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Paul Steinbok, ISPN President
Chandrashekhar Deopujari, ISPN Meeting Chair
John Kestle, ISPN Scientific Chair

ORAL PRESENTATIONS

Monday October 17, 2011

Session 1. CNS Infection

Endoscopic intraventricular lavage in children with pyogenic ventriculitis

Azmi Alias, Mohammed Saffari Mohammed Haspani
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Introduction: Pyogenic ventriculitis is a rare manifestation of severe intracranial infection. It is associated with high mortality and morbidity thus, requiring early diagnosis and aggressive treatment.

Objectives: This study aims to describe our experience in managing six children with pyogenic ventriculitis at the Department of Neurosurgery, Hospital Kuala Lumpur, Malaysia from August 2008 to October 2010; age range from 29 days to 6 months. The causative organisms include *Staphylococcus aureus* (3), *Escherichia coli* (1), *Salmonella* sp. (1), and *Serratia marcescens* (1)

Operative technique: Frontal horn of lateral ventricle is cannulated, followed by insertion of a rigid endoscope and control irrigation of the Ringer's lactate solution. Endoscopic examination of the lateral ventricle is done followed by intermittent aspiration of the intraventricular pus and debris under endoscopic control, using modified 5Fr infant feeding catheter. An average of 3L of Ringer's Lactate solution (gravity feed) is used for irrigation until clear return. Similar procedure is repeated on the opposite side. Either Ommaya reservoir or external ventricular drainage is inserted into one side of the lateral ventricle at the end of procedure.

Results: All six patients recovered well and completed 6–8 weeks course of intravenous antibiotics: four subsequently underwent ventriculoperitoneal shunting and two patients remained shunt independent. There is no specific complication related to the operative procedure.

Conclusion: Bilateral endoscopic transcranial transventricular lavage is safe and effective in treating children with pyogenic ventriculitis. It promotes faster recovery and improves outcome.

Craniovertebral tuberculosis: review of 21 cases in the paediatric age group

Sandip Chatterjee (Park Clinic, Kolkata, India)

Introduction: Craniovertebral tuberculosis constitutes a small percentage of caries spine. We report on our series of 21 paediatric patients who we treated with this condition in the last 8 years.

Materials and methods: Of these 21 children, two had tuberculous affection of the atlanto-occipital join as well, and the remaining 19 had atlanto-axial tuberculosis alone. All were treated initially with antitubercular drug therapy and had their neck immobilized. Digital X-ray (including open-mouth and lateral flexion/extension views) as well as MR scans of the craniocervical region were performed in all cases, with CT scan being reserved for the few that bony anatomical details were needed.

Results: The clinical presentations varied from neck pain with hypoglossal nerve palsy to frank spastic quadriplegia and instability at the atlanto-axial junction (5/21). Large cold abscesses were seen in 17 of 21 cases and severe torticollis in five of 21.

We graded the children with this condition into three groups:(1) those with instability and gross neurodeficit who required early operative intervention(one needed trans-oral decompression with posterior fixation and four

required posterior fixation); (2) those who had severe torticollis and large cold abscesses who were treated with trans-oral aspiration of cold abscess followed by neck immobilisation(6/21); and (3) those who did not have significant neck muscle spasm or torticollis and who were treated with immobilization alone(10/21). Only one child in group 3 required delayed intervention for instability.

Conclusion: We propose that this is a good classification to decide on modality of treatment for this condition.

Pott's puffy tumor in pediatric patients

Tatiana Protzenko, José Francisco Salomão, Antônio Bellas, Rene Leibinger (Instituto Fernandes Figueira, Rio de Janeiro, RJ, Brazil)

Pott's puffy tumor is a subperiosteal abscess of the frontal bone with inflammatory signs of the overlying skin. This unique pathology is more often associated to frontal sinusitis, but a few cases have been seen associated to forehead trauma. The purulent material can lead to osteomyelitis and intracranial complications, such as epidural, subdural, and brain abscesses. We retrospectively review four patients with the diagnosis of Pott's puffy tumor treated between 1999 and 2009. Three cases were related to frontal sinusitis and one to forehead trauma. There were three males and one female. The age at presentation ranged from 11 to 15 years and the median age was 13 years. All patients were treated by the same neurosurgeon. Intracranial extension consisted of epidural empyema in three patients and subdural empyema in one. Osteomyelitis was absent in only one patient. Three of them were operated on and treated with antibiotics for 6 weeks. One patient was treated conservatively with antibiotics and serial CT scans showed the resolution of the infection over time. All patients had good outcome, without further complications.

The utility of serial PCR assay in the surgical management of Pneumococcal intracranial sepsis

Robin Bhatia, Kathryn Harris, John Hartley, Owase Jeelani, William Harkness (Great Ormond Street Hospital for Children, London, United Kingdom)

Introduction: Aspirated intracranial fluid, in the surgical management of intracranial sepsis, may not culture an organism due to the previous administration of antibiotics. Polymerase Chain Reaction (PCR) detection of bacterial DNA does not require viable organisms and is a valuable method for determining the cause of culture-negative sepsis and monitoring response to therapy.

Methods: Samples were analysed using real-time quantitative PCR targeting the pneumococcal *lytA* gene and the number of genome copies/ml of sample determined. The assay is published and validated for use in the clinical microbiology laboratory.

Results: Streptococcus pneumoniae sepsis was diagnosed by PCR in five culture-negative cases comprising: ventriculitis (x3), subdural empyema and meningitis. Serial serum inflammatory markers (CRP and WBC) and number of genome copies were recorded over the duration of inpatient stay for cases requiring surgical drainage of recurrent collections or external ventricular drainage. A significant correlation was demonstrated between change in bacterial load and serum inflammatory markers, reflecting similar changes in clinical state.

Discussion: This is the first report of the use of serial quantitative PCR in monitoring the course of intracranial sepsis secondary to Streptococcus pneumoniae. Further work is required to determine the precise relationship between serum inflammatory markers, clinical state and bacterial load: do changes in one precede the other? Furthermore, a threshold value for number of genome copies in CSF/aspirate samples has yet to be defined. For instance, a higher likelihood of infective recurrence (requiring further surgical intervention) may be predicted based on the bacterial load in a fluid sample.

Session 2. Hydrocephalus 1

Adult outcome of pediatric-onset hydrocephalus

Matthieu Vinchon, Marc Baroncini, Isabelle Delestret (University Hospital, Lille, France)

The outlook of pediatric hydrocephalus has spectacularly improved over the past decades; however, the adult outcome is still poorly documented. Determining the healthcare profile of these patients is important in order to organize the management of this growing population.

We decided to review our pediatric hydrocephalus database for pediatric patients treated for hydrocephalus and followed into adulthood.

Our institution has a virtual monopoly for pediatric hydrocephalus serving a 4 million plus population; the transition to adult care is also managed in the same institution. We reviewed retrospectively patients younger than 18 treated for hydrocephalus since 1980 and followed beyond the age of 20. We reviewed 456 patients, with a mean initial age of 56 months, and followed for a mean of 24.2 years. In 65 patients (24.4%), the last shunt operation occurred after 20 years; 21 of these (5.8% of the total) having never been revised earlier. Fourteen patients (3.1%) could be weaned of their shunt. Twelve patients died in adult age, four (0.9%) of these dying of shunt-related causes. The most prominent sequels were motor (46%) and cognitive (48%); only 82 patients (18%) had no sequel at all. IQ values were ≥ 80 in 54% of studied patients, however schooling was normal in only 42%, and only 33% were employed in the competitive labor market.

Although late shunt-related mortality is low, morbidity is high, many patients require shunt surgery during adulthood.

The transition from child to adult neurosurgery needs to be organized for these vulnerable patients.

The retrograde ventriculo-sinus shunt: treating hydrocephalus pressure and not picture

Hassan El-Shafei (Cairo University, Cairo, Egypt)

Sequel after regularly implanted extracranial shunts; ventriculo-peritoneal or atrial, are familiar to most neurosurgeons. In this work, the clinical and radiological changes are studied to understand the natural sequel after normalization of the intracranial pressure (ICP) when treating hydrocephalus using the retrograde ventriculo-sinus (RVS) shunt.

Material and methods: This prospective randomized study since 2004, includes 45 cases, 15 open craniums, and 30 closed craniums. Pre- and post-operative clinical and radiological pictures are collected over 1 year at a regular schedule and includes head circumference, state of scalp veins, eye signs and radiological CT scan Ivan's ratio, signs of elevated intra-ventricular pressure (IVP), and transcranial Doppler resistive index (TCD RI) measurement.

Results: In patients with open craniums, the eye signs improve and the TCD RI returns to normal values, yet the head continues to grow and the Ivan's ratio is stationary indicating arrest of the hydrocephalic process. In patients with closed craniums, the eye signs improve including the papilledema and the TCD RI returned to normal values. The radiological signs of high IVP disappear and the Ivan's ratio is reduced relative to the duration of pre-operative hydrocephalus; the more chronic the hydrocephalus, the less the reduction in ventriculo-megaly, indications of arrest of the hydrocephalic process.

Conclusion: The results of the RVS shunt conform to the physiological sequel after treating hydrocephalus pressure and not picture, by arresting the hydrocephalic process and normalizing the ICP. Reversal of the ventriculo-megaly depends on the type of hydrocephalus and the age of the patient.

The constant flow ventricular infusion test is a reliable test for shunt malfunction in patients with equivocal clinical and radiological findings

Jenny Sacree, Anna Visca, Richard Edwards, Michael Carter, Ian Pople, Kristian Aquilina (Frenchay Hospital, Bristol, UK)

Objectives: Most episodes of ventricular shunt malfunction present with typical symptoms and changes in ventricular size. Some present with nonspecific symptoms and unchanged radiology. Traditionally, these patients were observed, or underwent overnight intracranial pressure monitoring and/or shunt exploration. In this study, we evaluate the role of the constant flow ventricular infusion test (VIT) in identifying shunt malfunction in this difficult patient cohort and correlate its results with surgical findings.

Design: Retrospective clinical study

Subjects: Patients with equivocal symptoms and radiology, between January 2009 and September 2010. Patients with normal pressure hydrocephalus were excluded.

Methods: Patients underwent constant flow VIT using the ICMp software (Cambridge, UK) by a dedicated hydrocephalus nurse practitioner. Patients with high resistance to cerebrospinal fluid outflow, B waves in the plateau phase or indication of shunt underdrainage on ICMp analysis were surgically explored.

Results: In addition to standard investigations, because of diagnostic uncertainty, between January 2009 and September 2010, 30 shunt revisions (23 < 16 years of age) were preceded by a VIT. Almost all were performed through a separate ventricular access device. None of these patients had a functioning shunt at surgery. Eleven VITs (7 < 16 years of age) were negative; symptoms subsequently resolved over clinical follow up (median 6 months). There was no morbidity related to VIT.

Cranial vault expansion as treatment for recalcitrant debilitating headaches in shunt-dependant patients with functioning shunts: a series of two patients

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Introduction: The brain and cranium normally develop in tandem. Open sutures accommodate the growing brain by responding to increases in brain size, cerebral pulse pressures, and cerebrospinal fluid circulation. Craniocerebral disproportion represents an aberration of this synchrony, resulting in a mismatch between the intracranial contents and cranial vault.

Methods: The authors present a series of two shunted pediatric patients who continued to complain of chronic, debilitating headaches, despite properly functioning shunts and normal ventricular calibers. Based on imaging studies and intracranial pressure monitoring, both patients were diagnosed with shunt-related craniocerebral disproportion. The patients subsequently underwent initial cranial expansion using the Rigid External Distraction (REDII) System (KLS Martin) followed by placement of an intracranial pressure (ICP) monitor. Each morning for 1 week, the skull was distracted a predetermined length while ICPs were monitored for the presence of plateau waves and the patient instructed to keep a "headache log." The distracted distance at which diminution of symptoms and cessation of plateau waves was achieved served as the target for permanent surgical fixation. Long-term follow-up demonstrated lasting resolution of symptoms and postoperative imaging coupled with volumetric analysis revealed that the

intracranial contents assumed the space created by the distractors.

Conclusion: The authors conclude that cranial vault expansion using REDII Systems is a viable treatment option for shunted patient with functioning shunts and craniocerebral disproportion.

Surgical management of the symptomatic, enlarging, isolated fourth ventricle

Yasser Jeelani, Hui-Ju Liu, Ira Bowen, Mark Krieger, J. Gordon McComb (Children's Hospital Los Angeles, University of Southern California, Los Angeles, CA, USA)

Background: A symptomatic, enlarging, isolated fourth ventricle is a difficult problem to surgically manage and is usually associated with multiple co-variables. Although each clinical scenario is unique, we reviewed our institutional series to see what strategies were more effective.

Methods: Under an IRB-approved study, a retrospective analysis was undertaken of patients who met the above criteria during the last decade.

Results: A total of 30 patients were treated. Underlying abnormalities included 18 preterm infants with grade III–IV intraventricular germinal matrix hemorrhage, five with history of posterior fossa tumors, four cases of congenital Dandy–Walker malformation with late presentation, one with congenital hydrocephalus, and one each secondary to meningitis and cerebellar hemorrhage. Primary operative procedures were: shunting, 16/30 patients; open microscopic fenestration, 12/30; and endoscopic fenestration, 2/30. Patients not requiring additional procedures were: shunting, 3/16; open microscopic fenestration, 5/12; and endoscopic fenestration, 1/2. A total of 149 procedures have been done with a median follow-up of 5 years. There were 15 incidences of cerebrospinal fluid (CSF) infections in 96 shunt-related procedures while there were no instances of CSF infections in patients without hardware. During five open fenestrations, all encountered Choroid plexi were excised and none have required another procedure. No new neurological deficits arose from the fenestration procedures.

Conclusions: These patients often require multiple procedures using a variety of operative procedures and techniques. Excision of any encountered choroid plexus is recommended as it may help decrease CSF formation in the area of involvement. Omitting hardware can significantly reduce the incidence of CSF infection.

Tumoral hydrocephalus in children: experience of Taipei VGH, Taiwan (1971–2008)

Tai-Tong Wong, Muh-Lii Liang, Robert Hsin-Hung Chen (Division of Pediatric Neurosurgery, Department of Neurosurgery, Neurological Institute, Taipei Veterans General Hospital, National Yang Ming University, School of Medicine, Taipei, Taiwan)

Purpose: We review our series of tumoral hydrocephalus in children, related managements, and the changes of shunting procedures in the past four decades.

Patients and methods: We perform a retrospective review of patients with tumoral hydrocephalus, from a series of 1,243 cases of primary brain tumors in children with age 18 years, collected from 1971 to 2008. Cases with uncertain records of hydrocephalus were excluded. Types of hydrocephalus occurred at diagnosis or in the whole course of the disease were highlighted and the related shunting procedures were studied. Changes of ratios of ventriculoperitoneal (VP) shunt insertion and endoscopic third ventriculostomy (ETV) for tumors in different locations were evaluated.

Results: In this series, 56.3% of patients developed hydrocephalus. However, 51.4% and 50.2% of patients presented hydrocephalus and obstructive diagnosis at diagnosis. Significant decrease of VP shunt insertion from 85.7% (1971–1980) to 35.3% (2001–2008) was observed. There were increasing ETV procedures for hydrocephalus with obstruction at the aqueduct and fourth ventricle regions.

Conclusion: For the management of tumoral hydrocephalus, VP shunt insertion has been decreased significantly in the past three to four decades. Avoiding VP shunt insertion through radical tumor resection temporal external ventricular drain and/or ETV is feasible and recommended for the management of obstructive tumoral hydrocephalus in children.

Quality and safety of home ICP monitoring compared to in-hospital monitoring in the pediatric population

Morten Andresen, Marianne Juhler, Tina Munch (Clinic of Neurosurgery, Copenhagen University Hospital, Copenhagen, Denmark)

Background: Since its initial introduction, intracranial pressure (ICP) monitoring has been conducted in-hospital using a stationary device severely limiting patient mobility. New mobile and telemetric ICP monitoring systems allow us to monitor patients in situations more closely resembling daily life. To prepare for widespread adoption of these devices, we evaluated patient safety as well as quality of technical data and adequacy for clinical evaluation in ICP monitoring in the home setting versus in-hospital monitoring.

Methods: We prospectively included all ICP monitoring sessions on pediatric patients from June 2007–November 2009. Patients were placed in two subgroups (home or hospital monitoring) based on clinical assessment by the attending physician. We reviewed technical complications as well as clinical curve quality for both subgroups.

Results: Thirty-seven monitoring sessions (home/in-hospital monitoring: 17/20) were reviewed based on evaluation of 34 patients (age 1–16, mean 9.22). Evaluating technical and clinical curve quality, no difference was found comparing percentage of measurement duration with valid curve ($p=0.73$), number of

interruptions ($p=0.44$), or when noting technical complications preventing accurate curve evaluation ($p=0.71$). No clinically detectable complications were encountered.

Discussion: Consistent with earlier results, home ICP monitoring is both safe and clinically useful when used in diagnostically difficult cases where previous procedures have failed, and where the patient's caregiver can adequately observe the patient during the monitoring session. The importance of a competent adult caregiver appear to make home monitoring an option more suited for the pediatric population.

What leads to a 0% infection rate in shunt surgery?

Roberto Jaimovich, Sebastian G. Jaimovich, Sergio Pampin (FLENI Neurological Institute, Buenos Aires, Argentina)

Several factors have been associated with infection in shunt surgery. Among others, number of persons in the operating room, time of the day when surgery is scheduled, shaving, preparation of the skin with different solutions, surgical time, and antibiotic prophylaxis have been issue for debate. We analyzed a continuous series of patients treated for hydrocephalus at three institutions by the same senior neurosurgeon between 1994 and 2010. Two hundred ninety-five patients were treated and 425 surgeries were performed. Ages ranged from neonates to 18 years old. Minimum follow-up was 6 months. Two hundred thirty-three patients underwent a first ventriculoperitoneal shunt surgery. Medical institutions where the surgeries were performed were a general hospital, an obstetrics/pediatrics hospital, and a neurological/neurosurgical center. Surgery was performed at any time during the day. Preoperative bath was routine. Number of persons in the operating room, shaving, skin preparation, antibiotic prophylaxis, surgical technique, draping, and postoperative care were standard, as in every neurosurgical procedure, and are described.

Infection rate for first implantation was 0.43% in the first 6 months and 0.86% in the first year after surgery.

In our experience, whenever a surgical team proposes itself to improve the infection rate in shunt surgery, the goal is reached. No clear-cut factors are involved, however, standard surgical technique should be strictly observed.

Overdrainage-related problems in shunts with gravitational valves—can adjustments end the era of reoperation?

Christian Sprung, Florian Freimann (Neurosurgery, Berlin, Germany)

Objective: After introduction of conventional differential pressure valves for shunting, the clinical course was often deteriorated due to overdrainage. The introduction of the first generation of adjustable valves improved the situation, but could not avoid all complications related to this

complication. Whether the combination of adjustable hydrostatic gravitational devices is capable to treat overdrainage-related complications successfully will be evaluated by an analysis of a proGAV series.

Methods: A series of 250 hydrocephalic patients of different aetiologies shunted including the gravitation-assisted proGAV with a pressure of 5 cm H₂O in the majority of cases was evaluated for clinical and/or radiological signs of overdrainage, two decompressed cases with hydrocephalus were excluded. Depending on severity of overdrainage, we elevated the opening pressure of the valve, sometimes in several steps. Clinical and radiological follow-up until resolution of overdrainage was documented. **Results:** Thirty-nine patients developed primary or secondary signs of overdrainage-related problems during follow-up of 3.3 years. Six patients suffered only hydrostatic headache, 14 slit-like ventricles, 16 cases developed hygroma and three subdural hematoma. Usually, we elevated the opening pressure in steps of 2 or 3 cm H₂O; but with distinct hygroma or hematoma, greater steps proved advantageous. All 39 cases of overdrainage could be treated successfully by adjustments without operation or ligation of the shunt.

Conclusion: Our series proves that also with the use of adequate adjustable hydrostatic valves overdrainage not be fully avoided. But there is the possibility of successful treatment of all overdrainage-related problems without increasing the incidence of underdrainage.

MRI morphometry of the third ventricle in hydrocephalus secondary to intraventricular haemorrhage and aqueductal stenosis and in children without hydrocephalus

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Introduction: The magnetic resonance imaging morphometry and morphological patterns of the third ventricle in children with and without hydrocephalus is not well known. In particular, it is unclear which parameters could be useful in evaluating the optimum ventricular shape and size in the management of hydrocephalus.

Aims and objectives: Using MR imaging, this study aims to perform quantitative and qualitative changes of the third ventricular surface area (TVSA), floor, and lamina terminalis and to report significant variations in the above parameters with different types of hydrocephalus.

Patients and methods: Medical records and imaging of 69 (40 boys, 29 girls) children treated consecutively at our institution were reviewed; M/F=1:4. The median age was 3.9 years (15 days–16 years). SPSS was used for statistical analysis.

Results: Twenty-three children had aqueductal stenosis (AS), 26 intraventricular (IVH) haemorrhage at birth.

Twenty MR images of children without hydrocephalus were used as controls (norm). The third ventricular, lateral (frontal horn, occipital) widths, lamina terminalis/third ventricle floor angle are significantly larger in children with hydrocephalus. No significant difference are noted between AS and IVH groups. After normalization with the biparietal width, the TVSA in the AS group (712 mm²) is significantly larger than control (497 mm²), $p=0.003$ and IVH group (451 mm²), $p=0.006$. There was no statistical difference in TVSA between the IVH and control groups.

Conclusion: The third ventricular surface area is significantly larger in aqueductal stenosis than in controls and IVH. Interestingly, the SA in IVH is smaller than controls but was not statistically different. Other measurements of the lateral and third ventricle do not differentiate between AS and IVH.

Paediatric ventriculo-atrial shunts in the modern era: an institutional experience

David Clark, Dominic Thompson, John Hartley, Derek Roebuck, William Harkness, Owase Jeelani, Jessica Ternier, Aabir Chakraborty (Great Ormond Street Hospital, London, UK)

Objective: Ventriculo-atrial (VA) shunts are commonly used as a second-line treatment of shunt dependent hydrocephalus. It has been the policy at our institution for over 10 years to use VA shunts using a percutaneous technique for insertion of the atrial catheter. We present our results.

Methods: A retrospective analysis of all patients who had VA shunts inserted between June 2000 and June 2010 was conducted using a prospectively updated surgical database and a case notes review. A review of the complete shunt history and the reasons for VA shunting were identified. Complications of VA shunting including the infection rate and shunt malfunction rate were calculated.

Results: Thirty-eight VA shunts were inserted (23 males, 15 females). The mean number of shunt-related operations prior to the first VA shunt insertion was 5.1. Reasons for VA shunting were abdominal infection, CSF malabsorption, abdominal pseudocyst, or perioperative difficulties with insertion. Perioperative complications included one haemopericardium. The 1-, 2-, and 5-year shunt survival rates were 68.4%, 42.1%, and 37.5% respectively. Fifty-four VA shunt revisions were performed in 20 patients. The mean number of revisions was 1.4 (range 0–8). The VA shunt infection rate following primary insertion and following VA shunt revision was 7.9% and 3.7% respectively. There were seven deaths; two were shunt related.

Conclusions: We propose that VA shunting remains a relatively safe, second-line alternative to the placement of a ventriculoperitoneal shunt when this route is unsuitable. Shunt survival rates are comparable to ventriculoperitoneal shunting.

Telemetric long-time ICP home monitoring in pediatric neurosurgery

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Objective: The knowledge of intracranial pressure (ICP) often is the basis of an appropriate neurosurgical treatment. As clinical, fundoscopic, or imaging data alone are often elusive, the exact measurement of ICP is desirable.

Methods: We would like to present our first long-time experiences with a new telemetric intraparenchymal pressure probe (Neurovent®-P-tel, Raumedic AG, Helmbrechts, Germany) whose transducer is placed over the calvaria under the galea. ICP can be monitored via a special telemetric reader, placed over the intact skin. ICP values are stored in a portable computer. The system does not require intensive care unit (ICU) environment but can be used at any ward or even at home.

Results: Since January 2010, the new system was successfully applied in 19 patients (mean age, 16.7 years; range, 2–56 years) without complications. Diagnoses were suspicion of hydrocephalus, shunt dysfunction, endoscopic third ventriculostomy (ETV) failure, pseudotumor cerebri, or craniostenosis. Implantation period of the probe was in mean 65 days (range, 8–209 days). In 11 patients, normal ICP values could exclude pathological ICP and further surgical measures could be avoided; in eight patients, repeated plateaus of raised ICP indicated ETV, shunt operation, or decompressive craniotomy—resulting in a normalization of ICP.

Conclusion: The system was effective for ICP monitoring over a long period outside the hospital, easy-to-handle even for parents at home, reliable over many weeks without drift problems, and highly helpful in surgical decision making or fine adjustment of shunt valves. Its use in difficult cases of suspected ICP alterations and desirable long-term monitoring outside the ICU is recommended.

CSF shunts—20-year series with long-term follow-up of patients with infantile hydrocephalus

Jivko Surchev, Yavor Enchev, Kiril Georgiev, Venceslav Busarski (Medical University, Sofia, Bulgaria)

The cerebrospinal fluid (CSF) shunts are the most common method for treatment of the hydrocephalus. We present a series of 242 cases of infantile hydrocephalus, operated for a period of 20 years (1984–2003). The follow-up was 5–25 years. Three hundred seventy-five revisions are made. The most frequent revisions are in the first 3 months after implantation. The patients without revisions are predomi-

nated, $n=91$ (38%); with one revision, $n=64$ (27%); and with two revisions, $n=34$ (14%). The rest, $n=53$ (21%), are subject of other study. We studied the dependence between the interval to the first revision and the number of the following revisions. We determinate the following “*criteria for survival of the shunt*”, which have evaluation, comparative, and predictive nature as follows:

- Percentage of shunts with only main operation
- Percentage of shunts with only one revision
- Average number of revisions to one shunt
- Percentage of shunts with three or more revisions

Conclusion: Among the youngest patients (up to 3 months old), the percentage of shunts without revisions is the lowest, the total number of revisions is almost two times higher than those shunted at the age of 3–12 months. The most frequent complications are “malfunction of the ventricular catheter” and “malfunction of the peritoneal catheter”.

Shunting at the age over 3 months increases the percentage of “malfunction of the cardiac catheter” almost double compared to those less than 3 months of age. The reversed trend was observed in “malfunction of the peritoneal catheter”, where its percentage is twice decreased.

Session 3. *Epilepsy/Functional*

Epilepsy surgery in pediatric population: study of 129 children with assessment of quality of life

Sarat Chandra, Amit Dagar, Rekha Diwedi, Kapil Choudhary, Achal Srivastava, Chandrashekar Bal, Shailesh Gaikwad, Ajay Garg, Chitra Sarkar, Ashok Mahapatra, Manjari Tripathi (All India Institute of Medical Sciences, Delhi, India)

Purpose: This study aims to assess the outcome of pediatric population operated for drug-resistant epilepsy from a large tertiary care centre in India.

Methods: Retrospective, quality of life (QOL) prospective: pre-operative assessment included inter-ictal electroencephalogram (EEG), magnetic resonance imaging (as per epilepsy protocol), video EEG. Ictal single-photon emission computed tomography (with subtraction) and positron emission tomography was performed when required. QOL scores assessed using the HASS or SSQ for Seizure Severity, Quality of Life in Childhood Epilepsy for quality of life, Child Behavior Check List for behavior, DASSI for Development assessment scale for Indian infants for measuring IQ in Infants, Binet–Kamat (Indian adaptation) test scale for cognition and intellectual ability.

Results: There are 129 patients operated from January 2000 to June 2009 by the senior author (corresponding). One hundred eighteen patients with least 1 year follow-up were included in study. Mean age at surgery was 9.8 (± 4.3) years. In addition, 40 patients underwent quality of life assessment prospectively both at pre- and post-surgery. Mean duration of epilepsy was 5.3 (± 3.3) years. Class I outcome (Engel’s) was seen in 79.5%

patients, class II in 8.6% patients, class III in 10.7% patients, and class IV in one patient. As per surgical procedures, class I outcome in 76%, 87%, and 72% in patients who underwent temporal resection, hemispherotomy, and extra temporal resection, respectively. QOL scores correlated with duration of seizures, epileptic encephalopathy, and outcome of surgery but not with side of surgery, age, and sex.

Conclusions: This study, the largest reported from India, has demonstrated satisfactory results for epilepsy surgery in children.

Clinical assessment score for children with CP: a practical tool to evaluate overall status before and after neurosurgical procedure for spasticity—preliminary data

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Introduction and goals: We have recently applied the “Clinical Assessment Chart for CP patients” (CACCP) in our spasticity clinic in order to obtain a thorough yet simple evaluation form of clinical status and performance of a child with cerebral palsy (CP).

The goals were:

1. To try to differentiate between different subgroups of children with “low performance” (GMFCS 5).
2. To identify which subgroups may benefit from ITB pumps.
3. To establish a clinical scoring that will include several major parameters which may influence the *general well-being* of the child.
4. To identify clinical parameters that may have impact on effectiveness of treatment by ITB.

Patients and methods: Thirty patients with CP were evaluated by the CACCP. The parameters included: communication, eye contact, hand function, ambulation, comfort versus pain, and co-morbidities (visual, hearing, cognitive, behavioral and feeding problems, seizures, dystonia).

Results: Preliminary results show that the CACCP enabled characterization of the patient in a simple but more thorough method than GMFCS. Within the group of “low performance”, mainly GMFCS 5, different subgroups could be identified, deferring by level of communication and interaction with the surrounding world.

Conclusions: several important parameters may influence the overall performance and status of a CP patient, some of which may be relevant for decision making of a neurosurgical procedure. Such scoring enables future follow up, simplified but thorough, of the patient. It may help conduct longitudinal

studies that determine the natural history of untreated patients as well as effectiveness of treatment in operated patients.

Analysis of the effectiveness of intrathecal baclofen therapy on the higher neurological function in a rat cerebral palsy model

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Objectives: Intrathecal baclofen (ITB) therapy is useful in the management of spasticity. In this study, we analyzed whether the therapy improves higher neurological function as well as motor function in an animal model of cerebral palsy (CP)-induced spasticity.

Methods: Sprague–Dawley rats were classified into control, CP model, and CP with ITB groups. In the case of the latter two, rats were placed in a hypoxic condition (8% O₂) for 150 min after ligation of the right common carotid artery at 7 days after birth. A spinal catheter was connected with an osmotic pump filled with baclofen and placed in the spinal subarachnoid space at 5 weeks after birth in the ITB therapy rat. A daily dose of 12 µg of baclofen was continuously administered for 14 days. Walking speed was measured as the parameter of motor function. Number of failures in a radial arm maze test was counted as the parameter of memory function.

Results: The time required for beam walking in the CP rat increased to 4.1 s (2.1 s in control rat) and shortened to 2.5 s in the ITB rat. Number of failures to chose correct arms of the maze in the CP rat increased to 2.27 (0.85 times in control rat) and did not decrease (2.00) in the ITB rat.

Conclusions: Although ITB effectively released spasticity and improved walking ability, higher neurological function did not change. The known amelioration of consciousness status by ITB should be due to indirect effect such as improved relaxation and comfort.

The use of intraoperative ultrasound and ultrasound elastography for the detection of epileptogenic areas

Aabir Chakraborty, Christopher Uff, William Harkness (Great Ormond Street Hospital, London, UK)

Introduction: Focal cortical dysplasias (FCD) and dysembryoplastic neuroepithelial tumours (DNT) are often very difficult to visualise on magnetic resonance imaging (MRI). Intra-operatively, these lesions often look very similar to normal brain. However, these lesions are often stiffer than normal brain; a property exploited by the surgeon to determine extent of resection. Ultrasound elastography (UE) is an objective method for identifying stiff regions within an ultrasound plane. We evaluated the use of UE intraoperatively during resection of these lesions.

Methods: Five patients were recruited into the study. Four patients underwent invasive electrode recording. One patient had no lesion demonstrable on MRI. UE was performed following dural opening and prior to resection. Comparison of UE with the surgical findings and MRI findings was performed. **Results:** There were two cases of FCD, two of DNT and one case where histology was inconclusive. In all cases, there was concordance between UE findings and surgical findings in terms of stiffness compared to brain. In the four cases where a lesion was demonstrable on MRI, the location of the lesion on UE was in concordance with the MRI findings. Furthermore, the lesion brain interface was far better defined on UE compared to MRI. It was possible to define the lesion shape as a wedge originating from the deep white matter in the periventricular area using UE.

Conclusions: Our results suggest that UE in combination with ultrasound is a useful adjunct when resecting these lesions possibly providing visualisation superior to MRI.

Outcome of epilepsy surgery in children with malformations of cortical development

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Objective: This study aims to assess outcome of epilepsy surgery in children with medically refractory epilepsy and malformations of cortical development (MCD), to determine the predictors of outcome.

Methods: Retrospective analysis of pre-surgical, surgical, and post-surgical data was performed in 40 children (2–16 years) with refractory epilepsy and MCD and at least one year post surgery follow-up. Pre-surgical evaluation was done by non-invasive protocol. Intra-operative electrocorticography was performed in 32, neuronavigation in six. Outcome was assessed according to Engel's outcome classification.

Results: Mean follow up was 34 (12–52) months; 20 (50%) were males. The age of onset of epilepsy was less than 2 years in 19 (47.5%). The commonest surgery performed was an extra-temporal resection in 17 (42.5) and temporal resection was done in 15 (37.5%). Functional hemispherectomy was done in six children with hemispheric malformations and two children underwent callosotomy. Pathology showed non-balloon cell type of focal cortical dysplasia in 15 (37.5%), hippocampal sclerosis with focal cortical dysplasia in 8 (20%), glioneural tumors in 6 (15%), and Taylor's type of dysplasia in 4 (10%). At last follow up, 26 (65%) were seizure free and 29 (72.5%) had favourable outcome. After stepwise regression analysis, variables found to be significant ($p < 0.05$) and predicting a favourable outcome were normal IQ, complete resection, Taylor's type of dysplasia while multiple spikes on interictal electroencephalogram and acute post-operative seizures were predictors of poor outcome.

Conclusion: Favourable outcome after epilepsy surgery can be obtained in children with refractory epilepsy and MCD after evaluation with non-invasive protocol. Normal IQ, complete resection and Taylor's type of dysplasia on pathology were predictors of favourable outcome.

“To Be or Not to Be?” The relationship between the clinicopathological diagnosis and neuroimaging in the identification of focal cortical dysplasia type IIB
 Va Elwell, R Gunny, S Jacques, B Harding, W Harkness (Great Ormond Street Hospital NHS Trust, London, UK)

Background: Focal cortical dysplasia (FCD) is a localised derangement of cortical organisation and is associated with medical-refractory epilepsy in children. Recently, the ILEA Diagnostic Methods Committee proposed an international consensus to improve the clinicopathological diagnosis in FCDs. In this study, the role of this novel histopathological classification system coupled with advanced neuroimaging has been evaluated.

Methods: A 6-year retrospective study of children with histologically confirmed FCD type IIB following surgical resection for epilepsy was conducted. All patients had brain magnetic resonance imaging (MRI) including 3D T1 and 3D fluid attenuated inversion recovery volumetric sequences. Analysis was performed by review of medical notes, electronic databases, neuropathological slides, and neuroimaging. MRI scans were evaluated based on a “seven point” classification scheme.

Results: During January 2005–April 2011, 62 children with histologically confirmed FCD underwent neurosurgery. Twenty-four patients met the diagnostic criteria of FCD type IIB. According to our classification, neuroimaging demonstrated subcortical white matter signal change (100%); well-defined margins (87.5%); blurring of gray-white matter junction, abnormal cortical gyration/sulcation, and single lobe involvement (83.3%); “transmantle sign” (70.1%), and apparent cortical thickening (54.2%). Signal intensities on T2- and T1-weighted images were variable (42% hyperintense on T2W and hypo-intense on T1W images, 33% hypo-intense on T2W and T1W sequences).

Conclusion: Characteristic features of FCD type IIB were seen in the majority, but not all patients. Patient selection and complete surgical resection improves long-term outcomes in patients with FCD type IIB. It is important to identify positive clinical and radiological characteristics to stratify patients prior to surgery to aid in their overall treatment.

Title: a pilot study on safety and efficacy of mesenchymal stem cells for children with cerebral palsy

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Background: Cerebral palsy is a group of disorders due to perinatal damage to the brain causing disabilities in motor, learning, and intellectual faculties. So far, they were treated symptomatically for seizures, dystonia, and other symptoms. Mesenchymal stem cells have recently been accredited to repair or regenerate the nervous system.

Methods: Eighteen children ranging from 1 to 17 years were recruited into the study. Parents voluntarily opted for this therapy after approval by institutional ethics committee with an informed consent. These 12 boys and six girls were evaluated clinically, brain magnetic resonance imaging and neurodevelopmental assessments screening for the infections was conducted.

Bone marrow-derived mesenchymal stem cells were transplanted into subventricular zone of the frontal areas through coronal burr holes under short general anesthesia. Post-transplant, they were evaluated every 3 months. The parents were interviewed in detail and all the improvements in the milestones were documented.

Results: The procedure was well tolerated. There were no serious adverse reactions. During the follow up of 1 year, majority of the children showed improvement in their learning capabilities and neuropsychological functions. Few of them have showed significant improvement in motor milestones.

Conclusion: Transplantation of mesenchymal stem cells is safe and the short-term results show encouraging improvement in the clinical deficits. With continued support and training, these children can be made self-learners. However, long-term safety and efficacy can be established by longer follow up and larger studies. This appears to be possible therapy for the future worth exploring.

Complications of intrathecal Baclofen therapy in children. A 12-year experience in Nottingham University Hospital, UK

Harshal Ingale, Ismail Ughratdar, Ahmad Moussa, Samiul Muquit, Michael Vloeberghs (Nottingham University Hospital, Nottingham, UK)

Aim: Intrathecal Baclofen (ITB) therapy has evolved into a standard treatment for severe spasticity. The aim of the study was to review complications of intrathecal Baclofen therapy in children from 1998 to 2011.

Method: Retrospective review of prospectively collected data of 221 patients who had Baclofen pump insertion done by single surgeon during the period of 1998 to 2011. A total of 273 patients underwent Baclofen test dose of which 21 failed to show any benefit and 31 declined the surgery. There were 245 Baclofen pump implants done for 221 patients during the period of study. Eighteen patients had cervical implants due to

previous spinal instrumentations. Since 1999, all pumps were implanted subfascially.

Results: Sixty patients had complications. Of the patients, 24 had their pump revised; 15 (6%) for infection, 7 (3%) for wound dehiscence, and 2 after expiration of battery life. Thirty-two (13%) patients had catheter revised; 16 patients had catheter migration, nine had fracture at connector site, and nine catheters were cut accidentally during spinal surgery (eight)/epidural anaesthesia (one). Eleven patients have died although deaths were not related to Baclofen pump. No adverse effects happened during test dose or refilling of pump. No pump migration has occurred.

Conclusion: Adverse effects were found to be comparable with current available evidence in literature. ITB is effective and safe for management of severe spasticity leading to reduction in pain and improved quality of life. This therapy has adverse effects and hence a robust infrastructure, meticulous attention to prevention of infection and programming errors is essential.

Pre and postoperative neuropsychological evaluation in children with occipitoparietal epileptogenic lesions

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Background: The number of pre- and postoperative neuropsychological studies in children affected by nonprogressive or slowly evolving parieto-occipital lesions is limited, also due to the relatively low proportion of cortical lesions in this area.

Patients and methods: All the children with lesional posterior epilepsy who underwent the removal of a malformative or low-grade tumor at the Pediatric Neurosurgery Unit of the Catholic University of Rome, were enrolled in the study. A full clinical and neuropsychological evaluation, a prolonged ictal and inter-ictal electroencephalogram (EEG) study, and a high-resolution MR study were performed in all cases before surgery; neuropsychological and EEG studies were repeated every 6 months after surgery.

Results: Twelve children were collected. Before surgery, neurological examination was normal in nine cases; three children presented a hemianopia, two of them also presenting a correspondent unilateral spatial neglect, and two prosopagnosia. In all but three cases, a total lesion removal was performed. Histopathology showed in one patient a type 1 Taylor dysplasia; in five, a DNET; in three, a ganglioglioma; and in three, a pilocytic astrocytoma. At a mean follow-up of 5.6 years, nine children were seizure

free (Engel IA), the remaining three, remaining respectively in class IIA (1 case) and IIIA (2 cases). Cognitive development was stable and in the normal range in 10 cases with no variation between pre-surgery and after surgery for what concerns verbal IQ; performance items showed a statistically significant improvement after surgery in 10 of 12 cases.

Role of limited, minimal resections in intractable epilepsy

Sanjiv Bhatia, Prasanna Jayakar, John Ragheb, Ian Miller (Miami Children's Hospital, Miami, FL, USA)

Surgical resections for intractable epilepsy in patients with nonlesional epilepsy or with malformations of cortical development tend to be large and may be associated with neurological sequelae. Use of high-resolution magnetic resonance imaging coupled with multimodality image registration techniques and detailed extraoperative electrophysiological monitoring have helped tailor resections in these patients with improved outcomes.

We present a select group of patients where seizure control was achieved with limited, minimal resections aimed specifically at the presumed epileptogenic area that was outlined using these techniques. This helped preserve visual fields, language cortex, and other eloquent areas.

Material and methods: We prospectively collected eight patients with intractable seizures that underwent minimal resections. Detailed video recordings, ictal and interictal single-photon emission computed tomography (SPECT) imaging, positron emission tomography scan, specialized image coregistration techniques coupled with extraoperative electrophysiological monitoring using subdural and depth electrodes, were used to precisely outline area of resection.

Results: Two of the eight patients had to be reoperated in the immediate postoperative period to extend resections which still remained minimal. One patient developed late recurrence of seizures after 2 years. All these patients had postoperative SPECT scans which were extremely helpful in the management. All the patients have been seizure free with a minimal follow up of 6 months. Cortical dysplasia was the most common cause of seizures.

Conclusion: While detailed electrophysiological analysis is the cornerstone of recognizing area of seizure onset, detailed image coregistration techniques can help further focus the precise area of abnormality, limit the area of cortical resection and associated sequelae.

Gyral mapping as a road map to target epilepsy surgery around the central sulcus: a technical note

Benoit Jenny¹, Jeremy Freeman², Simon Harvey², Wirginia Maixner² (¹University Hospital Geneva, Geneva, Switzerland; ²Royal Children's Hospital, Melbourne, Australia)

The good outcome of epilepsy surgery in children highly depends on the targeted resection of the epilepsy focus, without damaging eloquent areas. The identification of functional cortical regions of the brain is a determinant factor to preserve function. With surgery involving resection near the central region, identification of motor and sensorial cortex is a critical step. Different methods are available to localise these two eloquent regions in adult patients, including functional magnetic resonance imaging (MRI) and pre-operative electrophysiological studies using motor-evoked potential and somatosensory evoked potential. Unfortunately, these functional tests are not feasible in small children. The central sulcus has been shown in many studies to be a constant feature amongst patients and we believe that this information is sufficient to localise the motor and sensorial cortex in epilepsy surgery for children. We report here a series of 10 children aged from 3 months to 7 years who underwent a temporo-parieto-occipital disconnection. In all cases, the identification of the post-central sulcus was based only on anatomical landmarks. To identify the central sulcus during surgery, we used a gyral mapping program reproducing the cortical surface anatomy of the patient based on pre-operative MRI (MRI Cro program). This was used as a road map to identify gyri, sulci and veins once the craniotomy was done and before resection was performed. We believe that the identification of the central sulcus can be based purely on anatomical landmarks, and remains a safe technique for epilepsy surgery in children, when other electrophysiological methods fail.

Session 4. Vascular

Surgical outcome of intracranial aneurysms in children and adolescent

Ashish Pathak (P.G.I.M.E.R., Chandigarh, India)

Introduction: Paediatric intracranial aneurysms are rare entity and their presentation, location, and etiology is different from adults.

Method: Hospital data on paediatric and adolescent patients who were surgically treated between 1992 and 2009 for intracranial aneurysm were analysed. Demography, clinical presentation, details of imaging, and treatment details were reviewed and the outcome evaluated.

Results: There were a total of 69 patients who were less than 20 years old. There was male preponderance (M40/F29). Five patients were less than 10 years of age, the youngest being 3 years old. Majority were between 15 and 18 years (43) while five patients were between 19 and 20 years. Majority were internal carotid artery (ICA) aneurysms (37) most being at the ICA bifurcation (29). Other common locations were ACom in 20, MCA

bifurcation in three, and the rest in isolated different locations. There were three giant and four mycotic aneurysms. There was great variation in the time to ictus versus surgery (0–80 days) with only 22 patients presenting in the first 3 days after ictus. Forty-four patients (64%) were in Hunt and Hess grades I and II at presentation while 25 (36%) were in grades III and IV. Amongst patients, those in grades II and III (7%) had poor outcome (all died), whilst in grades III and IV group (28%) seven had poor outcome (five deaths).

Conclusion: Findings suggest that presentation, behaviour, location of paediatric aneurysms being different from adults. Vasospasm is minimal and the outcome can be good if patient is referred in time.

Surgical treatment of moyamoya disease by encephalo-duro-arterio-synangiosis (EDAS). A study of 52 cases

Trimurti Nadkarni (King Edward Memorial Hospital and Seth G.S. Medical College, Mumbai, Maharashtra, India)

Moyamoya disease is a rare disorder of uncertain etiology that leads to progressive occlusion and narrowing of cerebral vasculature at the base of the brain resulting in cerebral ischaemia.

The angiograms of 52 moyamoya patients were staged using Suzuki and Takaku classification (Fukuyama and Umezu subclassification) and Mugikura staging. A total of 76 cerebral hemispheres were revascularized by encephalo-duro-arterio-synangiosis (EDAS). The side of revascularization was selected based on symptoms of cerebral ischemia and angiographic findings. The follow-up ranged from 3 to 120 months. Post-EDAS angiogram specifically concentrated on exclusive external carotid artery (ECA) injections to note the development of any cerebral collaterals. Postoperative development of collateral vessels from the ECA system was assessed on the external carotid arteriogram following EDAS and classified as "Excellent", "good" or "poor".

Angiographic staging did not strongly correlate with the symptoms. Majority of the patients were in Suzuki stage 3 and in Mugikura PCA stage 2. All patients had bilateral moyamoya disease of which 32 patients had PCA involvement. Eighty percent of the patients showed evidence of vascularization of which 50% were excellent and 30% good.

In general, advanced stage of moyamoya was diagnosed at the time of presentation. Post EDAS, augmentation of blood flow was noted in the preexisting collateral and additional collateral formation was noted in 80% of the patients.

EDAS is a safe and effective procedure for cerebral revascularization in moyamoya disease, it appears to stabilize the progression of stenooclusive changes, but we could not identify regression in any of our patients.

Cerebral aneurysms. A pediatric perspective

Flavio Requejo, Roberto Jaimovich, Rolando Cardenas, Francisco Villasante, Jose Lipsich, Graciela Zuccaro (Pediatric National Hospital Prof. Garrahan, Buenos Aires, Argentina)

Objective: The purpose of this article is to share our experience and some reflections in the management of cerebral aneurysms from the perspective of pediatric neurosurgeons.

Material and methods: Twenty-three patients with 24 aneurysms were managed in our institution in the last years. The median age was 12 years. Every patient had a digital subtraction angiography, at least immediately after treatment.

Results: Of the lesions, 66.66% of the lesions were saccular, 25% were nonsaccular (fusiform and dissecting), there was a patient with a mycotic aneurysm and other with lenticulostriate artery aneurysm. Patients with saccular aneurysms were predominantly males and presented more commonly with intracranial hemorrhage. Nonsaccular aneurysms were dissecting in nature or chronic with intramural thrombus and mass effect, headache was the main symptom. Therapy was tailored for every patient. Six aneurysms were microsurgically clipped, 16 were managed by endovascular techniques, and two were not treated. Surgery was reserved for broad neck lesions to avoid the use of intracranial stents. Because of the size of vessel, we find in little children, the inadequacy of endovascular devices available.

Conclusions: Cerebral aneurysms in children are heterogeneous lesions. Endovascular therapy is a good option in many patients but a long-term follow up is recommendable. There are issues in the management of pediatric aneurysms that have to be considered: the potential growth of cerebral vessels, the physiologic changes in the cerebral circulation, the inadequacy of endovascular devices (created for adult patients) and the lack of experience of many of the pediatric neurosurgeons.

Pediatric subarachnoid hemorrhage: a review of 78 cases from 2001 to 2009

Pankaj Singh, P Sarat Chandra, Asish Suri, Ashok Mahapatra (AIIMS, New Delhi, India)

Introduction: Though subarachnoid hemorrhage (SAH) is a common cause of stroke in adults, it is uncommon in pediatric population. This area has not been much talked about in literature. We are presenting our data from 2001 to 2009 of pediatric SAH.

Materials and method: We have analyzed data of our SAH patients admitted from 2001 to 2009 and all patients with SAH below 18 years are included.

Results: Out of 1,433 patients admitted from 2001 to 2009 of SAH, 78 were of pediatric age group (below 18 years). Out of these patients, 18 had aneurysms, 51 had arteriovenous malformation, two were angioneegative SAH, four had arteriovenous fistula, one had tumour bleed, two had moyamoya disease. Out of 18 aneurysms, one was mycotic aneurysm.

Conclusion: In our institution study, the most common cause of SAH in pediatric population was arteriovenous malformation followed by aneurysms. Pediatric SAH are uncommon compared to adult population but they are to be looked for in case of presentation of stroke like presentation in stroke patients.

Session 5. Trauma

Electrical impedance tomography: a novel method of continuous imaging to improve outcomes in head trauma

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Background: Electrical impedance tomography (EIT) creates an axial image of the brain which, while limited in resolution, eliminates radiation risk, provides continuous imaging, and is exquisitely sensitive to cerebral edema and perfusional changes. We have developed a swine model to investigate these strengths toward human trials for the improved management of head injury, hydrocephalus, and stroke.

Methods: An external ventricular drain incorporating an intracranial pressure (ICP) sensor at its tip is a central electrode in an array including seven circumferential scalp electrodes. Impedance measurements 30 times per second of all combinations create an impedance tomogram where the 50 kHz very low current passes transcranial among electrodes. Six swine were so instrumented. Effects of hyper- and hypoventilation, mannitol, injection of IV blood, and inflation of a parenchymal 1 cm balloon were monitored in real time in anesthetized animals followed by monitoring of circulatory arrest and termination with KCL injection.

Results: The continuous axial tomogram allows immediate detection of physiologic shifts of intracranial hypertension, relief by mannitol, decreased perfusion with IVH, and loss of blood flow with circulatory arrest. Coupled with ICP, a real-time compliance map of the brain is achieved. Evolving mass effect can be observed in real time.

Conclusions: As a method of advanced ICP monitoring, EIT is a promising imaging technology to detect deterioration of the brain related to evolving mass effect from

edema or hemorrhage. EIT can show immediate regionalized or global loss of perfusion. Continuous sensitivity to critical changes for head injury, hydrocephalus, and stroke management appears practical.

Head injuries protocol in children in the Paediatric Emergency Department in Recife

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Introduction: The authors present a protocol for the initial management of brain injuries in children at the Paediatric Emergency Department at Hospital da Restauração (HR) in Recife. This protocol is based on studies conducted in 1999–2000 at the HR, in which 1,103 cases of head trauma in pediatric patients were analyzed.

Materials and methods: The protocol classifies patients into minor (Glasgow Coma Scale (GCS) 15), mild (GCS 14–13), moderate (GCS 12–9), and severe (GCS 8–3) trauma. According to the classification and neurological evaluation, a course of action is proposed. Minor trauma is referred for home observation. Mild trauma requires an observation period at the pediatric emergency department of at least 6 h. Moderate trauma requires immediate CT diagnosis and close monitoring in a trauma unit. Severe trauma should be assessed with a CT scan and then immediately referred to the intensive care unit. The protocol suggests that traumas considered minor and mild, may be attended by pediatricians, while moderate and severe cases require the presence of a neurosurgeon.

Results: From December 1999 until December 2010, 33,869 child victims of head trauma were treated at the HR. The main difficulty encountered in implementing this protocol has been the resistance on the part of the pediatricians, who insist on longer observation periods and systematic neurological evaluation for patients with minor and mild trauma. However, our experience has indicated the importance of this protocol in bringing about greater diagnostic accuracy, reducing the length of hospital observation, and the consequent reduction in costs with traumatized patients.

