomes are rarely described. We aim to describe our institutional results.

**MATERIAL-METHODS:**A retrospective review was performed for all children (<18 yrs) treated for a cAVM in the UMC Utrecht, The Netherlands since 1996. Functional outcome was telephonically updated with parents or patients in December 2015. We used the modified Rankin scale (mRs) adjusted for children. A very good outcome was defined as a 100% AVM occlusion and a mRs 0 or 1 at follow up.

**RESULTS:**In total 25 children were treated. Nineteen children presented with hemorrhage, of which 11 had a poor functional score (mRs 4 or 5) at admission. Five children were treated with surgery alone and 5 with endovascular treatment alone. In the other children (n=15) a combination was used of surgery, endovascular treatment and/or stereotactic radiosurgery. At follow up (mean 9,7 yrs, range 2,3-20 yrs) all children were alive. Twenty-three children (92%) followed a normal education, had no significant disabilities and showed age appropriate development. (mRs 0 or 1, Figure 1.) Eighteen children (72%) had a very good outcome. A very good outcome was in an univariate analysis associated with Spetzler Martin grade < 3.

**CONCLUSIONS:**Treating AVMS's in children requires a multidisciplinary approach. In this series we showed a good functional outcome with this approach.

**Keywords:** Intracerebral Arteriovenous Malformations

**FL-046**

**Vascular**

#### **Multimodality management of pediatric intracranial arteriovenous malformations**

Yasser Ismail Orz<sup>1</sup>, Mahmoud Alyamany<sup>1</sup>, Sultan Alqahtany<sup>2</sup>, Hussain Alhussain<sup>3</sup>, Ayman Albanyan<sup>1</sup>, Maqsood Ahmad<sup>1</sup>, Ahmad Lary<sup>1</sup>, Mohamed Bafaqeeh<sup>1</sup>, Abdullah Alobaid<sup>1</sup>, Ihtsham Ghani<sup>1</sup>

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**OBJECTIVE:**To describe our experience treating pediatric arteriovenous malformation (AVM) as intracranial AVMs represent the most frequent vascular pathology in the pediatric age. Successful management of these AVMs often requires a balanced application of embolization, surgery, and radiosurgery.

**MATERIAL-METHODS:**We analyzed 70 pediatric patients (<18 years of age) with AVMs treated with various combinations of radiosurgery, surgery, and endovascular techniques.

**RESULTS:**Between 2006 and 2016, 50 children with various Spetzler-Martin grade AVMs were treated in our institute. Presenting history has been constituted mainly by intracranial hemorrhage, followed by epilepsy. The arteriovenous malformation was located in noncritical cortical areas in 25 cases, in critical cortical areas in 7 cases, in the midline structures or in the basal ganglia in 8 cases, in the posterior fossa in 10 cases. Complete surgical resection of the lesion has been carried out in 30 cases, embolization in 12 case and radiosurgery in 8 patients. Disabling neurological complications occurred in 3 patients (6%), no mortality occurred in our series. At the final clinical follow-up (mean, 4 years), 35 patients (70%) had a modified Rankin Scale score (mRS) of 0 to 1. On multivariate analysis, significant risk factors for poor final clinical outcome (mRS  $\geq$  2) included baseline mRS  $\geq$  2, left-sided or posterior fossa locations, and high AVM grade.

**CONCLUSIONS:**Treatment of pediatric AVMs with multimodality therapy can substantially improve obliteration rates and may decrease AVM hemorrhage rates. The poor natural history and risks of intervention must be carefully considered when deciding to treat high-grade pediatric AVMs.

**Keywords:** arteriovenous malformation, pediatric AVM

**FL-047**

**Vascular**

#### **Pediatric intracranial aneurysms: characteristics, presentation, management and outcome**

Yasser Ismail Orz<sup>1</sup>, Mahmoud Alyamany<sup>1</sup>, Sultan Alqahtany<sup>2</sup>, Mohamed Bafaqeeh<sup>1</sup>, Abdullah Alobaid<sup>1</sup>, Ayman Albanyan<sup>1</sup>

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<sup>2</sup>Neurointerventional Department, King fahad medical city, Riyadh, Saudi Arabia

**OBJECTIVE:**The aim of our study is to characterize the clinical, imaging, presentation, management, and outcome of patients younger than 18 years diagnosed with intracranial aneurysms at our institute.

**MATERIAL-METHODS:**A retrospective review of patients presented with intracranial aneurysms managed in our center from 2007-2016.

**RESULTS:**Of 300 patients with intracranial aneurysms managed in our institute during that period, 20 pediatric patients (6.6%) harboring 30 aneurysms were treated using microsurgical or endovascular techniques. thirteen patients (65%) presented with subarachnoid hemorrhage. Most of the aneurysms located at the anterior circulation (80%). eleven patients (55%) had their aneurysms located at the carotid artery bifurcation (CAB). Most of the aneurysms were small in size with wide necks and irregular shape, 5 patients had giant non ruptured aneurysms.

One patient had 8 aneurysms located at different locations and he experienced 2 attacks of SAH from 2 different aneurysms at 4 years interval and he died due to the second attack, another patient had 2 SAH attacks with 7 years interval the second attack was from ruptured do novo right PCom aneurysm. Most of the patients (84%) treated by microsurgical clipping of their aneurysms. Fifteen patients (85%) had good outcome and only one patient died in our series.

**CONCLUSIONS:**Pediatric Intracranial aneurysms pose considerable diagnostic and therapeutic challenges. Good outcome was achieved with both surgical and neurointerventional management of pediatric patients with intracranial aneurysms. Long life follow up with either CTA or angiogram should be considered for these patients.

**Keywords:** pediatric aneurysm, intracranial aneurysm

**FL-048**

**Vascular**

#### **Ruptured intracranial aneurysms in children: management and outcome**

Matthieu Vinchon<sup>1</sup>, Rabih Aboulais<sup>1</sup>, Irene Stella<sup>1</sup>, Xavier Leclerc<sup>2</sup>, Lejeune Jean Paul<sup>2</sup>

<sup>1</sup>Neurosurgery clinic, Lille university hospital, France

<sup>2</sup>Department of interventional neuroradiology, Lille university hospital, France

**OBJECTIVE:**Intracranial aneurysms (IA) are rare in children, and aneurysm rupture often presents with management difficulties in emergency, on account of young age and complex vascular pathology.

**MATERIAL-METHODS:**We reviewed retrospectively consecutive cases of ruptured IA (RIA) in children treated in our institution.

**RESULTS:**Between 1998 and 2016, we treated 20 children for RIA, 12 male and 8 female (M/F ratio Between 1998 and 2016, we treated 20 children for

RIA, 12 male and 8 female (M/F ratio 1.5), aged 0 to 17 years, 6 patients (30%) being under 24 months. Sixteen IA were dysplastic, including 2 associated with a genetic disease; 2 were traumatic, and 2 were mycotic. Fourteen patients were treated surgically, 2 with embolization, and 4 patients in desperate status were treated conservatively and died. Among 16 patients treated actively, after a mean follow-up of 55 months, 10 (62%) had good outcome (GOS 1); 1 had mild sequelae (GOS 2), 4 (25%) had severe sequelae (GOS 3), and 1 died. Repermeation of the aneurysm occurred in two patients: one after surgery and one after embolization, requiring repeat surgery and embolization respectively.

**CONCLUSIONS:** Aneurysm rupture is a major emergency in children with favorable outcome in the majority of patients. However, the surgical and non-surgical management of IA in children is difficult because these lesions are rare, diverse, complex, and often occur in infants. Training and maintenance of a competent workforce is a challenge for the pediatric neurosurgical profession. This highlights the importance of tight links between adult and pediatric neurosurgeons and neuroradiologists.

**Keywords:** intracranial aneurysm, aneurysmal rupture, outcome

#### FL-049

##### Vascular

#### Paediatric intracranial aneurysms: transition of a unit into endovascular management

Andile Lungani Mbatha<sup>1</sup>, Duncan Royston<sup>2</sup>, Yusentha Balakrishna<sup>3</sup>, Mogwale Samson Motebejane<sup>1</sup>, R. Harrichandpersad<sup>1</sup>

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<sup>2</sup>Department of Radiology, Entabeni Hospital

<sup>3</sup>BioStatistics Unit South African Medical Research Council, Durban.

**OBJECTIVE:** Intracranial aneurysms are rare in the paediatric population ( $\leq 18$  Years). The reported prevalence is 0.5 – 4.6%. We report on the presentation, aetiology, management and outcomes of patients managed at our institutions in a period of transition into endovascular management.

**MATERIAL-METHODS:** Retrospective review of medical records of patients treated from January 2003 to February 2016. Data was analysed for demographics, clinical presentation, Glasgow Coma Scale (GCS), radiological features, management and outcomes.

**RESULTS:** Twenty three patients, with a total of thirty one aneurysms were recruited. Mean age was  $12.4 \pm [4.5]$ . Nine patient were tested for HIV, three positive, one had CD4  $< 200$ . Mean GCS was  $13.2 \pm [2.9]$  with headache and hemiparesis, the most common symptoms. The commonest cranial nerve palsy were third [7, 30%] and seventh [4, 17%]. Radiologically findings showed subarachnoid haemorrhage [11, 48%], infarcts [4, 17%], intracerebral haematoma [8, 35%], hydrocephalus [4, 17%].

**FISCHER Grade:** Four [3, 13%], Three [7, 30%], One [3, 13%]. Post traumatic were [4, 17%] and unruptured [6, 26%]. Diagnostic investigations were CT cerebral angiogram (CTA) [12, 52%], MRI angiogram [6, 26%], Digital subtraction angiogram [5, 22%]. Aneurysm location was 20 (65%) anterior and 11(35%) posterior circulation. Mean aneurysm size was  $8.2\text{mm} \pm [0.44]$ . Morphology was fusiform (14, 45%), saccular (13, 42%), dissection (1, 3%), pseudo-aneurysm (3, 10%). Aetiology showed infective [5, 22%], unknown [11, 48%], traumatic [4, 17%], vasculitis [3, 13%]. Ten (47%) were managed medically, four (17%) microsurgery, nine (39%) endovascular. Mortality was 2(9%) one in hospital, associated with re-bleed, one at one month discharge. Mean GCS at discharge was  $13.6 \pm 3.3$ , mean

hospital stay  $20.8 \pm 12.7$  and median Glasgow Outcome Score (GOS) of 17 patients followed up was (GOS5) at 12 months

**CONCLUSIONS:** Paediatric aneurysms in our review have a predominance of fusiform type, unidentified cause, present good grade and have good outcomes.

**Keywords:** Glasgow Coma Scale:GCS

#### FL-050

##### Vascular

#### Direct surgical treatment of pediatric intracranial pial arteriovenous fistula

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<sup>2</sup>Department of Neurosurgery, Higashiosaka General Hospital, Higashiosaka, JAPAN

**OBJECTIVE:** Pediatric intracranial pial arteriovenous fistulas (PIAVFs) are rare vascular lesions and the history and developmental mechanisms of these lesions remain unclear. Their high flow vascular characteristic and pediatric populations cause difficulty in the treatment. We report the efficacy of direct surgical procedure of PIAVFs.

**MATERIAL-METHODS:** This study reviewed a series of seven cases of PIAVFs treated between 2005 and 2012. A mean age was 5.6 years (1 month–14 years). Among the seven cases, three cases developed hemorrhagic events (ICH in two cases and SAH in one) and the remaining four cases were unruptured. All PIAVFs were associated with large venous varices with a mean diameter 20.2mm (12–26mm). We selected direct surgery under intraoperative monitoring (IOM) surveillances and intraoperative neuroimaging (IONI) evaluations.

**RESULTS:** In all cases, shunt points were directly identified and completely disconnected. Especially, four cases were conducted barbiturate coma therapy to prevent postoperative hyperperfusion events. None of seven cases encountered surgical-related complications. All patients were neurologically excellent with no deficits at discharge. With a mean long-term clinical follow-up of 7.4 years (3.8–10.7years), there were no signs of recurrences.

**CONCLUSIONS:** Endovascular treatment of these patients is very difficult due to the high flow vascular lesions and distal migrations of embolization materials have been frequently reported. We recommend the direct disconnection of these lesions if they are located at the superficial lesions and we can easily approach them. It is obvious that IOM and IONI might be essential for the direct surgery in PIAVFs.

**Keywords:** pial AVF, pediatric, direct surgery

#### FL-051

##### Vascular

#### Intracranial arteriovenous malformations treated with Gamma Knife radiosurgery in children

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**OBJECTIVE:** Treatment of arteriovenous malformations (AVMs) with Gamma Knife Radiosurgery (GKS) has become widespread. Most reported series compose in adult or in pediatric and adult patients together. There are

limited published series which are composed only pediatric patients. This study aimed to research the results of therapy by Gamma Knife Radiosurgery in children with intracranial AVM.

**MATERIAL-METHODS:**A total of 219 pediatric patients aged  $\leq 18$  years old were treated with GKS at our clinic between January 1990 and December 2014. A retrospective study was performed of follow up data obtained in 194 pediatric patients after GKS. Statistical analyses were performed to determine clinical presentation, treatment methods, obliteration rates, and outcomes. The median age was 13 years and the median follow up time was 54 months.

**RESULTS:**The most common clinical presentation was hemorrhage in 64%. The mean volume was 3,2 cm<sup>3</sup>. The median prescription dose was 23 Gy (50%). The median Spetzler Martin grade was 2. The AVM obliterations rates at 2, 3, 5, and 10 years were 16%, 27,3%, 37,6%, and 46%, respectively. The median obliteration time was 36 months (r:12-168). 14(7,2%) patients had hemorrhages during the follow-up period.

**CONCLUSIONS:**Gamma Knife Radiosurgery is an effective treatment for AVMs and provides a minimally invasive alternative to surgical resection. Pediatric patients require a special consideration of treatment-related late effects due to the longer life expectancy. Therefore we need further study to know long term effects of GKS in pediatric patients.

**Keywords:** Arteriovenous malformations, intracranial, gamma knife, pediatric

#### FL-052

##### Vascular

#### Clinical manifestations of infantile moyamoya disease: a comparison with schoolage and pubertal moyamoya disease

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<sup>3</sup>Department of Neurosurgery, St. Luke's International Hospital, Tokyo, Japan

**OBJECTIVE:**Pediatric Moyamoya disease (MMD) is characterized by repeated transient ischemic attacks (TIAs) and cerebral infarctions. MMD in infants tends to progress rapidly, develops repeated ischemic strokes and has a poor prognosis. The aim of this study is to elucidate the clinical manifestations of infantile MMD by comparing them with those of schoolage and pubertal MMD.

**MATERIAL-METHODS:**A total of 32 patients with MMD who underwent direct with or without indirect bypasses and had medical follow-up not less than 1 year were included. They were categorized into 2 groups according to their age, one is group I (n = 12,  $\leq 6$  years) and the other is group SP (n = 20, 7 - 15 years).

**RESULTS:**Cerebral infarctions at initial presentation and postoperative ischemic strokes were significantly more frequent in the group I than the group SP (P < 0.001 and P < 0.01, respectively). All the postoperative ischemic strokes occurred within 2 weeks in the group SP, whereas they occurred even 2 years after the surgery in group I. Preoperative modified Suzuki angiographic staging, posterior cerebral artery stenoses and occlusions, preoperative mean cerebral blood flow (CBF), postoperative mean CBF increasing rate were not significantly different between groups.

**CONCLUSIONS:**The present study showed that infantile moyamoya patients have much higher risks of the repeated ischemic stroke after the revascularization in the long period though the effect of bypass surgery on CBF was equal to schoolage and pubertal patients. These findings may be because infantile patients have higher demand of

CBF with the development of cerebral function. In contrast, impaired cerebral metabolism due to cerebral infarctions might be reduce demand for CBF and could be difficult to maintain the long-term CSF demand. Early diagnosis and surgical revascularization should be considered.

**Keywords:** Moyamoya disease, infant, clinical manifestations

#### FL-054

##### Vascular

#### Is temporal indirect revascularization enough for pediatric patients with moyamoya disease?

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**OBJECTIVE:**Indirect revascularization, mainly to the temporal region, is commonly used in children with moyamoya disease. The clinical response to indirect revascularization appears to be favorable in most cases. However, selective neuropsychological impairments have been noted in pediatric moyamoya patients that suggests the frontal perfusion is important. This retrospective study aims to evaluate the effectiveness of additional revascularization of the frontal region by using quantitative methods.

**MATERIAL-METHODS:**From 2007 to 2015, moyamoya patients younger than 20-year-old were included. The clinical records, surgical method, perioperative complications, cerebral angiographic and perfusion studies were reviewed. Patients receiving only temporal revascularization and both frontal and temporal revascularization were compared. The time-to-peak (TTP) prolongation areas, which mean perfusion impairment areas, on magnetic resonance perfusion (MRP) studies and Matsushima grading on cerebral angiography were used to evaluate the revascularization effect.

**RESULTS:**Forty four patients were included. Twenty six of them were female and eighteen were male. The clinical symptoms improved in all patients after both kinds of surgery. Satisfactory collateral formation on the MCA territory, Matsushima grade A or B on cerebral angiography, could be achieved in 84% and 87% of the patients in both groups, respectively. Frontal revascularization induced additional collateral formation in frontal lobe and the watershed zone between ACA and MCA territories that significantly reduced the TTP prolongation area on MRP studies as compared with the patients who only underwent temporal revascularization. One patient undergoing both frontal and temporal revascularization had mild subdural hematoma that resolved without permanent neurological deficit after conservative treatment.

**CONCLUSIONS:**Though the symptoms of all patients improved after temporal revascularization, frontal revascularization additional increased the frontal perfusion that may improve the neurocognitive function of pediatric moyamoya patients. Routinely revascularize frontal and temporal area is suggested. The postoperative complication due to long operation time and increased operative area should be taken care of.

**Keywords:** moyamoya disease, indirect revascularization, magnetic resonance perfusion

#### FL-055

##### Vascular

#### Utility of time-resolved imaging of contrast kinetics (TRICKS) MRI in evaluating pediatric direct and indirect bypass surgery for moyamoya syndrome

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**OBJECTIVE:**Children with Moyamoya syndrome (MMS) have an increased risk of stroke based on the clinical and angiographic severity of disease. We have sought to investigate the utility of TRICKS (Time Resolved Imaging of Contrast Kinetics) MRI for assessing pediatric MMS surgical patients. This new imaging modality is safe, non-invasive, reproducible, and has not been studied in pediatric bypass surgery.

**MATERIAL-METHODS:**We studied 4 children who underwent direct (superficial temporal artery/STA to middle cerebral artery/M4) and indirect bypass (encephalo-duro-arterio-myo synangiosis; pial synangiosis) surgery for MMS in the past 12 months. Preoperative cerebral angiography was performed in all patients. Intraoperative assessment of direct bypass included a combination of Doppler, indocyanine green, and flow probe measurements. Post surgery TRICKS imaging was studied with intraoperative correlation.

**RESULTS:**Four patients (3 sickle cell disease, 1 Neurofibromatosis Type 1) underwent combined direct and indirect bypass surgeries for MMS disease over 1 year (Average age = 12+/-5) with a follow up of 2-12 months. Of these patients, three received TRICKS MRI after surgery with good intraoperative STA-M4 bypass correlation noted in 2 (50%) patients. Doppler and indocyanine green direct bypass confirmation were noted in 4/5 superficial temporal artery branches in 4 hemispheres. One patient was lost to follow. One patient had no flow detected in the bypass up to 5 months after surgery with no clinical consequences. One patient (25%) developed an osteomyelitis requiring bone removal. No patients suffered a new stroke or hemorrhage since surgery.

**CONCLUSIONS:**Based on our preliminary experience, TRICKS MRI may be useful for obtaining immediate diagnostic information on direct cerebral bypass patency. Correlating the TRICKS data with clinical follow up may help avoid unnecessary radiation or invasive risk related to conventional angiography, and may help limit the total angiograms these children get over a lifetime.

**Keywords:** Bypass Surgery, Moyamoya, TRICKS mri

## FL-056

### Special topic: Neuro-imaging

#### Evaluation of cerebral blood flow after indirect revascularization in pediatric patients of moyamoya disease using multiphase selective arterial spin labeling MRI

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**OBJECTIVE:**The selective arterial spin labeling (ASL) magnetic resonance imaging (MRI) has been developed as noninvasive evaluation of cerebral blood flow (CBF). The purpose of this study is to evaluate CBF pre- and post-operatively in pediatric patients with moyamoya disease.

**MATERIAL-METHODS:**We included 10 patients of moyamoya disease treated with encephalo-duro-arterial synangiosis (EDAS) between March, 2010 to December, 2015. Selective ASL were undergone pre- and post-surgery. The labeling slab was positioned at the external carotid artery and 5 slices in each of the 10 sequential phases were acquired using a 3-T scanner.

**RESULTS:**The mean age at initial presentation was 115 months (ranged 74 to 172). EDAS was performed on 16 hemispheres of 10 patients. Frontal EGS

were added in 3 patients. In all patients, clinical symptoms were improved with no perioperative ischemic complications. ASL showed improvement of CBF on follow-up examination in 12 hemispheres out of 16.

**CONCLUSIONS:**Multiple selective ASL technique can be useful non-invasive diagnostic tool for pediatric moyamoya disease patients.

**Keywords:** moyamoya, arterial spin labeling, MRI

## FL-057

### Other

#### Current trends and issues in the initial treatment of myelomeningocele in Japan: analysis using clinical data from the Disease Procedure Combination database

Mayu Takahashi<sup>1</sup>, Yoshihisa Fujino<sup>2</sup>, Shinya Matsuda<sup>2</sup>, Shigeru Nishizawa<sup>1</sup>

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**OBJECTIVE:**Incidence of myelomeningocele is considered to be lower in recent years. It is getting more difficult to collect clinical data of the patients for each hospital in Japan. Therefore, we examined current trends of the initial treatment of myelomeningocele in Japan.

**MATERIAL-METHODS:**We used the Diagnosis Procedure Combination (DPC) database. For this study, we extracted the patient data with the diagnosis of myelomeningocele from 2011 to 2013. They were all newborns and had surgical treatment for the myelomeningocele. We examined average gestational age, average body weight at birth, the date of the first surgery, the treatment for hydrocephalus, ventriculoperitoneal (VP) shunt related troubles and length of hospital stay.

**RESULTS:**There were 137 patients (male 61, female 76). Average gestational age was 35.5 ± 8.9 weeks and average body weight at birth was 2751.0 ± 421.5g. Forty-five patients underwent the first surgery within 24 hours, 43 within 48 hours, and 18 within 72 hours. Seventy patients needed VP shunt for hydrocephalus. Thirty-five patients (51%) were underwent VP shunt placement within 2 weeks from the first surgery. Fourteen patients (28%) needed the shunt revision. There was no statistical significance between shunt malfunction and gestational age/body weight at birth. Average length of hospital stay was 53.3 ± 59.1 days.

**CONCLUSIONS:**Though myelomeningocele is well-known congenital malformation, it is not easy for each institution to collect enough cases for analysis because of its incidence. Considering the data provided rate, the estimated incidence of myelomeningocele in Japan was approximately 80 per year. This is much less than we expected. The clinical data that we could extract from the DPC database was limited, however; it provides current trends and issues for myelomeningocele in Japan. It needs to continue collecting those data to establish standard treatment and to improve the patients' prognosis.

**Keywords:** Diagnosis Procedure Combination, initial treatment, myelomeningocele

Tuesday, 25 October 2016

09:00 – 10:25

## Flash Session 5: Spine

### FL-058

#### Spine

#### Surgical management of myelomeningocele: a series of 102 cases

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**OBJECTIVE:**The myelomeningocele (MM) is one of the most common birth defect of the central nervous system. Its frequency is unknown in our country but its causes social and public health problems.

**MATERIAL-METHODS:**we report a series of 102 cases of MM operated in our department between 2004 and 2014, there were 53 females and 49 males with a mean age of 156,5 days. The neurological examination revealed a complete paraplegia in 44 cases, 25 haven't any motor deficit but had orthopedic impairment. Complete fecal and urine incontinence are present in all patient with complete paraplegia, congenital scoliosis is present in 22 cases. Hydrocephalus has associated in 75,5% of the cases. Chiari I malformation is present in 96,1% of cases

Our management strategy consisted in VP shunt as first step when hydrocephalus was present 66 cases or at the same time as the treatment of (MM) in 5 cases. The post operative complications consisted in local wound healing in 23,5% cases, 14,7% of meningitis responsible on 5 deaths, acute pleurisy 2 cases and lyell disease one case induced dead.

**RESULTS:**the mean follow up period was 37,58 months ranged, normal gait in 53 cases that haven't deficit or presented with para-paresis, 42 patients in this series have gait difficulties. Complete paraplegia persists in the series with complete paraplegic or spastic para-paresis. Most of the patients kept sphincters disturbance. 37 of children have normal IQ, 12 didn't reach schooling yet and 58 didn't go to school because of insufficient IQ or social problems.

**CONCLUSIONS:**Medical, surgical, and rehabilitation issues arise in the patients with myelomeningocele from birth through adulthood. Its results from failure of the neural tube to close in the developing fetus and can be prevented by folic acid supplements. We suggest some recommendations in our country to prevent this abnormality.

**Keywords:** spina bifida, hydrocephalus, Chiari II malformations,

## FL-059

### Spine

#### Myelomeningocele: contemporary postnatal treatment and related outcome perspectives

Elke Januschek<sup>1</sup>, Andreas Roehrig<sup>2</sup>, Sandra Kunze<sup>2</sup>, Bea Wiebe<sup>3</sup>, Christian Fremerey<sup>3</sup>, Martina Messing Juenger<sup>2</sup>

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**OBJECTIVE:**1-year survival rate of children with open spinal dysraphism is reaching 87%. 78% of all patients become 17 years of age. Life expectancy and quality of life improved significantly. In order to compare treatment and prognosis of recent series after prenatal myelomeningocele closure, we evaluated a contemporary group of postnatally treated children.

**MATERIAL-METHODS:**In order to assess MMC-associated comorbidities, treatment modalities and prognosis we analyzed retrospectively 49 consecutive newborns with open spinal dysraphism treated after birth in our highly specialized institution between January 2007 and December 2015. Children, who were previously operated in utero were excluded. Type of neural tube defect, anatomical level and demographic information were also screened

**RESULTS:**All 49 cases could be included into the study. Gender distribution was equal (25 males, 24 females). In 88% closure was performed on day of delivery (max 36 hrs). The follow-up period ranges from 5 months to 9 years (loss of follow up in 2 cases). 85.7% of the patients had shunted hydrocephalus. 33 patients(67%) required shunt revision. 36 children showed significant Chiari malformation type II with associated syringomyelia in 23. Neurourological management: intermittend catheterization(85.7%), bladder stoma(2). Secondary neurosurgical interventions: Gardner decompression(2), myelolysis(5), diastematomyelia(1). Orthopedic surgery: kyphoscoliosis(5), hip deformity(3), foot deformities(9), quadriceps tendon extension(1). 70% are ambulatory w/wo orthoses and devices. Infants were assessed separately. No severe neurocognitive dysfunction was observed in this series. Beside shunt revisions no major complications were seen.

**CONCLUSIONS:**Contemporary postnatal MMC treatment is safe and provides good functional outcome without major risks for complications or life threatening conditions. Quality of life and number of secondary surgery has improved over the years. No marked cognitive dysfunction or renal failure have been observed. All children survived. Hydrocephalus and Chiari malformation are the most important comorbidities, but do not seem to interfere with a good overall prognosis.

**Keywords:** Neural tube defect – hydrocephalus - Chiari malformation – neurogenic bladder disorder

## FL-060

### Spine

#### Management of myelomeningocele in the province of KwaZulu-Natal, South Africa

Mthandeni Nkosinathi Mnguni<sup>1</sup>, Basil Claude Enicker<sup>1</sup>, Thandinkosi Enos Madiba<sup>2</sup>

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<sup>2</sup>Professor of Surgery, Head: Department of Surgery, University of KwaZulu-Natal, Durban

**OBJECTIVE:**To investigate the clinical presentation, characteristics, surgical repair, management of hydrocephalus and factors contributing to outcome children with myelomeningocele treated at a single institution.

**MATERIAL-METHODS:**Retrospective analysis of 307 consecutive children with diagnosis myelomeningocele who were managed at Inkosi Albert Luthuli Central Hospital from January 2006 to December 2014. Multiple logistic regression analysis identified clinical, demographic and surgical variables that were associated with outcome.

**RESULTS:**There were 173 (57%) males. One hundred and thirty five (44%) were delivered via C-section. MMC location was at lumbar [175, 57%], sacral [21, 7%], thorax [17, 7%] and cervical 2 (0.7%). One hundred and fifty six (51%) presented with CSF leak. One hundred and forty one (48%) presented with complete paralysis of the lower limbs. The mean age at surgical repair was 4.7 ± 15.6 months. The dura was repaired primarily in all patients and 20 (7.9%) were operated in conjunction with plastic surgeons.

