

43rd Annual Meeting of International Society for Pediatric Neurosurgery, Izmir, Turkey, Oct 4-8, 2015

This supplement was not sponsored by outside commercial interests. It was funded entirely by the publisher.

Chandrashekhar Deopujari, ISPN President
Saffet Mutluer, ISPN 2015 Meeting Chair
Graham Fieggen, ISPN Scientific Chair

Monday, 5 October 2015
10:30 – 12:40

Platform Presentations

Oncology Free Papers

PF-001

Posterior fossa tumor

A probabilistic atlas of the cerebellar white matter

Kirsten Van Baarsen¹; Michiel Kleinnijenhuis²; Saad Jbabdi²; Stamatios Sotiropoulos²; Andre Grotenhuis¹; Anne-Marie Van Cappellen Van Walsum³.

¹Department of Neurosurgery, Radboud University Medical Centre, Nijmegen; ²Oxford Centre for Functional MRI Of The Brain (FMRIB), Nuffield Department of Clinical Neurosciences, University of Oxford, Oxford; ³Department of Anatomy, Radboud University Medical Centre, Nijmegen.

Objective: The Cerebellar Mutism Syndrome is a devastating disorder that may occur in about 25% of patients undergoing surgery for a posterior fossa tumor. In search for the anatomical substrate, differences in cerebellar white matter integrity should be investigated between patients and controls. A parcellation atlas would facilitate automatic and uniform segmentation and between-group analysis. In this work a digital three-dimensional probabilistic atlas of the cerebellar white matter is presented.

Methods: High quality 3 Tesla, 1.25mm resolution diffusion MRI data from 90 subjects participating in the Human Connectome Project were analyzed. Probabilistic tractography was performed in individual space based on automatic Region of Interest (ROI) segmentation in standard MRI space.

Results: The average tractography results were highly symmetrical and trajectories of superior, middle and inferior cerebellar peduncles strongly resembled the anatomy as known from anatomical studies. Binary parcellation maps were created at probability thresholds of 10, 25 and 50%.

Conclusion: This atlas will contribute to a better understanding of cerebellar white matter architecture. Moreover, it will facilitate the segmentation of the cerebellar peduncles, the localisation of lesions and the comparison of white matter integrity between patients and controls. It may eventually aid in defining structure-function correlations in patients with cerebellar disorders such as the Cerebellar Mutism Syndrome.

Keywords: Cerebellum, Mutism, Atlas, Tractography, Human Connectome

PF-002

Other

Bilateral lesion of the fastigial nuclei in juvenile rats – effects on social behaviour, vocalization and motor activity

Shadi Al-Afif¹; Frauke Helms¹; Svilen Angelov¹; Kerstin Schwabe¹; Joachim K Krauss¹; Elvis J Hermann¹.

¹Department of Neurosurgery, Medical School Hannover, Hannover.

Objective: Radical resection of malignant midline tumors of the posterior fossa in childhood followed by adjuvant therapies like chemotherapy or radiotherapy often leads to longterm survival and even healing of such patients. Therefore, quality of life becomes particular important. Postoperative neurological deficits, such as cerebellar mutism and ataxia, have been attributed to damage of cerebellar midline structures during surgery to remove these tumors. Here, we tested the effect of bilateral lesion of the fastigial nuclei in juvenile rats on social behaviour, vocalization and motor activity.

Methods: Juvenile male Sprague Dawley rats, aged 23 days, underwent bilateral thermocoagulation of the fastigial nucleus using stereotactically applied high-frequency electric current under general anaesthesia (lesioned group, n=16). In sham-lesioned rats, electrodes were inserted without application of electric current (n=16). Naïve rats served as controls (n=16). All groups were tested on day 0 (before surgery), and on days 1, 2, 3, 4 and 7, as well as 2, 3, 4, and 7 weeks after surgery for locomotor activity, motor coordination, social behaviour, and ultrasound vocalization during social interaction. Finally, lesions of the fastigial nuclei were histologically verified.

Results: Play behaviour and vocalization were reduced for 3 weeks after surgery in lesioned rats compared to sham-lesioned rats and controls, while young adults (10 weeks) no differences between groups were found. Locomotor activity was disturbed for 3 weeks after surgery in lesioned rats as compared to controls and for 3 days as compared to sham-lesioned rats. Motor coordination measured by rotarod and balance beam test was compromised until adulthood.

Conclusion: Deficient social behaviour and vocalization after surgery are related to lesioning of the fastigial nuclei in juvenile rats. These results indicate that similar to the human context damage of the cerebellar midline structures can reduce communicative drive and some aspects of motor activity.

Keywords: fastigial nucleus, juvenile rats, motor activity, social behaviour, ultrasound vocalization

PF-003

Oncology

Gli3 expression predicts favorable outcome and contributes to new therapeutic strategies for medulloblastoma

Junichi Yoshimura¹; Hiroaki Miyahara¹; Manabu Natsumeda¹; Ryosuke Ogura¹; Hiroshi Aoki¹; Kenichi Nishiyama¹; Masafumi Fukuda¹; Masafumi Fukuda¹; Akiyoshi Kakita¹; Yukihiko Fujii¹.

¹Brain Research Institute, University Of Niigata, Niigata.

Objective: Medulloblastomas (MB) can be subdivided into four distinct molecular variants. Abnormal sonic hedgehog (Shh) signaling is one of the mechanisms of these four molecular subgroups. We focused Gli3, a downstream protein of the Shh pathway, which is thought to regulate neuronal and glial differentiation and is speculated to play an important role for prognostic marker.

Methods: 32 newly diagnosed MBs receiving whole neuraxis radiation and/or chemotherapy from 1992 to 2010 were included. We assessed expression of Gli3 as well as histopathological findings, neuronal and glial differentiations, and also analyzed correlation between Gli3 expression and prognosis.

Results: 22 were classified as classic MBs and 10 were classified as desmoplastic/nodular (DN) MBs. There was no large cell/anaplastic MB. 17 were Gli3 positive, all cases showed neuronal differentiation and 3 showed both neuronal and glial differentiation. Histologically, 9 of 17 cases were DN MBs. However, among 15 Gli3 negative MBs, only one case showed neuronal differentiation, and another case showed DN type. 10-year overall survival (10yOS) of Gli3 positive and negative cases were 75.1% and 43.8%, respectively ($p < 0.05$). Furthermore, in the Gli3 positive cases, 10yOS of total/subtotal removal and partial removal cases were 90.0% and 60.0%, respectively. While, in the Gli3 negative cases, 10yOS of total/subtotal removal and partial removal cases were 62.5% and 29.6%, respectively.

Conclusion: The expression of Gli3 is associated with neuronal and glial differentiation in MB and histologically Gli3 expression is correlated DN MB. Gli3 expression is also correlated with good prognosis for patients with MB. Therefore, new risk stratification divided by Gli3 expression, extent of surgical resection and dissemination may contribute to new therapeutic strategies for MB.

Keywords: medulloblastoma, Gli3, prognosis

PF-004

Posterior fossa tumor

microRNA-192 suppresses leptomeningeal dissemination of medulloblastoma by modulating cell proliferation and anchoring through the regulation of DHFR, integrins, and CD47

Seung-Ki Kim¹; Seung Yeob Yang²; Seung Ah Choi¹; Kyu-Chang Wang¹; Ji Hoon Phi¹; Ji Youn Lee¹; Ae-Kyung Park³; Woong-Yang Park⁴.

¹Seoul National University Children's Hospital, Seoul; ²Dongguk University Ilsan Hospital, Ilsan; ³Sunchon National University, Sunchon; ⁴Samsung Genome Institute, Seoul.

Objective: The main cause of death in medulloblastoma is recurrence associated with leptomeningeal dissemination. Although the molecular basis of medulloblastoma has received considerable attention over the past decade, the role of microRNAs (miRs) in the acquisition of metastatic phenotype remains poorly understood. This study aimed to identify the miR involved in leptomeningeal dissemination and to elucidate its target mechanisms.

Methods: We analyzed the miR expression profiles of 29 medulloblastomas according to the presence of cerebrospinal fluid (CSF) seeding. Differentially expressed miRs (DEmiRs) were validated in 29 medulloblastoma tissues and three medulloblastoma cell lines. The biological functions of the selected miRs were evaluated using in vitro studies.

Results: A total of 12 DEmiRs were identified in medulloblastoma with seeding, including miR-192. The reduced expression of miR-192 was confirmed in the tumor seeding group and in the medulloblastoma cells. Overexpression of miR-192 inhibited cellular proliferation by targeting DHFR. miR-192 decreased cellular anchoring via the repression of ITGAV, ITGB1, ITGB3, and CD47.

Conclusion: Medulloblastoma with seeding showed specific DEmiRs compared with those without. miR-192 suppresses leptomeningeal dissemination of medulloblastoma by modulating cell proliferation and anchoring ability.

Keywords: medulloblastoma, microRNA-192, Integrins, dihydrofolate reductase, seeding

PF-005

Oncology

Pineal tumors in children. Experience of the French registry of pineal tumors.

Alexandru Szathmari¹; Blandine Grassiot¹; Pierre Aurelien Beuriat¹; Didier Frappaz²; Anne Jouvret³; Alexandre Vasiljevic³; Carmine Mottolese¹.

¹ Pediatric Neurosurgery, Neurological and Neurosurgical Hospital "Pierre Wertheimer", Lyon, Bron; ²Centre Léon Bérard Et IHOP, Lyon; ³East Neuropathological Center, Hospices Civils De Lyon, Lyon.

Objective: Tumors of the pineal tumors represent 1% of tumors of child's nervous system and represent a good model of multidisciplinary management. As they are rare, it seems interesting to as to report the pediatric series of French registry of these tumors with data collected from 26 centers.

Methods: 224 patients in pediatric age were included with 151 boys and 73 girls. 138 patients were treated for hydrocephalus: 109 by ventriculo-peritoneal or atrial shunt and 29 patients by endoscopic ventriculostomy. 70 patients had a biopsy, 23 by endoscopy, 42 in stereotactic conditions and 6 by direct approach. 124 patients underwent surgical resection by suboccipital-transtentorial either supracerebellar infratentorial paths. Histologically the analysis showed a glioma in 23 patients, a germ cell line tumor in 113, a pineal gland tumor in 54, a cyst in 20, a PNET in 1 patient, a papillary tumor of the pineal region in 6, an AT/RT in 3, an ependymoma in 1 and an undifferentiated tumor in 2 patients. In 1 patient the tumor has not been classified.

Results: Analysis of the results shows a relationship with histology with better survival for benign lesions. In patients with pinealoblastoma, survival does not exceed 25% with a particularly poor outcome for children younger than 3 years. For mixed secreting germ line tumors the overall survival is of only 15% despite additional treatment.

Conclusion: Pineal tumors represent a pathology that requires a multidisciplinary approach to improve outcomes in terms of survival and quality of life. The surgery requires a degree of expertise but the way of the approach has to be related with the surgeon's experience. As the pineal tumors are rare, a national registry of these tumors is a necessity in order to refine the knowledge, the treatment and to improve survival related their evolution and management.

Keywords: pineal tumors, pediatric brain tumors, tumor database

PF-006

Oncology

Clinical utility of serum versus CSF tumor markers in patients with CNS germ cell tumors

Arvind Krishnan¹; Edward F. Melamed¹; Erin N. Kiehna¹; J. Gordon McComb¹; Mark D. Krieger¹.

¹Division of Neurosurgery, Children's Hospital Los Angeles, Los Angeles.

Objective: The use of Alpha-fetoprotein (AFP) and Beta Human-chorionic gonadotropin (B-HCG) as markers is well established in the diagnosis and management of patients with CNS germ cell tumors (GCTs). This study aims to further characterize the predictive value of serum versus cerebrospinal fluid (CSF) levels of these markers in the diagnosis of non-germinomatous germ cell tumors (NGGCTs).

Methods: An IRB-approved retrospective review of 56 children with CNS GCTs was performed. Medical records, including serum and CSF AFP and B-HCG levels, were reviewed.

Results: All 10 patients who had elevated AFP levels in the serum also had elevated AFP levels in the CSF, and vice versa. All 9 patients who had B-HCG levels greater than 150ng/mL in the serum also had levels greater than 150 ng/mL in the CSF and vice versa. B-HCG levels less than 150ng/ml were less predictive of NGGCT pathology (i.e., these lower levels were also seen in histologically proven germinomas), but there was no differential benefit to CSF vs serum levels.

Conclusion: Serum and CSF AFP tumor marker tests are equally sensitive and specific in diagnosing germinomas and nongerminomatous germ cell tumors. The combined use of both serum and CSF levels for diagnosis does not help in the diagnosis of these tumors.

Keywords: germ cell tumor, Alpha-fetoprotein, Beta Human-chorionic gonadotropin, tumor markers, germinoma

PF-007

Oncology

Outcome and prognostic features in paediatric pineoblastomas: analysis of cases from the SEER database.

Senthil Selvanathan¹; Martin Elliot¹; Atul Tyagi¹; Paul Chumas¹.

¹Leeds General Infirmary, Leeds.

Objective: 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 clinical and pathological factors associated with outcome in paediatric pineoblastomas.

Methods: Paediatric patients (<16 years old) with pineoblastomas diagnosed between 1990–2007 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 spread and size, surgical resection and the use of adjuvant radiotherapy.

Results: Seventy-eight patients were identified from the database. Twelve patients were excluded as eleven had no surgery and one patient was excluded as the surgical status was unknown. Analysis of the remaining sixty-six patients revealed a median age at diagnosis of 5.5 years. Three patients underwent biopsy. Ten and seven patients underwent full and partial resection respectively. A further 46 patients underwent surgery the nature of which was not recorded. Thirty-nine patients (59.1%) received adjuvant radiotherapy. Eight patients (12.1%) had both surgery (Full or partial resection) and radiotherapy. The median overall survival was 40.5 months. Univariate analysis demonstrated that older age at diagnosis was the only positive predictor of overall survival.

Conclusion: This study represents the largest analysis of paediatric pineoblastomas to date. The only clinically relevant prognostic factor was older age at diagnosis. The role of surgery and adjuvant radiotherapy on overall survival remains to be defined.

Keywords: Pineoblastomas, SEER, Epidemiology, Survival

PF-008

Oncology

Identification of targets for rational pharmacological therapy in pediatric craniopharyngioma

Todd Hankinson¹; Andrew Donson²; James Johnston³; Richard Anderson⁴; Amy Rosenfeld⁵; Luca Massimi⁶; Lindsey Hoffman⁷; Michael Handler¹; Nicholas Foreman¹; Jacob Gump².

¹Childrens Hospital Colorado, Aurora; ²University Of Colorado, Aurora; ³Children's Hospital Of Alabama, Birmingham; ⁴Columbia University, New York; ⁵Phoenix Children's Hospital, Phoenix; ⁶Catholic University, Rome; ⁷Cincinnati Children's Hospital, Cincinnati.

Objective: Pediatric adamantinomatous craniopharyngioma (CPA) is a histologically benign but clinically aggressive tumor that arises in the sellar/suprasellar region. Despite a high survival rate with current surgical and radiation therapy (75–95% at 10 years), CPA is associated with significant morbidity, commonly resulting in poor quality of life (QoL) for survivors. An understanding of the unique drivers of CPA growth could help reveal effective pharmacological therapies, which could decrease morbidity and improve long-term QoL for children with CPA.

Methods: We completed mRNA microarray analysis of 15 CPA samples and 176 samples from other CNS tumors and normal brain tissue, as well as SNaPshot analysis for β -catenin and BRAF mutations. Protein levels of a selection of putative drug target genes in CPA were examined by Western blot analysis to validate the results of microarray analysis.

Results: Three patterns of significant and consistent gene product over-expression were identified in our panel of CPA relative to other pediatric brain tumors and normal brain tissue. These included 1) targets of the tyrosine kinase inhibitor Dasatinib: lymphocyte-specific protein tyrosine kinase (LCK), EPH receptor A2 (EPHA2) and v-src sarcoma (Schmidt-Ruppin A-2) viral oncogene homolog (SRC)); 2) EGFR pathway targets: amphiregulin (AREG), EGFR and ERBB3; and 3) Other pharmacological targets: Sonic hedgehog homolog (SHH) and matrix metalloproteases 9&12 Experimental drugs targeting each of these gene products are currently being tested clinically and/or pre-clinically for the treatment of other tumor types. We confirmed by western blot that a subset of these targets is highly expressed in CPA.

Conclusion: This is the first study to examine the transcriptome profile of CPA. The findings provide a rationale for further pre-clinical and clinical studies of pharmacological treatments that have previously been used for CPA.

Keywords: Craniopharyngioma, Microarray Analysis, Transcriptome, Rational Therapeutic Targets

PF-009

Oncology

Online survey on the management of paediatric craniopharyngiomas

Benedetta Pettorini¹; Barry Pizer¹; Imran Bhatti¹; Ganesalingam Narenthiran¹; Amedeo Calisto¹; Conor Mallucci¹.

¹Alder Hey, Liverpool.

Objective: Paediatric craniopharyngiomas pose a difficult challenge due to proximity of important neural structures as well as their potential for recurrence in spite of benign histology. Aggressive approach towards

surgical resection often leads to bad patient outcome in terms of quality of life whereas limited resection is fraught with its own drawbacks. There is always a question as to how much to do and what should be an acceptable outcome in each case. Although there are a number of modalities available to supplement surgery, there is no uniformity in utilising these since a lack of standardised protocols due to paucity of data on long-term outcomes of single or combination modalities.

Methods: The online survey includes two sections. One section will allow to collect basic information about neurosurgical units and the service provided for children with craniopharyngioma. The second section consists of 8 different clinical scenarios with related scans and involves a multiple choice questionnaire about management of hydrocephalus, surgical procedures, radiotherapy and recurrences treatment. The survey has been circulated in January 2014 through mailing lists involving the major international paediatric neurosurgery societies.

Results: We received 91 responses. Management of hydrocephalus received an overall agreement of 75%, surgical management of 70% and management of residual tumour of 45%. Transphenoidal approach is the first choice for resection in units with an on-site skull base surgeon. 57% of centres has departmental protocol, and 51% has designated surgeon. Majority of centres can provide an on-site skull base surgeon, endocrinologist, oncologist and neuro-ophthalmologist. Two third of units delivers a multidisciplinary clinic. Intracystic treatment is used in most centres.

Conclusion: The survey showed a significant lack of agreement in terms of choice of approach for resection and management of residual tumour. Intracystic treatment is widely used but with no agreement in terms of modality of treatment.

Keywords: craniopharyngioma, survey, surgery, radiotherapy, intracystic

PF-010

Oncology

The value of minimal invasive policy in treatment of pediatric craniopharyngioma patients

Seungwon Choi¹; Youngshin Ra¹.

¹Asan Medical Center, Seoul.

Objective: We applied minimal invasive policy in treatment of craniopharyngioma patients to improve and preserve the quality of life.

Methods: We retrospectively reviewed the medical records of children with craniopharyngioma who were treated from January, 2006 to December, 2014 in our institution (Asan Medical Center in Seoul, Korea). Twenty five patients were treated during this period and without a few exceptions, most patients had minimal invasive surgery since year 2008. We defined minimal invasive surgery as endoscopic transventricular or transsphenoid approach followed by radiosurgery. Radical surgery means conventional transcranial surgery. Patients were divided into two groups by their treatment policy, radical versus minimal invasive surgery. We compared treatment outcomes between the two subsets.

Results: Median age of patients is 8 years (1-16 yrs) and median progression free survival is 24 months (3-106 mos). No statistical significant difference is found in PFS between two groups (2yr PFS in radical surgery group 54.4% vs. minimal invasive surgery 58.3%, $p=0.315$). Tumor size is related to progression free survival (Exp (b)=1.05, 95% CI, 1.018-1.083, $p=0.02$): minimal invasive policy tends to be related in shorter progression free survival, however, this trend is not statistically significant (Exp(b)=2.387, 95% CI, 0.527-10.798, $p=0.25$). Only one patient complained of aggravated visual function and nine patients showed worsened hormonal deficiency after treatment. Hormonal function is better preserved in minimal invasive surgery group (Exp(b)= 0.001, $p=0.049$, Logistic regression analysis) and the number of hormone for replacement is fewer than the patients of radical surgery group.

Conclusion: Craniopharyngioma is a benign disease and many patients suffer from hormonal and visual deficit after radical treatment. Applying new policy – minimal invasive surgery- can help patients to have better quality of life after treatment and is also similar in disease control to radical treatment.

Keywords: craniopharyngioma, transsphenoidal, radiosurgery

PF-011

Brain malformation

Surgical management of Rathke's cleft cysts

Chandrashekar Deopujari¹; Subodh Patil¹; Nishit Shah¹; Vikram Karmarkar¹.

¹Bombay Hospital Institute of Medical Sciences, Mumbai.

Objective: Though diagnosis of Rathkes Cleft cysts (RCC) has increased due to the improvements in imaging techniques; symptomatic cases are uncommon and accurate preoperative diagnosis may sometimes be difficult. Indications of surgery are not clear and management is controversial. The aim of the study is to present the clinical and radiological findings, management options and their outcomes.

Methods: A retrospective analysis (2003-2015) of 51 consecutive cases with RCC seen in a surgical unit at our centre is presented. Twenty two surgically treated symptomatic RCCs were further evaluated for their clinical presentation, imaging characteristics, surgical approaches as well as intra-operative findings.

Results: Headache was the most common presenting complaint and was seen in almost all patients while 5 patients presented with visual deficit as the chief complaint. Hormonal abnormality was observed in 7 patients. The cyst had suprasellar extension in 17 patients. The cyst content showed hyper intense MRI signal in T1W as well as T2W images. Transsphenoidal or extended surgery for cyst excision was performed in all 22 patients. Pituitary stalk and the normal gland were preserved in all cases. Radical excision of cyst wall was not possible in 3 cases. Relief from headaches and visual deficits improved but preoperative hormonal deficiencies did not improve and new deficits were not observed.

Conclusion: Rathke's cleft cysts are an uncommon pathology with a wide spectrum of clinical and radiological features. Rathkes cleft cysts are typically benign, asymptomatic lesions that can be monitored. Although the underlying substrate is congenital, they appear to enlarge during life. In selected patients, transsphenoidal surgery provides excellent improvement in clinical symptoms and long term cyst resolution.

Keywords: Rathke's cleft cyst

PF-012

Oncology

Pediatric CNS tumors - a single Chinese center's experience

Jie Ma¹; Wei Meng¹.

¹Xin Hua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai.

Objective: The aim of this study was to describe a single centers experience in the management of pediatric CNS tumors over an 8-year period.

Methods: A total of 691 pediatric CNS tumor patients were treated in our center between January 2006 and December 2014. The presentation, location, histology, therapy, and outcome of these CNS tumors were analyzed.

Results: Female-to-male ratio was 1 : 2, and mean age was 5.9 years (range, 0.08-18). The main reason for treatment was increased head circumference in 0~3 years old children (42.3%) and headache in 3~16 years old (39.6%). The most common locations were cerebellum (21.3%),

suprasellar region (17.7%), ventricle (10.3%), brain stem (8.7%), pineal region (5.8). The most common histology were embryonal tumors (20.3%), pilocytic astrocytomas (13.2%), craniopharyngiomas (8.7%), germ cell tumors (7.1%). The main treatment methods were surgery-only (56%), surgery with chemotherapy (39%) in 0–3 years old children and surgery-only (46%), surgery with radiation (34%) in 3–18 years old. Mean follow-up duration was 49 months (range, 3–92). The 3 year PFS rate was 71.3% in 0–3 years old, and 85.9% in 3–18 years old.

Conclusion: Nearly 1/3 of pediatric CNS tumors arose below the tentorium. The suprasellar and pineal regions were frequent sites of origin for supratentorial tumors. Surgery with/without radiation or chemotherapy were the main treatment methods for pediatric CNS tumors. The 0–3 years old children had worse prognosis than bigger ones, and one of the reasons for that maybe delayed diagnosis due to short of chief complaints in infants.

Keywords: pediatric, brain, tumor, treatment

PF-013

Oncology

Assessment of emergency care for pediatric brain tumors in the Netherlands: always on a Friday

Eelke Bos¹; Oscar Eelkman Rooda¹; Eelco Hoving²; Hedy Folkersma³; Erik Van Lindert⁴; Sen Han⁵; Marie-Lise Van Veelen¹.

¹Erasmus University Medical Center, Rotterdam; ²University Medical Center Groningen, Groningen; ³Academic Medical Center Amsterdam, Amsterdam; ⁴University Medical Center Nijmegen, Nijmegen; ⁵University Medical Center Utrecht, Utrecht.

Objective: Brain and central nervous system tumors are the second most common types of cancer in childhood. In the Netherlands, approximately 120 children are affected each year. In the light of a current debate in the Netherlands on centralization of pediatric oncology, we performed a retrospective analysis of patient records from children with a brain tumor requiring neurosurgical intervention. Our aim was to assess the emergency load for pediatric neurosurgery in neuro-oncology and the differences in treatment strategy between the current five tertiary neurosurgical referral centers in the Netherlands.

Methods: Retrospectively, we collected data from all patient records of five tertiary neurosurgical referral centers in the Netherlands from 2011 to 2014. Referral pattern, symptomatology, patient and tumor characteristics, type of intervention, day of admission and time interval between presentation and surgery were analyzed.

Results: In total, 226 patient records were analyzed. Prehospitalisation delay for intra- and supratentorial tumors (respectively 10.0 weeks, SD 12.9; 9.8 weeks, SD 14.0 weeks) were not significantly different. Emergency intervention was associated with a shorter delay in high grade malignancies. 72 (30%) patients underwent emergency surgery because of raised intracranial pressure or clinical deterioration after admission. Most emergency interventions were related to hydrocephalus. Nearly half of the patients presented with hydrocephalus. 10 (4%) patients underwent immediate resection due to deterioration from tumor progression. From all patients, 156 (70%) underwent surgery within 1 week after referral. Interestingly, 23.5 % of children presented on a Friday, which was markedly higher than any other weekday (average of other weekdays 12.8 %).

Conclusion: A large proportion of pediatric neurosurgical oncology requires acute or semi-acute care which has implications for the organization of concentrated care. Our findings can guide the discussion on centralization of pediatric neuro-oncology in the Netherlands and abroad.

Keywords: emergency, oncology

Monday, 5 October 2015
14:30 – 15:30

Oncology Free Papers II

PF-014

Oncology

Neoadjuvant chemotherapy for infantile brain tumors

Hideki Ogiwara¹; Chikako Kiyotani¹; Keita Terashima¹; Kentarou Matsuoka¹; Hideto Iwafuchi¹; Nobuhito Morota¹.

¹National Center for Child Health and Development, Tokyo.

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

Methods: Retrospective review of infants who underwent tumor removal after neoadjuvant chemotherapy was performed.

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

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

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

PF-015

Oncology

The role of pre-operative plasma neutrophil-lymphocyte count ratio in predicting the histological grade of paediatric intrinsic tumours

Jamie Wilson¹; John Goodden¹; Atul Tyagi¹; Paul Chumas¹.

¹Paediatric Neurosurgery, Leeds Children's Hospital at The Leeds General Infirmary, Leeds.