The prevalence of cerebral ischemia in pediatric severe traumatic brain injury

Llewellyn Padayachy, Graham Fieggen, Anthony Figaji, Jonathan Peter (University of Cape Town, Cape Town, South Africa)

Background: Severe traumatic brain injury (TBI) is a leading cause of mortality in children. Outcome after

head injury is strongly affected by secondary insults, of which cerebral ischemia is a well-recognized contributor; however, the frequency of post-traumatic cerebral ischemia is often debated in adults, and even less is known in pediatric TBI.

Methods and materials: All children (<14 years old) who sustained a severe TBI (post-resuscitation Glasgow Coma Scale (GCS) <8), who were admitted to the intensive care unit and underwent PbtO₂ monitoring at the Red Cross Children's Hospital were included. Hourly recorded data was then used to analyse PbtO₂ values at two specific timepoints, (1) the 24-h post-injury time-point (early) and (2) 72-h post-injury timepoint. In addition, the first 24 h of PbtO₂ monitoring was examined, as well as the 24-h period around the 72-h mark.

Results: A total of 100 children with severe TBI underwent PbtO₂ monitoring between June 2006 and January 2010. PbtO₂ was monitored for 11700 hours. Mean PbtO₂ was 25.8±13.3 mmHg at the 24-h (early) time-point and 33.9±12.7 mmHg at the 72-h time-point. For the whole first 24 h and the 24-h period around the 72-h mark, the mean PbtOs was 27.4±11.7 and 33.0±12 mmHg, respectively.

Conclusion: Different methods for detecting cerebral ischemia are likely to yield different results concerning its prevalence. Results from this study suggest that selection of individual time-points for examination, especially for stable patients, may significantly underestimate the overall frequency of adverse cerebral events after TBI.

Therapeutic potential of cord blood stem cell in brain damage of an animal model

Mohammad Reza Nikravesh, Mehdi Jalali, Hossain Ali Ghaffaripoor, Javad Sanchooli (School of Medicine, Mashhad/Khorasan, Iran)

Neuronal loss is a common feature of many neurological maladies that affect the brain including traumatic brain injury, stroke, and hypoxic ischemic. On the other hand, stem cell therapy is a promising attempt to improve the recovery after these injuries. The present study was carried out on young Wistar rats. At the first, cord blood stem cells (CBSCs) were isolated, and labeled with Bromodeoxyuridine. Animals were subjected to internal carotid artery occlusion (ICA) for 20 min. After that, 2×10⁵ isolated CBSCs were injected into the tail vein of the experimental rats. The control animal group did not receive any injection and intact rats were assumed positive control group. The results were evaluated by the following parameters: behavioral analysis in animals and neurological study, in damaged brain areas especially in caudate–putamen region in experimental and negative control. It was shown that control animals did not return to their initial behavioral

while the experimental groups (CBSCs injection), within 2 weeks after ICA, had parameters like intact rats. In addition, histological study shown that, the size of the damaged (caudate–putamen) region in the control group was larger than in the experimental (CBSCs-treated) group. The positive control did not showed any damage in the caudate nucleus or other region. It is concluded that the CBSCs transplantation is a beneficial influence upon the brain tissue reparation after hypoxic ischemic cell death in hypothalamic area and caudate-putamen region specially.

Pediatric head injury—an initial experience

Shameem Ahmed, Basanta Baishya, Zakir Hussain, Rajesh Barooah, Panchanon Deori Bharali, Anshuman, Naba Borah, Mrinal Bhuyan, Poran Barman (Guwahati Medical College, Guwahati, Assam, India)

Abstract: The outcome of moderate and severe head injury in children at discharge, 3 and 6 months, were investigated in the age group of 1–18 years old, who were admitted to the neurosurgery department. Of 34 eligible children, the quality of recovery was assessed by the Glasgow Outcome Scale (GOS) at 3 months after injury in 29 patients (85% of series) and at 6 months in 24 patients (70% of series). The post-resuscitation Glasgow Coma Scale (GCS) score and pupillary reactivity were predictive of the 6-month GOS. Analysis of the initial computed tomographic (CT) scan revealed that bilateral brain injury with/without midline shift was related to a poor outcome as was the presence of mass lesions. On comparison of age-defined subgroups of patients, it was found that outcome was poorest in the 1–5 years old patients, as reflected by their mortality. Distinctive features of the injuries in the 1–5 years old included subdural hematomas (20%) and hypotension (32%). The most favourable outcome was attained by 6–10 year-old, whereas the GOS distribution of adolescents was intermediate. In summary, the GOS data reflect heterogeneity in the outcome after head injury depending on age group, neurological status, and CT scan findings.

Measuring school function in children after TBI: item generation and item reduction

Shobhan Vachhrajani, Dorcas Beaton, Maureen Dennis, Peter Rumney, Abhaya Kulkarni (University of Toronto, Toronto, ON, Canada)

Introduction: The sequelae of pediatric traumatic brain injury (TBI) pose significant challenges for children as they attempt to reintegrate into school settings. We have previously developed, using qualitative techniques, a concept of school function and we now describe the next steps (item generation and reduction) towards the development of a questionnaire to measure school functioning after TBI.

Methods: Seven focus groups of teachers and rehabilitation professionals were conducted to identify concept-specific observable behaviours and traits, which were converted into questionnaire items. Each item was placed into a putative domain of school function. Using a Delphi approach, 12 of the focus group participants were asked to rate the criticalness of each item and the ease of its observation in the school environment.

Results: Initial transcript review identified 208 potential items. These were placed into the domains of academic competence, cognitive function, psychosocial competence, and physical function. After eliminating redundancy, 168 items were subjected to the Delphi process and 54 of these items were deemed critical to measuring school function by greater than eight of 12 participants. Another 41 items were considered potentially important; in total, 95 items comprise the prototype school functioning questionnaire.

Conclusions: Using qualitative and clinimetric approaches, we have constructed a prototype questionnaire that measures school function in children after TBI. Field testing with teachers of afflicted children will psychometrically validate the concept and will further refine and reduce the number of items. The benefits of using such a disease- and context-specific instrument will provide immeasurable benefits to this vulnerable population.

Outcome of pediatric head injury patients admitted as unknown at a level-I Apex Trauma Centre

Haradhan Nath, Vivek Tandon, Ashok Mahapatra, Deepak Gupta (Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi, 110029, India)

Abstract

Objective: Patients with head injury who are not identified at admission are a challenge to manage and in this backdrop we decided to analyse our data of such pediatric patients for their outcome.

Method: It was a retrospective study conducted at the Level I trauma centre. Twelve consecutive pediatric (<20 years) age group patients whose identities were not known at the time of admission were included in the study.

Result: All 12 patients were male. The road traffic accident was the most common cause of injury (8; 67%). The majority of patients were in the age group of 16–20 years (8; 67%). Mean age of the patients were 16.75±4.45 years. Six (50%) of patients needed surgery and six (50%) were treated conservatively. During the course of hospital treatment, one (8%) patient died, two (16%) had good recovery, and four (33%) were moderately disable. Among the 12 patients, identity of eight (67%) could be ascertained. Seven (58%) of the patients were sent to home with their own relatives, one

(8%) were referred to district hospital, and three (25%) remained as unknown and referred to destitute home for rehabilitation.

Conclusion: Unidentified patients of pediatric age group have better outcome if proper care is provided in time.

Keywords: Head injury, intensive care unit, computerized tomography, Glasgow Comma Scale

Session 6. Pot Pourri

Worldwide survey for health system related to pediatric neurosurgery

Joong-Uhn Choi (CHA Bundang Medical Center, Sungnam, Republic of Korea)

Worldwide survey was done to review the different health system related to pediatric neurosurgery in each country.

Firstly, difference in health insurance system was reviewed in each country. Five out of 15 countries have national insurance system, which included Canada, Italy, UK, Singapore, and Taiwan. National insurance and partly private insurance system have been managed in other 10 countries including USA, Argentina, Brazil, Germany, Russia, Japan, Korea, India, Australia, and South Africa. Full coverage of medical expense was found in Canada, UK, Italy, Brazil, and Australia. But in other countries, medical fee is covered mostly by national insurance and partially paid additional expense by patients 10–30%.

Secondly, medical care system for pediatric patients was reviewed. To have medical treatment at University Hospital or Children's Hospital, patients must meet primary physician first in Canada, Germany, UK, and Australia. But patients may go to University or Children's Hospital directly in other countries. Thirdly, medical fee for treatment of pediatric neurosurgical patients was compared in each country. Total cost of hospitalization for ventriculoperitoneal shunt, endoscopic third ventriculostomy, lipomeningomyelocle, and brain tumor treatment was compared. Total cost of brain tumor treatment was between US\$ 3,500 and 85,000. Variety of medical fee and range of surgeon's fee will be presented.

Lastly, treatment for pediatric neurosurgeons was surveyed. Some pediatric neurosurgeons have basic salary and additional incentives in Singapore, part of India, and South Africa. The monthly salary is fixed in other countries. Range of annual income (before tax) are US\$ 20,000–380,000. Details will be discussed.

Carpal tunnel syndrome and mucopolisaccharidosis

Antonio Bellas, Jose Francisco Salomão, Tatiana Protzenko (IFF-FIOCRUZ, Rio de Janeiro-RJ, Brazil)

Carpal tunnel syndrome (CTS) is rarely seen in the childhood but its incidence increases in specific diseases,

such as mucopolisaccharidosis (MPS). The authors present five patients, with CTS, associated with MPS. In three, the basic disease was type 4 MPS while the other two presented with types 1 and 2 MPS. All patients had bilateral compression of the median nerve and pain was the most important complain. Neurophysiological studies were obtained in all cases, before and after surgery. The median age at surgery was 4.7 years and the mean follow up time was 14 months. All were operated under general anesthesia with orotracheal intubation performed with endoscopic assistance. Surgery consisted of section of the transverse carpal ligament under magnification and usually both sides were operated on the same session. Although no change in the neurophysiological findings has been observed, all patients experienced marked postoperative clinical improvement. The clinical improvement was attributed to a precece intervention (less than 30 days) after the diagnosis

The use of flexible CO2 laser for albumin solder-assisted dural tissue welding

Jayant Menon, Hal Meltzer, Michael Levy (University of California, San Diego, San Diego, CA, USA)

Introduction: Postoperative cerebrospinal fluid leakage is a concern in pediatric neurosurgical patients. We report a novel laser tissue welding technique for watertight dural closure using a flexible wave guide for a CO2 laser and human albumin solder.

Technique: Welding was employed after the dura was re-approximated with non-absorbable running suture. A 9.4–10.6 μm wavelength CO2 laser at 1.5–2 W was used to weld over the suture line with 25% human albumin solder. No cerebrospinal fluid (CSF) leakage was observed after Valsalva to 30 mmHg for 3–5 s.

Results: Patients ranged from 5 to 17 years of age. Welding was completed 12 times in 11 patients over a 9-month period. Patients included one laminectomy for extramedullary tumor. Eight patients underwent posterior fossa craniotomy (six for tumor resection, one vascular malformation, and one Chiari decompression with duraplasty) and two underwent supratentorial craniotomy (one frontal encephalocele resection with preoperative CSF rhinorrhea and one cavernous malformation with iatrogenic durotomy noted beneath the sphenoid wing). One patient had a subsequent CSF leak treated with secondary skin closure. One patient underwent re-operation for residual tumor that revealed the initial weld to be intact. One patient had an epidural abscess requiring re-operation that also revealed the initial weld to be intact, preventing intradural bacterial contamination. Seven patients had postoperative imaging from 7 days to 9 months after surgery. Only one patient's

MRI (s/p Chiari decompression) demonstrated a postoperative pseudomeningocele.

Conclusion: Our preliminary data suggest that laser tissue welding is a promising technique for dural closure in children.

Facing Africa

Safi Ur Rehman (Department of Neurosurgery, MCM Korean Hospital, Addis Ababa, Ethiopia)

There are a variety of neurosurgical pathologies, rarely seen in common neurosurgical practice, including congenital anomalies, tumours, horrendous infections, and untreated hydrocephalus.

Poverty, ignorance, lack of facilities, and inadequate health-care systems are some of the reasons responsible for such tragedies and human sufferings.

It is high time for the world of Neurosurgery, particularly paediatric neurosurgery to pay attention and reach out to these long neglected parts of the world, pay visits, make arrangements, not only to provide help and expertise but to teach and train the local surgical trainees.

Endoscopic endonasal approaches in pediatric neurosurgery

Graciela Zuccaro, Javier Gonzalez Ramos, Carlos Routaboul, Adrian Ratnoff, Martin Guevara (Hospital Nacional de Pediatria Juan P Garrahan, Buenos Aires, Argentina)

Objective: Endoscopic cranial base surgery is a minimal access, highly successful alternative to traditional transfacial, transcranial, or combined open cranial approaches. The collaboration between neurosurgeons and otorhinolaryngologists has resulted in the development of a new field of endoscopic endonasal cranial base surgery. The aim of this study is to describe our experience using an endoscopic endonasal approach (EEA) in different pathologies.

Methods: A purely endoscopic endonasal approach was used to solve different lesions in a series of eight patients. Each patient's record and imaging data was reviewed (pre- and postoperative). All cases were performed collaboratively by a neurosurgeon and an otorhinolaryngologist.

Results: From the eight patients, three were male and five female, with a mean age of 13 years (range, 19–216 months). The histopathological findings were: three Rathke cleft cysts, two sellar arachnoid cysts, one nasotmoidal encephalocele, one craniofaringioma, and one post traumatic cerebrospinal fluid leakage. There was postoperative improvement in all symptoms associated to the pathology. There were no complications. The immediate postoperation was comfortable with no need of nasal occlusion.

Conclusions: Endonasal endoscopic cranial base surgery is a minimal access, highly successful alternative to open transcranial base approaches for specific indications. The main advantages of EEA over the traditional approaches are a better view of the anatomical landmarks, the postoperative comfort of the patient and the reduction of hospitalization time.

A comprehensive approach to the management of birth injuries of the brachial plexus

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Birth injuries of the brachial plexus (aka OBPI) can be a cause for significant life-long morbidity. A thorough clinical assessment, followed by longitudinal follow-up assessments with regularized physical therapy are essential for ideal selection of a management strategy to achieve the best outcome vis-à-vis operative and non-operative means. In November 2009, a multidisciplinary approach was adopted at our institute with the establishment of the Brachial Plexus and Peripheral Nerve Clinic. Here, a team approach involving the neurologist, neurosurgeon, plastic surgeon, physiatrist, and orthopedician has led to more standardized management strategies, thereby optimizing results for all types of pediatric brachial plexus injuries.

Between November 2009 and March 2011, a total of 43 pediatric cases have been evaluated and managed. This included 26 Erb's and Erb's plus palsies and 17 Klumpke's or Erb's–Klumpke's pattern injuries (Louisiana State University Health Science Centre classification). Management of flail hand (Klumpke's or Erb's–Klumpke's injuries) required intraplexal/extraplexal microsurgical repair of the plexus in seven infants. The majority of Erb's and Erb's plus palsies were managed nonoperatively with regularized physiotherapy, including constraint-induced movement therapy, and botox injections for co-contraction release. Tendon release/transfers and derotation osteotomies were used in older children with long-standing deformities and functional restriction of proximal upper limb movements.

In summary, a multidisciplinary clinic is essential to optimize functional outcome in the management of OBPI.

The various aspects of our comprehensive approach including initial clinical and electrophysiological characterization, rehabilitative therapy, intraoperative electrophysiological monitoring, nerve repair strategies, and muscle/tendon transfer strategies will be discussed.

Tuesday October 18, 2011

Session 7. Brain Malformations

Craniopagus twins—a complex history and even more complicated diagnostic and treatment problem—a review of 10 cases of craniopagus twins

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The pediatric neurosurgical and plastic surgery team at the Children's Hospital at Montefiore/Albert Einstein College of Medicine has had the opportunity of reviewing and participating in either the surgical or clinical management of 10 sets of craniopagus twins. This series has been an interesting one to review as it reveals the complexity of this particular disorder along with the social, psychological, and social issues involved in making the decision to undertake a surgical separation. In two cases, the anatomical conjoining of the brains made the complexity of separating the brains too dangerous, entailing significant morbidity if not mortality. In three cases, presenting complex medical problems led to the death of the children before surgical separation could be undertaken. In two cases, social and cultural stigmas lead the families not to undertake any surgical intervention as these children were felt to be a “warning from God” and hence not a possibility for separation. In each case, the children expired due to complications of the conjoined brains and associated medical issues. In three cases, the conjoined twins underwent a successful staged separation leaving to survival of each of the twins. Surgical outcomes in this last group have exceeded what has been previously published. In this presentation, detailed analysis of each case, the issues involved in separation, and the long-term outcomes will be presented.

Enlarging arachnoid cyst: a false alarm for infants

Ji Yeoun Lee, Ji Hoon Phi, Seung-Ki Kim, Byung-Kyu Cho, Kyu-Chang Wang (Seoul National University Children's Hospital, Seoul, Republic of Korea)

Background: The natural history of arachnoid cyst (AC), especially regarding change in size is an unsettled issue. This brings confusion in deciding treatment strategies of incidental, asymptomatic AC in children, especially since the risk of

surgery is not negligible. The present study aimed to elucidate the change in size of AC in young children.

Method: Patients with documented AC in brain imaging from 2000 to 2008 were included. A total of 87 patients were included in the study and were divided into four different age groups (group I, 0–0.5 years old; group II, 0.5–1 years old; group III, 1–3 years old; group IV, 3–5 years old). The temporal changes in size of the AC on consecutive images were measured and plotted as graphs. The natural progression of AC in size was assessed by evaluation of the pattern of each graph.

Results: Eighteen patients showed enlargement of cyst size during follow up and 11 of them were in group I. Furthermore, 14 patients were less than 1 year old. None of the patients in group IV showed enlargement during follow up. Eleven of the 18 patients were not operated on; and in seven patients, increase in size seized. The remaining four untreated patients had shown asymptomatic increase in size at last follow up. Statistical analysis revealed age at diagnosis to be the only significant factor affecting cyst enlargement.

Conclusion: Asymptomatic size enlargement of AC in children in infancy may not be an indication for urgent surgical intervention. Close follow-up is a viable option for such children.

To open or not to open: is duraplasty necessary for treatment of syringomyelia and Chiari malformation?

Robert Keating, Kerri Thorn, John Myseros, Amanda Yaun, Suresh Magge (Children's National Medical Center, Washington, DC, USA)

Introduction: Long considered controversial in the treatment of Chiari malformation, the extent of posterior fossa decompression has involved bony decompression alone versus the efficacy of more extensive procedures including duraplasty, particularly in the setting of syringomyelia. In an attempt to reduce surgical morbidity, comparison of extradural (bone only) vs. intradural (duraplasty) decompressions was made with respect to treatment of associated syrinxes. The decision to extend surgical intervention to an intradural exposure has recently been guided by the expanded use of intraoperative ultrasound.

Methods: Over 16 years, 141 patients underwent surgical treatment for Chiari malformation at CNMC. Within this group, 55 patients (39%) manifested syringomyelia (33% asymptomatic). This cohort was comprised of 16 (29%) patients [10 male/6 female: 11.4 years old at time of surgery (6.6–16 years old)] undergoing simple bony-only decompressions vs. 39 (71%) patients [19 male/20 female: 9.06 (1.6–28 years old)] receiving duraplasties. Postoperative follow-up (serial magnetic resonance imaging) for bone-

only decompression was 11.1 m (3–24 m); duraplasty was 35 m (2.5–154 m).

Results: Syringes with bone-only decompression improved 10/16 (63%) versus 25/37 (68%) for duraplasty. Syringes were stable in 4/16 (25%) and 9/37 (24%) and worsened in 2/16 (12.5%) and 3/37(8%), respectively. Eleven patients (31%) of the duraplasty group required additional surgery (five ventriculoperitoneal shunting, four syringosubarachnoid shunting, and two posterior fossa decompression + stent). Overall morbidity was 28% in the duraplasty cohort (cerebrospinal fluid collections) and no discernable morbidity was seen in the bone-only group.

Conclusion: Introduction of intraoperative ultrasound has permitted tailoring our extent of posterior fossa decompression and reduced our need to open the dura resulting in less surgical morbidity. Despite limited numbers/follow-up, this appears to offer similar efficacy with respect to outcome of the associated syringes and thus should be considered a viable option for this group of challenging surgical patients.

Success of pure neuroendoscopic technique in the treatment of Sylvian arachnoid cysts in children
Hakan Karabagli¹, Völkcan Etus² (¹Selçuklu Faculty of Medicine, Department of Neurosurgery, Konya, Turkey; ²Kocaeli University Faculty of Medicine, Department of Neurosurgery, Kocaeli, Turkey)

Neuroendoscopic approaches to Sylvian arachnoid cysts (SAC) constitute an alternative treatment option to craniotomy for fenestration and shunting procedures. In this study, the authors discuss their experience on pure neuroendoscopic technique in the treatment of SAC in children.

The results of treatment of 20 children (range of age between 7 months and 17 years) with Galassi type II (*n*, 5) or III (*n*, 15) SAC who were subjected to pure neuroendoscopic fenestration procedure were presented. It was possible to perform the cysto-cisternostomy endoscopically in all children with several stomies. Three of the cases required repetition of the operation and two cases required “cysto-peritoneal-shunt” implantation. There was one minor complication in a patient who had an asymptomatic postoperative subdural effusion, which resolved spontaneously. Resolution of symptoms was achieved in all 18 cases, in which the neuroendoscopic procedures succeeded. Of those 18 cases, ten showed a reduction in cyst size. The mean follow-up period was 53 months. The site of the opening was between the optic nerve and the carotid artery in 19, between the carotid artery and the oculomotor nerve in 17, and below the oculomotor nerve in seven. The stomies were enlarged in all cases using the double balloon. Our results suggest that “pure neuroendoscopic” approach can be used safely in the management of SAC in children.

We recommend at least two fenestration sites for an effective marsupialization of the cyst within the basal cisterns. In pediatric cases, the use of a small diameter endoscope allows to reach safely the planned target areas.

Chiari malformation type 1 associated with complex multi-suture craniosynostosis

Nobuhito Morota, Hideki Ogiwara, Masahiro Joko (National Center for Child Health and Development, Setagaya-Ku, Tokyo, Japan)

Objective: The characteristics of Chiari malformation type 1 (CM1) associated with complex craniosynostosis (CS) was evaluated.

Methods: Data of 66 pediatric craniovertebral junction lesions confirmed 21 CM1 who underwent foramen magnum decompression (FMD) between March 2003 and March 2011. Association of complex CS was identified in seven patients (CS group), age ranged from 1 month to 2 years old. Strategy of surgery together with skull base morphological evaluation was compared with four CM1 under 6 years old (no-CS group).

Results: In CS group, all had bilateral lambdoid synostosis. Respiratory insufficiency was observed in four and delayed motor development in all. Ventriculomegaly was present in all and four had VP shunt and two endoscopic third ventriculostomy prior to FMD. FMD preceded cranial expansion in two with severe respiratory failure. FMD was technically more demanding by distorted bony anomalies. Cutting the outer membrane of dura was applied as dural plasty in six and the one had bony decompression only. Two patients with progressive syringomyelia received re-operation 8 and 6 years after the first one. In no-CS group, clinical presentations were typical as CM1. Tonsil coagulation and dural plasty were performed in three and one had bony decompression only. There was no mortality or morbidity in both groups. The clivo-dental angle was larger in CS group, suggestive of vertically elongated skull base.

Conclusions: CM1 associated with complex CS forms unique subgroup in CM1 among young children. Surgical strategy and timing of surgery should be individualized based on associated hydrocephalus, skull deformity and respiratory condition.

Cerebral dysgeneses and pediatric Chiari type I malformation

Marcelo Galarza^{1,2}, Juan F. Martinez Lage^{1,2}, Sandeep Sood^{1,2} (¹University of Murcia, Murcia, Spain; ²Children Hospital of Michigan, Detroit, MI, USA)

Objective: This study aims analyze the role of the association of cerebral dysgenesis in a series of pediatric patients with Chiari I malformation.

Methods: We reviewed the medical records of 20 patients (11 boys and nine girls, mean age 7.2 years; range, 2–16 years) out of 60 cases tested for Chiari I malformation (CM1) who had an associated cerebral dysgenesis. Symptoms of CM1 evolved over a mean of 12 months (range, 3 months–4 years). Syringomyelia was present in five cases. All patients underwent a posterior fossa decompression.

Results: We categorized associated cerebral dysgeneses as *Major* ($n=13$) when the anomaly was likely responsible for the Chiari development or *Minor* ($n=7$) when the anomaly was liable to be independent of the Chiari with respect to causation. Disclosed dysgeneses included: congenital hydrocephalus ($n=11$), cervicomedullary kinking ($n=5$), focal cerebral heterotopia with epilepsy ($n=4$), partial agenesis of corpus callosum ($n=4$), hypoplastic brain stem ($n=2$), holoprosencephaly ($n=1$), and subcortical dysplasia in the context of neurofibromatosis type I ($n=1$). Other malformations included: subcortical hamartoma associated with neurofibromatosis type I, craniofacial dysmorphism secondary to Noonan syndrome, congenital occipital plagiocephaly, OS odontoideum, craniofacial cleft, juvenile rheumatoid arthritis with platybasia, and osteogenesis imperfecta with bathrocephaly and scoliosis.

Conclusion: Cerebral dysgenesis in CM1 patients should not be considered merely as an incidental finding; instead, this may indicate that Chiari I malformation may constitute a complex but continuous spectrum of clinical manifestations.

Management of ruptured intracranial arachnoid cysts with symptomatic subdural fluid collections

Jeffrey Pugh, Jenny Souster, Vivek Mehta (University of Alberta Hospital, Edmonton, Alberta, Canada)

Arachnoid cysts are relatively common findings that generally do not require surgical intervention. Associated subdural hematomas (SDhe) or hygromas (SDhy) are rare. The pathophysiology and best treatment in these cases is poorly understood, owing to the uncommon occurrence of cyst rupture.

Retrospective chart review from August 2001 through February 2010 of patients presenting with neurologic deterioration secondary to ruptured intracranial arachnoid cyst, clinical presentation, treatment, complications, and outcome were evaluated.

Fourteen children (12 male, two female; age 17 months to 18 years) presented with subdural fluid collections with associated arachnoid cyst. Eleven of 14 (79%) were left sided. In 13 (93%), a clear history of trauma preceded neurologic deterioration. Average latency between head injury and clinical presentation was 5 weeks (1 day to 4 months). An equal number of children had an associated subdural hygroma (7) or subdural hematoma (7). Clinical

presentation included progressive headache (93%), emesis (50%), focal neurologic deficit (21%), and decreasing level of consciousness (7%). Six patients (four SDhe and two SDhy) required simple evacuation of the subdural collection as their only surgical intervention. Five patients, all with an associated subdural hygroma, required cyst fenestration. Two patients with associated subdural hematomas failed cyst fenestration. Only one patient has required a permanent cyst–peritoneal shunt.

Best management of patients with symptomatic subdural fluid collections and associated arachnoid cysts is uncertain. Simple drainage of the fluid collection was effective for half of our children. In patients with subdural hygroma, cyst fenestration was successful in all cases when simple drainage failed.

Session 8. Craniofacial

Near infrared spectroscopy (NIRS) analysis of cerebral perfusion and oxygenation during craniostomy surgery

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Introduction: Focal pressure-related changes in brain perfusion and metabolism are discussed in single-suture craniostomy and brachycephalic cases (bicoronal synostosis). Raised intracranial pressure levels could be measured in some cases. In order to find possible loco-regional brain tissue changes during plastic surgery, we investigated oxygenation and perfusion parameters using non-invasive near-infrared spectroscopy (NIRS) probes.

Methods: Twenty-two consecutively operated cases (mean age 7 months) with single-suture craniostomy were prospectively investigated using a NIRS probe (LEA[®], O2C, Wight Light 500–800 nm, laser NIR). Measurements for SaO₂, rHb, flow velocity, and blood flow of the bilateral frontal, temporal and parietal cortex were taken transosseously (prior to decompression) and directly after decompression as well as 15 and 30 min after decompression epidurally.

Results: Twenty-two patients with scaphocephaly (11), trigonocephaly (6), anterior plagiocephaly (3), and brachycephaly (2) were investigated. SaO₂ was improving in all patient groups, showing the highest levels in the temporal region. RHB improved in scaphocephalic, trigonocephalic, and brachycephalic patients. Flow velocity was increasing during surgery in some scaphocephalic, trigonocephalic, and brachycephalic children. Again, highest values were

found in the temporal region and in brachycephalic patients also in the frontal and parietal cortex.

Conclusion: These preliminary results of a new technology for brain tissue oxygenation and blood flow measurements suggest a regional compromise of brain metabolism in patients with craniosynostosis.

Endoscopic-assisted strip craniectomy for non-syndromic sagittal synostosis

Faizi Siddiqi, Robert Bollo, John Kestle (Primary Children's Medical Center, University of Utah, Salt Lake City, UT, USA)

Objective: This study aims assess the safety and efficacy of endoscopic-assisted strip craniectomy for non-syndromic sagittal craniosynostosis.

Methods: We retrospectively reviewed clinical and radiographic data from 59 consecutive patients with non-syndromic sagittal craniosynostosis treated over a 42-month period with endoscopic-assisted strip craniectomy. A standardized surgical technique was employed by a single craniofacial team. Postoperatively, all patients were placed in a post-craniectomy helmet for up to 12 months. Quantitative assessments of pre- and postoperative cephalic index and detailed analysis of perioperative variables were recorded.

Results: Mean age at surgery was 3.0 months. Mean operative time was 76 min. Average blood loss was 53 mL. Nine patients (15.3%) required intraoperative blood transfusions, averaging 71 mL and five patients (7.2%) required post-operative transfusions, averaging 94 mL. None required conversion to an open approach. One patient sustained a dural tear which was repaired without cerebrospinal fluid leak. There were no dural sinus injuries, no patient required reoperation, there was no mortality. The mean hospital stay was 2.1 days. The mean cephalic index was 0.71 preoperatively, 0.80 ($N=44$ of 49 possible) at 6 months and 0.80 ($n=29$ of 42 possible) at 1 year.

Conclusions: We demonstrate that endoscopic-assisted strip craniectomy is a safe and effective technique for non-syndromic sagittal synostosis and allows intervention at as early as 6 weeks of age with minimal risk and acceptable Cephalic index at 6 month and 1 year follow-up. Longer follow-up with more patients is required to ultimately determine if this approach is as durable as the open cranial vault procedure.

Syringomyelia in children with syndromic craniosynostosis and Chiari malformations: discriminant factors in a series of 134 children

Guirish Solanki, Desiderio Rodrigues, Ravi Vemaraju, Deb Roy, Paul Davies, Martin Evans, Nicholas White, Hiroshi Nishikawa, Stephen Dover (Birmingham Children's Hospital, Birmingham, UK)

Objectives: Chronic hindbrain herniation in children occurs in the Chiari malformations, syndromic craniosynostosis, lumbo-peritoneal shunts, and posterior fossa tumours. Thirty percent of paediatric Chiari malformations may develop syringomyelia at some stage. This may be associated with neurological deterioration. We aim to identify discriminant factors associated with syrinx formation in children with chronic tonsillar herniation.

Patients and methods: One hundred thirty-four cases and 32 age- and sex-matched controls were reviewed. Besides morphometric measurements, gender, age, tonsillar herniation, cerebrospinal fluid flow/presence at foramen magnum, surface area of foramen magnum, and volume of posterior fossa were evaluated and statistically analysed.

Results: Of the total 134 cases with chronic tonsillar descent, 57 cases had a syrinx on their radiological imaging. Discriminant analysis identified various factors associated with syrinx formation. Age, amount of tonsillar descent, ratio of the sagittal diameter of foramen magnum to posterior fossa height, and the ratio of the clival angle to the foramen magnum angle were statistically significant variables which could help predict syrinx formation in children with chronic tonsillar herniation. Syringomyelia was associated with age >8 years, tonsillar hernia >12 mm, a ratio of the sagittal diameter of the foramen magnum (FM) to the posterior fossa height of >0.6 and the ratio of the clival angle to the FM–Clivus angle of <0.8.

Conclusions: In this study, we have seen a continuum of morphological changes suggesting the hindbrain hernia from any cause may have a similar end point: syringomyelia.

It may be possible to identify patients who are at a higher risk of developing a syrinx.

Troubleshooting distraction osteogenesis for craniosynostosis

Takuya Akai, Hideaki Iizuka, Yasuo Sasagawa, Shunsuke Shiraga, Shigehiko Kawakami, Masanobu Yamashita (Kanazawa Medical University, Kahoku, Ishikawa, Japan)

We reported that distraction osteogenesis is less invasive due to less bleeding and a shorter operation time compared to conventional cranioplasty (Pediatr Neurosurg 2006). This method can give greater skull advancement and less wound trouble compared to conventional cranioplasty and would be especially suitable for syndromic craniosynostosis. We analyzed our patients treated by distraction osteogenesis and tried to identify the technical troubles and difficulties, and how to avoid or minimize these problems.

Patients and results: We operated on 18 patients by distraction osteogenesis between the years 1999 and 2011. During the treatment course, we had several technical

troubles, as follows; (1) dural laceration at the craniotomy, (2) skull fracture at sphenofrontal suture or coronal suture, (3) device dislocation, and (4) wound trouble.

Discussion: (1) We have to be careful to avoid dural damage at the placement of the burr holes. (2) We have to confirm the complete craniotomy by identifying the pulsation of the bone flap. The craniotomy line should be placed forward of the coronal sutures. (3) For patients younger than 2 years old, we recommend using a clamp type distraction device. (4) We have to cut the shaft short enough to prevent the tip from exerting pressure to the scalp from beneath.

Conclusions: Distraction osteogenesis has several advantages as a treatment modality for craniosynostosis. However, during the craniotomy and distraction, we need to pay special attention to the aforementioned points to avoid problems. These are problems that would not happen with conventional cranioplasty.

Clinical presentation and surgical therapy of multiple suture craniosynostosis

Masato Nagasaka, Mihoko Kato, Hirokatsu Osawa (Aichi Children's Health and Medical Center, Obu, Aichi, Japan)

Background: Multiple suture craniosynostosis is reported to be uncommon, but it often accompanies with features of raised intracranial pressure and/or developmental delay. However, early diagnosis in infants may be difficult because of its atypical cranial shape. So, our experiences are presented.

Materials: Between 1984 and 2010, 118 cases (44 females, 74 males) were operated on by the senior author. Of these, 87 (74%) were nonsyndromic and have been classified as: scaphocephaly, 30; brachycephaly, 12; plagiocephaly, 10; trigonocephaly, 5; lambdoid synostosis, 3; and multiple suture craniosynostosis, 27 (23%; 10 females, 17 males).

Results: (1) Chief complaints on admission: developmental delay, 18; and cranial and/or facial deformity, nine. (2) Suture involvement: sagittal and metopic, three; sagittal and uni-lambdoid, one; sagittal and bi-coronal, seven; sagittal and bi-lambdoid, four; sagittal and uni-coronal and uni-lambdoid, one; sagittal and metopic and bi-coronal, four; uni-coronal and sagittal and bi-lambdoid, two; holocalvarial, five. (3) Age at first operation: average 38.6 months, median 34.4 months. (4) Type and number of operations: the first operation; five conventional, 22 distraction osteogenesis including six multi-directional cranial distraction osteogenesis (MCDO). Ten patients needed a second operation. The second operation: three conventional, seven distraction including one MCDO. (6) Post-operative follow-up duration: average 89.1 months, median 70.5 months.

Conclusion: In our experiences, majority of multiple suture craniosynostosis were diagnosed in the children with

developmental delay. As for the surgical therapy, cranial remodeling and decompression by distraction osteogenesis including MCDO may be promising for the toddlers and the older children.

Mercedes Benz pattern craniosynostosis: diagnosis, management and outcome

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Background: "Mercedes Benz" pattern craniosynostosis caused by bilateral lambdoid and sagittal synostosis (BLSS) is a rare disorder, characterized by step-like deformity of the occiput, anterior turricephaly and mild brachycephaly. Associated anomalies include Chiari I malformation (CM), hydrocephalus and cranial venous anomalies. Surgical management typically comprises synostectomy and foramen magnum decompression (FMD). Outcomes after surgery are poorly described. The purpose of this study was to review the results of early FMD and synostectomy on hydrocephalus, CM and skull shape in BLSS.

Methods: We retrospectively reviewed one centre's experience with BLSS with respect to presentation, associated developmental delay, hydrocephalus, CM, anomalous venous drainage of the brain, management strategy and the outcome.

Results: Five children with BLSS were managed over 25 years. Four underwent single stage synostectomy and bony FMD between 3 and 6 months. Four had CM at presentation. At follow-up (1–9 years), all had asymptomatic tonsillar herniation below the foramen magnum. One developed asymptomatic syringomyelia. Two of four with venograms had cranial venous anomalies, which did not create problems during surgery. None of three patients with ventriculomegaly required ventriculoperitoneal shunting. Head shape in three of the four operated patients appears normal, although two had prominent frontal bossing in early childhood, which progressively became less obvious. One child, who is a year old, still has prominent frontal bossing.

Conclusions: Early synostectomy and bony FMD is a safe surgical option for BLSS and may stabilize CM and hydrocephalus and normalize head shape. MRI follow-up for development of syrinx is advisable.

Cranio-facial resections for skull base lesions in pediatric patients

Manmohan Singh, Vipin Gupta, Suri Ashish, Rakesh Kumar, Alok Thakker, Mahapatra Ashok Kumar (All India Institute of Medical Sciences, New Delhi, India)

Introduction: The lesions involving anterior and lateral skull base with intracranial and nasopharyngeal extension pose surgical challenge for their resection. Craniofacial resections are used for complete and safe excision of these extensive lesions.

Material and methods: Twenty-three patients who underwent craniofacial resections were reviewed retrospectively. Cases were analyzed with respect to age, sex, clinical presentation, extent of the lesion, pathology, surgical approach, peri-operative complications and their management, mortality, follow-up, and outcome.

Results: There were 23 patients in this study. There were 22 males and one female. The commonest tumor resected using craniofacial resection was juvenile nasopharyngeal angiofibroma in 20 patients followed by chordoma, fibrous dysplasia, and anterior nasoethmoidal encephalocoele in one patient each. The surgical approaches used were transbasal approach combined with lateral rhinotomy and maxillary swing for anterior skull base lesions. For lateral skull base lesions, temporal craniotomy along with orbitozygomatic osteotomy combined with maxillary swing or lateral rhinotomy were used for excising the lesions. All patients of juvenile nasopharyngeal angiofibroma underwent pre-operative embolization. There was one postoperative death. Complications included new cranial nerve deficits in four patients; wound infection in one patient, CSF leak in two patients, meningitis in one patient, and palatal wound gap in one patient. Eighty percent of the patients in whom follow up was available did not have any residual or recurrent disease.

Conclusion: Combined craniofacial resection is a safe and effective approach for patients with extensive lesions of anterior and lateral skull base with good cosmetic results.

Endoscopy-assisted strip craniectomy for syndromic and complex multisutural nonsyndromic craniosynostosis

Erik van Lindert, Wilfred Borstlap (Radboud University Medical Centre Nijmegen, Nijmegen, Netherlands)

Objective: Endoscopy-assisted strip craniectomy (EASC) followed by molding helmet therapy is increasingly recognized as a good surgical therapy for scaphocephaly in children <6 months, but not equally accepted for other types of craniosynostosis. Since this surgery is extremely well tolerated even in the very young (<2 months) and cranial remodelling in a later phase not impeded, we started to perform this surgery also in more complex cases of craniosynostosis in order to at least prevent progression of the deformity and thus simplify definitive remodelling, but also in the hope to give a definitive treatment.

Material and methods: From September 2005 until March 2011, 55 EASC for craniosynostosis were performed. Of these, five were on craniosynostosis syndromes and

three on complex multisutural nonsyndromic patients. The syndromic cases were three Apert syndromes and two Muenke syndromes. Surgery was performed between 6 weeks and 6 months of age on average of 3.5 months.

Results: The only complication in these cases was a single dura perforation. The average blood loss was 40 ml. One patient received a blood transfusion. The cosmetic results were considered to be satisfying in the syndromic cases and excellent in the nonsyndromic cases by both the parents and the surgeons. One Apert patient developed papiledema 2 years after surgery and required further treatment.

Discussion: Our results are preliminary since follow-up is very short and numbers are very small. Nevertheless, these results are promising and therefore, we will continue to explore the use of EASC for syndromic and complex craniosynostosis.

Factors affecting outcome in patients undergoing corrective surgery for craniosynostosis

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Background: Literature is scarce on factors responsible for blood loss, blood transfusion, hospital, and intensive care unit (ICU) stay during repair of craniosynostosis. The purpose was to identify these factors, which directly affect the outcome of craniosynostosis surgery.

Methods: Data for all patients who underwent corrective surgery for craniosynostosis from June 2000 to June 2010 were collected. Detailed review of records pertaining to preanesthetic evaluation, intraoperative and postoperative course was done. The correlation between different variables was evaluated using Spearman's rank correlation. A multivariable analysis was performed to evaluate associations between elements of intraoperative management and the clinical outcomes. The difference in medians of blood loss, blood transfusion, postoperative blood transfusion, ICU stay, hospital stay between the categories syndrome, type of surgery, type of induction, type of recovery were found using Wilcoxon rank sum/Kruskal–Wallis test syndrome. The p value <0.05 is considered statistically significant.

Results: Ninety-five patients underwent corrective surgery for craniosynostosis from June 2000 to June 2010. Blood loss was significantly associated with age, weight ($p<0.01$), duration of surgery ($p<0.001$), syndromes ($p<0.02$), and type of surgery ($p<0.001$). Hospital stay was significantly associated with type of surgery ($p<0.02$) and number of postoperative complications ($p<0.001$).

Conclusion: Short surgical duration and avoidance of modifiable factors may contribute to a low complication

rate and better outcome in patients undergoing craniostylosis surgery.

Volume measurements after extended strip and total calvarial remodelling for scaphocephaly

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Debate continues concerning the timing of surgery to correct scaphocephaly. Although early extended strip is less invasive, adversaries are concerned that remaining growth potential is not sufficient to achieve adequate intracranial volumes.

Intracranial volume measurements are usually performed on CT. To avoid radiation, we performed volume measurements on 3D surface imaging, taking the volume above a plane running through the tragus and the outer cantii. McKay et al. (2010) reported a good correlation between measurements on 3D surface imaging and intracranial volume on CT (Pearson, >0.91 ; $p < 0.001$) with a correction factor of 1.34. We correlated our volume measurements on 3D surface imaging to intracranial volumes in 10 patients and found a correction factor of 1.34 as well (SD 0.09). Interrater and intrarater reliability were high: 97% and 98%, respectively.

We included 51 patients with scaphocephaly who had 3D surface pictures, pre- and postoperatively up to 4 years. Extended strip craniotomy with outfracturing of the parietal flaps was performed in 34 patients at a mean age of 5.4 months; 17 patients, who presented after the age of 5 months, underwent total calvarial remodeling at a mean age of 10.5 months. Both patient groups were comparable as the preoperative cranial volume was 2 SD above norm in both groups.

We found no difference in postoperative cranial volume, defined by 3D surface imaging, between early extended strip and late total cranial vault remodeling. In all techniques, cranial volume remained above the norm but growth paralleled normal volume growth (Lichtenberg).

Spiral osteotomy in cranial augmentation and modeling for craniosynostosis

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Introduction: Several surgical techniques and procedures used for remodeling cranial bone have been previously described. These different methods are occasionally modified or combined in an attempt to provide optimal results. We present a modification of the classical radial osteotomy with the spiral technique to increase the width of the cranial vault

Patient and methods: We report the clinical course of 15 patients treated surgically with different types of cranial malformation due to craniosynostosis between August 2009 and January 2011. There are six patients with sagittal, five coronal, and four with two or more cranial sutures affected including three cases of Kleeblattschädel malformation.

Results: In all cases, the spiral osteotomy technique was used, mean age of the children were 10.8 months (6–30 months) at the time of surgery; the spiral bone flap was maintained with absorbable fixation plates and screws in all the frontal and orbital advancement and in some cases of parietal and temporal remodeling.

Conclusions: In all cases, this technique allows immediate correction of the cranial deformity increasing the vault convexity. This procedure also provided the resolution of cerebral compression by increasing intracranial volume.

Results of craniosynostosis surgery about 153 cases

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Background: The authors demonstrate through this study the superiority of fronto-orbital advancement and free flaps in the treatment of craniosynostosis in comparison to techniques of simple suturotomy.

Material and methods: One hundred fifty-three cases of craniosynostosis were treated in the Department of Neurosurgery Hôpital des Spécialités O.N.O between 1987 and 2010. They were 32 syndromic malformations (10 apert, 15 crouzon, four clover leaf skull, and three complex malformations) and 121 nonsyndromic (45 plagiocephaly, 14 oxycephaly, 47 scaphocephaly, and 15 trigonocephaly).

Results: The mean age of these children at time of surgery was 30 months (range, 3 months to 13 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. An electroencephalography and ophthalmologist evaluation was done in all the cases. The surgical treatment consisted in simple suturotomy (six cases). Free parieto-temporal bone flaps in scaphocephaly (44 cases) and technique of fronto-orbital advancement in 103 cases. These last techniques permitted very good functional and esthetical results with a mean follow-up period of 10 years. Some post-operative complications were observed with three deaths due to haemorrhage and to a complex cardiac malformation. Four cases or recurrence after suturotomy were observed, with one case of plagiocephaly who benefited from fronto-orbital advancement with good esthetical result.

Conclusion: Craniosynostosis deserve great interest. The fronto-orbital advancement technique and the free bone flap

permitted good functional and esthetical results in majority of cases.

Central sleep apnoea and associated Chiari malformation in children with syndromic craniosynostosis: treatment and outcome

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Purpose: The association of Chiari malformation (CM) in patients with syndromic craniosynostosis (SC) is well established. However, the incidence and management of central sleep apnoea in these patients remains poorly defined. We aimed to assess the efficacy of foramen magnum decompression in resolving central sleep apnoea in patients with CM and SC.

Method: The clinical data of four patients with CM and SC, who underwent foramen magnum decompression for central sleep apnoea at Alder Hey's Children Hospital between January 2008 and December 2009, were retrospectively analyzed. Outcomes were evaluated with respect to foramen magnum decompressions through pre- and post-surgical sleep studies. Of the four patients, two had a diagnosis of Crouzon's and two had Pfeiffer's syndrome.

Results: Patient age at time of surgery ranged between 1.1 and 12.6 years (median 2.6 years). Median postoperative follow-up was 2.9 years. Post-operative sleep studies revealed that one child experienced <80% reduction of central sleep apnoeas at 21 months after decompression. The remaining three children experienced <60% reduction in central sleep apnoeas when re-evaluated between 2 and 10 months later. The associated apnoea–hypopnoea index improved for all patients.

Conclusion: Our findings suggest that foramen magnum decompression is an effective treatment modality for central sleep apnoea due to CM, in syndromic craniofacial patients. The use of multimodality polysomnography technology may improve the evaluation and management of these patients.

Subtotal Calvarial vault remodelling in the treatment of scaphocephaly: aesthetic and MRI cerebral perfusion outcomes

Paul Sillifant, Ajay Sinha, Sasha Burn, Christian Duncan (Liverpool Super regional Craniofacial Unit, Liverpool, UK)

Aim: This study aims to present Liverpool Technique for the correction of scaphocephaly in children older than 6 months. **Background:** Scaphocephaly due to sagittal craniosynostosis presents a number of features which result in cosmetic

deformity. These include a long thin head, varying degrees of frontal bossing, occipital bullet, anteriorly positioned vertex, and bitemporal pinching.

Numerous surgical therapies have been utilized to manage these problems from strip craniectomy and micro barrel staving to spring assisted cranioplasty, subtotal vault remodelling, and total calvarial vault remodelling.

We will present our technique of subtotal calvarial remodelling and the results and complications in the first 45 patients and recommendations for timing and indications for the technique. We have also collected data on intracranial pressure generated during the PI squeeze procedure and the effect surgery has on MRI perfusion evaluated intraoperatively.

Results:

- Mean age=17 (7–75) months
- Mode age=10 months
- Mean Cephalic index (preop) 66 (58–76)
- Mean Cephalic index (postop) 76 (68–88), $P<0.05$
- Mean FU (months) 13 (1–72)
- Only six patients had a relapse in the CI by >3 points in follow-up period.

Conclusion: We believe that our results show that subtotal calvarial remodelling addresses most of the features of scaphocephaly namely normalising cephalic index, correcting bitemporal pinching, and frontal bossing correcting the location of the vertex and some improvement in the occipital bullet without the risk associated with total vault remodelling to the posterior venous sinuses.

How to evaluate the improvement of development after surgery for mild form of craniosynostosis

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Introduction: We have reported the surgical results in patients with mild form of craniosynostosis and clinical symptoms, previously. We mentioned the difficulties of statistical analysis of the improvement in development of those patients. In this report, the detailed analysis was performed in selected cases to assess the future direction of this clinical research.

Patients and methods: The indication of surgery is described previously. We measured intracranial pressure of those patients epidurally for few days. If the intracranial pressure is high, the cranial expansion was performed with gradual distraction method. We retrospectively analyzed 43 cases we treated in the last 10 years. In this paper, we analyzed more details of the relationship between the age of treatment and outcome. We also describe how we should evaluate the outcome.

Results: All patients showed improvement in some degree. If the patients were younger than 3, the improvement of speech was noticed more than 2/3 of the cases. If the patients were older, the improvement of behavior was recognized. On the other hand, the improvement in development was not shown numerically with well-known methods to evaluate the intelligence quotient (IQ) and/or developmental quotient (DQ).

Conclusion: Even though the family member of the patients with mild form of craniosynostosis and/or physician recognized the improvement in development after surgical intervention, the well-established IQ/DQ score did not show the improvement. We need to establish new system to evaluate the improvement of speech development and/or behavior in those patients.

Wednesday October 19, 2011

Session 9. Dysraphism

Pre- and post-untethering courses of syringomyelia which is associated with tethered spinal cord

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Background: There has been controversy regarding management of syringohydromyelia associated with tethered spinal cord. Patients of syringomyelia associated with closed spinal dysraphism, which was managed by untethering alone, were reviewed.

Method: Of 138 patients who were operated on from 2003 to 2008, 33 showed preoperative syringomyelia. The pre and post-untethering clinical data and syrinx index (ratio of the syrinx diameter and the cord diameter) were retrospectively reviewed.

Results: Five patients showed symptom progression during preoperative period and four of them had an additional magnetic resonance imaging (MRI) before the operation, in which all four of them revealed progression of the syringomyelia. Also, all of the seven patients with more than one preoperative MRIs with no progression of syrinx showed stability of symptoms. Postoperatively, 31 patients out of 32 with postoperative imaging available, showed long-term stability or decrease in syrinx index. In one patient with retethering, syrinx index increased, 4.5 years after untethering. All of the patients who were stable symptomatically did not show aggravation of the syrinx postoperatively. Also, four patients showed transient increase in the syrinx index during initial postoperative period which decreased spontaneously later with improved or stable symptoms.

Conclusion: Preoperative neurologic deterioration may be correlated with progression in syringomyelia. Untethering alone may be sufficient in management of syringomyelia associated with cord tethering. Transient increase in syrinx index during the initial postoperative period may be observed without additional surgery, if symptomatically stable. Also, retethering should be suspected in patients that show increase in syrinx size during follow-up.

Nonterminal myelocystoceles: an underdiagnosed entity?
Natarajan Muthukumar (Madurai Medical College, Madurai, India)

Aim: The aim of this study is to analyze the clinical presentation, radiological findings, surgical treatment and outcome of nonterminal myelocystoceles (NTM) treated by this author.

Methods: Between 1998 and 2010, 20 patients with NTMs were treated by this author. All children underwent neurological evaluation and magnetic resonance imaging evaluation. Surgical findings were recorded. The aim of surgery was to detether the cord and achieve watertight dural closure after sectioning the fibrovascular stalk in type I lesions; detethering, syrinx drainage and watertight dural closure in type II lesions. Follow up ranged from 3 months to 2 years.