Fifty-eight (21%) patients developed wound sepsis of those 35(60%) required surgical debridement, while chemical debridement was sufficient in the rest. The time to wound sepsis was 9.5 ± 3.6 days. Two hundred and eight (68%) children developed hydrocephalus requiring CSF diversion. In 143 (46%) children a VPS was used to treat HCP, while in 27 (9%) ETV was performed, 33 (23%) developed shunt malfunction due to infection [15, 45%]. The time to shunt infection was 176 ± 83.3 days.

The in-hospital mortality was 9.1% (27). Wound sepsis and meningitis were associated with death (p<0.002). The average hospital stay was 20,4 ± 16,93

days, children with wound sepsis and/or meningitis had longer hospital stay,  $38.1 \pm 22.3$  days and  $40.5 \pm 29.7$  days respectively.

**CONCLUSIONS:** Method of delivery, age at repair, CSF leak and hydrocephalus were not independently associated with infection. Infection was associated with adverse outcomes and yields longer hospital stay.

**Keywords:** ETV, Endoscopic Third Ventriculostomy MMC, Myelomeningocele IALCH, Inkosi Albert Luthuli Central Hospital

**FL-061**

## Spine

### Long-term follow up of 136 patients with myelomeningocele: predictors of good prognosis following postnatal surgery

Nelci Zanon<sup>1</sup>, Giselle Coelho<sup>2</sup>, Teresa Uras Belém<sup>3</sup>, Luanda Collange Grecco<sup>1</sup>

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<sup>2</sup>Santa Marcelina Hospital - São Paulo - Brasil

<sup>3</sup>Samaritano Hospital - São Paulo -Brasil

**OBJECTIVE:** To analyse retrospectively the results of 136 MMC patients and predictors of good prognosis

**MATERIAL-METHODS:** Retrospective study of a convenience sample of cases followed in the pediatric neurosurgery service. Complications, associated malformations, other surgeries and gait were analysed. Pearson's Chi Square test was used to analyze the association between categorical variables.

**RESULTS:** The analysis demonstrated that the main factor associated with better or worse neurological outcome and motor deficit was the level of MMC, with higher levels associated with worse prognosis. The MMC level was significantly associated with ambulation ( $X^2 = 21.9$ ,  $V = 0.40$ ,  $p = 0.001$ ) and scoliosis ( $X^2 = 17.2$ ,  $V = 0.36$ ,  $p = 0.001$ ), clubfeet ( $X^2 = 10.2$ ,  $V = 0.27$ ,  $p = 0.0016$ ). Although not associated incidence of hydrocephalus and Chiari malformation, the level of the lesion showed a significant association with symptomatic Chiari malformation ( $X^2 = 9.3$ ,  $V = 0.26$ ,  $p = 0.02$ ) and VP shunt revision ( $X^2 = 9.4$ ,  $V = 0.26$ ,  $p = 0.002$ ). The incidence of infection in the neonatal period was associated with the transfer between hospitals before surgery ( $X^2 = 7.1$ ,  $V = 0.23$ ,  $p = 0.029$ ) and death ( $X^2 = 8.7$ ,  $V = 0.25$ ,  $p = 0.013$ ).

**CONCLUSIONS:** The main predictor of neurological function is the level of MMC. Thoracic level was associated with more shunt revision and symptomatic Chiari malformation. Transfer between hospitals after birth and before surgery was associated with an increased risk of infection and this is the only preventable cause of infection detected by this retrospective analysis.

**Keywords:** myelomeningocele, prognosis, predictors, long term follow up,

**FL-062**

## Epilepsy and functional

### Comparative evaluation of somatosensory evoked potentials in children with occult spinal dysraphism and myelomeningocele in follow-up for tethered spinal cord

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<sup>3</sup>Department of Neurology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul, Turkey

**OBJECTIVE:** In this study, we investigate tibial somatosensory evoked potentials of children under follow-up in Spina Bifida Center. Patients were divided

into two groups; group 1: children with occult spinal dysraphism and group 2: children who underwent myelomeningocele repair after birth.

**MATERIAL-METHODS:** Seventeen children in group 1 and 44 children in group 2 were evaluated with tibial SEP recordings. Tibial The mean age of children in group 1 was 8.4 year old (10 female-7 male). The mean age of children in group 2 was 7 year old (18 female-26 male).

**RESULTS:** In group 1, we could obtain cortical P37 response bilateral in 15 patients (88%), and unilateral in 2 patients. Lumber responses were elicited in 8 patients (2 unilateral, 6 bilateral). In group 2, we could obtain cortical P37 response bilateral in 22 patients (50%) and unilateral in 5 patients. Lumber responses were elicited in 13 patients (8 bilateral, 5 unilateral). The mean latency of P37, N45 and lumber response in group 1 were; 36.2 (sd 8), 52.3 (sd 12.3), 17.2 (sd 3.3), in group 2; 38.6 (sd 8.2), 51.5 (sd 10.7), 17.8 (sd 4.6) respectively. The mean amplitude of P37-N45 was 7.8 (sd 7.1) in group 1, 8.4 (sd 13.7) in group 2. SEP were repeated in 10 patients (3 in group 1, 7 in group 2).

**CONCLUSIONS:** The response rate of cortical SEP was better in occult spinal dysraphism than MMC patients. The mean latencies and amplitudes has no difference between groups and are compatible with literature about SEP responses in children. However the amplitude of P37-N45 showed high variability which supports the latencies might be more reliable. The lumber responses might be more valuable to follow-up for tethered cord since we found changes in lumber responses without any change in cortical responses.

**Keywords:** Myelomeningocele, SEP, Occult spinal dysraphism, Tethered Cord

**FL-063**

## Spine

### Towards a pathophysiological understanding of lipomyelomeningocele: clues from histological and molecular analysis of an institutional case series of symptomatic lipomyelomeningocele

Gesa Cohrs<sup>1</sup>, Bea Kowitzke<sup>1</sup>, Ivo Leuschner<sup>2</sup>, Michael Synowitz<sup>1</sup>, Janka Held Feindt<sup>1</sup>, Friederike Knerlich Lukoschus<sup>1</sup>

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**OBJECTIVE:** Molecular and cellular mechanisms underlying symptomatic lipomyelomeningoceles (LMMC) are not well understood. A better understanding of these cascades probably enables modern management strategies for these conditions. We aimed for identifying specific mediators of inflammatory and proapoptotic molecular cascades potentially underlying symptomatic LMMC.

**MATERIAL-METHODS:** Ethical approval was obtained. Specimens from nine primary LMMC-surgeries (operated for "symptomatic" LMMC) harboring neuroepithelial tissue were investigated. Clinical data were collected. Pre-surgical MRI was reevaluated for lipoma type (classification by Pang 2013), conus position, and syrinx. Controls included normal adult spinal cords (n=4). Sections were analyzed histologically and by staining with neuroglial, neural crest, mesenchymal and epithelial markers. Immunohistochemistry and real-time RT-PCR for Interleukin-1beta (IL-1b), its receptor IL-1R1, tumor necrosis factor-alpha (TNF-a) and its receptor TNF-R1 were performed and analyzed qualitatively and semi-quantitatively. Cellular cytokine expression was confirmed via double-fluorescence-labeling with cellular markers. Hints for apoptosis were followed by Caspase-3-immuno-labeling. Finally, hypoxia induced factors (HIF-1a/2a) and cytokines Erythropoietin Receptor and its ligand (EpoR/Epo) were analyzed.

**RESULTS:** Four transitional intraspinal lipomas, 4 transitional lipoma with extraspinal extension, and one chaotic lipoma were included into the study. In all 9 cases, GFAP and Vimentin were detectable on significant induction level in intrathecal neural elements. IL-1R1, IL-1b, TNF-R1, TNF-a were significantly elevated in all cases (significance per case ranging between  $p < 0.05$  and  $p < 0.001$  vs. control). The investigated cytokines were co-stained with neural and glial cell

markers. Further, caspase-3 was found co-stained with the investigated molecules. Hypoxia-induced factors and consequently EpoR/Epo were found on elevated levels in the majority of investigated cases.

**CONCLUSIONS:** Our study identified pro-inflammatory and pro-apoptotic cytokines as potential mediators of pathophysiological cascades of LMMC. Their role as surrogate markers for symptomatic LMMC has to be proven in further studies.

**Keywords:** Lipomyelomeningocele, inflammatory cytokines, hypoxia inducible factors, immunohistochemistry, real-time RT-PCR

**FL-064**

### Spine

#### Recognition of limited dorsal myeloschisis and proper management yield excellent surgical outcomes: early experience

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**OBJECTIVE:** To emphasize the importance of recognising the entity "limited dorsal myeloschisis (LDM)" and rendering proper treatment, in preserving neurological function and preventing future deterioration from cord tethering. **MATERIAL-METHODS:** We present three patients with midline skin lesions who were diagnosed as having limited dorsal myeloschisis during the first Author's early experience as pediatric neurosurgeon in the Faculty of Medicine Siriraj Hospital, Mahidol University, Thailand. All three patients had no neurological deficit.

**RESULTS:** The first patient is a 2-year-old girl with a cervical saccular type lesion with a dimple at the base of the sac, and a history of leakage through the dimple during the newborn period. The second patient is an 8-month-old boy with a flat-type skin lesion and a history of possible fluid leakage once during the newborn period. The third patient is a newborn with a lumbar saccular type lesion. None of three patients had neurological deficit. Magnetic resonance images confirmed the diagnosis of LDM with stalks attached to the spinal cord. The first two patients had elective operations soon after diagnosis at the age of 2 years and 9 months, respectively, with resection of the LDM stalk attachments to the spinal cord and untethering of the spinal cord. Postoperatively both patients remain neurologically normal and recovered without complication. Pathology confirmed fibroneural stalks compatible with LDM. For the third patient, surgery is planned when the child reaches 5 months of age to avoid anesthetic complication.

**CONCLUSIONS:** The correct diagnosis of LDM and proper surgery in the early age yield excellent outcome. In developing countries that do not have formally trained pediatric neurosurgeon, the existence of this entity must be communicated to pediatricians and adult neurosurgeons to avoid missed or delayed diagnosis and inadequate surgery which will lead to future neurological deterioration.

**Keywords:** limited dorsal myeloschisis, flat-type skin lesion, saccular type lesion, fibroneural stalk

**FL-065**

### Spine

#### Limited dorsal myeloschisis: association with dermoid elements and its surgical implications

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**OBJECTIVE:** A histopathological hallmark of all Limited Dorsal Myeloschisis (LDM) is glioneuronal tissue in the stalk, as it originates from undisjointed neuroectoderm during primary neurulation. The majority of LDMs have only the neuroglial tissue within the stalk but a small percentage will have elements of a dermoid cyst or a dermal sinus tract either separate from the LDM stalk or incorporated within its fibroglial matrix, explainable by the original continuum of cutaneous and neural ectoderms in LDM's embryogenesis. The dermoid elements can be microscopic and escape casual observation, but could grow to large intradural or intramedullary dermoid cysts when left behind.

**MATERIAL-METHODS:** We present a pediatric series of 5 cases with LDMs associated with dermoids. We analyzed age and sex distribution, presenting neurological symptoms, cutaneous stigmata, level of spinal cord attachment of the fibroneural stalk, whether there was initial complete removal of the LDM stalk, levels of laminectomy, histopathology, recurrence of dermoid elements, postoperative complications, and neurological outcome.

**RESULTS:** 2 of 5 patients underwent revision surgery for recurrence of dermoid tumours because of incomplete resection of the entire fibroneural stalk at initial surgery, due to the surgeon's intention to minimize bone removal by using limited segmental laminectomies. Those cases with original complete removal of the fibroneural stalk and the 2 patients with secondary complete resection had excellent outcome.

**CONCLUSIONS:** The existence of dermoid cysts within the fibroglial matrix of a LDM stalk reflects the embryogenesis involving a continuum of cutaneous and neural ectoderms. We present our series of LDMs with associated dermoids and recommend excising the entire length of the intradural LDM stalk during the initial treatment to prevent recurrence of the dermoid remnants.

**Keywords:** dermal sinus tract, dermoid cyst, dysraphic malformation, limited dorsal myeloschisis, primary neurulation

**FL-066**

### Spine

#### Limited dorsal myeloschisis: our experience at Hopital Femme Mere Enfant of Lyon

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**OBJECTIVE:**Limited Dorsal Myeloschisis (LDM) is a recently identified form of spinal dysraphism.

Since 2012, we prospectively collected pre and post operative clinical and radiological findings from all children operated on for a LDM.

**MATERIAL-METHODS:**Between 2012 and 2015, 8 patients (4 females, 4 males) were operated on. The LDM was cervical in 2, thoracic in 1 and lumbar in 5 cases. Cutaneous signs were present in all subjects (saccular form in 3 cases, flat form in 5). The malformation was diagnosed prenatally in the 3 saccular cases. The flat form cases were identified at birth (2 cases), in the first year of life (2 cases) and at the age of 12 years in the remaining child. This last case was the only symptomatic one as the LDM was recognized following a MRI study carried out because of urinary disturbances and pain in the lower limbs. None of the 8 patients presented other associated CNS anomalies.

**RESULTS:**All subjects were managed surgically with spine detethering and removal of the fibrous tract. The mean age at operation was 20 months (range 0 to 12 years). There was no post-operative neurological deficit. The child who presented with abnormal urinary function and pain improved after the surgical procedure. We recorded only one surgical complication, that is infection of the surgical wound that required a second operation.

**CONCLUSIONS:**LDM is a rare spinal dysraphism which bears a good prognosis following the surgical correction. While the saccular form may be recognized in utero the antenatal diagnosis in flat forms is difficult. Consequently, the postnatal recognition requires particular attention for all the cutaneous stigmata that can herald the condition. The surgical repair should be carried out as early as possible due to the potential risk of the malformation to become symptomatic because of secondary tethering of the cord.

**Keywords:** Spinal dysraphism, tethered cord, spine malformation

## FL-067

### Spine

#### Investigation in our cases of congenital dermal sinus with the combination of morphological and pathological features

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**OBJECTIVE:**The classification of congenital dermal sinus (CDS) belonging to occult spinal dysraphism is still complicated with many arguments in literature. We investigate the pathological and morphological features of our CDS cases to propose our consideration of the classification.

**MATERIAL-METHODS:**The materials are 27 cases with CDS surgically treated in our hospital since 2006. We removed the whole length of CDS in all cases to confirm the morphological characters and to examine the pathological features.

**RESULTS:**As morphological characters, the cutaneous findings of lumbosacral region were; dimple in 8 cases, skin defect in 12, and human tail / tag in 7. Terminal end of the CDS penetrated into intradural space in 5 cases, continued to end of filum terminale at the bottom of dural sac in 7, terminated at dural surface in 4, reached to intradural lipoma in 4, and extinguished in extradural lipoma in 7. The pathological findings of CDS were; the lumen lined by a stratified epithelium in 3 cases, the sinus formed by fibrous or fibrolipomatous bundles occasionally with scattered clusters of meningeal (arachnoid-like) cells in 20, and dermal / epidermal inclusion cysts found subcutaneously in fat tissue combined with the fibrous sinus in 4. The two cases penetrating into intradural space proved to correspond to true CDS, while the other 3 cases were

equal to meningocele manqué in literature. However, we pathologically found one case of true CDS and another of meningocele manqué in 4 cases with the terminal end at dural surface. Seven cases with the continuity to filum terminale presented unique pathological features with scattered clusters of meningeal-like cells and epidermoid inclusion cysts. **CONCLUSIONS:**The discrepancy between morphological and pathological findings results that we should discuss about conventional classification with the etiological consideration.

**Keywords:** congenital dermal sinus, morphological and pathological features, classification in literature

**OBJECTIVE:**Filum terminale lipoma (FTL) is known to cause various spinal symptoms collectively referred to as tethered cord syndrome. With the increasing use of magnetic resonance imaging (MRI), there has been an increase in the identification of FTLs. The treatment for FTL is surgical untethering by sectioning the FTL, which can prevent symptom progression and often results in improvement of symptoms. We recently introduced a new surgical strategy for FTL untethering. In this report, we present a minimally invasive technique for FTL sectioning.

**MATERIAL-METHODS:**Forty-six consecutive patients were treated using this minimally invasive technique we refer to as an interlaminar approach (ILA). The detailed surgical approach and postoperative result are outlined. A midline skin incision was performed at the L5 vertebral level. After skin incision, dissection was extended to expose the ligamentum flavum. The ligamentum flavum was then incised and retracted medially to expose the dural sac. After dural incision, the FTL was identified and pulled out from the dural sac and then sectioned.

**RESULTS:**All patients underwent an untethering operation using ILA. Surgical complication was seen in only one patient who developed a subcutaneous fluid collection that resolved spontaneously. All patients remained stable or showed improvement in preoperative symptoms.

**CONCLUSIONS:**Tethered spinal cords can be safely and effectively untethered using an ILA. This technique provides the advantage of a small skin incision, reduced soft tissue injury, less postoperative pain, minimal blood loss, and minimal intradural procedures, which may also provide the added advantage of a reduced risk of retethering.

**Keywords:** filum lipoma, interlaminar approach, minimal invasive, surgical treatment

## FL-069

### Spine

#### The spectrum of caudal agenesis and associated malformations

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**OBJECTIVE:**To present a series of patients with caudal agenesis (CA), with emphasis on its clinical diversity and associated malformations.

**MATERIAL-METHODS:**44 patients with age at diagnosis ranging from 2 days to 18 years. Myelomeningocele patients and other with irregular

follow-up were excluded. The Reinshaw's classification for CA was used. Patients were sorted according to the conus position above (Group I) or below (Group II) L1 body.

**RESULTS:**Six mothers had diabetes mellitus. Agenesis of lumbar vertebrae was found in 8 patients. None had isolated coccygeal agenesis. All patients had neurogenic bladder. Among 68 skeletal malformations, 28 involved the girdle and 22 the feet. There were 28 anorectal anomalies, 11 kidney malformations and 5 VACTERL association. In Group I, 10 patients had the conus ending abruptly above or at L1 level and all had variable hypoplasia of the lower hemibody with gluteal hypotrophy and narrow hips. Agenesis of one or more lumbar vertebrae was seen in 6 and these patients were the most severely affected. In this group only 3/10 were operated. Group II had 34 patients. Anorectal and / or urogenital malformations were found in 19. The OEIS complex was seen in 12. Terminal myelocystocele (n=11), midline cutaneous stigmata (n=9) and subcutaneous lipomas (n=7) were also found. In this group the main neuro-orthopedic features were related to hip dysplasia and talipes deformity. All the patients were managed by a multidisciplinary team and several interventions were needed. Surgery for conus detethering, mostly due to fibrolipomas, and also for stenosis relieve were performed in 33/34 patients. From 36 operated patients, 23 were improved, 11 unchanged and 2 worsened. There were 8 postoperative complications in 5 patients.

**CONCLUSIONS:**Most of CA are associated with hindgut and urogenital malformations. However some may present as isolated malformations with tethered cord syndrome indistinguishable from typical closed spinal dysraphisms.

**Keywords:** Caudal agenesis, tethered cord syndrome, VACTERL association, OEIS complex, anorectal malformations

#### FL-070

##### Spine

#### The spectrum of childhood angular kyphosis: a multidisciplinary approach

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**OBJECTIVE:**Angular kyphotic deformities are relatively uncommon, most of them resulting from vertebral formation or segmentation failure. Approximately 10% develop severe kyphotic deformity and secondary compressive myelopathy. We illustrate the spectrum of childhood angular kyphosis from prenatal diagnosis to severe progressive deformity, and stress the importance of a multidisciplinary team approach.

**MATERIAL-METHODS:**We present 10 children with angular kyphosis, including one stillborn infant with severe angular kyphosis and other developmental anomalies. A multidisciplinary team evaluated all children, including extensive radiological work-up with magnetic resonance imaging and 3D computed tomography. Six children had decompressive surgery and instrumentation, while two more are scheduled at short term.

**RESULTS:**Etiologic diagnosis included failure of segmentation, hemivertebra, Ehlers-Danlos syndrome, chondrodysplasia punctata, and myelomeningocele, with kyphotic angles ranging from T8 to L3. Six children had associated anomalies, including hydromyelia, spinal lipoma, lipomatous filum, myelomeningocele, and talipes. Eight children manifested neurological and/or urological symptoms, including radicular pain, Babinski sign, progressive myelopathy with or without paraparesis, and neurogenic bladder. All children

presenting with neurological symptoms improved after surgery, while none of the children deteriorated during follow-up. Kyphosis was corrected, stabilized, or its progression slowed depending on individual etiology. Mean postoperative follow-up at time of writing was 30.7 months (10-78 months).

**CONCLUSIONS:**Surgical treatment of angular kyphosis is very complex and every case has its unique presentation. While spinal cord and/or cauda equina are always at risk even in those children not presenting with a progressive neurological deficit, relevant literature is largely limited to relatively small, exclusively orthopedic series. We strongly believe pediatric neurosurgeons should be more involved in a multidisciplinary team approach, including a thorough preoperative work-up, well-timed and carefully planned surgical intervention. Highly individualized treatment should only be offered in dedicated pediatric spine units with enough experience in anterior and posterior approaches and instrumentation.

**Keywords:** angular kyphosis, compressive myelopathy, decompressive surgery, multidisciplinary approach, segmentation failure

#### FL-071

##### Spine

#### Proatlas segmentation anomalies

Natarajan Muthukumar

Madurai Medical College, Madurai, India

**OBJECTIVE:**Proatlas segmentation anomalies are due to defective resegmentation of the proatlas sclerotome. These anomalies of the craniovertebral junction are rare and have multiple presentations. The aim of this study is to report this author's personal experience in managing five of these patients with different radiological findings necessitating different surgical strategies

**MATERIAL-METHODS:**Five patients, all in the second decade of life were treated between 2010 and 2013. There were three males and two females. All the patients presented with spastic quadriplegia and/or cerebellar signs. Patients underwent Plain radiographs, MRI and CT of the craniovertebral junction. CT of the craniovertebral junction was the key to the diagnosis of this anomaly. Postoperatively, patients were assessed with plain radiographs and CT in all patients and MRI in one.

**RESULTS:** Patient 1 had basilar invagination with an accessory ossicle posterior to the clivus with partially assimilated atlas. Patient 2 has assimilated atlas, basilar invagination, unilateral occipital condyle hypoplasia and Klippel-Feil anomaly. Patient 3 has Os avis with atlantoaxial dislocation. Patient 4 had abnormally elongated "comma shaped" clivus with atlantoaxial dislocation. Patient 5 had platybasia, horizontally oriented clivus with an "exuberant" hypertrophic apical dens located adjacent to clivus which was kinking the cervicomedullary junction along with Chiari 1 malformation. Two patients underwent craniovertebral realignment with occipitocervical fixation, two patients underwent C1-C2 fixation using Goel-Harms technique and one patient underwent craniovertebral realignment with C1-C2 fixation using spacers in the atlantoaxial joint and foramen magnum decompression. All patients improved during follow up.

**CONCLUSIONS:** Proatlas segmentation defects are rare anomalies of the craniovertebral junction. Routine use of thin section CT of the craniovertebral junction and an awareness of this entity and its multivarious presentations are necessary for clinicians dealing with abnormalities of the craniovertebral junction.

**Keywords:** Assimilation of atlas, atlanto-axial subluxation, basilar invagination, craniovertebral junction, occipito-cervical fusion, proatlas segmentation anomaly

**FL-072****Other****Posterior fossa decompression in pediatric Chiari 1 malformation: choosing the best technique**

Anton E. Korshunov, Yury V. Kushel

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**OBJECTIVE:**We aimed to investigate outcomes after pediatric Chiari-1 posterior fossa decompression surgery in connection to the surgical technique used. **MATERIAL-METHODS:**76 children (<18 y.o.) with Chiari-1 malformation had posterior fossa decompression in our department from 2001 till 2015. 52 (68%) had syringomyelia before surgery. Extradural decompression (EDD) was performed in 14(18%) patients; extraarachnoid duroplasty (EAD) in 21(28%); intraarachnoid dissection and duroplasty in 21(28%); foramen Majendie stenting and duroplasty in 20(26%). Surgical technique was selected by the surgeon's preference without protocol.

**RESULTS:**Choice of a surgical technique correlated with (1) presence of syringomyelia and (2) preoperative Chiari Severity Index grade.

Complications occurred in 15 patients (20%), one of them was lethal (mortality 1.3%). Complication rate was lowest after EDD (0%) and highest after stenting (40%). Complication rate was significantly increased after (1) intraarachnoid dissection ( $p=0.009$ ) and (2) stent insertion ( $p=0.02$ ), no other preoperative or intraoperative variable correlated with complications.

Median follow up was 17 months. Reoperations were necessary in 8(11%) patients. Reoperation rate was highest after EDD (21%) and lowest after EAD (5%), difference was not statistically significant.

Combined complication-and/or-reoperation rate was lowest after EAD (10%), and highest after stent placement (40%); difference was not statistically significant. No preoperative or intraoperative variable correlated with reoperation rate or combined complication-and/or-reoperation rate.

**CONCLUSIONS:**EAD is the most attractive option at primary surgery for pediatric Chiari-1 malformation with or without syringomyelia. EDD is the safest and least invasive primary option and is acceptable with proper patient selection. Intraarachnoid dissection with or without stent insertion is hardly justified at initial surgery, but may be inevitable at reoperation.

**Keywords:** Chiari-1 malformation, Syringomyelia, Posterior fossa decompression, Duroplasty, Stent

**FL-073****Other****Suboccipital decompression with duraplasty for Chiari malformation: experience with 106 consecutive pediatric patients**Eveline Teresa Hidalgo, Yosef Dastagirzada, Svetlana Kvint, Cordelia Orillac, Emily North, Christopher Hernandez, Ramona Bleda, Jeffrey H. Wisoff  
Division of Pediatric Neurosurgery, Department of Neurosurgery, NYU Langone Medical Center, New York, USA

**OBJECTIVE:**The need of duraplasty for adequate decompression of Chiari 1 malformation remains highly controversial. Although proponents of dural opening contend that duraplasty increases the likelihood of symptom and syrinx improvement while concurrently decreasing reoperation rates, opponents reference the risks associated with dural opening: CSF leakage, bacterial meningitis, aseptic meningitis, increased bleeding, pseudomeningocele, and hydrocephalus. In this study we retrospectively investigated the outcomes for pediatric patients who underwent surgery for treatment of symptomatic

Chiari Malformation. We limited our investigation to patients who were treated with bony decompression and duraplasty in an effort to describe the clinical outcomes and post-operative course associated with this posterior fossa decompression variant.

**MATERIAL-METHODS:**This is a retrospective chart review of all patients who underwent surgical decompression of symptomatic Chiari Malformation with dural opening by a single surgeon at New York University Medical Center between the years of 1985 and 2015.

**RESULTS:**107 patients, median age at surgery 10 years (range: 1-20).

Presentation was typical tussive headaches in 15.9%, mixed headache in 11.2%, atypical headache in 18.7%, serendipitous finding in 22.4%, scoliosis workup in 22.4%, neurologic deficit in 3.7%, ataxia in 2.8%, other in 3.7%.

Focal neurologic deficits at presentation was sensory in 19.6%, motor in 6.5%, mixed in 10.3%, CN involvement in 2.8%, none in 57.9%

Headache Resolved in 46/48 patients(95.8%).

Syrinx resolved in 63.8%, decreased in 28.8%, stable in 5%, progressive in 1.3%.

Complications were aseptic meningitis in 15%, Infection in 0.9%, dural leaks in 0.9%

No resurgery in 30 days, total resurgeries: 11 (10.3%). No mortality.

**CONCLUSIONS:**Suboccipital decompression with duraplasty for Chiari 1 malformation can be achieved with a high success rate and a low complication rate in selected patients. Headache resolved in 96% and Syrinx resolved or improved in 93% of the patients. The most common complication was aseptic meningitis.

**Keywords:** Chiari Malformation, Suboccipital Decompression, Duraplasty, Aseptic Meningitis, Syrinx

**FL-074****Other****Re-evaluation of the indications for foramen magnum decompression in achondroplasia based on the consensus guidelines**

Mihoko Kato, Daimon Shiraishi, Hirokatsu Osawa

Department of Neurosurgery, Aichi Children's Health and Medical Center

**OBJECTIVE:**Patients with achondroplasia often show sleep apnea syndrome (SAS). It is reported that SAS could cause death in infancy. Some relationships have been suspected between SAS and foramen magnum stenosis which is often observed in the patients. Therefore, foramen magnum decompression is very important surgical procedure to improve the outcome of achondroplasia. Recently, a consensus guideline was established by K.K.White et al. In this study, we retrospectively analyzed our cases and re-estimate operative indication using the guideline.