Objective: Neutrophil-Lymphocyte Count Ratio (NLCR) is an established prognostic marker for renal, lung and colorectal carcinomas, and has been suggested to be predictive of histological grade in adult intrinsic primary brain tumours. The aim of this investigation was to investigate whether the NLCR in paediatric intrinsic tumours can be predictive of the final histological grade.

Methods: A retrospective analysis of all tumour surgeries for 10 years in patients less than 18 was compiled for a single UK centre. Procedures for Craniopharyngiomas, recurrent tumours and metastases were excluded from the final analysis. Pre-operative full blood counts (FBC) collected before the administration of steroids were interrogated for each patient and matched with final histological diagnosis. Statistical analysis was used to determine any significance between the NLCR of low grade to high grade tumours.

Results: From a total of 214 tumour procedures, 134 procedures for primary intrinsic tumours were identified. Preliminary data available from 36 patients was analysed to include 19 patients with low grade tumours (WHO Grade I and II) and 17 patients with high grade tumours (WHO Grade III and IV). The mean NLCR in the low grade cohort (2.56, 1.21–3.96) was not statistically significantly different from the high grade cohort (2.67, 1.90–3.48). Further analysis on the remaining 98 patients will be presented at the conference.

Conclusion: Pre-operative NLCR may be used to predict the final histological grade of adult patients presenting with primary intrinsic brain tumours, but the evidence for the use NLCR in paediatric tumour remains to be published. At the conference, we will present the full analysis and conclusions regarding the effectiveness of this measure.

Keywords: Tumour, Neutrophil-lymphocyte Count Ratio, NLCR, Grade

PF-016

Oncology

Outcomes following the change towards chemotherapy alone for the treatment of optic pathway gliomas in patients with neurofibromatosis type 1

Frazer O'Brien¹; Ian Kamaly¹.

¹The University of Manchester, Manchester.

Objective: The risk of secondary tumour formation following radiotherapy for optic pathway gliomas in NF1 has previously been described. We wished to look at the change of practice towards chemotherapy to treat these tumours.

Methods: We conducted an assessment by note review of patients from a supra regional NF1 service with a diagnosis of an optic pathway glioma (OPG). We investigated the incidence and nature of subsequent tumour development along with ophthalmic outcomes.

Results: There were 79 NF1 patients with OPG identified. 12 tumours developed in the cohort subsequent to treatment of their OPG. Of the 12 tumours, 2 patients had received chemotherapy only and both of these tumours were pilocytic astrocytomas, 2 were secondary only to radiotherapy 1 MPNST and 1 glioblastoma multiforme. The other 8 post treatment tumours came after both radiotherapy and chemotherapy. All 3 malignant tumour patients had received radiotherapy. We saw that ophthalmic follow up was well regimented with 98.4% of patients being seen at or within the designated 1 year appointment interval. Poor visual outcome was seen in 7 of 11 patients who received chemotherapy only and 3 out of 9 patients who received radiotherapy only.

Conclusion: In patients with OPG treated with chemotherapy alone there were no malignant tumours subsequently identified. There was however a high rate of poor visual outcome in these patients. Further work is required to investigate the optimum timing of chemotherapy in these patients.

Keywords: neurofibromatosis

PF-017

Oncology

Management of supratentorial hemispheric tumors in pediatric age group: CCHE experience in 35 cases.

Mohamed A. El Beltagy¹; Mostafa M. E. Atteya².

¹Neurosurgery Department, Children's Cancer Hospital Egypt (CCHE, 57357). Neurosurgery Department, Kasr Al-Ainy School Of Medicine, Cairo University, Egypt, Cairo; ²Neurosurgery Department, Children's Cancer Hospital Egypt (CCHE, 57357). Neurosurgery Department, Faculty of Medicine, Helwan University, Egypt, Cairo.

Objective: Supratentorial hemispheric tumors are not uncommon in children. Many reports highlight prevalence of low grade tumors among this subtype of tumors. Epidemiological, pathological, clinical and operative profiles still need investigation.

Methods: We reviewed medical records of 35 children with hemispheric tumors operated at Children's Cancer Hospital Egypt (CCHE-57357) between July 2007 and January 2011. Male-to-female ratio was 1.5:1.

Results: Interestingly, nineteen cases (54%) were below three years old at presentation. Most common clinical presentation was disturbed consciousness level, motor deficits and seizures in 85.7%, 65.7% and 51.4%, respectively. Sixteen cases were operated on emergency base due to increased intracranial pressure and brain herniation. High grade tumors constituted 71.4% of this series. Tumors proved to be either anaplastic ependymoma or primitive neuro-ectodermal tumors in sixteen cases (45.7%). Other pathologies included astrocytomas, oligodendrogliomas, choroid plexus papilloma/carcinoma, craniopharyngiomas, meningioma and atypical teratoid/rhabdoid tumors. Total tumor excision was achieved in 25 cases. Adjuvant chemotherapy and/or radiotherapy were instituted according to tumor pathology, extent of resection and patient's age. Cases with low grade tumors which were excised totally showed better clinical outcomes. Longstanding subdural collections were evident in thirty cases (85.7%). There were no operative mortalities in this series. Tractography was performed in six cases. Fluorescence-guided surgery with 5-aminolevulinic acid was performed in three cases. Navigated intraoperative ultrasonography was utilized to follow step-wise tumor excision in 21 cases.

Conclusion: Majority of pediatric hemispheric tumors affect children less than years old. High grade tumors constitute the major bulk of pediatric hemispheric tumors in this series. Clinical outcome is affected by extent of resection as well as tumor grade. Available technology and operative adjuncts should be tailored to maximize preserving brain functionality in those growing kids. Difficult management of hydrocephalus and subdural fluid collections are stigmata for these tumors.

Keywords: hemispheric tumors, pediatric, brain tumors, 5-aminolevulinic acid

PF-018

Oncology

Chordomas in the pediatric population. The Sick Kids experience.

Vassilios Tsitouras¹; Dhruve Jeevan¹; William Halliday²; James Rutka¹.

¹Department of Neurosurgery, The Hospital for Sick Children, Toronto; ²Department of Neuropathology, The Hospital for Sick Children, Toronto.

Objective: Chordomas are rare malignancies with a predilection for the axial skeleton and their behavior is locally aggressive.

Less than 5% of chordomas appear in children making it difficult to propose strong treatment recommendations regarding their management in this population. Accordingly, we present our experience with pediatric chordomas at the Hospital for Sick Children.

Methods: This is a retrospective analysis. We reviewed the charts of the patients diagnosed with chordomas over the last 25 years. We examined demographic and clinical data, diagnostic approaches, modes of treatment and outcomes.

Results: Ten children were diagnosed with chordomas over these years. The mean age at presentation was 10.2 years (14 months - 13 years). In 9 patients the tumor was located at the clivus - upper cervical area, and in one case at the left cerebellopontine angle. The commonest neurological manifestation was lower cranial nerve palsies which were observed in 7 patients. The mean duration of signs and symptoms was 14.8 months. The commonest surgical approach was the transoral - transpalatal approach which was used in 5 cases. A gross total resection was achieved in 4 patients (40%). The main pathological diagnosis was "classical" chordoma in 6 patients. The rest had atypical features. Nine patients received post-operative radiation therapy - in two of them, as proton beam radiotherapy. Chemotherapy was given in 2 children. Three patients (30%) died of their disease and all of them had atypical chordomas. The mean follow up period for the group was 5.8 years.

Conclusion: The optimum management of these patients mandated a multidisciplinary approach. Maximal, safe neurosurgical resection followed by appropriate adjuvant therapy were required to achieve favorable outcomes in 70% of children in this series. Atypical chordomas are a pathological variant with an exceptionally poor prognosis in the paediatric population.

Keywords: chordoma, children

PF-019

Oncology

Pediatric meningiomas: clinico-radiological profile, management strategies and long-term follow-up results of 20 cases

Sahin Hanalioglu¹; Burcak Bilginer¹; Firat Narin¹; Ruslan Yunusov¹; Nejat Akalan¹.

¹Hacettepe University Faculty of Medicine, Department of Neurosurgery, Ankara.

Objective: Pediatric meningiomas comprise 2.2% of all CNS tumors in children and they account for 1.5% of all intracranial meningiomas. This study aims at defining the clinical-radiological features, management strategies, and the clinical outcomes of meningiomas in children.

Methods: Twenty patients under 18 years of age who underwent surgery for intracranial or spinal meningiomas during the period of at 1994-2015 at Hacettepe University Hospitals, Ankara, Turkey were included in this study.

Results: The study group included 11 (55%) males and 9 (45%) females. The mean age at presentation was 12.3±4.0 years. Five (25%) had evidence of NF2. The most common symptoms at presentation were headache in 6 (30%), seizures in 6 (30%), motor weakness in 5 (25%), visual disturbances in 4 (20%). There were 18 intracranial and 2 spinal meningiomas. Convexity meningiomas were seen in 10 (50%), skull base in 7 (35%). Two (10%) intraaxial and one (5%) intraventricular tumor was noted. Giant meningiomas were observed in 7 (35%) patients. Six (30%) patients had multiple meningiomas. Gross total excision was achieved in 13 (65%) and subtotal in 7 (35%). Pathological investigation revealed WHO grade I in 11 (55%), grade II in 7 (35%), and grade III in 2 (10%) patients. Patients with higher grade, residual tumor or recurrence underwent reoperation and/or adjuvant therapy. After a mean follow-up of 113 months, 16 patients (80%) were neurologically intact, 3 (15%) were having moderate disability, and 1 (5%) patient died.

Conclusion: Although rare, pediatric meningiomas may pose clinical challenges for neurosurgeons particularly in the setting of giant, complex tumors. Higher grade is seen more frequently compared with adults. Taking into account higher rates of recurrence, safe gross total resection should be the goal. However, in higher grade meningiomas and residual/recurrent tumors, radiotherapy may be beneficial to control the disease.

Keywords: meningioma, pediatric, children, childhood, surgery

Tuesday, 6 October 2015

08:30 – 10:00

Hydrocephalus Free Papers III

PF-020

Hydrocephalus

The cerebral surfactant system and it's role in brain water circulation

Preuss Matthias¹; Schob Stefan¹.

¹University Leipzig, Leipzig.

Objective: Recently, surfactant proteins have been detected as inherent proteins of the CNS, present in cerebrospinal fluid and in tissues of the central nervous system. Aim of the study was to determine normal values and differences in SP concentrations between hydrocephalus patients of various etiologies.

Methods: A total number of 159 CSF specimens of 125 patients have been examined after completion of clinical diagnostics. Patients with aqueductal stenosis, iNPH, pseudo tumor cerebri and CSF infection have been identified.

Results: Under normal conditions, SP-A, B, C and D are present in CSF independent from age. Concentrations of surfactant proteins did not show any association to age. Total CSF protein concentrations were not elevated compared to controls but showed a significant reduction in AQS and VIIP patients (p=0.01). Acute types of hydrocephalus showed higher SP-A concentrations than the chronic NPH group. SP-C levels were highest in pressure-active forms hydrocephalus and not elevated in NPH patients. SP-B was highly significantly elevated in patients with bacterial CSF infection. The findings of highly significant elevations of SP-A and SP-C in states of CSF disturbance are in concordance with the pulsatile vector theory and other CSF circulation concepts that base on pulsatile nature of CSF flow. The necessity of adequate inherent CSF viscosity characteristics and pulsatility within the CNS to maintain a brain water zero net balance underlines

Conclusion: A functional Cerebral Surfactant System exists. Surfactant Proteins A, B, C and D are present under physiological conditions in human CSF. Collectin-type SP-A is elevated in CSF disturbances with ventricular enlargement. Hydrophobic SP-C seems to increase when intracranial pressure elevation is present. SP-D did not show any significant changes in hydrocephalus. SP-B seems to be involved in inflammatory response when CSF infection occurs.

Keywords: brain water circulation, surfactant system, hydrocephalus, pseudotumor cerebri, CSF infection

PF-021

Hydrocephalus

Prosaika: the programmable gravitational device for hydrocephalus shunting: a prospective multicenter study

Uwe Kehler¹; Michael Kiefer²; Regina Eymann²; Wolfgang Wagner³; Christoph Tschan⁴; Niels Langer¹; Veit Rohde⁵; Hans Ludwig⁵; Jan Jan

Gliemroth⁶; Ullrich Meier⁷; Johannes Lemcke⁷; M. Javad Mirzayan⁸; Martin Schuhmann⁹; Alexandra Huthmann⁴.

¹Neurosurgery Hamburg, Hamburg; ²Neurosurgery Homburg, Homburg; ³Pediatric Neurosurgery Mainz, Mainz; ⁴Neurosurgery Meppen, Meppen; ⁵Neurosurgery Göttingen, Göttingen; ⁶Neurosurgery Lübeck, Lübeck; ⁷Neurosurgery Berlin, Berlin; ⁸Neurosurgery Hannover, Hannover; ⁹Neurosurgery Tübingen, Tübingen.

Objective: Cerebrospinal fluid (CSF) overdrainage is a major problem in shunt therapy for hydrocephalus. The adjustable gravitational valve proSA allows for the first time a targeted compensation for overdrainage in the upright position without interfering with the differential pressure valve. To evaluate benefit, safety and reliability, the multicenter prospective observational study PROSAIKA was conducted in 10 German neurosurgical centers.

Methods: Between March 2009 and July 2010, 120 hydrocephalic patients undergoing first time shunt implantation or shunt revision using proSA entered the study. 91 patients completed the 12 months follow-up.

Results: Hydrocephalus symptoms were improved in 86%, unchanged in 9% and deteriorated in 3%. In 51%, the proSA opening pressure was readjusted one or several times to treat suspected suboptimal shunt function, this resulted in clinical improvement in 55%, no change in 25%, and deterioration in 20% of these patients. The one year censored proSA shunt survival rate was 89%. Device related shunt failure was seen in 2 cases.

Conclusion: This is the first clinical report on the implantation of the adjustable gravitational valve proSA with a follow-up of 12 months in a substantial number of patients. Irrespective of different hydrocephalus etiologies and indications for shunt surgery, the overall results after 12 months were very satisfying. The high frequency of valve readjustments underlines the fact that preoperative selection of the appropriate valve opening pressure is difficult. The low number of revisions and complications compared to other valves proves that proSA implantation adds no further risk; this valve is reliable, helpful and safe.

Keywords: Hydrocephalus, Cerebrospinal fluid, VP-Shunt, Overdrainage, Gravitational valve

PF-022

Hydrocephalus

Re-evaluating weekend effect on hydrocephalus patients undergoing operative shunt intervention

Frank Attenello¹; Eisha A. Christian¹; Timothy Wen²; Steven Cen³; Gabriel Zada¹; Erin N. Kiehna⁴; Mark D. Krieger⁴; J. Gordon McComb⁴; William J. Mack¹.

¹Department of Neurosurgery, Keck School of Medicine, University of Southern California, Los Angeles; ²Keck School of Medicine, University of Southern California, Los Angeles; ³Department of Preventative Medicine, Keck School of Medicine, University of Southern California, Los Angeles; ⁴Division of Neurosurgery, Children's Hospital Los Angeles, Los Angeles.

Objective: Recently published data has suggested an increase in adverse outcomes in the pediatric ventricular cerebrospinal fluid diversion shunt population with same day weekend shunt revisions. We undertook an evaluation of the impact of weekend admission and time-to-shunting on surgery-related quality outcomes in pediatric patients undergoing ventricular shunt insertion or revision.

Methods: Pediatric patients with hydrocephalus undergoing ventriculoperitoneal, -atrial, and -pleural shunts were selected from the 2000-2010 Nationwide Inpatient Sample and Kids Inpatient Database. Multivariate regression analyses were used (adjusting for patient, hospital, case severity, and time-to-shunting) to determine differences in rates

of inpatient mortality and routine discharge among patients admitted on the weekday versus the weekend.

Results: There were 99,771 pediatric hydrocephalus shunt patients, with 16% admitted on weekends. Following adjustment for disease severity, time-to-procedure and admission acuity, weekend admission was not associated with an increase in inpatient mortality or change in percentage of routine discharge for ventricular shunt insertion ($p>0.05$). Additionally, associations were unchanged when evaluating patients who received shunt revision surgery ($p<0.05$). High volume centers were incidentally noted to show decreased inpatient mortality and increased rates of routine discharge in multivariate analysis ($p<0.05$).

Conclusion: Contrary to prior studies, our data suggests that weekend admission was not associated with poorer outcomes for ventricular shunt insertion or revision surgery. Better outcomes were noted at high volume centers.

Keywords: nationwide inpatient sample, time-to-surgery, weekend admission, ventriculoperitoneal shunt, cerebrospinal fluid

PF-023

Hydrocephalus

Success of repeat ETV procedure according to ventriculostoma reclosure pattern in children

Volkan Etus¹; Hakan Karabaglı²; Murat Geyik³; Umit Celakil¹; Aykut Gokbel¹.

¹Kocaeli University, Faculty of Medicine, Department of Neurosurgery, Kocaeli; ²Selcuk University Selcuklu, Faculty Of Medicine, Department of Neurosurgery, Konya; ³Gaziantep University, Faculty of Medicine, Department of Neurosurgery, Gaziantep.

Objective: The reclosure of the ventriculostoma is one of the major causes of failed ETV procedure in children. The present study investigates the relation between ventriculostoma closure patterns and success rate of the repeat-ETV procedure in pediatric cases.

Methods: The data of 51 pediatric triventricular hydrocephalus cases belonging to two centers which, have undergone repeat ETV due to failure of initial ETV procedure were retrospectively analyzed. Of the 51 cases 16 were under 2-years of age. All cases were examined with cine phase-contrast magnetic resonance imaging, which is regarded as an accurate method of detecting postoperative fenestration patency. Patient records on clinical features and video recordings of the endoscopic procedures were reviewed. The cases with a history of intraventricular hemorrhage, CSF-infection or CSF-shunt surgery were not included. The cases with incompletely penetrated membranes during the initial ETV procedure were also excluded from the study. Cases were grouped according to our previously described classification of “ventriculostoma closure pattern”. Type-1: Closure of ventriculostoma with gliosis or scar tissue that results in opaque floor; Type-2: Narrowing/closure of ventriculostoma by translucent membranes; Type-3: Patent ventriculostoma but CSF flow is blocked by newly formed reactive membranes in the basal cisterns.

Results: The interval to ETV failure was 12 days to 34 months (median 2.3 months). Repeat-ETV failed in 15 of the 51 cases (29.4%). According to our classification groups, success rates of the re-ETV procedure were as follows: Type-1: 3 in 14; Type-2: 7 in 20 and Type-3: 5 in 17.

Conclusion: The success of re-ETV was lowest in Type-1 closure pattern group. The cases with Type-2 closure pattern seem to benefit most from re-ETV procedure. Also our data reflect a higher tendency of Type-3 closure pattern in cases under 2-years of age.

Keywords: ETV, ventriculostoma, closure

PF-024

Hydrocephalus

Early endoscopic ventricular irrigation for the treatment of intraventricular hemorrhage and hydrocephalus in neonates

Volkan Etus¹; Hakan Karabaglı²; Gokmen Kahilogullari³; Agahan Unlu³.

¹Kocaeli University, Faculty of Medicine, Department of Neurosurgery, Kocaeli; ²Selcuk University, Selcuklu Faculty of Medicine, Department of Neurosurgery, Konya; ³Ankara University, Faculty of Medicine, Department of Neurosurgery, Ankara.

Objective: Neonatal intraventricular hemorrhage (IVH) usually results in posthemorrhagic ventriculomegaly which remains a challenge since this condition may evolve into loculated hydrocephalus. This multicenter study describes the approach of early neuroendoscopic lavage for the treatment of ventriculomegaly/hydrocephalus following IVH in newborns and compares the results to cases which have been treated conventionally.

Methods: The data of total 49 neonatal posthemorrhagic hydrocephalus cases were retrospectively analyzed. Between 2010 and 2015, 21 neonates with posthemorrhagic hydrocephalus underwent early endoscopic ventricular irrigation for removal of intraventricular blood remnants (Group-A). 28 neonates were treated conventionally, initially using temporary CSF diversion via lumbar punctures, a ventricular access device, or an external ventricular drain (Group-B). Complications, shunt dependency rates, and incidence of multiloculated hydrocephalus were evaluated retrospectively.

Results: Group-A and Group-B cases did not differ significantly regarding gestational age and birth weight. No neuroendoscopic procedure related complications were observed in Group-A. Among 21 cases which underwent early endoscopic ventricular irrigation 13 (62%) required a later shunt insertion, as compared with 27 (96, 4%) of the 28 infants who were treated conventionally. Group-A cases were associated with significantly fewer infections as well as significantly lower incidence for supratentorial multiloculated hydrocephalus development.

Conclusion: Results of the present study seem to demonstrate that early removal of intraventricular blood degradation products and residual hematoma via neuroendoscopic ventricular irrigation is feasible and safe for the treatment of posthemorrhagic hydrocephalus in neonates with IVH. This procedure seems to offer significantly lower shunt rates and fewer complications such as infection and development of multiloculated hydrocephalus in those cases.

Keywords: endoscopic, hydrocephalus, hemorrhage, intraventricular, neonate

PF-025

Other

A retrospective analysis of the limits of the endoscopic fenestration of arachnoid cysts

Paolo Frassanito¹; Gianpiero Tamburrini¹; Luca Massimi¹; Concezio Di Rocco²; Massimo Caldarelli¹.

¹Pediatric Neurosurgery, Catholic University Medical School, Rome; ²INI, Hannover.

Objective: Endoscopy plays a pivotal role in the treatment of the arachnoid cysts, though indication to this treatment option should be carefully evaluated on a case by case basis. Accordingly, factors affecting the outcome of the endoscopic treatment should be identified.

Methods: We performed a retrospective analysis of the consecutive cases affected by intracranial arachnoid cyst treated endoscopically at our Institution in the period 2004-2014.

Results: We collected 65 arachnoid cysts in 64 patients. Site of the cyst was: intraventricular (19), sylvian (16), interhemispheric (11), sovrasellar (5), infratentorial (5), cisterna ambiens (4), quadrigeminal plate (3) and cortical (2). The cyst was fenestrated in the cisternal spaces and/or in the ventricular cavities according to the site and the anatomical relationships of the cyst to the CSF pathway. In most complex cases, the endoscopic trajectory was planned with the aid of the neuronavigation system. No intraoperative complication was recorded. Radiological exams showed a decreased size of the cyst and/or patent fenestration in all patients except 6 cases: 5 required redo endoscopy and 2 finally the placement of a cysto-peritoneal shunt.

Conclusion: The impossibility to plan a safe and effective endoscopic trajectory remain the main contraindication to the endoscopic treatment. Other conditions that may negatively affect the outcome of the endoscopic fenestration are: infratentorial location (in particular retro-cerebellar and ponto-cerebellar), radiological and/or intraoperative evidence of scarring of the subarachnoid spaces, large size cysts involving more than one intracranial compartment with limited communication with the ventricles or communication with cistern other than basal ones (e.g. cisterna ambiens, cistern of the quadrigeminal plate).

Keywords: arachnoid cyst, endoscopy, fenestration

PF-026

Other

Efficacy of fenestration in pediatric middle fossa arachnoid cysts

Wei Meng¹; Jie Ma¹.

¹Xin Hua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai.

Objective: Middle fossa arachnoid cysts are the most common type in children. Fenestration using endoscopy or microscopy is now the main method of treatments, but its efficacy is not definite. This study aims at evaluating efficacy of fenestration in middle fossa arachnoid cysts through symptomatic and radiographic terms.

Methods: A retrospective analysis of 72 middle fossa arachnoid cyst patients who received endoscopic (40 patients) or microsurgical (32 patients) fenestration treatments from January 2009 to January 2014 in our hospital and have ≥ 1 year follow-up. We assessed symptomatic and radiographic changes before and after surgery, also compared outcomes between endoscopic and microsurgical treatments.

Results: Mean age was 4.77 years old. Mean follow-up duration was 24.11 months. Symptomatic improvement rate was 46% (23/50). Radiographic improvement rate was 76.39% (55/72), of which 34 cysts (47.22%) disappeared, 21 cysts (29.17%) were significantly reduced. Symptomatic and radiographic improvements were not significantly correlated. A total of 10 patients were seen with complications (7 patients had Galassi III type cysts), including two cases with subdural hemorrhage (one case improved with conservative treatment, another need draining), 8 patients with chronic subdural effusion (6 cases improved with conservative treatment, 2 cases need draining). Outcomes between endoscopic and microsurgical treatments is not significant. (P=0.53).

Conclusion: Fenestration using endoscopy or microscopy is an effective treatment for middle fossa arachnoid cysts. Symptoms can be improved to some extent, and radiographic improvements are obvious. There are some complications (mostly in Galassi III type), which can be relieved often by conservative treatment.

Keywords: arachnoid cyst, middle fossa, fenestration, treatment

PF-027

Brain malformation

Comparative analysis of bleeding risk by the location and shape of arachnoid cysts: a finite element model analysis

Chang-Hyun Lee¹; In Seok Han²; Ji Yeoun Lee³; Ji Hoon Phi³; Seung-Ki Kim³; Young-Eun Kim²; Kyu-Chang Wang³.

¹Konyang University Hospital, Daejeon; ²Dankook University, Yongin;

³Seoul National University Children's Hospital, Seoul.

Objective: Although arachnoid cysts (ACs) are observed in various locations, only sylvian ACs are frequently regarded to be associated with bleeding. The reason for this selective association of sylvian ACs with bleeding is not well understood. The objective of this study was to investigate the effect of the location and shape of ACs on the risk of bleeding.

Methods: A finite element model of the head/brain of a normal male was developed and modified for models of sylvian, suprasellar, and posterior fossa ACs. A spherical AC (ideal shape) of the same volume and outer wall area was placed at each location to compare the effect of AC location. Models of shallow bowl-shaped (actual shape) ACs were developed to compare the effect by shape. The shear force in the spot-welds (SFSW) was measured between dura and the outer wall of the ACs or the comparable arachnoid membrane in the normal model.

Results: By location, the SFSW of the posterior fossa was the highest among the normal models, and that of the sylvian AC was the highest among AC models except on posterior impact. By shape, the SFSW in the posterior fossa AC increased in proportion to its outer wall area, and that of the sylvian AC decreased in proportion to its outer wall area. In hierarchical regression analysis, presence of an AC contributed to 78.6% of SFSW, and the location and shape of the AC did not exhibit a significant contribution.

Conclusion: Among the risk factors tested, only the presence of an AC is a significant risk factor for subdural hemorrhage. The reason why AC bleeding is frequently observed in sylvian ACs may be related to factors other than location or shape, such as their increased incidence or abundance of bridging veins.

Keywords: arachnoid cyst, finite element, bleeding, location, shape

PF-028

Brain malformation

Support and management system for fetal brain malformation

Mami Yamasaki¹; Chika Teramoto²; Takumi Yamanaka¹; Atsuko Harada¹.

¹Department of Pediatric Neurosurgery, Takatsuki General Hospital, Takatsuki; ²Department of Nursing, Takatsuki General Hospital, Takatsuki.

Objective: Hydrocephalus related disease and brain malformation have been increasingly diagnosed prenatally due to the development of diagnostic technology. However, in the early stage of development morphological diagnosis is not still established and the long-term prognosis is still unclear. Therefore it is very difficult to make the precise diagnosis and inform the parents for proper outcome in prenatal period. In our hospital, we started a multi-discipline prenatal support team that consists of doctors, nurses, midwives, physical therapist, psych-therapist, medical social worker and perinatal coordinator. The purpose of this team is to discuss how to manage and support the fetus and families. From Apr 2012 till now, we conducted 90 cases. The aim of this paper is to analyze how to manage fetal brain malformation and to

clarify the benefits and problems of multi-discipline prenatal support team.