Results: Age, newborn to 3 years. Female/male ratio was 9:1. Lesion locations were as follows: cervical, 6; thoracic, 6; and lumbar, 8. All children except one were normal neurologically. Radiologically, two children had Rossi type II NTMs and the remaining 18 had Rossi type I NTMs. No patient with lumbar NTM had type II lesion. All children with lumbar NTMs had low-lying cords. Radiologically, type I lesions were characterized by posterior tenting of the cord at the site of the lesion. None of the patients had associated hydrocephalus. Six of the eight lumbar NTMs had radiological evidence of tonsillar herniation. No patient had postoperative CSF leak and there was no retethering during an average follow-up of 9 months.

Conclusions: NTMs are not rare lesions. They are often misdiagnosed as meningoceles. Improper diagnosis may lead to suboptimal treatment without adequately untethering the cord. Outcome of children with NTMs is good.

The genomic analysis in human myelomenigocele
Akihide Kondo, Kazuaki Shimoji, Masakazu Miyajima, Hajime Arai (Juntendo University, Tokyo, Japan)

Introduction: Myelomenigocele is supposed to be caused by complex genetic and environmental factors. Some studies have already implicated several chromosomal regions in myelomenigocele. However, there is no

consistent picture of susceptibility loci in myelomeningocele at this point.

Recently, the contribution of chromosomal structural variation to differences in genetic makeup within the human population has been highlighted. Advances in technology have enabled the screening of large cohorts of individuals for copy number variants (CNVs) genome wide. Here, we have performed this study to find the candidate genes or genomic regions for human myelomeningocele using this technology.

Methods: This study included 40 patients. All the patients had myelomeningocele treated at our institution between from 2006 and 2009. A blood sample was taken from all affected individuals. DNA was extracted from blood by means of a DNA purification kit. CNV analysis was carried out using Affymetrix® single nucleotide polymorphism arrays. To identify CNVs that are associated with myelomeningocele, we compared this data from control samples they are supposed to be disease free from a database publically available.

Results: We have detected several CNVs that are supposed to be associated with an increased the risk of myelomeningocele.

Conclusion: Further study is needed to identify the role of these candidate genes and genomic regions in the biomechanical pathway of the occurrence of myelomeningocele.

De novo in utero surgery for myelomeningocelis

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After Management of Myelomeningocele Study's (MOMS) results, the Fetal Department at the Federal University of Sao Paulo, Brazil, has resumed the program for closure of myelomeningocelis in utero. Five years ago, before MOMS started studies, six cases of myelomeningocele had been successfully operated in utero, all of them before 26 weeks of pregnancy. The patients were four female and two male. The Chiari II malformation disappeared in four cases. In two cases, it was necessary to place a shunt, which presented with a huge hydrocephalus, and should not have been operated in utero. Four of those six babies are now walking normally without a shunt. In two cases, some kind of device has to be used in order to help deambulation. Nowadays, 15 days after MOMS's publications another two cases have been operated without developing hydrocephalus. In conclusion, as shown by MOMS's results, the closure of myelomeningocelis in selected cases prevents hydrocephalus and perhaps improves movements in the lower limbs, besides being a safe procedure for both mother and baby. It would be

desirable that every country could develop fetal programs to treat this serious illness

Folic acid fortification in Costa Rica: impact on severity and incidence of neural tube defects

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The impact of folic acid fortification has been proved to dramatically reduce the incidence of neural tube defects (NTD). In 1997, a new health policy was passed as a law by the Costa Rica Congress and this enabled wheat flour, followed by corn flour fortification. This produced and average intake of folate of 124 µg/day. Within 3 years, there was a reduction of over 50% in the incidence of myelomeningocele which has remained constant ever since. On the other hand, the impact on the severity of these defects was also evident with a caudalization of the levels affected (27% vs. 14% thoracic lesion pre- and posttreatment) but also the diameter was greatly reduced and kyphotic defects disappeared from our series. About 35% of the cases after fortification have been able to become shunt independent within the first 5 years of age. Symptomatic Chiari decreased from 9% to 4% of the cases per year. Given these significant achievements, we discuss the importance of folic acid fortification health policies and their implementation in developing world countries.

Combined myelomeningocele closure and ventriculoperitoneal shunting is not associated with an increased risk of shunt infection or malfunction

Humphrey Okechi, Albright Leland, Susan Ferson (AIC Kijabe Mission Hospital, Nairobi, Kenya)

Introduction: Myelomeningocele (MMC) and hydrocephalus account for the majority of pediatric neurosurgical conditions in East Africa. Coexisting MMC and congenital hydrocephalus can be treated concurrently within the first day of life without any increase in shunt infection rate.

This study investigated the incidence of shunt infection and malfunction among patients undergoing combined MMC closure and ventriculoperitoneal shunting (VPS; CP) versus those undergoing staged procedures (SP).

Methods: Data were prospectively collected between October 2010 and March 2011 for infants (mean age, 15.38 days; range, 1 day to 3 months) undergoing MMC closure and VPS in a combined or staged fashion with a 30-day postoperative follow-up. All patients in the CP group preoperatively were afebrile, without leukocytosis and had

sterile ventricular cerebrospinal fluid (<5 white blood cells/mm³). Study endpoint was incidence of shunt infection and malfunction.

Results: Thirty-seven patients, 17 CP and 20 SP, underwent MMC closure and VPS in a combined and staged procedure, respectively. There was no significant intergroup difference in preoperative head circumference or ventriculomegaly. Also, no difference in the incidence of shunts infection CP ($n=1$) and SP ($n=2$) or shunt malfunction CP ($n=1$) and SP ($n=4$); $p=0.562$ and $p=0.225$ was observed. The SP group had a significantly longer duration of hospitalization mean, 14.10 day (CP, 7.31 days), $p<0.01$ with a mean staging interval of 6.75 days.

Conclusion: Combined myelomeningocele closure and VPS with preoperative sterile cerebrospinal fluid is not associated with an increased risk of shunt infection or malfunction in the immediate postoperative period.

Primary tethered cord syndrome—manifestations diagnosis and management: a prospective study

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Objectives: This paper aims to study the presenting clinical features of patients with primary tethered cord syndrome and to evaluate the indications of surgery in such patients and its outcome.

Methods: This is a prospective study carried out at All India Institute of Medical Sciences, New Delhi between July 2009 and December 2010. Study included patients admitted with clinical and radiological diagnosis of primary tethered spinal cord, for detethering. The indication for surgery in all cases was the presence of a tethered cord on magnetic resonance imaging, the criteria for tethering being low-lying conus (below L1–L2) and a thickened filum (>2 mm). Urodynamic studies were performed before detethering.

Results: Thirty-four patients were included in study. Female to male ratio was 1:8. Abnormality of curvature and gait were the most common complaints. Urological complaints were seen in 11 (32.3%) patients. On magnetic resonance imaging, L3 was commonest level of the low lying conus, seen in 58.85% of the total patients. Around 66.6% of the patients showed improvement post surgery at 6 months follow up. On follow up urodynamic study, bladder capacity improved by a mean of 18.8 ml. Of the patients, 37.5% showed improvement in bladder compliance, 35.7% showed improvement in involuntary bladder contractions whereas 50% patients showed improvement in dyssynergia of sphincter. On post operative urodynamic study, five patients showed involuntary bladder contractions and one showed dyssynergia of sphincter.

Effect of operation time on mortality and morbidity in neonatal myelomeningocele

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Objective: We aimed to evaluate the clinical features, short-term prognosis, and effect of operation time on mortality and morbidity in neonatal myelomeningocele (MMC).

Methods: This study was conducted between 1990 and 2010. Clinical features, defective area, existence of additional anomalies, and operation time on mortality and morbidity data were recorded. The effect of operation time on mortality and morbidity was evaluated.

Results: We analyzed data from the medical files of 19 patients with MMC that underwent surgery within 3 days after birth (group 1, $n=6$) and over 3 days after birth (group 2, $n=13$). Two groups were compared with clinical features, morbidity and mortality prospectively and existence of additional hydrocephalus. The preoperative rupture of the MMC occurred more frequently in group 1. The need for ventriculoperitoneal shunt was three in group 1 and four in group 2, and three of them were performed after the immediate MMC repair. After 1 year of follow-up, two of group 1 ($n=6$), showed neurodevelopmental delay and three of group 2 ($n=13$) showed also delay. Overall outcome of these neonatal MMC repair were improved in nine, stable in two, worse in none, and death one (mortality rate, 5.3%)

Conclusions: The surgical intervention performed in within 3 days after birth showed benefits regarding a lower incidence of preoperative rupture of the MMC, postoperative dehiscence (we simultaneously underwent repair of the MMC and ventriculoperitoneal shunt). And there were no differences of neurodevelopmental delay in two groups. During their follow-up period, 10 patients were has normal growth. Eight patients remained with low intelligence.

Tethered cord syndrome associated scoliosis benefit from sectioning of filum terminale

Mehmet Selcuki, Ahmet Sukru Umur, Deniz Selcuki, Mustafa Barutcuoglu, Yusuf Kurtulus Duransoy (Celal Bayar University, Manisa, Turkey)

Scoliosis may be the first sign of tethered cord syndrome (TCS). TCS has classically been defined as traction on the lower portion of the spinal cord and abnormally low position of the conus medullaris, which cause progressive neuromuscular and urological symptoms. It is no difficult to diagnose if classical signs are present or conus is lower than lumbar 1–2 disc space in MRI investigation.

In our 16 cases, although they had no previous signs of scoliosis, they developed scoliosis at a certain time of their life and progressive nonradicular low back pain. We perform lumbosacral MRI all patients for diagnose TCS and probable additional spinal anomalies. In six cases, magnetic resonance imaging (MRI) investigations were normal. In electrophysiological examinations, 13 of all 16 cases had no urodynamic abnormality. Somatosensorial evoked potentials (SSEP) could be performed in 14 cases. All SSEP investigations were pathological. The mean follow-up period is 5 years and there was neither progression of scoliosis nor deterioration of clinical outcome.

It seems SSEP is important tool for the diagnosis of tethered cord patients in the cases with normal MRI and normal urodynamic investigations. Sectioning of filum terminale is simple and safety operation for the treatment in TCS-associated scoliosis.

Intraoperative monitoring of motor-evoked potentials in children with spinal pathologies

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Introduction: Intraoperative monitoring during spinal operations is increasingly employed in children to minimize neurological complications of surgery. The efficacy of motor-evoked potentials is not fully ascertained in the pediatric population.

Material and methods: Intraoperative monitoring of motor-evoked potentials using transcranial (cork screw) stimulation was employed during spinal operations in 18 children (seven boys) in the period 2008–2011. Mean age at operation was 79 months (range, 6–206 m). Pathologies included spinal lipoma ($n=6$), thickened filum terminale ($n=5$), diastematomyelia ($n=3$), dermoid cyst ($n=1$), spinal arachnoid cyst ($n=1$), ependymoma of the cauda equina ($n=1$), and spinal neuroblastoma ($n=1$).

Results: In all patients, satisfactory recordings were obtained, regardless of patient age, including five patients with pre-existing weakness in the lower limbs. In three patients, the amplitude of the recordings was transiently reduced and complete recovery was observed after a period of cessation of dissection. In one patient with complex dysraphic state (myelomeningocele closed at birth, hydrocephalus with shunt, Chiari II, holocord syringomyelia and diastematomyelia at L1, which was being operated on), who was ambulant but with severe weakness of the legs, the recordings were preserved throughout the procedure but the patient woke up with neurologic deterioration of the legs (paraplegia) which never recovered 2 years from the operation.

Discussion: Motor-evoked potential monitoring is useful during spinal neurosurgical operations in children. Its main

benefit is in excision of intramedullary spinal lesions where the spino-thalamic tracts are directly threatened. In operations involving spinal roots and nerves probably its sensitivity is reduced.

The indication of endoscopic third ventriculostomy for the patients suffering from hydrocephalus due to myelomeningocele as a replacement of ventriculo peritoneal shunt

Takaoki Kimura, Kazuaki Shimoji, Akihide Kondo, Masakazu Miyajima, Hajime Arai (Juntendo University, Tokyo, Japan)

Objective: The efficacy of endoscopic third ventriculostomy (ETV) as a primary treatment for children having hydrocephalus due to myelomeningocele is controversial. However, some studies reported that ETV is well working for 50–80% of patients having shunt tube trouble by means of the replacement of ventriculoperitoneal (VP) shunt system in elder children.

Then, we have conducted a retrospective study to clear the surgical indication of ETV for patients suffering from myelomeningocele with hydrocephalus.

Materials and methods: This study is including 11 patients. All of them were performed ETV in our institution when they showed symptomatic hydrocephalus because of the trouble of VP shunt.

Results: Four patients needed shunt revision. Seven patients remained shunt free without any intervention for their hydrocephalus. Our MR imaging study revealed the fact that tight posterior fossa can be a restricting factor for the success of ETVs.

Conclusion: Successful ETV can be conducted in cases having aqueductal stenosis and mild Chiari malformation. Sufficient consideration of MR imaging is one of the key factor to determine the indication of ETV for patients suffering from myelomeningocele.

The surgical treatment of Chiari malformation type I

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The most appropriate surgical treatment for Chiari malformation type I (CM-I) remains controversial, especially in children.

Between 1989 and 2009, we operated on 46 children with CM-I. Pre-operatively, all patients were studied with magnetic resonance imaging (MRI) of the brain/cranio-cervical junction, 24 obtained a cine-MRI study, and 33 a spine MRI.

Surgical treatment included either bone decompression and dural stripping (group BD; $n=11$), duroplasty (group

DP; $n=13$) or duroplasty with tonsillar shrinking (group DP + TS; $n=22$).

Statistics included ANOVA and chi-square analysis to assess correlations between surgical treatment, age, hospital stay, surgical complications, and outcome.

Cine-MRI was abnormal in 19/24 children (79%). Syringomyelia was present in 26/33 children (79%). Mean age of group BD was significantly lower (7.1 year) as compared to group DP (10.2 years) or DP + TS (11.8 years; p value=0.001). Similarly, hospital stay was remarkably shorter (5.2 days) for group BD as compared to group DP and DP + TS (10 and 11 days; p value=0.019 and 0.001, respectively). There were no complications in group BD; one pseudomeningocele in group DP + TS; two pseudomeningoceles, one dural fistula, and one aseptic meningitis in group DP.

Mean follow-up was 8.7 years. All 20 children without syringomyelia improved after surgery, 90% becoming asymptomatic. Vice versa, only 52% of children with syringomyelia became asymptomatic, 40% improved, and 8% had no benefit from surgery.

We favour a more conservative surgery in younger children without syringomyelia, as this lowers both complications and hospital stay. This study, however, is not conclusive with regards to the value of a conservative approach in children with syringomyelia due to the small number of patients in this group.

Estimated cumulative effective dose from radiologic procedures in spina bifida, a retrospective cohort study of 135 children in the Netherlands

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Objective: Pediatric spina bifida patients undergo, as a result of multiple comorbidities, a large amount of radiologic examinations during childhood. Ionizing radiation is frequently used in these examinations, expressed in effective dose. A cumulative effective dose (CED) is used to predict the risk of cancer. Our aim was to estimate the CED from radiologic procedures in a cohort of pediatric spina bifida patients.

Method: A retrospective cohort study of 135 spina bifida patients. All procedures, in which ionizing radiation was involved, were recorded. Examination-specific effective doses were calculated using age-specific dosimetry data, derived from institutional research and published dosimetry literature. **Results:** Three patients were not exposed to ionizing radiation examinations. Individual CED estimates ranged from 0 to 103 mSv, the mean CED was 10.03 mSv. The mean CED was lowest in the meningocele subgroup (5.4 mSv) and highest in the myelomeningocele subgroup (14.97 mSv). The highest individual estimated CED (103.0 mSv) occurred in the SB occulta subgroup.

Discussion: Although this study gives a detailed insight in the characteristics of a spina bifida population, exposition to ionizing radiation was lower than expected. Nevertheless, radiologic examinations without ionizing radiation, like MRI and ultrasonography, should always be considered whenever possible to reduce the amount of ionizing radiation.

Keywords: spina bifida, neural tube defects, ionizing radiation, radiography, computed tomography, nuclear medicine, MRI, ultrasonography, effective dose, dosimetry

Stridor presenting at birth in neonates with myelomeningocele: preliminary results of an international survey

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Introduction: Stridor, associated with vocal cord paralysis, in neonates with myelomeningocele (MMC) usually appears after birth. Control of hydrocephalus and decompression of Chiari malformation (CM) usually improves symptoms. Occasionally, stridor presents at birth. There are five patients with stridor at birth at our institution and three reported in the literature died despite treatment. The purpose of this study was to determine the incidence and prognosis of stridor presenting at birth in MMC patients.

Methods: We conducted an international survey under the auspices of the Education Committee of the International Society for Pediatric Neurosurgery (ISPN). Survey was designed in two stages. A preliminary questionnaire, designed to obtain data on the incidence of stridor at birth in MMC patients were sent to ISPN members via e-mail. The members were also asked if they would participate in a multicenter study by providing more detailed information about these patients at a later time.

Results: Thirty-three centers answered our survey. Twenty-four centers agreed to participate in the detailed study. Sixteen centers had patients who presented with stridor at birth. Of 1,055 patients with MMC, 52 (5%) presented with stridor at birth. Most centers anecdotally reported poor outcomes for these patients.

Conclusion: Our initial survey reveals that stridor at birth in neonates with MMC is rare and probably has a dismal prognosis. A planned detailed multicenter study would provide a stronger scientific basis for making management decisions.

Session 10. Spine

Management of complex non-traumatic compressive spinal deformity in pediatric patients

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Introduction: Complex non-traumatic compressive spinal deformity in children is characterized by spinal deformity or instability co-existent with spinal cord or nerve root compression. The condition poses significant challenges in terms of preservation of neurological function and maintenance of stability in a growing spine.

Methods: We retrospectively retrieved from a prospective operative database all patients meeting these criteria from 2000 to 2010 treated by the primary author. Patient demographics, clinical presentation, operative procedures, outcomes, and complications were extracted from the hospital records.

Results: Of 38 patients, 20 boys (52.6%), age 1.4–16.7 years (mean 10.3 years), 55.3% presented with a neurological deficit ranging from mild spasticity to ventilator dependent quadriplegia. The most common etiologies were neoplasms (23.7%), trisomy 21 (18.4%) and Klippel–Feil syndrome (15.8%). The most common locations were C1–C2 (39.5%), cervico-thoracic (28.9%), and thoracic (15.8%). Pre-operative traction was used in 36.8%. All patients were operated by a combined neurosurgery and orthopedic team with intraoperative neurophysiological monitoring. Virtually all received spinal instrumentation and autologous bone grafts, 73.7% received halo immobilization, 13.2% required an unplanned second surgery, and 18.4% experienced a complication usually transient and/or minor.

Conclusions: Complex non-traumatic compressive spinal deformities represent a varied and generally high-risk group best managed in multidisciplinary teams. Pre-operative traction, selective halo immobilization, universal intraoperative neurophysiological monitoring may reduce perioperative complications and are generally well tolerated. Functionally important loss of spine growth and delayed progressive deformity has not been a significant problem to date.

Surgical outcome in paediatric atlanto axial dislocations: an institutional experience

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Atlanto axial dislocations (AAD) is challenging in pediatric age group and yet it is not very uncommon. This study was conducted to evaluate the surgical outcome in various operative techniques of paediatric AAD patients. Thirty patients diagnosed as AAD who underwent surgery at Nizams Institute of Medical Sciences, Hyderabad were analyzed retrospectively between June 2004 and April 2010. Details were taken from neurosurgical discharge

summary database and operation theatre records. There were 21 males and nine females. Age range was from 6 to 18 years. Etiologically, the atlantoaxial dislocation were divided as 19 congenital, 8 traumatic, 2 infective, and a single case of rheumatoid arthritis. Fourteen were reducible, 16 were irreducible. In this group, five patients were managed conservatively and 25 underwent surgery. Eight patients underwent combined trans oral and posterior fixation, 15 patients underwent only posterior fixation, and one patient only anterior approach. There were two cases of mortality in our series of 30 patients. Implant failure was encountered in three cases and a transient wound infection in one patient. Follow-up ranged from 3 months to 5 years and is available in 80% of patients all of whom improved significantly. In conclusion, AAD in paediatric age group is curable and has good prognosis. When properly judged and correctly fused, it gives good surgical outcome.

Craniovertebral junction stabilisation in children less than 5 years of age—limitations, options and outcomes Tony Elias, Nick Haliasos, Dominic Thompson (Great Ormond Street Hospital for Children, London, UK)

In young children with craniovertebral instability, deformity, or neuraxial compression, the treatment options are limited due to the combination of skeletal immaturity and small size particularly in the context of bone dysplasias.

This study examines the clinical effectiveness of craniovertebral stabilisation procedures in children less than 5 years of age. Fifteen children underwent posterior cervical fixation between 1997 and 2011. The median age at surgery was 27 months (range, 9–45 months), with mean follow-up of 3.5 years. The underlying diagnosis was mucopolysaccharidosis ($n=4$), spondyloepiphyseal dysplasia ($n=3$), congenital AAD ($n=2$), Down's syndrome ($n=1$), Marfan's syndrome predisposing to AARF ($n=1$), and AARF with os odontoideum ($n=1$). Three children had acquired lesions: one each for trauma, neoplasm and infection. Operations included 12 occipitocervical and three atlantoaxial fixations. All comprised a combination of sublaminar wires and autologous bone graft, except one which had rigid occiput to lateral mass fixation; and all patients were immobilised postoperatively in a halo orthoses for 3 months. Twelve children demonstrated radiological and clinical evidence of fusion. Two patients needed revision operations for initial nonfusion. One child died at 1 month due to systemic sepsis unrelated to surgery. One child developed an ischaemic injury to the spinal cord 24 h post-operative resulting in paraplegia. All the other children were mobile either independently ($n=12$) or with support ($n=1$).

Posterior cervical fixation can be performed effectively in very young children, and morbidity is largely related to neurological disability at presentation. An algorithm of management is presented for this group of children.

In over half of the children, gait abnormality remained after surgery. Eighty-eight percent of children with normal gait did no deteriorate after surgery. These findings need to be validated in a larger series.

Management of anteriorly placed intramedullary spinal cord tumours

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Spinal cord tumours need specific strategies for successful management. Anteriorly placed intramedullary tumours are especially challenging.

We describe our strategy for surgical management of these lesions in seven cases. Dorsal root entry zone approach was used in five patients and an anterior approach was used in two patients. Four lesions were in the cervical and three in the dorsal region. The extent of the lesion ranged from three to ten segments. One hemorrhagic lesion turned out to be a malignant mesenchymal tumour in a 6-year old who did not improve from his paraplegia. Three others were pilocytic astrocytomas. Two were completely excised and one subtotal excision with significant improvement. All three ependymomas were near totally excised with good neurological recovery. Follow-up ranged from 1 to 14 years.

The Vertical expandable prosthetic titanium rib (VEPTR) in the treatment of spinal deformity in growing children

Yusuf Ersahin, Tuncer Turhan (Division of Pediatric Neurosurgery, Ege University Faculty of Medicine, Izmir, Turkey)

The vertical expandable prosthetic titanium rib (VEPTR) has been developed to support multiple expansions of the thorax of children with chest wall and spine deformities, to prevent worsening scoliosis during body growth, and improve lung growth and function. We aimed to share our experience with the VEPTR in the treatment of spinal deformity in growing children.

The patients with spinal deformity who had undergone VEPTR implantation were retrospectively reviewed. All patients had MRI scan of the spine, plain X-rays of the spine. Those with spinal dysraphism had spinal cord release before VEPTR surgery. Scoliotic and kyphotic Cobb angles were measured before and after surgery. The expansion was done in every 6 months.

VEPTR was implanted in nine patients, age ranging from 4 to 12 years (mean 7.3 years). There were five boys and four girls. Seven patients had spinal dysraphism. Rib to iliac VEPTR was implanted bilateral and unilateral in five and one patient, respectively. Rib to lamina construct was used in the remaining patients. The constructs were removed in one of the patients because of skin breakdown.

The goals of fusionless methods to manage severe scoliosis in the growing child include curve maintenance and allowance for chest wall growth. Goals of treatment include stabilizing curve progression while allowing for normal spine, chest, and lung growth. Spinal fusion at an early age results in a short trunk and impaired lung development; in addition, posterior-only surgeries may lead to the crankshaft phenomenon. It seems that VEPTR can achieve these goals.

Intramedullary spinal cord tumors: analyses of 24 cases. A novel technique of "Micro" myelotomies is also highlighted

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Introduction: Intramedullary spinal tumors account for 2–4% of central nervous system (CNS) tumors. This study reevaluates aspects of the disease that contribute to their outcome. A novel surgical technique using multiple "micro" myelotomies is also highlighted

Methods: Twenty-four patients with intramedullary spinal cord tumors were treated in our institute from January 1997 till date. The patients were operated by the technique of multiple "micro myelotomies", when applicable, with piecemeal excision of tumor without using Cavitron Ultrasonic Surgical Aspirator or Somatosensory or Motor Evoked Potential. Two to 4 mm myelotomies are carried out in the posterior median sulcus at sites which are devoid of pial and dorsal spinal vessels. The patients were analyzed according to their clinical features, extent of resection, post-operative functional outcome, histological diagnosis, and adjuvant therapies.

Results: Total excision was performed in four cases, near total in eight cases, subtotal in six, and biopsy in six cases. Two cases were operated in a staged manner. Astrocytoma (six cases) ependymoma (four cases) were the most common histologies others included cavernomas, tuberculoma, schwannoma, and hemangioblastoma as well as 15 cases showed clinical improvement (64%). Six cases remained the same and three cases worsened. There was no mortality. Two patients recurred and two died in long-term follow up.

Conclusions: Technique of multiple myelotomy is a feasible option for radical excision. The tumor consistency and plane are most important parameters for total excision. Pre-operative neurological status and histology are the two important factors for favorable surgical outcomes.

Session 11. Hydrocephalus 2

Endoscopic aqueductoplasty in children with isolated fourth ventricle: approach, technique and endoscope selection

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Objective: An isolated fourth ventricle is rare but still a treatment challenge. Neuroendoscopic options encompass aqueductoplasty and aqueductal stenting. In this study, the authors present their experience with the use of rigid endoscopes in different surgical approaches in 11 neuroendoscopic procedures.

Methods: A series of seven patients (22–109 months of age) with an isolated fourth ventricle is reported. The patients underwent 12 endoscopic procedures at our department between 2005 and 2010. Rigid rod-lens endoscopes were used in all patients for aqueductoplasty and stent implantation.

Results: All patients underwent endoscopic aqueductoplasty. A supratentorial frontal approach was chosen in seven surgeries, a suboccipital infratentorial approach in five. No complications occurred. Successful opening of the aqueduct could be confirmed by transaqueductal inspection with a small rigid endoscope in all cases. Four patients initially underwent aqueductoplasty only without stenting (one of them because of cerebrospinal fluid (CSF) infection), three children had initially an aqueductoplasty with stenting. All four children with aqueductoplasty alone eventually underwent aqueductal stent implantation; three because of reclosure of the aqueduct, and one after recovery from CSF infection. One of the three children with initial stent implantation required revision for stent dislocation. The procedures were eventually successful by clinical and MR imaging criteria in all patients.

Conclusion: Endoscopic aqueductoplasty is an adequate treatment for isolated fourth ventricle. Transaqueductal inspection is possible even with a rigid endoscope without complications. After perforation of the obstructing membrane, primary stent implantation should be performed to prevent reclosure of the aqueduct.

Familial aggregation of congenital hydrocephalus in a nationwide cohort

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Objective: This study aims to investigate familial aggregation of idiopathic congenital hydrocephalus (CHC) in an unselected, nationwide population.

Methods: Based on the Danish Central Person Registry, we identified all children in Denmark born 1977–2009 and their family members (up to third-degree relatives). Information on a diagnosis of CHC was obtained from the National Patient Discharge Registry. Using binomial log-linear regression analysis the recurrence risk ratios (RRR) of congenital hydrocephalus was calculated for same-sex twins and first-, second-, and third-degree relatives of a CHC case.

Results: Of the 1,928,683 live born children, 2,302 had a diagnosis fulfilling the criteria for idiopathic congenital hydrocephalus (1.2%). Of these, 117 (5.1%) had at least one other family member with a diagnosis of idiopathic hydrocephalus. Significantly increased RRR of CHC were found for same-sex twins and first-, second-, and third-degree relatives: 40.2 (95% CI: 19.7–82.2), 7.6 (95% CI: 5.5–10.6), 2.3 (95% CI: 1.7–3.2), and 2.1 (95% CI: 1.3–3.5), respectively. The inheritance appeared to be transmitted equally in the maternal and paternal line of families.

Conclusion: This is the first population-based cohort study investigating familial aggregation of CHC. The results suggest a strong genetic influence on the occurrence of idiopathic CHC.

Phase contrast MRI of intracranial shunt tube: a valuable adjunct in the diagnosis of ventriculo-peritoneal shunt malfunction

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Introduction: Diagnosis of shunt malfunction is still a challenge. Erroneous shunt revisions lead to unwanted stress and economic burden to patient.

Aims and objectives: This study aims to assess the role of phase contrast magnetic resonance imaging (MRI) in children with clinically suspected shunt malfunction.

Material and methods: In this prospective study over a 1-year period, all children (<18 years of age) with clinically suspected shunt malfunction who were planned for shunt revision underwent phase contrast MRI of the brain preoperatively. All MRIs were done by a single radiologist using standard parameters and shunt tube was traced in its intraparenchymal course and a region of interest (ROI) defined.

Cerebrospinal fluid (CSF) flow velocities were measured across this ROI. A control arm of eight asymptomatic patients with documented well-functioning shunts (on serial computed tomographies of the head) were selected who also underwent phase contrast MRI under similar conditions

Observations: Seventeen patients with mean age of 5.2 ± 4.1 years (range, 0.25–15 years) were studied. Nine patients were in the shunt malfunction (test) group and eight in the control group. The mean area across which velocities were measured 1.65 ± 0.22 mm² approximately corresponded to internal cross-section area of shunt tube. The peak velocities (1.62 ± 1.41 cm/s) and mean velocities (0.78 ± 0.66 cm/s) in test group differ significantly from the peak velocities (0.38 ± 0.98 cm/s) and mean velocities (0.24 ± 0.66 cm/s) in the control group ($p=0.01$, $p=0.002$, respectively). Intraoperatively shunt malfunction was confirmed in all except one patient.

Conclusion: CSF velocities calculated across shunt tubing by phase contrast MRI reliably reflect the functional status of ventriculoperitoneal shunt.

The role of ETV in the management of myelomeningocele associated hydrocephalus

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The management of hydrocephalus is one of the most trying problems in children with myelomeningocele. The value of endoscopic third ventriculostomy (ETV) is frequently questioned in “myelomeningocele-associated-hydrocephalus” (MAH) even though this condition has mainly an obstructive basis. In this study, we compared the affectivity of ETV as the first choice of treatment with its use as an option for the management of shunt dysfunction in MAH.

The data of total 127 MAH cases belonging to two centers which have been treated with ETV were retrospectively analyzed. The patients who had been followed up for at least 1 year postoperatively were included in the study. The age of the patients ranged from 1 day to 16 years. Of those cases, 85 were treated with ETV as “the primary choice of management” (group A). Of the group A cases, 64 were “under 2 years of age” (group Au) and 21 were “over 2 years” (group Ao). ETV was used as an option for management of shunt dysfunction in 42 cases (group B). In group B, 12 cases were “under 2 years of age” (group Bu) and 30 were “over 2 years” (group Bo). The overall success rate of ETV was calculated as 48.2% for group A (39% for group Au and 76.2% for group Ao). The overall success rate for group B was calculated as 69% (41.6% for group Bu and 80% for group Bo).

Our results seem to suggest that the success of ETV is relatively higher when used as a treatment option in

shunt dysfunction compared to its use as “the-primary-choice-of-treatment” in MAH. In both conditions, it seems that the age of the patient “being less than 2 years” significantly lowers the success rate of the procedure.

Revisiting the endoscopic ventricular shunt catheter insertion and revision: risks factors for failure

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Introduction: Results of the endoscopic ventriculoperitoneal shunt insertion trial (2003) showed no reduction in the incidence of shunt failure. We analyze risk factors for failure in 31 endoscopically guided shunt insertions and revisions at our institution.

Methods: Retrospective review of charts and radiographic images for all endoscopic shunt placements and revisions performed by a single surgeon at Arkansas Children's Hospital from December 2008 to September 2010. Minimal follow-up was 6 months. Endpoint was failure of shunt requiring a subsequent revision. Risk factor analysis was performed with Cox proportional hazards regression and Kaplan–Meier survival curves.

Results: Thirty-one patients were included, two patients excluded due to unrelated death. Ten were initial insertions and 19 were proximal revisions. Median age was 5.65 years. Mean follow-up was 411 days. Overall, 27.6% of shunts failed (2/10 initial placements and 6/19 revisions). Considering endoscopic shunts with at least 1 year follow-up, the failure rate was 30.7%. No shunt infections were reported postoperatively. Analyzed factors included age, etiology of hydrocephalus, frontal vs. parieto-occipital approach, initial vs. revision shunt and pre-op ventricular width at the entry site. Cox regression analysis showed only pre-op ventricular width at entry site predicted shunt failure ($p=0.0265$). Subsequent Kaplan–Meier survival curves showed significant survival advantage when ventricular width >15 mm compared to width <15 mm ($p=0.0035$). All other risk factors did not show biostatistical significance.

Conclusions: In our small series, risk factor analysis demonstrated that only preoperative ventricular width is a significant predictor of shunt survival if larger than 15 mm at the entry site. Careful assessment of preoperative ventricular anatomy may be useful in predicting survival advantage of endoscopically guided shunt insertion or revision.

Efficacy of aqueductal reconstruction in obstructed aqueduct and compartmentalized hydrocephalus

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Introduction: Aqueductal obstruction is encountered in hydrocephalus associated with idiopathic aqueductal stenosis (AS), tectal tumors (TT), Dandy–Walker malformation (DWM) and isolated fourth ventricle (IFV). The efficacy of aqueductal reconstruction techniques (ART) have not yet been well established in the literature.

Methods: From 1999 to 2010, 57 patients with aqueductal obstruction underwent endoscopic ART procedures. Thirty-four had AS, 12 had IFV, 6 had TT, and 5 had DWM with AS. A frontal transventricular approach was used in all. Aqueductal stent (AST) was placed in 42 patients and aqueductoplasty (APL) was performed in 15. Patients with IFV and DWM simultaneously underwent either ETV or shunt procedure.

Results: The follow-up period ranged from 1 to 8 years with a mean of 46 months. AST was successful in 83%. The stent was found to be malpositioned in four, obstructed in one, and proximal migration was seen in one. Six patients (40%) with APL had a procedural failure with four failing early and two late in the follow-up due to restenosis. There was no mortality. Two patients with APL had oculomotor disturbances while another with AST had transient unresponsiveness. The stent failures were significant in patients with AS (27%) than the non-AS (9%).

Conclusions: ART are relatively safe and have an overall success rate of 77%. Aqueductal stents appear to have a better success rate than aqueductoplasty (83% vs. 60%) in the present series.

Endoscopic third ventriculostomy through lamina terminalis—a feasible alternative

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Objective: Endoscopic third ventriculostomy for treatment of obstructive hydrocephalus is commonly being done. Sometimes the ventriculostomy can be difficult due to anatomic variations in third ventricular floor. Here, we describe an alternative approach for this procedure through lamina terminalis.

Methods: We have demonstrated endoscopic third ventriculostomy through lamina terminalis in four clinical cases. The most common cause for obstruction being congenital aqueductal stenosis (in three out of four cases) and one patient had cerebellar tumor with meningitis causing obstructive hydrocephalus. Third ventricle anatomy including optic chiasma and lamina terminalis was completely delineated using both rigid and flexible neuroendoscope. The end point of ventriculostomy through lamina terminalis was visualization

of anterior cerebral arteries. An alternative approach through lamina terminalis was opted for ventriculostomy in cases whenever there is technical difficulty in doing third ventriculostomy like where the floor of third ventricle was very small or is in close contact with basilar artery or the prepontine distance was less and subarachnoid cistern is adherent and not visualized because of meningitis (videos).

Results: All patients except patient with cerebellar tumor showed significant improvement in symptoms and neurological condition. No patient required diversionary procedure or revision of third ventriculostomy.

Conclusion: Endoscopic third ventriculostomy through lamina terminalis is a feasible alternative approach which can be used in selected cases.

Skull growth in hydrocephalic children

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Background: Despite that abnormal skull size is often noticed in the shunt-treated children, we are not aware of any systematic study of skull growth after shunt treatment. Overdrainage is of major clinical significance and might cause complications. The aim of this retrospective study was to investigate the skull growth in children treated for hydrocephalus (HC) during infant period up to the age of 4 years.

Methods: After exclusions (tumors, malformations) 62 consecutive children from western Sweden, treated for HC before 12 months surgery were included. Current results of this study are based on 36 of these patients. For each child, we obtained the medical records to collect data for head circumference, length, and weight at nine defined ages up to 4 years. In case of prematurity, it was corrected to the expected day of birth (w 40+0). We also recorded aetiology, valve type, number of revisions, valve setting, number of adjustments, time, and type of first surgery. Reference data of normal head growth, excluding low-birth weight infants, were obtained from Swedish national reference values of postnatal growth.

Results: The skull growth in the shunt-treated boys was impaired with significantly smaller head circumferences than the reference population. Similar findings were seen in children with HC caused by haemorrhage, with non-adjustable shunt systems, without shunt revisions, and in children with early primary shunt placement.

Conclusion: Preliminary results of this study indicate risk factors for overdrainage. This is highly momentous in aspect of clinical follow-ups and choice of and level adjustments of the shunt valves in hydrocephalic children.

The accuracy of endoscopically assisted ventricular catheter placement in children

Christoph Wiegand, Niels Sörensen, Wolfgang Wagner (Ped. Neurosurgery, University Mainz, Mainz, Germany)

Ventricular catheter placement in children for shunting reasons can be challenging, e.g. in slit ventricles. We report a series of $n=43$ patients with non occlusive hydrocephalus from 2008 to 2010. The youngest was 2 months, the oldest 14 years old. Most of the catheters were placed occipitally, a minor rate was done from Kocher's point. Only a few of them received Rickham devices for post haemorrhagic or chemotherapeutic reasons.

Occipital or frontal ventricles were localised via ultrasound and punctured by a 3-mm peel away catheter (Braun/Melsungen). When intraventricular, the inner tube of the catheter was removed and a 2 mm 30° optic (Aesculap Minop 2) was introduced and the outer layer was then endoscopically guided either to Foramen of Monroi (frontal route) or forwarded to the frontal horn entering the ventricle from Frazier's burr hole. All procedures were complication free (no infection/no bleeding from plexus). All catheters were controlled postoperatively either with ultrasound or CT scan and showed high accuracy with no malpositioning.

In conclusion, this minimally invasive technique is secure and easy to perform, showing the point of entry as well as the final catheter position avoiding intraventricular lesion by guiding the peel-away catheter endoscopically above the plexus choroideus. In general terms, even postoperative CT scan can be withdrawn avoiding obsolete radiation for children. In difficult Chiari scenarios where ventricular catheter placement with slit ventricles might be difficult and tricky, this is a precise procedure under accurate endoscopic visualisation.

Endoscopic intervention for fourth ventricular neurocysticercosis in children: an experience of a tertiary level hospital from a developing country

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Introduction: Fourth ventricular neurocysticercal cyst (FVNCC) usually presents with acute hydrocephalus necessitating neurosurgical intervention. It is uncommon in children and there is no clear consensus on its management. Authors share experiences of the endoscopic intervention (EI) for FVNCC in children; to the best of our knowledge, this is the largest study in the English literature. **Materials and methods:** A retrospective review of nine pediatric patients, who underwent EI using Gabb (Karl–Storz) neuroendoscope for FVNCC at our center. Using single pre-coronal burr hole, FVNCC was approached through transventricular-*trans* aqueductal technique.

Results: The mean age of patients was 13 years with male predominance. Cyst excision (CE) with ETV was performed in six patients, while one patient underwent only CE. One patient had partial CE because of ependymitis. In two patients, cyst could not be excised because of intraventricular (IVH) and haziness of cerebrospinal fluid (CSF), both underwent ETV only. One patient had pre-operative VP shunt, which was removed during EI. Shunt was avoided in all except one patient, who required VP shunt secondary to failed ETV, the only EI because of introp IVH. All patients are doing well on mean available follow up of 27 months, irrespective of the kind of EI.

Conclusion: EI is safe, effective, and avoids shunt and associated complications in majority of these children. ETV with CE is preferable, CE should be avoided where seems difficult. Restoration of CSF pathway by ETV should be the primary goal as it is equally effective as a sole intervention in select cases, irrespective of the CE status.

Pre-resectional endoscopic third ventriculostomy in the management of pediatric posterior fossa tumours Taopheeq Rabi (Federal Medical Centre, Ido-Ekiti, Nigeria)

Background: Most children with posterior fossa tumors have clinically important hydrocephalus at presentation. Traditionally, many of them are treated with ventriculoperitoneal (VP) shunting prior to tumour resection. There are ongoing debates about the efficacy of pre-resectional endoscopic third ventriculostomy (ETV) in the care of these patients. This study evaluates the usefulness of ETV in this category of children as presented in current global literature.

Methods: A PubMed search was performed for studies reporting the use of ETV in the management of hydrocephalus prior to tumor resection in children with posterior fossa tumors. Data on the success rates of ETV as well as its complications were collated and analyzed.

Results: Nine studies from diverse geographical areas were identified. A total of 224 children were studied. The average reported success rate was 89.7% (range, 65.0–100.0%). The reported complications were meningitis, intraventricular hemorrhage, cerebrospinal fluid leak, reversed herniation, and orbital emphysema in two, four, one, one, and one patient, respectively. There was no reported mortality.

Conclusion: ETV is useful in the treatment of hydrocephalus prior to tumour resection in children with posterior fossa tumor. Its high success rates and extremely low morbidity rates make it ideal for symptomatic control before definitive tumor surgery in the pediatric population.

Session 12. Oncology 1

Low-grade glioma in children: surgical treatment, technological advances and outcomes over a 55-year period in North-West England

Radhika Jassal, Nii Addo, Ola Rominiyi, Ian Kamaly, Jillian Birch, Robert Alston, Edward Estlin (University of Manchester, Manchester, UK)

Purpose: Low-grade glioma (LGG) is the most common malignancy of the central nervous system in children. We aimed to examine the outcomes for children with LGG diagnosed over 55 years, to identify the possible importance of advances in neurosurgical technology and other treatments for prognosis.

Study design: The case notes of 554 patients presenting with a low-grade glioma from 1954 to 2008 in the North-West of England were analysed.

Results: The mean age of presentation was 7 years, the distribution was such that 18% of patients were under three, 49% were between 3 and 10, and 33% were over 10 years of age. The male to female ratio was 1:4 and 48% of tumours were supratentorial, 38% cerebellar and 9% arose from the brainstem. The number of total/nearly total resections has increased over the past 50 years, from an average of 10% to 20% of cases. The 5-year overall survival rates have improved from 24% in 1954 to 88% in 2008. Older age at diagnosis, a non-axial site of disease and a diagnosis made in the era of the MRI scan all relate to a more favourable prognosis. This may relate to surgical and supportive care factors as radiotherapy and chemotherapy has been used less in the past 15 years than their peak in the 1970s and early 1980s.

Conclusion: The prognosis for low-grade glioma in the north-west has improved steadily over the past 55 years. Changes in surgical techniques, technology and healthcare delivery could have led to the increase in survival seen.

Basal ganglia germ cell tumor in children: report of 42 cases

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Background: Germ cell tumor originating in the basal ganglia is rare. It is notorious for the diagnostic difficulty and the majority of patients present symptoms similar to multiple sclerosis or stroke.

Materials and methods: We retrospectively reviewed the clinical features, neuroimaging studies, tumor markers, management, and outcome of these 42 patients from 1985 to 2011 in Taipei Veterans General Hospital.

Results: Thirty-nine of them were boys and three were girls. The median age of onset of symptoms was 11 years. Thirty patients presented with hemiparesis, seven with headache and

nausea/vomiting, and five with precocious puberty. Twenty-nine patients have histopathological diagnosis: germinoma in 18, teratoma in 2, and other type of tumors in 9. Four patients had bilateral basal ganglia tumors. Postoperative primary adjuvant therapies included radiotherapy, chemotherapy alone, and combined chemotherapy and radiotherapy. Thirty-nine patients survived and three patients died. Local recurrence was observed in all of the patients received solitary chemotherapy only.

Conclusions: The basal ganglia is a significant locus for intracranial germ cell tumor and can be bilateral. The initial image finding may be subtle and it should be highly suspicious when hemiparesis or precocious puberty presented in children. Although rarely reported in Western countries, it does exist in Taiwan as well. Treatment of germ cell tumor in specific location is similar to germ cell tumor in other intracranial locations.

Intracranial tumors in infants: long-term survival, functional outcome, and its predictors

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Objective: Intracranial tumors are rare in the first year of life. This study evaluates the survival and functional outcome of survivors at least 5 years after diagnosis, in addition to the predictors of this outcome.

Methods: A retrospective chart review of infants (<1 year age) diagnosed to have intracranial tumor. Radiology and pathology were re-reviewed. Outcome was assessed at 5 years or more using Bloom's categories (Bloom 1–2=good, Bloom 3–4 and dead=poor) and Late Effects Severity Scoring. Age, tumor location, size, extent of tumor resection, type of adjuvant therapy given, and World Health Organization (WHO) grade of tumor histology were evaluated as predictors of outcome.

Results: Among 35 infants, 12 (34%) had a good outcome, and 23 had a bad outcome. Among the survivors, 14 of 20 (70%) had neurologic deficits, five (25%) had endocrine deficits, nine (45%) had visual deficits, and three (15%) had auditory deficits. Ten of the 20 survivors were either attending normal school or were engaged in a skilled job. Older age and infratentorial location of the tumor are predictors of poor outcome at presentation. After histopathological diagnosis, the WHO grading of tumor is the only independent predictor of outcome and survival ($p < 0.001$). **Conclusion:** About a third of the infants diagnosed with brain tumors (34%) will have good functional outcome and approximately a quarter of them (28%) will be able to attend regular school or take up a skilled job. After tissue

diagnosis, histological grade of tumor is the only independent predictor of outcome.

Paediatric brain tumour resection in a single centre—comparison of outcomes

before and after the advent of intraoperative MRI

Shivaram Avula, Laurence Abernethy, Barry Pizer, Dawn Williams, Sasha Burn, Conor Mallucci (Alder Hey Children's hospital NHS Foundation Trust, Liverpool, UK)

Aim: This study aims to compare the surgical and imaging outcome in children who underwent brain tumour surgery with the intention of complete tumour resection, prior to and after the commencement of intraoperative magnetic resonance imaging (ioMRI) service at a single tertiary level paediatric neurosurgical centre.

Method: Our ioMRI facility is a 3 T MR scanner located adjacent to the neurosurgical theatre (two room solution) and has been operational since October 2009. We evaluated 36 consecutive cases (group A) in whom the surgical aim was complete resection up to October 2009 and the following 27 cases (group B) with a similar surgical aim after implementation of the ioMRI service. The surgical and imaging data among the two groups were compared

Results: In group A, definite residual tumour was identified on the post-operative scan (within 48 h) in seven cases (20%); and in group B in six cases (22%) at the stage of the first ioMRI.

Five (14%) children with residual tumour among group A underwent potentially avoidable repeat resection (two as early second-look procedures within 2 weeks and three as delayed operations on progression of residual tumours within 6 months).

Of the six children in group B with residual tumour, the resection was extended in four cases following ioMRI. None of these six children have required repeat resections on follow up.

Conclusion: On comparison of surgical outcome among paediatric, brain tumour patients where complete resection was intended, 14% of patients with residual tumour required second resections prior to introduction of ioMRI whereas none among a similar group required repeat resection after the advent of ioMRI.

Quality of life in massive opticohypothalamic glioma. An analysis of an experience in 73 cases treated over 18 years

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Background: Gliomas of the visual pathways represent 1–5% of childhood brain tumors. About 43% occur in children <10 years old. The treatment approach is controversial. Although benign, opticohypothalamic chiasmatic gliomas can have aggressive course in young children.

Patient and methods: Seventy-three cases of opticohypothalamic chiasmatic underwent surgery in our department

over the past 18 years (1990–2008). A retrospective analysis is presented.

Results: The average age of presentation was 14.8 years and duration of complaints was 3 months. Headaches, vomiting, and diminution of vision were commonest complaints. The average size of the tumor was 7.4 cm. All patients underwent only biopsy of the tumor. The subfrontal approach was used in all patients. Ventriculoperitoneal shunt was required in 19 patients postoperatively. Postoperative morbidity was transient and minimal (diabetes insipidus and intraventricular hemorrhage in two patients each). There was mortality in two patients. Postoperatively, all patients above 4 years of age were administered radiotherapy. The average follow-up period was 7 years. Nonsymptomatic recurrence was noted in eight patients. Overall, the median time to tumor progression was 55 months. The 5- and 10-year survival probabilities were 93% and 74%, respectively.

Conclusion: Radical removal of opticochiasmatic gliomas offers little benefit in terms of progression-free survival or decreasing neuroendocrine complications. It improves the likelihood of controlling hydrocephalus without damaging vision on the contralateral side. Younger patients (<5 years) and the female gender were high risk factors for disease progression. Multimodality therapy (surgery with radiotherapy/chemotherapy) may be needed in the high-risk group.

Choroid plexus tumors in children: a review of 78 cases

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Objectives: Tumors of the choroid plexus are rare tumors of neuroepithelial tissue, accounting for less than 1% of all intracranial tumors. Most cases present in children less than 3 years of age.

Methods: Between 1980 and 2009, 78 children with choroid plexus tumors were treated at the Pediatric Department of the Institute of Neurosurgery. This presented 1.7% of all pediatric brain tumors diagnosed during this time period. Forty-two were males and 36 females.

Results: There were 63 choroid plexus papillomas, 13 atypical choroid plexus papillomas, and two choroid plexus carcinomas. The age of the patients ranged from 3 months to 17 years (57.6% in the infants). In 51 patients, the tumor was in the lateral ventricle (41 left, 10 right); in eight, it was in the third ventricle and 19 in the fourth ventricle.

The surgical removal of the lesion was performed in 60 (77%) patients: total removal, 59; subtotal removal, 9; and partial removal, 2. In 15 cases, after tumor excision were performed CSF-shunting operations. Operative mortalities the surgical failure seems to have due to large tumors and blood loss. Follow-up data from 2 months to 18 years is available for 77% patients.

Conclusions: The management of these children is frequently difficult and complicated by dilemmas associated with the choice of surgical procedure, the treatment of hydrocephalus and the value of adjuvant therapy. We believe the aim of treatment should be to achieve a complete resection.

Medulloblastoma in children: surgical treatment, technological advances and outcomes over a 55-year period in North West England

Ola Rominiyi¹, Nii Addo¹, Radhika Jassal¹, Eddy Estlin², Jillian Birch¹, Robert Alston¹, Ian Kamaly² (¹The University of Manchester, Manchester, UK; ²Royal Manchester Children's Hospital, Manchester, UK)

Purpose: The best current management of medulloblastoma using surgery, radiotherapy and chemotherapy has provided 5-year progression-free survival (PFS) of up to 85%. We aimed to evaluate the influence of various prognostic factors, including new imaging modalities and adjuvant therapies, on survival outcomes over a 55-year period.

Study design: Case notes of 220 patients diagnosed with medulloblastoma between 1954 and 2008 were evaluated retrospectively. Patients were stratified according to date of diagnosis into five 11-year time periods. Kaplan–Meier statistics were used to analyse survival data, with differences in survival assessed using the log-rank test. Multivariate analysis to determine factors affecting prognosis was performed using a Cox proportional hazards model.

Results: The age range was 0.3–15 years (median 6.6 years). The male/female ratio was 1.65. One hundred four patients received gross or near-total resection, 77 had subtotal resection, and 20 patients had only a biopsy. Five-year PFS improved from 34.1% to 88.5% for children diagnosed between 1954–1964 and 1998–2008, respectively ($p < 0.001$). Ten-year survival rates demonstrated similar trends.