**MATERIAL-METHODS:**We have treated 25 patients since 2010. Patients without preoperative MRI and FMD in other hospital excluded, 18 were eligible for the analysis. We focused on preoperative clinical and MRI findings especially for signal change (SC) and indentation (ID) of the spinal cord, CSF signal loss (CSL). Evaluation of the outcomes was done after FMD or conservative therapy. Development was estimated using a Milestone of the disease.

**RESULTS:**There were 12 boys and 6 girls. Mean age at initial diagnosis was 15 months. FMD was performed in 5. Three patients had SAS, and 4 had delayed development. Four showed SC. Five showed ID and CSL. Thirteen were conservatively treated. Three had SAS and one had delayed development. MRI showed ID in 3 and CSL in 6. Re-estimation using a new guideline revealed that 2 conservatively treated patients had operative indication. All patients receiving FMD showed improvement of SAS and got ability to walk until 3 years old except infant. Ninety percent of conservatively treated patients also got ability to walk until 2.5 years old. Two conservatively treated patients with operative indication also showed improvement of SAS and got ability to walk.

**CONCLUSIONS:**Compering with our operative indication, that of the new consensus guideline was more aggressive. However, all conservatively treated

patients with operative indication of the guideline showed the same improvement as other conservative patients.

**Keywords:** Achondroplasia, Foramen Magnum Decompression, Consensus Guidelines

**FL-075**

**Special topic: Neuro-imaging**

**Hypertrophy of the soft tissue at the retro-odontoid space in patients with Chiari type I malformation**

Yasuhiko Hayashi, Masahiro Oishi, Disuke Kita, Mitsutoshi Nakada  
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**OBJECTIVE:** Chiari malformation type I (CM-I) is a disorder in which the cerebellar tonsils protrude through the foramen magnum and into the spinal canal. The soft tissues, including the transverse ligament and the tectorial membrane at the retro-odontoid space, have a potential to compress the cervicomedullary junction, if they become hypertrophic. The purpose of this study is to determine whether the retro-odontoid soft tissue in CM-I may work as one of the ventral compressive factors to the cervicomedullary junction.

**MATERIAL-METHODS:** Twenty symptomatic CM-I patients (9 men and 11 women) from age 5 to 20 years were treated between 2004 and 2015 at Kanazawa University, Japan. Symptoms were as follows: headache in 8 patients, scoliosis in 7, sensory disturbance in 3, monoparesis and nystagmus each in 1. Soft tissue at the retro-odontoid space was evaluated using T2-weighted magnetic resonance (MR) images. Anterior-posterior (AP) distances and superior-inferior (SI) distances of the soft tissue were measured in CM-I patients and 48 normal control children. Modified clivo-axial angles (CAA) were also evaluated using the index of ventral compression of the cervicomedullary junction.

**RESULTS:** Of those CM-I patients, 15 were treated with ordinary foramen magnum decompression. Thirteen patients improved postoperatively, but 2 were unchanged. AP distances in the CM-I group (6.0 mm) were larger than in the control group (3.5 mm), whereas SI distances did not differ significantly (18.2 mm vs. 16.6 mm). Modified CAAs were significantly smaller in the CM-I group (131.5°) than in the control group (146.9°).

**CONCLUSIONS:** MR images of the retro-odontoid soft tissue in patients with CM-I showed significant hypertrophy and ventral compression of the cervicomedullary junction. CM-I has the potential to be symptomatic because of ventral compression not only in the osseous anomalies of the craniovertebral junction but also in the soft tissue at the retro-odontoid space.

**Keywords:** Chiari malformation type I, soft tissue, odontoid process, transverse ligament, craniovertebral junction  
Wednesday, 26 October 2016

16:15 – 17:15

## Flash sessions 6: Neuro-oncology

**FL-077**

**Neuro-oncology**

**Surgery of infantile brain tumors**

Tatsuya Nagashima<sup>1</sup>, Atsufumi Kawamura<sup>1</sup>, Junji Koyama<sup>1</sup>, Noriyuki Akutsu<sup>1</sup>, Hiroaki Nagashima<sup>2</sup>

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**OBJECTIVE:** Surgery plays a crucial role in the first year of life, because radiotherapy is not indicated and chemotherapy is limited. Treatment of the newborn and infantile brain tumors still remains the greatest challenge in pediatric neurosurgery. The present report aims at providing an understanding of the subject by analyzing the results of treatment in a personal series over a 12-year-period.

**MATERIAL-METHODS:** Twenty-five patients under the age of 1 year were treated for intracranial tumors during the last 12 years. Patients present with a variety of tumor pathologies; medulloblastoma 3, ATRT 4, PNET 1, Choroid plexus papilloma (CPP) 3, neuroblastoma 3, immature teratoma, 4 and others. **RESULTS:** Five-year overall survival rate was 75%. Five infantile immature teratomas were operated on; 4 were totally removed. After total removal, all 4 patients survive without chemo-radiotherapy, however, three show mild to severe developmental retardation. All of benign tumors such as craniopharyngiomas, CPPs were totally removed and the patients survive in good condition. All of 3 patients with medulloblastoma survive without significant neurological deficit; two of them were treated without radiotherapy and survive longer than 8 years, one patient who was treated by chemotherapy and reduced dose craniospinal radiation survives longer than 4 years without recurrence. Prognosis of medulloblastoma of infant have been considered to be poor, however, some of them can be cured without radiotherapy.

**CONCLUSIONS:** Treatment strategy for benign brain tumors of infant is total removal, though technically demanding. Treatment of newborn immature teratomas should be individualized because neoadjuvant chemotherapy may be effective. Malignant brain tumors in this age group are histologically heterogeneous. Risk stratification is necessary to treat medulloblastomas of very young children.

**Keywords:** Brain tumor, Infant, Newborn

**FL-078**

**Neuro-oncology**

**Management of pediatric brain tumors: strategies and long term outcome**

Abubakr Darrag Salim, Mohammed Awad Elzain Ahmed  
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**OBJECTIVE:** This study is aimed at shedding the lights on different patterns of presentation of Sudanese children with brain tumors and reflecting the experience of the national center for neurological sciences in setting strategies for management together with long-term follow up over 14 years period.

**MATERIAL-METHODS:** Retrospective, observational study for all Sudanese children with brain tumors operated in the National Center for Neurological Sciences in the period between September 2000 to March 2015. Data were collected and patients were followed throughout the entire 14.5 years study period. All patients with deficient clinical pre and post-operative data, patients with missed operative details, patients with missed histopathology reports and adult patients were all excluded from the study.

**RESULTS:** During this 174 months period, 54 patients were operated aging between 1-17 years with average presentation at 9 years of age. M:F 2:1. The commonest presenting symptoms are headache (90.7%), back pain (81.3%), vomiting (59.3%) and unsteady gait (48.1%). The average duration of symptoms was 1 year. Most patients were operated through posterior fossa craniectomy (n=30/54, 55.9%) and histopathology reports were mainly medulloblastoma (n=15/54, 27.8%) and pilocytic astrocytoma (n=11/54, 20.4%). Most patients improved or cured post-operatively (n=43/54, 79.7%) 1 deteriorated and 9 died.

**CONCLUSIONS:** Pediatric brain tumors are among the most challenging neurosurgical problems that needs stepwise multidisciplinary team. The lesions tend

to be infratentorial with obstructive hydrocephalus. In our center it is found that 2 steps surgery first with VP shunt followed by second stage tumor resection after few weeks is both effective and safe way with apparently good outcome.

**Keywords:** pediatric brain tumors, management, long term outcome

#### FL-079

##### Other

#### Endoscopic treatment for third ventricular and paraventricular tumors except for germ cell tumors: analysis of the nationwide investigation

Kenichi Nishiyama

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**OBJECTIVE:**Endoscopic biopsy and ETV for posterior third ventricular tumors with marker-negative is favored as a first step in its management. The author analyze the safety and validity of endoscopic treatment for third ventricular and paraventricular tumors except for germ cell tumors from the data of a nationwide investigation in Japan.

**MATERIAL-METHODS:**Retrospective data were collected from 123 centers in Japan performing endoscopic biopsies between 2005 and 2009. Among total 714 registered patients, 162 patients (91 male and 71 female, median age 35.5) were enrolled as a cohort. Data regarding pathology, mismatch between biopsy results and final diagnosis, perioperative complications, dissemination following the procedure, control of hydrocephalus and improvement of ADL were mainly analyzed.

**RESULTS:**Biopsies were informative in 139 patients (85.8%), while it was 92.8% in total 714 registered patients. Biopsy results were differed from final diagnosis based on subsequent craniotomy in 7 (3 pineal tumors, 2 gliomas and 1 choroid plexus tumor). Universal complications were seen in 10 (5 bleedings, 3 new onset of hydrocephalus and 2 meningitis with CSF leakage). The incidence of postoperative fever without infection or hemorrhage was 18.5%. It was relatively high compared with that in total registered cases (13.6%). ETV was required in 95 (68.8%), and long-term controlled hydrocephalus in 89 (success rate: 93.7%). The incidence of newly-formed dissemination after biopsy was 5.6%. The Karnofsky Performance Scale was increased after the procedures in 84 (52%). Among them, 75% of the patients were successfully treated with ETV for coexisted hydrocephalus.

**CONCLUSIONS:**Endoscopic biopsy should be considered as a treatment of choice for third ventricular and paraventricular tumors except for germ cell tumors. However, relatively low diagnostic accuracy and rather high incidence of post-operative fever are points to be improved. ETV plays a key role in making a daily performance better by means of improving hydrocephalus.

**Keywords:** Neuroendoscope, biopsy, third ventriculostomy, hydrocephalus, ventricular tumor

#### FL-081

##### Neuro-oncology

#### Surgical treatment of pineal region tumor in infants under three years

Jie Ma, Yang Zhao, Lianping Sun, Feng Jiang, Qifeng Li

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**OBJECTIVE:**Pineal region tumors in infants and toddlers (<3 years old ) are challenge for surgery, demonstrating dismal clinical outcomes. To explore a reasonable treatment plan, this paper reviewed the operative treatment of pineal region tumors under 3 years old in my hospital.

**MATERIAL-METHODS:**A retrospective study was done on 13 patients with pineal region tumors under 3 years-old.The clinical characteristic, surgical approach and prognosis were reviewed. All of the patients were operated on via infratentorial supracerebellar approach(n=8), trans-frontal-callosal-interforneal approach(n=2), paramedian infratentoria approach(n=1),combined approach(n=2)

**RESULTS:**Tumor was totally removed in 8 patients, subtotally resected in 3 and partially resected in 2. After surgery no patients fell in coma or died. Two patients died with abandoning treatment due to infection. Followed-up time was 3-30 months, 9 cases had resumed normal life, 2 cases died with recurrence.

**CONCLUSIONS:**Microsurgery is the prior option for children with pineal region tumor under 3 years, and favorable resection rate has been achieved in this series. Proper approach depended on every individual entitle successful resection.

**Keywords:** infant, pineal region tumor, microsurgery

#### FL-082

##### Neuro-oncolog

#### Cerebellopontine angle tumors in children

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**OBJECTIVE:**Cerebellopontine angle (CPA) and cerebellomedullary fissure (CMF) tumors are rare in children. This retrospective study reports their histological distribution, surgical resections and post-operative outcome based upon the authors' consecutive personal series

**MATERIAL-METHODS:**Clinical data of children 16 years old or younger of age treated from 2014 to 2016 by a single surgeon was retrospectively reviewed. All had histologically verified CPA/CMF tumors and underwent radical tumor resection through craniotomy. Tumors' pathological distributions, surgical approaches, and patients' outcome were reviewed.

**RESULTS:**There were 34 children with the age at diagnosis ranging from 1.1 to 14 years. Pathology showed 21 ependymomas, 5 benign gliomas (3 pilocytic astrocytomas, 2 ganglioglioma), 2 epidermoids, 1 medulloblastoma, 1 meningiomas, 1 inflammatory granuloma, 2 nerve sheath tumors, and 1 choroid plexus papilloma. For 36 tumor resections(two were relapsed 1.5 years after the first resection), all were approached through a posterior fossa craniotomy or far lateral approach. At tumor resection, 34 had a gross total or near total resection, 2 subtotal resection. There were no mortalities. 9 patients had unilateral vocal cord palsy or dysphagia. Of these,6 were treated with nasogastric (NG) feeding tube, 2 with a combination of tracheotomy. All had successful removal of NG feeding and tracheostomy from 2 weeks to 2 months

**CONCLUSIONS:**Various of tumor types occur in childhood at the CPA/CMF and our review indicated 62 % were ependymomas. Lower cranial nerves should be protected and carefully dissected from the tumors during the operation and most patients can avoid tracheotomy.

**Keywords:** Brain tumor. Cerebellopontine angle tumor. Children. Ependymoma. surgery

#### FL-083

##### Other

#### Endonasal endoscopic approaches in 28 pediatric cases: Ankara University series

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**OBJECTIVE:**To discuss the endoscopic endonasal approach in pediatric age group with its advantages and disadvantages.

**MATERIAL-METHODS:**Retrospective analysis of 28 pediatric patients that are operated via endonasal endoscopic approach for various indications except angiofibroma in Ankara University Medical Faculty between years 2010 to 2016 March.

**RESULTS:**19 patients (67%) were male and 9 patients were female (32%). Seven of the cases were pituitary adenoma (25%), 7 craniopharyngioma (25%), 4 traumatic CSF rhinorrhea (14%), 2 germinoma (7%), 2 meningocele (7%), 2 pituitary inflammation (7%), 1 odontoidectomy (3%), 1 fibrous dysplasia (3%), 1 hemangiopericytoma (3%) and 1 capillary hemangioma (3%). Among 20 patients with pathological results; total or gross total excision was achieved in 17 (85%), subtotal resection was achieved in 3 (15%). In 2 cases of pituitary inflammation (12%) only biopsy was made. Either traumatic or due to tumor resection, all dura lesions along the skull base (12 cases) was reached with endonasal endoscopic approach and watertight sealing was achieved. No patient had a postoperative CSF leak or meningitis. 7 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. There were no exits.

**CONCLUSIONS:**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.

**Keywords:** Endoscope, Transnasal approach, Child, Advantage-disadvantage

#### FL-084

##### Neuro-oncology

#### Investigating a thickened pituitary stalk in children: validating a prospective treatment paradigm

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**OBJECTIVE:**A thickened pituitary stalk (TPS) is a nonspecific finding on magnetic resonance imaging (MRI) with a wide differential diagnosis, ranging from idiopathic to neoplastic processes. Treatment of TPS may be time sensitive for neoplasia. We reviewed our institutional experience and validated a treatment paradigm.

**MATERIAL-METHODS:**This is a single institution retrospective IRB approved review of the electronic medical record and radiology database by ICD-9 codes and keyword queries from 2000-2015. MRI's were reviewed by neuroradiology and neurosurgery. Patient, endocrine and radiographic data were collected for analysis.

**RESULTS:**We identified 97 patients with TPS. Fifty patients had isolated TPS in the absence of systemic Langerhan's cell histiocytosis (LCH) or obvious tumor. Median age at presentation = 12.4 years (Range: 2.9-18 yrs) with female predominance. Median follow-up = 3.6 years. Patients presenting with

diabetes insipidus (DI, n=12) were more likely to have a neoplastic process ( $P<0.001$ ). Six developed neoplasia, including germ cell tumor (n=2), LCH (n=3) and low-grade glioma (n=1). Four of these had concurrent anterior pituitary hormone dysfunction (APD) at presentation and all patients with DI and APD had neoplasms ( $p<0.001$ ). Of the 38 patients without DI, 2 developed LCH during the follow up period. Any TPS  $>5$ mm was neoplastic (neoplasia 5.8mm vs idiopathic 3.6mm,  $p<0.001$ ).

**CONCLUSIONS:**A pituitary stalk  $>5$ mm, DI, and DI with APD are indicators of a neoplastic process, requiring additional evaluation for GCT or LCH, close surveillance, and often a biopsy. We further developed and validated a diagnosis and treatment paradigm to guide decision-making.

**Keywords:** Langerhan's cell histiocytosis, Germ cell tumor, Diabetes Insipidus, Neurosurgery, Pituitary gland,

#### FL-085

##### Neuro-oncology

#### Isolated gliomas of the optic nerve in children: a multi-center historical cohort study

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**OBJECTIVE:**Isolated gliomas of the optic nerve (IONG) constitute a rare subgroup of optic pathway gliomas (OPG). Due to the rarity of this condition, and the mix-up with other types of OPG in most clinical series, little is known about these tumors. Currently, due to lack of evidence, they are managed as any other OPG. Here, we conducted a bi-center, historical cohort study aimed at deciphering the natural history of IONG.

**MATERIAL-METHODS:**Included were patients with clear-cut glioma of the optic nerve and no posterior (chiasmatic / hypothalamic) involvement, with more than 1 year of follow-up and at least 2 MR studies and neuro-ophthalmological exams.

**RESULTS:**Thirty-four patients were included in this study. Age at diagnosis ranged between 6 months and 16 years (average 6 years). Follow up time was 5.6 years. Twenty-two patients were NF1. Eight patients had bilateral disease, most in the NF1 group. Eight of the NF1 patients had radiological abnormality in their other optic nerve. During the follow-up period, 47% of the patients progressed, while 17% patients experienced some degree of spontaneous regression. Forty-one percent of the patients presented with visual decline, 64% experienced further deterioration. Eight patients were treated with chemotherapy, out of which 5 improved visually. Eight patients were operated upon.

**CONCLUSIONS:**In conclusion, IONG are dynamic tumors, with 66% in our series that were active. In addition, they may warrant closer observation, and more aggressive treatment, as 64% deteriorated visually during the follow-up period.

**Keywords:** OPG, Optic pathway gliomas, NF1, neurofibromatosis

#### FL-086

**Neuro-oncology****The role of surgery in pediatric optic pathway gliomas: 156 cases results from one institute**

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**OBJECTIVE:** Optic pathway gliomas (OPGs) account for 3–5% of all pediatric CNS tumors. The management is often challenging and there was not broad consensus regarding the optimal treatment strategy, the role of surgery and radiotherapy in OPG is controversial. The role of different treatment options were analyzed based the patients outcome with long term follow-up.

**MATERIAL-METHODS:** All the cases diagnosed with OPG and treated at Beijing TianTan Hospital between April 2003 and November 2015 were retrospectively reviewed.

**RESULTS:** One hundred and fifty-six patients were included in this study, with a mean age of 6.7 years (ranges from 10 months to 28 years). OPGs were divided into three subgroups according to Dodge classification (Type I n=11; Type II n=89; Type III n=56. Four cases had neurofibromatosis type 1. One hundred and forty-nine cases underwent surgical debulking, and 2 were stereotactic biopsied. Among 149 patients received surgical intervention, 79 patients were followed with radiotherapy, 4 patients received chemotherapy. Histopathology revealed pilocytic n=52, pilomyxoid n=68, fibrillary astrocytoma n=1, and grade I papillary-glioneuronal tumor n=3. The overall survival rate was 84.8% with mean follow-up period of 43 months. The event free survival was 61.7% and 84.8 %, the quality of life was 40.4 % and 74.7 %, in surgery only group and surgery followed RT group, respectively. The preservation rate of both visual and endocrinal functions was 63.8 % and 67.1 % in this two groups.

**CONCLUSIONS:** The initial role of surgery in symptomatic OPG is get histological diagnosis and relief of hydrocephalus if there was. In these symptomatic patients, surgical debulking with radiotherapy decreases or stabilizes the tumor size in most cases with less complications after 3.6 years follow-up. The significant prognostic factor confirmed in this study was the age of the patient and radiotherapy. The OPG patients with NF1 was much lower in Chinese patients than the literatures

**Keywords:** pediatric; optic pathway gliomas; surgery

**FL-087****Neuro-oncology****Pediatric adamantinomatous craniopharyngioma cyst fluid demonstrates a pro-inflammatory milieu**

Andrew M Donson<sup>1</sup>, Andrea M Griesinger<sup>2</sup>, Vladimir Amani<sup>2</sup>, Richard C.e. Anderson<sup>3</sup>, Toba N Niazi<sup>4</sup>, Michael H Handler<sup>1</sup>, Nicholas K Foreman<sup>2</sup>, Todd C Hankinson<sup>1</sup>

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**OBJECTIVE:** The cyst component of pediatric adamantinomatous craniopharyngioma (ACP) often exerts substantial mass effect, grows unpredictably, and is poorly responsive to radiation therapy. Current cyst-directed therapies mandate surgical intervention. An improved understanding of the drivers of ACP cyst growth may allow for the identification of systemic therapies that can control cyst expansion and improve tumor control and/or our ability to safely undertake radical tumor resection.

**MATERIAL-METHODS:** Milliplex cytokine expression analysis was used to compare the cytokine milieu of ACP cyst (n=5) to another common cyst generating pediatric brain tumor, Pilocytic Astrocytoma (PA, n=5). The ratio of cytokine levels in the 2 tumor types was compared.

**RESULTS:** Compared to PA cysts, all ACP specimens showed a highly pro-inflammatory cytokine pattern. Among the cytokines with elevated expression level ratios in ACP, relative to PA, were IL-6 (353.9, p=1.23x10<sup>-5</sup>); IL-8 (57.6, p=0.04); IL-10 (35.5, p=0.003); TNF- $\alpha$  (8.21, p=0.01); and IL-1 $\beta$  (3.65, p=0.005).

**CONCLUSIONS:** ACP cyst fluid appears to be characterized by a highly pro-inflammatory cytokine expression pattern. Understanding the cellular source of these cytokines and the efficacy of selective cytokine blockade may represent an opportunity for therapeutic intervention.

**Keywords:** Adamantinomatous Craniopharyngioma, Cytokine Milieu, Cyst Fluid, Cytokine Ratio, Pediatric Craniopharyngioma

**FL-088****Neuro-oncology****Craniopharyngioma in children: a new classification of surgical relevance**

Venkataramana K Neelam<sup>1</sup>, Dr Adesh Jagadeesh<sup>2</sup>, Dr. Murali Mohan Selvam<sup>3</sup>, Dr. Sudheer Hegde<sup>4</sup>

<sup>1</sup>Dr Venkataramana Neelam

<sup>2</sup>Dr Adesh Jagadeesh

<sup>3</sup>Dr. Murali Mohan Selvam

<sup>4</sup>Dr. Sudheer Hegde

**OBJECTIVE:** A Retrospective analysis of imaging characteristics, surgical results and the outcomes based on the surgical approach in 185 craniopharyngiomas with an objective to identify an ideal surgical approach. **MATERIAL-METHODS:** Authors personal experience of 185 Craniopharyngiomas over three decades were retrospectively analysed. 93 (50%) of them were children. 55% were males and 45% were females. The predominant clinical presentation was endocrine, visual and raised intra cranial pressure. Imaging characteristics were analyzed in favour of total removal and better outcomes.

**RESULTS:** 74 craniopharyngiomas underwent surgery. In 1980s there were 40 children. 92% had cysts in C T which is primary diagnostic tool. 66% had total excision through trans cranial procedures, pterional, sub frontal inter hemispheric and intra ventricular (Microscopic). Post operative mortality was 25% and morbidity was significant. Inadequate imaging, neuro and endocrine monitoring were contributory. In 1990s Thirty three children underwent trans cranial procedures. 7 had total excision, the rest were near total removals. The mortality was reduced to 2%. Majority required endocrine replacement. After 2000, 20 children were treated. Trans nasal extended endoscopic approach in 17, Two trans cranial and one had combined approach. Total excision was achieved in 75% with zero mortality. Based on MR imaging they were classified as 1. Pre Infundibular 2. Post Infundibular 3. Trans Infundibular 4. Intra sellar 5. Ectopic / Exophytic 6. Intra ventricular 7. Multi compartmental and 8. Complex types. We recommend trans nasal extended endoscopic approach in Type 1. Type 6 and 7 requires trans cranial/ combined approaches.

**CONCLUSIONS:** Trans nasal endoscopic route is emerging as an ideal surgical approach in every Craniopharyngioma. MRI / CT Imaging helps in choosing the right surgical approach and the feasibility of total removal. The new proposed classification will be useful in deciding the surgical approach

**Keywords:** Craniopharyngioma, Endoscopic Surgery, Classification

**FL-089****Neuro-oncology****Craniopharyngioma in children: the experience of management in a cohort series in Taiwan**

**Tai Tong Wong**

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**OBJECTIVE:**Craniopharyngiomas originate in the sellar or suprasellar site with various extents of intracranial extensions in children. It constitutes of 8.6% in our cohort series. Demography, location of tumor origin, primary management, and outcome of this series were studied.

**MATERIAL-METHODS:**102 cases primarily treated by us from 1971 to 2015 were reviewed. Originate location, achievement of surgical excision, primary post-resection management, survival, and neuropsychosocial profile (25 cases) were analysis. The differences were compared between tumors of the two originate locations.

**RESULTS:**The mean age at diagnosis was 7 years and mean follow-up of 9.8 years. Tumors originated from sellar turcica and suprasellar location were 46 cases and 56 cases respectively. Extents of radical resection and event free survival were better in sellar origin group of tumors. Recurrence rate were higher in the suprasellar origin group of patients. Postoperative radiation therapy for residue tumor or salvage radiotherapy was more frequent in the suprasellar origin group. Gamma knife radiosurgery was applied more frequently than conventional fractionate radiotherapy since 1990s. Neurocognitive profile studies in 25 survivors showed deficits on processing speed, verbal memory, visual memory and sustained attention. Social skill in adaptive behavior and psycho health in quality of life was also disturbed.

**CONCLUSIONS:**In this retrospective hospital cohort study of craniopharyngioma in children, differences existed in extents of tumor resection, event free survival, treatment modalities for post-resection residue/recurrent tumor, event free survival, and psychosocial cognitive outcomes. The findings will provide evidences and reference for future management.

**Keywords:** craniopharyngioma, children, gamma knife radiosurgery, neuropsychosocial profile

**FL-090****Neuro-oncology****Transcranial surgery for recurrent craniopharyngiomas in children**

Reizo Shirane<sup>1</sup>, Tomomi Kimiwada<sup>1</sup>, Toshiaki Hayashi<sup>2</sup>, Teiji Tominaga<sup>3</sup>

<sup>1</sup>Miyagi Children's Hospital

<sup>2</sup>Sendai City hospital

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**OBJECTIVE:**Treatment options for recurrent craniopharyngioma include repeat surgery, radiotherapy, and radiosurgery. We present a series of recurrent craniopharyngiomas in children treated by transcranial surgery between 1998 and 2013 in Tohoku University and Miyagi Children's Hospital and discuss the points of surgical strategy.

**MATERIAL-METHODS:**A retrospective analysis was performed in 27 children younger than 20 years of age who underwent a total of 35 operations for recurrent craniopharyngioma. The mean age at the time of surgery was 9.3 years, and the mean follow-up was 9,1 years.

**RESULTS:**Total resection was achieved in 16 of 27(59%). Additional radiotherapy was applied in 10 cases. Another surgical treatment was needed in 5 cases during follow up periods. There was no operation related death. Favorable disease control was achieved in 24 patients. There were no significant differences in the neurological and functional outcomes between patients with primary and those with recurrent tumors in follow up.

**CONCLUSIONS:**Radical resection is possible in most of patients with recurrent craniopharyngiomas. Prior radiation therapy, hard calcified masses and adhesion to the vascular structure were risk factors for surgery related significant complications. Additional radiotherapy is a good option in order to obtain acceptable disease control.

**Keywords:** recurrent craniopharyngioma, surgery, prognosis

**FL-091****Neuro-oncology****Vascular abnormalities after treatment for craniopharyngioma**

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<sup>3</sup>Department of Radiation Oncology, Children's Hospital Colorado and University of Colorado, Aurora, CO, USA

**OBJECTIVE::** Craniopharyngiomas occur in proximity to vessels of the circle of Willis, which may require manipulation in the course of surgical dissection. The tumors are frequently irradiated which may predispose to vascular abnormalities. We reviewed a large center's experience.

**MATERIAL-METHODS:**Records of the brain tumor program, spanning 21 years, were reviewed for patients with craniopharyngioma. Demographics, details of treatment, clinical course and results of imaging over time are described.

**RESULTS:**50 patients with craniopharyngioma were identified, 8 of whom were excluded for inadequate data. 42 patients were followed over a mean of 7.2 years after treatment, 17 of whom eventually were found to have vascular abnormalities. Of these patients, 2 had received craniotomy alone with no radiation. The remainder had radiation, 4 after failure of initial intracystic therapy and either with and without further operation. 11 were treated with operation, either craniotomy or endoscopic biopsy or resection, followed by radiation. 3 patients developed cavernous malformations, 9 patients had aneurysmal dilatation of one or more vessels, and 10 had vessel narrowing or occlusion, which in some was documented to progress. Mean time to identification of the first abnormality was 3.4 years after radiation, and 1.4 years after operation alone. 2 patients with moyo moyo syndrome after radiation required cerebral revascularization procedures.