Methods: Of the 90 cases, 48 cases with CNS disorders are the materials in this paper.

Results: These include 16 cases of myelomeningocele, 7 case of hydrocephalus, 5 cases of cranial bifida, 5 cases of Dandy-Walker syndrome, 7 cases of lissencephaly, microcephaly and 7 other disease. Management was divided into three ways. The first way is to prepare and perform the aggressive treatment after birth. The second way is to refrain from the aggressive treatment after birth because of poor prognosis and the third way is to terminate the pregnancy before 21st weeks of gestations. In any ways, it is very important to make precise diagnosis using fetal sonogram, MRI and chromosomal molecular analysis. The full time perinatal coordinator is very important for keeping this work.

Conclusion: In every decision, accurate diagnosis and full support by a multi-discipline prenatal support team are required.

Keywords: brain malformation, prenatal diagnosis, fetal MRI, genetic analysis, family support

Tuesday, 6 October 2015

12:15 – 12:30

PF-029

Hydrocephalus

International infant hydrocephalus study: initial results of a prospective, multicentre comparison of endoscopic third ventriculostomy (ETV) and shunt for infant hydrocephalus

Shlomi Constantini¹; Abhaya Kulkarni²; Orna Friedman¹; Spyros Sgouros³.

¹Tel-Aviv Medical Center, Tel-Aviv; ²Hospital For Sick Children, Toronto; ³"Mitera" Childrens Hospital, Athens.

Objective: IIHS is an international, prospective, multicentre study to compare ETV and shunt in infants with symptomatic triventricular hydrocephalus from aqueductal stenosis.

Methods: IIHS utilized a prospective comprehensive cohort design. Patients received either an ETV or shunt, based on their randomization or parental preference. Patients were followed prospectively for time to treatment failure, defined as the need for repeat CSF diversion procedure (shunt or ETV). Survival analysis was used to compare time to failure for ETV versus shunt.

Results: A total of 158 patients met eligibility criteria (median age at surgery 3.6mths) across 27 centres in 4 continents. Since only 52 patients (32.9%) were randomized, all 158 patients were analyzed together (115 ETV, 43 shunt). Actuarial success rates for ETV vs shunt at 3, 6, and 12 months were: 68% vs 95%, 66% vs 88%, and 66% vs 83%. The 6 month ETV success rate of 66% was slightly higher than would have been predicted by the ETV Success Score (57%). The hazard ratio for time to treatment failure favoured shunt over ETV (3.17, 95% CI 1.45-6.96, p=0.004), after adjusting for age at surgery, history of previous hemorrhage or infection, continent, and randomization status. Patients younger than 6 months of age appeared to do relatively worse with ETV than older patients.

Conclusion: The IIHS has provided the first prospective comparison of ETV and shunt for infant hydrocephalus. These initial results suggest that shunting has a superior success rate compared to ETV, although the success rate for both was relatively high. This patient cohort continues to be followed in IIHS and we will await the results of the important primary outcome of health status at 5 years of age.

Keywords: Hydrocephalus

Wednesday, 7 October 2015
09:00 – 10:30

CVJ Free Papers IV

PF-030

Craniovertebral Junction

Role of neurosurgery for the management of chondrodysplasia punctata

Nobuhito Morota¹; Satoshi Ihara¹; Hideki Ogiwara².

¹Tokyo Metropolitan Children's Medical Center, Tokyo; ²National Center for Child Health and Development, Tokyo.

Objective: Chondrodysplasia punctata (CDP) is a clinically and genetically heterogenous group of rare bone dysplasias. Role of surgery still remains controversial.

Methods: The authors operated on 7 children with CDP between 2007 and 2012. Clinical features of those children and surgical outcomes were retrospectively analyzed.

Results: All 7 children were male, aged 1 month to 2 years (median: 6 months old) at the initial surgery. There were 6 CDPX2 (Brachy telephalangic type) and the lesion was craniovertebral junction (CVJ). Five occipitocervical fusion followed by Halo external fixation and 1 C1 laminectomy were performed. Staged surgery (C1 laminectomy at 2 months old and the posterior fusion at 1 year old) was required in 1 child who had diagnosed cervical cord compression prenatally. Newly developed cervical dislocation caudal to the occipitocervical fusion forced a child additional posterior fusion 2 years after the initial one. All 6 children with CDPX2 showed improved motor function after surgery. However, respiratory assist is necessary for 3 of them. Another 1 child with CDP tibia-metacarpal type had severe cervical stenosis. Despite decompression, he showed no functional improvement and died by respiratory insufficiency 1 year later.

Conclusion: Surgical decompression and fusion for CVJ lesions in CDP seems rewarded in terms of life-saving and functional improvement. Surgery is not straightforward, and staged or repeated procedure could be required. Postoperative management using Halo external fixation can face risk of skin complications. Since CDP has variety of clinical courses, relationship between surgical outcomes and the CDP subtype needs to be examined in future.

Keywords: Chondrodysplasia punctata, craniovertebral junction, cervical decompression, posterior cervical fusion

PF-031

Craniovertebral Junction

The natural history of cervical spinal cord compression in mucopolysaccharidosis: evidence for asymmetric disease progression

Gurish A. Solanki¹; Saikat Santra¹; Suresh Vijay¹.

¹Departments of Paediatric Neurosurgery and Inborn Errors of Metabolism, Birmingham Children's Hospital NHS Foundation Trust, Birmingham.

Objective: Patients with Mucopolysaccharidosis (MPS) are at high risk of neurological complications because of spinal involvement, principally cervical spine instability and compression leading to myelopathy. Atlantoaxial subluxation due to Odontoid hypoplasia and ligamentous laxity leads to instability while deposition of glucosaminoglycans (GAG) in ligaments, joints and dura lead to compression. Patients harbour a combination of abnormalities with

varying severity. There are currently no studies detailing the timing of progression from cord instability and compression to myelopathy. Aims: To evaluate the natural history of progressive spinal cervical cord compression in MPS estimated by regular clinical, functional and radiological evaluations.

Methods: Retrospective review between January 2003-December 2014 of MPS I (Hurlers), MPS IV (Morquio-Brailsford) and MPS VI (Maroteaux-Lamy) children. Of the 24(12 boys, 12 girls) children, 20 children had MPS IV, one had Hurler-Schie and three MPS VI. The average age at diagnosis was 3.6 (boys 3.9 and girls 3.3) years. Instability, compression, deformity and progressive reduction in the spinal canal was measured over time.

Results: Radiological instability and compression were present in 83%, deformity in 54%. Speed and severity of progression varied. Half of children underwent surgery. The time interval (in months) to development of instability (19 vs 51), cord compression (11 vs 43), CSF signal loss (21 vs 51), and cord signal change (31 vs 79) were significantly shorter in the operated group.

Conclusion: The radiological progression of spinal cord compression falls into two distinct groups. These findings correlate with clinical disease pattern, identify genotype-phenotype sub-groups and indicate the optimal window for intervention. We propose that this should ideally be at the stage of loss of CSF signal before development of cord compression or signal change within the cord in these children who have compromised spinal cord perfusion due to concomitant systemic disease.

Keywords: Natural History, cervical, spine, C1-C2, atlanto-axial

PF-032

Craniovertebral Junction

Reversed rotatory atlantoaxial subluxation due to lateral proatlas failure: introducing a new abnormality

Sandip Chatterjee¹.

¹Park Clinic Kolkata, Kolkata.

Objective: Anomalies at the craniovertebral junction need to be evaluated on the basis of their embryological development before a decision about appropriate treatment can be instituted. This is a review of 6 cases of torticollis with rotatory atlantoaxial subluxations occurring as a compensatory measure to a lateral tilt produced by non-development of the lateral mass of atlas/occiput on one side as a failure of the development of the lateral proatlas elements. Here the axis rotates round a fixed atlas as a compensatory mechanism, hence called "reversed atlantoaxial rotatory subluxation."

Methods: We encountered 6 cases of this anomaly which can easily be missed. 4 of these presented with progressive torticollis and neurodeficit within the first 5 years of life, and the other 2 had symptoms after trivial trauma. The first patient, a child of 1 months age had complete absence of the lateral mass of the atlas as well the occipital condyle on one side, associated with a very unusual location of a bony mass extending from the posterior arch of the axis to the clivus, whose embryological origin can best be postulated about, and was probably an abnormality of the hypochordal bow.

Results: The first 3 patients underwent occipito-cervical fusion using specially modified contoured rods. In all 3 cases, paraparesis improved after surgery. In 2 cases, opening up and fusion of the atlanto-axial joint on both sides were performed with over distraction on side, and in the 6th case atlantoaxial distraction was produced using laminar hooks on the posterior arch of the atlas with transpedicular screws into the axis.

Conclusion: The unifying factor in all these 6 children has been the maldevelopment of the lateral sclerotome of the proatlas with or without

accompanying maldevelopment of the C1 segment, resulting in the "reverse" rotatory subluxation.

Keywords: AARF, reverse atlantoaxial rotatory subluxation

PF-034

Craniovertebral Junction

Low velocity craniocervical junction injuries in children and young adults with Os Odontoideum

Ashraf Megahed¹.

¹Consultant Neurosurgeon, Mansoura University Hospital, Mansoura.

Objective: We looked at our patients harboring Os Odontoideum (O O) who sustained low velocity injury to the the craniocervical junction (CCJ) to test the hypothesis that; those patients have the propensity for more neurological damage with minor trauma than subjects who do not have O O.

Methods: We examined a cohort of 9 consecutive patients with O O who presented after having a low velocity CCJ injury e.g. (endotracheal intubation, carrying moderate weight over the head, mild blow of the head). The clinical presentation, radiological findings and surgical outcome were retrospectively evaluated. They were 4 females and 5 males, with a mean age of 18 years (14-29) with a median follow up period of 27 months (6-109). 7 out of our 9 patients had C1-2 fixation with bone graft (Harms technique), while the other 2 had occipitocervical fixation.

Results: Neck pain was present in all patients, prior history of neurological manifestations in 7/9 patients. Post traumatic neurological deficits were present in all patients (transient in 4 patients and permanent in 5 patients). Radiologically, C1-2 subluxation was present in 5/9 patients. Widened inter C1-2 interspinous distance was demonstrated in plain radiographs of 4/9 patients. Postoperatively, three out of the 4 patients who had permanent neurological deficits improved partially, while one improved completely. All the 5 patients with transient deficit remained neurologically intact. Solid fusion was achieved in all cases.

Conclusion: Patients with O O might be at a higher risk of developing neurological deficits following low velocity trauma than other subjects who do not have O O. C1-2 screw/rod fusion with bone graft seems to be an effective and safe procedure for this group of patients. Given the limitations of this retrospective small case series, a larger multicenter prospective trial to evaluate this cohort of patients is warranted.

Keywords: craniocervical, atlantoaxial, subluxation, odontoid, c1-c2 fusion

PF-035

Craniovertebral Junction

The transoral approach of the occipitocervical junction in children. Report of 15 cases in more than 20 years of surgical experience

Alexandru Szathmari¹; Blandine Grassiot¹; Pierre Aurelien Beuriat¹; Carmine Mottolese¹.

¹Pediatric Neurosurgery, Neurological and Neurosurgical Hospital "Pierre Wertheimer", Lyon, Bron.

Objective: The trans-oral approach allows direct access to the lower region of the clivus and the anterior part of the occipitocervical junction. Reports of pediatric series by this approach are scarce in the literature. We report our experience with 15 pediatric cases over a period of 23 years.

Methods: Between 1990 and 2013, 15 children were operated by a trans-oral route. The average age was 8 with limits between 2 and 17 (7M and 8F). The initial clinical presentation was represented by spinal pain in 12 cases and a stiff neck in 3 cases. The pathology was represented by a complex malformation of the atlanto occipital joint (hinge) in 12 cases (odontoideum bones (3), basilar impression (2) or dysplasia with instability (7)) and by tumoral lesions in 3 cases. All patients benefited of posterior fixation by wiring, plates or with rods-plate and inter-laminae hooks techniques. The interventions were performed widely under microscope but recently two patients were operated by pure endoscopic technique and 2 other by microscope assisted endoscopy

Results: In all cases the decompression has been satisfactory, allowing neurological improvement. In 3 patients with a tumor, complete resection was possible but followed by a LCS fistula in 1 case resolved with a temporary external lumbar drainage. We evidenced early disunion of the mucosa in 2 cases that required revision.

Conclusion: The trans-oral route may be feasible in children with very good results and low morbidity. Tumor pathology poses the same problems in adults for dural closure but mucosal repair and feeding is earlier in children. The endoscopic approach is possible but requires a good skill of the technique. We prefer the endoscope-assisted technique which ensures better control bleeding.

Keywords: craniocervical junction, transoral approach, endoscopy, craniocervical instability in children

PF-036

Craniovertebral Junction

Comparison of three different surgical techniques of craniocervical decompression in children with Chiari I malformation: the Edinburgh experience.

Pasquale Gallo¹; Drahoslav Sokol¹; Jothy Kandasamy¹.

¹Royal Hospital for Sick Children, Edinburgh.

Objective: A broad spectrum of posterior fossa decompression techniques (PFD) are employed for the treatment of Chiari I malformation (CM). They range from simple bony decompression to more invasive approaches involving dura mater opening with or without duroplasty and cerebellar tonsils resection. The aim of this study is to compare the clinico-radiological results of three different PFD performed in a single paediatric centre.

Methods: A retrospective analysis of children with CM treated between 2008 and 2014 was performed. Three different surgical techniques were randomly offered to the patients according to the surgeon's preference during this time period: a bony decompression (BD), a BD plus opening of the dura mater without duroplasty (DOWD) or with duroplasty and tonsils coagulation (DPTC). Presenting symptoms, radiological findings, clinico-radiological outcome and complications rate of the three different treatments were evaluated.

Results: Forty-six children underwent 51 PFD: 17 BD, 17 DOWD and 17 DPTC. Twenty-seven (58.6%) patients had a spinal cord syrinx and twenty-three (50%) had an associated scoliosis. The follow-up period ranged from 6 to 90 months. Preoperative symptoms improved in 94% DPTC patients, 82% BD patients and 53 % DOWD patients. Post-operative complications were detected in 7 DOWD patients (41%) including new syrinx development (2 cases), hydrocephalus (2 cases), meningitis (1 case), liquorrhoea (1 case) and bilateral subdurals (1 case). There were 2 cases of liquorrhoea in the DPTC group (11%) and no post-operative complications in the BD group. Overall the syrinx had a significant resolution in 90.3 % of the children: 100% (13/13) in the DPTC group, 85% (11/13) in the DOWD group and 80% (4/5) in the BD group.

Conclusion: Despite the syrinx outcome is comparable among the three techniques, the risk of post-operative complications and no improvement

of pre-operative symptoms performing a dural opening without duroplasty was significantly higher.

Keywords: Chiari Malformation, Bony decompression, Duroplasty, Syringomyelia, Outcome

PF-037

Craniovertebral Junction

Suboccipital craniectomy for Chiari I malformation: an update

Luca Massimi¹; Paolo Frassanito¹; Gianpiero Tamburrini¹; Concezio Di Rocco²; Massimo Caldarelli¹.

¹Pediatric Neurosurgery, Catholic University Medical School, Rome; ²INI, Hannover.

Objective: The surgical treatment of Chiari I malformation and associated syringomyelia includes several different techniques with various degrees of invasiveness. Less invasive procedures are found to provide good results with a lower rate of complications but a higher risk of reoperation compared with most invasive techniques. The goal of this study is to assess the effectiveness of suboccipital craniectomy alone in children with a long-term follow-up.

Methods: Forty children (23 females, 17 males; mean age: 6.5 years) operated on by bone decompression alone and with a minimum 5-years follow-up are retrospectively reviewed. All patients underwent foramen magnum decompression by means of suboccipital craniectomy and a resection of the epidural fibrous at occipito-cervical junction. Thirty-two children also required C1 laminectomy while a dural delamination was performed in a half of cases according to intraoperative ultrasounds.

Results: The mean current follow-up is 11.3 years (5-15 years). Head and/or neck pain was the most frequent preoperative symptom (56.7%), followed by upper and lower extremity weakness (20.0%), ataxia (20.0%) and vertigo (27.7%). Syringomyelia was present in 19 patients. A significant improvement of preoperative clinical symptoms and signs was observed in 33 patients (82.5%). Three children required adjunctive surgery for symptoms recurrence (7.5%). Neuroimaging revealed minor postoperative modifications in most cases regardless of tonsils location, while syringomyelia was reduced in size in 79% of the cases. Complication rate and length of hospital stay were significantly reduced compared with the literature data and our own experience using more invasive techniques.

Conclusion: Suboccipital craniectomy and C1 laminectomy is an effective, safe and long-lasting treatment for symptomatic children with Chiari I and associated syringomyelia. A certain risk of symptoms recurrence requiring new surgery exists.

Keywords: chiari I, syringomyelia, suboccipital craniectomy, C1

PF-038

Craniovertebral Junction

Bone-only Chiari decompression failure rate is no different than that of open duroplasty.

Robert Keating¹; Michelle Feinberg¹; Tiffani Defreitas¹; John Myseros¹; Suresh Magge¹; Chima Oluigbo¹.

¹Childrens National Medical Center, Washington.

Objective: Controversy continues to surround the optimum surgical approach to Chiari decompression and whether the dura needs to be opened. Assessment of long-term outcomes looking specifically at failure rates,

and associated factors, for bone-only decompression vs duroplasty was undertaken.

Methods: Retrospective review of patients undergoing decompression from 1996-2014(18yrs) at CNMC identified patients requiring additional Chiari decompression for worsening symptoms or persistent syringomyelia (IRB #Pro268). Preoperative symptoms, imaging studies, operative reports and post-operative follow up were available for all included patients.

Results: 19/195 (9.74%) patients were identified requiring additional Chiari decompression. Average age at initial surgery 9.4 yrs (1-17) and 10.3 yrs (3-20) for second surgery. Length of time between surgeries was 2.8 years (4m to 8 years) with follow-up 47m (1-224). Patients undergoing bone-only decompression demonstrated 10/70 (14%) need for additional Chiari surgery whereas 9/125 (7%) of patients s/p duroplasty required second operation (OR: 2.14, CI: 0.82-5.571, p=0.11). The syrinx cohort demonstrated a similar failure rate (OR: 2.04, CI: 0.577-7.21, p=0.26). Analysis of holocord syringes was also not significant (OR: 2, CI: 0.43-9.2, p=0.36). Factors contributing to reoperation for both surgical cohorts found inadequate bony decompression in 2/19, bone regrowth 3/19 and arachnoid scarring at 4th ventricular outflow in 17/19 patients. 10/19 (53%) required placement of 4th ventricular stent. 6/19 (32%) had craniofacial co-morbidity. Complications were seen in 2/70 (3%) for bone-only decompression vs 26% (CSF leak/14; pseudomeningocele/26; meningitis/4, p<0.001).

Conclusion: Comparison of Chiari failures does not appear to differentiate between open and closed decompression. The most common cause of failure was the presence of arachnoid scarring at the 4th ventricular outflow in both surgical cohorts. Craniofacial co-morbidity increased the likelihood of surgical failure, especially when hydrodynamic issues were involved.

Keywords: chiari malformation, duroplasty, surgery failures, open vs closed Chiari repair

Wednesday, 7 October 2015

11:00 – 12:00

Spine Free Papers V

PF-039

Spine malformation

Philosophy for management of split cord malformation

Prof. Ashok Kumar Mahapatra¹.

¹All India Institute of Medical Sciences, Bhubaneswar, Odisha, (India) 751019, Bhubaneswar.

Objective: Split Cord Malformation (SCM) is a rare condition and only few large series are published in the world literature.

Methods: Over 25 years, we have operated more than 300 cases of SCM of various types.

Results: In 2004-05, we sub classified SCM to A, B, C and D categories depending on the length of the split cord and position of the spur and space available below and surroundings of the spur. We also operated asymptomatic patients as the risk of deterioration is less than 5%. In 25 years, we have operated 35 patients asymptomatic cases. Improvement was recorded 55% and stabilisation 41% and in 4% cases there was deterioration. Amongst those deteriorated, the maximum deterioration was observed in 1 D Sub type.

Conclusion: In our experience, all SCM patients must be operated even the patient is asymptomatic. The chance of deterioration is less than 5%, hence it is better to take the risk and do the best.

Keywords: Split cord malformation, Good outcome

PF-040

Spine

Treatment of persistent urinary incontinence in children with possible occult tethered cord syndrome with section of the filum terminale: a randomized controlled pilot study

Paul Steinbok¹; Koroush Afshar²; Jeanne Landgraf³; Walter Hader⁴; Vivek Metha⁵; Andrew Macneily².

¹Division of Neurosurgery, BC Children's Hospital and University of British Columbia, Vancouver; ²Division of Urology, BC Children's Hospital and University of British Columbia, Vancouver; ³Healthactq Inc, Boston; ⁴Division of Neurosurgery, University of Calgary and Alberta Children's Hospital, Calgary; ⁵Division of Neurosurgery, University of Alberta and Stollerly Children's Hospital, Edmonton, Edmonton.

Objective: The concept of Occult Tethered Cord Syndrome (OTCS), in which there is normal neuroanatomic imaging in the face of clinical and urodynamic evidence of neuropathic bladder behavior is controversial. Uncontrolled series report improvement in bladder function following section of the filum for presumed OTCS. However, the natural history of bladder dysfunction is known to improve with time. We performed a pilot randomized controlled study to compare symptomatic medical treatment only to surgical section of the filum plus medical treatment in children with presumed OTCS.

Methods: Children refractory to standard medical management for at least 1 year under the care of a pediatric urologist with normal conus position on MRI and abnormal urodynamics were randomized. Exclusion criteria included: other neurologic conditions, spinal dysraphism, known bladder outlet obstruction, atonic bladder or other lower urinary tract pathology. Patients were assessed at randomization and 1 yr later with a validated dysfunctional elimination score (DES), a standardized urodynamic score (UDS), and a validated enuresis specific quality of life scale (PEMQOL).

Results: After 8 years, the study accrued 21 patients. DES improved in the medical and surgical arms (13% and 10% respectively) and UDS deteriorated slightly (16% and 5% respectively). These differences between groups were not statistically significant. The child and family impact scale of the PEMQOL improved slightly in both groups. A utility analysis indicated that >300 patients in each arm would be required to demonstrate statistical difference between the two groups with respect to DES and UDS.

Conclusion: There appears to be no objective difference in urologic outcome between medical management plus or minus filum section for patients with refractory dysfunctional voiding and a normal conus position. These data challenge the existence of the concept of OTCS, wherein urologic dysfunction is attributed to tethering by the filum despite a normally located conus.

Keywords: tethered cord, filum section, occult tethered cord syndrome, spina bifida

PF-041

Spine malformation

A rat model of chronic syringomyelia by epidural compression of the lumbar spinal cord

Ji Yeoun Lee¹; Saet Pyoul Kim¹; Seung-Ki Kim¹; Ji Hoon Phi¹; Dachling Pang²; Kyu-Chang Wang¹.

¹Seoul National University College of Medicine, Seoul; ²Oakland Medical Center, Kaiser Foundation Hospitals of Northern California, Oakland.

Objective: The research on the pathophysiology of syringomyelia has been focused on Chiari malformation, trauma, and

inflammation. To understand the pathophysiology of syringomyelia associated with occult spinal dysraphism, a novel animal model of syringomyelia induced by chronic mechanical compression of the lumbar spinal cord was created.

Methods: The model was made by epidural injection of highly-concentrated paste-like kaolin through windows created by partial laminectomy of L1 and L5 vertebrae. Behavioral outcome in terms of motor and urinary function was assessed serially for 12 weeks. Magnetic resonance images (MRI) were obtained in some animals to confirm the formation of syrinx. Immunohistochemistry (IHC) studies were done to evaluate the degrees of inflammatory reaction, demyelination, and cell death in the spinal cord surrounding the syrinx cavity.

Results: By 12 weeks after the operation, syringomyelia formation was confirmed in 85% of the rats (34 out of 40) on H & E staining and MRI. Without exception, the syrinx cavities were just rostral to the epidural compression. Motor deficit of varying degrees was seen immediately after the procedure in 28% (11 out of 40) of the rats. In 13 rats (33%), lower urinary tract dysfunction was seen. Motor deficit improved by 5 weeks after the operation, whereas urinary dysfunction mostly improved by 2 weeks. There was delayed mortality of 13% (5 out of 40) at 1 month or later and 3 of the 5 died from a new onset of urinary retention. At 12 weeks after the operation, IHC showed no inflammatory process, demyelination, or accelerated apoptosis in the spinal cords surrounding the syrinx cavities similar to sham-operated animals.

Conclusion: A novel experimental model for syringomyelia by epidural compression of the lumbar spinal cord has been created. It will serve as an important research tool to elucidate the pathogenesis of this type of syringomyelia.

Keywords: syringomyelia, animal model, epidural compression, chronic

PF-042

Spine malformation

Fetal surgery for myelomeningocele in Western Europe: some thoughts on ethical, legal, psychological issues based on the first 16 cases performed in our department

Frank Van Calenbergh¹; Mehrnaz Dingar¹; Katrien Jansen¹; Gunnar Naulaers¹; Luc Decatte¹; Roland Devlieger¹; Tim Van Mieghem¹; Jan Deprest¹.

¹University Hospital Leuven, Leuven.

Objective: To define and clarify some of the complex ethical, legal and psychological issues associated with fetal-maternal surgery for myelomeningocele (MMC) in a Western European country where over 75% of patients opt for termination of pregnancy (TOP) after prenatal diagnosis. TOP therefore almost seems to be the standard of care.

Methods: Since the publication of the results of the MOMS trial, pregnant women have three possible management options after prenatal diagnosis of MMC: prenatal surgery, postnatal surgery and termination of pregnancy. The MOMS trial has shown significant benefit for the child (smaller rate of hydrocephalus and Chiari type II malformation, higher rate of walking at 30 months of age), but treatment options obviously only included prenatal and postnatal surgery. We reviewed parental decisions in patients who were referred for counseling about prenatal surgery.

Results: In collaboration with the Childrens Hospital of Philadelphia and Kinderspital Zurich, we have started offering the option of fetal surgery since 2012. 40 mothers from 7 European countries and from Australia were assessed. After counseling 28 % of mothers opted for termination, whereas 36 % opted for postnatal and 38 % for fetal surgery. There was no relationship between the level of the lesion and parental decisions. The complex legal, ethical and psychological issues will be illustrated. It is

obvious that the relatively narrow time window, psychological pressures and unrealistic expectations of fetal surgery make parental decisions difficult.

Conclusion: Despite being a highly selected and referred population our transnational patients still do not lift the option of fetal surgery after counseling by a multidisciplinary fetal surgery team. Almost one out of three parents, despite considering fetal surgery, eventually opt for TOP.