Postoperative treatment had the most significant effect on prognosis ($p = 0.001$) with 5-year PFS of 83.9% for children diagnosed after 1998 receiving postoperative radiotherapy and chemotherapy; followed by the extent of surgical resection ($p = 0.006$). The presence of metastases at presentation was highly predictive of poor outcome.

Conclusions: Survival outcomes for childhood medulloblastoma are improving. For this long-term follow-up cohort, adjuvant therapy followed by extent of surgical resection had the most significant impact on survival.

A preliminary evaluation of the utility of soluble c-kit (s-kit) as a cerebrospinal fluid (CSF) marker for intracranial germinomas

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Aim: This study aims to determine the sensitivity and specificity of soluble c-kit (s-kit), a proto-oncogene encoding tyrosine kinase receptor for stem cell factor, which has been identified in germinoma, as a cerebrospinal fluid (CSF) marker for intracranial pure germinomas.

Method: CSF samples were obtained during initial treatment of tumoural hydrocephalus from consented patients whose tumours were subsequently confirmed histologically to be germinomas. The CSF samples were analysed for s-kit, using commercially available diagnostic enzyme-linked immunosorbent assay (ELISA) kits. Control CSF samples were drawn from consented patients who underwent surgical procedures for non-germinomatous tumours and for hydrocephalus. The diagnostic capacity of the ELISA test for s-kit were analysed using the receiver curve characteristic (ROC) curve analysis.

Results: The concentration of soluble c-kit in cerebrospinal fluid samples of four patients with pure germinomas, one with secretory germ cell tumour (GCT; raised β -HCG) and one with mixed GCT were analysed for s-kit using ELISA. Eight non-GCT cases were recruited as control. The sensitivity and specificity of CSF s-kit assay for pure germinoma is 66.7% and 90.9%, respectively, at a cut-off level of 7.3229 ng/ml. The area under the ROC curve, AUC is 0.667 ($p = 0.392$, 95% CI 0.133–1.200).

Conclusion: The concentration of soluble c-kit in CSF appears to be a suitable marker for the detection of intracranial pure germinomas. Nevertheless, the number of germinoma cases in this study are small and a larger study with more cases will be useful to determine the usefulness of this marker.

Posterior fossa tumours and gait abnormality in children

William Lo, Rebecca Davis, Giovanna Paternoster, Umar Farooq, Patrizia Pisano, Guirish Solanki (Birmingham Children's Hospital, Birmingham, UK)

Introduction: Normal gait is a complex movement, dependent on proprioception, balance, strength and coordination. Gait is often impaired in children with posterior fossa tumours. This relationship is poorly understood and requires elucidating.

Objectives: This study aims to determine the incidence of gait abnormality at diagnosis and post-surgery and identify contributing factors.

Methods: This was a retrospective review at a regional paediatric neurosurgery centre over 5 years. Patients with physiotherapy notes available underwent further analysis. Grading of gait was based on Brief Ataxia Rating Score.

Results: There were 35 patients (male 22, female 13; M:F ratio=1.7) with median age of 70 months. Thirteen were pilocytic astrocytomas (35%), 13 medulloblastomas (35%), 6 ependymomas (16%), and 3 others (8%). Main presenting symptoms were: 24 gait abnormality/motor regression (69%), 21 headache (60%), and 16 vomiting (46%).

Younger patients (56 vs. 147 months) and those with a longer history (68 vs. 47 days) were more likely to present with gait abnormality. Gender, histology, World Health Organization grading, tumour size, location and hydrocephalus were not associated with gait abnormality.

Subjects with physiotherapy notes were divided into those presenting with gait-related problems (group I=12) and those without (group II=8). In group I, five (41%) improved, six (50%) unchanged, one (8.3%) worsened. In group II, seven (88%) remained unchanged and only one (13%) deteriorated ($p=0.035$).

Conclusions: Gait abnormality is common in PFTs. Children with gait abnormality are younger and have a longer history.

CSF cytology and Spine MRI in diagnosis of metastatic seeding in posterior fossa tumours: do they identify different types of metastatic disease?

Patrizia Pisano, William Biu Lo, Giovanna Paternoster, Guirish Solanki (Birmingham Children's Hospital, Birmingham, UK)

Aim: Leptomeningeal metastatic disease (LMD) significantly affects the prognosis of posterior fossa tumours (PFT). Cerebrospinal fluid (CSF) examination for malignant cells and spinal MRI are routinely used to diagnose metastasis. It is unclear if CSF examination is useful for all tumour types and their current use is generally limited to primitive neuroectodermal tumors (PNET)/medulloblastoma.

Patients and methods: We studied 59 patients with PFT (33 medulloblastoma, 12 ependymoma, 12 pilocytic astrocytoma, 2 atypical teratoid/rhabdoid tumour (ATRT)). Median age was 7 (range, 1–14) years. Eight patients with metastasis at diagnosis were excluded: seven medulloblastomas, one pilocytic astrocytoma. CSF cytology and spinal MRI reviewed independently for the presence of LMD. Post-operative brain and spine MRIs are performed 6-monthly; median follow-up, 17 months. CSF for cytology obtained by lumbar puncture (47), ventricular (3), and intra-operative CSF (9).

Results: Five of 51 children, medulloblastoma (2), ependymoma (1), pilocytic astrocytoma (1), ATRT (1) were CSF-positive for malignant cells. Median time to diagnosis was 6.5 (range, 0.5–26) months post-operative. There was no LMD on MRI in all five cases. In six of 51 children, medulloblastoma (5), ATRT (1), MRI showed LMD at median interval of 17 (range, 4–32) months. Metastasis was synchronous with recurrence in two. CSF cytology was negative in all six cases. MRI noted subpial plaque/nodular mass lesions.

Conclusion: Overall, 11 patients (20%) had post-operative metastasis. CSF/MRI findings were discordant in all cases. Nodular/subpial lesions were detected by MRI. Leptomeningeal disease (without nodular lesions) was diagnosed by CSF-cytology. Interval to CSF-seeding was half that of MRI-detected lesions. CSF-cytology also identified metastasis in

different histology types (besides medulloblastoma), suggesting the need for CSF sampling of all PF tumours. All metastasis but one occurred in grade III and IV tumours.

Hyperthermia as an end-stage salvage treatment for recalcitrant pediatric malignant brain tumors

Charles Teo², Michael Jackson¹ (¹Univ. of NSW, NSW, Australia; ²Centre for Minimally Invasive Neurosurgery, NSW, Australia)

Complimentary alternative treatments (CATS) come and go like fads, some with miraculous anecdotes but sadly, most with a paucity of good results. Given the dismal survival times of children with malignant cerebral neoplasms, especially after failed standard chemo and radiotherapeutic options, CATS are frequently sought by desperate parents. Hyperthermia as a generic treatment option for all cancers, works on the basis that cancer cells are more susceptible to heat than normal cells. The heating may be administered in many forms with very few side effects. In the last 10 years, 12 patients, eight adults and four children, have been referred to the hyperthermia center in Germany, initially patient driven but recently physician instigated. This paper will only present the results of the pediatric patients. Of the four patients, three had dramatic and sustained results and are alive and well. The fourth child had no discernible benefit and died shortly after the completion of treatment. Patient 1 had widely metastatic primitive neuroectodermal tumors within the subarachnoid space after failed BST. Patient 2 had rapidly progressive pilomyxoid astrocytoma of the hypothalamus after failed radical surgery and BST and patient 4 had rapidly progressive widely metastatic malignant ependymoma after BST and experimental Avastin. All three patients are alive and well, 10 years, 4 years, and 6 months after treatment.

This paper will briefly discuss the mechanism of action, review the literature and show the pre and post-treatment imaging of all the patients.

Microsurgical treatment of giant pituitary adenomas in children and adolescents

Avijit Sarkari, Sumit Sinha, Ashok Kumar Mahapatra (Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi, India)

Background: Pituitary tumors are rare in childhood and adolescence, with a reported prevalence of one per million children comprising only 2% of pediatric brain tumors. Giant pituitary adenomas (>4 cm) are a surgical challenge. We present our experience in surgical management of these tumors in the pediatric age group.

Methods: Eleven patients (seven male and four female) with giant pituitary adenomas in the age range 12–20 years were managed surgically over a period lasting 5 years. Visual

deterioration (72.7%) was the most common presentation. Headache was a feature in 63.6% with pituitary apoplexy in 18.1%. Acromegalic features were present in 36.3%. Two females and one male had secondary amenorrhoea and delayed puberty respectively. One male had nonfunctioning tumor. Among functioning adenomas (90.9%), six patients (63.6%) had prolactinomas and four (36.3%) had growth hormone (GH)-secreting adenomas.

Results: Overall, 15 surgical procedures were performed. All 11 patients were operated trans-sphenoidally. Three had post-operative cerebrospinal fluid leak—two requiring re-exploration and packing. Two cases of GH-secreting adenomas had recurrence, reoperated transsphenoidally. Visual improvement occurred in 50% cases. One patient had post-operative meningitis managed conservatively. Three cases of GH-secreting adenomas required post-operative radiation.

Conclusions: Majority of giant pituitary adenomas in childhood and adolescent age groups are of functioning type, prolactinomas constituting majority (63.3%) as compared to adults (prevalence about 30%). Most are treatable by trans-sphenoidal route. The recurrence rate is high (18%). Surgery decompresses optic pathway improving vision and reduces tumor volume offering the best chances of tumor control with adjuvant therapies.

Orbital tumors in children: 121 cases

Suzana Serra, Izabel Eugenia Castelo Branco, Artur Da Cunha, Fatima Griz (Hospital da Restauração, Recife, Pernambuco, Brazil)

The object of this paper is to show 121 cases of infant orbital tumors over a 25-year period. In our series, the tumors were more frequent in girls and the ages of the children ranged from 2 months to 16 years. Pain, proptosis, palsy of the external movements of the eyes, blindness, and ulcers were the often symptoms. In the diagnostic evaluation, we used anamnesis dates, test of the 7 ps, neuro-ophthalmologic exam and nuclear magnetic imaging. The most frequent type of the tumors was pseudotumors (32), followed by optic glioma (18). Seventy-two operations were performed, 12 of which being biopsies, and in 57 cases the conservative management were the choice. The cases selected for surgery were the patients with progressive proptosis, blindness, external eyes movement deficits, and the cases with inconclusive diagnosis.

Initial experience with endoscopic side cutting aspiration system in pure neuroendoscopic excision of large intraventricular tumors.

Bobbye Thompson, Joel Patterson, Aaron Mohanty (University of Texas Medical Branch at Galveston, Galveston, Texas, United States)

Conventionally, neuroendoscopic excision of intraventricular tumors has been difficult and time consuming due to lack of an effective decompression system that can be used through the working channel of the endoscope. The authors report their initial experience in pure endoscopic excision of large intraventricular tumors with the minimally invasive NICO Myriad system.

The NICO Myriad is a side cutting soft tissue aspiration system which employs an inner reciprocating canula and an outer stationary sheath with a side port. During decompression, applied suction approximates the tumor into the lumen of the outer sheath with the inner canula excising the tissue by oscillation of the cutting edge. The tumor is then removed by aspiration through the inner sheath. Three patients with large intraventricular tumors were operated by a pure endoscopic approach using a Gaab's rigid endoscope and the NICO Myriad system. Of these, 2 had intraventricular craniopharyngiomas and one had a lateral ventricular subependymoma.

The tumor size varied between 1.9 and 4.5 cms in the largest diameter. A relatively firm and solid tumor was encountered in two and a multicystic tumor with thick, adherent walls in one. The tumor could be subtotally removed in one and near totally in two. The mild bleeding encountered during dissection and excision was controlled with irrigation. There were no complications.

The NICO Myriad is a highly effective tumor decompression system which can be effectively used in pure endoscopic approach to intraventricular lesions.

Pediatric lateral ventricle tumors: an institutional experience of 10 years

Yashpal Singh Rathore, P S Chandra, A K Mahapatra (All India Institute of Medical Sciences, New Delhi, India)

Tumors of the lateral ventricle comprise between 0.8% and 1.6% of all brain tumors and more commonly observed in children. Authors reviewed 47 children of lateral ventricle tumors operated at our Institute from Jan 2001 to Dec 2010. There were 31 boys and 16 girls. Age range was from 3 months to 20 years. There were 14 patients in age group 5–10 years and 16 patients in age group 15–20 years. The most common symptoms were headache, vomiting, vision loss, and seizure. There were 11 choroid plexus tumors (seven choroid plexus papilloma and four choroid plexus carcinoma), eight neurocytoma, six astrocytoma, seven ependymoma, five subependymal giant cell astrocytoma, five ganglioglioma, four meningioma, one epidermoid. One patient expired in post-operative period. Out of 47 patients operated, 19 patients required shunt. Surgical approach to the lateral ventricle was chosen on multiple factors, including localization of the tumor within the ventricle, presence or absence of hydrocephalus, whether the hemisphere involved is dominant, size of the

tumor, origin of the blood vessels supplying the tumor, and histopathologic features of the tumor. The most common site of tumor origin was in frontal horn, body and trigonal region of lateral ventricle. The most common complications were vision deterioration, hemiparesis, intraventricular hemorrhage, meningitis, seizure and development of subdural effusion. Lateral ventricle tumors in children are predominantly benign and are very large at presentation. Treatment of choice is gross total excision. The details will be presented.

Endoscopic microneurosurgery using rigid suction mounted endoscope

Sandeep Sood (Children Hospital of Michigan, Wayne State University School of Medicine, Detroit, MI, USA)

Improved magnification of an endoscope has been used in combination with traditional open microsurgical operation to locate and resect residual lesions not otherwise appreciated under microscope. However; a key limitation of traditional current endoscopic technology is inability to bimanually manipulate to allow for effective dissection. As a result, using instrumentation through the coaxial channel is effective only for biopsies and small relatively avascular tumors. These limitations are somewhat overcome by using a 1.5-cm port through the brain to the ventricle. Fixed position of the endoscope, small working space, scissoring of instruments, need to traverse brain tissue and small ventricles are some of the obstacles. To overcome these obstacles, we have developed a rigid suction mounted endoscope that is used in the left hand and a second instrument in the right hand similar to microsurgery. Approach to intraventricular tumors is inter-hemispheric through a small craniotomy and for pineal region tumors is through a subtorcular supracerebellar approach. Six patients were operated, three with large intraventricular and three with a pineal region lesion. Complete radical resection was achieved in all cases. Similarity to microneurosurgery combined with the smaller endoscopic approach and greater magnification are the major advantages of this technique.

Posterior fossa durotomy—clinical significance of occipital sinus

Manas Panigrahi, Shyamsundar K, Phaniraj GL, Venugopal Reddy, Krishna Reddy, Chandrasekhar Naidu (Krishna Institute of Medical Sciences, Hyderabad, India)

Introduction: Standard posterior fossa durotomy involves division of occipital sinus from the marginal sinus. Many variations of occipital sinus and complications due to ligation of occipital sinus have been reported in literature. The aim of this presentation is to analyse the normal anatomy and variations of sinus. Emphasis shall be on identifying non-invasive investigation to predict dominance

of occipital sinus. Finally, suggest alternative method of posterior fossa durotomy to preserve occipital sinus.

Materials and methods: Fifteen patients having posterior fossa tumors were included in the study. External ventricular drain was placed during surgery whenever they had hydrocephalus. The dura was opened parallel to the occipital sinus and extended down below the foramen magnum by cutting the marginal sinus in paramedian plane. The dural flap was reflected based on the occipital sinus and contralateral marginal sinus.

Results: The exposure of the tumor was adequate for total excision. Cerebellar edema was not encountered either intra or post-operative period. Dural closure was easier and none of them had any post-operative cerebrospinal fluid leak. Patients with hydrocephalous did not require external ventricular drain for more than a day.

Conclusions: Prophylactic preservation of occipital sinus does not compromise the extent of excision. One can avoid inadvertent damage of dominant occipital sinus and its complications. Visualisation of occipital sinus in magnetic resonance venography indicates dominance of the sinus.

Early diagnosis and combined treatments on sellar germinoma

Ge Jia (Beijing Tiantan Hospital, Beijing, China)

Objective: This study aims to summarize the early diagnosis and treatment on the germinoma at sellar region. **Methods:** Retrospective analysis on 23 cases of sellar germinomas, eight males and 15 females. Age ranged from 4 to 26 years old (mean, 12.4 year). The primary symptoms were diabetes insipidus (DI) in 21 and blurred vision in two cases. Eight cases (34%) showed serum β -HCG slightly increase. Operations were performed on nine cases confirmed by histopathology and 14 cases were diagnosed by the features of clinical manifestation (CM), imaging and tumor markers. All the cases followed chemo- and radiotherapy.

Results: The early imaging showed abnormal signal at stalk hypophysial and the only one CM was DI, and at the late stage the mass effect was shown. The prognosis was not significantly different between the group of operation combined with adjunctive therapy and the group of single adjunctive therapy. Blurred visions were improved and the DI and hypoevolutism were not relieved.

Discussion: When diagnosis was not clearly combined with vision impairment, operation was recommended to decompress the optic nerve compression and get the histopathological diagnosis. Otherwise, the followed criteria are important diagnostic evidences: (1) young girls, (2) diabetes, insipidus, (3) stalk swelling, (4) β -HCG slightly increase, and (5) hypoevolutism. The slight increase in β -HCG is the most important, if not, diagnostic radiotherapy is recommended, and if the volumes shrink more than 50%, the diagnosis of germinoma is definite. Combination of chemo- and radiother-

apy on intracranial germinoma are the best choice for the patients.

Intracranial tumors in the first year of life

J. Francisco Salomao, Antonio Bellas, Tatiana Protzenko, Rene Leibinger (Fernandes Figueira Institute, Oswaldo Cruz Foundation (Fiocruz), Rio de Janeiro, RJ, Brazil)

A retrospective study of 57 patients with intracranial tumors in the first year of life is presented. There were 15 perinatal tumors and in nine of them an intrauterine diagnosis was made. Intracranial hypertension was noticed in 36 patients and 32 had macrocrania. In 37 patients, the lesions were located above the tentorium. Topographically, the most common site was the posterior fossa ($n=20$), followed by the chiasmatic/hypothalamic region ($n=14$). Overall, 37 tumors were found in the midline. In 38, the greatest diameter of the lesion exceeded 5 cm and in 12 was greater than 10 cm. Fifty patients were operated on but a total removal was only achieved in 16. Seven had staged surgeries. Four patients received chemotherapy before definitive surgical treatment was attempted. There were four surgical-related deaths. Histopathological diagnosis was available in 53 patients and medulloblastomas/primitive neuroectodermal tumors (PNETs; $n=12$), astrocytomas ($n=12$), and ependymomas ($n=7$) were the most frequent tumor types. The follow-up ranged from 9 months to 21 years. At the present time, 27 patients died, 19 are stable, and 6 were lost for follow-up. An analysis of our series showed that PNETs and ependymomas behaved quite aggressively and that patients considered cured or stable were those with benign lesions completely resected or slowly progressive due to their own peculiarities. Due to difficulties in surgical treatment and limitations of adjuvant therapy, operative technique should be individualized and take into account the age of the patient, type, size and vascularity of the tumors.

Thursday October 20, 2011

Session 13. Oncology 2

Neuronal differentiation and immunosuppressive effect of human adipose derived stem cells

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Purpose and methods: This study was conducted to analysis of immunosuppressive effect of human adipose derived stem cells (ADSC) for allogenic transplantation for the therapeutic purpose. We obtained adipose tissue from donors and isolated and cultured ADSC. We have examined phenotypic and gene expression profile of the human ADSCs by the flow cytometric analysis and induced

ADSCs to differentiate into the mesenchymal cells and neuronal cells. Immunosuppressive properties of ADSCs were analyzed by mixed lymphocyte culture and lymphocyte proliferative response to mitogens.

Results: Cultured human ADSCs express CD34, CD45, MHC class marker, HAL-DR and negative for the CD29, CD44 which is similar to that of bone marrow stem cells. Under appropriate conditions, were differentiated into the osteoblast, adipocyte, and neuronal cells. ADSC did not provoke in vitro alloreactivity of incompatible lymphocyte and suppressed mixed lymphocyte reaction. ADSC also suppressed proliferative response of lymphocyte to the mitogen such as PHA, ConA, and IL-II.

Conclusion: These findings support that ADSC has properties of stem cells and has immunosuppressive effect. ADSCs might have potentials for cell-based regenerative therapies for various neurological diseases and also for allogenic transplantation because of their immunosuppressive properties.

Characterizing PI3K/Akt/mTOR signaling pathways associated with growth and dissemination of medulloblastoma

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Medulloblastoma is the most common primary CNS tumor in children, which appears to originate from cerebellar granule neuron precursors. Recent studies have demonstrated several subtypes of medulloblastoma based on their genetic profiles. The Hedgehog-Patch-Gli pathway activates the oncogene *N-myc*, often seen amplified in medulloblastoma, conferring a poorer prognosis. In addition, studies have shown that Gli1-transformed epithelioid cells require mammalian target of rapamycin (mTOR) activity for their survival. Modulations in the mTOR signaling pathway in medulloblastoma promote the status of the CDK inhibitor, p27. We aim to investigate the involvement of PI3K/Akt/mTOR pathway as it relates to the proliferation and motility of medulloblastoma cells. We observed that significant number of tumors had an amplification of *N-myc*. Inhibiting mTOR with rapamycin resulted in a time-dependent suppression of mTORC1 substrate, pS6K; however, MAPK suppression was ineffective in inhibiting pS6K levels. CDK inhibitor p27 levels and localization were also influenced by PI-3 K/Akt/mTOR suppression in a time-dependent manner. Prolonged suppression of mTOR resulted in activation of MAPK, Akt, and p27. Moreover, inhibition of mTOR suppressed *c-myc* expression. Inhibition of mTOR suppressed medulloblastoma growth and suppressed TPA-induced proliferation. Additionally, MAPK inhibition suppressed PDGF-induced cell growth. The PI3K/mTORC1/2 complexes inhibition suppressed the migra-

tion of medulloblastoma cells toward fibronectin. In conclusion, the PI3K/Akt/mTOR pathways may influence medulloblastoma growth and dissemination, and provides a basis for therapeutic targeting.

Distinct molecular genetic alterations in cortical and cerebellar pilocytic astrocytomas

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Pilocytic astrocytomas (PAs) are the most frequent brain tumors effecting children. These astroglial tumors correspond to World Health Organization grade I and are mainly localized in the cerebellum. As it has been reported that the rare cortical localized PAs could have a less favorable outcome, it has to be discussed if their molecular background is distinct from PAs localized in the cerebellum. To address this question, we analyzed 24 cortical PAs and 32 cerebellar PAs for genetic and epigenetic alterations within (1) the *O6-methylguanine DNA methyltransferase (MGMT)* gene promoter, (2) the *isocitrate dehydrogenase-1 and -2* genes (*IDH1/2*), and (3) the *BRAF* gene encoding for a component of the ERK/MAPK pathway using the quantitative pyrosequencing method.

Methylation of the *MGMT* promoter was found in 30% of the cortical PAs while none of the cerebellar PAs was methylated. Further, we identified a significant increase of *BRAF* V600E and 464–469 mutations in cortical PAs when compared to cerebellar PAs. *IDH1/2* mutations, which are frequent in low-grade astrocytomas were not detected in any case of both groups, indicating that cortical PAs are separated from diffuse astrocytomas.

Our data reveal distinct genetic and epigenetic changes of the *BRAF* gene and *MGMT* promoter in cortical and cerebellar PAs. Thus, implementation of molecular assessment of *MGMT* promoter methylation and *BRAF* mutations represents a novel trail in diagnosis and therapy of cortical PAs.

Investigation of the molecular bio-markers related to the recurrence of adamantinomatous craniopharyngiomas

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Objective: This study investigates the specific Molecular Bio-Markers related to the recurrence of adamantinomatous craniopharyngiomas (ACP) and analyze the correlation between the level of these markers and ACP recurrence.

Methods: The tissue samples of ACP were collected and divided into two groups (primary group and recurred group). Gene expression profiles in primary group and recurred group

were detected and compared by microarray. The expression of changed genes was identified by immunohistochemistry (IHC) and quantitative real-time PCR (qRT-PCR) in 40 cases of ACP, including 24 cases of primary group and 16 cases of recurred group. The score system combining clinical data with the level of CXCL12 was established to predict the recurrent possibilities of ACP according to 60 samples.

Results: The levels of MMP-9, CXCL12, CXCR4 and VEGF in recurred group were significantly higher than primary group detected by qRT-PCR ($p < 0.05$). It was found that CXCL12 showed more specificity for distinguishing primary samples from recurred samples. In further investigation, the expression of CXCL12 was tested by IHC in 60 cases of ACP. According to the IHC results, combined with clinical scale and the recurrent rate, the score system for predicting the recurrent possibilities of ACP was established. The patients with the higher score had the more recurrent risk compared with those with the lower score.

Conclusion: The changed genes may be the potential specific biomarker related to the recurrence of ACP. The score system that we established may be a useful tool to predict the recurrent possibilities of ACP.

Three-dimensional anisotropy contrast periodically rotated overlapping parallel lines with enhanced reconstruction (3DAC PROPELLER) MRI: application to preoperative evaluation for brainstem tumor surgery

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Objective: The goal of this study is to preoperatively evaluate the growth patterns of brainstem tumors and to detect tracts or nucleus adjacent tumors using three-dimensional anisotropy contrast periodically rotated overlapping parallel lines with enhanced reconstruction (3DAC PROPELLER) magnetic resonance imaging (MRI).

Methods: Ten patients with brainstem tumors aged 9–69 years were enrolled. The tumor histologies included seven gliomas, two cavernous angiomas, and one hemangioblastoma. We performed routine MRI to detect tumors as well as 3DAC PROPELLER sequence and compared detailed information between tumors and normal structures. Craniotomy was performed in eight patients and autopsy was performed in one patient. We also examined the surgical outcomes and tumor histologies.

Results: All the three diffuse brainstem gliomas showed infiltrative growth. Necrosis and cysts were not detected by

3DAC. Necrotomy was performed in one patient without deficit. Autopsy of another patient revealed infiltrative tumor growth. Three of the four focal brainstem gliomas showed demarcated growth. One partial resection and three biopsies were performed in patients without deficit. Of the two cavernous angiomas, one tumor showed demarcated growth, and total excision was performed. Pyramidal tract was encapsulated by the tumor in the other case; hence, biopsy was performed instead of radical removal. The one hemangioblastoma showed demarcated growth and total excision was safely performed.

Conclusion: 3DAC PROPELLER method can be used to evaluate the growth patterns of brainstem tumors and the tracts or nucleus into the brainstem. This new modality might be considerably useful for the preoperative evaluation of brainstem tumors.

Impaired cognitive and motor function of pediatric patients with posterior fossa tumor is associated with decreased fractional anisotropy of supratentorial tracts

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Objective: Tumor and therapy-associated neurotoxic effects cause brain damage in pediatric patients with posterior fossa tumors. Treatment-related toxicity factors are resection in pilocytic astrocytoma (PA) and additionally cranio-spinal irradiation together with chemotherapy in medulloblastoma (MB) patients. We tested whether MR diffusion tensor imaging (DTI) derived fractional anisotropy (FA) of white matter tracts correlates with results of ataxia, IQ, and fine motor assessment.

Methods: Twenty-three and 17 PA survivors (mean age, 14.25 years; SD, 4.83) were subjected to MR imaging and DTI on a 3 T-MR system equipped with an 8-channel headcoil (3 T Signa Excite, GE Healthcare, 25 directions). Skeletonized FA maps excluding the cerebellum were generated using FMRIB Software Library. Mean FA of the supratentorial tracts was determined. Ataxia was quantified using the International Cooperative Ataxia Rating Scale (ICARS). Wechsler Intelligence Scale for Children was applied to determine IQ. Movements of handwriting were analyzed using a digitizing graphic tablet; speed and automation were determined employing CSWIN software.

Results: Mean FA correlated significantly (Spearman correlation) with movement parameters of handwriting (speed: $r=0.48$, $p=0.012$; automation: $r=-0.4$, $p=0.038$). Furthermore, a significant correlation was found of mean FA and IQ ($r=0.39$, $p=0.038$). No correlation between mean FA and

ICARS scores were found. All significant correlations indicated functional loss when supratentorial mean FA was decreased.

Conclusion: DTI is capable to quantify brain damage and is linked with the functional outcome of this patient group. Mean FA of supratentorial tracts is a representative marker for cognitive and motor deterioration.

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Origins, diagnosis and prognosis of WNT-positive medulloblastomas

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In recent years, molecular expression profiles have demonstrated that medulloblastoma comprises a number of different subtypes with a common histological appearance. The anatomical location as well as outcomes of children with these subtypes is variable. New treatment strategies are being based upon the molecular expression profile of these subtypes. The authors present a series of 24 children with WNT-positive medulloblastoma, as demonstrated by nuclear beta catenin positivity by fluorescence in situ hybridization, studied prospectively on protocol and with at least 2 years of follow up. There were 14 female and 10 males, average age being 10.5 years. Twenty-one children were average risk and three were high risk. Twenty-three of 24 tumors occupied the fourth ventricle on MRI. This subgroup of children currently has 100% progression-free survival at 2 years follow up. The authors will present a discussion of molecular subtypes of medulloblastoma, prognostic factors associated with these tumors, and propose a treatment strategy aimed at reducing the toxic side effects of treatment in the WNT subgroup of medulloblastomas, given their excellent prognosis

Surgical complications of posterior fossa tumour surgery in children: improving reliability and validity for routine monitoring of outcomes

Ram Kumar, Conor Mallucci, Jothy Kandasamy (Alder Hey Children's NHS Foundation Trust, Liverpool, UK)

Introduction: Complications following posterior fossa tumour surgery in children are reported to be common, with previous reports of 32–62% of children affected. There is often difficulty in deciding whether a child who experiences an adverse event or outcome after surgery has had so as a result of surgery, or some other factor such as the tumour itself or other treatments. This decision may rely on the personal judgement of the neurosurgeon and others in the team. This makes reporting of complication rates unreli-

able, and thus identifying the predisposing risk factors, and methods of reducing these risks. A method for improving consensus agreement of whether a complication has occurred is required.

Method and results: The authors have developed a causality assessment tool for adverse events following posterior fossa tumour surgery in children. This is built around similar work in our institution on identifying adverse reactions to drugs in children. The causality tool has been tested and developed further on a retrospective review of post-operative course in 67 children with posterior fossa tumours who were operated on in our institution with specific reference to suspected complications of surgery. The review of case notes has resulted in the development of proxy indicators of complications that may be useful in future routine monitoring from administrative data.

Conclusion: The posterior fossa tumour surgery adverse event causality tool is necessary for future multi-centre research studies on surgical complications, and for comparative effectiveness studies. The proxy indicators of complications are necessary for quality improvement and provider comparison.

Diffuse Pontine PNET: confounding the differential diagnosis of DIPGs

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Introduction: Diagnosis of typical diffuse pontine tumors, as a standard, is by scan. This cannot distinguish primitive neuroectodermal tumors (PNETs) of the pons from diffuse intrinsic pontine gliomas (DIPGs). We present two cases of PNETs diffusely in the pons and show molecular evidence these are indeed PNETs and distinct from biopsied atypical DIPGs.

Methods: Pediatric brain tumor samples were collected and snap frozen in the operating room. All were analyzed histologically, and for cluster analysis and aberrant gene expression via Ambion Microchip HG-U133plus2. Biopsied diffuse tumors of the pons were selected, comparing them against other intracranial tumors.

Results: The histology of five atypical diffuse tumors of the pons were consistent with malignant gliomas, and in two young children (<3 years) was consistent with PNET. Clustering analysis demonstrated that pontine PNETs were most closely related to PNETs of the supratentorial region, not clustering with the DIPGs in the study. Over 200 genes, many of which are involved in transcriptional regulation, were uniquely upregulated in brainstem PNETs compared to 10 other types of pediatric brain tumors, including DIPGs and other PNETs, at an FDR statistical significance of <0.01.

Conclusion: As shown by cluster analysis and individual gene expression analysis, pontine PNETs are distinct from

DIPGs. This suggests that these tumors should be separated from DIPGs in terms of treatment. Since diagnosis by scans is not reliable and the biology of the tumors is disparate, biopsies should be performed to enable proper diagnoses and more effective treatments.

Outcome and prognostic features in paediatric pineoblastomas: analysis of cases from the SEER database
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Introduction: Paediatric pineoblastomas are rare central nervous system tumours. Patient and treatment factors associated with outcome are poorly defined and limited to small retrospective case series and single case reports. Using the Surveillance, Epidemiology, and End Results (SEER) cancer registry, we investigated clinicopathological factors associated with outcome in paediatric pineoblastomas. **Methods:** Paediatric patients (<16 years old) with pineoblastomas diagnosed between 1990 and 2007 were identified from the SEER database. Kaplan–Meier survival analysis and Cox models were used to examine the effect of variables on overall survival. The variables analysed included patient's age at diagnosis, gender, race, tumour location, uni-focal or multi-focal tumour, tumour size, surgical resection and the use of adjuvant radiotherapy.

Results: Seventy-eight patients were identified, with a median age at diagnosis of 6.3 years. Sixty-three patients (80.8%) underwent surgery and 39 patients (50%) received adjuvant radiotherapy. Thirty-nine patients (50%) had both surgery and radiotherapy. The median overall survival was 76 months. On univariate analysis, younger age at diagnosis emerged as the only important predictor of overall survival.

Conclusions: This study represents the largest analysis of paediatric pineoblastomas to date. Clinically relevant prognostic factor was younger age of diagnosis. Surgery and adjuvant radiotherapy did

¹⁸F-FET PET-guided tumor specimen acquisition in pretreated pediatric brain tumor patients

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Objective: Magnetic resonance imaging (MRI) might represent constraints in detecting relevant recurrent brain tumor tissue in pretreated pediatric patients. We retrospectively evaluated positron emission tomography (PET) with amino acid tracer ^{18}F -FET (FET), who received further surgical treatment in order to verify the histopathological diagnosis.

Methods: Twelve FET-PET examinations of pediatric pretreated patients with known brain tumor diagnosis and suspected tumor recurrence in MRI were performed (median age 13 years; range, 5–16 years). Lesion-based analysis of PET scans comprised visual appraisal and calculation of standardized uptake values (SUV) defined the hotspot activity within the tumor. Surgery was performed by image-guided specimen acquisition of tissue within the hotspot as predefined region of interest using neuronavigation (VectorVision², BrainLab) after fusing PET images with 3D-MR image datasets (iPlan 3.0, BrainLab). Histological diagnosis served as a reference.

Results: The mean ratio $\text{SUV}_{\text{max}}/\text{SUV}_{\text{brain}}$ for FET uptake in the biopsy target region accounted to 3.6 ± 1.3 (range, 1.6–5.6). The FET-PET-targeted biopsy revealed tumor tissue in 11/12 patients (sensitivity, 92%). Only one patient with a choroid plexus carcinoma showed a false negative result in FET-PET.

Conclusion: The interpretation of MRI findings in pretreated pediatric brain tumor patients can additionally be enhanced by FET-PET, which helps to define the appropriate biopsy targets. Future prospective studies with a larger sample size are warranted to estimate the value of FET-PET in pretreated childhood brain tumor patients.

Radiographic features that predict lack of response to chemotherapy in pediatric intracranial germinomas

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Introduction: Current management of intracranial germinomas in children consists of upfront chemotherapy followed by reduced dosage of radiation therapy where indicated. Residual radiographic disease after the initial sequence of chemotherapy often necessitates “second-look surgery”, to rule-out persistent non-germinomatous elements. The present study aims to evaluate radiographic features present at the time of initial diagnosis which might predict the need for “second-look surgery.” Such high-risk features might lead to alternate management of these patients.

Methods: An institutional review board-approved retrospective review was performed on all patients treated for intracranial germinoma at a major children's hospital over an 8-year period. Patients' charts and all radiographic

studies were reviewed in their entirety. MRIs and CTs were evaluated for tumor location, tumor size, calcifications, cysts, heterogeneity of enhancement, and invasiveness.

Results: Thirty patients with histologically confirmed germinomas were identified. Sixty-seven percent were male. Average time to follow-up was 4.3 years. The average age at diagnosis was 13.3 years. Nine (two females, seven males) required second look surgery within the first year of their diagnosis. At the time of second look surgery, one patient had residual germinoma, two had gliosis, three had teratomatous features, and three had mixed malignant germ cell tumors. Pearson chi-square statistical analysis was performed to compare all germinomas with and without second surgeries. Calcification was associated with a need for second-look surgery ($p=0.034$). Heterogeneity of enhancement correlated with persistent disease but did not reach statistical significance ($p=0.057$). Tumor location, tumor size, tumor invasiveness, and cyst presence did not impact on the need for second-look surgery.

Conclusions: Focal calcifications and possibly heterogeneous enhancement may predict which germinomas will require second-look surgery.

Proteomic analysis and variations of CSF peptides in posterior fossa tumors

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Objective: Cerebrospinal fluid (CSF) protein profile significantly varies between patients with central nervous system (CNS) diseases and healthy subjects, suggesting that CSF may be a source of disease-related biomarkers. Although some biomarkers have been identified by proteomic analysis of CSF, no specific studies on pediatric posterior fossa (PF) tumors have been carried out yet. The goal of this study is to assess the peptides composition of CSF children harboring PF tumors.

Materials and methods: CSF samples taken before and after the surgical excision of 14 tumors (six astrocytomas, five medulloblastomas, and three ependymomas) were analysed by liquid chromatography and mass spectrometry (LC-MS). Control CSF samples were obtained from five children affected by congenital hydrocephalus. The proteomic analysis was performed using a top-down approach where proteins are analysed according to their “natural” form and directly fragmented during mass spectroscopy analysis.

Results: LC-MS profiles of both preoperative and postoperative CSF samples led to the identification of a group of peptides presenting a characteristic behavior

in the different phenotypes if compared with controls. In particular, some small peptides called hemorphins showed a specific trend being found only in the controls and in the postoperative CSF samples but not in the preoperative ones, suggesting a possible role of the tumor in their suppression. The determination of the amino acidic sequences of these peptides allowed us to identify a group of hemorphins playing a specific role in CNS.

Conclusions: The identification of hemorphins may contribute to increase the knowledge on the tumorigenesis and tumor progression of PF tumors in children.

Brainstem gliomas—a precise scoring based surgical outcome: an experience from Northern, India tertiary care centre

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Background: Brainstem glioma occurs in children and is classified based on magnetic resonance imaging (MRI) findings. Surgery has been sought to have limited role. We tried to access precise scoring based surgical outcome in selected subgroups of brainstem glioma patients.

Material and methods: Forty-two patients of histopathologically proven brainstem glioma were evaluated for clinical features at presentation, radiological findings and surgical approach used, Kumar and Samir score (K&SS) was given both pre and postoperatively at 6 week.

Results: Pilocytic astrocytoma was the most common tumor in the operative group. Total K&SS decreased in three patients, was equal in seven patients, and showed improvement in 32 patients. The mean total K&SS from pretreatment level of 84.80 ± 10.4 increased to 88.7 ± 9.2 at 6 weeks follow-up that was statically significant.

Conclusions: The tumors amenable for surgery are usually low grade and present with slow progression of symptoms at a slightly older age. These patients are usually neurologically preserved in comparison to their malignant counterparts. Lesions appearing diffuse on MRI on rare occasions can be benign and all focal lesions may not be benign. Careful selection of cases gives good surgical outcome.

Results of multidisciplinary approach to pediatric intramedullary gliomas

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Introduction: Intramedullary gliomas account for 2–10% of pediatric CNS tumors. We aim to discuss the long-term results of safe multidisciplinary management.

Materials and methods: From 1976 to 2010, we treated 65 children harboring intramedullary tumors, of which 42 has gliomas. Patients age at presentation ranged from 7 months to 11 years, with 60% aged less than 5 years at presentation. While pilocytic astrocytoma was the most common histology (65%); four high-grade cases (three anaplastic astrocytomas, one glioblastoma) and four low-grade disseminated cases were encountered. All patients underwent pre-urgical multidisciplinary evaluation including brain and spine MRI. Follow-up ranged from 1.5 to 16 years (median 5.6 years). Cases were divided into those treated before 1990 (group 1, 17 cases) and after that date (group 2, 48 cases).

Results: Group 1 patients were treated with surgery followed by radiation therapy. In group 2, cases with complete surgical removal were closely monitored; chemotherapy was used if surgery was not radical, if recurrence was detected on follow-up, or in cases with dissemination or anaplastic histology, whereas radiotherapy was reserved to cases progressing under chemotherapy. The amount of complete surgical removals did not significantly change in the two groups (22% and 30%, respectively); however, group 2 patients had significantly reduced permanent neuro-orthopedic deficits (100% and 25%, respectively) and mortality (75% and 6%, respectively).

Conclusion: In pediatric intramedullary gliomas, safe multidisciplinary management is advantageous over conventional surgery-radiotherapy both in terms of long-term neuro-orthopedic outcome, disease control and quality of life.

Immediate post-operative imaging as an alternative to intraoperative MRI in the resection of pediatric tumors, our unique experience

Dhruve Jeevan, Jayson Neil, Avinash Mohan, Hasit Metha (New York Medical College, Valhalla, NY, USA)

The use of intraoperative MRI (ioMRI) to aid tumor resection has become increasingly prevalent. However, very little information has been published regarding the costs and benefits. Currently, a dedicated high-field ioMRI suite is a stand-alone structure with estimated construction costs at \$3 million, which limits availability, and can prolong operative times by up to a third.

Recently, the addition of a 3 T MRI suite, available for constant inpatient/outpatient utilization, was installed opposite to our operating room. This has provided us the unique opportunity to evaluate the cost and benefits of attaining an immediate post-operative MRI under a single anesthesia. Under our new protocol, patients attain a pre-operative 3 T planning MRI, and immediately following resection have repeat 3 T postoperative imaging under the same anesthetic sitting, while the operative room is

maintained sterile for possible return in cases of complication or residual tumor.

A comparison of 25 such cases to age-matched controls prior the institution of this policy. The results demonstrate that similar benefits can be achieved in terms of reducing length of stay, and reducing the need for repeat resections by simply attaining an immediate post-operative MRI (of high resolution), when compared to data emerging for ioMRI.

Our unique experience provides evidence that immediate post-operative imaging provides a similar benefit to ioMRI in the resection of pediatric tumors.

Impact of extent of resection on relapse and survival in primary spinal cord ependymomas: a 25-year review

Roberto Ramirez, Martin English, Guirish Solanki (Birmingham Children's Hospital, Birmingham, UK)

Aim: This study aims to report on the treatment and outcome including relapses in a series of paediatric spinal cord ependymomas carried out at a single institution.

Patients and methods: Between 1985 and 2009, 12 children (seven boys, five girls), with spinal cord ependymoma were identified. Median age at diagnosis was 8.3 (3.5–12.3) years.

Study type: Retrospective cross-sectional study.

Results: Average follow-up was 7.4 (1–19) years. Five children had myxopapillary ependymoma, six World Health Organization (WHO) grade II and one grade III. In one suspected case, the biopsy was negative. This child is being followed-up conservatively. Eleven of 12 (92%) patients are alive. The only death occurred at 4 months post-surgery in an anaplastic ependymoma WHO grade III. Four had gross total resection (GTR) and eight had subtotal resections (STR)/biopsy. None of GTR relapsed. Of the children, 41% (5/12) relapsed at a median interval of 2 (0.25–4.9) years following surgery. Extent of resection and relapses were significantly inversely related ($p < 0.02$), but not with histology type ($p < 0.19$, NS). Mortality (one case) occurred in the single case of malignant histology of the tumour ($p = 0.08$). Of the children, 75% (6/8) with subtotal resection/biopsy underwent primary adjuvant therapy. None of the GTR group received adjuvant therapy. Overall event-free survival (EFS) was 58.3% and overall survival was 92%. Three-year overall EFS was 75% (62.5% for STG), 5-year EFS was 66% (50% for STG). Overall EFS for STR group was 37.5%. 20% of myxopapillary lesions relapsed.

Conclusions:

- Gross total resection was associated with increased survival but was only possible in 33%. Resection rates have improved in the last two decades.
- Myxopapillary type showed a lower relapse rate than previously reported.

Improved GTR rates are keys to preventing relapses.

Posters

CNS Infection

Techniques in prevention of shunt infection

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Abstract: Shunt infections are the most common complication associated ventriculoperitoneal shunts. The average reported shunt infection rates is around 10% ventriculoperitoneal shunt surgery. Most of the shunt infections are believed to occur during the surgery.

Objective: The objective of the paper is to compare the shunt infection rates and the parameters associated with it, in our shunt series.

Material and methods: In our hospital, we have performed 300 shunt surgeries for hydrocephalus over the last 5 years from January 2005 till December 2009.

Results: In our series of patients, there were <1% shunt infections during our follow-up period of a minimum 1 year. The average duration of the surgery from the incision time to closure of wound was 18 min. We follow strict protocol regarding operation theatre traffic, priority for shunt surgery as the first surgery of the day and strict aseptic precautions.

Conclusion: Strict aseptic protocols followed by many neurosurgical centers have reduced the shunt infection rates considerably. In our experience, we found that keeping the surgery time less than 20 min along with routine strict aseptic precautions can reduce the risk of shunt infections further more than what is reported in the literature.

Unusual multiple intracranial hydatid cyst

Prakash Khetan (Jeevan Jyoti Hospital, Allahabad, India)

Multiple hydatid cysts of the brain are uncommon and may be either primary or secondary. A 10-year-old boy with a huge mass of intracranial hydatid cysts is presented. The first manifestation was headache and vomiting, which was followed by symptoms of raised intracranial pressure along with right hemiparesis. The patient underwent an urgent operation due to rapidly deteriorating neurological status, and 296 hydatid cysts were removed. Neuroendoscopic techniques were used to remove intraventricular portion of the cyst. This is the first case in English literature in which such a large number of intracranial hydatid cyst are removed in a single patient. Hydatid cyst is a benign lesion. Surgery is the standard and most effective treatment for intracranial hydatid cysts. Presentation, predictive factors, and management strategies are discussed in the present study.

Intracranial abscesses in children: the New Zealand experience

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Introduction: Intracranial abscesses are associated with high rates of morbidity and mortality. Congenital heart disease is historically implicated as the primary cause. We sought to characterize the incidence and predisposing factors behind the development of intracranial abscesses in New Zealand. **Methods:** Electronic neurosurgical logbooks and the Starship Hospital neurosurgery admission database were queried for pediatric patients with an “abscess,” “subdural,” “epidural,” or “empyema” from 2005 to 2010. Medical records were retrospectively reviewed to identify 40 pediatric patients with a documented intracerebral abscess, subdural or epidural empyema confirmed by magnetic resonance imaging and laboratory studies. Clinical, histological, and radiographic factors contributing to the pathology were documented.

Results: There were 15 intracerebral abscesses (38%; median age, 8.7 years) and 25 subdural/epidural empyemas (62%, median age=1.7 years, $p=0.01$). The majority (45%) were associated with mastoiditis (13%), sinusitis (30%), or orbital cellulitis (3%). With contiguous infections, eradication of the primary site of infection (sinus/mastoid) proved essential (89%) with reoperation rates of 80%. The infectious agent was primarily streptococci species (*Streptococcus anginosus* species or *Streptococcus pyogenes*, 44%). Up to 10% of the infections may have been avoided with scheduled immunizations. There were no deaths; developmental delay and/or seizures affected 38%.

Conclusions: New Zealand has a disproportionately high rate of pediatric intracranial abscesses associated with contiguous spread from sinusitis or mastoiditis. Successful management requires a multidisciplinary approach with otorhinolaryngology and infectious disease. Many of these infections (and associated morbidity) may be preventable with early detection and treatment of sinusitis and otitis media, along with adherence to vaccination schedules.

Hydrocephalus

Severe fetal asymmetrical ventriculomegaly: challenge for prenatal counseling regarding prognosis and treatment options

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Background/aims: Prenatal counseling for severe asymmetrical ventriculomegaly (SAVM) is a true challenge since the

prognosis of *isolated SAVM* is actually unknown. The aim of this paper is to describe the clinical outcome of fetuses presenting with SAVM and present our results with a shuntless endoscopic procedure in selected cases.

Methods: Five fetuses with fetal SAVM were followed after birth for a period of 12–84 months. None had associated malformations and no clear underlying pathology (by magnetic resonance) was diagnosed prenatally. All were born by elective Caesarean section in good clinical condition, without neurological deficit at birth. One had an abnormal EEG on the side of dilated ventricle. Three were operated (ages 1 day–4 weeks) for endoscopic septumpellucidotomy with or without opening of adhesions at the obstructed Monro that was documented intraoperatively. There were no intra/perioperative complications and hydrocephalus was well controlled, with normal motor and cognitive development in two, and mild developmental delay with controlled epilepsy in one baby (follow up 30–84 months). Two infants are conservatively followed with normal development and no signs of elevated ICP (follow up, 12 and 24 months each).

Conclusion: In cases without chromosomal, infectious or hemorrhagic risk factors, good outcome is possible despite severe dilatation of one lateral ventricle. Thus, severe asymmetrical VM has probably a better prognosis compared to severe bilateral VM as the pathogenesis may be different. We also suggest that prenatal counseling of isolated SAVM should include discussion about possible need for surgery, including the option of an endoscopic shuntless procedure.

Endoscopic third ventriculostomy for the treatment of osteopetrosis-related hydrocephalus: a case-based update

Boresh Dhamija, Benedetta Pettorini, Guirish Solanki (Birmingham Children's Hospital, Birmingham, UK)

Background: Osteopetrosis is a heterogenous group of disorders characterised by a failure of normal bone maturation, abnormal bone sclerosis secondary to the failure of osteoclasts to resorb bone. The most serious consequences of this disorder affect the nervous system. Patients with infantile osteopetrosis (also called malignant osteopetrosis) can develop a gradual occlusion of, or narrowing of the skull foramina at the skull base, resulting in the compression of vital nerves and vessels. Hydrocephalus has been identified in these patients, particularly those with the autosomal recessive variety of osteopetrosis. Although the exact aetiology is uncertain, it is possible that venous outflow obstruction at the cranial foramina along with a reduced intracranial space for CSF to flow around the hemispheres could be contributing factors. There are few reports in the literature on the management

of this unusual association, hydrocephalus secondary to osteopetrosis. The authors report one such case where this association has been successfully surgically treated with endoscopic third ventriculostomy as a form of cerebrospinal fluid (CSF) diversion.

Case report: We treated a 9-month-old girl with osteopetrosis and symptomatic hydrocephalus successfully treated with an endoscopic third ventriculostomy (ETV). She later went on to have stem cell transplantation to treat the osteopetrosis.

Conclusions: Most reports in the literature have identified ventriculoperitoneal (or other distal site) shunting as the treatment of choice for hydrocephalus in this setting. We would like to highlight that ETV is another effective and often very suitable method of CSF diversion in these patients.

Treatment of very low pressure hydrocephalus

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Introduction: Occasionally one encounters the situation where a patient with a normally functioning cerebrospinal fluid (CSF) diverting shunt develops clinical symptoms of shunt malfunction and progressive ventricular enlargement in the face of persistent low or even zero intraventricular pressure (IVP). How does one manage this condition?

Methods: With IRB approval, a retrospective review covering the past 10 years at our institution was undertaken to identify those patients who met the above criteria.

Results: Five patients ranging in age from 20 months to 17 years were found. Four patients had posterior fossa tumors and the fifth a cavernous malformation of the third ventricle. All five required CSF diversion following excision of their lesion. The initial treatment was to tap the shunt one or more times a day with removal of increasing volumes of CSF. When that failed an external ventricular drain (EVD) was placed with removal of CSF at a negative IVP up to -20 cm of water. Based upon clinical symptoms and interval imaging for ventricular size, the EVD was gradually raised until the outlet pressure was slightly positive (+5 cm of water or even less). At this point, a ventriculopleural or ventriculoperitoneal shunt without a valve was inserted. The entire process extended for weeks or even months but was successful in all five cases.

Conclusions: Treatment of very low pressure hydrocephalus can be very difficult but eventually successful over a protracted period of time. Why the compliance of the parenchyma is temporarily altered is unknown.

Neuroendoscopy in the treatment of hydrocephalus in Children. Our early experience

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Introduction: Through the years, brain endoscopy has been useful in the management of obstructive hydrocephalus. Still in pediatric population under 1 year of age, the value of the third endoscopic ventriculostomy is controversial; it is believed to cause more morbidity and to have higher failure rates.