**CONCLUSIONS:**A substantial percentage of patients develop vascular abnormalities after treatment for craniopharyngioma. They require long term surveillance and possible late intervention. Follow up imaging must include specific attention to vessels of the circle of Willis, with MR angiography.

**Keywords:** craniopharyngioma, aneurysm, cavernoma, moyo moyo, vascular abnormality

Wednesday, 26 October 2016

16:15 – 17:15

**Flash sessions 7: Neuro-oncology****FL-092****Special topic: Molecular biology****Genomic analysis of pilomyxoid astrocytoma**

Essam Al Shail<sup>1</sup>, Namik Kaya<sup>3</sup>, M. Anas Dababo<sup>2</sup>, Hindi Alhindi<sup>2</sup>, Anwar Ulhaq<sup>3</sup>

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**OBJECTIVE:**Pilocytic Astrocytoma (PA) is the most common pediatric brain tumors and auspiciously, they are not aggressive, typically slow growing brain tumors. Recently, a unique group of tumors previously diagnosed as PA were identified and named as Pilomyxoid Astrocytomas (PMA). The PMAs usually show unique histological characteristics and have a more aggressive clinical course. PMAs mostly appear in the optic-chiasmatic/hypothalamic region but sometime they are reported to be found in the posterior fossa, temporal lobe, and in the spinal cord. The occurrence and location of these tumors are helpful features for differential diagnosis as compared to PA. However molecular markers are still needed for more precision in the diagnosis.

**MATERIAL-METHODS:**We performed a molecular cytogenetic study on formalin-fixed paraffin embedded archived clinical samples of PMAs (n=20) using oncoscan arrays from Affymetrix Inc. (Santa Cruz, CA, USA). Few samples were excluded due to failed quality control (QC) during the cytogenetic analyses. Samples that passed the QC were analyzed for genome-wide aberrations.

**RESULTS:**Interestingly all the samples except for one have a duplication on chromosome 7. Interestingly nearly all the samples have quite similar proximal and distal end breakpoint starting from KIAA1549 gene (approximate genomic coordinate: ~138524080 bp) and reaching to BRAF (10th exon; approximate coordinate: ~140490180 bp). The duplication consists nearly 1.87Mb region and comprises 9 pseudogenes, 9 uncharacterized genes, and 10 genes with OMIM ID including KIAA1549 and BRAF. Other genes that may be of importance are as follows: NDUFB2, MKRN1, RAB19, LOC642355, ADCK2, DENND2A, SLC37A3, KOM7A, PARP12, TBXAS1, HIPK2, CLEC2L, KLRG2, UBN2, LUC7L2, C7orf55.

**CONCLUSIONS:**We will be describing the molecular markers of PMA with its unique features of duplication on chromosome 7. We feel that this will help in verifying the histological diagnosis of PMA.

**Keywords:** Pediatric Brain Tumors, Pilocytic Astrocytoma, Pilomyxoid Astrocytomas, Oncoscan Arrays, Cytogenetic Analyses

#### FL-093

##### Neuro-oncology

#### Proliferative index variability by location in pediatric pilocytic astrocytomas

Albert Tu<sup>1</sup>, Ian Buchanan<sup>2</sup>, Omid Hariri<sup>3</sup>, Harish Babu<sup>4</sup>, Edward Melamed<sup>1</sup>, Lynda Szymanski<sup>1</sup>, Mark Krieger<sup>1</sup>

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<sup>2</sup>University of Southern California

<sup>3</sup>Arrowhead Regional Medical Centre

<sup>4</sup>Cedar Sinai Medical Centre

**OBJECTIVE:**Describe the differences in proliferative indices measured by Ki-67 in pediatric pilocytic astrocytomas according to location of origin

**MATERIAL-METHODS:**A retrospective review of all pilocytic astrocytomas undergoing surgery at Children's Hospital of Los Angeles from 2003 - 2015 with complete radiographic, pathologic, and clinical outcome data. Tumor location was stratified into supratentorial, infratentorial, or suprasellar, and defined as the region from which the majority of the tumor arose. Proliferative index of the tumors was measured by Ki-67.

**RESULTS:**A total of 87 patients were included for study. 54 had infratentorial, 18 had supratentorial, and 15 had optic pathway gliomas. Average proliferative index was 3.58 for infratentorial, 6.98 for supratentorial, and 3.65 for optic pathway lesions. These differences trended towards statistical significance ( $p = 0.17$ ).

**CONCLUSIONS:**Supratentorial tumors trended towards a higher growth rate whereas optic pathway tumors trended towards a lower growth rate. This difference may be have multiple causes included specific tumor genetics,

tumor age, and sampling variability. The role of using proliferative indices in predicting the behaviour of low grade tumors is yet to be determined. Further study is necessary in order to validate these results and identify a role for measuring proliferative indices in clinical practice.

**Keywords:** proliferative index, pilocytic astrocytoma, pediatric brain tumor

#### FL-095

##### Neuro-oncology

#### Pilomyxoid astrocytomas: a single center experience of 20 consecutive cases and review of the literature

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**OBJECTIVE:**The goal of this study is to determine the epidemiology, clinical presentation and treatment outcome of Pilomyxoid astrocytomas treated in a single center at Saudi Arabia.

**MATERIAL-METHODS:**Clinical data of 20 patients of Pilomyxoid astrocytoma under 14 years of age managed at King Faisal Specialist Hospital and Research Center, Riyadh, Saudi Arabia., between January 2001 to December 2015 were reviewed.

**RESULTS:**The study included 15 females and 5 males. Mean age at presentation was 3 years. The most common location of tumor was Chiasmatic/hypothalamic area 12 patients. Other locations were: brainstem 3, spinal cord 1, cerebellum 1, optic nerve 1, thalamus 1 and temporal area 1. The common presentations were signs and symptoms of raised intracranial pressure followed by visual disturbances, focal neurological deficit, nystagmus. Diencephalic syndrome of emaciation and cranial nerve palsies. Gross total resection was achieved in 5, subtotal resection in 10 and biopsy in 5 patients. Histopathology was pure Pilomyxoid astrocytoma 10 patients, intermediate Pilomyxoid astrocytoma 10 patients. 18 patients received chemotherapy consisting of vincristine and carboplatin. Seven patients received focal radiation therapy. Follow up was 1 year to 15 years. Out of 12 Chiasmatic/hypothalamic Pilomyxoid astrocytomas, six patients showed significant clinical improvement with no residual disease, four patients had stable disease and two patients had progression of tumor with CSF metastasis and died within six months of diagnosis. Two brainstem gliomas showed improvement and one had severe neurological deficits with significant residual disease. Spinal cord Pilomyxoid astrocytomas improved with no deficit and no residual disease.

**CONCLUSIONS:**Pilomyxoid Astrocytoma is a glial tumor with aggressive clinical behavior. Most common location is Chiasmatic/hypothalamic area. Gross total resection is not achievable in most of cases. Overall prognosis for Pilomyxoid astrocytoma is poor. Further studies are needed to establish the standard of care and to improve the outcome of Pilomyxoid astrocytoma.

**Keywords:** Pilomyxoid astrocytoma, intermediate Pilomyxoid astrocytoma, mixed Pilocytic Pilomyxoid astrocytoma.

#### FL-096

##### Neuro-oncology

#### Pilocytic cerebellar astrocytomas in children: long-term outcome and quality of life

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**OBJECTIVE:** Cerebellar pilocytic astrocytomas (CPAs) are considered benign tumors, which allow prolonged survivals. We reviewed the long-term clinical outcome and quality of life (QoL) of children operated on for CPAs, at our institution since 1995.

**MATERIAL-METHODS:** The follow-up was available for 39 patients (55% males, 45% females). Clinical, radiological, and outpatient medical records were reviewed. Thirty-two patients were recently interviewed by phone and the PedsQL (Pediatric Quality of Life Inventory) questionnaire was used for QoL assessment. The PedsQL was administered to the parents of pediatric patients (Group A, 15 pts.), and directly to adult patients (group B, 17 pts.).

**RESULTS:** Mean age was 9.3 years at diagnosis, 12 years (group A) and 26 years (group B) at the follow-up. We achieved total resection in 74% of the children. Five patients (12%) were re-operated on for tumor recurrence, on average 3.2 years after the first operation. There was one post-operative mortality due to acute pneumocephalus. All other 38 patients are alive at a mean follow-up of 10 years (range 1–21), 31 free of disease, 7 with stable disease. At the follow-up 19 children (51.4%) had some neurological deficit but this was rarely impacting significantly on their QoL.

Patients in group A were attending regular school, 2 with support. In group B, 2 were attending high school, 4 were attending university, 9 were employed and 2 unemployed. PedsQL scores of both groups were in between those of the normal population and the reference values for patients with chronic diseases, excepted for the emotional scores that were below the scores of patients with chronic diseases.

**CONCLUSIONS:** As a group, children with CPAs have long-term QoL indicators similar to those of the general population. Larger studies may clarify the relative impact on QoL of other variables such as hydrocephalus, tumor location and socioeconomic factors.

**Keywords:** cerebellar tumors, pilocytic astrocytomas, quality of life, long-term outcome

## FL-097

### Neuro-oncology

#### Does tuberin delay the progression to SEGA?

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**OBJECTIVE:** 1. To test for expression of Tuberin protein, by Immunohistochemistry, in SEGA and to correlate its expression in Syndromic and Non syndromic SEGA.

2. Tuberin expression with age of presentation, other features of intracranial pathologies and surgical outcome of SEGA.

3. Role of tuberin in progression of Subependymal nodule to SEGA.

**MATERIAL-METHODS:** 15 patients of Subependymal Giant cell Astrocytoma (SEGA) were treated and followed up over 15 years. Immunohistochemistry was done in all cases for tuberin expression using indirect immunoperoxidase method. Appropriate normal positive, biological positive and negative controls were used. Patients were followed for a mean duration of follow up was 21.2 month (range from 3 – 48 months).

**RESULTS:** Two-thirds of SEGA expressed tuberin protein. Of these, 7 were syndromic (Tuberous Sclerosis Complex-TSC) SEGA, while 3 were Non syndromic (Non TSC) SEGA. Proportion of cases in Syndromic vs. Non Syndromic SEGA either lacking tuberin expression or retaining tuberin staining did not differ significantly ( $p$  – value = 0.593). Majority of SEGA with tuberin expression were >12 years with only one presenting at the age of 7

years. Majority of SEGA in which tuberin was not expressed were less than 10 years. Follow up was available for 11 patients. 2 had died in immediate post-operative period and 2 patient follow-up was not available. Average Glasgow Outcome Scale (GOS) outcome of patients with positive expression of tuberin was 3.875 and 3.2 in which tuberin was not expressed. Cystic changes in the tumour were seen only in cases with positive expression of tuberin.

**CONCLUSIONS:** Tuberin appears to have a role in delaying the manifestation of SEGA. Tuberin may have role in preventing progression of Subependymal nodule to SEGA. Studies with larger cohort is required to completely understand the role of tuberin in pathophysiology and prognosis of SEGA. Implications of cystic changes in SEGA are not clear.

**Keywords:** Subependymal nodule, Subependymal Giant Cell Astrocytoma (SEGA), Tuberin, Syndromic Tuberous Sclerosis Complex (TSC), Non Syndromic TSC, Glasgow Outcome Scale (GOS)

## FL-099

### Special topic: Neuro-imaging

#### The suitability of ADC-mapping to differentiate between recurrent tumor and radiation necrosis in malignant pediatric brain tumors

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**OBJECTIVE:** Malignant brain tumors in children may show recurrence after initial gross total resection and adjuvant treatment. To differentiate recurrent tumor from changes due to radio- and chemotherapy gadolinium enhanced MRI sequences are generally used. These sequences cannot conclusively differentiate between both situations. It is known that diffusion weighted images (DWI) with apparent diffusion coefficient mapping (ADC) may distinguish between benign and malignant tumor tissue in children. The goal of this study was to evaluate the role of ADC-mapping to differentiate recurrent tumor tissue from radiation necrosis.

**MATERIAL-METHODS:** We retrospectively reviewed all patients identified with a recurrent malignant brain tumor after initial treatment who presented during 2008 to 2015 in our department. MRI imaging, clinical presentation, operative procedures and pathology were analyzed. For ADC-mapping of the MRI scans the examiner was blinded to patient's data. In all cases at least two ROI were assigned. ADC-values between 500 and 800 were defined as malignant tumor and ADC-values between 1000 and 1200 as benign changes.

**RESULTS:** Of the 21 patients (11 male, 10 female) identified 17 (81%) had diffusion weighted images (DWI) with ADC-mapping. The most common tumor location was in the posterior fossa and in the parietal region (10 versus 7 cases) retrospectively. In 15 cases the mean ADC-value was 659 (group A). The mean ADC-value of the remaining two cases was 1090 (group B). 16 patients underwent second surgery. The histology of group A showed malignant tumor tissue whereas group B showed radiation necrosis ( $p < 0.05$ ).

**CONCLUSIONS:** This analysis demonstrates that ADC-mapping may distinguish between recurrent malignant brain tumor and radiation necrosis in the pediatric population. As it is not invasive this method may help in decision-making whether surgery is indicated or not. Further data in a larger prospective cohort is needed.

**Keywords:** ADC-mapping, recurrent tumors, children, ROI, MRI

**FL-100****Neuro-oncology****Secondary glioblastoma in patients with intracranial germ cell tumors treated with chemotherapy and radiotherapy**

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**OBJECTIVE:** Because of advances in treatment and improved overall survival of germ cell tumor, it is a concerned issue that radiation induced secondary tumor may happen in the patient treated for germ cell tumor. Here, we report 4 patients with germ cell tumor who developed radiation induced glioblastomas.

**MATERIAL-METHODS:** Since 1980 we treated 89 patients with intracranial germ cell tumor and experienced 4 patients with secondary glioblastoma. We investigated and evaluated the treatment procedure for the original tumor and the clinical courses in all patients with secondary glioblastoma. We also evaluated the characteristics of the secondary glioblastoma.

**RESULTS:** The diagnosis and primary site of germ cell tumor were 2 mixed germ cell tumor at right thalamic and pineal regions and hypothalamic region, choriocarcinoma at suprasellar region and hypothalamic region and germinoma in suprasellar and pineal regions. All patients with non germinomatous germ cell tumors underwent chemotherapy (cisplatin, etoposide ± ifosfamide) and radiotherapy (total 60 Gy). Two patients underwent second-look operations with total removal of the residual tumors. On the other hand, the germinoma patient underwent only radiotherapy (total 50 Gy). Since this germinoma case had been treated in 1989, high total dose radiotherapy was performed to germinoma patient. Residual tumor was not appeared after the radiotherapy, therefore chemotherapy was not performed. All cases remained free from the tumor recurrence. However, they developed glioblastoma in the field of radiotherapy 10–25 years later after initial treatments, respectively. All secondary tumors were pathologically diagnosed as glioblastomas developed in the field of radiotherapy such as basal ganglia and/or brain stem.

**CONCLUSIONS:** The total dose of radiotherapy may correlate with occurrence of secondary glioblastoma and the central part of brain may be the predilection site. The mechanism of the development for secondary glial tumor is unclear, however, radiotherapy but not chemotherapy may contribute to genetic toxicity.

**Keywords:** germ cell tumor secondary tumor glioblastoma

**FL-101****Neuro-oncology****Pediatric infratentorial ganglioglioma: results of a series of 35 children and literature review**

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**OBJECTIVE:** Pediatric infratentorial gangliogliomas are rare tumors, thus the prognostic factors are poorly understood and their optimal management has still to be defined

**MATERIAL-METHODS:** We reviewed the results of 35 children treated in our institution and reviewed the corresponding literature

**RESULTS:** The median age was 6.5 years. At diagnosis, 69% had signs of raised ICP, 60% a cerebellar syndrome and/or cranial nerve palsy. Half of them were transitional forms, 5 were developed in the tectal plate or the cervico-medullary junction (CMJ), 4 in the cerebellum and 3 in the pons. 86.5% had a contrast-enhancing part on MRI which could be totally resected for 24 children. The histology revealed 10% of malignant forms and 34% of BRAF mutations. After a median FU of 5 years, 3 deaths occurred, 1/3 was in complete remission, 1/3 had no more contrast enhancing part but a stable hyperT2 lesion and the remaining had still a contrast enhancing part. The overall survival (OS) was better if the contrast enhancing part has been removed (p=0.03) and a better progression free survival was correlated to the benign lesions (p=0.039) but not to BRAF status.

From our results and the literature review, a gross total resection should be attempted to remove the contrast enhancing part of the tumor, which may be possible in most of the cerebellar/transitional gangliogliomas and some of the brainstem lesions. CMJ ganglioglioma are the least amenable to complete resection. Chemotherapy has still a limited role. Radiotherapy exposes to risk of malignant transformation and should be reserved for progressive unresectable tumors

**CONCLUSIONS:** These tumors are rare and challenging. To date, adjuvant therapy has a limited role but targeted therapies against BRAFm are promising. The surgical resection of well-defined contrast enhancing parts should be attempted even in staged surgeries and balanced with the risks of neurological deterioration

**Keywords:** ganglioglioma, posterior fossa, pediatric

**FL-102****Neuro-oncology****The changing world in pediatric brain tumors: impact of advanced molecular analysis on clinical management**

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<sup>2</sup>Department of Pediatric Oncology, Hematology&Immunology, University Hospital Heidelberg, Germany

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**OBJECTIVE:** Over the past decades there are tremendous advances in the diagnostic and therapeutic approaches to pediatric brain tumors. A better understanding of the pathogenesis leads to the development of new stratifications and risk adapted individualized therapies. We show the increased importance of molecular analysis in diagnostic and therapeutic managements in 4 children.

**MATERIAL-METHODS:** We present 4 children aged between 3 month and 11 years. The first patient suffered from a metastasized medulloblastoma, second from glioblastoma WHO IV°, third from embryonal tumor with abundant rosettes (ETANR) and fourth from desmoplastic infantile ganglioglioma (DIG). Case 1: tumor resection followed by chemo-/radiotherapy with complete remission, local relapse 3 years later. Case 2: complete tumor resection followed by chemo- and radiotherapy. Case 3: total surgical resection, histology showed ETANTR, chemotherapy. Case 4: partial tumor resection and chemotherapy.

**RESULTS:**The molecular analysis in case 1 was GBM, that means a second tumor manifestation and not a relapse of medulloblastoma. Further therapy was adapted. Histology of case 2 was a pilocytic astrocytoma, molecular analysis showed GBM with low profile. 4 years follow up without relapse. Case 3: embryonal tumor with multilayered rosettes (ETMR), 1 year later tumor relapse, surgical resection, chemo- and radiotherapy followed. Case 4: showed a BRAF V600E mutation. 1 y after chemotherapy tumor progress and an individualized therapy with BRAF-inhibitor (Vemurafemib) resulted in an extensive tumor shrinking.

**CONCLUSIONS:**Molecular analysis enables a better disease classification and risk stratification of pediatric brain tumors. Identification of new molecular biomarkers could play a pivot role in further diagnosis and risk adapted therapeutic approaches to improve overall survival and decrease in long-term morbidity of pediatric brain tumors.

**Keywords:** molecular analysis, medulloblastoma, glioblastoma, ETANTR, DIG

## FL-103

### Special topic: Molecular biology

#### Histological subtypes of medulloblastoma and its correlation with genetic abnormalities

Jennifer S. Ronecker<sup>1</sup>, Raphael Salles Scortegagna De Medeiros<sup>2</sup>, Sidnei Epelman<sup>2</sup>, Michael Tobias<sup>1</sup>, Avinash Mohan<sup>1</sup>, Alex Braun<sup>3</sup>, Raj Murali<sup>1</sup>, Nelci Zanon<sup>4</sup>, Meena Jhanwar Uniyal<sup>1</sup>

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**OBJECTIVE:**Medulloblastoma (MB) is the most common primary pediatric malignant brain tumor, which presents with profound molecular heterogeneity with different developmental origins. Recent genetic analysis has identified four genetic subtypes, which correspond to distinct histologic subtypes: classic (WNT), sonic hedgehog (Shh), group 3 and group 4. Here, we studied specific immunohistological markers, their correlation with the amplification of the oncogene Myc and loss of tumor suppressor gene p53, and their association with metastatic potential.

**MATERIAL-METHODS:**IRB approved MB tumor specimens (n=51) were used for immunohistochemical and genetic analysis. Immunohistochemical analysis was done to evaluate the expression of glioma transcription factor 1 (GLI-1), Grb2-associated binding protein 1 (GAB-1), natriuretic peptide receptor (NPR), and voltage-gated potassium channel (KV1). FISH analysis using a DNA-based probe was done to determine the amplification of Myc or loss of p53 gene, in order to correlate with the histological findings. In addition, a specific p53-mutant medulloblastoma cell line was used to determine the signaling pathway leading to proliferation, migration, and drug resistance using Shh pathway and mTOR pathway inhibitors.

**RESULTS:**The results demonstrated that GAB-1 was highly expressed in the Shh group (82%) and less in other subtypes. Expression of KV1 was evenly distributed in all subtypes. No obvious correlation with expression of GLI-1, GAB-1, NPR and KV1 with metastatic potential was found. Analysis of loss of p53 and overexpression of Myc defined in these subtypes. Furthermore, in the cell line with p53 mutant, mTOR pathway was activated, which was inhibited by specific mTOR inhibitors.

**CONCLUSIONS:**The findings of this study infer that expression of GLI-1, GAB-1, NPR, and KV1 was important in defining the subgroups of medulloblastoma. However, their role in metastatic potential was unclear. Moreover, the association of loss of p53 and/or amplification of Myc may be a suitable marker in defining the subset of MB with metastatic

potential.

**Keywords:** p53, medulloblastoma, myc, sonic hedgehog, mTOR

## FL-104

### Neuro-oncology

#### The expression pattern of RBM5 in children with medulloblastoma

Hao Li

Children's hospital of Fudan university, shanghai, China

**OBJECTIVE:**To detect the expression pattern of RBM5 in medulloblastoma tissues and to explore the role as well as the mechanism of RBM5 in carcinogenesis and progression of medulloblastoma.

**MATERIAL-METHODS:**Exome-sequencing was used to detect the mutation of RBM5 in medulloblastoma tissues while IHC was used to analyze the expression pattern of RBM5 in medulloblastoma tissues. Then K-M analysis was employed to evaluate the correlation of RBM5 expression with survival time of patients. The role and molecular mechanism of RBM5 in medulloblastoma was studied by RNAi and Western blot technology.

**RESULTS:**We found that there was mutation in RBM5 in 4 of 40 medulloblastoma tissues. The expression level of RBM5 was significantly lower in medulloblastoma than that of adjacent normal control by RT-QPCR or IHC. And low RBM5 expression was correlated with a poor prognosis in medulloblastoma. Then knock-down of RBM5 induced cell proliferation and cell migration. On the contrary, over-expression of RBM5 in Daoy cells repressed cell proliferation and migration. Moreover, we found the mRNA levels of  $\beta$ -catenin, LEF1, CyclinD1, BCL-2 were upregulated while the expression of DKK1 and caspase3 was downregulated in Daoy cells with RBM5 knock-down. These data indicated that RBM5 may play an tumor suppressor role in medulloblastoma through Wnt/ $\beta$ -catenin signaling and caspase3-mediated apoptosis.

**CONCLUSIONS:**RBM5 functions as a tumor suppressor in medulloblastoma by regulating Wnt/ $\beta$ -catenin signaling and caspase3-mediated apoptosis. RBM5 could be developed as a marker for diagnosis and prognosis of medulloblastoma in future.

**Keywords:** children medulloblastoma RBM5 apoptosis Wnt prognosis

## FL-105

### Neuro-oncology

#### Protoporphyrin IX accumulation and sensitivity to photodynamic therapy of medulloblastoma cells after application of 5-aminolaevulinic acid

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**OBJECTIVE:**Fluorescence-guided resection using 5-aminolaevulinic acid (5-ALA) is well established in adult glioblastoma therapy. Moreover, protoporphyrin IX (PPIX), which accumulates selectively in tumor cells after application of 5-ALA, has been used as a photosensitizer in photodynamic therapy (PDT) to mediate phototoxicity after laser light exposure. In pediatric brain tumors, PPIX accumulation and sensitivity of tumor cells to PDT have not been studied in detail, although both fluorescence-guided surgery and PDT

might have clinical implications in children. We have therefore investigated the effect of 5-ALA in vitro in medulloblastoma cell lines.

**MATERIAL-METHODS:**The medulloblastoma cell lines D283, MED8A and ONS76 were treated with graded doses of 5-ALA (0–100 µg/ml). PPIX accumulation was detected by flow cytometry. After PDT with exposure to laser light of 635 nm wavelength, viability of cells was studied using the colorimetric WST-1 assay. Findings were compared with controls and with the glioblastoma cell line U373.

**RESULTS:**D283, MED8A and ONS76 medulloblastoma cells accumulated PPIX in a time- and dose-dependent fashion. After 6 hours of incubation with 100 µg/ml 5-ALA the mean frequencies of positive cells ( $\pm$ SEM) were 66.4  $\pm$ 3.9%, 91.2 $\pm$ 4.7% and 74.8 $\pm$ 7.5%, respectively (Fig. 1). Similar results were obtained with U373 (94.3 $\pm$ 2.3%), although maximum accumulation was achieved already at lower 5-ALA concentrations. Subsequent PDT resulted in a dose-dependent reduction in viability of D283 (72.2 $\pm$ 2.0%) and MED8A (64.7 $\pm$ 3.7%), but only to a lesser extent of ONS76 cells (19.1 $\pm$ 9.1%) (Fig. 2). **CONCLUSIONS:**Medulloblastoma cells accumulate PPIX when exposed to 5-ALA and are susceptible to PDT-induced phototoxicity. However, the individual cell lines differ substantially with regard to PPIX levels and response to PDT. Thus, our in vitro data suggest that fluorescence-guided resection as well PDT of medulloblastomas might be feasible, although a possible medulloblastoma subtype-specific response to 5-ALA as well as dose- and time-dependent effects warrant further studies in pediatric tumors.

**Keywords:** medulloblastoma, 5-aminolaevulinic acid, protoporphyrin IX, photodynamic therapy

#### FL-106

##### Neuro-oncology

#### High dose chemotherapy and autologous stem cell rescue for AT/RT of CNS: report of 10 cases

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<sup>3</sup>School of Medicine, National Yang-Ming University, Taipei, Taiwan

**OBJECTIVE:**The treatment of CNS AT/RT consisting of surgery followed by chemotherapy and radiotherapy are reported to have greater overall survival. However, the majority of AT/RT patients are under 2 y/o, who are too young for radiotherapy. Several studies have documented high dose chemotherapy with autologous stem cell rescue (HDC-ASCR) to be effective. We present the results of HDC-ASCR for CNS AT/RT at our institute.

**MATERIAL-METHODS:**From Mar, 2010 to Mar, 2016, there are ten CNS AT/RT patients received HDC-ASCR as part of treatment. All of the patients were under 2 y/o at diagnosis. The patients' characteristics, disease status, stem cell dose, engraftment status, post-transplant complication and outcome were analysed.

**RESULTS:**There were ten CNS AT/RT patients received total 16 cycles of HDC and ASCR at progressive disease (PD, n=4), partial response (PR, n=3), and complete response (CR, n=3) enrolled with mean follow-up period of 24.5 months. The male to female ratio was 4 to 6. Median age at diagnosis and transplant was 1.5 y/o and 2.1 y/o. 7 patients received radiotherapy and 4 patient died. The median survival time was 21.7 months. The 5-year overall survival and progression free survival rates were 44.4% and 38.1%, respectively.

**CONCLUSIONS:**With successful surgery, engraftment and manageable toxicity, the outcome of CNS AT/RT was improved as compared to our previous cohort.

**Keywords:** CNS tumors, Atypical teratoid/rhabdoid tumor, autologous transplant, Surgery

Thursday, 27 October 2016  
08:00 – 09:00

## Flash sessions 8: Hydrocephalus

#### FL-107

##### Hydrocephalus

#### Harvey Cushing's early treatment of congenital hydrocephalus

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**OBJECTIVE:**At the turn of the twentieth century, the importance of diagnosing and treating congenital hydrocephalus was apparent to clinicians, although the most effective methods for cerebrospinal fluid drainage had yet to be elucidated. While the details of specific and unconventional procedures have previously been described by this group, Cushing's broader contributions to operative management of congenital hydrocephalus have not been discussed. **MATERIAL-METHODS:**Following IRB approval, and through the courtesy of the Alan Mason Chesney Archives, the surgical files of the Johns Hopkins Hospital, from 1896 to 1912 were reviewed. Patients operated upon by Dr. Harvey Cushing were selected; from this cohort, pediatric patients with an admitting diagnosis of hydrocephalus were reviewed.