Keywords: ethical, myelomeningocele, fetal surgery

PF-043

Spine malformation

Prenatal myelomeningocele repairs in an existing fetal surgery center: oversight and initial experience

Michael Handler¹; Kenneth Liechty¹; Henry Galan¹; Michael Zaretsky¹; Nicholas Behrendt¹; Rony Marwan¹; Corbett Wilkinson¹; Mariana Meyers¹; David Mirsky¹; Timothy Crombleholme¹.

¹University of Colorado, Aurora.

Objective: The report of the Management of Myelomeningocele Study (MOMS) substantially increased the interest of both mothers to seek fetal surgery and centers to perform prenatal myelomeningocele (MMC) repair. Our institution began offering the operation at a busy maternal-fetal center in a large childrens hospital under protocol-driven evaluation and rigorous oversight. We report our experience to date.

Methods: Records of the Childrens Hospital Fetal Care Center were reviewed from 3/2013 to 4/2015, and from among the various fetal interventions, prenatal closures of fetal myelomeningocele were identified. Each had been approved for operation by an independent oversight committee, adhering to selection criteria from MOMS. Infants were delivered at this and other medical centers, with late follow up for babies outside of this region through local providers, and reported to us. Shunts were placed at the discretion of local neurosurgeons.

Results: In a 25 month period, we have performed 31 cases. Gestational age for the first 25 was between 24 and 25 6/7 weeks, with a change to earlier interventions (22 2/7 - 24 1/7 weeks) later in the series. 50% of fetuses delivered at ≥ 33 weeks. Fetal MRI two weeks after repair in 23 showed resolution of hindbrain herniation in 7 (30%), improvement in 9 (40%) and no change in 7 (40%). Shunts were placed in 9/24 (37.5%) infants delivered to date. There was one death from sepsis in an infant delivered at 26 weeks. One infant required reoperation at one day of age due to leak. In contrast to MOMS, there were no maternal complications, and 100% of hysteratomies healed completely.

Conclusion: A program for evaluation and oversight of prenatal myelomeningocele repairs may be safely instituted in the context of a pre-existing fetal surgery center. Results comparable to MOMS centers can be achieved, and in certain measures exceeded.

Keywords: myelomeningocele, fetal surgery, Chiari malformation, hydrocephalus

PF-044

Spine malformation

Percutaneous minimally invasive fetoscopic surgery for spina bifida Aperta: first cases of Turkey

Ibrahim Alatas¹; Huseyin Canaz¹; Ali Gedikbasi²; Kerem Ozel¹; Ayten Saracoglu³; Tolga Saracoglu³; Nursu Kara; Thomas Kohl⁵.

¹Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Department of Perinatology, Kanuni Sultan Suleyman Hospital, Istanbul; ³Department of Anesthesiology and Reanimation, Florence Nightingale Hospital, Istanbul Bilim University,

Istanbul; ⁴Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ⁵German Center for Fetal Surgery, Justus Liebig University Gießen, Frankfurt.

Objective: Myelomeningocele (MMC), one of the most common congenital malformations, can result in severe lifelong disabilities, including paraplegia, hydrocephalus, Chiari II malformation, incontinence, sexual dysfunction, skeletal deformations and mental impairment. MMC was the first nonlethal anomaly to be treated by fetal surgery. The first fetoscopic approaches for spina bifida closure in humans by maternal laparotomy, uterine exteriorization, and transuterine trocar placement were reported in the United States by Bruner et al. and Farmer et al. Both teams abandoned their fetoscopic approaches because early results were disappointing due to technical difficulties. Since then, fetoscopic fetal surgery for spina bifida has been replaced by an open surgical approach performed currently for several hundreds of human fetuses in North America and also for some fetuses in Brazil, Poland, Spain and Switzerland. Percutaneous minimally invasive fetoscopic approach was defined by Thomas Kohl, to minimize maternal trauma from the open fetal surgical approach. Our aim is to present the first two cases of Turkey and their short term results.

Methods: Percutaneous fetoscopic patch closure was attempted for repair of myelomeningocele in two fetuses with L5 and L4 lesions respectively, by a surgical team consists of a perinatologist, pediatric neurosurgeon and a pediatric surgeon. Procedures were performed at respectively 25+2 and 25+4 weeks of gestation. Fetuses were delivered respectively at 31+1 and 31+3 weeks of gestation.

Results: Their neural cords were completely covered although in small areas skin closure was incomplete. Complete skin closure occurred beneath an occlusive draping within 4 to 6 weeks.

Conclusion: Both neonates showed reversal of hindbrain herniation, near-normal leg function, and satisfactory bladder and bowel function. Ventriculoperitoneal shunt insertion was not required for both neonates.

Keywords: Fetoscopic surgery, short term results, spina bifida

Wednesday, 7 October 2015

14:50 – 15:00

PF-045

Education and Training

The role of mixed reality simulation in pediatric neurosurgical training

Giselle Coelho¹; Samuel Zymberg²; Marcos Lyra³; Benjamin Warf⁴; Nelci Zanon¹.

¹Pediatric Neurosurgery Center CENEPE, São Paulo; ²Department of Neuro Paulista School of Medicine, Federal University of São Paulo, São Paulo; ³Department of Gynecology, Federal University of Pernambuco, Recife; ⁴Department of Pediatric Neurosurgery - Boston Children Hospital / Harvard Medical School, Boston.

Objective: The Neurosurgical education is a long, laborious process, requiring many years of supervised directed, hands-on training. The development of surgical simulation platforms is essential to reduce the risk of potentially serious intraoperative errors arising from inexperience. The main goal: to propose a new tool for pediatric neurosurgical education, associating virtual and physical simulation (mixed reality) for craniostylosis correction and for neuroendoscopic training.

Methods: Tridimensional videos were developed by 3DS Max program. The simulated approaches were: the Reniers H technique,

choroid plexus cauterization and endoscopic thirdventriculostomy. The physical simulators were built with a synthetic thermo-retractile and thermo-sensible rubber which, when combined with different polymers, produces more than 30 different formulas. These formulas present textures, consistencies and mechanical resistance similar to many human tissues.

Results: Fiberglass moulds in the shape of the skull constitute the basic structure of the cranosynostectomy simulator. It is possible to perform the biparietal remodeling used in scaphocephaly correction. All aspects of the procedure can be simulated. The presence of superior sagittal sinus can simulate emergence situations with bleeding. Regarding the neuroendoscopic model, silicon and fiberglass moulds, in the shape of the cerebral ventricles. There is the possibility for practicing neuroendoscopic techniques such as: to navigate in the ventricular system, to perform thirdventriculostomy, choroid plexus cauterization, and to resect intraventricular “tumors”. Fifteen experienced neurosurgeons evaluated these simulators performance. The proportion of the answers were estimated (confidence intervals).

Conclusion: The experts conclude that this virtual simulation provides a highly effective way to work with 3D data and it significantly enhances teaching of surgical anatomy and operative strategies in neurosurgical field. A mixed simulation provided the desired attributes of both physical and virtual simulators in meeting the psychomotor and cognitive apprenticeship objectives. The combination of these tools has the potential to improve and abbreviate safely the non experienced learning curve.

Keywords: Virtual, Physical, Simulation, Neurosurgery, Training

Wednesday, 7 October 2015

16:10 – 17:30

Trauma and Vascular Free Papers VI

PF-046

Trauma

MRI in mild pediatric traumatic brain injury: diagnostic overkill or useful tool?

Gesa Cohrs¹; Johannes T. Hensler²; Michael Synowitz¹; Maximilian Mehdorn¹; Friederike F. Knerlich-Lukoschus¹

¹Department of Neurosurgery, Universitätsklinikum Schleswig-Holstein, Campus Kiel, Kiel; ²Department of Radiology and Neuroradiology, Universitätsklinikum Schleswig-Holstein, Campus Kiel, Kiel.

Objective: cMRI (cranial magnetic resonance imaging) is a sensitive well-established imaging tool lacking the burden of radiation exposure; yet it is not established as primary diagnostic tool in traumatic brain injury (TBI). This study is supposed to evaluate the usefulness and practicability of MRI in the setting of acute TBI in the pediatric population.

Methods: Our institutional electronic database was screened for patients (age up to 18 years) admitted for mild TBI (between 1/2009 and 4/2015). Following criteria were analyzed retrospectively: Initial GCS (Glasgow Coma Scale), presence of confusion or lethargy, loss of consciousness, post-traumatic amnesia, neurological abnormalities (i.e. focal signs, speech disturbances), vomiting, skull hematomas, GCS deterioration within two hours. Cranial MRI was obtained within 24 hours after trauma with T2-, T2*-weighted gradient-echo (“heme”-sequence), susceptibility (SWI)-, diffusion/apparent diffusion coefficient (DWI/ADC) and T2 FLAIR (fluid attenuated inversion recovery)-weighted sequences, and were evaluated for feasibility and time expenditure.

Results: 494 patients were included (female/male ratio 218/276). Age groups: 0-1 years: 2%, 1-6 years: 22%, 6-12 years: 39%, 12-18 years

36%. GCS was 15 in more than 90%, GCS-deterioration appeared in 1%. Trauma spectrum included falls (48%), sport injuries (20%), pedestrian struck by vehicle (10%), motor vehicle crash (7%), physical attack (3%), other mechanism of injury (12%). Sedation for cMRI was needed in 1%; intubation in 2%. Additional computertomography scans were obtained in 3.8% (mostly due to craniofacial injuries). In 98% axial T2- and in 89% coronal T2*-sequences could be obtained. In 13.5% MRI revealed trauma-associated findings. Incidental findings appeared in 4.9%.

Conclusion: The MRI-protocol, established in our clinic, is a feasible evaluation tool in mild pediatric TBI. Though diagnostic output in our examined collective was relatively low, in case of clinical doubt cMRI is a safe imaging alternative avoiding long-term risks associated with cranial computertomography.

Keywords: mild TBI, MRI

PF-047

Trauma

Surgical management and long term outcome of growing skull fractures

Muhammad Zubair Tahir¹; Dominic Thompson¹

¹Great Ormond Street Hospital, London.

Objective: Growing skull fracture (GSF) is a rare late complication of paediatric head injury. The predispositions for GSF are incompletely understood. The purpose of this study is to review an institutional experience of GSF in an attempt to better define at risk children and to review the results of surgical repair using full thickness autologous calvarial grafting.

Methods: The study is a retrospective review of prospectively collected surgical data from 1991-2014. All patients undergoing surgery for repair of GSF were identified. Demographic data, mode of injury, initial radiographic findings were assessed in each case. The operative technique was reviewed for each case and clinical and radiographic results at last follow up were recorded.

Results: Twenty-eight children underwent surgery for GSF. Age at time of surgery ranged from 2 months to 4 years. The initial injury leading to GSF included fall (8 cases), non-accidental injury (5 cases), and birth trauma (4 cases). The other less common causes were road traffic accident, TV trolley injury, and post-surgery. Out of 28 cases, 4 were located in frontal region while rest were parietal. Surgical techniques included split calvarial grafts in four patients while rest had full thickness bone grafting. In all cases with full thickness graft the donor site was covered with morsel bone chips mixed in fibrin glue. Three patients required VP shunt insertion. One patient required redo cranioplasty with titanium after failure of first operation and one required VP Shunt revision.

Conclusion: In this series the risk factors for the development of GSF were age < 2 years at time of injury and linear diastatic or comminuted fracture with underlying brain injury. Full thickness autologous graft and covering the donor site with morsel bone chips mixed in fibrin glue has not been previously reported and provided good bone integration and cosmetic results.

Keywords: Growing Skull Fracture

PF-048

Trauma

Surgical results of decompressive craniectomy in very young children- a level one trauma centre experience from India

Deepak Gupta¹; G Lakshmi Prasad¹; B S Sharma¹; Ak Mahapatra¹.

¹All India Institute of Medical Sciences, Delhi.

Objective: The purpose of this study was to quote our tertiary care institutional experience in dealing with such young children and highlight the timing, effectiveness and complications associated with surgical decompression

Methods: This study was conducted to identify very young children, all aged ≤ 3 years with TBI undergoing DC for intracranial hypertension at the apex trauma centre in India between Jan 2008 and April 2014. Analysis included demographic profile, trauma details, clinico-radiological details, surgical procedures, complications and outcomes. ICP monitoring was continued for a minimum of 72 hours and a maximum of 120 hours, if required. Enteral feeding through a nasogastric tube was started within 24 hours of surgery.

Results: Total patients were 71. Mean age was 1.6 years. Mean duration from injury to surgery was 11.9 hours (3–80 hours). Around 50% were severe head injury. Intracranial pressure (ICP) monitoring was done in 33 patients. Mean ICP was 22.2 mm Hg (9–50 mm Hg). The threshold ICP for surgery was 15 mm Hg. Perioperative mortality was 50% each for severe TBI (18/36) and diffuse cerebral edema (7/14) and 58% for infants (4/7). Ninety percent of expired patients had ICP >20 mm Hg. Mean follow up duration was 19.6 months (range 2–42 months). Except one, all survivors had good to excellent outcomes (GOS E–7,8).

Conclusion: Decompressive craniectomy offers survival advantage in 50% of young children with severe TBI and should be used judiciously. Highest mortality was within 1st week of surgery. The cut-off limit of 20 mm Hg for surgical decompression might not be applicable to young children and low threshold ICP needs to be considered. Factors associated with increased mortality are high opening ICP (>20 mm Hg), GCS <8 , diffuse cerebral edema and infant age group. Timing of DC remains crucial. Further prospective studies are necessary to optimize the timing and ICP limit for surgical decompression.

Keywords: severe head injury, pediatric, decompressive craniectomy

PF-049

Vascular

Utility of vessel wall MR imaging in pediatric neurovascular disease
Adele Ricciardi¹; Charles Matouk¹; Michael Diluna¹.

¹Yale University School of Medicine, New Haven.

Objective: High-resolution MR vessel wall imaging (MR-VWI) is increasingly being used to characterize intracranial vascular disease. Early reports have demonstrated its potential utility in differentiating quiescent from active intracranial atherosclerosis, reversible cerebral vasoconstriction syndrome from central nervous system vasculitis, and ruptured or “leaking” from unruptured aneurysms. We hypothesized that vascular wall enhancement would demonstrate utility in treatment of pediatric neurovascular disease.

Methods: High-resolution MR-VWI was routinely performed for pediatric patients treated in a combined neurovascular operating room immediately before definitive treatment of the ruptured intracranial aneurysm, AV fistula or AVM. MR imaging was performed by using a 3.0T scanner with a 32-channel head coil. The vessel wall protocol included time-of-flight MR angiography and T1-weighted black blood vessel wall sequence (turbo spin echo acquisition) before and after intravenous administration of gadolinium (with constant scan parameters). Each vessel wall sequence was performed in the axial and coronal planes. Pre- and postgadolinium subtracted T1-weighted black blood vessel wall images were also acquired.

Results: We report for the first time high-resolution MR-VWI in four pediatric patients with intracranial hemorrhage from ruptured AVMs and AVFs. Three patients harbored high risk features including intranidal aneurysm or arterio-venous fistulae. The suspected point of rupture demonstrated thick vessel wall enhancement in all cases. In one illustrative case, treatment of the high risk features (pre- and

intra-nidal aneurysms), but not the area of thick vessel wall enhancement (AVM nidus) resulted in immediate morbidity. None of the associated unruptured aneurysms demonstrated this MR imaging finding.

Conclusion: High-resolution MR-VWI identified the site of rupture in pediatric patients with ruptured AVMs, including those patients harboring multiple AVM high risk features. It may represent a useful tool in the investigation of intracranial hemorrhage from AVM rupture.

Keywords: vessel wall imaging, vascular malformations, rupture

PF-050

Vascular

Cavernous malformations of central nervous system in pediatric patients: a single centered surgical experience in 47 patients.

Dattaraj Sawarkar¹; Pankajkumar Singh¹; Deepak Gupta¹; Sumit Sinha¹; Deepak Agrawal¹; Gurudutta Satyarthee¹; Manmohanjeet Singh¹; Ashish Suri¹; P Sarat Chandra¹; Shashank Kale¹; Bhawani Sharma¹; Rajinder Kumar¹.

¹All India Institute Of Medical Sciences (AIIMS), New Delhi, New Delhi.

Objective: To assess the outcomes of micro-surgical excision of pediatric brain and spinal cavernous malformations (CMs).

Methods: We retrospectively enrolled all the operated CMs patients from 1st January, 2001 to 31st December 2014. Data was analyzed for their clinical features and outcome.

Results: Total 47 patients of CMs (25 supratentorial, 13 infratentorial, 3 intraorbital and 6 spinal) with mean age of 13.8 yrs. (3–18 yrs.) were enrolled into the study. Most of these patients (76.6%) were male. Size varied from 1.2 to 6.6 cm. Two patients had multiple CMs. Symptoms of CMs were site specific. Seizure was the most common symptom (68%) of CMs at supratentorial location followed by headache 48% & neurodeficit 20%, while all brainstem and spinal CMs had some form of neurodeficit. Intraorbital CMs had proptosis with vision impairment. History of acute hemorrhage was present in 20% of supratentorial CMs, 44.4% of brainstem CMs and 50% of spinal CMs. We had followup ranged from 6 months to 162 months. All supratentorial CMs patients showed improvement in these symptoms. Patients with preoperative seizure, showed good seizure control with Engel scale I in 16 (94.1%) patients and Engel scale II in 1 (5.9%) patient. In infratentorial & spinal CMs patients, there was 92.3% & 66.7% improvement in their neurodeficit respectively. McCormick score for spinal CMs patients improved in 4, declined in 1 and remained same in 1 patient. Post operative morbidity occurred in 6 patients (12.7%) including 3 patients (6.38%) with permanent new deficits, 2 patients with meningitis and one patient with pseudomeningocele. However, there was no mortality in our series.

Conclusion: Microsurgical excision of CMs results in excellent neurological outcome with excellent seizure control and improvement in neurological deficits with acceptable morbidity and no mortality.

Keywords: pediatric population, cavernous malformations, microsurgery, outcome

PF-051

Vascular

Surgical strategy of deep-seated large cavernomas in children

Reizo Shirane¹; Tomomi Kimiwada¹; Toshiaki Hayashi²; Teiji Tominga³.

¹Miyagi Children's Hospital, Sendai; ²Sendai City Hospital, Sendai;

³Tohoku University, Sendai.

Objective: Cavemoma was thought to be a benign vascular hamartoma caused by developmental malformations of the vascular bed, it is

recognized that lesions may increase in size. In children, subacute worsening of neurological symptoms is commonly observed. Such lesions should be removed totally in this paper, we discuss surgical strategy and present videos of deep-seated large cavernoma.

Methods: Six pediatric cases will be presented. Cavernoms were located in the thalamus, thalamus to midbrain, cerebellum to medulla and basal ganglia. Staged surgery was performed in two cases. The most commonly presenting symptom was progressive motor weakness (5 cases) and epilepsy was observed in one. Radiological signs of acute hemorrhage were observed in 5 cases. Trans cortical approaches through a small cortical window were applied to supra-tentorial cavernomas.

Results: Postoperatively, there was no additional neurological deficit, and all patients gradually improved. In a case of basal ganglia cavernoma, there was a need for reoperation due to rebleeding.

Conclusion: Trans cortical approach is a valid choice for the removal of deeply seated cavernomas. Via this approach, tumors can be removed without significant sequelae related to the surgery. Total resection is associated with patients' clinical improvement in deep-seated large cavernomas.

Keywords: cavernoma, surgery, children, deep seated, hematoma

PF-052

Vascular

Pediatric intracranial aneurysms – a single institution experience on 43 consecutive cases.

Alexandru Tascu¹.

¹"Carol Davila" University of Medicine and Pharmacy, Clinical Hospital "Bagdasar-Arseni", Bucharest.

Objective: Aneurysmal rupture is extremely rare in children. Cerebral aneurysms in pediatric population is not an usual pathology, therefore the management of these cases may be difficult sometimes. Facing such pathology needs a well-trained and experienced pediatric neurosurgical and anesthesiologist team. The authors present their experience in pediatric cerebral aneurysms' management.

Methods: A retrospective analyze of 43 pediatric cerebral aneurysms treated in the same institution in the last 15 years (2000-2014). Sex-ratio showed a male preponderance: 26 boys (60%) and 17 girls. Median age: 13,6 years old. The majority of our cases (34) have presented with subarachnoid hemorrhage. In our database we have used H&H scale for SAH evaluation having as results: H&H 1: 6 cases (17,6%); H&H 2: 18 cases (52,9%); H&H 3: 7 cases (20,5%); H&H 4: 3cases (8,8%) and no H&H5 cases. There were 2 cases presented as intracranial hypertension syndrome, 5 cases presented with intra-cerebral hematoma, and 2 incidental discovery cases. The main clinical features were represented by headache: 98%, neck stiffness: 94%, vomiting: 92%, neurological deficits: 78%, altered level of consciousness: 67%. Aneurysms' location – ICA: 5 cases (11,6%), ACoA: 14 cases (32,5%), MCA: 13 cases (30,2%), PCoA: 8 cases(18,6%), posterior circulation: 3 cases (6,9%).

Results: In our series 40 cases have been treated by microsurgical clipping, one case of multiple aneurysms underwent endovascular treatment, one case deceased during pre-operative evaluation due to a respiratory arrest, and one case was an incidental discovery witch refused any treatment. Follow-up and outcome database showed: good recovery in 34 cases (82,9%), no severe disabilities and no mortality.

Conclusion: Intracranial aneurysms in pediatric population represent a very rare pathology being a considerable challenge for all pediatric neurosurgeons. Early microsurgical approach and long-term follow-up, by clinical and imaging surveillance, should be mandatory in aiming best results.

Keywords: aneurysm, pediatric, subarachnoid hemorrhage

PF-053

Vascular

Moyamoya disease: clinical outcome after indirect revascularization in Saudi patients

Anwar Ul Haq Dr¹; Essam Al Shail Dr¹.

¹King Faisal Specialist Hospital and Research Centre, Riyadh, Riyadh.

Objective: Although moyamoya disease was first observed in Japan, it is increasingly being observed throughout the world, affecting children and adults. Only sporadic cases cases of moyamoya disease has been reported from Saudi Arabia. This is first study describing the surgical treatment of moyamoya disease from Saudi Arabia. Moyamoya disease, if left untreated, can result in overwhelming permanent neurological and cognitive deficits. Medical treatment of Moyamoya disease is ineffective. Only effective treatment is surgical revascularization of ischemic brain. Two types of procedure are used. Direct and indirect revascularization. We used indirect method of revascularization known as Encephaloduroarteriosynangiosis (EDAS). Objective of study is to describe the clinical presentation, diagnosis, surgical management and outcome of Moyamoya disease in Saudi patients.

Methods: The study was conducted at Department of Neurosciences, King Faisal Specialist Hospital and Research Centre. Clinical presentation, etiology, diagnosis, procedure of indirect revascularization and outcome of patients with Moyamoya disease between Jan 2006 to Dec 2014 were recorded.

Results: 20 patients were included in the study. 10 were males and 10 females. Most common age of presentation was 6-10 years. The most common presentation was TIA, recurrent strokes followed by seizures, neuro-cognitive decline and chronic headache. The cause of moyamoya disease was idiopathic in 11, sickle cell disease 3, thalassemia major 2, post radiotherapy 1, neurofibromatosis 1 and glycogen storage disease 1 and protein C deficiency in 1 patient. 27 EDAS were done. 8 patients underwent bilateral EDAS and 11 patients underwent unilateral EDAS. 12 patients return to normal life, 3 patients have activity with mild limitation, 2 patients have activity with severe limitation and 3 patients became vegetative with progressive disease. Seizures improvement was seen in 50% of patients

Conclusion: Encephaloduroarteriosynangiosis (EDAS) is simple, safe and effective method of surgical revascularization for patients with Moyamoya Disease.

Keywords: Moya Moya Disease, Moya Moya Syndrome, EDAS, pediatric stroke, sickle cell disease

Wednesday, 7 October 2015

16:10 – 17:30

Epilepsy and Functional Free Papers VII

PF-054

Epilepsy

Brain somatic mutations in MTOR cause focal cortical dysplasia type II in human and mouse

Dong-Seok Kim¹; Jae-Seok Lim²; Hoon-Chul Kang³; Eun Kyung Park¹; Heung-Dong Kim³; Jeong Ho Lee²; Seung Woo Park⁴.

¹Pediatric Neurosurgery, Severance Children's Hospital, Department of Neurosurgery, Yonsei University College Of Medicine., Seoul; ²Department of Biological Sciences, KAIST, Daejeon; ³Pediatric Neurology, Severance Children's Hospital, Department of Pediatrics, Yonsei University College of Medicine, Seoul; ⁴Department of Neurosurgery, Kangwon University College of Medicine, Chun Cheon.

Objective: Focal cortical dysplasia type II (FCDII) is a developmental malformation of cerebral cortex and an important cause of medically refractory epilepsy. FCDII sporadically occurs, and this condition is characterized by dysmorphic neurons and disrupted cortical lamination in affected cortical regions. It has been hypothesized that FCD is caused by somatic mutations in affected regions. However, no such mutations have been identified. Here, we report de novo somatic mutations of MTOR in the affected brains of FCDII patients.

Methods: Deep whole exome sequencing (the median read depth of 492×) of paired brain-blood DNA from 4 FCDII patients revealed brain somatic mutations in 3 patients including MTOR c.4448G>A (p.Cys1483Tyr), MTOR c.7255G>A (p.Glu2419Lys) and c.7280T>C (p.Leu2427Pro). We also performed deep targeted sequencing (the median read depth of 135,424×) of the codons encoding mTOR p.Cys1483, p.Glu2419, and p.Leu2427 residues in brain tissues obtained from an additional 76 FCDII patients.

Results: In total, we identified 13 FCDII patients carrying somatic missense mutations in MTOR including mTOR p.Cys1483Tyr or Arg, p.Glu2419Lys or Gly, and p.Leu2427Pro or Gln, accounting for 16.3% of all FCDII participants (13 of 80). The prevalence of the mutant allele in affected brain tissues ranged from 1.0% to 12.6%. The identified mutations induced the constitutive activation of mTOR kinase and cytomegalic neurons in affected brains carrying these mutations. Furthermore, the focal cortical expression of MTOR mutants in in utero electroporated mice was sufficient to interfere with proper neuronal migration and cause spontaneous seizures with epileptic discharge and cytomegalic neurons.

Conclusion: Therefore, this study provides the first evidence that somatic activating mutations in MTOR cause focal cortical dysplasia.

Keywords: malformation, mutations, MTOR, epilepsy

PF-055

Epilepsy

Simplified frameless stereoelectroencephalography in children: a pediatric series

Scavarda Didier¹; Trebuchon Agnes²; Millh Mathieu³; Pech-Gourg Gregoire¹; Lepine Anne³; Villeneuve Nathalie³; Girard Nadine⁴; Bartolomei Fabrice².

¹Department of Pediatric Neurosurgery, CHU Timone Enfant, Marseille, France, Marseille; ²Department of Neurophysiology, CHU Timone, Marseille, France, Marseille; ³Department of Pediatric Neurology, CHU Timone Enfant, Marseille, France, Marseille; ⁴Department of Neuroradiology, CHU Timone, Marseille, France, Marseille.

Objective: Localization of the epileptogenic zone is the goal in epilepsy surgery. SEEG, was developed in the 50s. Currently, modernization of the operative rooms allows operating under computer control. We developed an alternative methodology to implant children with deep electrodes, which is secure, decrease the radiations and the operative time and accurate. We report our frameless and simplified methodology of SEEG.