Patients and methods: Eighteen endoscopic procedures of rigid neuroendoscopy in the same number of pediatric patients (3 months to 11 years) with hydrocephalus of diverse etiology were treated in a 17-month period at the Hospital Regional de Alta Especialidad del Bajío in Mexico.

Results: Fifteen endoscopic third ventriculostomy procedures (ETV) were done, three biopsies of tumors occupying the posterior part of the third ventricle in only one patient, endoscopic navigation and placement of an external ventricular drain was done due to ventriculitis. ETV was successful in 33% of the children less than 1 year of age.

Conclusions: A careful patient selection and preoperative planning lead to better results of ETV, and should be considered as initial treatment in those cases with obstructive hydrocephalus, carries low morbidity with an experienced team of neurosurgeons, and represents a useful tool for obtaining biopsy samples.

Conceptual differences between valves for the treatment of hydrocephalus based on theoretical considerations and in vitro findings

Christoph Miethke (GmbH & Co.KG, Potsdam, Brandenburg, Germany)

Clinical studies on the in vivo performance of shunts for the treatment of hydrocephalus did not show significant differences between the available systems until recently. With the release of results from the so-called SWASONA study, highly significant differences are submitted for the first time: by the use of gravitational components, the risk of overdrainage in patients with normal pressure hydrocephalus can drastically be reduced.

Gravitational components belong to the group of hydrostatic valves, which claim to control overdrainage in the upright position of patients. Three different types are used clinically: anti-siphon devices, flow-reducing devices, and gravitational components. In vitro investigations demonstrate convincingly the significantly different performance of the different types. The reduction of flow introduces the risk of systematical under- or overdrainage. The concept of anti-siphon devices solves technically the problem of

overdrainage; however, this is counterbalanced by the unavoidably unpredictable dependency on subcutaneous pressure of these devices. Gravitational components introduce a posture depending beneficial increase of the opening pressure of the shunt systems, but the sufficient opening pressure cannot reliably predicted.

The conceptional analysis of the systems and the in vitro findings make two future tasks for the further improvement of shunt technology obvious: the adjustability of hydrostatic components and the noninvasive measurement of the intracranial pressure as a support for a reasonable adjustment.

Counterbalance of the hydrostatic pressure by an adjustable gravitational valve

Christoph Miethke (GmbH & Co. KG, Potsdam, Brandenburg, Germany)

The benefit of additional gravitational components for patients with normal pressure hydrocephalus (NPH) has been demonstrated in a prospective randomised clinical study recently (SVASONA). In addition to that, the adjustability of valves is seen to be superior to non-adjustable concepts due to the option to optimise the treatment for the individual patient. Up to now, no adjustable valve has been available which focuses on the upright position of the patient without interfering with the lying position. If a gravitational component improves significantly the complication rate in patients with NPH, the requirement of individual adjustment of the gravitational component becomes obvious.

The development of the proSA solves the problem of posture independent individual adjustment of the opening pressure. Gravitational valves are characterised by posture depending opening pressures. Whereas in the lying position, the opening pressure should be low this pressure has to be significantly higher in the upright position of the patient. Clinical problems during night and day might be related to insufficient pressure adjustments for one or the other position. The proSA introduces the option to increase or decrease the opening pressure just for the upright position by adjusting a spring force which is acting against a tantalum weight. The adjustment of the opening pressure does not change the setting for the lying position. In combination with a conventional adjustable valve, all adjustment options are given based on the individual patient

Neuroendoscopic laser-assisted choroid plexus coagulation: safety and feasibility

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Objective: We report our preliminary experience using neuroendoscopic laser-assisted choroid plexus coagulation (CPC) for treating severe forms of congenital hydrocephalus, focusing on its safety and feasibility.

Methods: We performed neuroendoscopic laser-assisted CPC in five children with severe forms of hydrocephalus (one hydranencephaly, three holoprosencephaly, and one extreme hydrocephalus), admitted to the Department of Neurosurgery of the Santobono Children Hospital in Naples, Italy.

Results: All patients underwent neuroendoscopic laser-assisted coagulation of choroid plexus. In two patients, coagulation of choroid plexus was performed after removing infected shunts. In one patient with extreme hydrocephalus and in one patient with semilobar holoprosencephaly, we performed also endoscopic third ventriculostomy. There were no surgery related complications. After a mean follow-up period of 10 months (range, 4–18 months), only two patients remained shunt free. One patient with holoprosencephaly did not require further hydrocephalus related procedures but died 5 months following CPC for pneumonia. The two patients with previously infected shunts required further surgery for reimplantation of ventriculoperitoneal shunts 30 and 45 days following CPC.

Conclusion: Neuroendoscopic laser-assisted coagulation of the choroid plexus could be a safe and feasible technique. Limited indications for this procedure may include desperate hydrocephalic condition such as holoprosencephaly or hydranencephaly.

Application of the Miethke ProGAV valve for children under 1 year old

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The purpose of the study is to answer whether applying Miethke ProGAV system in children under 1 year old is possible. We retrospectively reviewed 19 infants with hydrocephalus treated with Miethke ProGAV shunt system. In all cases, shunt catheter between burr hole and the reservoir–antigravitation unit complex was placed in S-shape to avoid displacement of the antigravitation unit during growth of the skull. Average age at the time of surgery was 2.6 months and mean observation period was 17.6 months. Average head circumference at the time of the surgery was 41.3 cm. Increase in head circumference during observation period varied from –2.6 to 11.2 cm (mean, 5.5 cm). Three patients had shunt obstruction and one patient experienced shunt infection, which required shunt revision. One-year shunt survival rate obtained from Kaplan–Meier analysis was 0.81. S-curve of the catheter became gentle after increase of the head circumference, while antigravitation unit kept upright position. Our early

experience indicate that placement of Miethke ProGAV shunt system is feasible for children under 1 year old.

Role of endoscopic third ventriculostomy in infants

B.K. Ojha, Anil Chandra, Sunil Singh, Chhitij Srivastava (CSM Medical University (KGMC), Lucknow, UP, India)

Introduction: The value of endoscopic third ventriculostomy (ETV) in infants less than 1 year of age is controversial. It is believed to cause more morbidity and to have higher failure rates. The aim of this study is to define the effectiveness of ETV for hydrocephalus in infants and to determine possible positive predictive factors.

Materials and methods: Forty-eight patients of hydrocephalus, who underwent ETV, were evaluated. Failure of procedure was defined as any subsequent surgical procedure for cerebrospinal fluid diversion or death related to hydrocephalus management in 6-month period.

Results: All failures occurred in the early postoperative period. The success rate varied with the etiology of the patient's hydrocephalus. Etiologies that were identified were idiopathic aqueductal stenosis, suprasellar arachnoid cysts, post-hemorrhagic hydrocephalus, post-infectious hydrocephalus, tumor-related hydrocephalus, and other developmental anomalies. Low birth weight and premature infants had higher failure rate compared to full-term infants with normal birth weight. The success rate of ETV increased with age.

Conclusion: Our results suggest that the selective use of ETV as the primary treatment in infants with hydrocephalus is safe and can lead to a reduction in the shunt-related complication.

MRI Morphometric features in patients undergoing endoscopic third ventriculostomy: quantitative and qualitative analysis

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Introduction: In the management of paediatric hydrocephalus, currently there is no reliable pre-operative radiological predictor for success of endoscopic third ventriculostomy (ETV), nor is there an early post-operative indicator to predict the likely failure.

Aims and objectives: This study aims to analyse conventional and new MRI parameters that reliably predict the immediate post-operative success of ETV and helps differentiate between successful and failed ETV cases.

Patients and methods: MRI imaging of 33 (21 boys and 12 girls) consecutive children with ETV were reviewed; M/F=1.75. The median age was 5.5 (15 days–15) years. Pre- and

post-operative MRIs were reviewed. T2W and FIESTA sagittal and axial sequences were used. Known and new parameters as well as qualitative features that help define ETV success were measured and are reported here.

Results: There are several immediate post-operative radiological indicators for success of ETV and these include a reduction of third ventricular width ($p=0.004$), third ventricular index ($p=0.007$), FOHR ($p=0.045$), MBLT ($p=0.017$), AC-TVF ($p=0.017$), convexities of lamina terminalis and third ventricular floor ($p=0.02$), significantly associated with clinical success. There was no reliable pre-operative radiological predictor of ETV success rate in this series. There was no statistically significant difference in any of these parameters when comparing successful ETV cases versus failed cases.

Conclusion: While no pre-operative radiological predictor of ETV success was found, this study has identified both quantitative and qualitative features which are of important prognostic value, although they have been found wanting in terms of differentiating success from failure of ETV.

Neurodevelopmental outcome in children with hydrocephalus—beyond a successful surgery

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It is not uncommon for children with hydrocephalus to have delayed developmental milestones and some degree of learning difficulty. Those most often associated with hydrocephalus are delayed social, language, motor development, nonverbal learning disorder, and difficulties in understanding complex and abstract concepts. Though surgically, the procedure required for hydrocephalus may be considered successful but these children may continue to have varying psychomotor deficits

This was a prospective study carried in 50 children less than 1 year of age. All the children underwent surgical treatment for hydrocephalus at our hospital between the period March 2008 and March 2010 and are under regular follow-up. All children are up to minimum follow up until the age of 2 years. Trivandrum developmental screening chart was used in this study and Fisher's exact test is a statistical test used in analysis of this study. Thirty-four patients underwent successful endoscopic third ventriculostomy and 16 patients underwent successful VP shunt. Out of 50 children, seven (14%) had normal milestones at presentation and were normal postoperatively also. In the remaining 43 children with delayed milestones, 27 (62.8%) improved; and in 16 (37.2%) children, delayed milestones persisted. Developmental outcome depends on etiology, duration of disease before treatment, associated central nervous system and other

anomalies, time of diagnosis, and treatment. Outcome was independent of type of procedure done (VP shunt/ETV).

Endoscopic third ventriculostomy for hydrocephalus in infants—improving surgical outcomes

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Endoscopic third ventriculostomy is considered a safe procedure and is now become a choice in treatment of obstructive hydrocephalus with high success rates. In infants, some authors have reported a lower effectiveness of endoscopic third ventriculostomy alone due to various reasons

At our institute, we try to evolve approaches and nuances in this procedure for bettering the outcome and success of the surgery. Simple steps like the type of incision to placement of the burrhole have also evolved. Proper perforation of third ventricular floor, avoiding use of cautery, and aqueductoplasty/exploration also contributes to success of the procedure. Cerebrospinal fluid (CSF) reservoir placement in the same burr hole has significantly contributed to the improving outcome by preventing a CSF leak and taking care of early raised pressure symptoms.

Results of 104 cases operated between July 2005 and July 2010 at our hospital are discussed. All cases were infants with hydrocephalus due to various etiologies; majority being aqueductal stenosis. Surgical success—patients do not require a second procedure and there is objective/subjective improvement. Overall success rate in our series of mixed etiology was 78% with success rate reaching up to 90% in cases of aqueductal stenosis. There were no deaths related to the procedure. Of the patients, 26% patients underwent initial CSF tapping from reservoir and there was no complication related to the Insertion.

Outcome was predominantly dependent on etiology rather than age at presentation. Adding flexible neuroendoscope to the armamentarium also has helped in taking care of multiseptate hydrocephalus, doing third ventriculostomy at alternate site (trans lamina terminalis) and exploring the fourth ventricle.

The role of intracranial pressure monitoring in the investigation of suspected shunt malfunction in children

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Intracranial pressure (ICP) monitoring can aid evaluation of shunt function when the diagnosis is uncertain clinically and radiologically. There is only one reported study of ICP monitoring in the investigation of suspected shunt malfunction in children.

Twenty-three children with an uncertain diagnosis of shunt malfunction underwent 25 episodes of ICP monitoring. Indication was suspected blockage in three cases, under-drainage in 15, and overdrainage in seven.

All three suspected blocked shunts had high pressures and were indeed blocked. Of 15 cases of suspected under-drainage, this was confirmed in three, although one had emergency revision for clinical deterioration rather than raised ICP. Eight had normal pressures. Four were overdraining. ICP monitoring confirmed overdrainage in six suspected cases. One had normal pressures, making overdrainage unlikely.

Nine children underwent shunt revision—eight for blockage, one to change to a different make of valve (shunt functioning, overdraining). Seven had valve pressure adjusted. Six had symptomatic treatment.

ICP monitoring confirmed clinical suspicion in 12/25 and changed the diagnosis in 13. One patient with suspected underdrainage had normal pressures but ultimately required shunt revision for blockage. ICP monitoring may have delayed surgery, or shunt may have blocked after monitoring period.

There were no complications.

ICP monitoring made a positive contribution in 23 cases, but may have delayed treatment in one. For the final patient, treatment decisions were made clinically, irrespective of ICP.

ICP monitoring can add considerable information when investigating suspected shunt malfunction. However, it is an invasive test so its use should be considered carefully.

Low-pressure hydrocephalus in a child: case report and literature review

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Low pressure hydrocephalus is a very rare event in children. Few literature cases were reported, since the first descriptions by D. Pang and H. Rekaté in the 1990s, but its occurrence should be remembered because it has a quite dismal outcome and its cure is long and complex. UE, a 2-year-old girl was treated by third ventriculostomy for hydrocephalus due to low-grade thalamic tumor; after 1 year, the lesion evolved and it was removed by a posterior approach in the sitting position; then there was a slow reappraisal of consciousness, necessitating only a mild right hemiparesis and oculomotor deficit; in a month's time, a

progressive ventricular dilatation occurred, treated by a programmable shunt, deserving progressive lowering of the output pressure, down to 30 mmH₂O; a progressive dilatation of the ventricles occurred, surrounded by broad white matter hypodensity, consciousness impairment until coma. External drainage was placed, revealing a quite low CSF pressure –20 mmH₂O, obtaining consciousness improvement and ventricle reduction; no wonder that when a flow control shunt was applied, the symptoms recurred. The definitive treatment was obtained by prolonged low pressure external drainage until ventricles shrinkage; this treatment obtained the improvement of cerebral compliance, with disappearance of periventricular radiolucency and normal ventricles size; only than the shunt was reconnected, and the girl slowly improved, residuating only right hemiparesis and oculomotor deficit. The low-pressure hydrocephalus may be misinterpreted as a parenchymal damage with no chance of recovery; the pathophysiology is correlated to excessive brain compliance leading to an increase of white matter permeability for the fluid; prolonging external drainage takes to recover, allowing application of normal valves.

Slit ventricle syndrome—is headache the clinical indicator for intervention?

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Text: Slit ventricle syndrome is a difficult and challenging entity. It is common to encounter children, who had undergone previous shunts, to present with acute, intermittent headaches. The shunt may be clinically working and normal sized ventricles on CT/MRI tempt the clinician to wait and watch. However, the underlying pathologies like intermittent catheter obstruction, normal volume hydrocephalus, or cephalocranial hypertension makes the child suffering and increases the risk of sudden decompensation. In settings where intracranial pressure monitoring is not available, meticulous clinical observations like timing, nature, and severity of headache are the sole armamentarium for the clinician to call for intervention. We present our series of five cases of slit ventricle syndrome, encountered in the last year, intervening on the basis of clinical judgment alone, which was later proven to be correct.

Advantages and drawbacks of valved shunts—a learning curve of 37 years

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Objective: Sixty years ago, the introduction of shunts with valves revolutionized the therapy of hydrocephalus. After

the first enthusiasm, a nightmare of different complications increasingly ruined the results of this new method. But various improvements in surgical techniques and by shunt technology lead to a decrease in patient morbidity and an increase of shunt survival. This will be illustrated by the gradual learning curve of the author

Methods: The advancements achieved over the years can be differentiated in those of "hardware" like improvements of valves, anti-syphon units, reservoirs and catheters, or miniaturized devices for in vivo pressure measurement on the one side and "software" increase of understanding regarding pathophysiology, optimal pressure, levels and avoidance of complications on the other. We tried to qualify and quantify the increase of knowledge in the most important of these parameters.

Results: Whereas operative advancements over the years were marginal and the increase of insight concerning unequivocal diagnosis, pathophysiology, and rational of shunt therapy of normal pressure hydrocephalus is limited, the avoidance of severe complications by improved adjustable hydrostatic valves and other technological progress is evident. The progress and avoidable errors will be illustrated by evidenced scientific results and by personal experience over the last 35 years.

Conclusion: Despite all critics of the industry, the improvement of hardware and the gain in understanding of valve function and pathophysiology of different forms of hydrocephalus lead to a significant better outcome in the therapy of hydrocephalus. But further efforts are essential necessitating cooperation between medical staff on the one hand and biomedical industry on the other.

Endoscopic third ventriculostomy in pediatric tubercular meningitis (TBM) hydrocephalus

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Introduction: The traditional shunt procedure provides effective treatment for tubercular meningitis (TBM) hydrocephalus but it has a number of limitations. The aim of this study was to assess the feasibility of endoscopic third ventriculostomy (ETV) in TBM hydrocephalus.

Material and methods: Twenty-five patients of TBM hydrocephalus treated with ETV were evaluated.

Clinical outcome were recorded as improved, unchanged, or deteriorated.

Failure of procedure was defined as any subsequent surgical procedure for CSF diversion or death related to hydrocephalus management in a 6-month period.

Results: The outcome of ETV showed a significant correlation with the presence of basal exudates on CT scan and intraventricular third ventricular floor anatomy.

Conclusion: ETV should be considered as the first surgical option for CSF diversion in patients with TBM hydrocephalus

External shunt should be reserved for those who fail with endoscopic CSF diversion.

Multiple shunt failures: an analysis of relevant factors

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We made a structural analysis of a group of patients shunted for infantile hydrocephalus with three and more revisions (called patients with multiple revisions) during the time of the existing shunt. We searched for possible relevant factors for the larger number of revisions.

We conducted a retrospective study of the shunt history of 242 children with infantile hydrocephalus (homogenous group) with 375 revisions. The follow-up was 5–25 years. The patients with multiple revisions are $n=53$ (21%) and have had 243 revisions (65%) of the total number of revisions. Of all the shunts, 151 of 242 were revised.

Summary: The large number of revision—243 (65% of all revisions) is due to the relatively small group of patients with multiple revisions (22% of all cases). The average number of revisions of one shunt for the entire series is 2.48 revisions, while for the patients with multiple revisions it is 4.58 revisions. The age at implantation, as well as the type of shunt operation (VPA or VAA) is not a factor that increases the percentage of patients with multiple revisions. A period from the implantation to the first revision that is shorter than 6 months correlates with a significantly larger percentage of patients with multiple revisions (47%) and with a low percentage (22%) of patients with one or two revisions. In the group of patients with multiple revisions, the larger percentage falls on the malfunction of the ventricular catheter, followed by a malfunction of the cardiac catheter as a first complication.

A new guide to simply apply accuracy in ventricular catheter placement for CSF shunts

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Objective: Neuronavigation has been introduced to overcome inaccurate freehand placement of cerebrospinal fluid catheters in narrow ventricles. We measured the insertion angles for ventricular catheters and developed an application tool to enhance accuracy with simple technical needs. **Methods:** 3D MR data sets of 21 children (9.5 ± 4.6 years) were used to virtually identify the ideal entry

point (EP) for ventricular catheters by defining the trajectory being perpendicular to the sagittal convexity targeting the foramen of Monro. The angle of freedom to target the ventricle relative to the median plane as well as the tangential angles in coronal direction was measured. Accordingly, a catheter guide was developed to apply these measurements in the implantation procedure.

Results: The ideal EP was located at 119.1 ± 6.4 mm from Nasion and 17.5 ± 0.6 mm lateral from midline. Lateral ventricle width was 7.6 ± 2.9 mm. While angles relative to the median plane showed high variation measurements, the tangential angle showed high similarity at ideal EP ($92 \pm 2.8^\circ$ to $99.6 \pm 4.1^\circ$) compared to EPs 5 mm lateral ($92.2 \pm 2.2^\circ$ to $99.9 \pm 3.6^\circ$) and 5 mm medial ($91.2 \pm 2.8^\circ$ to $98.4 \pm 3.6^\circ$), respectively. Consequently, the developed guiding tool uses the tangential angle as relevant parameter for accurate placement of ventricular catheters, which could successfully be proven in 12 consecutive patients.

Conclusion: The tangential angle in coronal direction is a robust parameter in terms of guiding the implantation of the ventricular catheter. Thereby, a simple tool can easily be used to apply these measurements to enhance quality in shunt procedures with limited technical effort.

Diverse arachnoid cyst morphology indicates different pathophysiological origins

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Background: Arachnoid cysts (AC) are fluid-filled malformations of arachnoid tissue found throughout the cranial–spinal axis. The origin of ACs as well as the mechanism for their expansion ultimately leading to neurological symptoms is unknown. Further, the morphology of the cyst wall has only been studied in a few cases. We aimed to study the variability of the cyst wall morphology to better understand the characteristics of the cysts.

Methods and material: Twenty-four patients (12 males, 12 females) with symptomatic AC were included. Mean age was 36.3 years (range, 10–79 years). Locations were cysts were temporal (14), posterior fossa (5), frontal (4), and one in the interpeduncular cistern. Before fenestration, the patients underwent clinical examination and an MRI. Samples from the cyst membrane were sent for routine neuropathological diagnostics, transmission electron microscopy (TEM) and scanning electron microscopy. As reference, arachnoid tissue was collected from five patients operated for Chiari type I.

Results: All patients improved 3 months after fenestration. All samples were diagnosed as typical arachnoid tissues at the pathological examinations. The AC membranes were found to be heterogeneous at TEM. Three different types each with similar morphological features could be identified. Twelve cysts type A demonstrated a structure typical for normal arachnoid matter. Type B (4 cysts) consisted of thick connective tissue layer. Type C (eight cysts) had a differentiated epithelium with one to three cell layers lining the cyst lumen.

Conclusions: Tissue from AC membranes shows a diverse morphology indicating different cystogenesis mechanisms.

“The Noodle sign”: a case of shunt migration in an infant.

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Background and importance: Ventriculoperitoneal shunts (VPS) are an established mechanism for cerebrospinal fluid (CSF) diversion in patients presenting with hydrocephalus. Mechanical malfunction of a VPS is a common occurrence and usually easily remedied. Patients with cerebral palsy and severe spasticity with opisthotonos may present with unusual shunt migration.

Clinical presentation: We present the case of an 11-month-old female with cerebral palsy and VPS insertion due to hydrocephalus that experienced a 2-day history of nausea, vomiting, and a bulging left occipital area overlying the proximal catheter insertion site. Imaging and intraoperative findings revealed extruded proximal and distal catheters into the subgaleal space and broken shunt valve retention sutures. Intraoperatively, the patient received a complete revision of the shunt, including anchoring of the shunt valve to the periosteal layer and securing of the burr hole with a subgaleal flap.

Conclusion: Mechanical failure of a VPS in this patient was due to a number of factors, including excessive arching, previous shunt revisions, fragile subcutaneous tissue, and absorbable retention sutures. Shunt revision involved manipulation of placement to circumvent all these factors.

Advantages of the new adjustable gravitational unit ProSA in pediatric hydrocephalus therapy

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Objective: Overdrainage and slit ventricle syndrome is a well-known complication in pediatric hydrocephalus. There was no adjustable gravitational unit available so far and there exist no experience for the optimal valve setting.

Methods: Forty-seven children with first diagnosis of hydrocephalus, arrested hydrocephalus, slit-ventricle syndrome or overdrainage (age range, 3–18 years old) underwent implantation of a new adjustable shunt assistant (ProSA, Aesculap Miethke, Tuttlingen/Potsdam, Germany) from May 2009 to December 2010. In 15 cases, the proSA was combined with an adjustable differential pressure valve (proGAV). In five cases, both shunt valves were adjusted under long-time telemetric ICP home monitoring.

Results: There were no infective or other operative complications as yet. In 32 cases, the valve pressure had to be adjusted after surgery, in 19 cases more than once. Forty-three children are symptom free, four children showed reduced cephalgia under ongoing fine adjustment. Follow-up MRI-scans showed slightly wider or normalized ventricles. The ICP home monitoring revealed first results about the effect of this new valve and was very helpful for the optimal fine adjustment.

Conclusion: The new adjustable shunt assistant is highly effective to avoid overdrainage and slit ventricle syndrome. Reoperations can be avoided and a normalisation of slit ventricles can be achieved stepwise. ICP home monitoring revealed first results of the working mechanism of the new gravitational unit under daily life conditions. The ICP monitoring helps to understand the optimal valve adjustment. The combination of an adjustable differential pressure valve and an adjustable gravitational unit seems to be the optimum for pediatric patients.

Shunt complications

Keki Turel (Bombay Hospital, Mumbai, India)

Ventricular shunt placement is one of the commonest neurosurgical procedure, yet most frequently associated with complications. Since the early days of shunt insertion, shunt failure has been considered a serious problem. There are many possible causes for shunt malfunction. These include over- and underdrainage, mechanical mismatch, occlusion, valve failure, growth effects in children, infection, catheter migration, and other less common complications.

Shunt infection remains the foremost problem of shunt implantation after mechanical malfunctions. Diversionary cerebrospinal fluid shunt implantation has a high complication rate, with 5–15% of such shunts becoming infected. Of these infections, 70% are diagnosed within 1 month after surgery and more than 90% within 6 months! Shunt infection in the vast majority of cases is therefore a complication of shunt surgery.

Attempts to address these problems include finding alternative drainage locations, ventricular catheter placements, and new valve designs. The latter include a variety of drainage pressures, antisiphon devices, programmable and flow-regulated valves, and catheter and shunt assembly designs. Nearly 400 types of shunt devices have entered the market. This shows that a perfect device is yet to be made or shunt complications may have little to do with advances in design. However, we have made progress in certain aspects of shunt function. For example, the proportion of shunt failures attributable to infection has probably decreased over the years.

We have studied shunt complications seen in our own operated patients and those treated elsewhere, and this analysis will be elaborated to address the issue of shunt complications.

Review of utility of ventriculosubgaleal shunt—an institutional experience

Suhas Udayakumaran, Ashok.B Pillai, Rajiv Kariyattil, Dilip Panikar (Division of Pediatric Neurosurgery, Department of Neurosurgery, Amrita Institute of Medical Sciences and Research Centre, Kochi, Kerala, India)

Introduction: Temporary methods of diversion of cerebrospinal fluid like ventriculosubgaleal shunt (VSG) is often required in patients who are not ideal candidates for a permanent method of cerebrospinal fluid (CSF) diversion.

Materials and methods: We retrospectively analyzed the data of 38 children for whom ventriculosubgaleal shunts were done for various indications between January 2003 to December 2010 at the Division of Pediatric Neurosurgery, AIMS, Kochi. The collected data viz. sex, age at insertion of VSG, aetiology of hydrocephalus, indication for VSG, complications associated with VSG, and the fate and life span of VSG.

The end point of the study was a permanent mode of diversion or nonrequirement of VSG.

Results: Our series comprised of 38 children. The ages ranged from 5 days to 7 years. Eighteen children (47.3%) were born premature. The commonest aetiology of hydrocephalus was prematurity-related posthaemorrhagic hydrocephalus (PHH) in 16 (42.10%), 14 meningitis (36.8%), 10 congenital aetiology, and six post-infective aetiology. Indications for VSG were PHH 16 children, active meningitis 14 children, five for shunt infection, ten VSG for post infective hydrocephalus.

The complications of VSG are three malfunctions, two revisions, three CSF leaks, one infection, and one had shunt tip slipping into the ventricular cavity. Thirty-four of the patients had conversion to ventriculoperitoneal shunt, two ETVs, and one have no shunt.

Conclusion: VSG shunts are a simple and efficacious temporizing measure for CSF diversion in younger children who are not ideal for a permanent method of CSF diversion even in presence of active meningitis and shunt infection.

A case of ‘compensated’ trapped fourth ventricle—long-term clinicoradiological follow up

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Background: A trapped fourth ventricle (TFV) is diagnosed when the inlet and outlets of the fourth ventricle is obstructed in association with a dilated fourth ventricle.

We postulate the concept of a “compensated” TFV with a long-term clinical and radiological follow up.

Case report: A 3-month-old male child had a ventriculoperitoneal shunt for post infectious hydrocephalus. At the age of 9 years, he had a ventriculogram when he became symptomatic with a large fourth ventricle. His ventriculogram revealed an aqueductal obstruction in association with an enlarged fourth ventricle. Although diagnosed as a TFV radiologically, his lateral ventricle shunt was revised with a diagnosis of shunt malfunction as there was a simultaneous dilatation of the lateral ventricle. He was never treated for the TFV, as he did not show any symptoms suggestive of compression at the level of fourth ventricle.

At follow-up at 24 years of age, his MRI brain showed well-decompressed lateral ventricles and a large fourth ventricle. On clinical examination, he had dysmetria of the limbs which according to the mother “was always there”. His MR flow study showed obstruction at the level of aqueduct with bidirectional flow at foramen of Magendie.

Discussion: The concept of compensated hydrocephalus has been well described. We demonstrate the concept of compensated TFV to explain the asymptomatic clinical situation in a radiologically proven TFV.

Conclusion: We demonstrate and describe a compensated TFV and reiterate that a TFV is functional concept with imaging being at the most only corroboratory.

Treatment of fetal and neonatal hydrocephalus and arachnoidal cysts by programmable shunts: pros and cons

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Introduction: The prognosis of fetal and neonatal hydrocephalus has been reported to be dismal and correlated with

the cause, being worse for posthemorrhagic and infective hydrocephalus than for malformations. Arachnoidal cysts of the same period seem to have a better prognosis, but still unpredictable. Fetal and neonatal cerebrospinal fluid (CSF) flow disturbances may cause postshunting synostosis due to overdrainage and consequent CMI.

Patients and clinical picture: Thirty children were shunted at the FINCB for hydrocephalus or arachnoidal cysts occurring between third trimester of pregnancy and the first 3 months after birth. The cysts were located in the posterior fossa (three), in the ventricles (four), fronto-temporal (two), and parasagittal region (two). All were submitted to programmable shunting for occurrence of symptoms or increase of CSF collection. Afterwards, all were followed at least 1 year with neuropsychological testing and RM

Results: There was no mortality and no post-shunting craniosynostosis; six were reoperated for malfunction. The outcome was favourable in all except two, one infection and one contralateral bleeding. All needed at least two or more pressure adjustment before 18 months.

Conclusions: The precocious treatment of CSF circulation disturbances and accurate adjustments of the programmable shunt is correlated with an early parenchymal re-expansion and favourable neurological outcome. A low pressure (70–90) was used until there a good re-expansion of parenchyma was observed; around 1 year of age, a progressive increasing of the output pressure was requested to avoid slit ventricles and excluded IV ventricle. In front of these unquestionable advantages, the main problems of the programmable shunts are represented by the need of multiple neuroradiological examinations.

An analysis of CSF shunting procedure requirement in children with posterior fossa tumors

Gopalakrishnan CV, Suresh Nair, Girish Menon, Amit Dhakoji

Objective: Management of hydrocephalus (HC) in children with posterior fossa tumors is controversial. In the present study, we analyse the factors that predispose to persistent HC and the need for a postoperative cerebrospinal fluid (CSF) diversion procedure.

Materials and methods: Children who underwent surgery for posterior fossa tumors with HC were retrospectively analysed. Patients having undergone CSF diversion preoperatively were excluded from the study. Eighty-four patients were included and the need for shunts after surgery was noted. The factors evaluated included age at diagnosis, symptomatology and duration, tumor type and location, ventricular size indices, extent of surgical resection, need and duration of external ventricular drain (EVD) in the post-operative period and postoperative complications that could be related to CSF circulation disorders.

Results: Eighty-one of 84 (96.4%) patients had symptomatic hydrocephalus. Of the patients, 29.7% required a CSF diversion procedure in the postoperative period. Evan's ratio and frontal-occipital horn ratio were found closely to correlate with the need for postoperative shunt. The requirement for shunt was higher in patients with midline tumors, tumors with brainstem infiltration, medulloblastoma and ependymoma, and in those who underwent postoperative EVD.

Conclusion: The low postoperative shunt insertion rate led us to believe that the routine use of preoperative CSF diversion procedures is not entirely justified. Factors such as degree of ventriculomegaly, tumor subtype, location of tumor, presence of brainstem invasion, and need for EVD in the postoperative period, which showed a statistically significant association with the postoperative shunt requirement in our study, should be considered when the decision regarding treatment is made.

Epilepsy/Functional

Acute crossed cerebellar diaschisis in a pediatric patient with nonconvulsive status epilepticus mimicking acute shunt failure

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Objective: Abnormalities on computed tomography and magnetic resonance imaging (MRI) associated with convulsive or nonconvulsive status epilepticus have been reported in the literature. Crossed cerebellar diaschisis (CCD) has been rarely observed in this context and refers to signal abnormality in one cerebral hemisphere and the contralateral cerebellum. However, in the pediatric literature, CCD is only described as a chronic state related to long-standing cerebral damage.

Methods: We report the case of a 3-year-old girl with ventriculoperitoneal shunt for post hemorrhagic hydrocephalus who presented with an acute CCD. In addition, a literature search was performed.

Results: The young patient was admitted to our department after a short episode of absence. The initial investigation revealed no neurological deficits and MRI demonstrated no new radiological abnormalities. Four days later, the patient developed acute neurological deterioration (GCS 7) with bilateral mydriasis. Repeated MRI demonstrated a new increased signal intensity involving the whole right cerebral hemisphere and the left cerebellum especially on T2-sequences. Additionally, both areas were signal intensified in diffusion-weighted sequences with corresponding apparent diffusion coeffi-

cient abnormality. An external ventricular drain was inserted due to suspicion of shunt failure. However, electroencephalography showed abnormalities. Treatment with levetiracetam was started and neurological status recovered quickly. Follow-up MRI showed regression of the abnormalities.

Conclusions: The present case report demonstrates a pediatric patient with an acute CCD mimicking shunt failure. To our knowledge, CCD has not been previously reported as an acute phenomenon in a child with non-convulsive status epilepticus.

Auditory brainstem implant

Chinmay Bhalke, Pranav Kumar, Ameet Kishore (Indraprastha Apollo Hospital, New Delhi, India)

Aim: We describe our experience with the auditory brainstem implantation for a non-NF2 indication in a 4-year-old child.

Objective: Auditory brainstem implantation is classically indicated for adults with bilateral auditory-vestibular nerve tumours, as in NF2, where the implant is placed at time of tumour removal. Lately the indications have broadened to include non NF2 conditions such as congenital hypoplasia/aplasia of the cochlea or cochlear nerve.

Results: This child had bilateral cochlear and cochlear nerve hypoplasia. ABI surgery was carried out successfully by our own Indian team of ENT and neurosurgeon. This presentation highlights key surgical steps for optimum positioning of the electrode and its outcome.

Conclusions: Auditory brainstem implantation is a complex and intricate surgical procedure requiring rigorous planning, close team work among skilled surgeons and audiologists experienced with sophisticated intraoperative electrophysiological testing to achieve a successful outcome.

Preliminary experience using intraoperative ultrasound during surgery for intractable epilepsy in children

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Introduction: Surgical resection of epileptogenic tissue offers a high chance of seizure control but the recognition of the margins of the epileptogenic zone can be challenging. Intraoperative electrocorticography and neuronavigation have been used in outlining the epileptogenic zone and outline the boundaries of specific lesions associated with epilepsy.

Intraoperative ultrasound imaging can provide information regarding the location and extent of lesions. We

describe our preliminary observations using intraoperative ultrasonography (US) to localize epileptic pathology, guide resection, and assist in placement of depth electrodes.

Methods: Preoperative and postoperative imaging studies, surgical, pathology reports, and hospital records were reviewed retrospectively for 22 procedures where intraoperative ultrasonography was performed during surgery for intractable epilepsy in patients, aged 3 months to 17 years.

Results: The procedures included lesion resection and electrode placement. Final diagnosis included cortical dysplasia in eight cases, cortical tubers in six, tumors in five, and other lesions in three. Intraoperative imaging provided valuable information on the localization of the lesion, extent of resection, and electrode placement. It was of special value in guiding entry to the lateral ventricle during hemispherectomies. No untoward events attributable to the imaging were encountered. Postoperatively, all patients had documented successful resection of the lesion or placement of electrode.

Conclusion: Intraoperative US can safely assist pediatric neurosurgeons during treatment of patients with intractable epilepsy. Its ease of use and ability to distinguish areas of cortical dysplasia from normal brain can help achieve precise resections, guide placement of depth electrodes, and assist in performing hemispherectomies.

Seizure control in dysembryoplastic neuroepithelial tumours

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Dysembryoplastic neuroepithelial tumours (DNTs) are developmental tumours, usually associated with refractory epilepsy. We present our series of 100 children who underwent surgery for these tumours between 1991 and 2010. This is an update to a previous presentation, after a thorough histological review.

There were 62 boys and 38 girls, with age range of 6 months to 17.5 years (mean 10.5 years). Tumours fell into two broad groups: those with classical diagnostic features of DNT (classic), e.g., the specific glioneuronal element, and those with features suggestive but not diagnostic of DNT (probable DNT). There were 60 in the classic group and 40 in the probable group. Of the 100, only five underwent repeat procedures for poor seizure control. The major complications included three with temporary motor deficits, three with subdural haematoma that needed evacuation, and two children with CSF leaks. The follow-up ranged from 1 month to

18.3 years (mean of 3.75 years). Five were lost to follow-up. In the classic group, 38 children (63%) were seizure-free (Engel class 1A) and discontinued the anti-epileptic medication, while 11 children (18%) were in 1B, two in class 3, and six in class IV. In the probable group of 40 patients, complete seizure control was obtained in 26 (65%), while eight children (20%) were in class 1B, two in class III, and two in class IV. Resection of DNTs is associated with good seizure control. Residual lesions may be followed up with frequent scans and reoperations may be undertaken if seizures are not adequately controlled.

A new hemispherotomy technique that might reduce motor impairment. preliminary experience in three cases

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Anatomical hemispherectomy for epilepsy was often followed hydrocephalus and hemosiderosis. Functional hemispherotomy has been described to avoid gross hemisphere removal and achieves a complete disconnection by sectioning the principal commissural fibers through a transventricular approach. Hence, the risk of hydrocephalus and subdural collections still exists. On this basis, we have developed a new technique aiming to get the same disconnection via paramedian extraventricular route.

Three children affected by drug-resistant epilepsy due to hemispheric pathology were submitted to extraventricular hemispherotomy. Mean age was 7.2 years (range 2.5–15 years); there were two males and one female. Two were post-traumatic epilepsy, one a diffuse hemispheric cortical dysplasia. At pre-operative neurological examination, all children showed hemiparesis contralateral to damaged hemisphere. Mean follow-up was 14.7 months (9–24 months).

Surgery was always computer-assisted. After a 4×4 cm fronto-parietal right craniotomy with exposure of middle line a multi-step section of anatomical commissures was made: (1) little corticectomy and section of gyrus rectus, hemispheric white matters fibers, hippocampal commissure, and posterior commissures through transcortical vertical paraventricular approach to interpeduncular and perimesencephalic cistern; (2) complete corpus callosotomy through interhemispheric approach. In the early post-operative period, all patients worsened the pre-existing hemiparesis. At last follow-up, all children are seizure free and able to walk. They still show a slight hemiparesis in resolution. None of them developed hydrocephalus.

This new technique achieves the same good results in seizure control and seem to enable an earlier motor recovery and to reduce the risk of hydrocephalus.

Incidence of hydrocephalus following hemispheric surgery in the surgical management of uncontrolled epilepsy

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A retrospective case note review was performed on all patients undergoing hemispheric surgery for the surgical management of their medically uncontrolled epilepsy between 1991 and 2010. All surgery was carried out at a single institution and by the senior author. A total of 144 patient records were reviewed, of whom 20 patients had hydrocephalus. Six of these patients (3%) had hydrocephalus prior to their hemispheric surgery whilst 14 patients (10%) developed hydrocephalus subsequent to their epilepsy surgery. In the hydrocephalus group, there were 15 males and five females.

Of the patients with hydrocephalus the diagnosis at surgery was as follows:

- Hemimegalencephaly 7/20 35%
- Cerebrovascular acc 6/20 30%
- Rasmussen's encephalitis 3/20 15%
- Cortical dysplasia 2/20 10%
- Sturge Weber 1/20 5%
- Encephalitis 1/20 5%

Time from hemispheric surgery to the development of hydrocephalus varied from 7 days to 6 years.

Following shunt insertion in those that developed hydrocephalus, 7/14 (50%) required subsequent shunt revision for shunt malfunction, three patients requiring more than one shunt revision. Of the six patients who had treated hydrocephalus before their hemispheric surgery, three patients (50%) needed shunt revision.

The potential factors influencing the development of hydrocephalus will be discussed particularly the underlying diagnosis and the extent of resective surgery performed. This retrospective review provides valuable data in the pre surgical counseling of families prior to surgery.

Seizure control after epilepsy surgery: experience in children and adolescents

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Objective: Surgery for refractory medical epilepsy is well accepted as efficient treatment for children suffering the

debilitating consequences of repetitive seizures. The goal of this study was to analyse seizure outcome and complications after epilepsy surgery in our center.

Methods: Data were retrospectively reviewed from 66 patients aged from 5 months to 16 years old who underwent epilepsy surgery at the Geneva University Hospital and Lausanne University Hospital between 1997 and 2010. Epilepsy surgery included temporal surgery (27 cases), extra-temporal surgery (29 cases), and hemispherotomies (10 cases). Three aged groups were identified: under 5 years old (22 cases), between 6 and 11 years old (23 cases), and between 12 and 16 years old (21 cases). Mean follow-up was 27.25 months (range, 4–96 months).

Results: The overall seizure-free rate for the period of follow up was 63.3% (42 out of 66 cases). Interestingly, the seizure free outcome was significantly better for temporal epilepsy (81.4%) compared to extra-temporal epilepsy (55.1%; $p=0.027$, <0.05). At least 56% of patients (37 out of 66 cases) had a reduction in their post-operative antiepileptic medication. Complications included five transient hemiparesis, two wound infections, one cerebral oedema after hemispherotomy, one extradural hematoma and one subdural hematoma after insertion of subdural grids.

Conclusions: Efforts to promote epilepsy surgery in children are crucial since it may significantly impact on the cognitive development of these children. A better understanding of extra-temporal epilepsy is needed to help improving seizure free outcome in this large group of patients.

Transcallosal resection of hypothalamic hamartoma for intractable epilepsy

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Purpose: This study aims to present the results of transcallosal surgical resection of hypothalamic hamartoma (HH) in 37 patients with refractory epilepsy in a prospective outcome study.

Methods: Patients with refractory epilepsy symptomatic to HH were referred for surgical resection of their HH (mean age, 8.7 years; range, 1.25–35 years). A transcallosal transeptal interforniceal approach was used to remove the hamartoma. Outcome assessment included determination of postoperative seizure frequencies in comparison to baseline and the incidence of postoperative complications. Follow-up data were obtained through clinical records and phone calls.

Results: Thirty-two patients were followed-up and five patients were lost. The average postoperative follow-up interval was 56 months (range, 30–108 months). Twenty-

two (68.7%) patients were completely seizure free, and four (12.5%) had at least a 90% improvement in total seizure frequency, four patients (12.5%) had a reduction in total seizure frequency to 50–90% of preoperative baseline, and two patients (6.25%) experienced no change in seizure frequency. Transient postoperative memory disturbance was seen in 12 (37.5%) patients, but persisted in only four (12.5%). Transient hypernatraemia developed in 10 patients and hyponatraemia in 19 patients. Two patients had transient diabetes insipidus.

Conclusions: Refractory epilepsy associated with HH can be safely and effectively treated with surgical resection by a transcallosal, interforniceal approach. Short-term memory deficits, hyponatraemia, and hypernatraemia appear to be transient for most patients. Complete resection or subtotal resection yields the best result.

Extratemporal epilepsy surgery around eloquent cortex using noninvasive presurgical evaluation. An analysis of 14 cases

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Objective: This study aims to assess surgical outcome in refractory epilepsy (RE) patients with extratemporal lesions (ETL) around eloquent cortex

Method: Surgery was performed in 14 patients having RE due to ETL around eloquent cortex at King Edward VII Memorial hospital Mumbai, India in year 2008–2010. Presurgical evaluation included dedicated MRI brain (1.5/3 T), ictal and interictal video EEG, neuropsychology, and psychiatric assessment. Preoperative invasive monitoring neither was not performed. Fourteen patients having concordant data were selected. Surgery was performed using intraoperative electrocorticographic guidance and neuronavigation. Corticectomy with or without multiple subpial transections was performed. Postsurgical outcome was assessed according to modified Engel's classification.

Results: Fourteen patients with age range 8–21 years were assessed. Average seizure duration till surgery was 12.2 years. The follow-up period ranged from 6 to 18 months. Seven patients had a new transient postoperative neurological deficit during immediate post-operative period which recovered completely in the following 2–3 weeks. All patients showed improvement, 11 in Engel class I and 3 in class II. The seizure outcomes were comparable in patients with neoplasm and cortical dysplasia. The cost of entire treatment was (300–400\$).

Conclusion: Patients with RE due to ETL around eloquent cortex can achieve good seizure outcome following surgery. Optimal presurgical evaluation is paramount. Good outcome

was possible in whom the lesion was well defined on MRI brain, concordant EEG and MRI data, and intraoperative identification of epileptic zone by corticography. The seizure outcome in patients in whom invasive presurgical evaluation is not possible can be gratifying in well selected patients.

Epilepsy surgery in children—outcome and its predictors

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Objective: This study aims to assess outcome of epilepsy surgery in children with medically refractory partial epilepsy and to determine the predictors of outcome.

Methods: Retrospective analysis of pre-surgical, surgical and post surgical data was performed in 118 children who had at least 1 year post-surgery follow up and were evaluated with non-invasive protocol. Outcome was assessed according to Engel's outcome classification. Stepwise regression followed by logistic regression analysis was employed in data analysis.

Results: Mean follow up was 42 (12–62) months; 60(51%) were males. The age of onset of epilepsy was less than 2 years in 34 (28.88%). The commonest surgery performed was a temporal resection 70(60%). Extra temporal resection (commonest—frontal) was performed in 28, hemispherectomy in 14, and callosotomy in 5. The commonest pathology was hippocampal sclerosis in adolescents and developmental, tumoral lesions, and gliosis in children. At last follow-up, 72 (61%) were seizure free and Engel's favourable outcome was noted in 86 (72.8%). After stepwise regression analysis, variables found to be significant ($p < 0.05$) and predicting a favourable outcome were lesion on MRI, normal IQ, and partial seizures without secondary generalisation. Bilateral spikes on interictal EEG and acute post operative seizures were predictors of poor outcome.

Conclusion: Favourable outcome after epilepsy surgery can be obtained in children with temporal lobe epilepsy with HS and lesion related epilepsies in developing countries with limited resources, after evaluation with a non-invasive protocol.

Anesthetic management in pediatric epilepsy neurosurgery: a retrospective analysis of perioperative morbidity and mortality in a level-4 center in India

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Introduction: More and more pediatric aged group of patients is undergoing epilepsy neurosurgery in developing countries. Since starting comprehensive epilepsy program,

we have operated 311 pediatric patients for various epilepsy surgeries. During this period, a total of 1,200 patients underwent epilepsy surgery in our institution. The aim of this study is (1) to find perioperative morbidity and mortality due to anesthesia in pediatric patients undergoing epilepsy surgery and (2) to compare the results with that of adult patients undergoing epilepsy surgery.

Material and method: Three hundred nine patients underwent epilepsy surgery under general anesthesia. Two patients were operated using conscious sedation and monitored anesthesia. The youngest child was of 11 months old who underwent epilepsy surgery. General anesthesia consists of induction with either Sevoflurane or Thiopental or Propofol. Muscle relaxants facilitated endotracheal intubations and intermittent positive pressure ventilation. Anesthesia was maintained with oxygen, nitrous oxide and Isoflurane or sevoflurane or propofol. A central vein cannulation and invasive blood pressure was measured in all the patients. At the end of operation, an attempt was made to extubate all the patients. Electrocorticography were performed successfully in all these pediatric patients

Results: There was no morbidity or mortality in pediatric patients in the perioperative period. However, there were two adult patients that had mortality during postoperative period following epilepsy neurosurgery.

Conclusion: We conclude that anesthesia is safe for pediatric patients undergoing epilepsy neurosurgery. Perioperative outcome is comparable with that of adult patient undergoing epilepsy neurosurgeries.

The importance of subdural electrode recordings in the planning of cortical resection for epilepsy

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Cortical resection for epilepsy can be planned on concordance of seizure semiology, MRI findings, and video telemetry. However, telemetry using subdural electrodes provides far more accurate information, as illustrated in our patient.

Our patient had medically intractable epilepsy, with several major seizures every day. Semiology suggested left frontal and temporal seizures. Surface EEG was non-localising. MRI showed left frontal and temporal gliosis. These gliotic areas were resected age 12, with confirmatory follow-up MRI. Initially, seizures improved slightly, then deteriorated to worse than pre-operatively. Repeat MRI showed only post-operative changes. External telemetry was again non-localising. Insertion of intracranial electrodes and further video telemetry

was performed. This captured seizure activity in frontal and temporal lobes with discrete ictal onset zones. Neurostimulation was performed through electrodes to obtain map of functional eloquence. On basis of this, further surgical resection was planned at age 14. Three frontal lobe areas and temporal ictal onset zone were resected, combined with anterior temporal lobectomy and amygdalohippocampectomy. Post-operatively, he developed homonymous hemianopia, nominal dysphasia and initial memory problems. Memory has improved to pre-operative state and speech has improved significantly. Visual field defect persists. At 15 months post-operatively, he has occasional absences but no major disabling seizures.

This case suggests invasive monitoring with subdural electrodes provides accurate information about ictal onset zones. Neurostimulation through electrodes allows eloquent areas to be charted very precisely. This is essential when planning cortical resection for complex epilepsy and in revisional surgery. Although invasive, the technique is safe, reliable and well tolerated in children.

Limited resection of epileptic focus in a child with bilateral Sturge–Weber disease offers control of epileptic seizures

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Introduction: Sturge–Weber disease (SWD) causes progressive damage to the cerebral hemispheres and severe epilepsy, often poorly controlled with medication. In unilateral disease hemispherectomy improves epilepsy control. In the presence of bilateral disease, hemispherectomy may not help as the contralateral affected hemisphere will continue to cause epilepsy.

Case report: This is a report of an 18-month-old girl affected by SWD suffered by a mild left hemiparesis and progressively difficult to control complex partial seizures requiring four medications. On imaging, there was extensive disease in the entire right hemisphere, with calcification, pial angiosis and atrophy, and disease in the left frontal and occipital lobes. Video EEG monitoring demonstrated bilateral interictal epileptic activity but several subclinical seizures were recorded arising predominantly from a right frontal focus. Decision was made to attempt surgical excision of this focus. During craniotomy, intraoperative EEG recordings obtained from all the hemisphere surface localized the right frontal epileptic focus, extending over two adjacent gyri. A limited resection of the

mapped area was performed. Recording at the end of resection did not show any epileptic discharge in any area of the exposed hemisphere. There were no complications. There was a significant improvement of epilepsy and a corresponding improvement of behavior and development. Reduction of medication is in progress.

Discussion: In the presence of bilateral SWD hemispherectomy, with all its potential complications, is unlikely to offer control of epilepsy. Limited cortical resection of electrically active areas can improve epilepsy and leaves the option for further limited resections or hemispherectomy at a later stage if needed.

Bilateral globus pallidus internus placement of deep brain stimulation electrode using intraoperative MR control in a 12-year-old child with dystonia

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Introduction: Deep brain stimulation (DBS) therapy with globus pallidus internus (GPI) targeting is effective in children with dystonia. Accuracy of electrode placement is very important and several methods have been employed to ensure it, including microrecordings and fluoroscopy screening. We describe the use of intraoperative MR control.

Case report: A 12-year-old girl suffered by progressively deteriorating secondary dystonia with severe episodic dystonic head and neck hyperextension and dystonic movements of the right arm and torso. Oral medication and BTX-A treatment failed to control the symptoms. On CT scan, there were bilateral calcifications of the caudate nuclei heads. On MR scan, the GPI was visualized anatomically intact. DAT scan showed intact dopaminergic system. It was decided to proceed to bilateral DBS implantation with GPI targeting. The cranial part of the procedure was performed in a Philips 1.5T open MR suite, using the Leksell frame and a T-1 volumetric sequence for target calculation. Post-placement imaging demonstrated that the electrodes were too deep compared to the intended target. The wound was reopened and the electrodes were withdrawn to the calculated length. Final imaging showed good electrode placement. Subsequent IPG implantation was performed in the regular operating suite. There were no complications. The clinical result was very good at 2 months, with good control of head posture and drastic reduction of dystonic movements.