**RESULTS:**In total, 11 patients underwent operative intervention for congenital hydrocephalus. The mean age at the time of first operation was 1.01 years. Operative procedures for permanent CSF drainage included 6 intra-abdominal drainage procedures, 1 vascular shunt, 2 ventriculostomies, and 1 callosal puncture. Temporary CSF drainage procedures, occasionally performed in the operating room, included 17 lumbar punctures, 20 ventricular punctures, and 4 ventricular aspirations.

**CONCLUSIONS:**The broad range of CSF drainage procedures employed by Cushing in these patients demonstrate his commitment to improving the quality of life in his patients, as well as helping to drive the field of pediatric neurosurgery forward.

**Keywords:** hydrocephalus, CSF drainage, CSF diversion, Harvey Cushing

#### FL-108

##### Hydrocephalus

#### Shunt-assistants with fixed versus adjustable opening pressures: comparison of clinical parameters in the course of the treatment of pediatric hydrocephalus

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**OBJECTIVE:**The overdrainage of shunts in the treatment of pediatric hydrocephalus is - in the long term - one of the most frequent and most challenging complications. Since many years, anti-gravity valves are available to treat overdrainage. We studied retrospectively possible differences in the course of hydrocephalus treatment when using shunt-assistants with fixed opening pressures (SA; 10 - 35 cm H<sub>2</sub>O) versus shunt-assistants with postoperatively adjustable opening pressures (proSA; 0-40 cm H<sub>2</sub>O).

**MATERIAL-METHODS:**The data of 46 children, aged 0-16 years and operated on in the Department of Pediatric Neurosurgery Mainz between 2003 and 2012, were evaluated. The studied parameters were the type of shunt-assistant

(SA versus proSA), the time of recovery from complaints and the frequency of necessary secondary operations.

**RESULTS:**In 27 children, a shunt-assistant with fixed opening pressure (SA) and in 19 patients, a shunt-assistant with postoperatively adjustable opening pressure (proSA, both devices Miethke, Potsdam, Germany) was implanted. From the 27 children with SA, 15 (55%) had to undergo a revision surgery (change of the valve) because of ongoing slit ventricle problems. In the 19 cases with proSA, the opening pressure had to be adjusted in the postoperative period up to 9 times per patient (average of adjustments during the first year: 2.3 per patient), but no revision surgery was necessary. The mean time until recovery from complaints was 6 years in SA patients and 2 years in proSA patients.

**CONCLUSIONS:**The results confirm the apriori plausible assumption that the frequency of necessary revision surgeries and the recovery time are markedly lower when using a postoperatively adjustable shunt-assistant. In particular the reduction of secondary surgeries overweighs the primarily higher costs of adjustable shunt assistants.

**Keywords:** Hydrocephalus, Overdrainage, Shunt Assistant, Adjustable Valve

#### FL-109

##### Hydrocephalus

#### Standardized protocol for perioperative managements of CSF shunting in hydrocephalic infants decreases infectious complications: single-center study

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**OBJECTIVE:**Shunt infection and shunt malfunction are common complications for CSF shunting in hydrocephalic infants. Prevention of these complications in shunt surgery is important for improvement of clinical outcome and reduction of hospital cost. In Japan, many young neurosurgeons and other medical staffs participate in shunt surgeries and perioperative management at university hospitals like our institution. In an effort to improve the clinical outcome of ventriculoperitoneal shunting, a standardized protocol was developed with the cooperation of some expert pediatric neurosurgeons and pediatricians. We analyzed clinical parameters associated with shunt infection and shunt malfunction and estimated the effect of the protocol associated with perioperative management.

**MATERIAL-METHODS:**Standardized protocol containing preoperative and postoperative management was introduced at shunt surgeries in children from 2007 at Osaka University Medical Hospital. 131 ventriculoperitoneal shunting procedures for 83 children were performed from 1996 to 2016. Forty-six children underwent surgery between 1996 and 2006 (group A) and thirty-seven between 2007 and 2016 (group B). We analyzed the influence of the protocol on infection rate, shunt survival time and other clinical parameters including administration period of antibiotics and highest value of CRP.

**RESULTS:**Total infection rate of all shunting procedures was 10.2%. Infection rate was 13.0% in infants under 12 months of age. Infection rate for infants significantly decreased from 18.6% (group A) prior to the protocol to 6.3% (group B) while using the protocol ( $p < 0.01$ ). In group B, both administration period of antibiotics and highest value of CRP remarkably improved due to introduction of the protocol, compared with A. Shunt survival time calculated by Kaplan-Meier analysis was not influenced by the protocol.

**CONCLUSIONS:**Standardized protocol for perioperative practice can improve shunt infection rate and reduce hospital cost, should be used in other

educational institution. Further multicenter study is needed to verify this study.

**Keywords:** CSF shunting, hydrocephalus, infant, perioperative management, standardized protocol

#### FL-110

##### Hydrocephalus

#### Age as a novel risk factor for revision of ventriculopleural shunt in pediatric patients

Edward Frederick Melamed, Eisha Anne Chrisitan, Mark Darren Krieger, Cherisse Berry, Parham Yashar, J. Gordon McComb  
Division of Neurosurgery, Children's Hospital Los Angeles, California, USA

**OBJECTIVE:**The first choice for distal end location of a cerebrospinal fluid diverting device is the abdomen. However, for patients in whom the peritoneal cavity is not suitable, a ventriculopleural shunt (VPL) is an alternative.

**MATERIAL-METHODS:**With IRB approval we performed a retrospective review of all patients with ventriculopleural shunts inserted at our institution from 1977-2013.

**RESULTS:**131 (78 M) patients were identified. Mean age at insertion of VPL was  $14 \pm 5$  years. Prior to VPL insertion, 58 patients with available preoperative data had experienced of mean of  $2 \pm 3$  revisions. These patients underwent a mean of  $1 \pm 1$  subsequent revisions of their VPL ( $p < 0.01$ ).

59/131 (45%) patients underwent revision of VPL; malfunction 32/59 (54%), pleural effusion 18/59 (31%), and infection 9/59 (15%). Median revision free duration was 3.6 years. All effusions required the distal end of the shunt be removed from the pleural space, in contrast to 20% of other indications ( $p < 0.001$ ).

Binary regression found that for each additional year in age at the time of VPL insertion, patients experienced an almost 10% reduced risk of revision (Exp B=0.91, CI 0.84, 1.00). More precisely, eleven-years-of-age was the threshold value at which revision rate differentiated. Among 112 patients with minimum one year follow up, 38/82 (46%) patients eleven or older underwent revision in contrast to 21/30 (70%) under eleven ( $p < 0.05$ ). Strengthening this finding, binary regression found a risk ratio for revision of 2.9 (CI 1.1, 7.5) for patients under eleven.

**CONCLUSIONS:**The mean number of revisions dropped significantly after VPL shunt insertion. Malfunction accounted for the majority of indications for VPL revision, but the site of distal drainage was changed mainly in the case of pleural effusions. Younger patients in our population experienced a higher risk for revision, with the rate differentiating at 11 years.

**Keywords:** Ventriculopleural shunt, hydrocephalus, cerebrospinal fluid, shunt revision, age, pleural shunt

#### FL-111

##### Hydrocephalus

#### Ventriculoperitoneal vs percutaneously-placed ventriculoatrial shunts: which is better? A single institution's 13-year experience

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**OBJECTIVE:**The use of VP shunts have long been considered the standard treatment for hydrocephalus, relegating VA shunts as a secondary procedure when unable to utilize the peritoneal cavity as a distal site. However, there is a paucity of data evaluating the long-term effectiveness and safety of VA shunts as well as comparing the two treatment approaches. Review of our 13 year experience with both VP and VA shunts was undertaken to help clarify this question.

**MATERIAL-METHODS:**The authors retrospectively analyzed all CSF shunting procedures performed from January 1st, 2000, until December 31st, 2013, comparing both VP and VA shunt cohorts for demographics and outcome

**RESULTS:**459 VPS patients and 85 VAS patients were reviewed(F/U VPS:2178 d, VAS:1936 d). 984 VPS and 172 VAS procedures were performed. Median survival for VPS:1991 days,VAS:940 days(not excluding elective lengthening). 46% of VPS were never revised vs 40% for VAS. 7% of VPS had > 3 revisions in contrast to 1% for VAS. 80/85 VAS patients were initially treated with VPS. VPS demonstrated a significantly greater overall survival compared to VAS < 7 years of age( $p=0.001$ ) but no difference seen > 7y ( $p=0.42$ ). There was no difference in failure rate from infection, or malfunction of the proximal/ distal catheter/valve. Rates of failure at 90 days, 1 year and 2 years were equivalent. Long-term survival rates are equivalent when VAS lengthening procedures are excluded ( $p=0.38$ )

**CONCLUSIONS:**VA shunts, placed percutaneously, demonstrate an excellent long-term efficacy with minimal morbidity and appear to have similar long term outcomes for the older patients(excluding elective lengthening) when compared to VP shunts. The number of patients undergoing multiple revisions (>3) was significantly less for VAS. These findings suggest that VA shunts should be considered as a valuable and viable option in difficult, shunt-dependent patients, with limited abdominal access.

**Keywords:** hydrocephalus, VA shunts, percutaneous VA shunts, VP shunts

#### FL-112

##### Hydrocephalus

##### Impact of CSF parameters on decision making in the management of bacterial shunt infection: results from a global online survey

Adrian Caceres

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**OBJECTIVE:**CSF shunt infection remains a challenge despite recent advances in shunt technology, surgical techniques and antibiotic prophylaxis. It remains the most expensive medical device related infection in developed countries. Despite its global occurrence, there are great swings in the initial approach, length of treatment and specially the decision to consider the patient viable for reinsertion of the CSF shunt based on the various parameters of CSF, the isolated microorganism and the length of treatment.

**MATERIAL-METHODS:**An online survey was conducted among pediatric neurosurgical centers where attitudes, beliefs and decision making for management of CSF shunt infection was explored. 75 responses were obtained from all 5 continents, representing 75 institutions where initial management after csf parameters indicated infection was obtained as well as current beliefs regarding CSF output, cellularity with special attention to the number of leucocytes, glucose, protein and globulin presence

**RESULTS:**Most institutions will perform CSF shunt removal with EVD drainage, length of treatment varies between 10 to 21 days depending on the isolated microorganism. There is great difference between institutions as to what should be an adequate number of CSF WBC, protein, globulins and

**CONCLUSIONS:**The pattern of management for CSF shunt infection seems to follow a trend towards VP shunt removal and EVD placement, however there exists great discrepancy as to which number should be the ideal standard when it comes to WBC, protein, glucose

and globulin contents of CSF. Further studies should be conducted to measure the statistical impact of these choices on the incidence of CSF shunt reinfection in this particular complex scenario.

**Keywords:** CSF shunt infection, treatment, antibiotic, pediatric,

#### FL-113

##### Hydrocephalus

##### Ventriculo-subgaleal shunt using an antibiotic-impregnated catheter: a retrospective analysis of our institutional experience

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**OBJECTIVE:**To assess the safety and effectiveness of ventriculo-subgaleal shunt (VSS), using an antibiotic-impregnated shunt, in the management of hydrocephalus.

**MATERIAL-METHODS:**We performed a retrospective analysis of VSS performed at our Institution in the period 2013-2015. Antibiotic-impregnated catheter (Bactiseal, Codman®) along with a right angle connector were used to manufacture a patient-tailored shunt. In case of collapsed or tense subgaleal pouch with signs of elevated intracranial pressure, a revision of the VSS was performed while tapping of the pouch was always avoided to reduce the risk of infection. Patients were monitored with serial ultrasounds.

**RESULTS:**We collected 36 patients receiving VSS. Main indication was post-hemorrhagic hydrocephalus in 32 preterm babies, followed by post-infectious in 3 patients and systemic conditions contraindicating the implant of permanent shunt in a single case. One patient also received subduro-subgaleal shunt and another patient received a IV ventricle-subgaleal shunt. Time of permanence of VSS ranged from 15 days to 9 months, with a median time of 32 days. Revision was required in a half of the case. Complications were: one case of hemorrhage secondary to abrupt ventricular drainage, that did not require additional surgery, and two case of infection in patients who underwent multiple surgical procedures (including endoscopy for multiloculated hydrocephalus). All patients were converted to a permanent shunt except for 2 patients who died because of complications of prematurity and one patient who was made shunt-free with endoscopic third-ventriculostomy.

**CONCLUSIONS:**VSS is a safe and effective method to control hydrocephalus in critical babies. Management of patients harboring VSS resulted easier than other options, such as external ventricular drainage, to nurses and intensive care physicians. Our policy, based on the use of antibiotic-impregnated catheter and avoidance of tapping the subgaleal pouch, contained the rate of infection to about 5% in this particular subset of patients.

**Keywords:** antibiotic-impregnated catheter, hydrocephalus, post-hemorrhagic, post-infectious, ventriculo-subgaleal shunt

#### FL-114

##### Spine

##### Ventriculoperitoneal shunt complications and scoliosis surgery

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**OBJECTIVE:**Some populations of patients with spinal scoliosis are associated with hydrocephalus. Especially many patients with myelomeningocele have ventriculoperitoneal shunts. Shunt complications can occur after the surgical procedure of spinal deformity. The purpose of this study was to predict the risk of shunt complications reviewing our experience at one of the largest scoliosis centers in Japan.

**MATERIAL-METHODS:**We retrospectively reviewed a single institution's series of 994 patients with severe spinal deformities under 18 years old at surgery. 2031 surgical procedures were performed between 2006 and 2015.

**RESULTS:**34 patients had functioning shunts. 1 mortality case had a history of Dandy-Walker malformation. 6-year-old girl suddenly deteriorated two days after the anterior release and growing rod insertion. She was not able to recover from uncontrollable intracranial hypertension due to shunt obstruction. 6 patients with myelomeningocele developed symptomatic shunt malfunction. 4 of them required surgical procedures. 2 of them who were conservatively treated showed fluctuating symptoms related to the positions under halo traction or bed rest after scoliosis correction. The ventricular catheters were not in appropriate status radiologically in 4 failed cases.

**CONCLUSIONS:**21% complication rate including 3% mortality had a significant impact on the patients with ventriculoperitoneal shunts having scoliosis correction. Neurosurgeons should evaluate the patients' shunt status carefully before they undergo scoliosis surgery.

**Keywords:** scoliosis, ventriculo-peritoneal shunt, hydrocephalus, complication

#### FL-115

##### Hydrocephalus

##### Late paediatric ventriculoperitoneal shunt failures: an institution's experience

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**OBJECTIVE:**The introduction of ventriculo-peritoneal shunts radically influenced the treatment of hydrocephalus in the pediatric population. Much information has been published reviewing the causes of early shunt failure in the first few years after shunt insertions; however, there is a paucity of data regarding the causes of late shunt failures. We conducted a review to elucidate the causes of late shunt failures in our institution.

**MATERIAL-METHODS:**A 10 year retrospective analysis of all the patients who were treated in our institution from 2006 to 2015 was conducted. Patients who had to undergo shunt revision more than 5 years after their initial shunt insertion were included in our study. Patient's casenotes and scans were reviewed to obtain patient's age, time taken till shunt fails, reason for failure and patient's follow-up.

**RESULTS:**48 patients in our institution had late shunt failures in the last 10 years. Their ages ranged from 7 to 26 years old (mean  $12.23 \pm 4.459$ ). Time taken for shunts to fail occurred between 6 and 24 years (mean  $10.25 \pm 3.77$ ). Reasons for failure included shunt fracture in 23 patients (47.9%), shunt blockage in 15 patients (31.2%), shunt tract fibrosis in 5 patients (10.4%), proximal shunt migration in 3 patients (6.3%) and shunt erosion in 2 patients (4.2%). Follow-up ranged from 6 to 138 months (mean  $45.15 \pm 33.26$ ). The degradation of barium plays an important role in shunt tract fibrosis which results in pain and discomfort. This phenomenon when coupled with patient's growth spurt, also plays an important role in shunt dislodgement and fracture.

**CONCLUSIONS:**Reasons for late shunt failure differs from shunts that fail within the first few years of insertion. The use of barium lined instead of barium coated shunts may help to prevent many of the late

shunt complications in this study.

**Keywords:** Ventriculoperitoneal shunt, Hydrocephalus, Shunt fracture

#### FL-116

##### Hydrocephalus

##### Long-term outcome of shunted pediatric patients in Eastern Finnish population 2003-2013

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**OBJECTIVE:**Shunting can be life-saving procedure and reduces morbidity of hydrocephalus (HC) but can also cause multiple complications that may require several surgical procedures during a patient's lifetime. The main objective of this study is to review outcomes of pediatric patients in our single institution.

**MATERIAL-METHODS:**A total of 80 shunted pediatric patients were analyzed in a population-based setting, between 2003 and 2013. All patients were under 16 years old at the time of initial shunt placement.

**RESULTS:**The mean follow up time was 3.3 years. The mean age at the time of initial shunt placement was 3.2 years. A total of 41 (51 %) patients underwent shunt revision with a mean time to first revision being 8 months. Most common reasons for HC requiring shunting were tumors 22 (27.5 %), congenital defects (CD) 18 (22.5 %), intraventricular hemorrhage (IVH) 15 (19 %) and aqueductal stenosis 10 (12.5 %). Patients with CD ( $p=0.017$ ) or IVH ( $p=0.016$ ) had higher revision rates as compared with tumor patients. There was no difference in revision rates between patients with programmable vs. non-programmable valves ( $p = 0.632$ ). The imaging analysis showed lesser change in the mean biparietal measure change between pre- and postoperative images for the patients that underwent shunt revision as compared to the ones who did not require further shunt surgery ( $p = 0.003$ ).

**CONCLUSIONS:**Just over half of the patients (51 %) underwent at least one revision during follow up. Most of the revisions were made during the first year after the initial shunt surgery. IVH and CD etiologies for HC were associated with higher risk for shunt revision.

**Keywords:** Hydrocephalus, ventriculoperitoneal shunt, shunt revision, shunt survival, intraventricular hemorrhage, tumor

#### FL-117

##### Hydrocephalus

##### ShuntScope in pediatric neurosurgery: ventricle catheter positioning into the third ventricle

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Department of Neurosurgery, Krankenhaus Ludmillerstift, Meppen, Germany

**OBJECTIVE:**In pediatric hydrocephalus insertion of the ventricle catheter can be challenging. Furthermore, increase of head circumference may lead to dislocation of the ventricle catheter. To achieve the best shunt function and to avoid revisions, correct placement of the ventricle catheter is crucial. Here we present a strategy of placing the catheter into the third ventricle to avoid dislocation in growing children.

**MATERIAL-METHODS:**The semi-rigid ShuntScope with an outer diameter of 1 mm and a resolution of 10.000 pixels was used in 24 cases of hydrocephalus. The clinical indications were slit ventricles, placement of CSF reservoirs, shunt implantations in preterm and newborn infants. In young children with

ongoing head growth and in cases of slit ventricles, the catheter was planned to be placed into the third ventricle. Neuronavigation was used to plan and optimize the trajectory. In order to gain an insight view into the ventricles through the otherwise closed catheter, the catheter tip was slit.

**RESULTS:**A correct positioning of ventricle catheters according to pre-operative trajectory planning was possible in all 24 children. Additionally, intra-operative neuronavigation was complementary used to optimize the entry point and trajectory in order to avoid bleeding. No complications like intraventricular or parenchymal hemorrhage, infections or catheter occlusions occurred. The image resolution of this endoscope is astonishing, especially in combination with full HD cameras. However, already slight bleeding degrades the image quality.

**CONCLUSIONS:**The ShuntScope is a safe and effective instrument to solve challenging cases of ventricle catheter placement, especially when placement into the third ventricle is desired. Positioning of the ventricle catheter to the third ventricle can avoid dislocations to the brain parenchyma caused by physiological head growth and thereby prevent occlusion of catheters in slit ventricles. This helps to decrease the number of shunt revisions. Thus, the ShuntScope is recommended for all difficult applications of ventricle catheters.

**Keywords:** ShuntScope, Endoscopy, Hydrocephalus, Slit Ventricle

## FL-118

### Hydrocephalus

#### Endoscopic third ventriculostomy and shunt removal for pediatric hydrocephalus previously treated by ventriculoperitoneal shunt

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<sup>2</sup>Department of Pediatric Neurosurgery, Takatsuki General Hospital, Takatsuki, Japan

**OBJECTIVE:**Pediatric patients with hydrocephalus who were treated by ventriculoperitoneal shunts were thought to be shunt dependent for their lifetime. However, using endoscopic third ventriculostomy, some patients have possibility to be shunt-free, especially if their form of hydrocephalus is noncommunicating.

**MATERIAL-METHODS:**From 2002, 33 pediatric hydrocephalus cases underwent ETV and shunt removal (mean age 21.0 years). All cases had ventriculoperitoneal shunt previously, and underwent imaging study to assess aqueductal stenosis, or a bowing of the third ventricular floor to assure the noncommunicating hydrocephalus. Mean follow-up period after the procedure was 42.8 months. The 33 patients treated included those with myelomeningocele (12), aqueductal stenosis (14), brain tumor (4) and other conditions(3). 26 patients (group A) underwent ETV and shunt removal when their shunts were blocked. In other 7 patients, prior to endoscopic intervention, distal end of the catheter was externalized, and opening pressure was gradually elevated up until enlargement of the ventricle was documented. ETV and shunt removal was then performed. **RESULTS:**In 24 cases (73.7%), shunt was successfully removed. On the other hand, 9 cases failed to remove shunts. In group A, 5 cases failed to remove shunts. Sagittal T2-weighted image in all failed cases in this group demonstrated CSF flow in their ETV stoma, indicating that they had problems in cisternal absorption of CSF. In group B, 4 cases were unsuccessful. Re-occlusion of the stoma was documented in one patient in group B.

**CONCLUSIONS:**Shunt removal with ETV is effective for patients with non-communicating hydrocephalus previously treated by VP shunt.

**Keywords:** hydrocephalus endoscopic third ventriculostomy

## FL-119

### Hydrocephalus

#### Endoscopic management of intraventricular shunt-related cystic compartment in pediatric patients

Ahmed Awad Zaher, Hatem Badr, Amr Farid

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**OBJECTIVE:**intraventricular cystic compartments occasionally develop after insertion of ventriculoperitoneal shunts which may continue to enlarge despite the functioning shunt system. Shunt infection with the resultant ventriculitis, direct ependymal trauma during shunt insertion, collapse of shunted ventricular compartment and over drainage of existing shunt are considered to be contributing factors that may stimulate the growth of shunt related compartments. Our goal is to create a free communication between the isolated cysts and the ventricular cavity to be drained by a single catheter with the aid of endoscopy

**MATERIAL-METHODS:**We retrospectively reviewed fourteen previously shunted children presented with shunt related cystic compartments who underwent nineteen endoscopic procedures at Neurosurgical Department of Mansoura University Hospital during the period from March 2007 to February 2016. Endoscopic fenestration of the cyst wall was done in all patients. Moreover, endoscopic third ventriculostomy was done in three patients, septum pellucidotomy in five patients and foraminoplasty in three patients. Clinical, radiological and surgical data were reviewed.

**RESULTS:**Our work included eight boys and six girls ranged in age between four months and eight years (mean=3.9 years). Endoscopic procedures succeeded to restore the communication between the cyst and the ventricular cavity in twelve patients(85.7%) with ten patients(71.4) continued to have a single shunt device and two patients (14.3) became shunt independent. The rate of shunt revision per year had been dropped from 2.98/year prior to endoscopic cyst fenestration to 0.27% after the previous endoscopic procedures during the mean follow up period of 52 months.

Five repeated endoscopic procedures were considered necessary in four patients and finally two patients (14.3%) required insertion of an additional shunt during our follow up period.

**CONCLUSIONS:**Cranial endoscopy is a useful operative alternative that control intraventricular cystic compartments related to previous shunt insertion.

**Keywords:** endoscopy-hydrocephalus-intraventricular cystic compartment

## FL-120

### Hydrocephalus

#### Endoscopic treatment of posthemorrhagic hydrocephalus in preterm infants

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<sup>1</sup>Department of Pediatric Neurosurgery, Severance Children's Hospital, Yonsei University College of Medicine, Seoul, Korea

<sup>2</sup>Department of Neurosurgery, Kangwon National University hospital

**OBJECTIVE:**Posthemorrhagic hydrocephalus is a significant problem arose from germinal matrix hemorrhage in preterm infants, complicating their neurologic outcome in long term. Early ventricular peritoneal shunt insertion is associated with high failure rate and multiple measures were carried out to control increased intracranial pressure while the infants become suitable for shunt placement such

as serial lumbar tapping, ventricular tapping, external ventricular drainage (EVD), ventriculostomy (VSG) and ventricular access devices (VADs). In our institute, endoscopic exploration with EVD was applied to preterm infants with posthemorrhagic hydrocephalus.

**MATERIAL-METHODS:** From January 2008 to December 2014, 82 patients underwent for endoscopic exploration of ventricle and among them 16 patients were preterm infants with GM IVH with hydrocephalus. Their charts and imaging studies were reviewed to see the outcome of patients and their shunt after endoscopic exploration and EVD.

**RESULTS:** There were 9 male and 7 female patients. They underwent 1st endoscopic exploration at age of 79.6 days after birth. They were born between intrauterine period of 23+6wks to 34+5wks on average 26+2wks. 57 explorations were done and only one case of infection was found (1.74%). 15 patients had ventriculoperitoneal shunt and one patient had two shunts inserted. One patient did not require for shunt insertion.

**CONCLUSIONS:** Endoscopic exploration and EVD is effective treatment to clear hemorrhagic debris and stabilize CSF constituents for shunt placement. This method with long subcutaneous tunneling of EVD catheter makes long term maintenance of EVD possible without infection and malfunction.

**Keywords:** prematurity, posthemorrhagic, hydrocephalus, endoscopy, germinal matrix

#### FL-121

##### Hydrocephalus

##### Efficacy of ETV in the treatment of hydrocephalus after removal of posterior fossa tumor in children

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Department of Pediatric Neurosurgery, "Mitera" Childrens Hospital, Athens, Greece

**OBJECTIVE:** The purpose of this study was to analyze the efficacy of Endoscopic Third Ventriculostomy (ETV) in the management of hydrocephalus after posterior fossa tumor surgery in children.

**MATERIAL-METHODS:** A retrospective review was performed of the management of hydrocephalus after 47 posterior fossa tumor resections in 42 patients performed by one surgeon between 2008-2016 (one patient had 4 operations and another two patients 2 operations in total, for recurrence). All patients had tumour excision as the first operation, regardless of the size of the ventricular system. Management of hydrocephalus was instituted after tumour excision when this became clinically necessary due to symptoms of intracranial hypertension in the presence of large ventricles, regardless of pseudomeningocele.

**RESULTS:** Hydrocephalus was present before tumour resection in 19 patients (41%). After tumour excision, 14 patients in total (30%) required permanent treatment for hydrocephalus, 4 of those did not have hydrocephalus at presentation, but developed after tumour resection (2 midline and 2 had laterally placed tumours). 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, and one of those developed frank CSF infection.  
**CONCLUSIONS:** 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.

**Keywords:** Posterior fossa tumour, hydrocephalus, Endoscopic Third Ventriculostomy, ventriculo-peritoneal shunt

Thursday, 27 October 2016

08:00 – 09:00

## Flash sessions 9: Hydrocephalus

#### FL-122

##### Hydrocephalus

##### Differences in cerebrospinal fluid motion in the ventricle and subarachnoid space

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**OBJECTIVE:** Cerebrospinal fluid (CSF) is thought to be mainly absorbed into the arachnoid granules and drained into the sagittal sinus. However, some observations conflict with this hypothesis; 1) There are no arachnoid granules in lower level animals and even in human fetuses at gestation before 32 weeks, 2) The mean pressure in the subarachnoid space is not high enough to push CSF into the venous sinus. In this study, we aimed to determine the movement of CSF and disclose the CSF movement regulation system in fetuses.

**MATERIAL-METHODS:** Fluorescent probes with differing molecular weights (MW) were injected into the lateral ventricle or subarachnoid space in mice with gestation of 13 days. The movements of the probes were monitored under fluorescent microscope.

**RESULTS:** Following intra-ventricle injection, the probes dispersed into the 3rd ventricle and aqueduct immediately, but did not move into the 4th ventricle and spinal canal. By compression of the brain, the probes moved into the spinal canal transiently and returned into the aqueduct. With an injection of low and high molecular weight (LMW, HMW) conjugated probes, both probes dispersed into the brain but only the LMW probe dispersed into the whole body within 15 minutes. Following intra-subarachnoid injection, the LMW probe moved faster compared with the HMW probe. The probes diffused into the spinal canal gradually. Neither probes dispersed into the brain and body.