Methods: We prospectively studied our children investigated for refractory epilepsy in our institution with a simplified frameless implantation methodology that we developed since 2011 for deep electrodes implantations. Our technique is based on a neuronavigation system and a biopsy arm (Stealthstation Medtronic) without any added software. The number of electrodes, the side of implantation, the age of the children, the

accuracy of the entry point, the accuracy of the target point, the operative time, the complications and the ability to define the EZ are reported.

Results: from March 2011 to April 2015 we performed 52 SEEG in children. Age varied from 20 months to 21 years old. 35 implantations were unilateral. 539 electrodes were implanted with a mean of 10 electrodes/child. Implantation time varied with experience from 12 to 7 mn/ electrode. Mean time under general anesthesia is about 240 minutes. Accuracy has varied during this period after modification of our imaging protocol. We report 3 hemorrhages (2 minor/1major). Complication rate/ electrode: 0,8%.

Conclusion: Our methodology was developed to decrease the amount of radiation to the children and to decrease the time under general anesthesia. These goals were achieved but we needed to perform a CT scan to improve the accuracy. This technique is very intuitive and less expensive for the institutions with the same amount of complications. We did not add any software to plan the trajectories. Finally, accuracy matches positively with the few data available in the literature.

Keywords: epilepsy surgery, SEEG, children, Computer assisted surgery, frameless implantation

PF-056

Epilepsy

Multilobar resections for epilepsy in children from Sweden 1990-2013

Daniel T Nilsson¹; Bertil Rydenhag²; Kristina Malmgren².

¹Department of Neurosurgery, Sahlgrenska University Hospital, Gothenburg; ²Epilepsy Research Group, Institute of Neuroscience and Physiology, The Sahlgrenska Academy at The University of Gothenburg, Gothenburg.

Objective: Reports on outcome after multilobar resection (MLR) are scarce and are either retrospective single-centre studies or small case studies. This is the first prospective study of results after MLR providing robust data on seizure outcome and complication rates after this procedure in children.

Methods: The Swedish National Epilepsy Surgery Registry (SNESUR) provides prospective population-based data on outcome and complications after epilepsy surgery from 1990-2013. We report the seizure outcome (2 year follow-up) and complications after MLR from the SNESUR in children <19 years.

Results: Thirty-eight children underwent MLR, of which 15 were classified as partial hemispherotomy. Resections were right-sided in 25 (65.8%). Mean age was 7.7 years (range 0.3-16.7 years). Mean age at seizure onset was 2.2 years range 0-12 years, and mean duration of epilepsy before surgery was 4.8 years (range 0.2-15.9 years). Preoperative neurological deficits were seen in 23 children (60.5 %). Mental retardation (MR) was seen in 11 children (29%), four had severe MR (IQ<50). Seizure outcome after two years was available for 35 children, nine (26%) were seizure free and five (14%) had >75% seizure frequency reduction. Minor complications resolved within three months were seen in seven patients including three wound infections/osteitis, two transitory neurological deficits, one urinary tract infection and one cerebrospinal fluid leak. One major complication was seen, a permanent neurological deficit from infarction of the middle cerebral artery.

Conclusion: In this severely affected group of children, seizure outcome after MLR was considerably worse than for other resective epilepsy surgery procedures in the SNESUR, but 40% gained significantly. Major complications were rare. MLR can be considered also in children with IQ<50, but expectations for seizure freedom should not be high and parents should be counseled appropriately.

Keywords: epilepsy surgery, multilobar resection, seizure outcome, complications

PF-057

Epilepsy

Tailored frontal lobectomy after posterior quadrantectomy versus functional hemispherotomy for hemispheric pediatric epilepsy patients

Ju-Seong Kim¹; Eun Kyung Park¹; Kyu-Won Shim¹; Heung-Dong Kim²; Dong-Seok Kim¹.

¹Department of Pediatric Neurosurgery, Severance Children's Hospital, Yonsei University College of Medicine, Seoul; ²Department of Pediatric Neurology, Severance Children's Hospital, Yonsei University College of Medicine, Seoul.

Objective: To study the outcome of Tailored Frontal lobectomy after posterior quadrantectomy for hemispheric pediatric epilepsy patients, compare to functional hemispherotomy

Methods: A retrospective analysis of the Severance Children's hospital's epilepsy surgery database was done in all children who underwent a Functional hemispherotomy (FH) and Tailored frontal lobectomy after posterior quadrantectomy (FLPQ) from February 2006 to December 2012. All patients underwent a detailed pre surgical evaluation. Seizure outcome was used by the Engel's classification. And complication related to surgery were compared with each group. FLPQ group was underwent second staged operations. 1st surgery was performed posterior quadrantectomy (behind motor cortex and temporo-occipital lobe) and subdural grid insertion on the frontal area. After surgery, patients underwent intracranial EEG monitoring using subdural grid for a week. Based on that result, we decided the Frontal resection margin. 2nd stage surgery was Frontal lobectomy along the determined resection margin.

Results: There was 39 patients (50 operation cases including revision of hemispherotomy) in FH group. Epilepsy etiology was due to Lennox-Gastaut syndrome, Rasmussen's encephalitis (RE), Infantile hemiplegia seizure syndrome (IHSS), Hemimegalencephaly (HM), Sturge-Weber syndrome (SWS) and due to post-encephalitis or post-traumatic sequelae (PES or PTS). Seizure control rate of functional hemispherotomy was 85.7% (42/49 cases, Engel classification I, II). 7 patients were inserted shunt after hemispherotomy and 7 patients were in need re-operation due to post-operative adhesion, incomplete disconnection. Another 5 patients were underwent 2nd staged Tailored Frontal lobectomy after Posterior quadrantectomy. 100% seizure control rate was seen in this group. (Engel classification I, II). Just 1 case had post-operative complication, hemiparesis. FLPQ group did not need to sacrifice the unilateral motor function.

Conclusion: Tailored Frontal lobectomy after posterior quadrantectomy was shown excellent seizure outcome. This new procedure could be treat the hemispheric epilepsy patient without sacrifice of motor function.

Keywords: Hemispherotomy, Quadrantectomy, Epilepsy, Outcome

PF-058

Epilepsy

Surgical outcome in refractory epilepsy (RE) patients with extratemporal lesions (ETL) around eloquent cortex. An analysis of 14 patients

Dattatraya Muzumdar¹; Sangeeta Ravat²; Yogesh Ghodse²; Urvashi Shah²; Neena Sawant².

¹Department of Neurosurgery, Seth G.S. Medical College and King Edward VII Memorial Hospital, Mumbai; ²Department of Neurology, Seth G.S. Medical College and King Edward VII Memorial Hospital,

Mumbai; ³Department of Psychiatry, Seth G.S. Medical College and King Edward VII Memorial Hospital, Mumbai.

Objective: To assess surgical outcome in refractory epilepsy (RE) patients with extratemporal lesion (ETL) around eloquent cortex, who underwent epilepsy surgery based on data from noninvasive presurgical evaluation.

Methods: 14 patients having RE due to ETL around eloquent cortex, who underwent surgery at KEM hospital Mumbai, India in year 2008-10, was performed. Presurgical evaluation included dedicated MRI brain (1.5/3 tesla), ictal and interictal video-EEG, neuropsychology and psychiatric assessment. Preoperative invasive monitoring neither was not performed. 14 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: 14 patients with age range 8-20 years were assessed. Average seizure duration till surgery was 12.2 years. The follow-up period ranged from 6 to 18 months. 7 patients had a new transient postoperative neurological deficit during immediate post-operative period, which recovered completely in the following 2 to 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 & 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.

Keywords: eloquent cortex, epilepsy surgery, extratemporal

PF-059

Epilepsy

Endoscopic corpus callosotomy and hemispherectomy

Sandeep Sood¹; Abilash Haridas¹; Steven Ham¹.

¹Children Hospital of Michigan, Wayne State University, Detroit.

Objective: Corpus Callosotomy and hemispherotomy are traditionally done using a large craniotomy with a microscope for children with intractable epilepsy. Main considerations are completeness of disconnection and blood loss, Author describes an endoscopic technique done through a micro-craniotomy for these procedures.

Methods: Seven patients with drop attacks and 4 patients with intractable seizures underwent endoscopic complete corpus callosotomy and hemispherotomy respectively. The surgery was done using a novel method allowing bimanual two handed tissue handling with an endoscope mounted on a suction in one hand and dissecting instrument, bipolar or CUSA in the other hand. Through a pre-coronal 2 cm micro craniotomy, inter-hemispheric dissection to the corpus callosum is done using standard technique. Subsequently, an endoscope with a mounted suction is used to perform complete corpus callosotomy; interforncial disconnection and disconnection of the anterior commissure. In patients who had hemispherotomy, in addition the fornix is resected posteriorly with temporal horn un-roofing. A modification of Delalande's vertical hemispherotomy incorporating cortisectomy done along the ipsilateral anterior cerebral artery to reach the bifurcation of the internal carotid artery is used to compete the

disconnection. Patients underwent post-operative MRI scans and Diffusion Tensor Imaging (DTI) to confirm completeness of the disconnection.

Results: The procedure was accomplished successfully in all patients. Excellent visualization was achieved. The microscope was used intermittently in the first 2 hemispherotomies, and 3 corpus callosotomies. None of the patients required a blood transfusion. Postoperative MRI and DTI confirmed completeness of the disconnection.

Conclusion: Authors have demonstrated that endoscopic corpus callosotomy and hemispherotomy are surgically feasible procedures with minimal blood loss and risk.

Keywords: endoscopic, hemispherectomy, corpus callosotomy, intractable seizures

PF-060

Spine

Selective dorsal rhizotomy - experience over our first 50 cases

John Goodden¹; Alec Musson²; Katie Davis²; Kate Mccune²; Ian Smith³; Michael Clarke⁴; Rajib Lodh⁵.

¹Paediatric Neurosurgery, Leeds Children's Hospital at The Leeds General Infirmary, Leeds; ²Paediatric Physiotherapy, Leeds Children's Hospital at The Leeds General Infirmary, Leeds; ³Neurophysiology, Leeds General Infirmary, Leeds; ⁴Paediatric Neurology, Leeds Children's Hospital at The Leeds General Infirmary, Leeds; ⁵Paediatric Neurorehabilitation, Leeds Children's Hospital at The Leeds General Infirmary, Leeds.

Objective: Selective Dorsal Rhizotomy (SDR) aims to improve function and mobility through reduction in abnormal lower limb tone. We present Functional and Quality of Life (QoL) outcomes from the first 50 cases performed in our centre in England. We also report on our experience developing this new service.

Methods: We performed our first SDR procedure in October 2012. Post-operative outcomes are prospectively recorded on a spreadsheet database. SDR was performed according to a standardised protocol. Through a single-level laminectomy, using intra-operative neurophysiology, approximately 66% dorsal rootlets are cut from L1-S1. Standardised selection criteria were used - spastic diplegia with dynamic spasticity limiting function, no dystonia, typical MRI changes, and GMFCS Level 2 / 3. All patients had standardised pre & post op assessments with 3D Gait Analysis, GMFM-66, Ashworth grading, muscle power & joint range of movement. QoL was assessed using the CPQoL questionnaire.

Results: Fifty patients have had SDR (35 male, 15 female). Mean age at surgery was 6.75 years (range 2.6-13.8). Patients were GMFCS 2 (22) or GMFCS 3 (28). Mean follow-up is 528 days (range 108-1081). Over time the patient pathway has improved with shorter operation duration & hospital stays, better patient information, improved analgesia and developments in physiotherapy. Our learning points will be discussed. All patients have reduction in tone after SDR. GMFM-66 results demonstrate improvement. Range-of-movement change was not significant. CPQoL demonstrated improvements in all domains, with particularly large gains in Participation & Physical Health. There were no significant complications - specifically no instances of paralysis, numbness, or bladder/bowel dysfunction.

Conclusion: SDR is a safe and effective treatment for appropriately selected patients with spastic diplegic cerebral palsy. Our service has developed and grown as we have gained experience over these 50 cases. Important improvements in function and QoL are demonstrated.

Keywords: Selective Dorsal Rhizotomy, Cerebral Palsy, Spastic Diplegia, Quality of Life, Outcome

PF-061

Functional

Selective dorsal rhizotomy without neurophysiological monitoring for management of spasticity, the first ten year experience and results at the National Childrens Hospital of Costa Rica

Adrian Caceres¹; Juan Luis Segura¹; Justiniano Zamora¹; Edgar Jimenez¹.

¹Neurosurgery Service, National Children's Hospital of Costa Rica, San Jose.

Objective: Retrospective review of the outcome of selective dorsal root section without the use of neurophysiological monitoring in a 10 year period with a mean follow up of 4.9 years.

Methods: The medical records of 165 patients who underwent SDR between 2004 and 2014 were reviewed. 54% were male and 46% female. Ages ranged from 4 to 14 years (mean 5.6) All patients underwent a multilevel "en block laminectomy" with random selection of dorsal rootlets without the use of neurophysiological monitoring. Laminoplasty was performed in all patients. All patients had Ashworth and Gross Motor Function Assessment pre and postoperatively. Indications for SDR included gait abnormalities, pain due to hip luxation and restriction of daily activities by spasticity.

Results: All patients had a significant and persistent decrease of muscle tone in adductors, hamstrings and dorsiflexors ($p < 0.001$) as evidenced by Ashworth and GMFM scores. Patients also reported less spasticity and increase of dexterity of upper extremities in 46% with less sialorrhea in 40% of cases who drooled preoperatively. The degree of improvement was felt to be similar by caretakers at 1 and 5 Years postop. We found a 13% of patients who had CSF fistula and or wound dehiscence. 5% reported transient bladder control issues. Only one patient developed postoperative pneumonia.

Conclusion: Selective dorsal rhizotomy using a multilevel laminotomy and a 60% section of randomly selected roots without the use of neurophysiological monitoring provides short and long term results similar to other series. Complications were exclusively local (CSF fistula, wound infection/dehiscence) in all but one patient who developed pneumonia.

Keywords: spasticity, rhizotomy, cerebral palsy

Thursday, 8 October 2015

08:30 – 08:40

PF-062

Hydatid disease

Hydatid cysts of the central nervous system in pediatric population: critical review of literature and analysis of 188 pediatric patients

Abdelaziz Tariq M Sagga¹; Ahmed N Al-Ahmari²; Faris Alebdi¹; Khaloud K Alghamdi¹; Samer K Elbabaa³.

¹Department of Pediatric Neurosurgery, National Neuroscience Institute, King Fahad Medical City, Riyadh; ²Department of Neurosciences, King Faisal Specialist Hospital and Research Centre, Riyadh; ³Department of Neurological Surgery, Division of Pediatric Neurosurgery, Saint Louis University, Saint Louis, MO.

Objective: This study aims to provide an epidemiological, clinical, surgical, and prognostic analysis of pediatric patients with hydatid cysts "Echinococcosis" of the central nervous system (CNS) from published literature.

Methods: Review of the literature was performed using PubMed. Patients who were ≤ 18 years were included. For included patients, we have reviewed the Demographic information, presentations, locations of

the cysts, the presence of multiple cysts inside or outside the CNS, the use of Albendazole therapy, surgical techniques, complications, and prognosis.

Results: 188 patients were included. 120 (63.8%) were males and 62 (36.2%) were females. The median age was 10 years (range: 3–18). The most common geographical location of patients was in Turkey (52.7%). Most patients presented with history of headache (67%), vomiting (52.1%), and weakness (39.8%). The locations of the cysts were highly variable, but the most common locations were found in Frontoparietal (13.8%), Parieto-occipital (11.7%), and frontal (11.1%) lobes. Among patients who had excellent prognosis, 89.6% had single cyst compared to 10.3% with multiple cysts in CNS (p-value <0.001), 83.4% underwent removal of cysts using Dowlings technique compared to 16.6% with other techniques (p-value: 0.001), and 92.2% underwent removal of cysts without rupture (p-value <0.001). 73.3% of patients with poor prognosis did not receive Albendazole therapy compared to 26.6% who did (p-value: 0.002). However, excellent and good prognoses were not higher among patients who received Albendazole therapy compared to the other group (43.2% vs. 55.4%) (p-value: 0.002).

Conclusion: Hydatid cyst of the CNS among pediatric patients is endemic in certain countries. Excellent prognosis was associated with single cyst, removal of cysts without rupture, and the utilization of Dowlings technique. The efficacy of Albendazole therapy should be further validated using prospective studies.

Keywords: Hydatid, Pediatric, Central Nervous System, Prognosis

Thursday, 8 October 2015
08:40 – 09:10

Technology Free Papers VIII

PF-063

Hydrocephalus

Intraoperative ultrasound guidance for shunt placement in hydrocephalus

Marcel Kullmann¹; Marina Khachatryan²; Martin U. Schuhmann¹

¹University of Tuebingen, Tuebingen; ²Yerevan State Medical University, Yerevan.

Objective: Ventricular shunting operations are one of the most effective options for treatment of hydrocephalus in all age groups. However, there are few complications, leading to misplacement of the ventricular catheter. Intraoperative Ultrasound Guidance (IOUSG) has been increasingly suggested for better visualization of catheter positioning. The aim of our study is to evaluate the benefit of IOUSG utilization during ventricular catheter (VC) placement.

Methods: A retrospective analysis of patients data admitted to the University of Tübingen, Germany from June 2007 to July 2014 with hydrocephalus was performed. 69 patients were included in the study. The mean age was 2.5 years old (0 to 17). 50.7% females (n=35) and 49.3% males (n=34). Patients were divided into two groups: Group 1 received OUSG, Group 2 did not. The number of reoperations, reason of shut dysfunction, satisfactory position of VC on follow-up imaging have been recorded.

Results: The total number of operations was 114 including 69 (60.5%) primary shunting operations, 23 (20.2%) VC revisions, 9 (7.9%) valve revisions, 7 (6.2%) revisions of the peritoneal

catheter and 6 (5.2%) cases of total shunt replacement or removal. Group 1 included 84 (85.7%) operations, Group 2 14 (14.3%) operations. 23 of 45 shunt revisions were due to VC replacement. In 10 cases the reason was obstruction of VC, in other 13 cases general shunt malfunction. When comparing the rate of reoperations, Group 2 required 10 revisions out of 14 patients (71.4%) whereas the number of revisions in Group 1 was only 18 cases out of 84 patients (21.4%).

Conclusion: Utilization IOUSG in VC placement does not affect the rate of radiographic satisfactory results (83.3% vs 85.7%). However, the rate of required VC revisions is dramatically lower if IOUSG is performed (71.4% vs 21.4%).

Keywords: Hydrocephalus, Intraoperative Ultrasound, VP-Shunting, Revision

PF-064

Oncology

Tumor resection margins guided by intra-operative ultrasound

Llewellyn Padayachy¹; Anthony Figaji¹; Graham Fieggen¹.

¹University of Cape Town, Cape Town.

Objective: Improved tumor resection margins have been associated with better outcome in paediatric patients, IOUS offers a safe and effective technique for detecting residual tumor and defining resection margins. This study aimed to evaluate the benefit of IOUS in guiding tumor resection margins.

Methods: This is a prospective study evaluating the benefit of IOUS as an adjunctive tool in the resection of primary central nervous system tumors. These lesions were imaged according to a standardised intra-operative protocol, acquiring images for tumor localisation prior to dural opening, intermittently during the resection, and after surgical resection of the tumor. Tumor localisation, delineation, resection margins, vascularity, echogenicity, size, degree of resection were evaluated and analysed.

Results: 35 patients, with a median age 12 months (range 5 - 96) were included. Adequate IOUS imaging and tumor localisation was possible in all cases. Astrocytomas accounted for 49%, embryonal tumors 25%, ependymomas 14%, choroid plexus tumors 6%, and craniopharyngiomas 6%. IOUS confirmed gross total resection in 27/35 patients (77%) which correlated well with the pre-operative strategy ($t=0.65$, $p=0.08$). Tumor vascularity, residual, density differentiation (cystic vs solid) was adequately defined by echogenicity and power Doppler criteria in 33/35 (94%) cases. IOUS was particularly helpful in detecting residual tumor in low grade astrocytomas, where the margins were ill-defined on pre-operative imaging, and improved the initial grade of resection in 5/7 (71%) of these lesions.

Conclusion: IOUS is useful real-time guidance tool in tumor resection. Portability, improved image quality, ease-of-use and relative cost-effectiveness are areas of distinct advantage in a resource setting such as our own. It has proven most beneficial in improving resection margins in low grade astrocytomas and identifying vascularity in malignant tumors.

Keywords: tumor, ultrasound, resection

PF-065

Technology

MRI guided laser interstitial thermal therapy (MRgLITT) in pediatric brain lesions: lessons learned and future perspectives

Zulma Tovar-Spinoza¹.

¹SUNY Upstate Medical University, Syracuse.

Objective: Magnetic-resonance-guided-laser-interstitial-thermal-therapy (MRgLITT) as a novel minimally invasive therapy has convenient

benefits in pediatric neurosurgery. We present the lessons learned on the application of this technology on the patients treated with MRgLITT at the Golisano Childrens Hospital in Syracuse, NY, USA.

Methods: We reviewed the records of the 38 MRgLITT procedures (Visualase Thermal Laser System, Medtronic, Minnesota, USA) performed between February 2012 and April 2015. We identified the advantages of the use of MRgLITT in pediatrics, the factors attributable to the learning curve on the application of the technology, different procedures that can be performed, variations on the laser's use according to the type of lesion, limitations of the current software and future directions.

Results: MRgLITT advantages include its use at any age as de-novo or as a subsequent treatment, alone or as an adjunctive therapy; it has no-dose-limitations and no reported significant side effects. A planned injury is performed in one-single-session, for single or multiple-lesions with one or multiple-lasers and with a single or multiple-thermal ablations. The procedure is done under MRI and diminishes hospital-stay, it does not require hair-clipping, the incision is closed with single-absorbable-stitch and it produces minimal pain. However, different types of procedures (tumor ablations vs focal-epileptogenic-lesionectomies vs cortical disconnections vs multiple-laser-ablations for tuberousclerosis) require different plans for surgery and use of the laser(s). The current software's restrictions on the volumetric-view and limits of the ablation zone and pitfalls are described. Future applications of MRgLITT as a coadjuvant-therapy for targeted-tumor treatment are presented.

Conclusion: MRgLITT as a novel-technology has an associated learning curve. Presenting our learned-lessons will assist the new users on their own learning-process. We present our experience with the aim of compiling multi-institutional data and create and improve "protocols" that will allow further analyses and improvements.

Keywords: Laser-induced-thermal-therapy, pediatric, brain lesions, technology

Thursday, 8 October 2015
09:40 – 10:30

Craniofacial Free Papers IX

PF-066

Craniofacial

Quantitative analysis of Crouzon mouse calvarial morphology by synchrotron tomography

Alessandro Borghi¹; Alessandra Carriero²; Michael Doube³; Andrew Bodey⁴; Silvia Schievano¹; Naiara Rodriguez Florez¹; Erwin Pauws¹; David Dunaway⁵; Owase Jeelani⁵

¹Institute of Child Health, London; ²Florida Institute of Technology, Melbourne; ³Royal Veterinary College, London; ⁴Diamond Light Source, Oxford; ⁵Great Ormond Street Hospital, London

Objective: Heterozygous Fgfr2C342Y mice show premature fusion of craniofacial sutures which causes mid-face shortening and coronal craniosynostosis. We investigated the skull bone morphology of two weeks old mutant mice and compared with age-matched wild type using high resolution synchrotron CT (sCT).

Methods: Calvaria from 2 week old wild type (WT, n = 5) and mutant (C342Y, n = 5) mice were collected and skin and soft tissue removed. C342Y calvaria showed clear phenotypic features such as coronal synostosis and brachycephaly. sCT was performed at Diamond Light Source (Oxford, UK). Cross sections were processed in MIMICS (MATERIALISE) for 3D reconstruction. Image processing was performed on a 1mm x 1mm region of interest (ROI) from the frontal bone:

porosity was extracted using morphological operations. Bone volume (BV), bone surface (BS) and TV (volume of the ROI) were measured.

Results: The mutant calvaria showed features typical of Crouzon syndrome: a visual inspection showed a higher curvature and undulation in the sagittal as well as interfrontal suture of the mutant mice. The mutant population showed a lower bone volume fraction (BV/TV 54.79%±13.59% vs 87.34%±4.45%, p<0.01) and a higher specific bone surface (BS/TV 12.01±1.20% vs 7.54±0.93%, p<0.01). This was most pronounced in the frontal bone, where abnormalities in the vasculature were also observed (as compared to WT frontal bone and mutant parietal bone).

Conclusion: Skulls of Fgfr2C342Y mice show skull defects, similarly to children affected by Crouzon syndrome: a novel observation is that the bone architecture of the frontal bone is affected specifically in the C342Y mutant. This may reflect the different embryonic origin of the frontal and parietal bones. The quantitative analysis of the microscopic morphology constitutes an important tool for understanding the skull development, which will provide critical information for surgical planning.

Keywords: crouzon, craniosynostosis, tomography

PF-067

Craniofacial

Venous anomalies in syndromic craniosynostosis.

Andrea Copeland¹; Caitlin Hoffman¹; Vassilios Tsitouras¹; Dhruve Jeevan¹; James Drake¹; Christofer Forrest².

¹Department of Neurosurgery, The Hospital for Sick Children, Toronto; ²Division of Plastic and Reconstructive Surgery, The Hospital for Sick Children, Toronto.

Objective: The pattern of venous drainage in syndromic craniosynostosis is unpredictable and not adequately understood. Collateral and subcutaneous channels substitute for stenotic venous sinuses and pose potential risk for planned surgical intervention. We examined the frequency of these anomalous patterns and their influence on operative planning and morbidity.

Methods: We performed a retrospective study of patients treated for syndromic craniosynostosis from 2000 until 2013. Phenotypical diagnoses and associated pathologies (ICP, hydrocephalus, hindbrain hernia, syrinx and sleep apneas) were reviewed. We focused on relevant imaging (CT/CTV, MRI/MRV) and examined venous sinus stenosis, emissaries, collateral vessels and persistent fetal sinuses. We categorized the anomalous venous drainage and analyzed its relationship with surgical morbidity using appropriate analyses for non-parametric variables.

Results: There were 41 patients with complete preoperative imaging. The most frequent phenotypes were Crouzons (46%) and Aperts (27%) and the most frequent associated pathology was hindbrain herniation (51%). The median age at first operation was 14 months and the commonest procedure was total cranial vault remodeling (n=14). Venous anomalies were present in 31 patients (77%); dural sinus stenosis in 28 (68%), dilated emissaries in 26 (63%), and fetal sinuses in 8 (20%). The syndrome with the most frequent anomalies was Pfeiffers (100%). In 4 cases, the planned procedure was altered based on venous anatomy. Post-operative complications occurred in 19.4% of the cases, all of which had anomalous drainage patterns. Risk factors for highest mean estimated blood loss (EBL) were Aperts phenotype, 1 to 2 emissaries and a more distal venous sinus stenosis.

Conclusion: Venous anomalies were frequently encountered in our series in variable patterns. Their presence affected surgical morbidity and outcome, with higher EBL, alteration of procedure, and post-operative complications. Detailed preoperative imaging of the venous drainage is therefore recommended in cases of syndromic synostosis.