Discussion: Despite the technical difficulties of operating within the MR suite, it is possible to place DBS electrodes

under intraoperative MR guidance. Further refinement of the technique will reduce operating time and improve electrode placement accuracy

Our experience with 100 pediatric patients at Epilepsy Surgery Center, Prague

Michal Tichy, Pavel Krsek, Vera Sebronova, Alena Jahodova, Vladimir Komarek (University Hospital, Prague, Czech Republic)

During the period of 10 years, we have operated on 100 pediatric patients with refractory epilepsy. In these patients, we have performed 111 resective procedures. In eight patients, the second operation and in two cases, the third more extensive resection was indicated.

The seizures started from 0 to 17 years of age (average 5) and the age at surgery was from 4 months to 20 years (average, 11.5). The most frequent etiology of refractory epilepsy in our group was focal cortical dysplasia (isolated pathology in 33 cases, in 14 cases associated with hippocampal sclerosis, in 22 cases associated with benign tumors). In 75 patients, we have performed a single-stage operation based on intraoperative corticography, 29 patients were operated after the previous implantation of invasive electrodes. In four children, the awake craniotomy was performed.

According to the extent of surgery, we have performed 42 focal resections, 41 lobectomies, 20 multilobar resections, and eight hemispherectomies. In 60 children, we have the follow up period of more than 2 years. In this group of patients, 47 children (77%) are seizure free; in 10 of them, it was possible to terminate the automated external defibrillator treatment. The follow-up period of more than 5 years was in 32 children, 22 are seizure free (69%). We have observed the temporary neurological changes with complete recovery in 29 patients. In 13 cases, the expected deficit and in four cases, unexpected deficit was observed postoperatively.

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Effect of intrathecal Baclofen on diffuse spasticity and generalized dystonia in nonprogressive disorders (a prospective study)

Purohit AK, Vár Prasad

Methods: The study (September 2004–December 2008) included patients with generalized dystonia ($n=8$) and spasticity ($n=8$) of various nonprogressive causes. The patients were selected after positive results from a trail dose (50 or 100 mcg) injection of baclofen intrathecally. A Synchronmed type 1 or 2 pumps was placed with the catheter tip passed up to T4–T6 level. Doses were regulated by telemetry using a programmer along with regular

refilling of the drug. Response to the treatment was judged based on change in various impairments and functional scales. The follow-up period was 1–45 months.

Results: Spasticity and dystonia improved in all patients from grades 4–5 to 3 and from 19 to 13.62, respectively. The response was with higher doses in dystonia (18.365 $\mu\text{g}/\text{kg}/\text{day}$) compared to spasticity category (11.0071 $\mu\text{g}/\text{kg}/\text{day}$). Spasm frequency reduced from 3 to 1. functional improvement by WeeFIM Scale in spasticity patients was 00.75 points on the average and 5.25 in dystonic category. The GMFM score improved by 0.82 points in spasticity and 0.75 points in dystonic patients. A total of six pumps were removed due to various causes. One pump was successfully reimplanted after wound healing in patient with flap necrosis.

Conclusion: Continuous intrathecal baclofen offers a good modality of treatment for selected cases of generalized dystonia and spasticity. Most of the complications are minor and could be managed conservatively.

Vascular

Surgically managed paediatric intracranial aneurysms: how different are they from adult intracranial aneurysms?

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Paediatric intracranial aneurysms are rare; about 0.5–6.8% of intracranial aneurysms occur in patients aged less than 19 years. We retrospectively reviewed all intracranial aneurysms in children less than 19 years of age that were surgically treated over the last 25 years at our institute. From our aneurysm data base maintained at our institute, records of the adult intracranial aneurysms operated during the same time were also reviewed. Thirty-three children under 19 years of age with 34 aneurysms were managed at our institute over the last 25 years. Age range was 11 months to 18 years with a female to male ratio of 1.5:1 (20/13). Twenty-five patients out of the 33 were treated surgically. Twenty-four (73%) patients presented with subarachnoid haemorrhage, two presented with intracerebral haematomas, four had cranial nerve deficits. Seizures were seen in six (18%), limb weakness in seven (21%) and associated heart diseases in two. Twenty-six patients (79%) patients were in good clinical grade. ICA bifurcation was the commonest site (11/34–32%) followed by distal MCA (6/34) and MCA bifurcation (5/34). Large and giant aneurysms were seen 14 cases. Twenty-five patients were treated surgically (21, clipped; four, proximal ligation or trapping). Vasospasm was

detected on DSA in five. There was 8% morbidity and 0% mortality in the operated group. Overall good outcome following surgery was seen in 20 cases (80%). The results are then compared and contrasted with those of adult aneurysms operated in our institute and in literature.

Comparison between adult and pediatric patients with moyamoya disease concerning the postoperative complications

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From 2007 to 2010, 39 patients with moyamoya disease (MMD) were operated upon at National Taiwan University Hospital. Twenty-two of them were older than 18 years of age. The remaining were pediatric patients. The male/female ratio is 1:2 in both adult and pediatric patients. All adult patients except one received direct revascularization. All pediatric patients except one received indirect revascularization, mainly encephalo-duro-arteriosynangiosis.

The associated diseases of MMD was present in eight (36%) and six (35%) of the adult and pediatric patients, respectively. In adult patients, DM (five patients), hypothyroidism (two patients), and SLE (one patient) were the concomitant diseases. In pediatric patients, neurofibromatosis (NF-I, 2 patients), hypothyroidism (one patient), Jeune syndrome (asphyxiating thoracic dystrophy, one patient), paroxysmal nocturnal hemoglobinuria (one patient), and thalassemia (one patient) were the concomitant diseases. The postoperative complications occur in six (27%) and two (12%) of the adult and pediatric patients, respectively. In adult patients, the complication included acute subdural hematoma (three patients), subarachnoid hemorrhage (one patient), cerebral infarction (one patient), and systemic infection (three patients). In pediatric patients, wound infection happened to one patient undergoing direct bypass surgery, and one mortality caused by respiratory failure due to preexisting asphyxiating thoracic dystrophy.

In conclusion, direct revascularization surgery may induce more complications such as new intracranial hemorrhage and wound infection in both adult and pediatric patients. The associated systemic disease is common in MMD patients. The presence of the associated disease may complicate the postoperative condition that even caused postoperative mortality in one case.

Pre- and post-operative cerebrovascular reactivity to acetazolamide measured by the dual-table autoradiographic method to quantify two sequential rCBFs in a single 123I-IMP SPECT session in younger patients with moyamoya disease

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Using positron emission tomography and single-photon emission computed tomography (SPECT), normal value and 3D distribution for regional cerebral blood flow (rCBF) in adult are well established, but not for children. In consideration of revascularization surgery for patients with moyamoya disease, measurement of cerebrovascular reactivity (CRV) to acetazolamide (ACZ) provides useful and essential information. The dual-table autoradiographic (ARG) method is reliably used for adult to quantify rCBF before and after an ACZ challenge. In the present study, to investigate the usefulness of the dual-table ARG method for younger population, the authors analyzed rCBF and CRV to ACZ before and after surgery in younger patients.

Between 2008 and 2010, 14 patients younger than 19 years of age were analyzed. The mean patient age was 14.5 years. Direct and indirect bypass were performed in all patients; 1–2 months before and 3–6 months after surgery, the dual-table ARG method was obtained. Data analysis was performed using quantitative SPECT, iSSP5, SEE-JET, and NEURO FLEXER.

Lower values in rCBF and impaired CRV were observed in all cases preoperatively. Revascularization surgery was performed mainly over the brain surface with impaired CRV. After operation, normalization of CRV was observed in all cases and quantitative values and 3D-distribution of rCBF in each patient were feasible to those in normal young adult.

The dual-table ARG method can be reliably used in younger patients. Evaluation of CRV to ACZ by the dual-table ARG method provides useful information for the surgical planning of younger patients with moyamoya disease

The role of miR-196a2 SNPs in moyamoya disease

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Recent evidence suggests an association between the SNP rs11614913 in *hsa-mir-196a2* and malignant tumors in a brain model of ischemia and reperfusion. The

possibility of a correlation of SNP with moyamoya disease is compelling, but not proven despite the many possible genes investigated.

We recruited 107 patients with moyamoya disease and 240 healthy controls from a Korean study population and determined their genotype from whole blood. We compared the genotypes and allele frequencies of rs2910164, rs11614913, and rs3746444, and investigated the association of SNPs with age and clinical characteristics.

Rs11614913 in *hsa-mir-196a2* was significantly associated with moyamoya disease. Association of this SNP with adult age and cerebral infarction was statistically significant compared to the control group, but not with hemorrhagic moyamoya disease. The CT genotype of the *hsa-mir-196a2* sequence was represented at an increased frequency among patients with moyamoya. The CC and TT genotype distribution of the *hsa-mir-196a2* was not significantly different between healthy participants and moyamoya. The distribution of *hsa-mir-146a* and *hsa-mir-499* genotypes was not statistically different between groups.

Our data indicate that the SNP rs11614913 is significantly associated with moyamoya disease, as well as cerebral infarction and adult age in patients with moyamoya disease. This study demonstrates a higher frequency of a C/T genotype of the SNP rs11614913 in *hsa-mir-196a2*, which suggests that *miR-196a2* may play a role in the pathogenesis of moyamoya. Future studies investigating the role of Annexin 1 regarding this SNP in endothelial proliferation will be helpful in further elucidating the cause of moyamoya disease.)

Trauma

Transforming pediatric neurosurgical healthcare delivery in rural areas by use of integrated call centre(s)

Deepak Agrawal (All India Institute of Medical Sciences, New Delhi, India)

Background: There is poor penetration of neurosurgical healthcare delivery in rural areas. People have to travel long distances for neurosurgical consultations as well as for follow-up visits. Also, a significant proportion of population is illiterate and without internet access.

Aims and objectives: This study aims to assess the implementation and usefulness of a call centre in providing healthcare delivery for rural patients with head and spinal injuries.

Materials and Methods: This was a prospective observational study carried out over a 3-month period at a level I trauma Centre in New Delhi, India. An 11-seater call centre was outsourced to a private company and the hospital's Electric Medical records were integrated with the call centre operations. The Call Centre was given responsibility of

maintaining appointments and schedule clinics for the whole hospital as well as ensuring follow-up visits. Trained call centre staff handled simple patient queries and referred the rest via email to concerned doctors. Telephonic survey was done prior to the start of call centre operations and after 3 months to assess for user satisfaction.

Observations and results: There were 484 patients admitted in the neurosurgery during the study period. Of these, 63% ($n=305$) were from rural areas. The initial cost of outsourcing the call centre was US\$ 12,000 with a recurring cost of US\$ 2,609/month. User satisfaction increased from a mean of 52–96% following implementation of call centre operations.

Conclusions: As shown by our case study, call centres have the potential to revolutionize delivery of neurosurgical healthcare to rural areas in an extremely cost-effective manner.

Experience with mobile CT in neurosurgery ICU in a level 1 trauma centre in india

Deepak Agrawal, D Gupta, S Sinha, GD Satyarthee (All India Institute of Medical Sciences, New Delhi, India)

Background: Neurosurgical patients, particularly of severe head injury require frequent CTs of the head, usually at short notice. A mobile CT may prove to be invaluable for these patients.

Aims and objectives: This study aims to review the usefulness of mobile CT (Ceratom[®]) in Neurosurgery ICU at a level 1 trauma centre in India

Materials and methods: This review was carried out over a 32-month period. For the first 6 months, only the number of CTs done was available. However, data was collected prospectively from 1 Jan 2010 include variables like Glasgow Coma Scale (GCS) ventilator status and pressor support (dopamine and non-adrenaline) at the time of CT scan.

Observations: A total of 3,522 head CTs were done during the study period with an average of 3.6 CTs daily. Prospective data was available on 2,263 patients. Of these patients, 82.4% ($n=1,865$) were on ventilator, 81.1% ($n=1,843$) were on sedation, and 6.9% ($n=1$) were on pressor support at the time of CT head. The mean GCS at the time of CT was 8.2 (range, 3–15). The average time taken to do a CT scan (from ordering to transmission into PACS) was 11.4 min (range, 7.6–47 min). Image quality was judged to be excellent by all neurosurgical faculty in the ICU.

Conclusions: Mobile CT was found to be extremely useful for neurosurgery department and was found to be very easy to use, with fast scanning time and excellent image quality. The mobile CT is strongly recommended for any high volume neurosurgery department in the country.

Profile of paediatric head injury in a level 1 trauma centre in India

Deepak Agrawal, D Gupta, S Sinha, GD Satyarthee (All India Institute of Medical Sciences, New Delhi, India)

Background: Due to lack of dedicated facilities, paediatric head injuries continued to be managed in adult neurosurgery departments in India. This may have an adverse impact on the outcome in this population group.

Aims and objectives: This study aims to evaluate the demographic profile and outcome of head injuries in the paediatric population in one level 1 trauma centre in India

Materials and methods: In this retrospective study of over 1-year period (January–December 2010), records of all patients of head injury who visited the emergency department were evaluated and patients who were ≤ 18 years were included in the study. Demographic variables as well as in-hospital mortality (for admitted patients) was assessed for this group.

Observations: A total of 4,479 patients presented with head injury during the study period. Of these, there were 1,088 paediatric patients. The majority of children were in 0–5 age group ($n=419$), followed by 6–12 age group ($n=355$) and 13–18 age group ($n=314$). Of these, 912 were minor head injury, 52 were moderate head injury, and 124 were severe head injury. In-hospital mortality was 34% ($n=42$) for pediatric severe head injury as compared to 17% for adults and 35% for elderly (>60 years of age).

Conclusions: Mortality for pediatric head injury is almost double that of the adult head injuries in our study. One of the causes could be lack of dedicated facilities for paediatric head injuries.

Study of factors responsible for mortality amongst children with craniospinal trauma in neurointensive unit: Apex Trauma Centre experience from India

Deepak Gupta, Akash Mishra, Ashok Kumar Mahapatra (All India Institute of Medical Sciences, New Delhi/Delhi, India)

Back ground: Children account for one third of neurointensive admissions in apex trauma centres ICU. They have a high mortality/morbidity resulting from respiratory and septicemic complications directly related to duration of ICU stay.

Methods and material: A retrospective review of pediatric mortality was conducted using a prospective database for quality improvement during 2009–2010 at level I apex trauma centre of India.

Results: A total of 65 children (<18 years old; mean age, 6.7 years, 21 male/44 females) expired in trauma centre (1 year period). Most ($n=61$) were head injured and only

four cases had spinal trauma. Of these, 30 cases were operated for cranial/spinal trauma (45%), while the rest were conservatively managed. Mean duration of stay in ICU was 8.25 days. Eight patients had positive tracheal cultures. Sixteen cases developed systemic inflammatory response syndrome (SIRS) and nine patients had ventilator associated pneumonia. Meningitis was noted in two cases and in 15 children, associated co morbidity like pneumothorax and polytrauma accounted for mortality. Two patients developed acute respiratory distress syndrome (ARDS). One patient had acute renal failure from the initial time of presentation. Out of 61 patients of head injury, 14 patients died due to cause directly attributed to head injury. **Conclusions:** Duration of stay and ventilatory support is directly related to pulmonary complications in form of ARDS and VAP. SIRS was responsible for major cause of death. Misuse of antibiotics should also be avoided as it leads to rise of multidrug-resistant organisms. Pediatric neurotrauma outcome can be improved by minimising ICU stay and selective use of antibiotics.

Management of growing skull fracture: our experience about 15 cases

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Introduction: Growing skull fractures (GSF) are the rare complications of head injury in childhood. This entity consist of the skull fractures with an underlying dural tear that courses with a progressive enlargement of the fracture to produce a crania defect. The purpose of this study is the analysis of the aspects of this pathology as well as the evolution of its treatment.

Materials and methods: It is a retrospective study of 15 GSF cases treated in our institution; the average age of trauma is of 18 months, 13 patients were aged under 3 years old at the moment of the trauma. The average delay between the head injury and the diagnosis of GSF is 14 months.

Results: The cause of injury was fall. The initial plain X-ray of 13 patients showed a diastatic skull fracture. The diagnosis was made by palpation of the cranial defect and confirmed by skull X-ray, CT scan with 3D reconstruction and MRI. The leptomeningial cysts and brain tissue herniation were noted. All the children underwent a dural repair with pericranium or fascia lata. The cranial defect was covered by local calvarial bone fragments. One case developed meningitis and treated. After an average recession of 74 months, all the children are schooled with normal psychomotor development, one patient is always under antiepileptic drugs. The radiological control shows a good ossification of the osseous defects.

Conclusion: Children under the age of 3 years with a diastatic fracture more than 3 mm are more prone to develop GSF and have to benefit from a clinical and radiological surveillance. A premature repair allows avoiding progressive brain damages.

Pot Pourri

An experience of hemorrhagic disease of newborn [HDN] presenting with intracranial bleed in children: report of 10 children

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Introduction: Hemorrhagic disease of newborn [HDN] is an unusual form of coagulation disease commonly seen in children associated with acquired vitamin k deficiency.

Methods: Clinical presentation, neuroimaging findings, and surgical outcomes were reviewed for 10 children treated for HDN presented with intracranial bleed.

Results: Ten children (age range, 14 days–6 months) underwent surgery for HDN presenting with intracranial bleed with raised intracranial pressure. All children presented with raised ICP and abnormal INR. Seven children underwent decompressive craniectomy, one craniotomy and evacuation of bleed and two burr hole and drainage of the subdural bleed. All children improved neurologically except one who continues to have visual deficit.

Conclusion: HDN presenting intracranial bleed carries good prognosis if diagnosed at time and appropriated treatment is instituted.

Acute presentation of a Chiari I in an infant: case report

Sandrine de Ribaupierre, Maria Macdonald, Keng Yeow Tay, Narayan A Prasad (University of Western Ontario, London, Canada)

Symptoms of cough, drooling, respiratory distress, and progressive dysphagia are common in pediatric practice, and usually associated with respiratory illnesses. However, sometimes, they are related to an underlying neurological problem and early diagnosis is critical for timely intervention.

A 12-month-old male, previously healthy, presented with a short history of respiratory difficulty, drooling, choking and coughing episodes, coinciding with symptoms of an upper respiratory tract infection. He was treated as such, but over 5 days, he continued to worsen with fluctuating level of consciousness, supposedly worse after a lumbar puncture. A CT scan done subsequently showed ventriculomegaly, and the infant was transferred. The neurological assessment at admission showed an awake infant with a flat but full

fontanelle, and some signs of bulbar weakness. He was sedated for an MRI, which showed significant cerebellar tonsillar ectopia. After the imaging was done, the child was kept intubated, and progressively became tetraparesis. The placement of an external ventricular drain followed by posterior fossa decompression led to a gradual resolution of symptoms. At 2 months after discharge, near-complete restoration of feeding and resolution of respiratory symptoms and motor deficit occurred.

While this case represent an extremely rare presentation of a Chiari I, in infants, such presentation should be kept in mind as being compatible with a Chiari, as suggested also by a review of the literature.

The role of spatial abilities in learning neurosurgical procedures

Sandrine de Ribaupierre, Tim Wilson, Ladan Ashrafi, Roy Eagleson (University of Western Ontario, London, Canada)

Most surgeries require good spatial reasoning, especially neurosurgery, where anatomical landmarks may be subtle or nonexistent. For example, while performing a temporal lobectomy, the only visual landmarks are at the cortex, therefore finding the ventricle without neuronavigation requires knowledge of the anatomical structures and then estimate their position with respect to the position of the head. Without good spatial reasoning, the surgeon or trainee would be groping blindly to find the ventricle.

In the literature, it has been suggested that training can overcome any shortfall in spatial abilities. In our study, novices, residents and experts were administered a modified Sheppard and Metzler mental rotation task, where the subject was able to interact with the virtual object to change the point of view in order to respond in a spatial matching task. Half of the subjects were exposed to our virtual temporal lobectomy model, while half were shown 2D anatomical diagrams. Subsequently, everybody was tested on spatial anatomy, and their ability to target the ventricle in a brain was measured.

Experts were better both in the anatomy questions and in finding the ventricle; while novices with better spatial abilities were better in localizing the ventricle than matched controls. And exposure to the virtual lobectomy model enabled novices to perform better in the anatomical questions and ventricle targeting, as compared with others at their level without the virtual training.

Endoscopic endonasal resection of Rathke cleft cysts

Javier Gonzalez Ramos, Carlos Routamboul, Adrian Ratinoff, Martin Guevara, Graciela Zuccaro (Hospital Garrahan, Buenos Aires, Argentina)

Objective: Rathke cleft cysts (RCC) are benign lesions that can be diagnosed as an incidental finding, and sometimes

they grow and cause visual impairment, hypothalamic–pituitary dysfunction and headache. The aim of this study is to show our experience with this entity using an endoscopic endonasal approach (EEA).

Methods: A retrospective analysis was conducted in three patients with a diagnosis of RCC. Each patient's record and imaging data was reviewed (pre- and postoperative).

Results: All three patients underwent a purely EEA with the assistance of an otorhinolaryngologist. Complete removal of the cyst contents and its capsule was achieved. The patients' ages were 15, 16, and 17 years, respectively. All of them presented headache and pituitary dysfunction and one patient also presented visual impairment. There was postoperative improvement in all symptoms. There were no complications. The immediate postoperative was comfortable since the patients did not need nostril occlusion like in the traditional transphenoidal approach. The control MRI did not show residual lesion. The average hospital stay was 2.6 days (range, 2–3 days).

Conclusions: The EEA is a safe and effective approach in the treatment of RCCs. Improvement in visual and endocrine dysfunction can be expected after surgical decompression of the optic apparatus and the hypothalamic–pituitary axis. The main advantages of EEA over the traditional transphenoidal approach are a better view of the anatomical landmarks, the postoperative comfort of the patient, and the reduction of the hospitalization time.

Pediatric footsteps in the Department of Neurosurgery, SGPGIMS, Lucknow—a tertiary care centre

Raj Kumar (S.G.P.G.I.M.S., LUCKNOW/U.P., India)

The referral trend of pediatric cases reported between 2000 and 2007 revealed complex spinal and cranial pediatric references. The cranial references include neoplasm in which 78% were infratentorial. Medulloblastoma (48%), ependymoma (25%), choroid plexus papillomas (15%), and brain stem glioma (7%) were most frequently referred infratentorial neoplasm. Barring brain stem glioma, all showed good prognosis of about 75–80%. Craniopharyngioma (28%), primitive neuroectodermal tumor (PNET; 20%), Chiasmatic glioma (18%), DNET (10%), and ganglioglioma (9%) were most referred supratentorial neoplasm in which the prognosis was about 80–85%. In vascular disorders, we had most number of cases of AV malformation (43%), followed by aneurysm (37%), moyamoya (7%), and idiopathic (7%). Twenty percent of these children reported with intracerebral hemorrhage while 8% each reported with SAH and IVH. Overall, we had a mortality rate of 6.67%. We encountered significant percentage of inflammatory lesions and referral for endo-

scopic intervention in hydrocephalus as well. We are also part of International Infantile Hydrocephalus Society. The spinal reference includes CV junction anomalies, spinal dysraphism, and spinal tumors with frequency of 43%, 38%, and 17%, respectively. Complex spina bifida and multiple tethering were among them. Among spinal tumours, most common intramedullary spinal tumors were astrocytoma (25%) and ependymoma (16%), while neurofibroma (15%), neurolemmoma (4%), and schwannoma (4%) were the most encountered intradural extramedullary spinal tumors. Aneurysmal bone cyst, histiocytosis and PNET were few of referred extradural spinal tumors. As per the outcome, one-third of the referred patients which were initially in the poor grades of neurological deficits were reduced to one-tenth after the surgery.

How evolutionary anthropology informs the ontogeny (growth/development/maturation) of the immune response relevant to the brain/cranial base (CB)

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Introduction: General somatic growth slows down, 4.4 months gestation, yet growth/development/maturation of neural brain–CB/somatic immune system continues, full ontological pace.

Methods: This paper explores ontological significance of brain/CB and immune system in concert in fetus/neonate.

Results/discussion: CB studies show second trimester anterior CB begins faster elongation rate, $p < 0.001$, matching midline extension brain enlargement. A significant phylogenetic identity shift, early fusion of *intrinsically* controlled mid-sphenoid synchondrosis, is an added elongation enhancement, comprising crista galli/olfactory receptors. Unexpected link between immune/olfactory systems by major histocompatibility complex (MHC) transmits information about MHC genotypes/influences behavior/reinforces *new* human identity.

Maturation of human immune response/brain growth follows the same developmental pattern in the first year of postnatal life. Conserved genome plasticity of immune response is 515 million years old; basic strategy for antigen presentation, MHC molecules, with protein-binding region polymorphism was established. Conserved gene plasticity for neocortex development is 57 million years young; genes *RFPL1,2,3* duplication/cluster arrangement began neural expression for neocortex size/organizational changes.

Life history theory, focusing on nutrition/pathogens, favors trade-offs for privileged brain/immune systems, investment associated with increased society longevity. Embryonic

gene assembly confers vulnerability/responsiveness to environmental context the immature brain relies on to establish/guide pathways/connections. Maximized nutrition investment/outcome: 60% for brain growth on the third trimester/60% BMR neonate brain results in brain 3:1/neocortex 3.6:1 larger than expected. Fetal capacity for immune responsiveness develops on the first trimester; driven by omnipresent environmental pathogens, infancy/growth investment is maximization by rapid development of individually refined/costly specific immune-repertoire.

Conclusion: Evolutionary principles inform central design features for brain-CB/immune defenses, constrained by genetic programming with intergenerational effects: identity, plasticity, and fitness.

Ferromagnetic dissection: a new energy modality to improve precision of neural tissue incision, sealing, and coagulation

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Background: Most brain and spinal cord pathologic conditions are managed surgically with either electrosurgery, ultrasound cavitation and aspiration, or laser. Each method has some risk of "beyond the tip" effects related to tissue based energy pathways. Ferromagnetic (FM) dissection is a new method which incises by pure heat conduction delivered from the tip of the surgical handpiece with no adjacent energy absorption.

Methods: A variety of tissues in swine, goat, and rabbit were incised including skin, muscle, dura, and brain using the FM technique. These were compared to adjacent identical cuts with RF cutting and coagulation modes (monopolar cautery) as well as plain scalpel cuts. Incisions were subjectively ranked for hemostasis, drag, and conformity. In rabbits, acute and 14-day delayed histology as well as MR imaging were performed to determine depth of injury, reactive edema, and healing patterns. Data was analysed by a blinded radiologist and pathologist.

Results: FM dissection was hemostatic and produced the least damage at the cellular level. Similarly, MRI imaging demonstrated the least reactive edema in the FM incised tissue.

Conclusions: Brain and spinal cord incisions exhibit effective hemostasis and the least histological tissue injury with FM method. Similar results have been seen in muscle and dura. Tactile feedback is maintained in contrast to laser. Due to the temperature of superficial sealing of this method, minimal tissue tractional distur-

tion was seen and no tissue sticking was encountered. FM dissection shows promising qualities to improve outcomes of neurosurgical procedures.

Considerations about the cooperation between industry and scientific organisations

Christoph Miethke (Christoph Miethke GmbH & Co.KG, Potsdam, Brandenburg, Germany)

Medical devices are needed to treat patients. Whenever these devices are developed, the commercial background interferes with the promise of improvements. However, it seems to be more than questionable that technological innovation can be established without this commercial background. The good proposal of a surgeon will be introduced in clinical praxis only if a company takes the commitment and responsibility for this product.

The collaboration between clinical and "industrial" scientists is undoubtedly necessary. The development of new devices follows the rules of engineering sciences. A medical device without a clear scientific justification should rather be viewed as an esoteric approach. Especially in the medical field, it is the responsibility of the manufacturer to explain his concept. The introduction of new devices for the treatment of hydrocephalus happens preferably in closed meetings organised or sponsored by the company with the obvious economical interest. A public discussion based on scientific means about different technical concepts, which is at least partially led by the responsible and competent representatives of the manufacturing companies, could help to reveal clear differences and therefore insights. Many of the devices used in the field of hydrocephalus have never been conceptually discussed.

Cui bono? This is the outstanding central question whenever humans are acting or talking. Medical devices have an outstanding importance for the daily life of patients with hydrocephalus. The choice of technology used should be driven on scientific aspects: clinical and technical knowledge. The technical statements by the manufacturer should critically be valued by the neurosurgeons.

A regional paediatric neurosurgery referrals database: patterns of referral in more than 900 cases and evidence for shortfalls in resource provision

Roberto Ramirez, Amir Ramezani, Jatinder Singh, Bhoresh Dhamija, Desiderio Rodrigues, Guirish Solanki (Birmingham Children's Hospital, Birmingham, UK)

Brain malformations

Objectives: Following a review of specialist services at our institution supported by SHA and HCC a database of referrals was set up in 2009. This database captured the

referral source, provisional diagnosis, urgency, and ability to admit within a stipulated time frame for each clinical scenario.

Subjects: Nine hundred twenty-seven (368 girls, 552 boys, seven unknown) children were included in the database. The M/F ratio is 1.5:1, ages ranged from 1 day to under 18 years. Median age was 7 years.

Methods: We present a preliminary retrospective analysis of 927 patients referred to a regional paediatric neurosurgery unit from April 2009 to December 2010.

Results: There were 938 database entries. Eighteen (1.8%) blank/incorrect entries were excluded from the analysis; 4.5% had missing age data. About 20% were less than 1 year old. Twenty percent were 1–3 years, 7% 3–5 years, 28.6% were 5–12 years, and 21% were older than 12 years. The top five conditions (HI, HCP, tumours, infections, and spinal conditions) accounted for about 90% of emergency admissions. HCP and HI accounted for 40% of the referral workload and for the highest proportion of immediate (admit 51 h) and emergency referrals (admit 54 h). Life-critical immediate and emergency referrals were refused in 14 cases for lack of psychiatric intensive care unit/ward beds. Forty-nine children suffered additional transfer delays including retrieval. Of these, 29 (60%) needed immediate/emergency admission.

Conclusions: A paediatric neurosurgery referrals database is essential to identify patterns of care/delivery and resource gaps. This report identified local resource shortfalls which on addressing should lead to improved delivery of care and its use is highly recommended.

Endoscopic management of temporal arachnoid cysts: surgical technique and complication avoidance

Michael J. Fritsch (Dietrich Bonhoeffer Klinikum, Neubrandenburg, Germany)

Objective: Temporal arachnoid cysts present different challenges: indication, surgical technique, complication avoidance.

Methods: In a retrospective analysis of 21 pediatric (nine male, 12 female) patients treated endoscopically within a 4-year period (2006–2009), we evaluated patients for clinical outcome, complications, and revision rate. The mean age the time of surgery was 4 years and 8 months (6 weeks–14 years), the mean follow up was 3 years (minimum 1 year). We utilized neuronavigation for approach planning in all patients.

Results: We had to converse the endoscopic approach to a microsurgical procedure in 5/21 patients. The reason was either impaired vision due to bleeding or thick multilayer arachnoid membranes which could not be fenestrated by an endoscopic approach. Two patients developed subdural fluid collections after surgery which did not require further

intervention. Two patients developed hydrocephalus after cyst surgery and required a VP shunt. One patient was treated for a subgaleal fluid collection with repeated lumbar punctures.

Clinical results were: some reduction in headaches in half of the symptomatic patients, almost no reduction in cyst size after surgery, good flow void on T2 weighted flow sensitive MRI studies postop. No neurologic impairment occurred due to the surgery.

Discussion/conclusion: The indication for temporal arachnoid cyst surgery remains controversial. Navigation is helpful in approach planning. It is important to avoid bleeding into the cyst during surgery. The endoscopic approach is a valuable treatment option. Risks and benefits have to be considered carefully.

Encephaloceles—study of 110 patients at AIIMS over all 8-year period (2002–2009)

Ashok Kumar Mahapatra (AIIMS, Ansari Nagar, New Delhi, India)

Over an 8-year period (2002–2009), 110 encephalocele were treated at the Neurosurgery Department, AIIMS. This retrospective data is analyzed from computer data base to study age incidence, type of encephalocele and the outcome of surgery. Vast majority patients were under 2 years of age. Eight patients were between 5 and 10 years. Ten patients were neonate less than 30 days. All the patients had CT scan and over 30% also had MRI, in addition.

Sixty of these patients had occipital encephaloceles, 30 patients had anterior encephalocele, and seven had parietal encephaloceles. Eleven patients had giant encephaloceles (10%), of which nine were occipital and two anterior encephaloceles. Patients with hydrocephalus were subjected to CSF diversion in the form of VP shunt. All the patients except one were not operated. Anterior encephalocele patients, in addition to repair of encephaloceles also underwent corrective osteotomies for hypertelorism, when the hypertelorism was significant.

Overall results were good. There were five postoperative deaths (4.5%). Two were neonates with giant occipital encephaloceles, who developed hypothermia during surgery and did not recover. One patient of occipital encephalocele developed wound break down followed by fulminant meningitis, leading to death. The fourth mortality was in a 7-month-old child, who underwent surgery for anterior encephalocele, and post op aspiration pneumonia leading to death.

Encephalocele are rare and accounts less than 0.5% of our patients undergoing surgery. Occipital encephaloceles accounted for 65% followed by anterior encephaloceles 29%. Overall, result of surgery was good with a mortality of 4.5%.

Incidence of hydrocephalus after surgical correction of sincipital, parietal and occipital meningoencephalocele
 Panu Nacharong, Charoempong Chatdokmaiplai (Children's Hospital, Bangkok, Thailand)

Objective: Many articles have described about the incidence or rate of complication especially postoperative hydrocephalus, but no one have explain or identified factor that associated with these complication. The purpose of this study was to determine the rate of hydrocephalus after elective surgical repairs meningoencephalocele and to identify factors that related to the occurrence

Methods: A retrospective analysis of all surgical repaired meningoencephalocele patients performed between January 2004 to December 2010 at Children hospital (90 cases) and Ramathibodi hospital (five cases) was identified using medical records. Postoperative hydrocephalus was diagnosed when the patients have signs and symptoms of increased intracranial pressure (irritability, bulging of the fontanelle, prominence of scalp veins, and bulging of the wound with CSF leakage). Imaging was performed to confirm diagnosis. **Statistical analysis:** Univariate analysis with Fisher's exact test and ranksum test were performed to analyze individual factors and their impact on postoperative complications.

Results

- A total of 95 patients were treated by surgical corrections that were performed in over 6 years. Eighteen patients were excluded from this study due to loss of follow up. Seven patients (9.0%) had preoperative hydrocephalus and all have preoperative VP shunt before repair meningocele. The incidence of hydrocephalus after surgical correction overall was 7.79% (6 patients) and univariate analysis demonstrated that parietal type (three from six of postoperative hydrocephalus patients (50%)) had significantly more risk for postoperative hydrocephalus with 0.013 *P* value at 95% confidence interval. In all six patients, no one have documented that they have preop hydrocephalus.

For the establishing of criteria in prenatal diagnosis of intractable fetal brain malformation

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Recent advance in molecular biology made a significant advance in diagnosis for congenital disease. We made the protocol for prenatal diagnosis of XLH. For proper counseling, it is very important to develop the methods of prenatal diagnosis for isolated ventriculomegaly.

Methods: We made the data bank of the patients with intractable fetal brain malformation. Data include clinical information, images, molecular genetic information, pathological findings and biological samples such as tissues, cells, and DNA. Every doctor entry the cases by website protected in high security.

Result: Seventeen institutes over nationwide have been registered. Eight molecular biologists are registered as a team performing genetic analysis. One institute is registered as pathological evaluation. Four pediatric radiologists are nominated as radiological evaluation team. Fetal ultrasound evaluation are perfuming by experts. All of these brain malformation are included as various disease, each of these disease are orphan diseases and have rare cases and difficult to be exactly diagnosed. From Dec 2009 to Aug 2011, almost 108 patients are registered. Forty-three cases are diagnosed exactly by neuroradiological reports. In 13 cases, the pathological evaluation was performed. Molecule analysis was performed about several genes related to development central nervous system. Mutations of *LICAM* gene, *DCX* (double cortin), *ARX*, *CASK*, *Zic2*, and *TUB1A* were identified in each several cases.

Conclusion: Consultation system through website is useful for diagnosis of orphan disease. Not only classical methods of gene analysis technique but also comprehensive molecular analysis are required for the establishment of criteria for prenatal diagnosis in fetal brain malformation.

Outcome of posterior fossa decompression, duraplasty and tonsil reduction for Chiari malformation type 1

Xianlun ZHU, Hoitung Wong, Waisang Poon (Chinese University of Hong Kong, Hong Kong, China)

Background: Suboccipital craniectomy is the essential part of posterior fossa decompression for Chiari malformation type 1 (CM1). However, whether or not to open the dura and proceed with intradural maneuvers remains the subject of debate.

Objectives: An audit of syrinx improvement outcome after posterior fossa decompression for CM1.

Materials and methods: Sixteen consecutive patients with CM1 underwent posterior fossa decompression in our unit from 2000 to 2010. The mean age at operation was 24 (median, 21; range, 7–56, five cases age <18), with a male to female ratio of 6:10. All cases are symptomatic. Five (31%) cases had scoliosis. All but one had syringomyelia on MRI (92%).

All patients received suboccipital craniectomy and duraplasty. All but one underwent tonsil reduction.

Results: Of the 15 cases with syringomyelia, 13 showed significant and one showed moderate improvement on post-

operative MRI. The overall improvement rate was 93%. One case, who was 48 years old with an extensive syrinx from C1 to the conus, required syringoplural shunt eventually. One case with diplopia, nystagmus, and ataxia without syringomyelia also completely recovered after the operation. There is no surgical complication for all the cases.

Conclusion: For CMI with syringomyelia, high improvement rate can be achieved with suboccipital craniectomy plus duraplasty and tonsil reduction without surgical complication.

The neurosurgical treatment of an infant with encephalocraniocutaneous lipomatosis and extensive spinal cord involvement

Robert Ayer, Alexander Zouros (Loma Linda University Medical Center, Loma Linda, CA, USA)

Encephalocraniocutaneous lipomatosis (ECCL) is a rare neurocutaneous syndrome whose hallmark lesions are benign lipomas of the brain and spinal cord. We present a new case of a male infant with ECCL who has extensive brain stem and spinal cord lipomas. The management of this patient's hydrocephalus, cervicomedullary compression, tethered cord, and scoliosis over the course of the patient's first 2 years of life is described. During the course of his treatment, we were the first to perform endoscopic third ventriculostomy, and complete successful debulking of an intramedullary lipoma without post-operative neurological deficits, in a patient with this syndrome. This case is presented as an example of the problems likely to be encountered by neurosurgeons that treat patients with this syndrome.

Multidisciplinary approach for a large fronto-basal encephalocele

Dr. V. Velho, Dr. S. Khan, Dr. D. A. Palande, Dr. R. Mally, Dr. V. Agarwal, Dr. M. Sharma, Dr. Abhishek V (Grant Medical College and Sir JJ Group of Hospitals, Byculla, Mumbai)

Fronto basal encephaloceles are rare congenital lesions which form only 1.5% of all encephaloceles. Large cranial base defects, herniation of functional brain tissue, and craniofacial anomalies makes their management challenging. In our case report, we highlight a rare variety of large frontobasal encephalocele in an 8-year-old boy, in whom a multidisciplinary approach was used in the management, requiring the expertise of a pediatric neuroanaesthetist, neurosurgeon, pediatric surgeon and plastic surgeon.

Primary scalp AVM In paediatric population. Report of two cases and review of literature

Professor Rakesh Gupta (Department of Neurosurgery, Sri Aurobindo institute of Medical Sciences, Indore (MP), India)

Scalp AVM is rare condition. Very few cases are reported in paediatric population.

The vascular malformation of scalp is an abnormal arteriovenous communication located within the subcutaneous fatty layer of the scalp. Arterial feeders are usually from the vessels supplying the scalp.

Different nomenclatures are given to this condition namely cirroid aneurysm, aneurysm racemosum, and aneurysm serpentinum.

The etiology is controversial. It may be congenital or traumatic in origin.

Author reports two cases of scalp AVM in paediatric age group. Both the cases presented with progressively increasing pulsatile swelling. Surgical excision was done in both the cases.

Different treatment options and literature is reviewed.

Epidermoid cyst of the brainstem in children: case based update

Gopalakrishnan CV, Suresh Nair, Girish Menon, Amit Dhakoji

Objective: Brainstem epidermoid cysts are extremely rare in children. We report two additional cases of pediatric brainstem epidermoid cysts, underlining their clinical characteristics, the difficulties faced in the diagnostic work-up, and the surgical treatment adopted.

Materials and methods: In a retrospective analysis of pediatric brainstem tumors operated over the last 10 years, we report two cases of epidermoid cysts one being purely intraaxial. The cases were analyzed for their clinical presentation, imaging characteristics and surgical outcome. The results were compared to available data in literature.

Results: A 6-year-old girl presenting with headache was diagnosed to have a prepontine lesion with insinuation into the brainstem. A 2-year-old boy presented with hemiparesis and MRI revealed an intrinsic brainstem mass. Both the tumors were resected; subtotally in the first case. Histopathology was suggestive of epidermoid with infection. With presence of hyperintensity on T1-weighted images with restriction on DWI in a pediatric brainstem cystic lesion, one should consider the possibility of an infected epidermoid cyst.

Conclusion: The very existence of true intraaxial brainstem epidermoid is doubtful. Liquefaction of the cyst content due to infection may result in rapid increase in size with presentation in early childhood. The paucity of clinical signs in brainstem epidermoid can be attributed to neural plasticity. Brainstem epidermoids pose a surgical challenge and attempts at aggressive removal are fraught with increased risks of morbidity and mortality. Neurosurgical judgment at the time of surgery is warranted to ensure maximum resection while minimizing postoperative neurological deficits.

Craniofacial

Surgical treatment for the coronal synostosis: a review of 40 cases in 5 years

Naresh Biyani, Nitin Mokal, Uday Andar, Chandershekar Deopujari (BJ Wadia Children Hospital, Mumbai, India)

Craniosynostosis is an uncommon disease seen in children due to premature fusion of the cranial sutures. Premature fusion of coronal suture leads to coronal synostosis.

Aim: This study aims to review the 40 children with coronal synostosis operated in past 5 years in a tertiary care centre in Western India.

Material and methods: There were 25 male and 15 female. Age at surgery was 4 months to 5 years [mean of 12 months]. There were 30 children with unicoronal synostosis and bicoronal synostosis was seen in 10 with named syndrome in five children. Indication for surgery was raised intracranial pressure and cosmetic. There was no perioperative mortality. Post operative course was uneventful in all except three children who had superficial wound gape. Mean follow-up was 36 months

Conclusion: Coronal synostosis can be safely treated surgically with excellence. Various surgical technique are discussed.

Surgical approaches to the orbit: a study of 33 cases

Aadil Chagla¹, Devendra Tyagi² (¹King Edward Memorial Hospital, Mumabi, Maharashtra, India; ²B Y L Charitable Nair Hospital, Mumbai, Mhahrashtra, India)

Introduction: Approaches to the orbit are rare. The pediatric orbit is different from the adult. Wider bony exposures are often necessary to provide safe access to be orbit. We describe various surgical approaches for orbital tumors and other lesions.

Materials and methods: Thirty three cases (20 females and 13 males) operated by the senior author at the King Edward Memorial Hospital, Parel Mumbai is described over the past 11 years. The patients were investigated using plain X-rays, computerized tomography, and magnetic resonance imaging for most of the cases. (Excluded in this study are cases that underwent orbital exenterations as well as tumors that secondarily involved the orbit.)

Results: Seven cases underwent lateral orbitotomy; sub-frontal superior orbitotomy, five cases; supero lateral orbitotomy, 10 cases; fronto-orbito-zygomatic, eight cases; and fronto temporo-orbito-zygomatic craniotomies, three cases; and one case of anterior orbitotomy (one cases was operated twice). The histology included neurofibroma, five; optic nerve glioma, four; cavernous hemangioma, three; tuberculoma, three; fibrous dysplasia, three; lymphan-

gioma, meningioma, and aneurysmal bone cyst were also seen. The malignant cases included rhabdomyosarcoma and non-Hodgkin's lymphoma. The one case of foreign body was an air gun pellet. Complications included deterioration in vision in a case of optic nerve glioma and optic sheath meningioma; mild lateral rectus paresis in one case; and partial relief of proptosis in two cases of tumors. There was no mortality or infection.

Conclusion: Superolateral orbitotomy was the commonest approach. Adequate exposure is the key. Standard microsurgical skills produce excellent surgical outcomes.

Occipital decompression for recurrent non-syndromic craniosynostosis

Hassan Kadri (Clinic of Neurosurgery, Damascus, Syrian Arab Republic)

Introduction: Posterior decompression was described in many cases of syndromic craniosynostosis, we try in this study to evaluate this technique for recurrent non-syndromic craniosynostosis.

Patients and methods: Eight patients aged between 2 and 4 years suffering from non-syndromic craniosynostosis were operated on for recurrence of raised ICP signs by posterior decompression.

All the patients had had a cranial remodeling during the first year of life for their initial malformation. Later on, five of them developed a visual deterioration with recurrence of the synostosis while three developed visual deterioration with partial recurrence on the X-ray examination.

The eight kids underwent a posterior decompression by a bilateral occipital skin incision and a bone flap across the sagittal and the transverse sinuses.

Results: The five patients who were suffering from recurrence were partially ameliorated (three ameliorations and two stabilization of signs) while in the group of partial recurrence, two patients stabilize their symptoms and one continue to deteriorate.

A case of Apert's syndrome with concurrent occipital encephalocele

Ema Knight¹, David David², Stephen Santoreneos¹, Peter Anderson² (¹Department of Neurosurgery, Flinders University of South Australia, Adelaide, Australia; ²Australian Craniofacial Unit, Women's and Children's Hospital Adelaide, Adelaide, Australia)

Apert's syndrome (a form of acrocephalosyndactyly) is a rare genetic condition. Global prevalence approximates one in 200,000 live births, with affected individuals displaying characteristic craniofacial anomalies, syndactyly and variable intra- and extra-cranial abnormalities. Primary central

nervous system abnormalities have previously been reported in association with this condition, including megalencephaly, gyral abnormalities, pyramidal tract abnormalities, hypoplasia of cerebral white matter, and heterotopic gray matter. Similarly, rare cases of frontonasal encephalocele have been noted, as well as one case of occipital encephalocele in conjunction with agenesis of the corpus callosum. Here we present a further case of Apert's syndrome with concurrent primary occipital encephalocele. The patient described is an 11-month-old Indonesian female referred to the Australian Craniofacial Unit with features characteristic of Apert's syndrome. In Australia, she underwent fronto-orbital advancement for craniosynostosis and subsequent excision of the encephalocele as a separate procedure. She recovered without complication. The probability that this concurrent pathology represents a chance occurrence (combined prevalence estimate one in 1,000 million live births) is low. Alternative explanations involve a low-incidence association consequent to the underlying genetic condition, or a secondary phenomenon related to the altered intracranial pressures during development. The presence of the corpus callosum in this case is in contrast to that reported previously, and the proposed association with a short cranial base was also not found here.

Perioperative management of craniosynostosis

Pratima Kothare, Ranjana Das (Bombay Hospital Institute of Medical Sciences, Mumbai, India)

Introduction: Craniosynostosis is a premature closure cranial sutures in children that often require surgical correction. We review perioperative anesthesia management of craniosynostosis surgery in 20 children.

Material and method: Twenty children were reviewed operated for craniosynostosis.

A detailed review of preoperative, perioperative, and post operative records was done.

Observation and results: Age ranged from 6 weeks to 20 months [mean, 7 months] and bodyweight of 5–14 kg [mean, 8 kg]. Male/female ratio was 12:8. Balanced anaesthesia technique was followed with standard monitoring. Mean estimated blood loss was 15% of blood volume and was replaced meticulously. Core body temperature was maintained. Difficult intubation was anticipated in all syndromic children. All children were extubated immediate postoperative period and managed at psychiatric intensive care unit for further care. Mean operative duration was 3 h. Haemoglobin and hematocrit was repeated and blood collected in the drain was measured and replaced as per requirement.

Conclusion: Craniosynostosis can be safely operated with present perioperative anaesthesia management. Main concerns during the surgery are hypothermia and hypovolemia. Blood transfusion to be started with skin incision.

Management of fronto-nasal dermoids in children: an 8-year experience from a supraregional centre

Benedetta Pettorini, Giovanna Paternoster, Nicholas White, Hiroshi Nishikawa, Guirish Solanki, Peter Noons, Stephen Dover, Martin Evans, Desiderio Rodrigues (Birmingham Children's Hospital, Birmingham, UK)

Introduction: Dermoid sinus is the most common midline nasal anomaly. Intracranial extension has been reported in up to 45% of cases. Complete surgical excision is the only therapeutic option.

Objectives: The purpose of this study is to determine effectiveness and safety of a transcranial approach for treatment of fronto-nasal dermoids in children and discuss the merits of CT versus MRI in their pre-operative evaluation. **Design:** A retrospective review of fronto-nasal dermoids treated surgically over an 8-year period in a single paediatric centre. We analyse the presentation, surgical approach, post-operative complications and recurrence rate with our procedure.

Methods: Database was used to identify all children diagnosed as nasal dermoids. Case notes and radiology was used to identify and analyse the subgroup of children treated for fronto-nasal dermoids.

Results: We identified 34 patients with nasal dermoids of which 32% were transcranial. The median age at presentation was 2 years. Four patients presented with intermittent discharge while seven had swelling over the bridge of the nose or glabella. Radiology demonstrated evidence of intracranial extension in these cases.

No major complication was noted in our series. No patients had intracranial recurrence but extracranial (nasal) recurrence occurred in three cases.

Conclusions: Combined one-stage intracranial approach for treatment of nasal dermoids with intracranial extension is effective and safe. Preoperative evaluation is essential to rule out intracranial extension and we discuss the relative merits of CT versus MRI in this. The role of pericranial flaps, fibrin glue and duragen to prevent CSF leaks is discussed.

The growth of the skull in the first 18 months of life: findings from an MRI-based morphometric study

Christian Siegmund, Kohmal Solanki, Jan Poloniecki, Guirish Solanki (St. George's Hospital NHS Trust London, Birmingham, UK)

Objective: The morphometric cranial changes in the first years of life are poorly understood. The aim of this study is to introduce a new curvilinear skull measurement which allows describing skull growth more accurately.

Design: Cross-sectional study measuring universally accepted and new parameters of skull dimensions in MRI scans of healthy children at 1 monthly intervals.

Subjects: Three hundred seventy-six anatomically normal children aged between 1 and 540 days (179 girls and 197 boys). Infants with syndromic and single suture synostosis, hydrocephalus, Chiari malformations, trauma, metabolic diseases, and other obvious or diagnosed skull abnormalities were excluded.

Methods: Anteroposterior (AP), glabella-occipital (GOP), and biparietal (BP) distances were measured by three operators using IMPAX software (AGFA). GOP length is defined as the maximum distance between the occipital protuberance and the frontal bone measured along the skull contour of the vertex.

Results: Intra- and interobserver variance was calculated to be <1% (Student's *t* test, $p < 0.0001$). AP, GOP, and BP increase steeply in the first 9 months of life. Subsequent monthly growth changes reduce to less than 1% thereafter. More importantly, all three parameters show the same growth characteristics.

Conclusion: There is a statistically significant difference between male and female measurements in the first 18 months of life ($p < 0.0001$). The new curvilinear measurement of GOP might be an important adjunct to describe normal skull growth more accurately.

The shape of the skull in the first 24 months of life: findings from an MRI-based morphometric study

Christian Siegmund, Kohmal Solanki, Jan Poloniecki, Guirish Solanki (St. George's Hospital NHS Trust London, Birmingham, UK)

Objective: The most widely accepted measurement in assessing the shape of the human skull is the cephalic index (CI). The current normal range is 0.73–0.81. The aim of this study is to determine the change in CI in the first 24 months of life and determine if it conforms to normality. **Design:** Cross-sectional study measuring universally accepted parameters of skull dimensions in MRI scans of healthy children at 1 monthly intervals.