**CONCLUSIONS:** 1) CSF does not appear to circulate. At the occasion of high intracranial pressure, ventricle CSF may move into the spinal canal rapidly. 2) The movement of metabolites in CSF might be restricted by MW. 3) CSF in the lateral ventricle and CSF in the subarachnoid space move differently. This new information will contribute to the treatment of hydrocephalus, and the development of medicines for actively transferring brain metabolites that could prevent neurodegenerative disease and brain aging.

**Keywords:** cerebrospinal fluid, ventricle, subarachnoid space, hydrocephalus, molecular weight

#### FL-123

##### Hydrocephalus

##### Clinical and radiological analysis of macrocrania

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<sup>3</sup>Department of Neurosurgery, Kyoto Prefectural University of Medicine, Kyoto, Japan

**OBJECTIVE:**Macrocrania in infants can be caused by or associated with hydrocephalus, a benign extracerebral fluid collection, a subdural effusion, familial macrocephaly, syndromic macrocephaly, and other clinical conditions. This study was conducted to analyze the clinical and radiological characteristics of macrocrania.

**MATERIAL-METHODS:**We retrospectively screened 42 cases diagnosed with macrocrania (a head circumference in the 98th percentile or above). Cases of internal hydrocephalus and traumatic subdural hematomas were excluded. Cases were divided based on the size of the subarachnoid space on CT or MRI, into group A (6 mm) or group B (under 6 mm). We reviewed the clinical findings and developmental states.

**RESULTS:**In group A (12 cases), no surgery was required, and the average developmental quotient (DQ) was 88.9 (60–109). In group B (30 cases), four cases were associated with polymicrogyria, hemangioma, polydactyly, ventriculomegaly, and/or cerebellar tonsillar herniation (CTH), which are characteristics of megalencephaly capillary malformation (MCAP). Three of the four infants with MCAP required surgery such as foramen magnum decompression or a ventriculo-peritoneal shunt; the one infant who was not treated surgically died suddenly at 5 months of age due to CTH. The average DQ in the four cases with MCAP was 53.5 (50–57); the remaining 26 cases in group B had an average DQ of 93 (63–108).

**CONCLUSIONS:**Because MCAP has a poor prognosis and sometimes requires surgery, we should differentiate MCAP from other, less dangerous conditions. Radiological findings are important for a precise diagnosis of macrocrania.

**Keywords:** Macrocrania, Megalencephaly capillary malformation, Subarachnoid spaces, MRI

#### FL-124

##### Other

#### Temporal arachnoidal cysts: 15 years of neuroendoscopic experience

Sergio R Valenzuela

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**OBJECTIVE:**>Neuroendoscopic treatment of arachnoidal cysts has proved to be an option among others for the management of this pathology showing favorable outcomes. The objective of the current presentation is to report our personal experience in children under 15 years old.

**MATERIAL-METHODS:**This is a retrospective chart review of pediatric patients under 15 years old undergoing Neuroendoscopic fenestration of temporal arachnoidal cysts. A total of 90 children were included harbouring intracranial arachnoidal temporal cysts out of a total series of Intracranial cysts of 125 cases. Outcome is analyzed according with the need of a secondary procedure and clinical evolution on the long run with an average of 8 years follow up being the oldest case of 15 years as and. Video recordings were reviewed as well as CT scanners and brain MRI to achieve and to classify results.

**RESULTS:**Good results (93 %) were considered as long as no other procedure was needed to treat the patient ( 7 %).

Regarding Cyst size our series showed a complete reduction only in 10 %, partial reduction in 65 % and no reduction in 25 %.

No mortality was observed and Morbidity was mainly CSF leaks in 10 cases, wound infection in 4, Subdural hygroma in 3, III nerve palsy in 2 cases.

**CONCLUSIONS:**Neuroendoscopic fenestration of Arachnoidal temporal cysts in our center has proven to be a successful therapy. Proper selection of patients, Operator experience, tools quality, diagnostic equipment and careful surgical technique are the base of good results comparable with other techniques

**Keywords:** neuroendoscopy, arachnoidal, cysts, fenestration, c.s.f. leak

#### FL-125

##### Hydrocephalus

#### Midline intracranial arachnoid cyst in children: what is the best treatment?

Essam A Elgamal

Sheikh Khalifa Medical City

**OBJECTIVE:**Midline intracranial arachnoid cysts are rare. While may be asymptomatic, some cause symptoms mostly related to hydrocephalus and warrant surgical treatment. In this retrospective review, the author aimed to study different methods of treatment of symptomatic cases and the outcome after surgical intervention in children with midline intracranial arachnoid cysts. **MATERIAL-METHODS:**Retrospective chart review of 18 consecutive, prospective cases presented with midline intracranial arachnoid cyst. Clinical presentation and radiological findings were analyzed, and methods of treatment of symptomatic cases and the outcome of surgical intervention were studied.

**RESULTS:**Eighteen children, (11 male & 7 female) presented with midline intracranial arachnoid cyst, 8 of them were diagnosed antenatally. Follow up period ranged between 2 months and 5 years (mean 2.37 years).

Hydrocephalus was present in (n. 12), seizures (n. 2), papilloedema (n. 1), sixth nerve palsy (n.1), motor weakness/ataxia (n. 4), and delayed menarche (n. 1). Arachnoid cyst was suprasellar (n. 7), quadrigeminal (n. 6), supracerebellar (n. 2), retrocerebellar (n. 3), and 4 patients had large cyst extending between quadrigeminal cistern and posterior fossa.

Twelve patients underwent 16 endoscopic procedures (cyst fenestration, ETV, and septostomy), temporary EVD after endoscopy (n. 4). VP shunt (n. 5), Ventriculo-cysto-peritoneal shunt (n. 3), and posterior fossa decompression (n. 1).

Complications of treatment included hemorrhage (n. 2), CSF leak (n.1), seizures (n.2), temporary respiratory failure (n.2), and shunt malfunction (n. 5). Outcome was considered good (n. 10), fair (n. 7), and one patient had near fatal hemorrhage during endoscopy and recovered in a vegetative state.

**CONCLUSIONS:**The natural history of midline intracranial arachnoid cyst is not well understood. There is no standard treatment that agreed upon. There is no association between radiological size of the cyst, and clinical presentation / clinical improvement after treatment.

**Keywords:** Midline intracranial arachnoid cyst, Hydrocephalus, Endoscopic fenestration, ETV, outcome

#### FL-126

##### Trauma

#### Sport-related structural brain injury associated with arachnoid cysts in the pediatric population: a systematic review and quantitative analysis

Scott Zuckerman, Colin Prather, Aaron Yengo Kahn, Gary Solomon, Allen Sills, Christopher Bonfield

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**OBJECTIVE:**It is postulated that children with arachnoid cysts (AC) demonstrate higher rates of structural brain injury (subdural hematoma, hygroma, hemorrhage) after trauma, including while playing sports. Given the potential neurologic consequences of a structural brain injury requiring neurosurgical intervention, we performed a systematic review of pediatric sport-related structural-brain injury associated with AC with a corresponding quantitative analysis.

**MATERIAL-METHODS:** Titles and abstracts were systematically searched for studies that reported a structural brain injury due to a sport or recreational activity with associated AC. Descriptive statistics were summarized. Univariate logistic regression assessed for predictors of neurologic deficit, open craniotomy, and cysto-peritoneal shunt.

**RESULTS:** Forty-one studies reported 49 cases of sport-related structural brain associated with AC in children. Median age at presentation was 14 years. Soccer (27%) and football (12%) were the most common sports. Nearly all (90%) presented greater than 7 days after the injury. Headache was the most common presenting symptom (98%), followed by nausea/vomiting (50%). One-quarter presented with a neurologic deficit, most commonly hemiparesis. Open craniotomy and burr holes were performed in 53% and 37% respectively. Seven patients (14%) received a cysto-peritoneal shunt. No significant predictors were found for neurologic deficit or open craniotomy. However, the odds of receiving a cysto-peritoneal shunt decreased as age increased ( $p=0.004$ ). All reported outcomes were good.

**CONCLUSIONS:** Pediatric sport-related structural brain injuries associated with AC are rare. The majority of cases presented with chronic symptoms, most commonly headache, and recovery was good. Injuries occur in team/non-team and contact/non-contact sports. Most are treated with open craniotomy or burr hole. We did not find a contraindication to participate in sports in patients with an AC, although parents and children should be counseled appropriately. Further studies are necessary to evaluate AC characteristics that could pose a higher risk of adverse events after trauma.

**Keywords:** sports concussion, traumatic brain injury, arachnoid cyst, subdural hemorrhage, return to play

#### FL-127

##### Hydrocephalus

##### Efficacy and safety of endoscopic third ventriculostomy and choroid plexus cauterization for hydrocephalus: a systematic review and meta-analysis

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**OBJECTIVE:** Infantile hydrocephalus is a significant neurosurgical issue, and endoscopic third ventriculostomy / choroid plexus cauterization (ETV/CPC) has gained popularity in its treatment over the past decade. The objective was to perform a systematic review meta-analysis to determine the efficacy and safety of ETV/CPC, and to compare the procedural outcomes between North American and sub-Saharan African cohorts.

**MATERIAL-METHODS:** Systematic review was performed using 4 electronic databases and bibliographies of relevant articles, with no language or date restrictions. Cohort studies of participants undergoing

ETV/CPC that reported outcome were included using PRISMA guidelines. The outcome was time to repeat CSF diversion or death. Forest plots were created for pooled mean and its 95% CI of outcome and morbidity.

**RESULTS:** Of 78 citations, 11 retrospective reviews (with 524 participants total) were eligible. Efficacy was achieved in 63% participants at follow-up periods between 6 months and 8 years. Adverse events and mortality was reported in 3.7% and 0.4% of participants, respectively. Publication bias was detected with respect to efficacy and morbidity of the procedure. The efficacy of ETV/CPC in 6 studies from sub-Saharan Africa was 71%, compared to 49% in the 3 studies reported in North America.

**CONCLUSIONS:** In the literature, the reported success of ETV/CPC for infantile hydrocephalus is higher in sub-Saharan Africa than developed nations. These efficacy rates are likely an overestimation and the low morbidity rate is likely an underestimation. Large long-term prospective multicenter observational studies addressing patient-important outcomes are required to further evaluate the efficacy and safety of this re-emerging procedure.

**Keywords:** hydrocephalus, endoscopic third ventriculostomy, choroid plexus cauterization

#### FL-128

##### Hydrocephalus

##### External ventricular drainage: indications and outcome among Sudanese children

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National Center for Neurological Sciences, Department of Neurosurgery, Khartoum, Sudan

**OBJECTIVE:** In this study we would like to reflect our experience in managing Sudanese children with different cranial conditions through EVD; indications, pathologies faced and early outcome.

**MATERIAL-METHODS:** Prospective review of cases who have been operated at the National Center for Neurological Sciences during the period from February 2014 to February 2016. The patients were closely assessed till the EVD removal and patient discharge. All patients with deficient operative or post-operative data or those operated outside the center or adult patients who underwent EVD operation were excluded from the study.

**RESULTS:** Forty-one cases were operated aging between 6 days to 7 years. Most of them were below one year of age. Male to female 2:1. The majority of the cases have posterior fossa tumor with obstructive hydrocephalus ( $n=19$ , 46.3%). Twenty patients (48.8%) presented with decreased level of consciousness while 28 patients presented with symptoms and signs of raised ICP (68.3%). The decision for EVD made preoperatively based on positive CT/MRI findings in 10 patients (24.4%) while in the rest the decision for EVD was made intra-operatively for the CSF being unclear. Most patients ( $n=28$ , 68.3%) respond to single injectable antibiotic therapy. The average duration for antibiotics use was 22 days approximately. Complications were encountered in 13 patients. Most patients improved or cured ( $n=25$ , 61%) 5 deteriorated and 11 died

**CONCLUSIONS:** EVD can be used for many indications including obstructive, post-infectious, post-meningitic hydrocephalus and IVH. Most patients may present with either deteriorated level of consciousness or symptoms and signs of raised ICP but few of them may have positive brain imaging findings and therefore in most of them the decision for EVD may be done intra-operatively. The average duration for EVD use is 3 weeks with single antibiotic therapy use which was found as effective as combined and intraventricular therapy.

**Keywords:** External ventricular drainage, indications, outcome

## FL-129

**Special topic: Neuro-imaging****Surgical site infection and intra-operative magnetic resonance imaging: a single centre experience**

Santosh Mohan Rao Kanangi<sup>1</sup>, Dawn Williams<sup>1</sup>, Linda Marshall<sup>1</sup>, Stephan Paulus<sup>2</sup>, Richard Cooke<sup>3</sup>, Christopher Parks<sup>1</sup>, Sasha C Burn<sup>1</sup>, Ajay Kumar Sinha<sup>1</sup>, Conor Mallucci<sup>1</sup>, Benedetta Pettorini<sup>1</sup>

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**OBJECTIVE:**The use of Intraoperative magnetic resonance imaging (IoMRI) has recently dramatically expanded. Due to the nature of its design, it involves shifting the patient whilst under anaesthesia and with an open craniotomy to the scanner. Due to this, there is a potential higher risk for surgical site infections. The aim of this paper is to assess if the use of io-MRI is related to increased risk of surgical site infections.

**MATERIAL-METHODS:**In this retrospective study, we analysed data of patients operated on in the period between 2009-2015 using the ioMRI. The patients were identified by our neurosurgical database and cross-checked with the radiology database. Surgical Site Infections (SSI) were identified by a 30 days round check and medical notes. SSI were defined based on Public Health of England classification.

**RESULTS:**We used IoMRI in 196 patients in a 6 years period.

We showed low rate of SSI (1.5%) with 3 patients with superficial site infection which were treated with antibiotics. The pathogens were Staphylococcus Epidermidis in all cases.

SSI rate in standard craniotomies for tumours in literature is 2%.

The risk of surgical site infection is not increased by the use of ioMRI.

We also shed light on the technique used in Alder Hey for closing the patient prior to shifting to the ioMRI. The low rates of infection in our ioMRI is proof that not only is our preparation technique safe but also goes on to show that there is no added risk of infection whilst using the ioMRI. The infection rates in other intra-operative MRI data series were compared with our study.

**CONCLUSIONS:**Intra-operative MRI is safe and there is no added infection risk with the use of ioMRI.

**Keywords:** intra-operative MRI (ioMRI), craniotomy, open wound, infection

## FL-130

**Craniofacial****Single dose versus multiple dose perioperative prophylactic antibiotics in post-operative wound infection in pediatric cranial procedures with implanted skull materials: a single center clinical trial**

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**OBJECTIVE:**No evidence-based protocol is available for the administration of perioperative prophylactic antibiotics in children undergoing cranioplasty or cranial remodeling with implanted materials. The purpose of this study is to evaluate the effectiveness of single versus multiple dose prophylactic antibiotics on the prevention of wound infection following cranial surgeries using mini-plates, plates and screws.

**MATERIAL-METHODS:**Between March 2014 and March 2016, 52 patients who underwent cranioplasty or cranial remodeling using

implanted materials were randomly allocated to one of the intervention groups; Group A: those who received a single dose of 30 mg/kg cefazolin just 30 minutes before surgery; Group B: those who also received extra doses for the next 24 hours. The wounds were assessed on the 1st, 3rd, and 14th days after surgery, and any wound erythema, discharge, dehiscence, and high grade fever (38.5<) with signs of sepsis were considered as failure. Children with history of immunodeficiency, congenital heart disease, or those who needed antibiotics for other reasons were excluded from the study.

**RESULTS:**A total number of 52 patients have been enrolled in this trial [Group A: 29 (55.8%); Group B: 23 (44.2%)]. This is the preliminary report and the 5 trial is open. There were 24 girls and 28 boys, with the age ranging between 4 and 56 months. The procedures consisted of 4 cranioplasty, 8 syndromic craniosynostosis, and 40 non-syndromic craniosynostosis surgeries. The failure rates were as follow; 3 wound superficial discharge (5.7%), and 1 erythema plus fluctuation (1.9%), all were successfully managed with oral antibiotic without hospitalization. There was no significant difference between two intervention groups in terms of wound infections.

**CONCLUSIONS:**The current study demonstrates that a single dose administration of preoperative antibiotic can be effective to prevent wound infection in pediatric patients who undergo cranial surgeries with implanted materials in skull.

**Keywords:** Cranial remodeling, Cranioplasty, Implanted Materials, Prophylactic antibiotics

## FL-131

**Other****Acute cerebellitis: what a paediatric neurosurgeon should know**

John Amaechi Emelifeonwu

Department of Neurosurgery, Western General Hospital, Edinburgh

**OBJECTIVE:**To provide an overview of Acute Cerebellitis (AC) and its relevance to paediatric neurosurgeons

**MATERIAL-METHODS:**A Literature review of all cases of AC in the post-MRI era. The PubMed and Ovid database were searched to determine all cases of AC in the paediatric population. Search terms included "acute cerebellitis" AND "pediatrics". Generated abstracts were reviewed and only cases in which MRI had been used as part of the investigations of AC were selected.

**RESULTS:**A total of 124 cases of AC have been reported in the post-MRI era. Most causes are idiopathic or of unknown aetiology. Most cases are managed medically and have a benign course but a handful of cases run a 'fulminant' course, requiring surgical intervention such as CSF diversion or posterior fossa decompression.

**CONCLUSIONS:**'Fulminant' AC is rare but difficult to predict. We therefore propose that cases of AC should be managed in tertiary neurological centres with access to Paediatric Neurosurgery as emergency surgical interventions may be required as part of the treatment. A prospective study is needed to determine the true incidence of fulminant AC.

**Keywords:** Acute cerebellitis, CSF diversion, Posterior fossa decompression

## FL-132

**Hydrocephalus****How evolution informs the fate of ancient inhibitory interneurons, in relationship to the cerebrospinal fluid contacting neurons, in the human neocortex**

Alexandra R Kunz

Harvard University (Extension) student, Cambridge, MA, USA

**OBJECTIVE:**600 Mya marks an evolutionary milestone: 1st internal fluid brain-tissue environment: the chordate lancelet's hindbrain inhibitory interneurons (INS) in direct contact with cerebrospinal fluid-contacting (CSF-c) neurons, whose cilia transduced diffusible growth-promoting, non-synaptic signals to INS' progenitor cell bodies. In time with evolutionary increases of INS in phylogenetically differentiated vertebrates, the CSF-c neurons migrated from the ventricles, communicating now synaptically, their cilia extending into intracellular fluid.

**MATERIAL-METHODS:**This paper explores the ontological fate of evolutionary ancient interneurons/their significance for human neocortex functioning.

**RESULTS:**Energy metabolism sets humans apart from primates: evolutionary increases in synaptic signaling/connectivity, quadrupled glial cells, an unexpected 46% greater glial:neuron density,  $p < 0.001$ . INS' energy efficiency exceeds excitatory neurons': 85% energy consumption associated with excitatory glutamate recycling, using both glycolytic/glycogenolytic processes, only glycolytic ATP for INS' synaptic-cleft recycling.

Key in evolution's INS' origins is recruitment of other mechanisms of greater number/diversity for primates'/human's neocortex: primitive vertebrate lampreys' (450Mya) INS' circuits devoid of sense organs/pallium/ geniculate eminence (GE); vertebrate gnathostomes' (350Mya) INS' tangential migration from GE to pallium highly conserved; INS' competence to enter neocortex subventricular zone (SVZ) established in amniotes (310Mya); competence to enter cortical plate (CP) from GE, mammalian unique (185-210Mya). 40Mya primates' INS' number/diversity/complexity increased more than excitatory neurons': a pre-existing developmental mechanism's boosting, a bipartite process: INS' lateral ventricular neuroepithelium progenitors migrating radially. Relaxed phylogenetic brain/body constraints to a behavioral evolutionary shift was the adaptive force for anthropoid primates' social acumen. An extrinsic supply neuromodulators for behavioral flexibility, dopamine(DA), acetylcholine(ACh), serotonin(5-HT) with slower/longer neuromodulation, altered INS' terminal neocortical axon patterns; humans'/chimps' axonal density increased as DA/5-HT/ACh "coils"/"clusters" for plasticity; subtle human evolutionary shift favored cortex layers V/VI's increased innervation,  $p < 0.05$ . **CONCLUSIONS:**Evolutionary ancient INS were vitally important to brain function 600 Mya; their legacy today ontologically as neocortical INS help define our preeminent human identity.

**Keywords:** evolution inhibitory neurons cerebrospinal fluid-contacting neurons energy efficiency bipartite process social acumen

#### FL-133

##### Hydrocephalus

##### Pattern of congenital craniospinal anomalies among neurosurgical patients in a Nigerian tertiary hospital

Hammed Abiola Oshola, Taopheeq Bamidele Rabiu

Division of Neurological Surgery, Department of Surgery, Ladoke Akintola University of Technology Teaching Hospital, Osogbo, Nigeria.

**OBJECTIVE:** This study evaluates the patterns of various craniospinal anomalies and their possible risk factors in a new neurosurgical centre in Nigeria

**MATERIAL-METHODS:** This prospective study included all patients with congenital craniospinal anomalies managed at the LAUTECH Teaching Hospital Osogbo from January 2013- December 2015. Frequency and pattern of these anomalies were recorded. All patients were assessed for other associated anomalies. Data related to age, gender, type of anomaly, location and maternal details including social economic class, febrile illness in pregnancy and drug use in pregnancy were recorded on a pre-designed proforma. Cranial CT/MRI and

TFUSS were obtained according to the type of the lesion. Surgical interventions were performed as required.

**RESULTS:**Nineteen patients with congenital craniospinal anomalies were seen during the study period. Ten of the cases were males (52.6%). The most common anomaly observed was hydrocephalus (n:10, 68.4%) which was either in isolation (n:7,70%) or combined with spina bifida(n:3,30%), followed by spina bifida which was found in 5 patients (36.8%). Of the spina bifida cases, myelomeningocele accounts for 4 (80%). Encephalocele was seen in 3 (17.6%) and all were occipital. There was a case of craniosynostosis. Majority of patients with hydrocephalus had VP shunt (n:7, 70%) while 1(10%) had ETV. Surgical repair was performed for the neural tube defects. None of the mothers had periconceptional folic acid supplementation, three (15.8%) of them had febrile illness in pregnancy while 1 (5.3%) had exposure to irradiation. Four (21.1%) of the mothers used anti-malarial early in pregnancy. None had family history of CNS anomalies. Talipes equinovarus constituted 50% of the associated anomalies. **CONCLUSIONS:**Congenital Craniospinal anomalies represent an important group of neurosurgical conditions seen at our centre. Public awareness of periconceptional use of folic acid to reduce the risk of mothers having babies with congenital CNS anomalies need to be promoted.

**Keywords:** hydrocephalus, risk factors, spina bifida, nigeria

#### FL-134

##### Vascular

##### Gamma knife for paediatric patients less than 12 years: the AIIMS experience

Deepak Agrawal, Chetna Banga, Renu Saini, Manmohan Singh, Rajinder Kumar, Shashank S Kale, Bhawani S Sharma

All India Institute of Medical Sciences, New Delhi, India

**OBJECTIVE:**Radiation therapy may have deleterious effects on the developing brain. However stereotactic radiosurgery provides a high level of protection to normal brain and can be given in children as small as 4 years for vascular malformations as well as variety of neoplastic lesions. The objective of this study was to review our experience of Gamma Knife radiosurgery in children less than 12 years of age

**MATERIAL-METHODS:**In this retrospective chart review over 4 years (Jan 2011- September 2015), records of children ( $\leq 12$  years) who underwent gamma knife radiosurgery were reviewed to see for demographic data, diagnosis and whether gamma knife was done in local or general anaesthesia.

**RESULTS:**A total of 41 children underwent gamma knife radiosurgery in the study period. The mean age was 9.5 (range 4-11) with the male: female ratio of 2.4:1. The most common indication was AVM (n= 26) followed by tumours (n=15). All children except two were done under local anaesthesia. Mean follow up was 4.3 years (1- 8.2 years). 82% of all AVM had obliteration and 8 tumor control rate was 89% at last follow up

**CONCLUSIONS:**Gamma Knife radiosurgery offers a compelling alternative to standard radiation therapy in children and may be especially useful in vascular malformations like AVM's which are not amenable to surgery or embolization. In addition as our experience shows that more than 95% of children can be given Gamma Knife under local anaesthesia. Excellent AVM obliteration rate & tumor control rate were achieved in our study

**Keywords:** Gamma Knife, Pediatric, AVM, Tumor, Outcome, Obliteration rate

## FL-135

## Trauma

## The use of Custombone for repair of cranial defects in children

Carmine Mottolese, P. A. Beuriat, A. Szathmari, B. Grassiot, Federico Di Rocco

Hôpital femme Mère Enfant, Lyon France

**OBJECTIVE:** Repair of cranial defects can be difficult in children mainly if cranial defects are of large surface for the potential risks of hemorrhagic complications and risks of sequels.

The use of foreign material became important and we report our experience with synthetic hydroxyapatite to repair cranial defects.

**MATERIAL-METHODS:** From 2006 to 2013 we have operated 19 patients for a cranial plasty. The age of these patients varies from 8 months, the more young, to 13 years the oldest, the median age is 6 years and two months.

Three patient were operated because the bone flap was invaded by tumoral tissue, two patients presented a bacterial osteitis, 14 patients were operated after a decompressive craniectomy.

All patients have been submitted to a CT-scan with 3D reconstruction.

Three patients have been operated in age inferior to three years.

All patients have been followed in outpatient consultation.

**RESULTS:** The plasty was well positioned in all patients. The esthetical result was judged acceptable in all patients also in the fronto-orbital location.

The CT-scan showed the presence of bone tissue invading the hydroxyapatite after 13 months (6–22 months).

Only a patient presented a fracture of the plasty after a cranial trauma.

No infectious complications have been reported

**CONCLUSIONS:** The synthetic hydroxyapatite can be used for cranial plasty in children because it has good proprieties of osteoinduction and osteocondensation.

This plasty can be used in patients under the age of two years.

The problem can be the prize but the good esthetical results reducing the number of rehospitalization for good results justifies its utilization.

**Keywords:** Pediatric Neurosurgery

## FL-136

## Other

## Should I spend my summer doing research? A quantitative and qualitative study evaluating student achievement in a surgical summer research program

Ash Singhal, Jennifer Liang

British Columbia Children's Hospital; University of British Columbia, Vancouver, Canada

**OBJECTIVE:** Over the last few decades, the documented decline in the number of physician-scientists has been an ongoing concern for health services and universities. Although studies have suggested the utility of exposing students to research, few have examined the tangible long-term benefits of this exposure and answered questions regarding future research interest or research output. We sought to explore the value of a surgical summer student research program.

**MATERIAL-METHODS:** We identified all students who participated in a surgical summer student research program (SSRP) over a 10 year period. Semi-structured phone interviews to further explore the SSRP experience were conducted. A separate online survey was sent out to research supervisors who provided mentorship to students in the SSRP over the same time period.

**RESULTS:** 89 students were successfully surveyed. Seventy eight (88%) students indicated that they were satisfied/very satisfied with the SSRP. Seventy

one (80%) participants have had an abstract published and 53 (60%) have had a paper published. Fifty four (61%) participants are currently involved in research while 40 (45%) indicated an interest in incorporating research into their future career. Analysis of the 12 qualitative interviews showed a saturation of themes after 4 interviews. Prominent themes included career exploration opportunities and clinical research exposure. Seventeen (89%) supervisors were satisfied/very satisfied with the SSRP. In addition, 16 (85%) supervisors indicated that students helped them achieve their research goals and contributed to their own learning. **CONCLUSIONS:** Students and supervisors both indicated benefits from the SSRP in their careers. Early saturation of themes from the semi-structured interviews suggests that participants shared similar views about their experience. The large majority of participants who participated in the SSRP currently have an abstract and/or a paper published and over half continue to be involved in research, suggesting a great value for summer surgical research exposure.

**Keywords:** Education, Research, Medical Student, Qualitative Analysis

## NURSING SYMPOSIUM ABSTRACTS

Monday, 24 October 2016

11:00 – 12:20

## Nursing Symposium - I

## NS-001

## Articulating the paediatric neurosurgical nursing practice model of care at CURE children's hospital of uganda: challenging current thought with emerging evidence

Lydia Nabuduwa Ssenyonga<sup>1</sup>, Angela Louise Leonard<sup>2</sup>, Minette Coetzee<sup>2</sup>

<sup>1</sup>CURE Children's Hospital of Uganda, Mbale, Uganda

<sup>2</sup>Department of Paediatrics and Child Health, University of Cape Town, Cape Town, South Africa

**OBJECTIVE:** To articulate the clinical practice model of care at CURE Children's Hospital of Uganda (CCHU) by describing the care pathway from admission until discharge. To gain a detailed understanding of who provides care and what care is provided at CCHU. This specialist paediatric neurosurgical teaching hospital provides treatment, support and care for children with hydrocephalus, neural tube defects and brain tumours.