Keywords: syndromic, craniosynostosis, veins, anomalous

PF-068

Craniofacial

Hydrocephalus in craniosynostosis – is bigger better or does diversion matter?

Desiderio Rodrigues¹; Melanie Sharp¹; Jagadeeshan Jagajeevan¹.
¹Birmingham Children's Hospital, Birmingham.

Objective: To present our experience in the management of hydrocephalus in the presence of craniosynostosis. Hydrocephalus with craniosynostosis is well recognised, especially in syndromic multisuture craniosynostosis. The aetiology is multifactorial. While diversion procedures are reliable in treating non-synostotic hydrocephalus, they are less so in craniosynostosis is variable and can be damaging.

Methods: Of the 1169 craniosynostosis patients treated in our unit over the last 12 years, 169 (14%) had syndromic craniosynostosis; 21 of these had hydrocephalus (N=21). We divided them into two groups, and compared their outcomes. Group A – Those who had Cerebro-spinal Fluid (CSF) diversion as a primary procedure prior to calvarial expansion procedures (N=8). Group B – Those who underwent calvarial expansion procedure as a primary procedure with or without subsequent CSF diversion (N=13).

Results: Group A were associated with increased complications when compared to Group B (87% vs 8%). Majority of these complications were a consequence of CSF diversion procedure and sometimes resulted in a delay in definitive treatment or required multiple calvarial remodeling interventions.

Conclusion: Primary aim of treatment should address the craniosynostosis with calvarial expansion surgery. Further close monitoring should be undertaken to evaluate the need for further calvarial surgery or a CSF diversion. This may avoid the need for any CSF diversion procedure and their associated complications.

Keywords: Hydrocephalus, Craniosynostosis

PF-069

Craniofacial

Characteristics of intracranial cerebrospinal fluid collection in scaphocephaly.

Kenichi Usami¹; Eric Arnaud¹; Francesca Nicolini¹; Federico Di Rocco¹.
¹Craniofacial Unit, Department of Pediatric Neurosurgery, Necker-Enfants Malades Hospital, Paris.

Objective: Some patients with scaphocephaly can present with an intracranial cerebrospinal fluid (CSF) collection, as pericerebral effusion (PE) in frontal region and as ventriculomegaly. However its etiology and development are still unknown. In this study, we clarify the characteristics of intracranial CSF collection in scaphocephaly, consider the surgical effect on CSF collection, and analyze the prognostic factor of CSF dynamics.

Methods: The subjects were 60 patients with scaphocephaly who underwent craniotomy with decompression of superior sagittal sinus (SSS) at the age of less than 6 months between 2008 and 2014. PE was measured before and after surgery, and compared to expected average of estimated value corrected for age (normal control). Evans index (EI) was calculated as ventricular indices. Correlations among these values and indices were analyzed.

Results: Forty-seven of 60 patients (78%) had larger PE than normal control preoperatively. PE improved more than natural course of normal control in 56 of 60 patients (93%) postoperatively. Thirty-four of 60 patients (57%) had pathological EI (EI >0.3) preoperatively. PE and EI were significantly reduced in the postoperative period (p<0.001). The earlier intervention, the more PE improved.

Conclusion: Surgery for scaphocephaly with decompression of the SSS reduced PE and EI significantly. Particularly, PE improved faster than the expected natural course. These may indicate that the decompression of SSS is important to hydrodynamics of CSF in scaphocephaly.

Keywords: scaphocephaly, cerebrospinal fluid, surgical effect, pericerebral effusion, ventriculomegaly

PF-070

Craniofacial

Bilambdoid and sagittal synostosis: report of 39 cases

Nathalie Chivoret¹; Eric Arnaud¹; Leslie Pamphile¹; Philippe Meyer¹; Dominique Rénier¹; Federico Di Rocco¹
¹Necker Hospital, Paris

Objective: Bilambdoid and sagittal synostosis or « Mercedes pattern » is a multisuture craniosynostosis that has been described as a specific entity. However this synostotic pattern can also be found in syndromic craniostenosis. In order to better define this entity we reviewed our cases of bilambdoid and sagittal synostosis.

Methods: We searched our prospective database for cases of bilambdoid and sagittal synostosis among all types of craniosynostosis. Two groups were identified: the patients with isolated BLSS and the group of syndromic craniostenosis for whom BLSS was observed in initial presentation. We reviewed the clinical findings, associated diseases and their management more specifically for isolated BLSS patients.

Results: 39 patients were diagnosed with bilambdoid and sagittal synostosis among 4250 cases of craniosynostosis treated in our department over a period of 37 years. Among them, 8 were finally diagnosed as Crouzon syndrome. Of the 31 patients identified with isolated bilambdoid and sagittal synostosis, 16% of population presented papillary oedema and 55% posterior digitate impressions at diagnosis. Two types of pattern of cranio-facial features were observed: a pattern with narrow occiput and a pattern with dolichocephaly. Tonsillar herniation malformation was the most frequent associated malformation. Several surgical techniques were used during time to treat patients with BLSS: isolated biparietal vault remodeling, posterior vault remodelling, posterior vault expansion with internal or external distraction. In some cases, a craniovertebral junction decompression was also performed. The mean follow-up was 84 months. The overall mental prognosis was burdened by a mental delay in 25% of the cases, especially in case of late treatment.

Conclusion: Bilambdoid and sagittal synostosis constitute an isolated entity in almost 80% of the cases. Two types of phenotype may be found. For this relative heterogeneous population, several techniques can be used. Tonsillar prolapse is present in majority of cases but seldom requires a specific treatment.

Keywords: Complex craniostenosis, bilambdoid and sagittal synostosis, surgical strategie, outcome

Thursday, 8 October 2015

11:00 – 12:20

Craniofacial Free Papers X

PF-071

Craniofacial

The effects of craniectomy width and age at surgery in endoscopic strip craniectomy for sagittal craniosynostosis: what leads to better outcomes?

Suresh Magge¹; Arthur Bartolozzi Bartolozzi²; Gary Rogers¹; Robert Keating¹.

¹Children's National Medical Center, Washington, DC; ²Harvard Medical School, Boston, MA.

Objective: Endoscopic-assisted strip craniectomy has been used to treat sagittal craniosynostosis. There is significant variation amongst surgeons regarding surgical technique. The aim of this study was to examine whether the width of craniectomy or age at surgery affects outcomes.

Methods: This IRB-approved, retrospective study included 30 patients diagnosed with sagittal craniosynostosis who were treated with endoscopic strip craniectomy followed by helmet therapy. When examining craniectomy width, patients were divided into 2 groups based on the maximum width of craniectomy: Group A: 14 mm or less (mean width 10.9 mm), and Group B: 15–20 mm (mean width 17.2 mm).

Results: When studying craniectomy width, the mean age of patients were similar in the two groups (Group A: 3.3 months; Group B: 2.8 months; $p=0.25$). Operative times were similar (Group A: 68 min; Group B: 68 min). Hospital stays were roughly similar (Group A: 1.06 days; Group B: 1.29 days; $p=0.19$). Mean follow-ups were 20 months for Group A and 15 months for Group B. Each group started with a similar mean cranial index (CI) at the start of treatment (Group A: 0.693; Group B: 0.672; $p=0.15$). Group B had a greater mean increase in CI [CI=0.672 to CI=0.783 (16.7% increase)] compared to Group A (CI= 0.693 to CI=0.765 (10.5% increase)) ($p<0.01$). When examining age at surgery, there was a trend that endoscopic patients had improved CI outcomes when their surgeries were done at younger ages (approximately 1.4 % increase in CI per month younger).

Conclusion: Larger craniectomy widths correlated with improved CI measurements for patients undergoing endoscopic strip craniectomy. In addition, endoscopic patients had a trend of improved CI results when their surgeries were done at younger ages.

Keywords: craniosynostosis, sagittal, endoscopic, craniectomy

PF-072

Craniofacial

Can molding helmets improve head shape in patients having sagittal craniosynostosis without surgery?

Sandeep Sood¹; Abilash Haridas¹; Arlene Rozzelle¹; Neena Marupudi¹; Steven Ham¹.

¹Children Hospital of Michigan, Wayne State University, Detroit.

Objective: To assess improvement in head shape with a molding helmet alone without surgery in patients with sagittal craniosynostosis.

Methods: Twenty one patients with sagittal craniosynostosis underwent placement of a molding helmet at presentation with the intention of performing calvarial reconstruction at 6–8 months age. Patients were monitored at 4 weekly interval with clinical parameters such as head circumference, Cranial Index (CI), milestones and development, fundus and fontanelle assessment and a clinical photograph was obtained. Amongst 10 patient who were studied prospectively under IRB approval, 8 were under 3 month's age at the time of placement of the helmet.

Results: No patient had any observable adverse effects during the use of helmet. Mean duration of helmet use prior to surgery was 4.3 months. In these patients, the Cranial Index had increased from a mean of 0.69 ± 0.03 to 0.75 ± 0.03 (significant at $p < .000$) with use of the molding helmet alone. Due to proximity to surgery, in one patient who outgrew the helmet, the replacement helmet was given only following the surgery. This patient's CI worsened. Even though the intent was to operate all these patients at around 6 months, 4 parents refused surgery since parents felt the patients head shape appeared acceptable to them.

Conclusion: This report challenges the conventional thought that molding helmet alone cannot improve head shape in patients with sagittal craniosynostosis. We question the need for surgery in some of these patients who would otherwise have significant improvement in head shape with just a molding helmet.

Keywords: sagittal craniosynostosis, molding Helmet

PF-073

Craniofacial

Posterior cranial vault distraction in patients with craniosynostosis. Leonid Satanin¹; Alexander Ivanov²; Vitaly Roginsky²; Sergey Gorelyshev¹; Alexander Sakharov¹; Andrej Evteev³; Ivan Teterin¹; Natalia Lemeneva¹.

¹Moscow Burdenko Neurosurgery Institute, Moscow; ²Central Institute of Stomatology and Maxillo-Facial Surgery, Moscow; ³Anuchin Research Institute of Anthropology, Moscow.

Objective: Patients with complex syndromic craniosynostosis have a severe craniocerebral disproportion. Fronto-orbital advancement (FOA) cannot achieve an adequate increase of the intracranial volume. Occipital expansion allows avoid these problems.

Methods: In Moscow Burdenko Neurosurgery Institute from 2009 to 2015 52 patients have had posterior cranial vault expansion using distraction osteogenesis. The mean age was 19 months (6–96 months). Patients have Kleeblattschadel anomaly (6 cases), Pfeiffer (7), Apert (17), Saethre-Chotzen (5), Craniofrontonasal syndrome (2), and unspecified syndrome (15) were operated. Arnaud type distractors (KLS Martin) were used in 32 patients and internal distractors (Conmet, Russia) in 20 cases. In 19 cases 4 distractors were used, in 10 cases – 3 distractor, and in 23 cases only 2 devices. The latency period was 5 days and distraction rate was 0,5 mm/day. The average advancement was 23,7 mm (range 17,4–30 mm). There was no complication during distraction. Satisfactory callus formation was found on postoperative CT. Distractors were removed after consolidation period (6 months) and no any additional fixation was necessary. After 6 months FOA were performed as a second stage of the treatment.

Results: Measures of volumetric changes of intracranial volume after posterior cranial vault expansion revealed significant increase of intracranial volume (mean +250 ml). That differs significantly from changes in intracranial volume after FOA (mean +175 ml). As a result of surgery there were changes in anterior part of skull morphology, with reduction of bulging in anterior fontanelle region, temporal protrusion. Comparison of the morphology of the cranial vault before and after surgery by geometric morphometrics was performed.

Conclusion: Expansion of the posterior cranial vault using distraction osteogenesis is effective method of treatment of complex syndromic deformities of the skull accompanied by hypoplasia of parietal-occipital region.

Keywords: Craniosynostosis, Vault distraction, Craniofacial surgery

PF-074

Craniofacial

100 cases of posterior vault expansion surgery - technique, complications, intracranial volume changes and its effect on ophthalmic assessment

Owase Jeelani¹; William Breakey¹; Alessandro Borghi¹; Silvia Schievano¹; David Dunaway¹.

¹Great Ormond Street Hospital, London.

Objective: Children with multisuture craniosynostosis require cranial vault expansion to ameliorate the effects of raised intracranial pressure and treat turribrachicephaly. Of the various techniques described, posterior cranial vault expansion offers the greatest volume increase. Our unit moved from rigid posterior vault expansion to spring mediated expansion in 2008 and

we have since undertake 116 cases using this method; the largest cohort using this technique. The purpose of this study is to demonstrate our novel technique, results, complications and the volumetric changes achieved using this method and correlating this with their ophthalmological findings.

Methods: 100 consecutive cases of spring mediated posterior vault expansion surgery are described. Preoperative and postoperative Computer-Tomography imaging with 6 months of each other were available for 32 patients and these was analysed using OsiriX Medical Image software. Segmentation of the entire cranial vault was performed and volume calculated. Post operative volumes were compared to published normative data. Preoperative and serial postoperative ophthalmological assessments were collated and used as a surrogate measure for procedural efficacy. Fundoscopy and Electrodiagnostic studies were employed.

Results: All patients were operated on by the same primary surgeon. The mean preoperative intracranial volume was 976.1cm³. The mean postoperative intracranial volume was 1273.0cm³. The mean change in volume was 296.9cm³. (30% increase) Mean age at insertion of springs was 548 days. Springs remained in situ for an average of 219 days. We had 11 grade 1, 15 grade 2 complications and one mortality in our series. 78% of the cases showed a resolution in their ophthalmological signs over a 6 month period post operatively.

Conclusion: Spring assisted posterior vault expansion results in significant increases intracranial volume resulting in improvement on ophthalmological assessments. It is the preferred method of undertaking calvarial volume expansion for raised Intracranial Pressure within our unit.

Keywords: Vault Expansion, ICP

PF-075

Craniofacial

Strategic cranial reformation with distraction osteogenesis in sagittal craniosynostosis

Kyu Won Shim¹; Myung Chul Lee²; Eun Kyung Park¹; Dong Seok Kim¹; Yong Oock Kim²; Joo Seong Kim¹.

¹Department of Pediatric Neurosurgery, Severance Children's Hospital, Yonsei University, College of Medicine, Seoul; ²Department of Plastic & Reconstructive Surgery, Craniofacial Reforming and Reconstruction Clinic, Yonsei University College of Medicine, Seoul.

Objective: Distraction osteogenesis (DO) is a less daunting procedure than extensive cranial vault remodeling and has been in use for over a decade to correct sagittal craniosynostosis. We have performed three different surgical procedures in accordance with cranial index and report excellent results herein.

Methods: Between June 2002 and June 2013, 45 patients (>5 months) with non-syndromic sagittal synostosis were recruited retrospectively, each assigned to one of three therapeutic groups: EC, expansion-compression (n=27); BE, bitemporal expansion (n=9); and EE, expansion-expansion (n=9) methods. Circumferential baseline, mid-sagittal, and bicoronal osteotomies were performed in addition to four-quadrant bone flap procedures via distraction with dural attachment. Cranial index (CI), intracranial volume (ICV), and head circumference (HC) were analyzed.

Results: Preoperative CI of the EC group (68.85±3.61) differed significantly from that of BE (76.75±2.4, p<0.05) and EE (85.63±4.31, p<0.05) groups. Postoperative CI in all groups converged to the mesocephalic CI value. Preoperative ICV determinations were within the normal for the majority of participants in EC and BE groups, nonetheless falling below -1SD in the EE group (n=7, 77.8%). Postoperatively, 77.8% (n=7) of EE participants had ICV values within the normal range. All patients in the EC and BE groups displayed a range of HC values (-1SD to 2SD).

Preoperative HC values in 66.7% (n=6) of EE were below -1SD, but 77.8% (n=7) showed normal results postoperatively.

Conclusion: Symmetric sagittal synostosis can be treated effectively through DO reformation, guided by strategic categorization. The fundamentals of our approach remain tentative to further refinement.

Keywords: Sagittal craniosynostosis, distraction osteogenesis, cranial index, intracranial volume, head circumference

PF-076

Craniofacial

Patient related outcome measurements (PROM) to evaluate the result of surgical correction for sagittal suture synostosis

Marie-Lise Van Veelen¹; Nathalie Kamst¹; Hester Lingsma¹; Irene Mathijssen¹.

¹Erasmus University Medical Center, Rotterdam.

Objective: Good outcome measures for sagittal synostosis surgery are lacking. Cosmetic results are difficult to objectify, while cephalic indices seem insufficiently representative. Patient Related Outcome Measurements (PROMs) reflect the patient's own satisfaction and may be the most valuable type of outcome parameter. This study evaluated a PROM specifically designed for sagittal synostosis surgery.

Methods: The questionnaire contained 9 questions. Answers were rated along a five point Likert scale. 145 questionnaires were sent to patients aged six and older who were operated for sagittal synostosis. This study evaluated the PROM by looking at type of surgery and the correlation with the CI.

Results: 94 questionnaires were returned (mean age 11,6 ± 4,1 years). Patients underwent an extended strip craniotomy (n=50) at a mean age of 4,5 ± 2,0 months or a complete remodelling (n=44) at a mean age of 11,9 ± 6,1 months. Most respondents (84%) considered the shape of their head as being similar or slightly different from others, 95% never or very rarely received remarks about the shape of their head. The PROM was not significantly correlated to age, except the question on scars, which were considered more noticeable in older children (p=0.012). The PROM showed a trend but was not significantly correlated to CI. Dissatisfaction was more likely to consider the forehead after complete remodelling (11% vs. 2%, p=0.064) and the occiput after extended strip craniotomy (28% vs. 2%, p=0.001).

Conclusion: This first evaluation of the PROM shows that the majority of patients is satisfied with the outcome of their intervention. The PROM is mostly robust to the age of completion, has additional value as it is independent of cranial index, and shows to discriminate between different techniques. The PROM is a valuable outcome parameter for evaluating sagittal synostosis surgery and may influence technique or extent of remodelling.

Keywords: PROM, scaphocephaly

PF-077

Craniofacial

A 3D morphometric and volumetric follow up analysis

after frontoorbital advancement in non-syndromic craniosynostosis
Markus Martini¹; Mirjam Schulz¹; Andreas Röhrig²; Jennifer Nadal³;
Martina Messing-Jünger²

¹Clinic for Maxillo-Facial Surgery, University Hospital Bonn, Bonn;

²Pediatric Neurosurgery, Asklepios Klinik Sankt Augustin, Sankt Augustin; ³Institute for Medical Statistics, Bonn University, Bonn.

Objective: The frontoorbital advancement (FOA) in patients with non-syndromic craniosynostosis is mainly addressing esthetical and functional correction of the frontoorbital region. So far no objective criteria exist to evaluate the surgical outcome. Basing on a 3D morphometric analysis new parameters for the documentation of changes of the frontoorbital bandeau have been developed in a prospective study. Volumetric measurements have been included into the assessments.

Methods: In a prospective series 13 children with non-syndromic craniosynostosis treated with frontoorbital advancement (metopic 7, unilateral coronal 4, bilateral coronal 2) underwent detailed morpho- and volumetric evaluation using a 3D light-optic scan system (3D-Shape®, Erlangen, Germany). Measurements were obtained preoperatively as well as 3, 6 and 12 months postoperatively with newly developed parameters (frontal angle, frontoparietal angle) generated by a cephalometric analysis software (Onyx Ceph®, Image Instruments, Chemnitz, Germany).

Results: After frontoorbital advancement surgery most cases showed stable and constant longterm results without growth inhibition and with normalization or improvement of the following skull development. The mean frontal angle was 145° and the frontoparietal angle 137°–140°. Cephalic index was normalized or markedly improved. Head circumference and head height increased significantly ($p = 0.001$, $p=0.002$). These changes were confirmed in all postoperative measurements. Initial median skull volumes varied amongst the different synostoses groups (984cm³ in brachycephalus, 1205 cm³ in anterior plagiocephalus, 1214 cm³ in trigonocephalus). In all 3 groups a steady increase of skull volumes over 3, 6 and 12 months postoperatively could be observed. After 12 months brachycephalic children had a median volume of 1340 cm³, plagiocephalic cases of 1598 cm³ and trigonocephalic children of 1596 cm³. The volume increase in brachycephalic patients was significantly lower.

Conclusion: During the 12 months follow up period all angle parameters proved to be stable and no major impairment of harmonic skull growth could be observed after FOA. The frontoorbital angle is a useful parameter to evaluate longterm outcome. The frontoparietal angle is important for the stability of the frontoparietal region, in which a certain growth inhibition has been discussed in former series. In our series the classical FOA technique does not inhibit a constant increase in skull volume over a one year period, although brachycephalic children have a significantly lower increase in volume.

Keywords: Frontoorbital advancement, cranioplasty, morphometric analysis, volumetric analysis, 3D scan, craniosynostosis

PF-078

Other

Separation of craniopagus twins: experience with three pairs.

Ahmed Alferayan¹.

¹KAMC Hospital, Riyadh.

Objective: To describe the management of complicated craniopagus twins and the necessity for co-operation with other specialties such as radiology, plastic surgery, anaesthesia and paediatrics.

Methods: Three pairs of craniopagus twins have been managed and separated successfully with no neurological deficits. The first was performed in 2005 using one stage of separation. A staged strategy was used for the other two, utilizing interventional radiology, which facilitated separation at minimal risk.

Results: Successful separation of three pairs of conjoined twins, resulting in 6 healthy girls.

Conclusion: Appropriate planning and co-operation with other members of the team made it possible to achieve this excellent outcome.

Keywords: Craniopagus, Siamese Twins, Conjoined twins, Interventional neuroradiology, Separation

Monday, 5 October 2015

09:00 – 12:40

Nursing Symposium

PF-079

Nursing Symposium

The health related quality of life of children with lumbar sacral lipoma

Lindy May¹; Adam Kuczynski¹; Jo Wray¹.

¹Great Ormond Street Hospital For Children NHS Foundation Trust, London.

Objective: To compare the Health Related Quality of Life of Children with lumbar sacral lipoma, with that of children with spinabifida Aperta and with normative values.

Methods: 54 children between the age of 5 and 18 years old with lumbar sacral lipoma and their parent/ proxy were recruited at one study centre and provided with an age appropriate Health Related Quality of Life (HRQL) questionnaire- the Pediatric Quality of Life Inventory (PedsQL). The results were compared with that of children with spinabifida Aperta and with normative values. Secondly, each child and parent were asked independently, what were the 5 most important features of their disease / their child's disease and asked to rank them in order of importance.

Results: The children with lumbar sacral lipoma and their parent / proxy scored higher in all domains of the PedsQL than the children with spinabifida and parent / proxy. When compared with normative values the children with lumbar spinal lipoma reported a lower HRQL in the physical domain of the PedsQL, however their parent / proxy scored lower on the physical, psychosocial, social and emotional domains when compared to normative values. The majority of children and parents reported that managing urinary continence was the most important aspect of their lives in relation to their disease, followed by mobility and pain respectively.

Conclusion: The results suggest that many children with lumbar sacral lipoma have a reduced HRQL due to their condition and that this may in part, be associated with urinary dysfunction. This has not been recognised in the literature and as such, points the way to further research for this rare anomaly.

Keywords: lipoma

PF-080

Nursing Symposium

Measuring gait in children with lipoma: can we detect early changes in motor function?

Lucy Alderson¹; Christina Rampota²; Dominic Thompson³; Lindy May³.

¹Physiotherapy & Neurosciences Department, Great Ormond Street Hospital NHS Trust, London; ²Advanced Physiotherapy, University College London, London; ³Great Ormond Street Hospital for Children NHS Foundation Trust, London.

Objective: To describe gait characteristics of children with lipoma. Children with lipomyelomeningocele (spinal lipoma) have a spectrum of physical presentations affect-ing neurological and urological function, musculoskeletal deformity, pain and motor deficits (Kanev et al., 1990; Kulwin et al, 2013; Segal et al, 2013). Evidence to support prophylactic

untethering is conflicting. Current practice at GOSH is to monitor within an MDT clinic and intervene promptly when function deteriorates. Identifying deterioration is challenging as baseline function and tethering presentations vary. Standard mobility classification is often insensitive to symptom progression. This study explores identification of subtle walking deficits in this population.

Methods: 30 children who attended dysraphic clinic at GOSH between Jan 2013–June 2014 completed a GAITRite assessment as part of routine clinic follow up. Children walked across a 6 m pressure sensitive carpet 3 times at preferred, slow and fast speeds. Approval for retrospective review was granted by GOSH/ICH Research and Development Office.

Results: Mean age was 8.6years (range 3.8–17.9). Preferred velocity mean (sd) was 118.2cm/s (21.0). Data was plotted on normal gait centiles, from 650 typically developing children, and converted to Z scores using LMS-Growth. Preferred walking velocity fell within the 25th and 75th centiles (Figure 1) with no significant difference between Z scores and predicted age means ($p>0.05$). Preliminary analysis of fast walking data shows reduced centiles for a subgroup of children.

Conclusion: Preferred walking velocity of children with lipoma falls within normal ranges. Progressive symptoms are associated with reduced endurance, falling behind peers, with slowing of gait when walking fast. Multi-speed analysis identified gait deterioration in children with Talipes who present with similar muscle imbalance (Kemp, 2015). Longitudinal gait measurement in children with Lipoma at fast and pre-ferred speeds may identify subtle deteriorations, aiding surgical decision making.

Keywords: Lipoma, lipomyelomeningocele, gait, walking, velocity

PF-081

Nursing Symposium

The tethered-cord-syndrome and its possible challenges for nursing care after myelolysis

Kristina Düngen¹.

¹Gesundheits- Und Kinderkrankenpflegerin, B.Sc in Health Care Studies, Sankt Augustin.

Objective: To discuss tethered-cord-syndrome, the reasons for surgery and nursing care pre and post operatively. (TCS) describes a congenital or acquired tissue connection between the spinal cord and/or nerve roots within the spinal canal and the dura or other surrounding tissues. A functional state of stress with reduced blood supply and metabolism in the spinal cord results in various clinical symptoms and signs, like progressive neurological deficits or bladder/bowel dysfunction.

Methods: In order to minimize or prophylactically prevent symptoms and signs, early surgery is advocated. It consists of a microsurgical cutting of adhesions between the neural structures and the surrounding tissues. These procedures are called detethering. In case of a secondary tethered cord in patients with spina bifida this surgery is known as myelolysis. This intervention is supported by adequate care centered on the patient and his or her parents and this is discussed below.

Results: Specialized nurses conduct the specific care and include also the parents. This is how many intricacies are located and minimized in time. During the postoperative phase, the most important components of the nursing care is keeping patients in bed in a flat position for 3 days and monitoring the micturition with possible essential intervention procedures.

Conclusion: This presentation provides an insight into the pre- and post-operative care of children with tethered cord after detethering surgery or myelolysis and into various postoperative aspects, which imply an interdisciplinary teamwork for best possible patient support.

Keywords: Nursing management for tethered cord

PF-082

Nursing Symposium

The long-term implications of a child with myelomeningocele following neurosurgical repair and the subsequent preparation and support provided to families

Sheree Dean¹; Jenny Savage¹.

¹Great Ormond Street Hospital, London.

Objective: To discuss the evolving medical treatment of a child with myelomeningocele, the pre and post-operative nursing care currently delivered at Great Ormond Street Hospital; in addition the psychological preparation/support provided to parents/carers by the neuroscience ward nurse regarding potential long term difficulties, will be discussed.