Subjects: Four hundred sixty-eight anatomically normal children aged between 1 and 729 days (218 girls and 250 boys). Infants with syndromic and single suture synostosis, hydrocephalus, Chiari malformations, trauma, metabolic diseases, and other obvious or diagnosed skull abnormalities were excluded.

Methods: Anteroposterior (AP) and biparietal (BP) diameters were measured by three operators using IMPAX software (AGFA). AP diameter is defined as the maximum distance between the occipital protuberance and the frontal bone. BP diameter is the maximum distance between the parietal eminences. CI is defined as BP/AP.

Results: Intra- and inter-observer variance was calculated to be <1% (Student's *t* test; $p < 0.0001$). AP and BP increase

steeply in the first 9 months of life. Subsequent monthly growth changes reduce to less than 1% thereafter.

Conclusion: There is a statistically significant difference between male and female CI in the first 24 months of life ($p < 0.0001$). All CI are outside the range of normality indicating the need to redefine the current accepted range for CI. The growth deceleration of AP and BP might have implications for the timing of craniofacial surgery.

Electromagnetic image guidance based complex posterior vault craniofacial remodeling procedures: our initial experience in Liverpool, UK

Ajay Sinha, Paul Sillifant, Sasha Burn, David Richardson, Christian Duncan (Alderhey Childrens' NHS Foundation Trust, Liverpool, UK)

Background: The availability of an intra-operative MRI in our unit has widened its application to complex craniofacial procedures.

Aim: This study aims to share our experience with electromagnetic (EM) image guidance in complex posterior skull remodeling and distraction procedures.

Materials and methods: All patients underwent a GA MRI the day of surgery. The image guidance was performed using a Treon plus (Medtronic) image guidance system. EM guidance helped us avoid a rigid head fixation. Guidance registration was done in supine position and then patients were turned prone in order to improve registration accuracy. Image guidance was used to mark the position of Torcula and transverse sinuses and to determine the limits of bony exposure while protecting major sinuses.

Results: A total of nine posterior skull remodeling/distraction procedures were performed. Four cases underwent a posterior vault distraction osteogenesis, three cases had total vault remodeling and two had re-do posterior vault remodeling. In two cases, foramen magnum decompression was also carried out and image guidance proved useful in mapping out the extent of bony decompression. There were no surgical complications.

Conclusions: Our technique of image-guided posterior vault remodeling procedure is a new technical development and increases the safety and efficacy of posterior vault procedures. The technical details will be discussed.

Development of a radiographic criteria for surgical assessment of outcome following posterior calvarial augmentation

Guirish Solanki, Desiderio Rodrigues, Martin Evans, Nicholas White, Hiroshi Nishikawa, Stephen Dover (Birmingham Children's Hospital, Birmingham, UK)

Aim of the study: Posterior calvarial surgery was introduced by our unit 27 years ago. The standard morphometric evaluation of the outcome of such surgery was to evaluate the increase in calvarial size, initially by X-rays and later using 3D CT scans. We report on the use of MRI for such evaluation and report a new set of criteria that may be helpful in improving consistent reporting of surgical outcomes.

Methods: Based on our experience with changes following surgery, we developed a set of 10 radiological criteria to evaluate surgical outcome. The criteria included CSF distribution in sulci, venous hypertension, tentorial angle, bowing of corpus callosum, ventriculomegaly, cervicomedullary kink, the "standing-up" cerebellum, CSF flow at CCJ, tonsillar descent and syrinx. We then applied it to a set of children that underwent posterior calvarial augmentation to evaluate outcomes and report the findings.

Results: Lambdoid synostosis (5) and pansynostosis (5) were most frequent followed by Crouzon (3), Aperts (3), Saethre-Chotzen (2), other (2). Median follow-up was 2 years. Thirteen cases had hindbrain hernia. All children improved symptomatically. Ninety-two percent had improved radiological criteria. Hindbrain hernia regressed in 75%. Regression time averaged 27 months. There was no progression to syrinx and no mortality.

Conclusions: Standardization was useful to assess calvarial augmentation outcome. Fixed posterior calvarial augmentation is effective in the management of raised ICP and leads to regression of the hindbrain hernia in craniosynostosis. It is now preferred over foramen magnum decompression for this group of patients.

Use of molding helmet as primary treatment of sagittal craniosynostosis

Sandeep Sood, Arlene Rozzelle, Steven D Ham (Children Hospital of Michigan, Wayne State University School of Medicine, Detroit, MI, USA)

Introduction: Sagittal craniosynostosis is traditionally considered a surgical condition. Poor results of simple suturectomy are from early reclosure of the suture. Post operative use of molding helmet has improved results. Since suturectomy reunites within 8–12 weeks of surgery, we questioned if the improved outcome was primarily related to use of helmet.

Methods: In nine patients, who opted for calvarial reconstruction at 4–6 months instead of endoscopic suturectomy, molding helmet was used with an intention to minimize compensatory changes in the interim. Patients underwent 3D CT scan to confirm craniosynostosis. Follow-up was at 4 weekly intervals for adjustment of helmet, head circum-

ference measurements, clinical photograph and Cranial Index (CI) measurement.

Results: Molding helmet was placed at a mean age of 2.8 ± 1.2 months. Overall, CI improved from $68.1 \pm 3.1\%$ at presentation to $75.1 \pm 3.4\%$ after a mean interval of 10.4 ± 5.7 weeks of helmet use. There was significant improvement in the head shape in seven patients. In two patients, lack of improvement related to poor helmet compliance with use of less than 10–12 h per day against our recommended 23 h. Three of the patients had an ICP monitoring and were found to have normal pressures. All patients had maintained the head circumference growth rate and developmental milestones during use of helmet.

Conclusions: These cases demonstrate that molding helmets improve head shape even without a suturectomy in patients with sagittal craniosynostosis challenging the traditional view.

Craniosynostosis—techniques and outcome:

an institutional study

Dwarakanath Srinivas¹, Girish Rao², Dhawal Shukla¹, Sampath Somanna¹, Indira devi¹, Chandramouli Ananthakrishna¹ (¹NIMHANS, Bangalore, India; ²RV Dental College, Bangalore, India)

Introduction: Craniosynostosis is an eminently correctable problem though oft-neglected in our country. In this article, we discuss the experience at our institute regarding the surgical management of craniosynostosis.

Material and methods: This retrospective study included 36 patients who underwent surgery over a period of 7 years. The patient's records were obtained from the medical records. The demographic profile, clinicoradiological features, various aspects of surgical management, outcome and long-term follow-up were analysed.

Results: There were a total of 36 patients (nine, sagittal suture; 18, coronal suture; two, metopic suture; seven, multisutural including one case of Aperts and two of Crouzon syndrome) operated at our institute. All patients with brachycephaly/plagiocephaly/syndromic craniosynostosis underwent fronto-orbital advancement. We used either titanium or bioresorbable plates. The average duration of surgery was 176 min and blood loss 268 ml. The average age of surgery was 9.8 months. Eight patients had dural tears which were repaired. There were no postoperative CSF leaks. One patient had delayed wound infection (after 1.6 years and was managed with antibiotics and dressings). There were no long-term complications. The average duration of follow-up was 2.6 years.

Conclusion: In this paper, we present our experience with craniosynostosis, review the surgical techniques used, and present operative videos and review long term follow-up and outcome.

An extreme case of aplasia cutis congenita in Adams Oliver syndrome

Mariel ter Laak-Poort, Rene van der Hulst, Erwin Cornips (MUMC+, Maastricht, Netherlands)

Adams–Oliver syndrome is a rare congenital condition involving a cutaneous and bony skull defects and limb abnormalities to a variable extent. We describe an extreme case with a very large skin, skull and dural defect involving the frontal, parietal and occipital regions. Extensive prenatal ultrasound investigation because of intrauterine growth retardation did not reveal structural abnormalities. The child was born a terme by C-section after an unsuccessful attempt at natural birth with forceps extraction. At birth, the large defect became apparent with only a very thin translucent membrane covering the brain. The child was bleeding profusely through an occipital parasagittal laceration caused by the forceps. The bleeding was controlled and the tear in the membrane repaired with a dural graft and microsutures.

The child was admitted to the neonatal intensive care unit and treated under strict aseptic conditions, in close collaboration between neurosurgeons, plastic surgeons, neurologists, neonatologists, and microbiologists. Long-term prophylactic antibiotics and low-dose acetazolamide were started. We noticed a sagittal sinus thrombosis without apparent venous infarction. Several subsequent tears of the membrane with brain herniation and CSF leakage occurred, which were carefully patched, microsutured, and covered with Tissucol and Tachoseal. The entire defect was covered with fatty gauzes and abundant Fucidin ointment. The gauzes were changed, and the wound rinsed every 2 days. Over the next 12 weeks, the defect rapidly shrunk and was covered with skin. We discuss our successful treatment strategy and multidisciplinary approach in this extreme case of aplasia cutis congenita.

Developmental analysis of skull base after fronto-orbital advancement in patients with unilateral frontal plagiocephaly

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Introduction: The role of skull base in the pathogenesis of craniosynostosis is still unknown. We analyzed skull base deformity in the patients with unilateral frontal plagiocephaly who underwent fronto-orbital advancement (FOA) during the last decade.

Methods: We assessed the treatment results and outcome of FOA performed in four patients, two girls and two boys younger than 2 years old, at Kagoshima University. Also, the basal cranium's angles were measured by 3D reconstruction images on computed tomography (CT) scan.

Results: Four patients with unilateral frontal plagiocephaly were treated with FOA one time each. Patients' ages at FOAs ranged from 9 to 23 months. The postoperative follow-up in all cases was uneventful, with good cosmetic results. On the preoperative 3D-CT images, there was clear synostosis of the affected coronal suture of each case. The ethmoidal axis was deviated with a mean angle of 12.6° (4–23°) to the affected side. The mean values of angles between the petrosal pyramids and the midline (posterior-petrosal-sagittal angle, PPSA) were 63.3° (60–66°) on the affected side and 59.0° (53–65°) on the normal side. The mean difference of PPSA was 5.8°. On the follow-up CT images 5 years after surgery, both the mean deviation of the ethmoidal axis and the mean difference of PPSA clearly decreased to 7° (4–12°) and 1.5° (0–2°), respectively.

Conclusion: The midline distortion of skull base should be considered to be spontaneously corrected for the follow-up periods in patients undergoing FOA for unilateral frontal plagiocephaly.

Dysraphism

"Tethered Cord Syndrome" our experience

Vivek Agrawal, Vernon Vélho, Deppak Palande, Rahul Mally (Grant Medical College and Sir J.J. Group of Hospitals, Mumbai, Maharashtra, India)

Introduction: Tethered spinal cord represents a clinical entity in the paediatric age group which has neurological morbidity, producing motor and sensory deficits in lower limbs, as well as bladder and bowel incontinence. The advances in imaging like CT scan and MRI have made visualization of the tethering and underlying pathology more evident. Early surgery allows complete neurological recovery.

Aims and objectives: This study aims (1) to evaluate the clinical profile of patients with tethered spinal cord and (2) to study the associated anomalies and co-relate the underlying pathology with the overall surgical outcome.

Materials and methods: Twenty-six patients were studied from 1994 to 2001. All patients underwent X-rays, MRI and nCTscan of spine. Detethering was done under general anaesthesia. Patients were followed up for 3–6 months.

Observations and results: Of the 26 patients in our study, 42% were males and 58% were females. Patients presenting at an early age (<1 year) had a lesser neurological deficit

than those presenting between 1 and 15 years of age. Common symptoms at presentation were neuro-orthopaedic deformity, gait abnormalities, bladder and bowel disturbances. The commonest cause of tethering was intradural lipoma, myelomeningocele and neural placode. Four patients had good bladder function recovery.

Conclusions: (1) At birth, children with a lumbosacral mass or cutaneous stigmata of occult spinal dysraphism should be dealt with a high index of suspicion despite a normal clinical examination. (2) MRI/CT imaging is essential to plan the surgical management. (3) Early surgical detethering of cord offers the best chance of complete neurological recovery.

The correlation between the degree of Chiari II in myelomeningocele patients and the clinical symptoms related to brainstem dysfunction

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Background: The anatomical complex of myelomeningocele (MMC) includes Chiari II malformation and hydrocephalus in most babies born with this severe defect. Chiari II malformation is very common at birth unless fetal surgery has been performed. It may cause symptomatic pressure on the brainstem or on low cranial nerves, resulting in breathing and swallowing difficulties and in mortality due to massive aspiration or apnea. Central apneas may be detected by performing sleep test, which can be pathological even in the absence of reported clinical symptoms.

Goals: This study aims to study the correlation between the Chiari level (according to MRI) and (1) the clinical symptoms of brainstem compression (including central breathing disorder according to a sleep test), (2) the level of the spinal defect (level of MMC), and the (3) presence of hydrocephalus.

Methods: The study was based on 66 children with MMC that underwent surgery in “Dana Children's Hospital” during the years 1998–2007. A detailed database of the patients was created for the research (Filemaker pro 8). Chiari II level was determined while reviewing MR images.

Results: A clear correlation was found between (1) *Chiari level* and brainstem compression clinical symptoms; (2) severity of Chiari II and presence of hydrocephalus. No correlation was found between the levels of Chiari and results of sleep test (central breathing disorder).

Conclusions: It is possible to identify patients at high risk to develop brainstem compression and hydrocephalus by defining severity of Chiari II by MRI. Early detection may lead to early successful treatment by decompression.

Spectrum of spinal dysraphism surgically treated at tertiary care hospital from Western India: a review of 250 cases in 3 years

Naresh Biyani, Uday Andar, Chandershekar Deopujari (BJ Wadia Children Hospital, Mumbai, India)

Spinal dysraphism is neural tube defect commonly seen in children if untreated have devastating neurologic deficit. We present spectrum of children that were surgically treated at a tertiary care hospital in Western India over period of 3 years.

Two hundred fifty children with spinal dysraphism were operated in 3 years. Clinical details, operative management and post operative follow-up was reviewed of all the children.

Results: Neurocutaneous stigmata was present in all the children with spinal dysraphism. About 40% of children presented with neurological deficit.

Conclusion: Spectrum of spinal dysraphism in tertiary care center is discussed. Prophylactic detethering surgery is recommended for all spinal dysraphism with tethered cord.

Myelomeningocele in premature babies

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It has been suggested that prenatal repair of myelomeningoceles (MMC) improves outcome. We describe the cases of two babies that were born premature and presented a MMC at birth not previously diagnosed.

S was born at 33 weeks gestational age due to delayed intrauterine growth. She presented a lumbosacral MMC that was operated on the first day of life. Her transfontanel ultrasonography showed no ventriculomegaly. She presented no complications due to her MMC repair but developed respiratory complications related to prematurity and one episode of sepsis. She was discharged 6 weeks after birth. At age 2, she is developmentally normal and has not developed hydrocephalus.

I was born at 28 weeks gestational age due to premature amniorrhexis. He presented a lumbar MMC that was operated on the second day of life. AVP shunt was inserted 8 weeks postnatally after his transfontanel ultrasonography showed increasing ventricles. He developed severe respiratory complications related to prematurity and was treated for one episode of sepsis and one episode of urinary tract infection. He was discharged 11 weeks after birth. His development is adequate for his age at 7 months.

Small size was not an issue for the repair of the MMCs. However, their hospital stay was significantly longer and

morbidity higher when compared to babies that were born and operated on at term. One of the babies did not develop hydrocephalus.

Non-operated myelomeningocele: case report and literature revision

Artur Henrique G B Da Cunha, Fernando Henrique M de Souza, Silvia N Bernardino (Hospital Da Restauração, Recife, Pernambuco, Brazil)

Spinal dysraphism remains one of the most frequent pathologies in our pediatric neurosurgery daily work. The early surgical approach remains a practice routine. Myelomeningocele (MMC) in an adult patient is a rare condition with only a few cases reported in the literature.

The authors present a 44-year-old male patient, suffering from not operated lumbar MMC. He was born in rural area and his family never sought medical care. The patient is paraplegic. Trophic disturbance in lower limbs and neurogenic bladder are present. He moves with aid of a wheel chair adapted to the presence of 50 cm circumference lumbar sac.

This case allows a rare opportunity to observe, discuss and revise about a possible natural evolution of a non-operated MMC.

Identifying environmental risk factors for human neural tube defects in the central region of Mexico

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Introduction: Congenital malformations represents an important cause of mortality and disability in many countries. The neural tube defects (NTD) are one of the most common birth defects. In Mexico, the prevalence myelomeningocele represents 1/1,000 of the live births. An environmental cause is any non-genetic factor that increases the risk of a birth defect for the exposed individual. Such factors include fetal infection, maternal illness, nutritional deficiencies, drug ingestion, chemical exposures, air pollution, radiation, etc.

Patients and methods: A retrospective review of the clinical, socioeconomic, and geographical factors of a series of patients with neural tube defects referred to the Hospital de Alta Especialidad del Bajío in Mexico, where they receive integral (pediatric, neurosurgical, orthopedic, and rehabilitation) management between June 2009 and January 2011.

Results: Fifteen newborns (11 with myelomeningocele and four with encephalocele) of the southeast region in the state of Guanajuato, Mexico were treated surgically. Young

mothers with irregular prenatal control with deficiency in the intake of supplementary preconceptional folic acid, live in an area with high exposure to environmental contaminants due to the industries located there.

Conclusions: Birth defects have a substantial public health impact on mortality, morbidity, disability, and to the cost of health care provision. This pilot study attempts to record for the first time in a region of central Mexico the relation between exposure to environmental risk factors (teratogenic chemical products) and the occurrence of neural tube defects.

Myelocystoceles: a retrospective study of 13 consecutive cases at All India Institute of Medical Sciences, New Delhi

Vivek Tandon, K Garg, Ashok Kumar Mahapatra (AIIMS, Ansari Nagar, New Delhi, India)

Objective: The study aims to analyse the presentation, treatment, surgical complications, and outcome in patients with myelocystocele.

Method: It was a retrospective analysis of 13 patients. All patients underwent neurological examination, plain X-ray films of the spine, computed tomography scans of the brain, and magnetic resonance (MR) imaging of the spine to confirm the diagnosis.

Result: The mean age of the patients was 22 months, 10 (76.9%) were males. All had swelling in the lower back and varying neurological deficits, weakness of lower limbs was the most common complaint in 12 (92.3%). All patients underwent excision of the meningocele sac and drainage of the syringocele with detethering of the spinal cord. Conus was low lying in 12 (92.3%), thickened filum was seen in 10 (76.9%), syrinx in four (30.8%), and Chiari malformation and hydrocephalus in two (15.4%) each. Post-operative CSF leak occurred in two (15.4%). During the last follow-up visit (range, 3 months–5 years), there was no change in the neurological status of these children, except one patient presented with increased neurological deficits; that patient had paraparesis with incontinence. During the follow-up period, no patient presented with retethering. **Conclusion:** Myelocystoceles are dissimilar from other skin-covered masses of spine. A proper imaging evaluation is required to differentiate myelocystoceles from other lesions in this area because the surgical treatment and prognosis may differ for these group of patients.

Postpartal and long-term clinical outcome after intrauterine myelomeningocele repair

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Introduction: Intrauterine myelomeningocele repair should minimize pathological features related with spina bifida syndrome (hydrocephalus, secondary tethered cord, Chiari malformation). Functional outcome and the necessity of repeated postpartal surgery is a target for improvement.

Methods: An ongoing uncontrolled prospective observational study was performed in two spina bifida centers independently from intrauterine surgery center. All newborn babies received intrauterine endoscopic patch closure in the second trimester. Postnatal follow-up results were recorded (radiological, clinical, surgery, and complications).

Results: Seventeen children after intrauterine surgery (2004–2010) were investigated. All were preterm births (GW, 22–36; mean GW, 29.6). Defect levels ranged between L1 and S3 (one level, 4; multilevel, 13). Three intrauterine repair complications occurred (patch defect, two; bleeding, one). All children developed hydrocephalus (shunt, 12) and Chiari malformation type II (tonsillar herniation in at least six). Secondary tethered cord syndrome (TCS) was obvious in 14 children. Primary postpartal surgery was necessary in nine patients (secondary wound closure, patch removal, and myelolysis). Secondary postpartal surgery was performed in 11 patients (shunt surgery, myelolysis, dermoid, Gardner decompression). One child (GW, 29) developed intraventricular hemorrhagia. Motoric and bladder function was pathological in more than 50%.

Conclusion: Although functional outcome is slightly better, hydrocephalus and Chiari malformation type II cannot be prevented in intrauterine MMC patch closure. Tonsillar herniation incidence seems to be lower, but without influencing HC development. Secondary TCS and frequency of subsequent complication surgery seems to be as high as in postpartal MMC repair. Preterm delivery and related complications could not be prevented so far.

Neural activity generated in the neural placode and nerve roots in the neonate with spina bifida

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Objective: This study aims to determine the neurophysiological capacity of the neural placode in spina bifida neonates. It also aims to determine if the spinal nerve roots in these neonates had normal stimulation thresholds and whether closure of the placode and associated defect affected these stimulation thresholds.

Methods: A case series of two neonates born with open neural tube defects who underwent neural tube closure within 24 h of birth. Neurophysiological monitoring and electrical stimulation of the placode and nerve roots was performed before and after closure of the neural tube.

Results: Stimulation of nerve roots results in evoked electromyographic responses in distinct muscle groups, indicative of the myotomal innervation pattern. Stimulation threshold did not change significantly with closure of the placode. Stimulation within the placode generated an alternating pattern of activity in the left and right legs.

Conclusions: Closure of the neural tube does not affect the stimulation threshold of the nerve roots, and they remain easily excitable. The viability of the nerve roots suggests that they may be candidates for neural prostheses in the future. The neural placode contains basic neural elements for generating a locomotor-like pattern in response to tonic neural inputs.

Complications of CSF shunts in children with myelomeningocele

Suresh Sankhla, Gulam M. Khan (Dr. B. Nanavati Hospital, Mumbai, Maharashtra, India)

Objective: Children with myelomeningocele (MMC) are believed to have a higher risk of shunt complications. In this retrospective study, we analysed the rate of shunt complications in patients with MMC who underwent shunt surgery concurrent with MMC surgery and compared it with those who had shunt placement as a separate procedure.

Methods: Outcome of shunt surgery was reviewed in 66 consecutive patients of MMC who were followed for more than 1 year after shunt placement. Shunt insertion was done prior to MMC surgery in 13 patients, concurrently with MMC surgery in 27, and after MMC surgery in 19 patients. In seven patients, shunt surgery was the only procedure performed.

Results: The overall rates of shunt infection and shunt malfunction were 17% and 36%, respectively. There was a high rate of shunt malfunction in those patients who were treated with cerebrospinal fluid shunting only. No significant difference in complication rates was observed in patients in whom the two procedures were performed concurrently and those who underwent separate operations. **Conclusions:** The order in which MMC repair and shunt placement were carried out did not have a significant effect on the rate of shunt complications. We therefore recommend that when indicated, shunt surgery should be done concurrently with MMC repair.

Low-lying spinal cord and tethered spinal cord syndrome in children with anorectal malformations

ATK Teo, BK Gan, WT Seow (KK Women's and Children's Hospital, Singapore, Singapore)

Aim: Anorectal malformations (ARM) and low-lying spinal cord (LLC), (conus >L2), have common embry-

onic origins and are commonly associated. The main radiological investigations to diagnose LLC are lumbar ultrasound (US) and MRI. LLC may result in tethered cord syndromes (TCS) and hence need early surgery. This study aimed to review the incidence of LLC in children with ARM, the presence of any urodynamic dysfunction (hence TCS) and their surgical outcomes.

Method: An institutional review board-approved retrospective study of children who underwent surgery for ARM from 2002 to 2009 at our institution. Patient records including radiological and urodynamic studies were reviewed.

Results: One hundred one patients underwent surgery for ARM. Of these, 17 (16.8%) had LLC. 12/17 (70.6%) had abnormal US and MRI findings. Five (29.4%) had normal US but abnormal MRI. In three of these five patients, MRIs were performed because of new symptoms; while two underwent MRIs because of equivocal US findings. These 17 patients subsequently underwent surgical detethering. Pre-operatively, 15/17 underwent UDS; of these, six UDS were abnormal. Post-operatively, abnormal UDS in three of six patients improved. None had new complications or deterioration. Twelve of these 17 LLC (70.6%) were intermediate or high ARMs.

Conclusion: In our study, 16.8% of children with ARM had LLC. Most ARMs were intermediate or high. Lumbar US is a useful first-line screen for LLC in ARM. Abnormal or equivocal US, onset of new symptoms require MRI. Surgical detethering can improve outcomes in TCS while prophylactic surgery in asymptomatic patients had very low surgical risk.

Secondary effects of spinal cord anomalies

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Occult congenital spinal cord anomalies can present with deformities of spine and limbs. These secondary effects have been found to be treated erroneously by repeated corrective surgeries of spine and foot ignoring the primary cause at the spinal cord.

From 14 June 2003 to 15 December 2010, 114 patients presented to Hospital and Rehabilitation Centre for Disabled Children with different deformities of foot and spine. Those with spina bifida manifesta were excluded from the series. On further investigation, they were detected to have spinal cord anomalies. Male/female ratio was 3:4 with age ranging from 3 months to 19 years. There were foot deformities in 45%, gait disturbance in 22%, and 33% had spinal deformities.

MRI showed tethered cord in 33%, 24% tethered cord with syringomyelia, 12% diastatomyelia, 23% Arnold Chiari malformation with syringomyelia and 8% Arnold Chiari malformation, syringomyelia and tethered cord. Posterior fossa decompression was done in 33%, tethered cord release in 76%, and syringotomy in 5%. Postoperatively, pseudomeningocele developed in 16% and increase in neurological deficit in 30% which continued to progress in three patients due to retethering. Correction of the spinal and foot deformities was undertaken subsequently by orthopaedic surgeons. There was mortality in three patients.

Refractory, resistant, or progressive foot and spinal deformities must always alert physicians to the possibility of occult spinal pathology. Surgery done timely prevents further neurological deterioration. A multidisciplinary effort for possible prevention, early detection, and management could reduce and prevent many morbidities in these patients.

Reconstruction of the skin defect in open myelomeningocele surgery

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Objective: Five-layer closure of open myelomeningocele is a well-established surgery. However, in terms of the final layer, skin defect, we need special consideration to close. Since there are factors to take account such as the size of the skin defect and the body weight of the child, also the associated malformation such as deformation of the spine in each case. In our institute, we have started to collaborate with plastic surgeons to close the skin defect in cases that was difficult to reconstruct. In this study, we have reviewed these cases to find out the benefit of the skin reconstruction method.

Materials and methods: A retrospective analysis was conducted with a chart review from 1999 to 2010. Sixteen patients were included in this study. Mean follow-up term was 5 years and 9 months.

Result: All 16 cases followed a satisfactory perioperative course. However, only one case suffered from pressure ulcer when the child was 1 year old because of his deformation of the spine. This child underwent another skin reconstruction.

Conclusion: Satisfactory outcome of a primary closure of open myelomeningocele could be established using V–Y skin flap method. Multidisciplinary collaboration at the moment of the primary closure is favorable to treat these children.

Case adapted operative strategies in children with Chiari malformation

Christoph Wiegand, Niels Sörensen, Wolfgang Wagner (Ped. Neurosurgery University Mainz, Mainz, Germany)

Chiari malformations during childhood represent an interdisciplinary challenge. Whereas Chiari I malformation can remain clinically mute for some time, Chiari II malformations might come up with a much more acute scenario, which requires prompt surgical intervention. Classical surgical technique like Gardner's still has its stand nowadays. In fact, many centers of pediatric neurosurgery rely on intraoperative ultrasound for posterior fossa decompression.

From August 2009 to August 2010, we operated on $n=7$ children harboring Chiari type I and $n=6$ type II. The youngest kid was 5 months, the oldest was 14 years. $N=3$ (Chiari I) were treated by third ventriculostomy endoscopy successfully alone. Children from the "usual" Chiari II group were operated by intraoperative ultrasound-guided decompression of the posterior fossa without opening the dura, incl. C1 laminectomy. All of these children showed a freed cerebrospinal liquid passage during ultrasound. The dura was left untouched in all patients. All Chiari II kids with bad prognosticators like bilateral vocal chords paresis lost their tracheal canula within 4 weeks after surgery and were on spontaneously breathing pattern. In addition to that, cerebellar symptoms were improved as well. The grading of cerebellar herniation was not significant for surgical technique or postoperative outcome.

Chiari malformations require a case adapted therapy at the right time. Postoperative results of the Chiari II group clearly indicate the importance of ultrasound during surgery combined with an intraoperative "step-by-step" strategy. They encourage ultrasound guided decompression of the posterior fossa, leaving the pediatric dura intact.

Spine

Spinal abnormalities in Down syndrome

Essam Alshail, Ahmed Alkhani (Department of Neurosciences, King Faisal Specialist Hospital and Research Centre, Al Faisal University, Riyadh, Saudi Arabia)

Introduction: Down syndrome, trisomy 21, is the most common autosomal trisomy. Birth prevalence continues to be around 1/1,000 live birth. Patients with Down syndrome have an increased risk of spinal abnormalities that may lead to neurological manifestations. These include congenital anomalies of the upper cervical spine like atlanto-axial abnormalities that may lead to cervical instabilities and possible sudden or progressive neurological deficits. We reviewed our experience in the surgical treatment of atlanto-axial instability in patients with Down syndrome treated at our institute.

Method: A retrospective chart review was carried out to all patients with Down syndrome who underwent surgical fusion for C1–C2 instability in our institute between 1990 and 2011. Clinical and surgical data were reviewed. Pre and post operative neuroimages were also reviewed when available.

Results: Eleven patients with Down syndrome phenotype were identified for the review. These include six females and five males. The mean age of the patients was 10.7 years (range from 3 to 25 years). Nine patients presented with severe clinical myelopathic features. The surgical procedures varied from C1–C2 or occiput-C2 fusion with sublaminar wiring and most recently occiput-C4 craniocervical fusion with rods, screws, and wiring. These data will be presented with a special focus on the surgical techniques and peri-operative care.

Conclusions: Early and aggressive surgical interventions are highly recommended for Down syndrome patients with cervical instability.

Congenital cervical spine anomalies presenting with deficit or deformity: analysis of 67 cases

Sandip Chatterjee, Agnivesh Tikoo (Park Clinic, Kolkata, India)

Introduction: Developmental anomalies of the cervical spine vary widely and often go unnoticed till they present with major neurological deficit or marked deformity. Identifying the lesion is important as prompt recognition often prevents disasters in later life.

Material and methods: This is a retrospective analysis of 67 cases with congenital anomalies of the cervical spine which presented to our unit in the last 10 years either with significant neurodeficit or deformity. They include three with malformations of the occipital condyles, occipitalisation of the atlas in 12 cases of which nine had atlanto-axial subluxations, three with atlas arch anomalies, 10 with basilar invagination, one with posterior arch anomaly of the axis, 20 with anomalies of the odontoid. In the subaxial spine, Klippel–Feil anomalies were found in 13 patients, with segmentation defects and scoliosis in five children.

Conclusion: Since these constitute a heterogeneous group of conditions, no treatment plan can be drawn but appreciation of the exact anatomy is the guide to planning of treatment. This will be illustrated with a few examples.

Awake fiberoptic intubation and perioperative anaesthesia management in congenital cerebrovertebral anomaly with fused C2 C3 posteriorly for transoral endoscopic odontoidectomy

Ranjana Das, Pratima Kothare (Bombay Hospital Institute of Medical Sciences, Mumbai, India)

Aim: This study aims to perform awake fibreoptic intubation as a safe option to prevent neurological deterioration during induction for anaesthesia and minimal neck movements in a,

Material and method: A 9-year-old child with atlantoaxial instability with CV anomaly and fused C2 C3 posteriorly with bonegraft was given Philadelphia collar.

Anticholinergic was given to reduce secretions. Awake fibreoptic endotracheal intubation under local anaesthesia without sedation was performed and a bite block in form of cut 20 cc syringe was used to protect the endoscope being bitten by awake child.

Observation and results: IV Glycopyrrolate reduced the oral secretions aiding in scope; 4% xylocaine nebulisation, 1 cc of 2% xylocaine injection transtracheal, successfully blocked the cough and gag reflexes, 10% xylocaine spray orally further reduced pharyngeal gag reflex. Syringe Biteblock was used to avoid biting of the scope.

Conclusion: Awake Fibreoptic Endotracheal Intubation is recommended in children for craniovertebral instability surgery. We report successful perioperative anaesthesia management in a child with complex CVJ Anomaly without neurological deterioration.

Extended endoscopic skull base approaches—endoscopic odontoidectomy

Vikram Karmarkar, Chandrashekhar Deopujari, Nishit Shah (Bombay Hospital Institute of Medical Sciences, Mumbai, India)

This objective of this study is for acceptance with extended skull base procedures being used for anterior, middle, and even posterior skull base pathology.

We describe our technique and experience with the less commonly used endoscopic odontoidectomy in a young patient with ventral cervico-medullary compression.

Occipitocervical and atlantoaxial fusion for OS odontoideum: case reports and review of the literature

Willard Kasoff, Alyssa Zupon, Michael Diluna (Yale University School of Medicine, New Haven, CT, USA)

Purpose: This study aims to present our recent experience with OS odontoideum and to review the current surgical management of pediatric craniovertebral junction abnormalities.

Methods: We reviewed our operative cases of pediatric craniocervical fusion over the preceding 12 months. A MEDLINE search for recent surgical literature was performed and reviewed.

Results: Two cases of OS odontoideum were included. The first, a 13-year-old male with severe spinal cord compres-

sion at C1, underwent decompression and occipitocervical fixation using C1 lateral mass screws and C2 translaminar screws. At 8-month follow-up, he had excellent fusion with preserved cervical range of motion (ROM). The second patient, a 4-year-old female atlantoaxial instability, underwent posterior atlantoaxial fixation with C1 lateral mass screws and C2 translaminar screws. Five months later, she had also achieved excellent fusion with preserved ROM.

Review of the literature demonstrated growing use of instrumentation in the pediatric cervical spine at experienced centers, with excellent outcomes and low complication rates.

Discussion: Rigid fixation of the craniovertebral junction was achieved, allowing bony fusion and treatment of the underlying pathology. The relative mobility of the pediatric spine resulted in compensatory preservation of near-normal ROM in both cases.

Conclusion: Instrumented fusion of the pediatric craniocervical junction is becoming increasingly accepted and is being safely performed in increasingly young patients. Careful preoperative planning and imaging review are critical for complication avoidance. When fusion is obtained, ROM preservation is far greater than that in the adult population. Careful follow-up data are needed to assess long-term outcomes.

Morphometric characterization of the cervical spine and spinal cord in children with MPS IVA (Morquio–Brailsford) disease

William Biu Lo, Giovanna Paternoster, S Vijay, A Chakrapani, Guirish Solanki (Birmingham Children's Hospital, Birmingham, UK)

Introduction and aims: Morquio–Brailsford disease (MPS IVA) is an autosomal recessive disorder with abnormal glycosaminoglycan metabolism causing spinal deformities including odontoid dysplasia, kyphosis and scoliosis, with associated spinal cord instability and/or compression. MRI morphometric measurement of the spine in these children may differ significantly from normal. We aim to quantitatively characterize these radiological findings.

Methods: Spine MRI of 17 (nine boys, eight girls) affected children were reviewed. DICOM axial and sagittal images in T1-weighted sequence were studied. The anteroposterior (AP) distance of the vertebral bodies and spinal canal, cross-sectional area of spinal cord and canal at every segment were measured. The cervical spinal canal–body ratio (CBR or Torg) and volume were calculated.

Results: The median age at time of MRI was 6 years (range, 1–17 years). Mean AP canal diameters were: C1=8.3 mm, C2=8.5 mm, C3=8.6 mm, C4=8.8 mm, C5=10 mm, C6=10.7 mm, and C7=10.2 mm. The median CBRs were: C2=0.83, C3=0.80, C4=0.81, C5=0.93, C6=0.97, and C7=

0.85. The median cross-sectional areas for CSF space were: C2=66.2 mm², C3=56.5 mm², C4=46.2 mm², C5=55.8 mm², C6=68.4 mm², and C7=68.9 mm². The estimated mean and median cervical spinal cord volumes were of 3,481 and 3,685 mm³ (range, 2,187–4,457mm³). Median cord–canal ratios were C1=0.45, C2=0.40, C3=0.55, C4=0.51, C5=0.45, C6=0.45, and C7=0.50.

Conclusions: MPS IVA children have lower than normal Torg ratio suggesting upper cervical canal stenosis. The cord/canal ratio and AP diameter were smaller at C1–C2. This preliminary data suggests that the Morquio child has an "inverted funnel-shaped" cervical spine with narrowing of cervico-thoracic region, the opposite of a normal cervical spine. These findings may have prognostic significance and may help determine a pattern of progression.

Oncology

Ependymoma in children: surgical treatment, technological advances and outcomes over a 55-year period in North West England

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Purpose: Surgical resection has always been the mainstay management for childhood ependymoma. We aimed to evaluate the influence of various prognostic factors on survival outcomes over a 55-year time period.

Study design: We retrospectively reviewed the medical records of 129 patients diagnosed with ependymoma between 1954 and 2008. Patients were stratified into five 11-year time periods according to their date of diagnosis, with age at presentation ranging from 3.5 months–15 years (mean age, 5.5 years). The male/female ratio was 0.9:1. Kaplan–Meier statistical analysis, the log-rank test and a multivariable analysis assessed survival differences between prognostic groups.

Results: The 5-year overall survival (OS) rate steadily improved from 20% in 1954–1964 to 71% in 1998–2008 ($p<0.001$). The trend in 5-year progression free survival (PFS) differed; PFS for 1954–1997 was less than 30%, and then rapidly improved to 51% for the 1998–2008 time period ($p=0.009$). The extent of surgical resection was the most significant prognostic factor ($p<0.001$); gross total resection rates followed a similar trend to PFS, with rates of 33% and 56%, respectively, for the latter time periods. Age and tumour site were also key prognostic factors ($p<0.001$). Patients aged over 10 years achieved a 65% 5-year OS rate, compared to those aged 3–10 years (OS=39%) and those under 3 years (OS=14%).

Conclusion: Survival outcomes for childhood ependymoma are improving. For this long-term follow-up cohort, surgical resection followed by age and site of lesion had the most significant impact on survival.

3 Tesla intraoperative MR evaluation of paediatric pilocytic astrocytoma resection

Shivaram Avula, Laurence Abernethy, Barry Pizer, Dawn Williams, Conor Mallucci (Alder Hey Children's Hospital NHS Foundation Trust, Liverpool, UK)

Background: The value of intraoperative MRI (ioMRI) in adult brain tumour resection is gaining recognition. There is limited documentation of its efficacy in children. In our initial experience, we have often encountered challenges in the evaluation of ioMRI in children with pilocytic astrocytomas (PA), a tumour that is less common in adults.

Aim: To present the intraoperative findings in children who underwent ioMRI during PA resection, its influence on the surgical management and follow-up.

Method: IoMRI was performed in 14 children with pilocytic astrocytomas using a 3 T MR scanner between November 2009 and December 2010. The scans were reviewed with the clinical records and follow-up imaging.

Results: Among the 14 children, the surgical aim was complete resection in ten and partial resection in four. Among the complete resection group, the surgical aim was achieved in five children at first attempt. Residual tumour was identified on ioMRI in three children, two of who underwent further resection achieving complete tumour resection. In two children, the ioMRI findings were equivocal for residual tumour and no further resection was performed. Among two children in the partial resection group, surgery was extended after ioMRI. Follow-up imaging revealed tumour recurrence in one child and reduction in enhancement in patients with equivocal findings and one patient with residual disease.

Conclusion: Surgical strategy was altered in four out of 14 patients with PA following ioMRI. Post-surgical contrast enhancement, sometimes compounded by brain shift can pose challenges and collaborative imaging analysis by the radiologist and neurosurgeon is helpful.

Short term surgical outcome of pediatric craniopharyngiomas

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Craniopharyngiomas as tumors still remains an enigma. This is a prospective study to evaluate the surgical outcome in pediatric craniopharyngiomas. Twenty-four patients who underwent surgery between 2007 and 2010 were included

and recurrent ones were excluded. The age range was 1–20 years. Headache was present in 74% of patients and along with vomiting in 54%. Vision was near normal in 37%, 4% had no useful vision. Diabetes insipidus was noted in 25% of patients. Of the patients, 58% had no significant hormonal abnormality. Focal neurological deficits were present in 17% and 29% of the patients were functionally independent. Thirty percent of the patients had mild to moderate hypothalamic disturbances. CSF diversion was done in eight patients. Subfrontal and transsylvian approach were commonly used. Transphenoidal approach was used in one patient. Retromastoid suboccipital craniectomy was required in one patient. Gross total resection was done in 33% and subtotal resection in 67% of patients. Post-operatively, DI and electrolyte imbalance were the common problems and seven patients required ventilator support. Four patients expired in the post-operative period. All tumors were adamantinomatous craniopharyngiomas. Mean follow-up period was 17.4 months. Visual improvement noted in 50% of patients and 62% were on hormonal supplementation. Increased BMI was noted in 32% of patients. Of the patients, 55% were functionally independent at the time of last follow up. Data was analyzed using SPSS software [13]. In conclusion, this study reveals that raised ICP, hypothalamic involvement, vertical extension of tumor is significantly associated with surgical morbidity and mortality.

Long-term outcome of focal brain stem gliomas of childhood

Paul Klimo¹, Frederick Boop¹, Greg Armstrong¹, Atman Pai¹, Alex Chavez¹, Robert Sanford¹ (¹St Jude Children's Research Hospital, Memphis, TN, USA; ²LeBonheur Children's Hospital, Memphis, TN, USA)

Focal brain stem gliomas (FBSG) are an uncommon entity, presumed to have a good prognosis compared to diffuse brain stem tumors. The authors submit a single institution retrospective study of 66 biopsy proven focal brain stem tumors treated between 1985 and 2010, the largest study of its type to date.

Patients presented with hydrocephalus, cranial nerve palsies or hemiparesis. Cerebellar signs were unusual. Histology was consistent with juvenile pilocytic astrocytoma in two thirds of patients and astrocytoma (NOS) in another one third.

Tumors were categorized as mesencephalic, pontine or medullary, intrinsic or exophytic. Some were treated with biopsy and irradiation, others with resection.

Analysis of subgroups demonstrated that the majority of children enjoyed an excellent quality of life with over 85% 5-year survival.

Conclusion: FBSG are a benign group of tumors in which gross total resection may be curative but partial resection is

also associated with excellent long-term survival. Radiation therapy is very effective in those which progress. Therefore, the surgeon must be able to offer curative surgery with minimal morbidity or else consider very limited surgery followed by adjuvant therapy and not risk injuring the child

Anterior fontanelle tumors: surgical results in a 10 year series

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Anterior fontanelle masses are benign lesions usually detected during the first 2 years of life when they are evident due to progressive growth, the lack of ossification of the anterior fontanelle and the scarcity of hair in this region. In this 10-year series, we describe 21 cases of anterior fontanelle tumors, with 20 dermoid cysts and one lipoma. Our series describe 13 females and eight males with ages ranging from 2 months to 10 years with an average age of 21.7 months. The clinical presentation included progressive enlargement of a painless mass over the fontanelle with sizes ranging from 2 to 4 cm in diameter and one late referral case who had repeated scalp infections in an ossified fontanelle during a 10-year period. Surgery provided total removal of these dermoid cysts and lipoma with no complications and no recurrences. This pathological entity has been underreported in the literature and was previously described only in population of African origin.

Surgical outcome in 65 consecutive cases of pediatric craniopharyngiomas: an evaluation of fronto-temporal basal approach

Aadil Chagla, Sandeep Patil (King Edward Memorial Hospital, Mumbai, Maharashtra, India)

Objective: Craniopharyngiomas arise from the remnants of Rathke's pouch, which are critically located in the region of the tuber cinereum. The aim of this study is to evaluate the versatility and safety of the Fronto-temporal basal approach to achieve radical excision.

Methods: Sixty-five consecutive cases of craniopharyngiomas were treated surgically over the past 12 years. All cases underwent enhanced computerized tomography/magnetic resonance imaging. The symptoms were headache/vomiting in 58 patients, 12 were totally blind, eight were blind in one eye, 37 had visual impairment; 25 cases had endocrinological manifestations, eight suffered generalized tonic-clonic seizures; five were in altered sensorium. All patients underwent a wide fronto-temporal craniotomy with a basal surgical approach to the tumor.

Results: Eleven patients underwent total excision, 23 near-total excisions (mortality, three) and 24 subtotal excisions (mortality, two); seven underwent partial excision. Thirty-one patients with visual impairment improved, five remained same and one worsened. Twenty-nine patients suffered from temporary diabetes insipidus and two cases needed long-term hormonal replacement. Thirty-two received post-operative radiotherapy. Six recurrences were seen till date and only one within 4 years. No patient underwent any post-operative ventricular drainage.

Conclusion: The wide craniotomy allows access to all corridors and lateral sylvian fissure to achieve radical safe excision. Rock hard calcification is a deterrent to total excision. Though total excision is the goal, care must be taken to prevent damage to the perforators and the pituitary stalk at all cost. Hormonal replacement is an expensive proposition and not suitable for our patients.

Antro-lateral mini fronto-orbito zygomatic (MFOZ) craniotomy via an eyebrow incision in pediatrics. Technical notes and evaluation

Hassan El-Shafei (Cairo University, Cairo, Egypt)

This study aims to describe and retrospectively analyze and evaluate the anterolateral mini fronto-orbito-zygomatic (MFOZ) approach via an eyebrow incision in treating pediatric pathologies especially those related to the sellar region.

Method: Between January 2003 and December 2008, 18 patients with lesions in the sellar region or the anterior corridor leading to it were operated upon via the same approach. There were 10 males and eight females. The age ranged between 11 months and 15 years (mean, 7.9 years). The pathologies were craniopharyngiomas in six cases, hypothalamic and chiasmal pilocytic astrocytoma in seven, and five miscellaneous cases including two pituitary adenomas, one extradural hematomas, hypothalamic hamartoma and one arachnoid cyst in the retrosellar and prepontine area. Surgery was performed from the right side in 15 cases and from the left in three cases.

Results: Total resection was achieved in nine cases (50%), subtotal resection in four cases (22%), and partial in five cases (28%). Complications related to the approach like CSF rhinorrhea, supra-orbital hyposthesia and loss of upward elevation of the eyebrow were temporary. There were no mortality in this study and no significant added morbidities related to the approach. The approach gathers the advantages of the small cosmetic eye brow incision and the skull base trajectory thus reducing brain retraction and achieving the desired exposure with good outcome.

Conclusion: The MFOZ craniotomy using an eyebrow incision in pediatric patients is safe, effective, and both suitable and convenient for treating lesions especially at the sellar region.

Juvenile xanthogranuloma supra and infra tentorial (non-Langerhans cell histiocytoses). What we know, what we can and what we must do. A new case

Juan Bosco Gonzalez (Hospital Infantil Manuel de Jesus Rivera, La Mascota, Managua, Nicaragua)

The non-Langerhans cells histiocytosis (non-LCH) is a rare group of histiocytic disorders in which the most common is the juvenile xanthogranuloma (JXG), this is a tumor that commonly affects the skin but can sometimes occur within the intracranial compartment, solitary or multiple, This location is extremely rare, perhaps fewer than 20 reported cases; for this reason, there are controversies in the management of this intracranial lesions, not having a protocol or centers that have the expertise to create it, but the backlog of cases worldwide will provide us with knowledge to improve treatment options for that is considered important to contribute to the provision of a new case, the surgical experience, the clinical course of patients and adjuvant therapies as well as with the review of the available information regarding this disease when it has implications in neurosurgery

Intracranial teratomas in children: a series of ten cases

Nishant Goyal, Pankaj Singh, Mehar Sharma, Ashok Mahapatra (All India Institute of Medical Sciences, New Delhi, India)

Background: Intracranial teratoma is a rare entity, accounting for 0.5% of all intracranial tumors and 2–4% of intracranial tumors in children.

Objective: This study aims to study the demographic profile, clinico-pathological features and outcome of pediatric intracranial teratomas.

Methods: All cases of central nervous system (CNS) teratoma diagnosed over a 10-year period (2001–2010) were retrieved from records of the Departments of Neurosurgery and Neuropathology. Out of 37 cases of CNS teratomas, eight were adults and 29 were children. Of the pediatric cases, 19 were spinal and ten were intracranial in location. Patients' demographic profile, clinical and radiological features; and intra-operative findings were recorded.

Results: Ten cases of pediatric intracranial teratoma were identified (six males, four females). Seven occurred in the first decade and three in the second decade (age range, 3 days–16 years). Third ventricle was the most common site (five cases); these presented with features of raised intracranial pressure. Three cases were seen in association with occipital encephalocele. All patients underwent sur-

gery except for a newborn, who died 3 days after birth and was autopsied. Intra-operatively, most of the tumors were solid–cystic. A tooth and hair were identified in one case each. Histologically, seven cases were classified as mature teratomas and three as immature teratomas.

Conclusion: A diagnosis of teratoma should be considered in young children presenting with a solid–cystic tumor in the region of the third ventricle. In view of the co-existence of teratoma with occipital encephalocele, thorough histopathological sampling of the excised sac and contents is advised.

Tractography and neuronavigation in a posterior fossa residual ependymoma

Nelci Zanon, Giselle Coelho, Marcia Noriko Oliveira Homa, Graciele Prianti, Silvana Brito, Rita Delgado Villora (Hospital Beneficência Portuguesa, São Paulo, Brazil)

This study aims to preserve function in a posterior fossa reoperation, we describe the diffusion tensor image (DTI) and neuronavigation. A 12-year-old-boy had 15 days of headache and vomiting, followed by diplopia. Clinically was detected bilateral papilledema. The MRI findings show a huge posterior fossa tumor with supratentorial hydrocephalus. The lateral displacement of the brainstem was evident, the obex and cervical canal was filled by tumor until C3. The surgeon impression was a total resection. The CT scan next day shows a residual hematoma in the posterior fossa. The external ventricular drain was maintained for 14 days and he does not need other treatment for hydrocephalus. The MRI, after the hematoma absorption shows a residual tumor in the superior medullary velum. In the reoperation, the neuronavigation and tractography was used to minimize lesion. The tractography was performed by fiber tracking software (Brainlab company). The choice of the tracts was made based on region of interest, in this case fossa posterior tracts like middle cerebellar peduncle and superior medullary velum. The selection of the correct tract is based on anisotropy and length of fiber parameters. To prevent shift, DTI evaluation during surgical procedure, the intraoperative acquisitions was updated. The surgical approach was telovelotonsilar and there was not any kind of shift detectable. The residual lesion localization was precise and accurated with tractography and neuronavigation. The complete resection was possible and the neurological examination of the patient was maintained. The anathomopatology confirmed ependimoma OMS grade II.

The possible effects of stem cell implantation on damaged hypoglossal motoneurons nucleus in rat

Mehdi Jalali, Mohammad Reza Nikravesh, Ali Reza Fazel, Daruosh Hamidi, Masoomah Seghatoleslam, Shahin Saeedinejat (Mashhad University of Medical Sciences, Mashhad, Iran)

Neuronal loss such as ischemia, traumatic brain injury and Alzheimer disease is a major pathological outcome of many common neurological disorders which affect the brain. Investigation have been shown that although adult mammalian brain contains neural stem cells with self-renewal and multilineage potential in the hippocampus and subventricular zone, neurogenesis from these areas does not compensate for neuronal loss in age-related neurodegenerative. The aim of the present study was to investigate whether stem cells could be transplanted in the rat hypoglossal nucleus to improve neurologic disabilities.

In the present study, right hypoglossal nerve of 1-month-old Wistar rats were either injured or sham operated and nor nerve injury occurred. Four days later, all groups received labeled human umbilical cord blood stem cells (HUCBSc) via tail vein injection. One month after injection, cresyl violet and immunohistochemistry study as well as behavioral test were carried out.

Our immunohistochemical evaluations after transplantation revealed that transplanted (HUCBSc) were detected in the lesioned area. Our finding also indicated that there was significant difference in the amount of (HUCBSc) transplants between the lesioned, nonlesioned and control groups. This data indicate that HUCBc are enriched for stem cells that have the potential to initiate and maintain tissue repair. This potential is especially attractive in neural diseases for which no current cure is available.