**MATERIAL-METHODS:** The intentionally participative methodology used the lens of appreciative inquiry approach and qualitative methods. Data was collected through focus groups, interviews, participant observations, unit and bed space layout. It included how a child moves through the hospital; who they see; what happens along each part of the neurosurgical pathway; communication and how parents are involved in care. Graphic facilitation, a visual tracking method captured multidisciplinary staff discussion and contributions to the neurosurgical pathway during the focus groups.

**RESULTS:** Tracking and exploring the neurosurgical pathway allowed staff and researchers to articulate the paediatric neurosurgical nursing practice model of care. Core to this care is that mothers are admitted as the primary patient. The mother maintains the fundamental care of her child throughout her stay. The pathway showed that all staff direct their care of the child through the mother.

**CONCLUSIONS:** While this approach may seem like a necessity in the scarce nursing resources of Uganda and other resource constraint countries, the emerging neuroscience evidence confirms that the presence of the mother determines physiological stability of the child. This presentation will include data from Uganda and South Africa to confirm the value of zero separation of mothers.

**Keywords:** neurosurgical nursing practice model of care, care pathway, graphic facilitation, mothers, zero separation

## NS-002

**Gaining service users perspectives to develop the training and education of nurses on a neurosurgical ward**

Nicola Wilson, Francesca White

Neurosciences Department, Great Ormond Street Hospital, London, United Kingdom

**OBJECTIVE:** In the United Kingdom there are professional standards set by the Nursing and Midwifery Council for undergraduate education; however, when it comes to postgraduate nurses there is less guidance on how to support their continued professional development. We are told that we must ensure we have the right people, with the right skills, in the right place at the right time, and the identification of these people and the methods of developing their skills is often decided by professionals (Cummings, 2013). In recent years there has been a move to listening to, and working in partnership with, service users so as to improve and transform services (Needham, 2012). This presentation will highlight how the views of service users can be used to develop paediatric neuroscience nurses' knowledge, skills and attitudes.

**MATERIAL-METHODS:** A qualitative survey of children, young people and parents' will be used to identify their perspectives on neurosurgical nursing

**RESULTS:** A thematic approach will then be used to present the results of the children, young people and parents' perspectives.

**CONCLUSIONS:** This presentation will conclude by outlining how these themes have helped us adapt and shape our educational strategy to ensure that the child and family's needs remain at the centre of all our nursing care. Examples from both individual teaching sessions and the shaping of nurses' education as a whole within our Neurosurgical Unit will be examined.

**Keywords:** Nursing Education Service Users

## NS-003

**A multi-disciplinary prenatal support team**

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**OBJECTIVE:** Although the prenatal diagnosis of CNS malformations has improved due to advances in diagnostic technologies, there is no sufficiently established system for supporting and caring for the family after the diagnosis is made.

**MATERIAL-METHODS:** To address this issue, we formed a multi-discipline prenatal support team (MDPST) at our hospital in 2012. The team includes obstetricians, pediatricians, neonatologists, pediatric neurosurgeons, nurses and midwives from an outpatient clinic, staff members from a neonatal intensive care unit (NICU), a growing care unit (GCU), and a maternal fetal intensive care unit (MFICU), physical therapists, clinical psychotherapists, medical social workers, and secretarial staff from the various medical departments involved. The team meets every two weeks. To date, we have provided support for 126 families. I was appointed to the MDPST as the first full-time perinatal coordinator in Japan. The coordinator guides and informs the parents, coordinates the related staffs, and manages the MDPST meetings.

**RESULTS:** I will describe cases that involved preparing for treatment after delivery, cases that required selective or palliative treatment

while withholding or withdrawing life-prolonging medical treatment, and cases that involved family support through decisions regarding the termination of pregnancy (TOP).

**CONCLUSIONS:** I described cases that involved preparing for treatment after delivery supported in MDPST.

**Keywords:** family support

## NS-004

**Treatment of intraventricular hemorrhage and post hemorrhagic hydrocephalus in preterm infants**

Angela Forbes

Seattle Children's Hospital, Seattle, Washington, USA

**OBJECTIVE:** Premature infants, specifically those born at a very low birth weight (<1000 kg) are at a very high risk of bleeding of the germinal matrix of the brain, resulting in intraventricular hemorrhage (IVH). IVH is the most frequent and severe neurological complication of premature birth. Approximately 25-50% of infants with IVH go to develop posthemorrhagic hydrocephalus (PHH). IVH and PHH are known to have considerable neurological sequelae, including cognitive delays, visual impairment, behavioral problems, epilepsy and cerebral palsy.

**MATERIAL-METHODS:** Oral presentation

**RESULTS:** There is no standard of care or management of IVH/PHH and the risks and benefits of each intervention.

**CONCLUSIONS:** We will discuss the history of treatment of IVH and PHH and review the current evidence regarding with risks and benefits of current surgical options. We will discuss timing and types of surgical intervention. We will also discuss pre-operative and post-operative management of premature infants with IVH/PHH. We will discuss management and care of subgaleal reservoirs/shunts, including indications and procedure for subgaleal reservoir tap. Through 3 specific case studies patients will be reviewed to include clinical presentation, imaging, treatment and outcomes. We will then summarize current nursing management based on the current evidence.

**Keywords:** Hydrocephalus, nursing, management, intraventricular hemorrhage, post hemorrhagic hydrocephalus

## NS-005

**Physical therapy for neonates with myelomeningocele**

Takahito Iitsuka<sup>1</sup>, Masato Yamashita<sup>1</sup>, Atsushi Keyaki<sup>2</sup>, Daisuke Sakamoto<sup>3</sup>, Atsuko Harada<sup>3</sup>, Mami Yamasaki<sup>3</sup>

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<sup>3</sup>Department of Pediatric Neurosurgery General Hospital, Osaka, Japan

**OBJECTIVE:** The purposes of physical therapy for neonates are to stabilize respiratory and cardiovascular conditions, to protect the infant from stresses, and to promote normal development. Physical therapy programs evaluate development, maintain an ideal posture and physiological posture, improve joint movement, and provide respiratory support and breastfeeding exercise. We here report cases in which neonates were treated by physical therapy after undergoing surgery for myelomeningocele at our hospital.

**MATERIAL-METHODS:** Between April 2013 and February 2016, 14 neonates (seven male and seven female) were diagnosed with myelomeningocele by prenatal MRI and underwent surgical myelomeningocele repair immediately after cesarean delivery at Takatsuki General Hospital. The mean gestational age at birth was 37.3 weeks  $\pm$  5.6 days; the mean body weight was 2453.8  $\pm$  249.6 g (2030–2872 g). Hydrocephalus was present in 10 cases. Physical therapy was begun 8.4  $\pm$  8.3 (2–8) days after birth. Motor development was evaluated by the Dubowitz neurological examination. A prone position was enforced immediately after surgery to prevent surgical-wound

problems, with periodic changes to a lateral position. When the restriction to a prone position was lifted, aggressive physical therapy began with a focus on good limb posture, rolling over, and passive joint and limb movement. Self-motion was induced by sensory stimulation.

**RESULTS:** Position-correcting physical therapy is reported to prevent limb deformities and joint contracture. In the present study, range of joint motion improved in four of 10 cases with lower-leg deformities and limited joint motion. None of the cases had any deterioration of general condition or motor function. Handling techniques to maintain the neonate's head and trunk alignment also promoted normal development.

**CONCLUSIONS:** Early intervention through physical therapy can be safe and effective after the surgical repair of myelomeningocele in neonates.

**Keywords:** Physical therapy, myelomeningocele, neonates

#### NS-006

##### **Is there concordance between child and parent ratings of health related quality of life reports, in children with spinal dysraphism?**

Lindy May<sup>1</sup>, Adam Kuczynski<sup>2</sup>, Jo Wray<sup>2</sup>

<sup>1</sup>Department of Neurosurgery, Gt Ormond St Hospital, London, UK

<sup>2</sup>Department of Psychology, GT Ormond ST Hospital, London, UK

**OBJECTIVE:** To assess the level of concordance in Health Related Quality of Life (HRQL) reports between the child with spinal dysraphism and their parents

**MATERIAL-METHODS:** Firstly, the HRQL of 54 children with lumbosacral lipoma and their parents was assessed using standardised questionnaires. Secondly, a literature review was undertaken to obtain data regarding the HRQL of children with spina bifida aperta and their parents.

**RESULTS:** Self-report HRQL was higher than the proxy report particularly with regard to psychosocial, social and total scores in children with lumbosacral lipoma. Assessing the HRQL of children with spina bifida aperta is more difficult due to the complexity of the condition and the neurocognitive level of the child. However, many publications suggest that self-report of HRQL is higher than proxy report, particularly with regard to physical and emotional scores (Sawin and Bellin, 2010)

**CONCLUSIONS:** Further research is required to understand the HRQL of children with spinal dysraphism in order to evaluate inequalities in health care provision and plan resources accordingly

**Keywords:** Spinal dysraphism, Health Related Quality of Life.

#### NS-007

##### **Non-germinoma germ cell tumors (NGGCT) masquerading as seasonal allergies: subsequent NP management following oncologic diagnosis**

Kamilah A Dowling

Department of Neurosurgery, The Childrens Hospital at Montefiore Medical Center, Bronx New York, USA

**OBJECTIVE:** Germ cell tumors of the central nervous system have varying prognosis depending on the subtype. NGGCT is unfavorable when compared to pure Germinoma Cell Tumors (GCT), however NGGCT are chemo sensitive and have demonstrated good response with combination radiation therapy. A significant proportion of patients with GCT experience a delay in time to diagnosis, in some cases despite evaluation by general pediatricians and specialists. This delay increases the risk of disseminated disease. Early detection and proactive diagnosis of these tumors are required because diagnosis delay may negatively influence patient survival. This presentation will describe the management of an adolescent male patient, who was diagnosed with NGGCT. The patient presented with progressive symptoms for over one year. Presentation of symptoms work-up, diagnosis and treatment will be discussed. The role of the advanced practice nurse will be outlined.

**MATERIAL-METHODS:** Retrospective chart analysis of a patient with diagnosis of NGGCT

**RESULTS:** Germ cell tumors of the central nervous system have varying prognosis depending on the subtype. NGGCT is unfavorable when compared to pure Germinoma Cell Tumors (GCT), however NGGCT are chemo sensitive and have demonstrated good response with combination radiation therapy. A significant proportion of patients with GCT experience a delay in time to diagnosis, in some cases despite evaluation by general pediatricians and specialists. This delay increases the risk of disseminated disease. Early detection and proactive diagnosis of these tumors are required because diagnosis delay may negatively influence patient survival. This presentation will describe the management of an adolescent male patient, who was diagnosed with NGGCT. The patient presented with progressive symptoms for over one year. Presentation of symptoms work-up, diagnosis and treatment will be discussed. The role of the advanced practice nurse will be outlined.

**CONCLUSIONS:** Early detection and proactive diagnosis of these tumors are required because diagnosis delay may negatively influence patient survival.

**Keywords:** NGGCT

#### NS-008

##### **Nursing of patient with aphasia after brain tumor surgery: how to care for the patient the way they used to live**

Yuka Nouji, Aki Tashiro, Hitomi Kawabata, Masahiro Nonaka  
Kansai Medical University, Hirakata, Japan

**OBJECTIVE:** Postoperative complication such as hemiparesis and a decline of activity of daily life may occur after brain tumor surgery, and may not recover soon, and remains for a long period. This situation is stressful for both parents and patients. And sometimes, they fall in the mental critical state.

**MATERIAL-METHODS:** We experienced a patient who became aphasic and required total assistances due to postoperative cerebellar mutism after medulloblastoma surgery. Her mother was the only one who could tell what her daughter wanted without words. We set a goal with her mother, not to restrict her daily life, and try to maintain their daily life according to the way they used to live. For example, we removed her urinary catheter to encourage her to go to the restroom, and assisted her to attend at the school class in the hospital. For this purpose, we asked her mother to participate in nursing care with nursing staff.

**RESULTS:** The patient and her family successfully escaped from the mental critical state due to her bed-ridden, aphasic condition.

**CONCLUSIONS:** This nursing intervention to maintain patient's daily life is thought to be effective.

**Keywords:** nursing mutism aphasia

#### NS-009

##### **The importance of assessing for papilledema in children with raised intracranial pressure. (case study)**

Michele Maree

Red Cross War Memorial Children's Hospital, Cape Town, SOUTH AFRICA

**OBJECTIVE:** Papilledema, defined as swelling of the optic disc, occurs frequently in the setting of increased ICP due to a variety of conditions, including hydrocephalus, intracranial neoplasms, pseudotumor cerebri and sinus thrombosis. The optic nerve can be visualised non-invasively using an ophthalmoscope, and this is referred to as fundoscopy. The typical findings occurring in the setting of papilledema include flattening of the posterior sclera and protrusion of the optic disc. Although the

pathogenesis of papilledema is not completely understood, an important factor is an increase in intracranial pressure that is transmitted to the subarachnoid space surrounding the optic nerve, thereby disturbing the metabolic processes of the nerve and consequently leading to oedema, ischemia, and ultimately visual impairment or loss. Numerous studies have established a link between elevated intracranial pressure and the development of papilledema. The importance of papilledema as a useful indicator of increased intracranial pressure is a valuable guide to the necessity for surgical intervention. Furthermore, early recognition of papilledema and elevated intracranial pressure is of paramount importance for ensuring restoration of vision. The essential etiological factor in papilledema is increase in intracranial pressure, relief of which will diminish the risk of blindness or serious deterioration of vision if it can be effected before secondary changes have developed in the disc. In attempting to detect and diagnose papilledema as early as possible in paediatric neurosurgical patients, fundoscopic examination is an essential non-invasive method.

**CASE STUDY:** 5 ½ girl referred with history of headaches, weight loss, change in behavior and loss of appetite, but an otherwise normal neurological examination. She was referred for neuroimaging which revealed a 4x3 cm inhomogeneously enhancing suprasellar lesion with compression of the chiasm and third ventricle, hydrocephalus with transependymal fluid shift and and leptomeningeal spread. The patient deteriorated acutely with the Glasgow Coma Score dropping to 7/15. Fundoscopic examination showed papilledema with micro hemorrhages and CT scan showed acute hemorrhage into tumor with increased ventricular size, necessitating an external ventricular drain.

**CONCLUSION:** Optic pathway tumors may be associated with loss of vision, degeneration of the optic nerve and papilledema. The finding of papilledema, that is, a swollen optic nerve-head usually reflecting elevated intracranial pressure, is a medical emergency.

**Keywords:** Papilledema, Pilocytic astrocytoma, Subarachnoid space. Optic nerve, Intracranial pressure, Ophthalmoscope.

#### NS-010

#### Managing complex hydrocephalus with the help of telemetric shunt reservoir pressure readings- challenges and future strategies : a case study

John Preston

Sheffield Children's Hospital, Sheffield, UK

An evaluation of the first six months after insertion of a Miethke sensor reservoir® into a 3 year old boy with slit ventricle syndrome. Using pressure readings to guide adjustments to a combined proGAV® and proSA® ventriculoperitoneal shunt system, we aim to improve his management and reduce the number of neurosurgical procedures and CT scans. Management of complex hydrocephalus in children can be challenging and can present diagnostic dilemmas. We replaced a standard reservoir of an indwelling proGAV/proSA® combined ventriculoperitoneal shunt system with a Miethke sensor reservoir® in a 3 year old boy with difficult to manage slit ventricle syndrome. The child had undergone numerous neurosurgical interventions including shunt revisions, extraventricular drains and ICP monitoring. By combining telemetric pressure sensor readings along with reported symptoms we were able to guide his management without the need for further surgical intervention. We took pressure readings from the reservoir in the horizontal, vertical and sitting positions using two different device modes. These readings were compared and used alongside symptoms reported. We will present data from the pressure sensor and discuss our experience of its use in this paediatric case study. We acknowledge the limitations of our study, namely that we currently only have six months of data. However this is a pilot study evaluating the use of the Miethke sensor reservoir® in paediatrics and our initial results have been encouraging. We believe the use of this device can improve the

management and reduce the need for surgical intervention and number of CT scans in children with complex and difficult to manage hydrocephalus.

Monday, 24 October 2016

14:00 – 14:10

## Nursing Symposium - II

#### NS-011

#### improved skin care procedure during distraction osteogenesis for craniosynostosis

Ayana Uegaki<sup>1</sup>, Yusaku Kikuchi<sup>1</sup>, Kayoko Asada<sup>1</sup>, Satomi Hayashi<sup>1</sup>, Mieko Hayakawa<sup>1</sup>, Hirokatsu Osawa<sup>2</sup>, Mihoko Kato<sup>2</sup>

<sup>1</sup>Faculty of Nursing, 31 Ward, Aichi Children's Health and Medical Center

<sup>2</sup>Department of Neurosurgery, Aichi Children's Health and Medical Center

**OBJECTIVE:** Some problems such as erosion, crusting and ulceration of the skin are frequently occurred during the distraction osteogenesis. Previously we reported the importance of coherent procedure during hospitalization and at home. For three years after the last evaluation, patients have kept stable skin condition. In this study, we tried to analyze factors that contributed to the improvement.

**MATERIAL-METHODS:** Twenty one patients received cranioplasty from April, 2013 to March, 2016. The outcomes of the patients were compared with formerly reported 30 patients. We clarified that nursing staff thought what was the most important factor and checked how to teach patients family about shampooing and crust removing.

**RESULTS:** They recognized that removing crust procedure was one of the most important factors. All of the nursing staff had stopped using a brush and used gauze for cleaning the wound gently. They taught the same shampooing manner for parents. The occurrence of skin complications was less often in recent group than the former group.

**CONCLUSIONS:** We previously reported the importance of the coherent guidance for parents and the successful reduction of the skin complication. We could reduce complications more by introducing a procedure that senior nurses used to practice.

**Keywords:** Skin Care, Distraction Osteogenesis, Craniosynostosis

#### NS-012

#### Distraction for distraction

Yuko Hirano<sup>1</sup>, Chie Fuse<sup>1</sup>, Yoshimi Tanase<sup>1</sup>, Ayana Uegaki<sup>2</sup>, Yusaku Kikuchi<sup>2</sup>, Hirokatsu Osawa<sup>3</sup>, Mihoko Kato<sup>3</sup>

<sup>1</sup>Child Life Support Team, Aichi Children's Health and Medical Center

<sup>2</sup>Faculty of Nursing, 31 Ward, Aichi Children's Health and Medical Center

<sup>3</sup>Department of Neurosurgery, Aichi Children's Health and Medical Center

**OBJECTIVE:** Cranial vault distraction osteogenesis has been performed actively in the Neurosurgical Department of Aichi Children's Health and Medical Center, hereafter called "the Center." The children need to take the skull expansion by distracting the shafts approximately for a month after the operations. However, it is quite difficult for the children to stay calm during the distraction because they tend to cry and move acutely due to the stress from their fear and anxiety. In order to alleviate their stress and shorten treatment time, the doctors, nurses, and Hospital play specialists (HPS) of the Center together have made a "DISTRACTION" method, which was based on the idea

that a good distraction focused on their thoughts from the procedure. Here, we would like to report the result as well as the process to develop the method.

**MATERIAL-METHODS:** For preschool children, we used action toys with sound effects or lightning toys when children took distraction procedure in the ward. On the other hand, school age children played with mobile tablets like iPad during the treatments.

**RESULTS:** The school age children were often into their plays, especially with iPad before and after the treatments. Infants were crying from fear or pain even during DISTRraction, however, the children tended to calm down soon after the treatments

**CONCLUSIONS:** It is considered that DISTRraction is remarkably effective for the school age children, though for the infants the method is insufficient. We would like to make an effort to improve the efficacy of DISTRraction especially for infants with cooperation among the doctors, nurses, and HPS together.

**Keywords:** Distraction, Hospital play specialist, Distraction osteogenesis

**NS-013**

**Seeing double: craniopagus conjoined twins**

Bindu Peter

Montefiore Medical Center

**OBJECTIVE:** Craniopagus twins represent a rare phenomenon of congenital misfortune. Craniopagus is of two types, partial and total. In the partial form, the union is of limited extent, particularly as regards to its depth, and separation. It can be expected that both children can lead normal lives. In this review, I will explore the history of craniopagus as well as our current understanding of the malformation. With the increase in modern neurosurgical technology and techniques, numerous opportunities for successful separations can be anticipated. Normalcy can be achieved in these children. When venous drainage from the 2 brains are connected, it has been documented that staged separation is preferable, as gradual alteration of hemodynamics may be safer than a single-stage procedure.

**MATERIAL-METHODS:** Power point presentation

**RESULTS:** Explanation & definition

**CONCLUSIONS:** With increased technological aids patients can lead a normal life.

**Keywords:** craniopagus, conjoined twins

**NS-014**

**Knowing a thing or “TWO” about proper positioning for conjoined craniopagus twins**

Cecile Edwards

Montefiore Medical Center

**OBJECTIVE:** Proper positioning of a patient for any neurosurgical procedure is an important part of the perioperative nursing care that should not be under-emphasized. When positioning two patients that are joined at the head, “there is no two ways about it”, it can be difficult. Conjoined twins are rare and only about two percent of all conjoined births are what's called craniopagus twins, those joined at the head. In this presentation, collaboration among the entire OR team was used with creative methods to protect both patients from mechanical injury, by maintaining normal body alignment without excess

flexion, extension, or rotation which are consistent with proper principles of positioning for surgical patients.

**MATERIAL-METHODS:** Power point presentation

**RESULTS:** Risks and Benefits of Patient Positioning During Neurosurgical Care

**CONCLUSIONS:** Positioning of the patient for neurological surgery is an important part of nursing care and poses many technical and physiological challenges especially when dealing with conjoined twins.

**Keywords:** Neurosurgical positioning

**NS-015**

**Systematic approach to improved acute postoperative management of patients following functional hemispherectomy**

Herta Yu, Kathryn Sebastien, Nicole Baer

Department of Neurosurgery, Hospital for Sick Children, Toronto, Canada

**OBJECTIVE:** Functional hemispherectomy is a well accepted procedure for surgical treatment of selected patients with medically refractory epilepsy. The surgery promotes hope for cure of seizures but is also a source of stress and anxiety for these families. Children undergoing this surgery generally demonstrate a range of acute, transient post-operative symptoms as a result of the technical procedure; however, these same symptoms may indicate more concerning complications such as infection, meningitis, stroke, or hydrocephalus that require extensive investigation and treatments. These events compound the stress and anxiety for these families thereby making the hospitalization a difficult experience.

**MATERIAL-METHODS:** We performed a retrospective review of all patients who underwent functional hemispherectomy at our center between June 2000 and March 2014.

**RESULTS:** During the identified period, 47 patients underwent a total of 54 hemispherectomies with 6 patient having repeat surgeries. Common post-operative symptoms included: fever (33); motor/sensory deficits (13); persistent irritability (12); and lethargy (10). There were five positive cultures however, none were from CSF samples. More persistent complications resulted in two patients developing hydrocephalus that required VP shunt insertion and two other patients had strokes.

**CONCLUSIONS:** The findings of our review provided information to change practice and improved quality of care. The information allowed for enhanced nursing education, family preparation, and parental guidance during the pre-operative phase. The clinical data improved focused care, earlier use of steroids, limited the lab work required, and promoted greater collaboration between members of the health care team. Although the presence of acute post-operative symptoms are inherent to the technical operative procedure of functional hemispherectomy, the approach to systematically obtain data via the retrospective review provided evidence to support changes in practice that promoted quality of care and methods to minimize the stress and anxiety to improved overall hospital experience for these families.

**Keywords:** hemispherectomy, post-operative symptoms, nursing care

**NS-016**

**A case of successful functional posterior rhizotomy with multidisciplinary collaboration**

Masato Yamashita<sup>1</sup>, Takahito Iitsuka<sup>1</sup>, Daisuke Sakamoto<sup>2</sup>, Atsuko Harada<sup>2</sup>,

Mami Yamasaki<sup>2</sup>, Atsusi Keyaki<sup>3</sup>

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<sup>3</sup>Department of Rehabilitation Medicine, Takatsuki General Hospital, Osaka, Japan.

**OBJECTIVE:** We report the case of a 7-year-old boy with severe cerebral palsy (CP) who underwent functional posterior rhizotomy (FPR) with multi-disciplinary collaboration before and after surgery.

**MATERIAL-METHODS:** The boy was born at 23 gestational weeks with a birth weight of 558 grams. He was diagnosed with athetotic cerebral palsy. He was classified as level V in the Gross Motor Function Classification System (GMFCS). We evaluated the degree of spasticity using the Modified Ashworth Scale (MAS) prior to surgery. Flexion and abduction of the hip joint was 3 points and flexion of the knee joint was 3 points on MAS. Unpleasant stimuli and mental stresses caused respiratory wheezing and increased muscle tone, and the boy could not remain in a buggy for more than five minutes once he became hypertonic.

**RESULTS:** We held a multidisciplinary meeting with doctors, nurses, and a physical therapist before the surgery. The patient underwent FPR of L2-S1 without any complications, and began riding in the buggy eight days after the surgery. We prepared a handout to help the family and nurses properly position the boy to sit in the buggy. Another multidisciplinary conference, this time including the home-based staff, was held before the boy was discharged from the hospital. A physical therapist assigned to help the boy at home visited the hospital several times before the discharge to become acquainted with the boy's condition and current physiotherapy. Finally, the boy was discharged home after surgery and two months of rehabilitation.

**CONCLUSIONS:** Sharing information between the various disciplines involved, both before and after surgery, avoided complications and promoted successful rehabilitation. The family felt confident in having the child discharged home because of the communication and continuity of care between the hospital-based and home-based physical therapists.

**Keywords:** functional posterior rhizotomy, GMFCS level V, multidisciplinary collaboration

NS-017

**A review of the VNS device ASPIRE SR with Auto stimulation function, versus standard VNS therapy**

Kate Watts<sup>1</sup>, Michelle Seymour<sup>1</sup>

<sup>1</sup>Lead Nurses for Children's Epilepsy Surgery, Bristol Children's Hospital, UNITED KINGDOM

**INTRODUCTION:** Vagal Nerve Stimulation (VNS) therapy is considered for use in children who have been evaluated on the Children's Epilepsy Surgery Service (CESS) programme at Bristol Children's Hospital and whom are not suitable candidates for epilepsy surgery. Bristol Children's Hospital were the first to implant the Aspire SR in a paediatric patient in the UK in July 2014. Since then a total of 41 implants have been performed (41 new implants, 18 generator replacements). The Aspire SR is the latest model of VNS device which has an additional function which responds (provides an automated stimulation) to an increase in heart rate which often occurs during a seizure (ictal tachycardia). This provides reactive stimulation which can be particularly beneficial to those patients experiencing nocturnal seizures, patients that are non-verbal and patients that don't experience an aura.

**AIM:** To establish if the VNS device ASPIRE SR with auto stimulation function provides additional benefit to patients compared to standard VNS therapy.

**METHOD:** 18 children underwent a VNS generator replacement from standard VNS therapy to Aspire SR with auto stimulation. 12 out of the 18 patients were included in the review. 6 patients were excluded from the review. 2 patients did not have the auto stimulation function on, 2 patients had been transitioned to adult services so follow up information was not available. 1 patient was out of region so again follow up information was not available.

The patient's medical notes were reviewed +/- a parental survey which had been sent to the families beforehand. Seizure type, frequency, duration and recovery were reviewed. Changes in quality of life measures including mood, sleep, alertness, behaviour, memory were also evaluated. Changes in anti-epileptic medication, rescue medication usage and A&E attendances were also documented when available.

**RESULT:** Preliminary findings show a decrease in seizure frequency and improvement in a number of quality of life measures such as sleep, mood and alertness. Other benefits noted were a decrease in rescue medication and reduction in A&E attendances (health care utilisation).

NS-018

**The holistic nursing care of a child with vein of Galen aneurysm**

Francesca Ann White, Nicola Wilson

Neuroscience Department, Great Ormond Street Hospital, London, United Kingdom

**OBJECTIVE:** Vein of Galen aneurysm is a rare neurovascular malformation (Hoang et al 2009). Vein of Galen aneurysm is associated with a significant morbidity and mortality due to the risk of intracranial bleeding (Hoang et al 2009; Smith & Martin 2009). Therefore, in light of the safe and sustainable approach in the United Kingdom all children with a vascular disorder are referred to a tertiary centre where range of treatments can be offered and healthcare specialists are available to support the child and family. Prompt recognition and early treatment may improve the survival outcome for the child. The objective of this presentation is to demonstrate the fundamental part nurses play in the process of delivering holistic care and shaping the patient experience.

**MATERIAL-METHODS:** For this reason, this oral presentation is to provide an insight of the nursing care for a child with Vein of Galen aneurysm at Great Ormond Street Hospital. The immediate and long term outcomes will be discussed for this condition.

**RESULTS:** This presentation will summarise the clinical presentation of a child with Vein of Galen aneurysm. Also describe how as nurse working at a tertiary neurosurgical unit, support the child and families through the diagnostic investigation, management and treatment for this condition by relating it to a specific case study.