Methods: Senior members of the nursing and neurosurgical team will be informally interviewed regarding the evolving management of myelomeningocele patients. Following surgical treatment, parents/carers will also be asked how they felt nursing care impacted upon personal experience and contributed towards emotional adjustment whilst resident on the ward.

Results: Yet to be determined and subsequently discussed in the presentation.

Conclusion: Advances in treatment and specialist neurosurgical intervention continue to positively reduce mortality rates for myelomeningocele patients. However, these infants often endure a repertoire of life limiting disabilities as a consequence of associated physiological and cognitive complications. Psycho-social support is therefore deemed imperative, particularly considering the optimistic prognosis now associated with the condition in comparison to historical statistics. Published literature should be utilised to amplify understanding, provide rationale for clinical intervention and ultimately promote a sense of trust within the health care professional. Guiding parents through the devastating diagnosis and management of a myelomeningocele can evidently prove challenging for the neuroscience nurse, justifying the relevance of the specified topic.

Keywords: Myelomeningocele

PF-083

Nursing Symposium

Post-operative management of pain for children undergoing neurosurgical procedures

Tom Kennedy¹.

¹Great Ormond Street Children's Hospital NHS Foundation Trust, London.

Objective: Management of Pain for children post Neurosurgical Procedure can be a complex issue. Each individuals experience of pain and ability to cope with pain can differ which in turn can affect what medical or non-medical interventions are appropriate or effective. Effective Post-operative management of Pain aids in the recovery process and healing of the patient. The Objective of this presentation is to discuss the current practices used at GOSH, looking at the analgesic ladder, use of NCA/PCA medication and the effectiveness of this method. Nurses are involved in this way of managing pain. The Presentation will also report on the findings of a qualitative piece of research looking at pain management effectiveness. There will also be discussion of what non-medicinal alternatives are available to staff that could be useful in practice for some patient groups.

Methods: A qualitative approach will be taken, utilising a questionnaire that both parents and the child can complete asking them how they felt their pain was managed post procedure.

Results: Results will be discussed once available as the study will be done over the coming summer months. The presentation will

also include discussion on what methods of pain management are effective for our patient group.

Conclusion: Findings of the study will be reported and the nurses role in effective pain management discussed. Aims for the future will also be highlighted.

Keywords: Post operative pain management

PF-084

Nursing Symposium

Unusual presentation of osteosarcoma as a posterior fossa lesion

Kamilah Dowling¹.

¹The Children's Hospital at Montefiore, Bronx.

Objective: To discuss an unusual presentation of a large posterior fossa osteosarcoma in an asymptomatic 7-year-old male.

Methods: Using MRI, CT scan images and intra-operative photos, information will be presented on the presentation and treatment for this previously healthy patient, who presented with a 2-3 fingerbreadth wide, raised bony mass over the occipital area just below the inion. By parents report, mass developed after a fall in school where he hit that area of the head.

Results: Imaging revealed a large heterogeneous mass that expands the right occipital bone and exerts mass effect on the right cerebellum, the mass extends medially and superiorly to the inion where the outer table is destroyed and the mass extends in to the soft tissues with areas of posterior calvarial destruction. Pre-op diagnosis - large right cerebellar holohemispheric tumor involving the neck, inion, posterior fossa bone with dural invasion and compression of cerebellum and brainstem. Patient underwent a right cerebellar craniectomy with resection of a very large bone and intra-dural tumor. Neuropathology revealed a high-grade osteosarcoma with giant and pleomorphic cells.

Conclusion:

Osteosarcomas account for 2% of pediatric cancers, most occur between the age of 10 and 30. Although rare, osteosarcoma should be included in the differential for patients presenting with a raised lesion with or without a trauma history. A specialized medical center with an experienced multidisciplinary team is key to achieve optimal survival and quality of life.

Keywords: osteosarcoma, posterior fossa, scalp mass

PF-085

Nursing Symposium

Endoscopic third ventriculostomy in conjunction with choroid plexus coagulation - a useful treatment for post haemorrhagic or meningitic hydrocephalus in neonates, where shunting is not possible

Jenny Sacree¹.

¹Bristol Royal Hospital For Children, Bristol.

Objective: Whilst VP shunting is the treatment of choice for Cerebral Spinal fluid (CSF) diversionary therapy in neonates with post haemorrhagic or meningitic hydrocephalus, there are scenarios where this is not possible due to: size, both physical size of the baby and the size of the vessels (for VA shunting); or having a hostile abdomen, due to scarring, infection or abdominal wall anomalies. This talk will explore the concept of endoscopic third ventriculostomy in conjunction with choroid plexus coagulation to treat post haemorrhagic or meningitic hydrocephalus where shunting is not possible. It will look at the evidence, past and present, and present several clinical cases where this technique has been utilized.

Keywords: hydrocephalus

PF-086

Nursing Symposium

Management of post-operative care for cranioplasty patients

Cyprienne Coen¹.

¹Asklepios Children's Hospital, Sankt Augustin.

Objective: To discuss the management of cranioplasty patients following surgery.

Methods: An understanding of the reasons for undertaking cranioplasty are discussed: Cranioplasty is a surgical technique to correct defects of the skull of various etiologies. Most often typical skull deformities resulting from premature closures of the skull sutures are treated with cranioplasty. Possible causes of cranial defects or deformities include; birth trauma, other trauma mechanisms, tumors, and craniosynostosis.

Results: The pathological fusion of the cranial sutures can result in cosmetic as well as functional problems including impaired eye movements and increased intracranial pressure {intracranial hypertension}. Postoperative nursing care is essential in achieving optimal results in patients after cranioplastic surgery and this is discussed below.

Conclusion: This includes continuous monitoring, Hb-control, care of intravenous lines, care of wound sites, and avoiding pressure on the skull surface through special positioning and support of the patient. Optimizing the cranial structure through positioning is another very important aspect of the postoperative care. It can avoid adjuvant helmet therapy in minimal invasive cranioplastic procedures. Possible accompanying drug therapies will also be discussed. This presentation should give you an insight into the postoperative care of cranioplasty patients.

Keywords: Cranioplasty post operative care

PF-087

Nursing Symposium

Pre-surgical work up for intractable epilepsy: road to the operating room

Esther Kho Uy¹.

¹Montefiore Medical Center, Bronx.

Objective: From the first seizure experience to the visit with neurologist/epilepsy specialist to choosing the appropriate seizure treatment. Advance diagnostic techniques play a very important role in making accurate diagnosis to help in the management of intractable epilepsy. This presentation will provide information on the different diagnostic techniques used for Intractable Seizure that can lead to surgical treatment when medication and other methods of treatment fails. The objective of this presentation is to help nurses take good care of seizure patients with necessary knowledge.

Keywords : diagnostic technique, intractable seizure

PF-088

Nursing Symposium

A critique of the epilepsy patient experience from diagnosis to surgery: the role of the children's nurse in the provision of holistic quality evidence based care

Claire Gubby¹.

¹Neurosciences Unit, Great Ormond Street Hospital For Children, London.

Objective: To determine how nursing practice influences the patient experience throughout the surgical pathway from diagnosis

and identification as a potential surgical candidate through to the surgery. To analyse current practice at Great Ormond Street Hospital identifying where improvements can be made regarding the impact on the patient experience.

Methods: Questionnaires to be given to patients/families following epilepsy surgery on the Neurosciences Unit at Great Ormond Street Hospital regarding their patient experience. Informal interviews with the key health care professionals who have an impact on the process and patient experience. Analysis of the current practices and pathway used at Great Ormond Street Hospital and the influence of the nurse on the patient experience.

Results: Reflective nursing practice and ongoing assessment of the patient experience will always identify areas for development and improvement. The outcome of the current methodology is yet to be determined and will be discussed during the presentation.

Conclusion: With constantly evolving practices and advancing surgical techniques it is likely that the incidence of potential surgical candidates increases over time. The pathway for epilepsy surgery is a lengthy process with regular hospital visits and admissions for telemetry and investigations. Many of these investigations can be invasive both clinically and psychologically and the impact this process has on the patient and family is not often discussed or evaluated. It is reasonable to suggest that, as frontline health care professionals, nurses have a significant influence on the patient experience throughout. Through careful examination of the patient experience throughout their surgical pathway process, it may be possible to identify how nurses can best support and prepare the patient and family for epilepsy surgery.

Keywords: Nursing care following epilepsy surgery

PF-089

Nursing Symposium

The management of vagus nerve stimulation therapy in children

Nicola Barnes¹.

¹Great Ormond Street Hospital, London.

Objective: Vagus Nerve stimulation therapy (VNS) is a non-pharmacological adjunctive treatment of refractory epilepsy where epilepsy surgery may not be a viable option, several antiepileptic medications may have failed or unacceptable side effects of medications present. Patients are seen in nurse led clinics for the first year following commencement of therapy. We however are unable to predict outcomes prior to insertion and how as nurses can we best manage parental aspirations for therapy when efficacy may take time. Can we improve how we select and counsel patients prior to insertion of device, to ascertain which patient groups on evaluation for suitability of therapy may respond more positively with minimal potential side effects of hoarseness to voice, cough or sleep apnoea.

Methods: At Great Ormond Street we support approximately 50 children from age 3-19 years with VNS therapy. Recent studies have shown VNS can provide 50% of patients with >50% seizure reduction, improved post ictal recovery from seizure, improved mood and alertness. (Healy et al 2013). The recent introduction of the Aspire SR VNS has been exciting as may lead to greater benefit for our patient groups that experience ictal tachycardia. Review of clinical data from recent implants will be made as to effect on seizure control, tolerance of therapy, effectiveness of magnet and quality of life.

Results: Discussion of case studies will be made of two cases to illustrate the potential effectiveness of VNS therapy.

Conclusion: Vagus nerve stimulation therapy continues to be an effective choice of treatment for young people with epilepsy, careful counselling of patient and family can be completed to assist in managing parental aspirations when many medications may have failed.

Keywords: VNS, nursing, children, epilepsy

PF-090

Nursing Symposium

A case study of a surgical treatment of pediatric intractable epilepsy

Jinyoung Glynn¹; Esther Uy¹.

¹Montefiore Medical Center, Bronx.

Objective: Intractable epilepsy (IE) is a seizure disorder that is difficult to control despite treatment of 2 or more antiepileptic drugs (AED). Children with IE are faced with negative impact on their quality of life physically, emotionally, and socially. Surgical treatment may provide them with a sense of normalcy when medications and other treatments fail. A group of specialists work together in the operating room to ensure the best result for patient with Intractable epilepsy.

Keywords: pediatric, intractable epilepsy

Monday, 5 October 2015

16:00 – 17:15

Flash Presentations

Oncology Flash Parallel Presentations I: Surgical approaches

FL-001

Posterior fossa tumor

Vermian saving approaches for surgery of pediatric tumors involving the IVth ventricle

Elvis J Hermann¹; Shadi Al-Afif¹; Joachim K Krauss¹.

¹Department Of Neurosurgery, Medical School Hannover, Hannover.

Objective: Midline tumors of the posterior fossa involving the fourth ventricle are one of the most common tumors in childhood. The aim of the surgery is to remove as much tumor as possible while preserving neurological function. To avoid vermian-splitting some surgical approaches are increasingly used like the telovelar approach (TA), infratentorial-supracerebellar approach (ITSCA) or a combination of the infratentorial-supracerebellar and telovelar approach (CA). We present our experience in the using of the vermian-saving approaches.

Methods: Forty three pediatric patients with posterior fossa tumors involving the fourth ventricle were operated microsurgical with avoiding the splitting of the cerebellar vermis. Data were collected retrospectively for all patients with respect to the degree of resection achieved, complications and the incidence of posterior fossa syndrome.

Results: Telovelar approach (TA) was performed in 60% of the patients, (ITSCA) in 9% of the patients, (CA) in 7% of the patients. An amount of 23% of the tumors could be accessed directly because of tumor breaking through the cerebellar cortex. A complete resection was achieved on postoperative MRI in 72% of the patients. Residual tumor was found in 28% (average diameter 0.9 cm, range: 0.5-1.6 cm). In 65% the cerebellar vermis was completely intact in postoperative MRI, in 35% of the patients there was limited lesion to the vermis. Seven percent of the patients had evidence of posterior fossa syndrome in the postoperative follow-up period, all of them with lesions of the cerebellar vermis. All had resolved at the last follow-up.

Conclusion: Vermian-saving approaches offer the possibility of safely removing pediatric midline tumors. Splitting of the cerebellar vermis is not necessary for removal of such tumors. However, lesion of the cerebellar vermis cannot be always avoided, when invaded by the tumor.

Keywords: IVth ventricular tumors, approaches, children

FL-002

Oncology

Telovelar approach in giant 4th ventricular medulloblastomasAlp Özgün Börcek¹; Ayfer Aslan¹; M. Kemali Baykaner¹.¹Division of Pediatric Neurosurgery, Gazi University Faculty of Medicine, Ankara.**Objective:** Literature has a long going debate on the limitations of this telovelar approach. This study aims to present the results of this approach in patients diagnosed with medulloblastoma after resection of their giant tumors.**Methods:** From August 2011 to April 2015, 33 patients presenting with midline posterior fossa mass were operated in Gazi University Faculty of Medicine Division of Pediatric Neurosurgery. A retrospective analysis was performed to demonstrate main characteristics, surgical and radiological results of the patients.**Results:** Of these 33 patients, 11 patients were diagnosed to harbor a medulloblastoma postoperatively. Mean age of the study population was 7.36 years. There were 8 (72.7%) males and 3 (27.3%) females. Mean tumor volume was 39.1 cm³. According to the intraoperative judgement of the surgeon, telovelar approach resulted in 2 (18.2%) subtotal resections and 9 (81.8%) gross total resections. Mean follow-up duration was 12.3 months (Range: 1-28 months). All tumors that judged as gross totally resected intraoperatively were demonstrated as gross totally resected at the postoperative follow up imaging. There were 3 postoperative neurological complications all of which were related to increased cerebellar ataxia and gait difficulties. There were no cerebellar mutism in this series. All patients were alive and under oncological treatment at the last follow-up.**Conclusion:** In all of the cases, using bilateral telovelar approach, no significant difficulty was observed in reaching to any parts of the fourth ventricle including the aqueduct of sylvii and the roof of the ventricle. The two subtotal resections in this series were due to the invasiveness of the tumors, rather than the limitations of the approach. Presence of a giant tumor in the 4th ventricle facilitates the approach in almost all cases. We recommend usage of telovelar approach for midline fourth ventricular giant tumors**Keywords:** Fourth ventricle, Medulloblastoma, Telovelar Approach

FL-003

Posterior fossa tumor

The role of telovelar approach in fourth ventricular surgery: a new perspectiveEhab M Eissa¹.¹Neurosurgical Department-Kasr Alainy Medical School -Cairo University, Cairo.**Objective:** The aim of this study is to evaluate the efficiency and safety of the telovelar approach for removing 4th ventricular lesions through identifying and preserving important neurovascular structures.**Methods:** This is a combined retrospective and prospective study on forty children (≤12 years) suffering from fourth ventricular tumors using the telovelar approach from 2005 to 2014.**Results:** This approach provided adequate exposure in all cases and the narrow working angle was efficiently compensated by changing the angle of the microscope and operating table. The brainstem and posterior inferior cerebellar artery (PICA) were early identified and preserved in all cases. Potential tumor attachment was observed at the floor of the fourth ventricle in twenty two (55%) cases. Out of brain stem cases which constitute five cases, 16 of the remaining 17 cases (94%) had focal attachment at any area of the caudal fourth ventricular floor e representing an inverted triangle with the obex inferiorly and the level of lateral recesses bilaterally and two cases (11,7%) was attached at any area of the lateral aspect of the rostral fourth ventricular floor which was the only point point of attachment in one of them. None of these tumors infiltrated the area of the cerebral aqueduct.Gross total excision was achieved in 45% of cases and near total excision (leaving < 1.5 cm²) was possible in 25% due to focal tumor attachment at one or more of the previously mentioned areas. However, debulking was only possible in 30%, because in other cases, the tumor either originated from the brain stem or was attached extensively to the floor.**Conclusion:** The main advantage of the telovelar approach is the early identification and preservation of the brain stem**Keywords:** telovelar approach, cerebellomedullary fissure, fourth ventricular tumors

FL-004

Posterior fossa tumor

Telovelar approach for midline posterior fossa tumors in pediatrics: 25 cases experienceMohamed Refaat¹; Ehab Elrefae¹; Walid Elhalaby¹.¹Neurosurgery Department, Cairo University, Cairo.**Objective:** The classic transvermian route for excision of fourth ventricular tumors could be associated with post-operative cerebellar mutism. The telovelar approach leads to the fourth ventricle via the cerebellomedullary fissure, avoiding retraction on the dentate nuclei and vermian incisions, hence lowering the incidence of cerebellar mutism. The aim of this study is to evaluate the telovelar approach for excision of fourth ventricular tumors.**Methods:** This retrospective study was conducted on successive 25 cases with midline posterior fossa tumors collected between 2012-2014. All cases were operated upon via the telovelar approach succeeded by microscopic tumor excision. Follow up data were collected regarding the degree of excision (total, near total and subtotal), and post operative complications. Mean follow up period was 5 months.**Results:** Age ranged from 3 to 8 years (mean 5.6 yrs). The most common was medulloblastoma (13 cases), followed by ependymoma (10 cases), and then choroid plexus papilloma (2 cases). Gross total excision was achieved in 8 cases (32%), near total excision (>80% of tumor volume) in 14 cases (56%), and subtotal excision (<80% of tumor volume) in 2 cases (8%). We experienced postoperative cerebellar mutism in 2 cases (8%) which improved in one case within the long term postoperative follow up. None of the cases had truncal ataxia. Two cases (8%) had postoperative facial palsy, while 3 cases (12%) had postoperative bulbar affection. There were two mortalities in this study, that were related to bulbar palsy and chest infection.**Conclusion:** Telovelar approach to the fourth ventricle is becoming a widespread technique for removing midline posterior fossa tumors. It gives excellent access and visualization to the fourth ventricle, with a low incidence of postoperative cerebellar mutism.**Keywords:** Fourth ventricle, telovelar, cerebellar mutism, medulloblastoma, ependymoma

FL-005

Posterior fossa tumor

Efficacy of ETV in the treatment of hydrocephalus after removal of posterior fossa tumor in childrenGeorge Papaevangelou¹; Christos Chamilos¹; Spyros Sgouros¹.¹"Mitera" Childrens Hospital, Athens.**Objective:** The purpose of this study was to analyze the efficacy of Endoscopic Third Ventriculostomy (ETV) in the management of hydrocephalus after posterior fossa tumor surgery in children.**Methods:** A retrospective review was performed of the management of hydrocephalus after 33 posterior fossa tumor resections in 29 patients performed by one surgeon between 2008-2015 (one patient had 4 operations

and another 2 operations for recurrence). All patients had tumour excision as the first operation, regardless of the size of the ventricular system. Management of hydrocephalus was instituted after tumour excision when this became clinically necessary due to symptoms of intracranial hypertension in the presence of large ventricles, with or without pseudomeningocele.

Results: Hydrocephalus was present before tumour resection in 17 patients (51%). After tumour excision, 12 patients in total (36%) required permanent treatment for hydrocephalus, 2 of those did not have hydrocephalus at presentation, had laterally placed tumours but developed it after tumour resection. In 10 patients ETV was performed as first procedure; 8 of those required subsequent ventriculo-peritoneal shunt. Hence the success rate of ETV was 20%. All patients that ETV failed had pseudomeningocele post-operatively. Of those who had ETV that failed, 2 patients had wound CSF leak postoperatively, and one of those developed frank CSF infection.

Conclusion: ETV when employed in the management of hydrocephalus after removal of a posterior fossa tumour in children has a poor success rate. Presence of pseudomeningocele is a negative predisposing factor for ETV success.

Keywords: posterior fossa tumour, hydrocephalus, ETV, shunt

FL-007

Oncology

Combined pre-operative OsiriX 3D topographic cerebral surface anatomy and intra-operative motor strip mapping for resection of motor eloquent area lesions in pediatric patients

Ivan Verdu-Martinez¹; Pablo Gonzalez-Lopez¹; Samer Elbabaa²

¹Miguel Hernandez University, Alicante; ²Saint Louis University School of Medicine, Saint Louis.

Objective: Microsurgical resection of neoplastic and vascular brain lesions in the motor eloquent area represents a great challenge for neurosurgeons. Recent literature confirmed the usefulness of OsiriX software as a pre-operative surgical planning tool. In this study, we analyze the combined usefulness of preoperative 3D topographic cerebral surface anatomy to identify the central sulcus and motor strip, with intra-operative motor strip mapping in pediatric patients with motor strip lesions.

Methods: Clinical and radiological data were reviewed for five pediatric patients (3M/2F) with motor strip lesions (3 cavernous vascular malformations, 1 GBM and 1 Oligoastrocytoma). Average age was 14 years. Intra-operative motor strip mapping was performed using phase reversal of somatosensory evoked potentials to identify the central sulcus. The results were correlated with pre-operative 3D Topographic Cerebral Surface Anatomy surgical planning obtained using OsiriX free software for Mac computers. 3D FSPGR (fast spoiled gradient echo) and 3D T1 with gadolinium sequences for OsiriX planning were obtained. Clinical motor and rehabilitation outcomes were assessed post surgery and at conclusion of study.

Results: All surgical procedures were performed by the senior author. Using SSEP phase reversal technique, the central sulcus was successfully localized intraoperatively in all cases. Motor strip and lesional localization had an excellent accuracy in all cases when compared to 3D Topographic Cerebral Surface Anatomy and reconstruction of neurovascular structures. Gross total resection achieved in all cases with no recurrence noted (average follow-up 16 months) except in GBM case which recurred after 11 months. All preoperative motor deficits were present after the surgical procedures, but all of them improved within 1-2 months after surgery and all patients were functionally independent and ambulatory at time of follow up.

Conclusion: Combined use of 3D topographic reconstruction method and intra-operative motor strip mapping can be useful in planning safe gross total resections of eloquent motor lesions in pediatric patients.

Keywords: OsiriX, operative planning, eloquent brain, motor strip mapping, 3D topographic surface anatomy

FL-008

Oncology

Optic pathway Glioma in children with neurofibromatosis 1

Hamilton Matushita¹; Daniel Cardeal¹; Fernanda Andrade¹.

¹Pediatric Neurosurgery - Dept. of Neurosurgery São Paulo University, São Paulo.

Objective: The aim of this study was to present our experience in the management of Optic pathway Glioma (OPG) with neurofibromatosis-1 (NF1) in children and contribute to the knowledge of this unpredictable entity.

Methods: Clinical charts and imaging studies of 24 consecutive cases of OPG with NF1 in children, were retrospectively studied.

Results: The age at diagnosis varied from 1-16 years (mean = 5,9 years). Ten patients were asymptomatic for visual complaint, although 6 demonstrated visual impairment on ophthalmologic evaluation. All other 14 children presented with some visual complaint (visual blurring 9, proptosis 4, strabismus 2, and palpebral swelling 1). Four out of 9 with visual blurring, had amaurosis of at least one side. Three patients developed a secondary tumor. MR imaging demonstrated that the main pattern of optic pathway involvement was tubular thickening of optic nerve and chiasm. Chiasmatic/hypothalamic mass were present in 3 cases, and with optic tract extension in only 2 cases. Treatment consisted of: observational in 13, chemotherapy in 8, debulking of chiasmatic-hypothalamic mass in 3, total removal of unilateral orbital tumor in 2, and chemotherapy + radiotherapy in 1 case. Mean follow up ranged from 1- 15 years (mean = 4,4 years). During follow up all patients were alive, and from 13 patients initially in surveillance, 2 demonstrated visual deterioration.

Conclusion: OPG in NF1 were diagnosed in very young age, most of them was asymptomatic for visual complaints, although ophthalmologic evaluation may demonstrated visual impairment in many of these patients. Severe visual deficit may also be demonstrated even in symptomatic children. Anterior visual pathways were more frequently affected with thickening of the optic nerve and chiasm. Best treatment is still controversial, but initial observation is advisable, mainly for those asymptomatic cases, with no visual impairment.

Keywords: optic pathway glioma, neurofibromatosis, children, astrocytoma, brain tumor.

FL-009

Oncology

The surgical treatment of pineal region tumors in infants under three years

Xiaozheng Ling¹; Baocheng Wang¹; Feili Liu¹; Jie Ma¹.

¹Xin Hua Hospital Affiliated To Shanghai Jiaotong University School Of Medicine, Shanghai.

Objective: Pineal region tumors in infants and toddlers (<3 years old) are a challenge for surgery, demonstrating dismal clinical outcomes. To explore a reasonable treatment plan, this paper reviewed the operative treatment of pineal region tumors under 3 years old in my hospital.

Methods: A retrospective study was done on 13 patients with pineal region tumors under 3 years-old. The clinical characteristic, surgical approach and prognosis were reviewed. All of the patients were operated on via infratentorial supracerebellar approach (n=8), trans-frontal-callosal-interforniceal approach (n=2), paramedian infratentorial approach (n=1), combined approach (n=2).

Results: Tumor was totally removed in 8 patients, subtotally resected in 3 and partially resected in 2. After surgery no patients fell in coma or died. Two patients died with abandoning treatment due to infection. Follow-up time was 3-30 months, 9 cases had resumed normal life, 2 cases died with recurrence.

Conclusion: Microsurgery is the prior option for children with pineal region tumor under 3 years, and favorable resection rate has been achieved in this series. Proper approach depended on every individual entitle successful resection.

Keywords: pineal region tumor, microsurgery, infant

FL-010

Oncology

Limitations in surgical excision of craniopharyngioma: personal experience of 68 cases

Ashis Pathak¹.

¹Director of Neurosurgery, Fortis Hospital, Mohali, India, Mohali (Chandigarh).

Objective: To explore the adaptation and modification in surgical approaches in excision of craniopharyngiomas based on anatomical considerations, morphology, size, consistency and relationship to vital structures.

Methods: Sixty eight cases of craniopharyngiomas operated upon by a single surgeon were analysed retrospectively. There were 45 males and 23 females with age ranging between 7 and 16 years. The anatomical location, size, morphological appearance on imaging and proximity to important structures were noted. The lesion was confined to a single compartment in 32 cases, dual compartment in 26 cases and multiple fossa in the remaining. The invasiveness of the lesion was decided based on its adherence to hypothalamus, engulfment of vital structures. Consistency of the lesion was divided into solid, partially cystic, purely cystic or predominantly calcified. Lesions were classified into small (<3 cm), medium (3–5 cm) and large (>5 cm). The various approaches initially planned and modifications made during surgery for better visualisation and approach to the lesions were evaluated from the operation notes. The amount of residual lesion was calculated. The factors leading to suboptimal excision were identified.

Results: Total excision was possible in 47 cases which were predominantly smaller in size, located mostly in a single compartment and had no adherence to vital structure. Excision was possible using single corridor in 27 cases, 2 corridors in 16 cases and > 2 corridors in remaining. The limiting factors for total excision were: 1. adherence to vital structure (common in cystic lesion), 2. calcification & 3. multi compartment lesions. There was 1 mortality in the series. Endocrinopathy was new in 10 and worse in 35 which were given adequate support.