Two cases report of brainstem pilomyxoid astrocytoma Dongwon Kim (Keimyung University, Deagu, Republic of Korea)

Objective: A pilomyxoid astrocytoma (PmA) is considered to be either a more aggressive variant of a pediatric pilocytic astrocytoma (PA) or a tumor of a separate entity. Brainstem lesions are rarely amenable to gross total resection and are associated with high operative risks. We present two cases of pediatric brainstem PmA.

Method and result (case report): We report one female and one male pediatric patient. The first patient presented at age 8 with facial asymmetry and progressive right-side motor weakness. A biopsy and cyst aspiration was made stereotactically. Although she received a radical course of radiotherapy, the lesion regrew rapidly, necessitating a second operation. After completion of chemotherapy, the patient achieved a clinical and radiological response which was maintained during the 4-year follow-up period. The second patient presented at age 5 with left eye ptosis and left hemiparesis. He underwent stereotactic biopsy and cystic aspiration and demonstrated complete response to chemotherapy for 3 years.

Conclusion: The optimum management of PmA still remains controversial. Our report indicates that tissue diagnosis and chemotherapy is good option for management of midbrain PmA. The author reviews the literature on PmA and discuss the specific issues associated with the update management.

Key words: astrocytoma, pilomyxoid, midbrain, pediatric, chemotherapy

Pediatric brain tumours—Oman experience

Santosh Lad (khoula Hospital, Muscat, Oman)

The Sultanate of Oman has achieved great success in primary health care over the last 30 years. Due to rapid socioeconomic development of the country, changes in lifestyles have also increased. Pediatric population contributes significantly to this overall population. In this presentation, the pediatric age group has been taken to include age 0–12 years. A review of the cases of pediatric brain tumour cases from 1985 till 2008 has been done. I will discuss the evolution of various facilities like investigations, operation theatres and other paramedical facilities that have occurred from 1985 till 2008.

In this study, we could find total cases of 259 from the available data from 1985 till 2008. Male patients were 153 and female 106. In this presentation, initially patients were diagnosed with CT scan, later with MRI and DSA. Initially, we did VP shunts but with the availability of endoscope, we did third ventriculostomy. As the facilities for dedicated operation theatres with dedicated ICU were created, more complex surgeries were undertaken. Also, there was a rapid increase in the number of surgeries as investigative facilities increased in the peripheral hospitals. With the advent of stereotaxic and neuronavigation facilities, we are now able to undertake major and complex pediatric brain tumour surgery. Some illustrative cases will be referred to in this lecture. The location of these tumours as well as the histological types of the tumours will be discussed in this lecture.

Abducens nerve schwannoma in an 8-year-old boy—case report

Muh-Lii Liang, Sanford PC Hsu, Hsin-Hung Chen, Tai-Tong Wong (Taipei Veterans General Hospital, Taipei, Taiwan)

Schwannomas of abducens nerve are extremely rare tumors and only 20 cases had ever been reported. The case report is an 8-year-old boy with a symptom of diplopia on the right size horizontal gaze for 3 months. The neurological examination showed solitary sixth nerve palsy of the right eye without paresis of other cranial nerve. The images study disclosed a well-defined spherical tumor mass about 2.6 cm in size located on right petro-clival junction. The mass caused lateral

displacement of trigeminal nerve and obliteration of abducens nerve, and mild compression of right ventral pons. The initial differential diagnosis included meningioma, hemangiopericytoma and neurogenic tumors. We performed a pretemporal and subtemporal approach and identify the tumor originated from the cisternal portion of abducens nerve attaching on the dura layer of cavernous sinus. Near-total removal of tumor and preservation of abducens nerve were achieved. The final pathology reported schwannoma.

After operation, the patient had transient numbness of right face but the symptom relieved after 1 month. The abducent palsy of right eye had not yet recovered in 1 month post-operatively. Therefore, the case is the youngest one till now who sustained of the typical symptoms of abducent palsy and had been proven surgically and pathologically. Even though it is rare, the differential diagnosis for tumor locating on this area should be included.

Pituitary adenomas and endoscopic transphenoidal surgery

Luca Massimi¹, Viola Custodi², Mario Rigante¹, Paolo Castelnovo³, Massimo Caldarelli¹, Gianpiero Tamburrini¹, Davide Locatelli², Concezio Di Rocco¹ (¹Catholic University, Rome, Italy; ²San Matteo Hospital, Pavia, Italy; ³University Of Insubria, Varese, Italy)

Introduction: Pituitary adenomas and endoscopic transphenoidal approach represent an unusual association in pediatric neurosurgery. Pituitary tumors are actually uncommon and transphenoidal surgery is rarely performed in children for anatomical and pathological reasons (big tumors versus small anatomical pathways).

Materials and methods: Herein, we present a pediatric series composed by 16 children affected by pituitary adenomas operated on in the last 10 years. The mean age at surgery was 14.2 years. Hypopituitarism and hormone hypersecretion were detected in 11 and eight subjects, respectively. An endoscopic trans-nasal parasseptal approach was carried out in all cases through the four-hands-two nostrils technique using a dedicated 18-cm-long rigid endoscope (2.7 and 4 mm diameter).

Results: All patients are alive after a 6.3 years mean follow-up (range, 2–11 years). Gross total resection of the adenoma was possible in all but one case (93.5%). However, three recurrences were noticed during follow-up; all of them were successfully managed by redo endoscopy. Hypopituitarism improved in six cases (55%) and was stable in the remaining five. Transient insipid diabetes occurred in four patients. CSF leakage was the main complication (18.7%); no neurological or visual morbidity was noticed.

Conclusion: Pituitary adenomas remain rare tumors in children where they are mainly burdened by endocrinolog-

ical sequelae. In our opinion, endoscopic surgery is the safest and most effective way to approach these neoplasms.

Choroid plexus tumors in children

Hamilton Matushita, Daniel Cardeal, Adriana Espindola, Umbertina Reed, Manoel Jacobsen (São Paulo University, São Paulo/SP, Brazil)

Background: According to previous review of 1,195 pediatric tumors of the nervous system from our institution, reported by Rosemberg et al in 2005, choroid plexus tumors constituted 3% of the tumor.

Objectives: This study aims to evaluate the clinical presentation and results of treatment of 34 children with choroid plexus tumors treated consecutively from 1985 to 2010.

Methods: Thirty-four cases of choroid plexus tumor in children were analyzed retrospectively. The mean age at admission was 31.9 months. Males were slightly more affected (19/15). Topographic distribution showed: lateral ventricles 26, III ventricle 4, and IV ventricle 4. All patients presented with signs and symptoms of increased intracranial pressure, except one, that presented with convulsion. Impaired level of consciousness was observed in five children including one which was admitted in coma due to bleeding of the tumor. Pathology examination demonstrated 20 papillomas and 14 carcinomas. Radical removal was obtained in all papillomas and in 10 of 14 carcinomas. Two deaths occurred in patients with carcinoma during admission: one which had previous intraventricular hemorrhage and other related to massive intraoperative bleeding. The mean follow-up was 6.5 years. The long-term follow-up demonstrated that 18 of 20 papillomas and only six of 14 carcinomas were alive. Apparently, the survival of some of the carcinomas was not related to the adjuvant therapy. Recurrence and CSF dissemination of the tumors were the main cause of fatality of the cases.

Conclusions: Prompt diagnosis and gross total resection of choroid plexus tumors in children may be the main prognostic factors.

Histopathological variety of a selective pediatric tumor population

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Objective: Does the pediatric tumor spectrum of a general neurosurgical department differ from a specialized pediatric neurosurgical service?

Methods: Retrospective single center study with evaluation of consecutive tumor cases.

Results: Since 2007, 104 consecutive patients with CNS tumors have been treated surgically in a pediatric neurosurgical unit of a maximum care pediatric hospital. Fifteen suffered from spinal tumors and three had craniospinal involvement. The 89 pediatric patients had a mean age of 8 years, the six adults of 35 years. Most frequent histologies were pilocytic astrocytoma I in 23 (3 spinal, 20 cranial; 22%), anaplastic astrocytoma III in 6 and glioblastoma IV in six cases. Medulloblastoma was found in seven cases, myxopapillary ependymoma of the cauda/filum in six. Furthermore, an unusual series of rare tumors was found: atypical choroid plexus papilloma (six), cranio-pharyngeoma (three), ATRT (three), subependymal giant cell astrocytoma (three), germinoma (two), choroid plexus papilloma (two), pilomyxoid astrocytoma (two), angiocentric glioma, anaplastic pilocytic astrocytoma III, atypical neurocytoma in a child, rosette forming glioneuronal tumor, primarily metastasising spinal anaplastic ependymoma III, pineocytoma and pineoblastoma. Beside the relatively high rate of rare pathologies, also tumor localization was unusual compared to a statistical tumor population: 53 tumors were located in the midline or intraventricularly, 12 of them were intrathalamic. Only 21 tumors were located in the posterior fossa.

Conclusions: This selective patient series of a dedicated pediatric neurosurgical service shows different tumor entities compared to series from general neurosurgical departments. A dedicated pediatric service reflects a more elective tumor series.

Treatment of optic pathway hypothalamic gliomas (OPHG): experience with 12 cases in Tokyo

Hideki Ogiwara, Nobuhito Morota (National Center for Child Health and Development, Tokyo, Japan)

Objective: This study aims to present our 7-year experience (from 2004 to 2011) in the treatment of optic pathway/hypothalamic gliomas (OPHG).

Methods: Retrospective review of 12 children treated for an OPHG was performed.

Results: Mean age was 82.4 months. Nine patients had newly diagnosed tumors, two had recurrent tumors. One had evidence of neurofibromatosis-1. Partial resection was performed in five and gross total resection was performed in one. Endoscopic transventricular biopsy was performed in five patients. Stereotactic ventricular drainage was used for facilitating the biopsy for three patients without ventriculomegaly. Postoperatively, none of the patients deteriorated neurologically. Histopathological studies revealed nine pilocytic astrocytoma (grade I), two diffuse astrocytoma

(grade II), and one pilomyxoid astrocytoma (grade II). Chemotherapy was performed in nine patients. Local radiation therapy was performed in one (total, 45 Gy). The mean follow-up was 31.4 months. None of the patients died. Two patients progressed 27 and 12 months after the original surgery (one without any adjuvant therapy and one with chemotherapy). The second surgery (partial resection) was performed for one patient and had been stable for 36 months since then. The other tumor stopped progression and had been stable for 41 months with chemotherapy. Nine patients underwent chemotherapy, of which eight patients (88.9%) did not show progression (four decreased in size and four stable) with a mean follow up of 28.3 months (range, 4–74 months).

Conclusion: Progression of OPHGs is relatively slow and chemotherapy is considered to be useful for the control of these tumors.

Magnetic resonance spectroscopy profiles of pediatric mixed glioneuronal tumors

Joffre Olaya, Mark Krieger, Yasser Jeelani, Mark Fedor, Brian Lee, J. Gordon McComb, Marvin Nelson, Stefan Bluml (Childrens Hospital Los Angeles, Los Angeles, CA, USA)

Introduction: Mixed glioneuronal tumors are rare, slow growing tumors seen most commonly in children. In vivo MR spectroscopy characterizes tissue at a cellular level by quantifying intracellular metabolites. The goal of this study was to review the metabolic features of these tumors.

Methods: A retrospective review of the MR spectroscopy database at Children's Hospital Los Angeles was performed for a 9-year period (2001–2010). All MR spectra were acquired with a standard single-voxel PRESS sequence with an echo time of 35 ms. Metabolite levels were compared with absolute levels in nontumoral regions, normal controls, and other pediatric brain tumors.

Results: Four children with histologically confirmed mixed glioneuronal tumors underwent MR spectroscopy prior to any intervention. Spectra showed features typically seen in other tumors, such as elevated choline (Cho) and lipids. However, an unusually high *N*-acetyl-aspartate (NAA) peak was also observed in all patients with mixed glioneuronal tumors, distinguishing from all other tumors. The NAA to Cho (NAA/Cho) was 2.1 ± 0.5 in mixed glioneuronal tumors vs. 0.6 ± 0.6 ($p < 0.001$) in 60 other pediatric tumors.

Conclusions: Mixed glioneuronal tumors have a unique MRS pattern with elevated NAA, in addition to elevated Cho. NAA is present in neurons and axons. The metabolic features of mixed glioneuronal tumors could be useful in distinguishing these tumors from other tumors, and for evaluating residual/recurrent disease.

Brain tumors in the first 3 years of life

Yrii Orlov, Ludmila Verbova, Andrii Shaversky (Institute of Neurosurgery named after ACAD. A.P.Romodanov, AMSU., Kiev, Ukraine)

Objectives: Brain tumors in infants differ sufficiently from those of older children in presentation, distribution, and some morphological characteristics to be considered separately.

Methods: Between 1980 and 2009, 541 infants with brain tumors and less than 3 years were treated at the Pediatric Department of the Institute of Neurosurgery. Those patients represent 12% of the tumor of childhood seen in this unit. Of the tumors, 49.9% were supratentorial and 43.1% with lateral location.

Results: The lesions were verified in 427 (79%) cases. In order of frequency, the most common types were astrocytic tumors (all grades, 32.5%), embryonal tumors (22.9%), choroid plexus tumors (10%), ependymal tumors (all grades, 14%), craniopharyngiomas (3.5%), germinomas (1.1%). Malignant tumors accounted for 48% of all neoplasms. Choroid plexus papillomas are the most common tumor during the first year of life (31%).

The surgical removal of the lesion was performed in 355 (65.6%) patients: total removal, 164; subtotal removal, 115; and partial removal, 76. In some cases, CSF-shunting operations were performed after tumor excision. Forty of the patients who survived surgery received postoperative chemotherapy. Follow-up data from 6 months to 18 years is available for 56.8% patients.

Conclusions: Surgical removal is the first choice treatment, chemotherapy does not seem to offer hope of cure but can at times provide control of tumors growth, radiation therapy is delayed until 3 years of age. Children with brain tumors under 3 years of age have a worse prognosis than older children.

Diffusion-weighted imaging and pathological correlation in pediatric medulloblastomas—"They are not always restricted!"

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Introduction: Some investigators have suggested that medulloblastomas can be distinguished from other cerebellar neoplasms by demonstrating "restricted diffusion" on the apparent diffusion coefficient (ADC) map obtained from diffusion-weighted imaging (DWI) sequences on magnetic resonance imaging (MRI). Previous authors have postulated

that this observed restricted diffusion is a reflection of very high cell density. There has been a tendency to assert that pediatric medulloblastoma uniformly demonstrates restricted diffusion on DWI. However, our clinical observation has been that there are pediatric medulloblastomas that exhibit normal or even increased diffusion on DWI. The current study was undertaken primarily to determine whether restricted diffusion is uniformly present in pediatric medulloblastoma and secondly to look for pathological features that might distinguish medulloblastomas with and without restricted diffusion.

Methods: The DWI characteristics of pathologically confirmed medulloblastomas diagnosed at our institution were retrospectively reviewed. The ADC was obtained in two non-overlapping, solid, non-hemorrhagic, non-necrotic regions of tumor and averaged. An ADC below $1 \times 10^{-3} \text{mm}^2/\text{s}$ was considered to represent restricted diffusion. A detailed pathologic review of each tumor was conducted. **Results:** Ten cases of medulloblastoma were reviewed, of which two demonstrated average ADCs above $1 \times 10^{-3} \text{mm}^2/\text{s}$ (1.223 and 1.169 mm^2/s , respectively), indicating no restricted diffusion. Pathologic review revealed that both of these non-restricting cases displayed a lack of reticulin deposition by light microscopy.

Conclusion: DWI does not appear to be an entirely reliable means of distinguishing medulloblastomas from other cerebellar neoplasms. Histologically, restricted diffusion in medulloblastomas may be related to reticulin deposition.

The extent of surgical resection is correlated with outcomes of pediatric intracranial germ cell tumors: single-institute analysis for 20 years

Young-shin Ra, Ho-Jun Lim, Seung-Do Ahn, Shin-Kwang Kang (Asan Medical Center, Univ of Ulsan, Seoul, Republic of Korea)

Objective: Primary intracranial germ cell tumor (GCT) is not uncommon pediatric brain tumors in Asian countries and its treatment options are complicated for various histological subtypes. In this study, the authors analyzed outcomes of intracranial GCTs in order to identify long-term outcome according to histology and treatment strategy.

Methods: The authors retrospectively reviewed 79 children with newly diagnosed intracranial GCTs from 1990 to 2010 in a single institute. Mean age at diagnosis was 12.7 years (range, 20 days–18 years), and 59 patients were male. GCTs were mostly located at pineal region (43%) or suprasellar (34%). The pathology of GCTs was pure germinomas (60%) and non-germinomatous GCTs (40%) in which eight choriocarcinomas, five endodermal sinus tumors, teratomas (three mature, four immature) and 12 mixed GCTs were included.

Results: Forty-seven patients with pure germinoma underwent combined chemotherapy and radiotherapy (37), radiotherapy alone (six), and chemotherapy alone (four). Their 5-year event-free survival rate was 82.6% and overall survival rate was 92.3%. In case of non-germinomatous GCTs treated with surgery and combined chemo-radiation therapy, 5-year event-free survival rate was 52.2% and overall survival rate was 77.0%. Disease control of Non-germinomatous GCTs was correlated to the extent of surgical resection; biopsy only (50%), subtotal resection (50%), and total resection (91%).

Conclusions: Outcome of primary intracranial GCTs apparently depends on their histological characteristics; pure germinomas showed good results with various treatment options, but non-germinomatous GCTs need more aggressive treatment options including radical resection of tumor before or after adjuvant therapy.

Primary spinal epidural Burkitt's lymphoma in a child: a case report

Taopheeq Rabi (Federal Medical Centre, Ido-Ekiti, Nigeria)

Introduction: Burkitt's lymphoma primarily involving the spine is very rare and is often associated with poor prognosis. This is a report of such a case in a young Nigerian boy.

Case summary: A 10-year-old school boy presented with rapidly progressive paraparesis following a fall from a cashew tree. He had T8 paraparesis (Frankel C). Spinal X-rays showed no evidence of trauma. Thoracic and lumbosacral magnetic resonance imaging revealed contrast-enhancing dorsal epidural masses at T6–T11 and L2–L4 levels with severe cord and cauda equina compression. There was no clinical evidence of abdominal or thoracic disease. T6–T11 laminotomy was performed for gross total excision of the thoracic mass. A soft epidural mass with multiple areas of hemorrhagic necrosis was found. Histology revealed Burkitt's lymphoma. He (and his parents) defaulted from care but was re-admitted 2 months after surgery with complete paraplegia, multiple abdominal masses, and renal failure. He died before chemotherapy could be instituted.

Conclusion: This case illustrates the abysmal outcome of children with primary Burkitt's lymphoma. Early institution of chemotherapeutic trial in children with multilevel spinal tumour with no evidence of extra-spinal disease might be beneficial.

Timing of imaging following resection of pediatric posterior fossa tumours and effect on achieving complete resection

Jacqueline Reaper, John McMullan, Hesham Zaki (Sheffield Children's Hospital, Sheffield, South Yorkshire, UK)

Prognosis of pediatric posterior fossa tumours is improved with complete surgical resection. In our institution, we perform MRI on the day of surgery whilst still under anaesthetic. If residual tumour is seen, we consider further debulking on the same day.

Eleven children underwent posterior fossa tumour resection over a 2-year period (ten new diagnoses, one recurrence). Median age was 3 years (0–14). There were five medulloblastomas, three ependymomas, two pilocytic astrocytomas, and one clear cell meningioma. One had spinal metastases at presentation.

Intra-operative gross total resection was achieved in ten and subtotal excision in one (close proximity of vessel, complete resection unsafe). Post-operative MRI was performed on the day of surgery in all 11. Eight had no residual tumour. Two of those thought to have complete excision intra-operatively but with residual tumour on MRI returned to theatre immediately from scan. The child with subtotal excision returned at later date. Complete excision was achieved (demonstrated on follow up imaging) in all three.

Post-operatively, two developed mutism that completely resolved. One developed mild cognitive problems, which are improving. There were no other new deficits.

Eight patients have no recurrence to date. One had radio-surgery for recurrence and is now also disease-free. The child with spinal metastases at presentation and the child operated on for recurrent tumour died. Mean length of follow-up is 23 months (15–44).

Performing imaging on the day of surgery allowed us to achieve complete resection in 10 of 11 patients in a single sitting, with very low complication rate.

Is it safe to leave head open, after craniotomy or craniectomy

Safi Ur Rehman¹ (¹Department of Neurosurgery, MCM Korean Hospital, Addis Ababa, Ethiopia, Ethiopia; ²Department of Neurosurgery, Kwong Wah Hospital, 25 Waterloo Road, Kowloon, Hong Kong)

Inspired by the dramatic effects of leaving the abdomen temporarily open for abdominal compartment syndrome and even for reducing intracranial pressure in traumatic brain injury (TBI), the idea of leaving the head open for a few days may be plausible.

Bearing in mind the serious risks of CSF leak and infection, leaving the head open seems risky and fraught with danger but does it invariably lead to disaster.

We present a case of a young girl with a massive tumour of the forehead with invasion of dura, skull and brain. After removing the grossly infected tumor, dural closure was achieved with fascia lata graft only, leaving the scalp open.

The second operation, 5 days later consisted of a large acrylic cranioplasty and pedicle skin graft. The patient did not suffer any serious consequences from delayed closure. Two similar cases done previously for tumour and TBI will be briefly outlined.

Details of this dramatic case and promising outcome will be presented and critical review of the literature discussed.

The procedure cannot be recommended as a routine procedure but in exceptional circumstances, when the situation demands, such drastic measures may be the last resort. Further clinical and laboratory studies are needed to determine the safety and efficacy of the procedure.

Single center experience in the application of intraoperative high-field magnetic resonance imaging in pediatric neurosurgery

Thomas Schmidt, Marie Schuler, Klaus Seitz, Michal Hlavac, Christian-Rainer Wirtz, Ralph Werner Koenig (Neurosurgical Department, University of Ulm, Ulm, Germany)

Objective: Intraoperative MRI (iMRI) and neuronavigation are established in the treatment of brain tumors. Previous studies have shown the value of iMRI in pediatric neurosurgery mostly for low-field systems. The authors summarize their experience using an intraoperative high-field iMRI a 1.5 T with integrated neuronavigation.

Methods: Between January 2009 and April 2011, 15 consecutive cases were treated in a BrainSuite 1.5 T iMRI. Eighteen intracranial neurosurgical procedures were performed in 15 children (eight male and seven female). Focussing on whether intraoperative imaging and neuro-navigation affected surgical strategy the entire surgical procedure was evaluated for feasibility, image quality, accuracy and safety.

Results: The median age at surgery was 11 years (5–20 years). Treated lesions included pilocytic astrocytomas, anaplastic astrocytoma, medulloblastomas, ependymoma, adenoma, ganglioglioma, cortical dysplasia, inflammatory lesions and gliosis. Neuronavigation was applied in all cases with high accuracy. iMRI led to further surgical resection in 33% of the cases. Thus, gross total resection was achieved in 12 surgeries. iMRI during four procedures for the treatment of non-neoplastic disease did not lead to any change in surgery but confirmed the surgical goal of the surgeon. Average image quality was excellent. Permanent morbidity was 7%, whereas mortality was 0%.

Conclusions: Highfield- iMRI in pediatric neurosurgery is safe and allows less invasive surgical exposures. Its potential is greatest when its high image quality is coupled with its superior neuronavigation capabilities, which permits tracking

of the extent of resection of intracranial tumors and, to a lesser extent, other lesions during the surgical procedure.

Vestibular schwannomas in children—study of 11 cases

Manish Tawari, Sanat Bhagwati, Geeta Parulekar, Mahesh Chaudhary, Suneel Shah, Nirav Mehta, Anoop Thomas, Amit Mukherjee (Bombay Hospital Institute of Medical Sciences, Mumbai, Maharashtra, India)

Introduction: Vestibular schwannomas in children are rare accounting for 0.8% of all brain tumors in children.

Objectives: This study aims to retrospectively analyze the epidermological profile, clinical features, radiological findings, type of excision, histopathological findings and overall management profile of these patients and comparison of our study with the world literature.

Material and methods: Eleven consecutive cases of vestibular schwannoma in children under 18 years of age operated at our institute between 1972 and 2010 are included in this study.

Results: Mean age of presentation to our hospital was 14.9 years. Male/female ratio was 3:8. Average duration of symptoms was 7.18 months. B/L vestibular schwannoma was present in 18.18% and neurocutaneous marker was present in 9.09%. Increased incidence was seen as sporadic vestibular schwannoma rather than associated with neurofibromatosis. Total tumor excision was achieved in all cases with no postoperative mortality.

Conclusion: Vestibular schwannoma in children present surgical challenges in view of large size tumors but retromastoid suboccipital approach is an effective approach for safe removal of all types of vestibular schwannoma in children.

Intracranial meningiomas in infants—experience at PGIMER, Chandigarh

Manoj Tewari, Rajesh Chhabra, Sunil Gupta, Bishan Radotra, Niranjan Khnadelwal (PGIMER, Chandigarh, India)

Intracranial meningiomas are rare tumors in infants and children accounting for 0.4–2% of all intracranial tumours. Usual presentation in features of raised intracranial tension and focal neurological deficits. They can be in any location supra or infratentorial. We report our experience of two infants with very big meningiomas. Clinical features, imaging, histopathological aspects, surgical strategies, intraoperative problems and outcome will be discussed and relevant literature will be reviewed.

Clinical characteristics and treatment of optic pathway gliomas in Han nationality in China

Yongji Tian, Deling Li, Ge Jia, Zhenyu Ma (Beijing Tiantan Hospital, Beijing, China)

The aim of this study is to describe clinical characteristics of optic pathway gliomas (OPG) in Han nationality in China and evaluate the treatment of microsurgical management followed by postoperative radiotherapy for these patients. Fifty-three pediatric OPG cases treated in Beijing Tiantan Hospital from April 2003 to January 2011 were retrospectively reviewed. All patients' clinical data were collected and analyzed. Mean age at presentation was 7.6 years (range, 10 months–28 years). The ratio of male to female equals 1.5: 1. There were two cases of NF1 in the OPGs patients. Based on the Dodge classification, type I covered 5.6% (three cases), type II covered 67.9% (36 cases), type III covered 26.4% (14 cases). All tumors were partial resected (50–70%) and pathology showed fibrillary astrocytes in 27 cases (51%), pilocytic astrocytomas in 24 case (45%), and there were one oligodendrogliomas and one ganglioglioma case. Average period of follow-up was 31 months (range, 3 months–8 years). Of these follow-up, symptoms were improved in 11 cases and stable in 30 cases. The mean progression-free survival (PFS) was 26 months (range, 3–64 months). The patients received radiotherapy have a significant better PFS than those without radiotherapy ($p < 0.01$), while radiotherapy side effect occurred only in one patient with hypothyroidism. Partial resection of tumor followed by radiotherapy was efficient to control OPGs with less complication. There were less NF1 in Han nationality in China compared with OPGs patients in existing literatures from other countries.

The contribution of the neurosurgeon to highly conformal radiotherapy in standard risk medulloblastoma: a modelling approach

Erik van Lindert¹, Corrie Gidding², Stefan Rutkowski³, Peter van Kollenburg⁴, Geert Janssens⁴ (¹Neurosurgery, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; ²Pediatric Oncology, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; ³Pediatric Oncology, University Medical Centre Hamburg-Eppendorf, Hamburg, Germany; ⁴Radiation Oncology, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands)

Objective: Multiple retrospective single centre studies do support the use of boost volumes limited to the tumor bed to reduce potential severe late toxicity in standard risk medulloblastoma patients. Based on current MR information, the volume of the tumor bed is probably overestimated. **Material and methods:** A neurosurgical report with emphasis on growth patterns in relation to reference structures, visible on MR imaging, was developed. MR datasets (T1±gadolinium, T2) of the pre-operative, immediate post-operative and the situation, 1 week before onset of the

boost irradiation, were matched. Based on neurosurgical and MR data, the tumor bed (GTV) and clinical target volume (GTV+1.0 cm) was shaped and compared to a situation where neurosurgical information was not available (ongoing standards). Organs at risk were delineated. A boost dose of 30.6 Gy in 1.8 Gy fractions was prescribed. Results: A patient, who underwent a macroscopic complete resection was selected. Identification of the tumor bed based upon pre-operative extent and anatomic shifts after surgery resulted in a volume of 7.9 cm³ compared to 1.7 cm³ when neurosurgical information was used. Corresponding CTVs were 66.9 and 30.1 cm³, respectively. The model demonstrated a biological boost dose reduction of 30–45% for the contralateral cochlea, 33–42% for the pituitary gland, and 51–62% for the hippocampus compared to the traditional approach with 1.0 and 1.5 cm margins, respectively.

Discussion: Integration of neurosurgical information can further reduce the target volume and consequently the dose to the critical surrounding tissues. A prospective analysis is needed before introducing this highly conformal approach in daily practice.

Deep sequencing analysis identifies the oncogenic role of miR-221-222 in pediatric embryonal brain tumors

Jui-Yu Hsieh¹, Chen-I Lin², Chan-Yen Tsai¹, Hsin-Yi Tsai³, Chih-Yi Hsu⁴, Meng-En Chao², Da-Jung Liu², Muh-Lii Liang², Shih-Chieh Lin⁴, Donald Ming-Tak Ho⁴, Tai-Tong Wong², Hsei-Wei Wang¹ (¹Institute of Microbiology and Immunology, National Yang Ming University, Taipei, Taiwan; ²Division of Pediatric Neurosurgery, Neurological Institute, Taipei Veterans General Hospital, Taipei, Taiwan; ³Institute of Clinical Medicine, National Yang-Ming University, Taipei, Taiwan; ⁴Department of Pathology and Laboratory Medicine, Taipei Veterans General Hospital, Taipei, Taiwan)

Atypical teratoid/rhabdoid tumor (AT/RT) is a highly malignant central nervous system tumor often misdiagnosed as other pediatric embryonal tumors such as medulloblastoma (MB). AT/RT prognosis is much worse than MB, but the underlying mechanisms are unclear. To enhance current prognostic and therapeutic systems, tumor molecular properties were examined on fresh AT/RT and MB tissues by gene expression microarray and RNA-Seq technologies. Novel miRNAs were found, and their very existence could be confirmed by wetlab validation such as RNA-immunoprecipitation (RNA-IP). Genes and miRNAs responsible for stemness (S100A4 and SOX1), proliferation/tumorigenesis (miR-221, miR-222, IL7 and IL18) and angiogenesis (FGF2 and VEGFA) were abundant in AT/RT, while those associated with differentiation and neurogenesis (miR-301 and

ZIC) in MB. AT/RT possesses a dedifferentiated transcriptome pattern, partly due to the overexpression of embryonal genes. This study discloses the molecular makeup of embryonal brain tumors and suggests candidate drug targets for AT/RT and MB. Our current effort is to deduce the relationships between filtered genes and stem cell features. Further challenge will be to apply cloud computing technologies to facilitate next-generation sequencing (NGS) data, as well as to further integrate NGS information with systems biology algorithms.

Neurosurgical endoscopic approaches and operative strategies in pediatric craniopharyngioma

Christoph Wiegand¹, Niels Sörensen¹, Hermann Müller² (¹Ped. Neurosurgery, University Mainz, Mainz, Germany; ²Ped. Oncology Klinikum Oldenburg, Oldenburg, Germany)

Craniopharyngioma defines an intracranial embryonal, non-glial dysplastic tumor of the sella in close anatomical vicinity to hypothalamus, brain stem, optic nerve and pituitary gland.

Neuroendoscopic routes to craniopharyngioma have been defined as transnasal-transsphenoidal, transventricular as well as the supra-orbital approach.

Our series consists of $n=17$ pediatric patients, operated upon from April 2009 to November 2010. The youngest child was 4, the oldest was 14 years, showing a mean age of 5.6 years. $N=11$ patients faced surgery via a fronto-orbito-lateral approach. All of them had intraoperative neuroendoscopic rectification of the resectional tumor amount, $n=4$ presented with craniopharyngioma as first time diagnosis, whereas $n=3$ were recurrent cases. $N=3$ underwent transventricular endoscopic surgery alone, consisting of mainly cystic lesions. One patient, 14 years old, was followed in excellent clinical condition without hormonal substitution. The approach fitted best for a child suffering complex heart failure, hemiparetic, blind due to chiasmatic cystic tumor compression and on complete hormonal substitution. This case highlighted nicely the value of minimally invasive neuroendoscopy being highly effective.

$N=1$ Patient received gross total resection of a solid craniopharyngioma. She was operated upon via the transsphenoidal route 2 months later presenting acute loss of vision due to chiasmatic compression from tumor progress. The histologic specimen showed an immature teratome.

To our understanding, preoperative hypothalamic involvement by craniopharyngioma, especially invading the posterior part of hypothalamus predicts a limited tumor resection to avoid focal deficits. The hypothalamus represents the resectional neurosurgical landmark.

*Nursing Symposium***The management of the patient of brain tumor for better quality of life and the role of nurse for psychological care**

Mariko Fukui, Aki Tashiro, Natsuko Sakaguchi (Kansai Medical University, Hirakata, Osaka, Japan)

Abstract

Background: The Delhi city alone witnessed 7,516 (2009) road traffic accidents and many were admitted to hospitals as unknown or unattended.

Aims: This study aims to assess the morbidity and mortality of unknown or unattended patients and problems faced during nursing care.

Materials and methods: This is a retrospective analysis from February 2010 to August 2010 wherein all unknown or unattended with head injury (Glasgow Coma Scale (GCS) 1–15) admitted in neurosurgery was included. The duration of hospital stay, admission GCS, and outcome were assessed and an attempt was made to analyse the problems faced by nursing personnel during their hospital stay.

Observations: Total patients enrolled during the study period were 38. Of these, 22 were unknown and 16 were unattended. Average hospital stay of unknown and unattended was 35 (1–151) and 21 (7–120) days, respectively. Mean GCS of unknown during admission was 9 (3–15) and during discharge was 8 (3–15). Mean GCS of unattended during admission and discharge was 12 (13–15) and 14 (3–15), respectively. Of the 22 unknown, 24% became known during hospital stay, 33% shifted to rehabilitation homes, and 43% expired as unknown. Of the 16 unattended, 25% went home, 63% shifted to rehabilitation homes, and 12% expired. The most common problems faced during nursing care were contractures (8%) and pressure sores (11%) mainly because of long hospital stay.

Conclusion: Patients remaining unknown/unattended is a unique problem as far as developing countries are concerned. Managing these patients is difficult as they occupy hospital beds for longer duration and require more nursing care with higher mortality and morbidity. It remains surprising that in spite of advancements in the field of mass communication, almost 76% of the unknown remain unidentified.

Does PEEP affect central venous pressure reading in mechanically ventilated head injury patients?

Category: 10. Trauma,

Author's preference: oral presentation

Metilda Robin, Deepak Agrawal (JPN Apex Trauma Center, AIIMS, New Delhi, India)

Background: Monitoring of central venous pressure (CVP) is important so as to ensure adequate Volume status in

severely injured patients. In routine practice, positive end-expiratory pressure (PEEP) is subtracted from CVP reading in ventilated patients.

Aims and objectives: This study aims to assess the effect of PEEP on CVP in mechanically ventilated head injury patients. **Methods:** This prospective study was carried out in neurosurgery ICU at JPNATC, AIIMS over a 2-month period. Hemodynamically stable mechanically ventilated head injured patients with CVP monitoring were enrolled for the study. CVP was measured manually with ventilator support and with ventilator disconnected with a time gap of 5 min. The difference in both reading was recorded. The PEEP was also recorded whenever CVP was measured with ventilator connected. Student's two-tailed test was used to compare the two groups. **Results:** A total of 40 patients were taken for the study, in which 32.5% were females and 67.5% were males. In the study, 22.5% patients were on PSV mode of ventilation and 77.5% patients were on SIMV mode. After analysis, it was found that no significant difference in CVP reading in mechanically ventilated patients when the PEEP was set at <10.

Observation	T test	P value
1st CVP measurement	2.03×10^{-8}	0.99
2nd CVP measurement	2.12×10^{-6}	0.99
3rd CVP measurement	0.0004	0.99
4th CVP measurement	0.0016	0.99

Conclusion: It is not necessary to subtract PEEP from the CVP reading if PEEP is set below 10 mmHg.

Study on support of prenatally diagnosed children with congenital hydrocephalus by occupational therapists

Status: Pending

Category: 12. Other

Author's preference: Oral Presentation

Miyoko Takebayashi, Chika Teramoto, Masahiro Nonaka, Mami Yamasaki (National Hospital Organization Osaka National Hospital, Osaka, Japan)

More than 60% of patients with congenital hydrocephalus are diagnosed prenatally. Among the patients with prenatally diagnosed hydrocephalus, 80% show some developmental retardation. Therefore, it has been suggested that these patients should receive the rehabilitation from an early age. However, to date, no reports on the rehabilitation with these prenatally diagnosed patients have been published. Moreover, no studies have assessed parents' opinions about the need for early rehabilitation.

We sought to determine the types and timing of support needed by patients prenatally diagnosed with congenital

hydrocephalus and their parents, by identifying such cases retrospectively and surveying the parents.

We investigated the questionnaire by mailing. We chose 150 parents of patients with congenital hydrocephalus: 80 were prenatally diagnosed, 70 were postnatally diagnosed. All patients were diagnosed and treated at our hospital.

Effective answers were collected from 77 parents (43, prenatal; 34, postnatal). The average time from the diagnosis to the delivery was 8.6 ± 5.1 weeks. The explanation and support to the parents were performed mainly by doctors. However, 58% of the respondents wanted to hear from the therapeutic personnel, indicating that an explanation of the available therapeutic approaches is greatly desired. Many parents wished that occupational therapy start within a year of birth.

The time between prenatal diagnosis and delivery can be thought of as a time for the family to prepare to accept their child. This time is necessary to decrease anxieties and provide the appropriate information to the family. In the future, therapeutic personnel should actively join this support group.

The management of the patient of brain tumor for better quality of life and the role of nurse for psychological care

Mariko Fukui, Aki Tashiro, Natsuko Sakaguchi (Kansai Medical University, Hirakata, Osaka, Japan)

Introduction: Pediatric brain tumors are the most common solid tumor and the second most common neoplasm in childhood. Recent advances in neuroradiology, neurosurgical techniques, and neuro-oncology have begun to impact the length and quality of survival of children diagnosed with brain tumors. However, the prognosis of those patients are often bad. In this study, we reported a case of brain tumor and discussed the importance of patient's management.

Case presentation: The patient was a 14-year-old boy. When he was sent to our institute, he had the left hemiparesis. The tumor was thought to be aroused from the right thalamus. The biopsy revealed the tumor was cerebral glioma grade III. Because of the nature of tumor, we decided the goal should be to send him home after the combined treatment with irradiation and chemotherapy.

Results: He became dependent during hospital stay. He was also losing his self-confidence to come home. Because of those problems, we started to have conference between team nurses to determine how to deal with those problems. We also discuss with social medical workers and teachers from school. After discussion, all team nurses communicate with him in the same attitudes. Since then, he started to live positively.

Discussion and conclusion: Team approach to the patients with brain tumors seems effective. However, the continuous psychological care had not been carried out after discharge.

We need to establish the system to follow those patients after discharge from the hospital.

External ventricular drainage and posterior fossa tumors: the crucial role of nurse management

Paola Leonardi (Catholic University, Rome, Italy)

Background: Endoscopic third ventriculostomy (ETV) is the best treatment of hydrocephalus in posterior fossa tumors (PFT). ETV can be performed either preoperatively or postoperatively. Both options have specific advantages and limits. Postoperative ETV requires the maintenance of an external ventricular drainage in the first postoperative days in order to discriminate those patients who will require the ETV procedure from those undergoing spontaneous compensation of the hydrocephalus. Consequently, the main limit of postoperative ETV is the risk of contamination of the external drainage, for the prevention of which the role of the nurse word is crucial.

Materials and methods: The series is composed by 67 children operated on for PFT between 2005 and 2010 and harboring preoperative hydrocephalus. All of them underwent perioperative external ventricular drain by means of antibiotic impregnated catheter, which was left in place for no more than 7–10 days. In the postoperative period, both ICP and the amount of daily produced CSF were recorded.

Results: Twenty-eight children showed persistent postoperative hydrocephalus (42%) as demonstrated by high ICP values and large amount of produced CSF. This amount, which varied according to the child's age, was found to progressively decrease in children with resolution of hydrocephalus (average: 365 ml/day in the first postoperative day vs. 20 ml/day in the seventh postoperative day) compared with those with persistent hydrocephalus (374 vs. 190 ml/day). Two children developed CSF infection (3%); one of them experienced failure of ETV.

Safe and sustainable paediatric neurosurgery in the UK—are there lessons to be learnt?

Lindy May (Great Ormond Street, London, UK)

The National Specialised Commissioning Group (NSCG) has been undertaking a review of the provision of children's neurosurgical services in England with a view to reconfiguration. The pressures prompting the need for a review of the current configuration of services in the interests of delivering a safe, high quality service include the fact that centres vary in the range of neurosurgery services they provide and in access to subspeciality expertise. The infrastructure of some centres may be considered sub-optimal in that compliance with on-call consultant cover may not be in place/achievable and

associated paediatric services including nursing may not be adequate.

Can the lessons we have learnt from this process be adapted and utilised in international settings?

Outcomes following a decompression of a severe traumatic brain injury

Helena Willans (Great Ormond Street Hospital, London, UK)

One of the factors that can affect the outcome following a traumatic brain injury (TBI) is cerebral oedema the increase of intercranial pressure (ICP).

A study was taken on 155 patients with TBI where 73 underwent an early decompression craniotomy and 82 patients received standard care. The study showed how the patients with decompressive craniotomies had less time in intensive care and required less intervention to decrease ICP. If ICP becomes elevated, it can compromise the metabolism of the brain and therefore causes secondary brain injury, which in turn could affect the outcome of the patient. By having a decompressive craniotomy, it allows the brain to swell in a larger area. The research behind the outcomes varies and whether they are beneficial to the patient and their long-term care.

In this presentation, I will analyse the literature available to find the best treatment required for these group of patients and the care provided by neurosurgical nurses.

Paediatric neurosurgery in Christchurch: providing specialty care in a general hospital

Becky Conway, Alison Duggan (Christchurch Hospital, Christchurch, New Zealand)

Christchurch, New Zealand, is a city known mostly of late for its devastating earthquakes. Christchurch, population approximately 400,000, is the largest centre in the South Island of New Zealand. Christchurch Hospital is an acute publicly funded teaching hospital where children and adults are admitted for specialist care.

There is no separate children's hospital in Christchurch. Paediatrics is a subset of the main hospital within a division known as Women's and Children's Health.

When children need neurosurgical care, they may be admitted to either of the three wards, as an arranged admission or acutely through the Emergency Department or the Child Acute Assessment Unit.

The challenge for the nurses who provide the care in these units is to have specialist neurosurgical nursing skills in order to provide an excellent standard of assessment and care, within a multispecialty environment. How do we ensure that all nurses with a range of experience are skilled enough to think as good neurosurgical nurses? And how do

we tailor an essentially adult-focussed surgical service to the needs of children?

There is little available literature about paediatric neurosurgical nursing within a general paediatric ward or unit, situated in an adult hospital. Yet this must be a common experience in many parts of world. By describing the services for children needing neurosurgical care at our hospital, there is an opportunity to review and discuss how our service meets this challenge.

Looking back on relations to spina bifida patients for CISC; clean intermittent self-catheterization

Terumi Fukai, Mami Yamasaki (National Hospital Organization Osaka National Hospital, Osaka, Japan)

Spina bifida clinic is established in Osaka National Hospital. For the patient of the SB clinic, there were 14 patients who began clean intermittent self-catheterization (CISC). It was investigated with a past clinical record whether CISC had been started.

At first, we taught the mothers about clean intermittent catheterization (CIC). Usually, when they were 6-year-old children, we taught about CISC. They were in the hospital and they have undergone the operation on the untethering or the VP shunt revision. Diagnosed as the spinal lipoma by dysuria, it was about 10 years old when CIC was begun, and the diagnosed infants who had become CISC subjects were four patients.

Three patients were hospitalized for CISC. Two patients could do CISC, but a patient could not do CISC. Afterwards, the point with the problem was in the interpersonal relationship formation received by the mother from the psychology counselor.

SB patients become independent when they could do CISC. Many of the SB patients who cannot do CISC should visit the school with the mother and do CIC. The example executed by the nurse who works for the nurse-teacher and the school is a very unusual case.

As for SB patients who were following it in this hospital, they could finally become independent. It seemed that it was necessary to intervene positively for the SB patient who was not able to do CISC.

Study to assess the cost-effectiveness of reuse of expensive disposable items in neurosurgery ICU

Metilda Robin, Deepak Agrawal (JPN Apex Trauma Centre, AIIMS, New Delhi, India)

Background: Disposable items like percutaneous tracheostomy (PT) sets and ICP monitoring sets are expensive and drain the resources of public-funded hospitals.

Aims and objectives: This study aims to assess the use and cost-effectiveness of reusing expensive disposables (PT and

ICP sensor) in neurosurgery ICU. Another objective was to assess the importance of bedside tracheostomy and ICP insertion in ICU rather than in OT.

Methodology: An observational, retrospective study was done from January 2009 to November 2010 in the neurosurgical department of JPNATC. Retrospective data on surgeries performed in neurosurgery OT was taken for the year 2009 and following introduction of PT and bedside ICP monitoring sets in ICU, prospective data was collected from November 2009. A procedure book was maintained in which each case with the set used was mentioned.

Observations: Of the 1,209 surgeries done in neurosurgical OT in 2008–2009, 257 were minor procedures (238 open tracheotomy and 19 ICP transducer placements). In 2009–2010, 236 percutaneous tracheostomies were done in ICU. Of these, 79 (33.4%) were new and 157 (66.5%) were reused sets. The cost of new PT set is Rs 15,000. With reuse, the average price per set came down to Rs 5,033/set. In the same period, 231 disposable ICP transducers were placed with an average of 19 cases per month (range, 5–28). Of these, 142 (61%) were new ICP sets and 97 (42%) were ETO sterilized. The cost of new ICP set is Rs 35,000. With reuse, the average price per set came down to Rs 2,515/set.

Results: Nearly 40% of minor procedures are now excluded from the neurosurgery OT statistics as they are being performed in ICU. Our study shows that expensive disposable items can be effectively reused bringing immense cost savings to hospitals.

Craniopharyngiomas: pre- and post-operative care from a nursing perspective

Sarah Wallington (Great Ormond Street Hospital, London, UK)

Abstract

Brain tumours are the second most common childhood cancer accounting for 24% overall. At Great Ormond Street Hospital alone, 80–110 brain tumours are newly diagnosed a year. Craniopharyngiomas make up 6–13% of all childhood tumours; this makes them the most common childhood tumour found in this area of the brain. There are approximately 25 new cases a year in the UK.

Craniopharyngioma are classified as benign lesions. However, due to regrowth and the potential damage they can cause to the brain, they are considered as potentially damaging as a malignant tumour. Craniopharyngiomas often invade the pituitary and hypothalamic glands and can cause optic nerve damage. Due to the location of the tumour, complications may occur pre- and post-operatively, this requires specialist nursing care and a multidisciplinary approach. As an example, 9–17% of craniopharyngioma

patients will have diabetes insipidus pre-operatively and 60–80% post-operatively. Management of Di can be a challenge.

It is essential that appropriate and high-level nursing care is provided for these patients both pre- and post-operatively to help minimise potential complications. This presentation will look at the role of the neurosurgical nurse, discussing the baseline assessment including the basal endocrinology and recommendations for pre-operative care. Post-operative management including the importance of comprehensive fluid management will be examined with recommendations for best practice.

Pediatric neurorehabilitation—nursing issues

Yee Yit Cheng (Kuala Lumpur Hospital, Kuala Lumpur, Malaysia)

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Pediatric neurorehabilitation is crucial for their ongoing learning and development and can help develop new skills and compensatory strategies to decrease the effects of the brain injury.

It is effective using a multidisciplinary approach and nurses play an important role in training and educating the families and caregivers and retraining of patients to help them regain function and recover physical and cognitive skills.

Nurses face a lot of problems dealing with these children: due to shortage of staff and trained personnel, e.g., pediatric neuropsychologist, speech therapist, physiotherapist, nurses are so burn out with the routine care that needed for these children, therefore we, nurses find it difficult to carry out proper rehabilitation for these children.

Full support and cooperation from families or caregivers is crucial. Families often cannot emotionally accept the futility of the situation even they intellectually understand it; this will interfere with the progress of therapy.

We believe, even with all the limitation we have, rehabilitation gives as many neurologically challenged individuals as possible the opportunity for success and the need for every child to have access to his potential to get back into as normal life as possible.

To assess the effect of camera surveillance in improving compliance with hand washing practices in the ICU setting

Shallu Chauhan, Deepak Agawam, M.C. Mishra (All India Institute of Medical Sciences, New Delhi, India)

Neurosurgical patients, particularly of severe head injury require frequent CTs of the head, usually at short notice.

Aims: This study aims to review the usefulness of mobile CT in Neurosurgery ICU at a level 1 trauma centre in India
Methods: This review was carried out over a 14-month period in AIIMS. Administrative and clinical data was reviewed and analyzed. For the first 6 months, only the number of CTs done was available. However, data was collected to include variables like GCS ventilator status and pressor support at the time of CT scan. The average time to do a CT was 150 min. The mean number of people required for shifting the patient was four whereas for mobile CT, the average time to do a CT was 27.4 min and mean for manpower required is three.

Observations: A total of 1,752 head CTs were done during the study period with an average of 4.78 CTs daily. Detailed data was available on 1,023 patients. Of these patients, 75.85% were on ventilator, 72.3% were on sedation and 5.57% were on pressor support at the time of CT head. The mean GCS at the time of CT was 8.09. The average time taken to do a CT scan was 12.6 min.

Conclusions: Over a short period of 14 months the mobile CT managed a record 1,752 head CTs and acted as a back-up CT for the casualty. Mobile CT was found to be extremely useful for neurosurgery department

Is routine use of arterial lines in ICU feasible for blood sampling?

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Background: Patients in ICU require repeat blood sampling for various investigations. This results in repeated arterial and venous puncture leading to avoidable complications like thrombophlebitis and sepsis.

Aim and objectives: This subject aims to assess the feasibility for keeping arterial lines in ICU patients for sampling. A secondary objective was to assess the problems associated with sampling using traditional methods.

Materials and methods: A prospective study was carried out over a period of 1 month in Neurosurgery ICU JPNATC AIIMS. Arterial lines placed during surgery in head injured patients were retained in the postoperative period. They were kept patent by flushing with normal saline and using pressure infusion bag. The control group was assessed for

number of arterial and venous punctures per day basis and complications at the site.

Observations: Thirty patients were included in the study which included 10 in the arterial line group and 20 in the non-arterial line group. Average duration of arterial line was 2.4 days (1–7 days). Average number of arterial and venous puncture in the controlled group was 2.7/day (1–5) and 1.1/day (1–2), respectively. Complications seen in the arterial line group was blockage (50%) followed by kinking (40%) whereas complications in controlled group was thrombophlebitis (40%), followed by haematoma (20%).

Conclusions: Arterial lines eliminate the requirement of repeated arterial or venous puncture with the potential of dramatically reducing local complications. The patency of arterial lines can be significantly prolonged using pressure infusion bags. Our study shows that arterial lines should be the standard of care in ICU patients.

Ventricular Infusion studies are a useful and cost effective tool for diagnosing shunt malfunctions in patients with unchanging neuroradiology and equivocal symptoms

Jenny Sacree (Frenchay Hospital, Bristol, UK)

Hydrocephalus and CSF diversion with shunts is a mainstay of paediatric neurosurgery. Shunt malfunction and subsequent revision is an inevitable consequence of shunt dependence. Every paediatric neurosurgery unit will have patients admitted on a regular basis with more subtle signs of shunt malfunction and unchanging CT imaging. Often, there may be behavioural issues or developmental delay that further cloud the picture.

At Frenchay Hospital, Bristol in the UK, we have a nurse practitioner run ventricular infusion studies service for the diagnosis of shunt malfunction in those patients without neuro-imaging change and subtle shunt malfunction symptoms.

In this paper, I would like to discuss the service along with the cost and time benefits to patients, staff and institutions.