**CONCLUSIONS:** Therefore, this will give an insight to all professionals the impact and the journey the child and family experiences. This presentation will conclude by discussing our current outcomes of morbidity and mortality and comparing this against the wider literature.

**Keywords:** Nursing Neurovascular Family experiences Vein Galen aneurysm

**List of posters presented at 44th ISPN Annual Meeting**

Monday, 24 October 2016, 15:20 – 16:00

**Poster session 1: Craniofacial | Epilepsy and Functional | Spine | Other**

PP-001

The RUNX2 master gene in nonsyndromic craniosynostosis: insights from molecular biology towards the prediction of site and time of suture closure  
Gianpiero Tamburrini (Roma, Italy)

PP-002

New Tool for Neurosurgery Teaching: The Craniosynostosis Puzzle  
Giselle Coelho (Sao Paulo, Brazil)

**PP-003**

The computed cranial focal point  
Guido de Jong (Nijmegen, Netherlands)

**PP-004**

Evaluation of craniocerebral disproportion in children with craniosynostosis  
Aleksandr Kim (St. Petersburg, Russia)

**PP-005**

Advantages of Cranial Orthotics as an Adjuvant to Cranial Vault Remodeling in Infants with Craniosynostosis  
Volkan Etus (Kocaeli, Turkey)

**PP-006**

Phi Flap Craniectomy for Cranial Vault Reshaping in Children with Multiple Non-Syndromic Craniosynostosis: A Case Series  
Fadzlishah Johanabas Rosli (Kuala Lumpur, Malaysia)

**PP-007**

Cranial index change in scaphocephaly – analysis of seventy seven cases  
Santosh Mohan Rao Kanangi (Liverpool, United Kingdom)

**PP-008**

Fronto-orbital advancement for bicoronal synostosis: technique and results  
Matthieu Vinchon (Lille, France)

**PP-009**

Posterior cranial vault expansion using distraction osteogenesis in patients with multiple synostosis  
Tatsuki Oyoshi (Kagoshima, Japan)

**PP-010**

Multi-directional Cranial Distraction Osteogenesis (MCDO) procedure for craniosynostosis cases  
Masahiro Kameda (Okayama, Japan)

**PP-011**

Robotic assisted-(ROSA) midface distraction using transfacial pin in patients with craniofacial dysostosis  
Suhas Udayakumaran (Kochi, India)

**PP-012**

Diagnosis, management and outcome of Mercedes Benz Pattern craniosynostosis  
Oscar García González (León, Mexico)

**PP-013**

Surgical Treatment of Life Threatening Intraoperative Complications of Cranial Expansion Surgery in Complex Syndromic Craniosynostosis  
Azmi Alias (Kuala Lumpur, Malaysia)

**PP-014**

Intra-operative dural tears and their impact in craniofacial procedures: analysis of 410 consecutive cases

Santosh Mohan Rao Kanangi (Liverpool, United Kingdom)

**PP-015**

Two cases of bilateral lambdoid and sagittal craniosynostosis required ventriculo-peritoneal shunt  
Daimon Shiraiishi (Obu, Japan)

**PP-016**

Utilization of Computer-Based Navigation for Anteromesial Temporal Lobectomy with Amygdalo-Hippocampectomy in Pediatric Epilepsy Surgery: A Case Series  
Fadzlishah Johanabas Rosli (Kuala Lumpur, Malaysia)

**PP-017**

Cost utility analysis of treatment strategies for drug-resistant epilepsy in children with Tuberous Sclerosis Complex  
Aria Fallah (Los Angeles, United States)

**PP-018**

Hypothalamic Hamartomas in Children  
Hamilton Matushita (São Paulo, Brazil)

**PP-019**

Surgical management strategies and seizure outcome in medically refractory pediatric epilepsy with large gliotic foci  
Siddharth Saurav (Kochi, India)

**PP-020**

Posterior quadrantic scar epilepsy: Excellent seizure and cognitive outcomes with regional disconnective surgery  
Siddharth Saurav (Kochi, India)

**PP-021**

Extratemporal refractory epilepsy: surgical outcome  
Suchanda Bhattacharjee (Hyderabad, India)

**PP-022**

Neurocognitive Function in Children after Anterior Temporal Lobectomy with Amygdalohippocampectomy  
Ju Seong Kim (Seoul, South Korea)

**PP-023**

Improvement of Health Related QOL after Vagus Nerve Stimulation (VNS) for pediatric Drug Resistant Epilepsy (DRE)  
Satoko Ochi (Sapporo, Japan)

**PP-024**

Electromagnetic stimulation therapies for refractory status epilepticus  
Colin John Kazina (Winnipeg, Canada)

**PP-025**

Gpi DBS for Dystonia in Pediatric Patients  
Huseyin Canaz (Istanbul, Turkey)

**PP-026**

Intrathecal baclofen for chronic visceral pain in children - A case report  
Santosh Mohan Rao Kanangi (Liverpool, United Kingdom)

**PP-027**

Neurosurgical procedures for pediatric patients with congenital bleeding disorders  
Hiroshi Yokota (Kashihara, Japan)

**PP-028**

Early versus late repair of myelomeningoceles in Kuwait: an institutional case review  
Alya Hasan (Safat, Kuwait)

**PP-029**

Atypical Dermoid in upper cervical junction- a case report  
Ashok Gupta (Jaipur, India)

**PP-030**

Pyomyelia in pediatric patients with dermal sinus tract  
Sheng Che Chou (Taipei, Taiwan)

**PP-031**

Preoperative and Postoperative Urodynamic Differences of Children with Spina Bifida Aperta  
Huseyin Canaz (Istanbul, Turkey)

**PP-032**

Vertebral Deformity Surgery in Spina Bifida Patients  
Huseyin Canaz (Istanbul, Turkey)

**PP-033**

Spina Bifida in Sudan  
Mohammed Awad Elzain Ahmed (Khartoum, Sudan)

**PP-034**

A new approach in the repair of a myelomeningocele using autologous amnion and a sensate perforator flap. A case report  
Kristin Sjøvik (Tromsø, Norway)

**PP-035**

Sequential morphological change of Chiari malformation type II following surgical repair of myelomeningocele  
Kimiaki Hashiguchi (Fukuoka, Japan)

**PP-036**

Effective dural tenting with a little trick on foramen magnum decompression for Chiari malformation  
Shunsuke Ichi (Tokyo, Japan)

**PP-037**

Non contiguous double spinal lipoma with tethered cord and polydactyl: A clue to multimodal origin of spinal lipomas  
Shashank Ravindra Ramdurg (Kalaburagi, India)

**PP-038**

Does large syrinx with spinal lipoma need syrinx-subarachnoid shunt?  
Masato Nagasaka (Kasugai Shi, Japan)

**PP-039**

Management of CSF leak post detethering in spinal dysraphism  
Suhass Udayakumaran (Kochi, India)

**PP-040**

Is the outcome of surgery for Spinal Cord Lipomas worse in the presence of additional Split Cord Malformation?  
Shibu Vasudevan Pillai (Bangalore, India)

**PP-041**

Symptomatic congenital tethered cord developed in adulthood  
Yumiko Someno (Hirakata, Osaka, Japan)

**PP-042**

Spinal lipoma without skin lesions  
Kunitoshi Otsuka (Tokyo, Japan)

**PP-043**

Lumbosacral cutaneous lesions and spina bifida occulta  
Kazuhiisa Yoshifuji (Sapporo, Japan)

**PP-044**

Proposal of an additional entity: 'probable' limited dorsal myeloschisis  
Ji Yeoun Lee (Seoul, South Korea)

**PP-045**

Limited dorsal myeloschisis: Is it truly a benign form of spinal dysraphism??  
Shashank Ravindra Ramdurg (Kalaburagi, India)

**PP-046**

Cranial vault expansion for management of intracranial hypertension due to osteopetrosis  
Shaun D Rodgers (New York, United States)

**PP-047**

Long term follow up of a complex pygopagus Conjoined Twins after successful separation, Case report in Mansoura University Hospitals  
Hatem Ibrahim Badr (Mansoura, Egypt)

**PP-048**

A multidisciplinary approach to a girl with Williams-Beuren syndrome and possible tight filum syndrome  
Erwin M.j. Cornips (Maastricht, Netherlands)

**PP-049**

Augmentation Laminoplasty in Paediatric Achondroplasia: Towards Avoiding Instrumented Fixation while Stabilising Deformity Progression  
Guirish A Solanki (Birmingham, United Kingdom)

**PP-050**

Minimally Invasive Approaches to the Spine in Pediatric Patients: Direct Pars Repair, Technical Note and Literature Review  
Hasan Raza Syed (Woodbridge, United States)

**PP-051**

Microsurgical Excision of Herniated Lumbar Disc in Pediatric Population  
Ahmed Awad Zaher (Mansoura, Egypt)

**PP-052**

Pediatric spine surgery for Non congenital Spinal Conditions  
Mohammed Awad Elzain Ahmed (Khartoum, Sudan)

**PP-053**

Factors affecting recovery of neurological deficits in surgical management of intraspinal ganglioneuroma: report of 2 pediatric cases and literature review  
Yoshinori Maki (Kyoto, Japan)

**PP-054**

Midline Ventral Intradural Granular Cell Tumor of the Cervical Spinal Cord Resected via Anterior Corpectomy with Reconstruction: Technical Case Report and Review of Literature  
Hasan Raza Syed (Woodbridge, United States)

**PP-055**

A Case of Sacral Extradural Meningocele  
Yoshiki Uemura (Takatsuki, Japan)

**PP-056**

Underestimated midline nasal pimple in children turn out as intracranial, intradural fronto nasal dermoids  
Stephanie Anetsberger (Heidelberg, Germany)

**PP-057**

Rare fronto-basal intraparenchymal dermoid cyst mistaken for cavernoma  
Stephanie Anetsberger (Heidelberg, Germany)

**PP-058**

Complicated Congenital Dermal Sinus, Diagnosis and Management  
Mohame Ali Kassem (Mansoura, Egypt)

Tuesday, 25 October 2016, 10:15 – 11:00

## Poster Session 2: Hydrocephalus | Vascular | Trauma | Other

**PP-059**

Determining Cerebrospinal Fluid (CSF) Ventricular Volume with a Single Mouse Click  
J Gordon McComb (Los Angeles, United States)

**PP-060**

Expression of aquaporin 1 and 4 in the choroid plexus and brain parenchyma of kaolin-induced hydrocephalic rats  
Sung Kyoo Hwang (Daegu, South Korea)

**PP-061**

Posthemorrhagic Hydrocephalus in Preterm Babies: Treatment Options  
Ömer Can Yildiz (Magdeburg, Germany)

**PP-062**

Hydrocephalus in Sudan Diagnosis and Management  
Mohammed Awad Elzain Ahmed (Khartoum, Sudan)

**PP-063**

Analysis of hydrocephalus management in Tallinn Children's Hospital between 2010 and 2015  
Andres Asser (Tallinn, Estonia)

**PP-064**

Report of 3 cases of Mucopolysaccharidosis type II (MPS II; Hunter syndrome) with communicating hydrocephalus treated by ventriculoperitoneal (VP) shunting  
Chisato Yokota (Osaka, Japan)

**PP-065**

The relationship between clinical data and progressive hydrocephalus in patients with myelomeningocele  
Nayuta Higa (Kagoshima, Japan)

**PP-066**

Acute MR examination in pediatric patients with programmable drainage system  
Eva Brichtová (Brno, Czech Republic)

**PP-067**

Outcome analysis of newborn patients with hydrocephalus occurred after meningomyelocel repairing: a comparison of ventriculoperitoneal and ventriculopleural shunting  
Gökmen Kahiloğulları (Ankara, Turkey)

**PP-068**

Long-term outcomes of ventriculoperitoneal shunt in pediatric patients with more than 15 years of follow-up  
Noritsugu Kunihiko (Osaka, Japan)

**PP-069**

Strategy for prevention of shunt infection and shunt malfunction in neonates and infants with hydrocephalus  
Hirokazu Nakatogawa (Hamamatsu, Japan)

**PP-070**

Ventriculoatrial Shunt Complication Avoidance and Management- A Single Center Experience

Abilash Haridas (Detroit, United States)

**PP-071**

ValveViz - a novel method of adjustable shunt valve visualization  
Sergey Abeshaus (Haifa, Israel)

**PP-072**

Surgical result of endoscopic third ventriculostomy for pediatric hydrocephalus: analysis of failure cases  
Shinji Iwata (Matsuyama, Japan)

**PP-073**

The feasibility of endoscopic surgery combined with electromagnetic-guided neuronavigation and narrow-band imaging for intractable multiloculated infantile hydrocephalus  
Koji Fujita (Wakayama City, Japan)

**PP-074**

A new device for anchoring external CSF catheters  
Luca Massimi (Rome, Italy)

**PP-075**

Hydranencephaly: to treat or not to treat and if- how to treat  
Stephanie Anetsberger (Heidelberg, Germany)

**PP-076**

Simultaneous ventriculoperitoneal shunt removal and endoscopic third ventriculostomy for 3 patients previously treated for intracranial germ cell tumors more than 20 years ago  
Daisuke Kita (Yokohama, Japan)

**PP-077**

Endoscopic Third Ventriculostomy as a Possible Measure for the Treatment of Hydrocephalus of a Premature Baby – Case Report  
Marcia Cristina da Silva (Belo Horizonte, Mg, Brazil)

**PP-078**

Endoscopic resection of a choroid plexus papilloma in an infant  
Nida Thamenah Kalyal (Bognor Regis, United Kingdom)

**PP-079**

Complete resection of a giant colloid cyst in a child using a monoportal endoscopic approach  
Nida Thamenah Kalyal (Essex, United Kingdom)

**PP-080**

Endoscopic ultrasonic aspiration for third ventricle colloid cyst  
Paola Hernandez Ponce (León, Mexico)

**PP-081**

Endoscopic fenestration for the symptomatic cysts of septum pellucidum  
Takaoki Kimura (Tokyo, Japan)

**PP-082**

Surgical management of pediatric Interhemispheric arachnoid cyst, An Institutional experience  
Hardik Sardana (New Delhi, India)

**PP-083**

The ‘invisible cyst’: tri-ventriculomegaly due to a third ventricular ependymal cyst  
Mahum Imran (London, United Kingdom)

**PP-084**

Cysts of the Cavum Septi interpositi - how to manage?  
Uli Barcik (Moralzarzal, Madrid, Spain)

**PP-085**

Giant prepontine – bitemporobasal- multicompartmental arachnoid cyst complex – theories on origin and surgical strategies  
Suhass Udayakumaran (Kochi, India)

**PP-086**

Incidence of subcallosal midline cysts by brain computed tomography in pediatrics  
Kyoji Tsuda (Tsukuba, Ibaraki, Japan)

**PP-087**

Thoracoscopic-assisted ventriculoatrial shunt placement  
Jennifer Sachiko Ronecker (Ossining, Ny, United States)

**PP-088**

Moyamoya Disease and Stroke Post-Hematopoietic Stem Cell Transplant in Patient with Sickle Cell Disease: A Case Study  
Abilash Haridas (Detroit, United States)

**PP-089**

Indirect Revascularization in Adult Moyamoya Disease  
Hao Chun Hsu (Taipei, Taiwan)

**PP-090**

Moyamoya Disease and Indirect Revascularization  
Hakan Karabagli (Konya, Turkey)

**PP-091**

Encephalo Duro Arterio Synangiosis (EDAS) in Childhood Moyamoya Disease (MMD): Our Initial Experience in NINS &H, Bangladesh  
Sudipta Kumer Mukherjee (Dhaka, Bangladesh)

**PP-092**

Histopathological features of pediatric intracranial cavernous malformation  
Naoyuki Ohe (Gifu, Japan)

**PP-093**

Clinical evaluation of nontraumatic intracranial hemorrhage in pediatric patients  
Takao Tsurubuchi (Tsukuba, Japan)

**PP-094**

Efficacy of indocyanine green videoangiography in direct surgery of pial arteriovenous fistula  
Nobuyuki Akutsu (Kobe, Japan)

**PP-095**

Intracranial Aneurysms in Children under One Year of Age: an Update  
Dennis R. Buis (Amsterdam, Netherlands)

**PP-096**

Rupture brain aneurysm: What to do when the parents (and child) refuse blood transfusion?  
Leandro Oliveira (Braga, Portugal)

**PP-097**

Clinical features of chronic subdural hematoma in pediatric patients  
Ayumi Narisawa (Sendai, Japan)

**PP-098**

Cranial Vault Reduction Cranioplasty for Severe Hydrocephalic Macrocephaly  
Wihasto Suryaningtyas (Surabaya, Indonesia)

**PP-099**

Characteristics of anterior transpetrosal approach in pediatrics  
Tomoru Miwa (Tokyo, Japan)

**PP-100**

Hybrid surgery in pediatric neurosurgery  
Yu Cheng Chou (Taipei, Taiwan)

**PP-101**

Usefulness of an electromagnetic neuronavigation system for pediatric neurosurgery  
Kazuhiko Kurozumi (Okayama, Japan)

**PP-102**

Epidemiology of Pediatric Neurosurgery in a Third Level Hospital in México City: A report from a 16 year retrospective chart review  
Abraham Ibarra De La Torre (Mexico City, Mexico)

**PP-103**

Endoscopic removal of neonatal acute epidural hematoma via Strip-Bending osteoplastic craniotomy  
Hirokazu Nakatogawa (Hamamatsu, Japan)

**PP-104**

Our experience of the cranioplasty of autologous bone in the pediatric population  
Janna Semenova (Moscow, Russia)

**PP-105**

Distinguishing Accident from Abuse in Acute Subdural Hematoma  
Ruka Nakasone (Osaka, Japan)

**PP-106**

Traumatic Posterior Fossa Subdural Effusion associated with secondary acute hydrocephalus in a child  
Yoshinori Omori (Sapporo, Japan)

**PP-107**

A case of the infant head injury that the seamless emergency system led to lifesaving  
Jun Sakuma (Fukushima, Japan)

**PP-108**

Navigation Guided Trans-cerebellar Endoscopic Removal of Bullet in a Child with Gun Shot Head injury  
Azmi Alias (Kuala Lumpur, Malaysia)

**PP-109**

Successful Management of a Pediatric Case with Complicated Calvarian Depression Fracture due to Dog Bite Injury: A Case Report and Literature Review  
Abdülkerim Gökoğlu (Kayseri, Turkey)

**PP-110**

Selective serotonin reuptake inhibitors in the early period of recovery after severe traumatic brain injury in children  
Yuliya Sidneva (Moscow, Russia)

**PP-111**

Cranioplasty after external decompression in pediatric severe head injury: long-term outcome of cranioplasty using custom-made artificial bone flap and bioresorbable plates  
Naoki Kagawa (Osaka, Japan)

**PP-112**

Update of Skull Implants in very Young Children  
Angela Brentrup (Muenster, Germany)

**PP-113**

Infected Cephalhaematomas: Case Report and Review of Literature  
Samantha Ashworth (London, United Kingdom)

**PP-114**

A Large Calcified Subdural Empyema  
Saumitra Sarkar (Dhaka, Bangladesh)

**PP-115**

Brain Abscess in a Patient with Glycogen Storage Disease Type Ib  
Youngha Kim (Yongsan, South Korea)

**PP-176**

Head Injury Secondary to Rotor Blades in Children  
Rakesh Rethinasamy (Kuala Lumpur, Malaysia)

Wednesday, 26 October 2016, 15:30 – 16:15

### Poster Session 3: Neuro-oncology | Neuro-imaging | Molecular biology | Other

#### PP-116

Infantile Brain Tumours: A Tale of Two Cities  
Sandip Chatterjee (Kolkata, India)

#### PP-117

Incorrect diagnosis of “Cerebral palsy” in representative cases of an infantile tumor and a space occupying lesion, and postoperative reversible “chronic spasticity”  
Liana Adani Beni (Tel Aviv, Israel)

#### PP-118

BrainStem Leisons Stereotactic Procedure -Novel Approach for Histopathological Diagnosis in Paediatric Patients  
Bagathsingh Karuppanan (Madurai, India)

#### PP-119

Paediatric tectal plate ‘gliomas’: a decade’s single centre experience  
Nida Thamenah Kalyal (Essex, United Kingdom)

#### PP-120

Pilomyxoid Astrocytoma of Brainstem: Case report and literature review.  
Anwar ul Haq (Riyadh, Saudi Arabia)

#### PP-121

Surgery for diffuse intrinsic pontine glioma as palliation therapy  
Satoshi Ihara (Tokyo, Japan)

#### PP-122

True Aqueductal Tumors: A Unique Entity  
Shlomi Constantini (Tel Aviv, Israel)

#### PP-123

5ALA use in pediatric brain tumors  
Shlomi Constantini (Tel Aviv, Israel)

#### PP-124

Fluorescence-guided surgery with 5-aminolevulinic acid for resection of pediatric brain tumors  
Takafumi Wataya (Shizuoka, Japan)

#### PP-125

5-Aminolevulinic acid in paediatric brain tumours: The UK’s first case series  
Nida Thamenah Kalyal (London, United Kingdom)

#### PP-126

Malignant embryonal brain tumors occupying the cerebellopontine angle: report of four surgical cases  
Junji Koyama (Kobe, Japan)

#### PP-127

Management of Paediatric Lower Cranial Nerve Schwannoma Institutional Experience  
Gaurav Singh (New Delhi, India)

#### PP-128

A Rare and Interesting Case of Cerebellar Pleomorphic Xanthoastrocytoma, Chiari 1 Malformation in a patient of Neurofibromatosis type 1  
Rohan P Shah (Kalaburgi, India)

#### PP-129

The comparative review of the juvenile vestibular schwannoma between neurofibromatosis type 2 and non-neurofibromatosis type 2 patients  
Maya Kono (Tokyo, Japan)

#### PP-130

Hydrocephalus in children with posterior fossa tumors  
Sergei Kim (Novosibirsk, Russia)

#### PP-131

A 5-year experience in the surgical treatment of posterior fossa tumors in children: impact of interventional timing and preoperative neurological condition on clinical outcomes  
Gökmen Kahiloğulları (Ankara, Turkey)

#### PP-132

Reconstruction and closure of posterior fossa dura in pediatric patients using “inner flap” method: surgical technique and single center clinical experience  
Gökmen Kahiloğulları (Ankara, Turkey)

#### PP-133

Management of MB in children: 85 cases report from the department of pediatric neurosurgery, Xinhua Hospital  
Jie Ma (Shanghai, China)

#### PP-134

Outcomes of pediatric medulloblastoma in one institution  
Shigeo Ohba (Toyoake, Japan)

#### PP-135

Clinical factors and prognosis of children with medulloblastoma  
Hao Li (Shanghai, China)

#### PP-136

Proteome characterization of medulloblastoma DAOY cell line  
Luca Massimi (Rome, Italy)

#### PP-137

Two cases of radiation-induced tumor occurred after treatment for childhood medulloblastoma  
Yukiko Kotani (Nara Prefecture, Japan)

**PP-138**

A single-institution study of treatment outcomes for pediatric ependymomas  
Koji Yoshimoto (Fukuoka, Japan)

**PP-139**

Safety and tumor inhibitory effect of Ketogenic diet for pediatric patients with anaplastic ependymoma: Two case reports  
Hiroaki Matsuzaki (Kumamoto, Japan)

**PP-140**

Leptomeningeal seedings correlate with higher mortality in pediatric ependymomas  
Muh Lii Liang (Taipei, Taiwan)

**PP-141**

PF-A ependymoma = plastic ependymoma  
Akira Gomi (Tochigi, Japan)

**PP-142**

The long-term survivor of the recurrent pediatric ependymoma after gamma knife radiosurgery  
Atsufumi Kawamura (Kobe, Hyogo, Japan)

**PP-143**

Treatment outcome in childhood intracranial ependymomas: Experience from a single institution  
Keishi Makino (Kumamoto, Japan)

**PP-144**

A small series of pediatric Hamangioblastomas  
Laura Valentini (Milano, Italy)

**PP-145**

Pediatric Choroid Plexus Tumors, CCHE-57357 Experience  
Mohamed El Beltagy (Cairo, Egypt)

**PP-146**

Case report of synchronous pilocytic astrocytomas of right parietal lobe and cerebellum in a child of 9 years  
Sergei Kim (Novosibirsk, Russia)

**PP-147**

Cellular Proliferation is Not a Predictor for Progression Free Survival after Resection of Pediatric Pilocytic Astrocytomas  
Albert Tu (Los Angeles, United States)

**PP-148**

Analysis of BRAF V600E mutation gene expression in pediatric pilocytic astrocytomas: a retrospective review of 16 cases  
Kentaro Chiba (Tokyo, Japan)

**PP-149**

Antitumor effect of PEG-ZnPP in Rat Glioma Cells, F98 and C6, and in Brain Stem Tumor Models  
Young Sill Kang (Berlin, Germany)

**PP-150**

Angiogenic glioma: an Australasian Case Series  
Shinuo Liu (Auckland, New Zealand)

**PP-151**

Quantitative tumor metabolite analysis based on MR spectroscopy to predict patient prognosis of pediatric and young adult gliomas  
Shunsuke Nakae (Toyoake, Aichi, Japan)

**PP-152**

Spindle Cells expressed by Stem Cells as the Origin of Rat Glioblastoma Multiforme (GBM) – Equivalent to Precursor in Human GBM  
Shoko Merrit Yamada (Kawasaki, Japan)

**PP-153**

Subsequent Glioma a few decades after the initial Radiation Therapy for Pediatric Brain Tumor  
Tomoo Matsutani (Chiba Shi. Chiba, Japan)

**PP-154**

Pediatric high-grade brain tumors treated with an irradiation-sparing protocol - which subtype has long-term survival?  
Eveline Teresa Hidalgo (New York, United States)

**PP-155**

A rapid progressive left basal ganglia malignant mixed germ cell tumor had safe tumor resection with multimodality intraoperative neuromonitor assistance  
Szu Yen Pan (Taichung, Taiwan)

**PP-156**

Childhood pineoblastoma: clinical and therapeutic aspects and reported 18 cases  
Yongji Tian (Beijing, China)

**PP-157**

Pineal Anlage Tumor: Case report  
Hakan Karabagli (Konya, Turkey)

**PP-158**

Surgical tips of occipital transtentorial approach for large pineal and juxta-pineal region tumors  
Junichi Yoshimura (Niigata, Japan)

**PP-159**

Rapid Growth of Immature Teratoma: Case Report  
Harold Westley Philips (New York, United States)

**PP-160**

Extratemporal Pediatric Ganglioglioma and Epilepsy  
William Gump (Louisville, United States)

**PP-161**

Pediatric temporal tumors with and without epilepsy: review of 25 patients  
Daniel Dante Cardeal (São Paulo, Brazil)

**PP-162**

Anterior skull base tumours, endoscopy, intra-operative MRI, pituitary, craniopharyngioma, recurrence  
Santosh Mohan Rao Kanangi (Liverpool, United Kingdom)

**PP-163**

Malignant transformation of a craniopharyngioma to a high grade sarcoma seven years following gamma knife  
May Lian Leong (Singapore, Singapore)

**PP-164**

Spinal involvement in pediatric patients with neuroblastoma: the role of neurosurgical intervention  
Shih Hung Yang (Taipei, Taiwan)

**PP-165**

Spinal and para-spinal plexiform neurofibromas in NF1 patients, a clinical-radiological correlation study  
Liat Ben Sira (Tel-Aviv, Israel)

**PP-166**

Treatment of Symptomatic Epidural Spinal Masses in Children: Remarkable recoveries with urgent surgery in ASIA A patients  
Erin N Kiehna (Los Angeles, United States)

**PP-167**

Primary intracranial Ewing's sarcoma: report of three cases  
Michael H Handler (Aurora, United States)

**PP-168**

Multiple Recurrent Hand-Schuller-Christian disease of the skull bone: A case report  
Joon Khim Loh (Kaohsiung, Taiwan)

**PP-169**

Improved cosmetic outcome by bone remodeling in the chemotherapy for pediatric Langerhans cell histiocytosis  
Koji Fujita (Wakayama City, Japan)

**PP-170**

Quality of life outcomes in children who have undergone surgery only for benign intracranial neoplasms  
Conor Mallucci (Liverpool, United Kingdom)

**PP-171**

Intracerebral rhabdoid meningioma with brain and spine dissemination in a 7-years-old patient  
Tatiana Protzenko Cervante (Rio de Janeiro, Brazil)

**PP-172**

Molecular biology of neonatal and infant brain tumours - literature trends and current standing  
Conor Mallucci (Liverpool, United Kingdom)

**PP-173**

Physical Therapy for a Case of Trisomy 18  
Tadashi Kosugi (Amagasaki, Japan)

**PP-174**

Relationship between Retinal Nerve Fiber Layer Thickness and Spina Bifida  
Huseyin Canaz (Istanbul Turkey)