Conclusion: Surgical excision of craniopharyngioma needs detailed evaluation and pre-operative planning with adaptive strategies during surgery to enable better approach and visualisation of the lesion in order to achieve the maximum excision.

Keywords: craniopharyngioma, endocrinopathy, suprasellar lesion

FL-011

Oncology

Management of craniopharyngiomas in pediatric age group: CCHE experience in 70 cases

Mohamed A. El Beltagy¹; Mostafa M. E. Atteya².

¹Chief Neurosurgeon, Children's Cancer Hospital Egypt (CCHE, 57357), Consultant Neurosurgeon And Professor, Neurosurgery Department, Kasr Al-Ainy School Of Medicine, Cairo University, Egypt, Cairo; ²Assistant Consultant of Neurosurgery, Children's Cancer Hospital Egypt (CCHE, 57357). Lecturer of Neurosurgery, Faculty of Medicine, Helwan University, Egypt, Cairo.

Objective: Management of pediatric craniopharyngiomas remains a challenging zone in neurosurgery. The aim is to assess management strategies adopted at our center.

Methods: Medical records of seventy children who had microsurgery for craniopharyngiomas at Children's Cancer Hospital Egypt (CCHE-57357) from January 2008 to June 2014 were reviewed. Based on management strategy, 2 patient groups were identified; group I included 41 cases (undergone gross total resection (GTR) or near total resection (NTR) with residual less than 1 cm) that had post-operative expectant follow-up and group II included 29 patients (undergone biopsy or subtotal resection (STR) with residual more than 1 cm) who received postoperative radiotherapy. Age ranged from 1 to 13 years (mean age 6.8 years).

Results: In group I, GTR and NTR were achieved in 29 cases and 12 cases, respectively. In this group, tumor recurrence occurred in eleven of the totally excised tumors and in six of subtotally excised tumors. In group II, STR and biopsy were achieved in 27 cases and 2 cases, respectively. In this group, tumor recurrence occurred in seven of subtotally excised tumors and in one of biopsied tumors. 5-years overall survival and event-free survival were 92.5% and 23.9% in group I as compared to 96.4% and 64.5% in group II, respectively. Vision had stabilized or improved in 89% of affected cases. Endoscope-assisted microsurgery was utilized in 33 cases (47%). Postoperative endocrine deficits occurred in 69% of patients (40% of which had endoscope-assisted microsurgery). Endoscope-assisted cases had better clinical outcomes without statistical significance.

Conclusion: Whenever craniopharyngioma was not dissectible from hypothalamus, STR and subsequent radiotherapy provided good outcomes. The recurrence of craniopharyngioma after surgery was less common in cases which received postoperative radiotherapy. Endoscope-assisted surgical exposure may play a role in decreasing morbidity and complications.

Keywords: craniopharyngioma, pediatric, brain tumors, endoscope-assisted, microsurgery

FL-012

Oncology

Evolution and therapeutic strategy for craniopharyngioma.

About 17 pediatric cases.

Blandine Grassiot¹; Alexandru Szathmari¹; Pierre-Aurélien Beuriat¹; Pascale Berlier²; Didier Frappaz³; Carmine Mottolese¹.

¹Service De Neurochirurgie Pédiatrique, Lyon; ²Service D'endocrinologie Pédiatrique, Lyon; ³Institut D'hématologie De D'endocrinologie Pédiatrique, Lyon.

Objective: Craniopharyngioma is a benign and rare hypothalamo-pituitary tumor with risk of long term morbidity. Since 2000, several changes of management of these lesions have been discussed combining initial intracystic chemotherapy, conservative surgical resection and the increasing use of proton radiotherapy. The aim of this study is to evaluate tumor evolution and morbidity of patients depending on the therapeutic strategy.

Methods: Between 2000 and 2012, we treated 17 children with craniopharyngioma aged from 3 to 17 years (8F and 9M). Treatment was initial surgery in solid forms for 8 patients or initial puncture and intracystic chemotherapy on Ommaya inserted catheter for large cystic forms in 9 patients. Chemotherapy used was Bleomycin in 5 patients and alfa-2A interferon in 4 patients. Secondary proton radiotherapy was used for residuum in 5 patients. Overall, 14/17 had a direct approach (8 at the time of diagnosis, 6 after chemotherapy) with complete removal of lesion in 6 cases and partial for 8. Among these patients, 3 were re-operated for recurrent lesion.

Results: The average follow-up is 5.8 years for this series. All except one patient is still alive. Overall life quality is good with only 1 patient with secondary narcolepsy. Global hormonal substitution is necessary in 10 (58%) and only 8 (47%) are substituted for desmopressin. Cognitive analysis showed 5 children with difficulties at school. 4 of 5 patients

submitted to proton therapy have a good cognitive evolution, but all of them developed endocrine deficiency.

Conclusion: The management of craniopharyngioma is complex and need a multidisciplinary approach. Initial intracystic therapy seems to allow good control of tumor with almost normal hormonal activity in the majority of patients. However, surgical resection is advocated in case of progression or solid lesions. Proton radiotherapy is necessary in case of residuum albeit it seems to accentuate hormonal deficiency.

Keywords: craniopharyngioma, proton therapy, intracystic chemotherapy

FL-013

Oncology

Risk of endocrinopathy in pediatric craniopharyngiomas treated with surgical intent to cure

Peter Jang¹; Edward F. Melamed¹; Erin N. Kiehna¹; J. Gordon McComb¹; Mark D. Krieger¹.

¹Division of Neurosurgery, Children's Hospital Los Angeles, Los Angeles.

Objective: The optimal management of childhood craniopharyngiomas is dependent on consideration of the risks versus benefits of treatment approach. High rates of surgical cure have been associated with high rates of postoperative endocrinopathy. This study examines the effect of surgical approach on the development of postoperative endocrinopathy.

Methods: We retrospectively analyzed under an IRB approved protocol a series of craniopharyngiomas managed at single institution with complete data sets available. Only cases treated with surgical intent to cure were included.

Results: 52 cases treated surgically with a goal of gross total resection (GTR) were identified. Surgical approaches included transsphenoidal (n=10), interhemispheric (n=23), subfrontal (n=10), and pterional (n=9). Extent of hypothalamic involvement (grade) was not uniform across the varied surgical approaches. 30 patients underwent GTR and 22 patients underwent subtotal resection (STR). Post-operatively, those who had GTR had a higher rate of anterior pituitary dysfunction: 26 (87%) versus 11 (50%) for those who had STR ($p < 0.01$, chi-squared). This result was consistent across all surgical approaches ($p < 0.05$, Fishers exact test). Higher grade was associated with increased incidence of anterior and posterior pituitary dysfunction ($p < 0.05$) regardless of surgical approach.

Conclusion: There is a higher rate of postoperative anterior pituitary dysfunction when there is complete resection of childhood craniopharyngiomas. This effect persists when controlling for the extent of hypothalamic involvement (grade) of the tumor and for the surgical approach.

Keywords: Craniopharyngioma, Endocrinopathy, Surgical Approach, Gross Total Resection

FL-015

Other

Endoscopic transnasal approach in pediatric cases: advantages and disadvantages - a review of 20 cases

Gokmen Kahilogullari¹; Cem Meco²; Suha Beton²; Murat Zaimoglu¹; Agahan Unlu¹.

¹Ankara University Faculty of Medicine, Department of Neurosurgery, Ankara; ²Ankara University Faculty of Medicine, Department of Otolaryngology, Ankara.

Objective: To discuss the endoscopic endonasal approach with advantages and disadvantages.

Methods: To analyse 20 pediatric patients that are operated via transnasal endoscopic approach in Ankara University Medical Faculty between years 2012-2014.

Results: 15 patients (75%) were male and 5 patients were female (25%). Median age for males and females were 5 and 17 respectively. Overall median age was 13. 6 of the cases (30%) were hypophysis adenoma, 3 craniopharyngioma (15%), 3 rhinorrhea (15%), 2 germinoma (10%), 2 meningocele (10%), 2 inflammation (10%), 1 angiofibroma (5%), 1 normal hypophysis tissue (5%). Among 13 patients with pathological results; gross total excision was achieved in 9 (69.2%), subtotal resection was achieved in 2 (15.3%). In 2 cases only biopsy was made. Among 20 patients that are operated, there was 10 cases of CSF leakage. CSF leakage was repaired with reconstructive methods such as nasoseptal flap, fascia lata flap or using fibrin products. There were no patients with rhinorrhea or meningitis. 5 patients had transient diabetes insipidus, 1 patient had temporary loss of lateral gaze and 1 case of pneumocephalus. There were no exits.

Conclusion: Endoscopic Transnasal approach is an effective method for surgery in pediatric patients. The main reason for this statement is that it is less invasive, and satisfactory treatment can be achieved. Sphenoid sinus pneumatization is the main handicap for this approach.

Keywords: Endoscope, Transnasal approach, Child, Advantage-disadvantage

FL-016

Oncology

Endoscopic resection of solid intraventricular tumors in children

Eveline Teresa Hidalgo¹; Aryan Ali¹; Jeffrey H. Wisoff¹; Howard L. Weiner¹; David H. Harter¹.

¹NYU Langone Medical Center, New York.

Objective: We report the feasibility and outcomes of endoscopic resection of select intraventricular tumors in children.

Methods: The clinical characteristics of 11 children with solid intraventricular tumors who underwent tumor resection were reviewed. 12 procedures were performed.

Results: Gross total resection was achieved in 11 of 12 cases (92%). Maximal diameter ranged from 9-26 mm (mean 16.6 mm). Pathology included subependymal giant cell astrocytomas (SEGA), ependymomas, non-germinomatous germ cell tumor (NGGCT) and pilocytic astrocytoma. Mean follow-up was 35 months (range 10-109 months). All patients returned to their neurological baseline following surgery. Local tumor recurrence occurred in one patient and distant recurrence in another. Complications occurred in one patient, no permanent morbidity or mortality occurred. Hydrocephalus was present preoperatively in 5 cases and was treated with tumor removal alone or with additional endoscopic third ventriculostomy. No patient required a ventriculoperitoneal shunt.

Conclusion: Neuroendoscopic gross-total resection of solid intraventricular tumors is a safe procedure in carefully selected pediatric patients.

Keywords: Neuroendoscopy, Intraventricular Tumors

Monday, 5 October 2015
16:00 – 17:15

Oncology Flash Parallel Presentations II: Clinical Science

FL-017

Posterior fossa tumor

Preliminary results of integrated proteomic platforms for posterior fossa tumors

Luca Massimi¹; Claudia Martelli²; Luca D'angelo¹; Gianpiero Tamburrini¹; Concezio Di Rocco³; Massimo Caldarelli¹; Irene Messina⁴; Maria Teresa Sanna⁴; Federica Iavarone²; Morena Arba²; Federica Vincenzoni²; Ilaria Inserra²; Diana Valeria Rossetti²; Massimo Castagnola²; Claudia Desiderio⁵.

¹Pediatric Neurosurgery, Catholic University Medical School, Rome; ²Biochemistry And Clinical Chemistry, Catholic University Medical School, Rome; ³INI, Hannover; ⁴Life And Environment Science, University of Cagliari, Monserrato; ⁵Molecular Biology And Chemistry, CNR, Rome.

Objective: The goal of the present study is to apply an integrated proteomic approach for the comparative tissue profiling of tumors of different grade, as medulloblastoma and pilocytic astrocytoma.

Methods: A top-down/bottom-up integrated proteomic approach based on LC-MS and 2-DE analysis was applied for comparative characterization of 4 cases of medulloblastoma and 4 cases of pilocytic astrocytoma of the posterior fossa.

Results: The results showed different proteomic profiles of the two tumors and evidenced interesting differential expression of several proteins and peptides. Top-down proteomics of acid-soluble fractions of brain tumor homogenates ascribed a potential biomarker role of malignancy to β - and α -thymosins and their truncated proteoforms and to C-terminal truncated (des-GG) ubiquitin, resulting exclusively detected or over-expressed in the highly malignant medulloblastoma. The bottom-up proteomics of the acid-soluble fraction identified several proteins, some of them in common with 2-DE analysis of acid-insoluble pellets. Peroxiredoxin-1, peptidyl-prolyl cis-trans isomerase A, triosephosphate isomerase, pyruvate kinase PKM, tubulin beta and alpha chains, heat shock protein HSP-90-beta and different histones characterized the medulloblastoma while the Ig kappa chain C region, serotransferrin, tubulin beta 2A chain and vimentin the pilocytic astrocytoma.

Conclusion: The two proteomic approaches well complemented each other in characterizing the proteome of brain tumor tissues and in disclosing potential disease biomarkers to be validated in a future study on individual samples of both tumor histotypes.

Keywords: proteomic, medulloblastoma, pilocytic astrocytoma, top down approach

FL-018

Posterior fossa tumor

Medulloblastomas in young children: what is new for prognostication?

Raghav Singla¹; Deepak Gupta¹; Kavneet Kaur¹; Chitra Sarkar¹; Bhawani S Sharma¹; Ashok K Mahapatra¹.

¹Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi.

Objective: Medulloblastoma (MB) is the most common malignant brain tumor in childhood. Currently, it is classified on the basis of morphological characteristics. However, recent molecular studies have shown that it is not a single disease entity, but a cohort of various

molecular subgroups. There are four core molecular subgroups, namely WNT, SHH, Group C and Group D. This subgrouping has significant prognostic relevance and is expected to guide the treatment of MB patients in the future. The objective of the study is to establish the feasibility of molecular subgrouping of medulloblastoma by using immuno-histochemistry (IHC) and Fluorescence In-Situ Hybridisation (FISH) and see if it has the same implication on the prognosis as that by gene expression profiling.

Methods: A retrospective study was performed on the cases of biopsy-proven pediatric (age < 18 years), range: 6-16 yrs, mean: 10.2 years, sex ratio : 2 :1) medulloblastoma, who underwent surgery at our center between 2003 and 2011. Clinical and radiological data was obtained from the Neurosurgery records. Slides were reviewed and molecular subgrouping was done using IHC and FISH.

Results: A total of 52 pediatric cases (40 males, 12 females; mean age 9.4, range 1-18 years) were studied. On molecular subgrouping, nine (17.3%) were WNT, twelve (23.1%) were SHH, 5 (9.6%) were group C and 24 (46.2%) were group D.

Conclusion: The authors advocate that IHC and FISH can be used for molecular subgrouping of MB in economically constrained set up with the same prognostic bearing, as the much more expensive gene expression profiling.

Keywords: medulloblastoma, molecular subtyping, prognostic markers, posterior fossa tumors

FL-019

Oncology

Artesunate inhibits glioma growth through inhibition of HDAC6

¹Xiaozheng Ling; Feili Liu¹; Jie Ma¹.

¹Xin Hua Hospital Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai.

Objective: To investigate the effects of Artesunate (AST) on glioma cells and identify the mechanisms underlying its potential anticancer effects.

Methods: AST was investigated for its in vitro and in vivo antitumor effects and its effects on the protein expression of HDAC6.

Results: We showed that AST inhibited the viability and proliferation of human glioma cells in a dose-, time -dependent manner through MTT and CFDA SE assays. We observed that 8 h after being AST-treated, compared with control group, the migratory cell numbers were significantly decreased. Flow cytometry assays revealed G0/G1 phase arrest and apoptosis in U251 and A172 cells. Western blot analysis of AST-treated cells revealed the down regulation of HDAC6. Moreover, AST also inhibited the tumor growth in nude mice carrying subcutaneous A172 tumor xenografts and IHC analysis suggested the hypo expression of HDAC6 after AST-treated.

Conclusion: These data demonstrated that AST inhibits glioma growth through inhibition of HDAC6. These findings indicate the promising antitumor potential of AST for further glioma treatment applications.

Keywords: Artesunate, gliomas, HDAC6, inhibition

FL-021

Oncology

Association study of susceptibility to pediatric embryonal brain tumor and its clinic management in Chinese Han population

Baocheng Wang¹; Jie Ma¹.

¹Xinhua Hospital, Shangha.

Objective: To investigate the genetic susceptibility to pediatric EBT in 89 Chinese patients through a case-control association study and to explore all combination of genotypes in positive loci as potential biomarkers for the clinical prognosis of pediatric EBTs.

Methods: We recruited 89 patients with EBT and 190 healthy adults as the control in Chinese Mainland. A total of 105 germline variants of 22 candidate genes involved in histone modification and three core signal pathways was investigated using the high-throughput Sequenom MassArray genotyping platform, which would provide a renewed interest in strategies for the early risk detection and prevention of pediatric EBT. Finally, we explored their clinical prognosis and the association with all kinds of genotypes through phenotype-genotype association analysis.

Results: After statistical case-control analysis, eleven SNPs in six genes explain the association with the risk of pediatric EBT. Some haplotypes in positive susceptible genes are significantly associated with pediatric EBT in the case-control haplotype analysis. The genotype of rs20551 shows a trend towards association with shorter survival times. According to the process of tumor progression in 89 pediatric EBTs, the cross-case comparative analysis manifested that six loci locating in five genes were associated with their clinical prognosis. Two haplotypes "GCCAA" and "GCTGA" in PTCH1 gene reaches statistical significance with the different progression of pediatric EBT in the cross-case haplotype analyses. The condition with these haplotypes predict the possible postoperative progression of pediatric EBTs and the stable, respectively.

Conclusion: A series of genetic variations of hot gene are associated with individuals at high risk of pediatric EBT, germline genetic variants associated with AKT pathway play a complex role in the development of pediatric EBT. The haplotypes of PTCH1 and genotype of rs20551 may show true association as potential biomarkers with the clinical prognosis of pediatric EBTs.

Keywords: embryonal brain tumor, pediatric, germline, susceptibility, prognosis

FL-022

Oncology

Brain tumors in patient under the age of 1 year

Zohreh Habibi¹; Farideh Nejat¹; Sara Hanaei¹; Ehsan Moradi¹.

¹MD, Tehran.

Objective: Brain tumors in infants have different clinical presentations, anatomical distribution, histopathological diagnosis, and clinical prognosis compared with older children.

Methods: A retrospective analysis was done in patients less than 12 months old who were operated on for primary brain tumor in Children's Hospital Medical Center since 2008 to 2014.

Results: Thirty one infants, 20 males and 11 females, with the mean age of 7.13 months (0.5 - 12) were enrolled. There were 16 supratentorial and 15 infratentorial tumors. The presenting symptoms included increased head circumference (16); bulge fontanel (15); vomiting (15); developmental regression (11); sunset eye (7); seizure (4); loss of consciousness (4); irritability (3); nistagmus (2); visual loss (2); hemiparesis (2); torticollis (2); VI palsy (3); VII,IX,X nerve palsy (each 2); and ptosis (1). Gross total and subtotal resection was performed in 19 and 11 cases, respectively. Fourteen patients needed external ventricular drainage in perioperative period, from whom four infants required ventriculoperitoneal shunt. One patient underwent ventriculoperitoneal shunting without tumor resection. The most common histological diagnoses were PNET (7), followed by anaplastic ependymoma (6) and grade II ependymoma. The rate of 30-day mortality was 19.3%. Eighteen patients are now well controlled with or without adjuvant therapy (overall survival; 58%), from whom 13 cases are tumor free (disease free survival; 41.9%), 3 cases have residual masses with fixed or decreased size (progression free survival; 9.6%), and 2 cases are still on chemotherapy.

Conclusion: Brain tumors in infants should be treated with surgical resection, followed by chemotherapy when necessary.

Keywords: Brain tumor, Infants, Surgical treatment

FL-023

Oncology

Infant brain tumors in Belgium: a multicenter pilot study

Edward Baert¹; Bart Depreitere²; Vera Vanvelthoven³; Mania De Praeter⁴; Jacques Born⁵; Gaus Koerts⁶; Mike Huylebroeck³; Cristo Chaskis⁷.

¹UZ Gent, Ghent; ²UZ Leuven, Leuven; ³UZ Brussel, Brussels; ⁴UZ Antwerpen, Antwerpen; ⁵CHR De La Citadelle, Liège; ⁶UCL - Cliniques Universitaires Saint-Luc, Brussels; ⁷CHU De Charleroi, Charleroi.

Objective: Infant brain tumors represent a rare entity. In order to map its frequency and characteristics in Belgium, the BSN pediatric section launched a joint effort pilot study to assemble a multicenter retrospective database. This is a preparatory initiative to a more profound study in collaboration with the Belgian Cancer Registry and with the Pediatric Oncologists.

Methods: Retrospective data on brain tumors diagnosed before the age of 1 year from 1995 until present could be retrieved from 7 large neurosurgery units.

Results: 67 infant brain tumor diagnosis were identified. Histology was available in 67 patients: 22 low grade and 14 high grade gliomas, 9 PNE T tumors, 8 plexus tumors, 2 hamartomas, 2 teratoid, 1 pineal/sarcoma/meningeoma and 7 undefined tumors. Of 49 cases with location data, 40 were supratentorial (22 hemispheric, 9 diencephalic, 8 visual pathway and 1 sellar) and 9 were infratentorial (6 cerebellar and 3 brainstem). Surgery details were available in 49 cases: total resection in 15, subtotal in 11 and partial resection in 7, biopsy only in 11 and no surgery in 5 patients. There were 21 documented deaths at a median interval of 4 months after diagnosis. Of 43 patients with documented ongoing follow up (median FU 41 months), 29 are alive to date (67,4%). For high grade tumors survival is 9/18 (50,0%). Outcome data was available in 28/29 survivors: 14 (50,0%) have cognitive impairment and 8 (28,6%) have visual deficits.

Conclusion: Low and high grade gliomas are the most frequent histology in our infant brain tumor series, followed by PNET and plexus tumors. Survival rate at ongoing follow up is 67,4%. The rate of cognitive impairment in surviving infants is high, which will be subject of further investigation.

Keywords: infant, brain, tumor, Belgium

FL-024

Posterior fossa tumor

Multicenter retrospective study of pediatric posterior fossa embryonal tumors in Belgium

Gaus Koerts¹; Bart Depreitere²; Edward Baert³; Mania De Praeter⁴; Cristo Chaskis⁵; Eveleen Buelens⁶; Raf Van Paesschen⁷.

¹Cliniques Universitaires St-Luc, Brussels; ²KUL, Leuven; ³UZ Gent, Ghent; ⁴UZ Antwerpen, Antwerp; ⁵CHU Charleroi, Charleroi; ⁶ZOL, Genk; ⁷St Augustinus, Antwerp.

Objective: Tumors of the central nervous system are the 2nd most frequent tumors in children and adolescents. Between 2004 and 2009 621 new diagnoses were registered in Belgium. There were 83 malignant embryonal tumors including 60 medulloblastomas. Since pediatric neurosurgery in not centralized in Belgium case load per center is relatively low compared with many foreign centers. This is the first attempt to perform a multicenter retrospective study of pediatric posterior fossa embryonal tumors in Belgium.

Methods: Retrospective multicenter study of the BSN pediatric section concerning pediatric posterior fossa embryonal tumors. Seven centers participated in this retrospective patient chart review. Thirty-eight patients could be identified between 2004 and 2009.

Results: Thirty-eight patients with an embryonal tumor in the posterior fossa were operated between 2004 and 2009. There were 33 medulloblastomas, 4 atypical teratoid rhabdoid tumors (ATRT), and 1 embryonal tumor with abundant neuropil and true rosettes (ETANTR). There was a male preponderance. Age ranged from 1.1 to 18.5 years (mean 7.7 years). Overall two-year survival rate was 71%. Overall five-year survival rate was 56%. In the medulloblastoma group the 2-year and 5-year survival were 75% and 58%, respectively. There was no surgical mortality. Three out of four ATRT died during follow-up confirming dismal prognosis of this entity.

Conclusion: Embryonal tumors, especially medulloblastomas are the most frequent malignant brain tumors in children. Although there is no centralization of pediatric brain tumors yet, survival rates are comparable to those of the literature. This study stimulates us to perform further analysis of morbidity and long-term side-effects in surviving patients and to develop multi-center prospective data collection for future research.

Keywords: posterior fossa tumor, Belgium, multicenter, embryonal tumors

FL-025

Oncology

Correlation of ADC map with histological grade in paediatric brain tumours

Aimilia Moraiti¹; Georgia Papaioannou¹; Spyros Sgouros¹.

¹"Mitera" Childrens Hospital, Athens.

Objective: The purpose of this study was to investigate the possible correlation between ADC values and histological grade of pediatric brain tumors.

Methods: The study included only patients with full MRI imaging with ADC sequences, treated between 2008–2015, in whom the original digital data were available. ADC maps were calculated. Minimum ADC signal intensity values were measured in the darkest area of the lesion. The values were correlated with benign / malignant tumour histology type.

Results: 20 patients with age between 4 months and 16 years old were included. Histology types were: 3 medulloblastomas, 3 pilocytic astrocytomas, 3 glioblastomas, 2 ependymomas, 2 germinomas, 5 diffuse astrocytomas grade II, 1 atypical teratoid rhabdoid tumor and 1 pineocytoma. Hence there were 9 benign and 11 malignant tumours. Minimum average ADC signal intensity values were: benign tumours: 1027,20 mm²/sec (range:378,5–1583), malignant tumours: 638,42 mm²/sec (range: 135,8–910,7). There was statistically significant difference (p=0,03, chi-square).

Conclusion: There is a strong correlation between minimum ADC signal intensity value and histological grade of the tumour. This may prove particularly useful in evaluating possible tumour recurrences.

Keywords: brain tumours, MRI, ADC map, malignancy

FL-026

Posterior fossa tumor

Posterior fossa syndrome after resection of medulloblastoma - can it be predicted from preoperative magnetic resonance imaging?

Haffenden Verity¹; Kim Phipps¹; Kshitij Mankad¹; Paul Smith¹; Owase Jeelani¹; Richard Hayward¹; Kristian Aquilina¹.

¹Great Ormond Street Hospital, London.

Objective: Posterior fossa syndrome (PFS), classically described after resection of medulloblastoma (MB), remains a challenging complication resulting in long-term speech, cognitive and motor deficit. Although

known to be related to cerebellar efferent pathway injury, it has not been possible to predict or prevent. Our objective was to determine whether PFS may be predictable from pre-operative magnetic resonance (MR) imaging in children with MB from a single-institution.

Methods: Retrospective review of medical records and MR images (1.5 T) was carried out for children presenting with MB to Great Ormond Street Hospital between 2003 and 2012. PFS was established through a scoring system incorporating mutism, ataxia, behavioural disturbance and cranial nerve deficits. Pre-operative and early/late post-operative imaging were blindly assessed by two experienced neuroradiologists. Structures related to the cerebellar efferents were evaluated.

Results: 56 consecutive children were identified. 12 (21.4%) developed PFS. All had mutism; 8, 7 and 9 children also had ataxia, behavioural change and cranial nerve dysfunction. On pre-operative scans children who developed PFS had similar tumour size to those who did not. Distortion of the dentate or red nuclei, superior cerebellar peduncles (SCP) or decussation was not statistically different. In both early (median 4 days) and late (median 31 months) post-operative scans, hyperintensity along the SCP on T2-W images was commoner in the PFS group (p < 0.001, chi squared test). There were no significant differences in diffusion characteristics of dentate and red nuclei, decussation or SCP in the early post-operative scans, and no significant changes in the inferior olivary nucleus on late scans.

Conclusion: Expert evaluation of pre-operative MR scans was unable to predict PFS in these children. T2-W hyperintensity in the SCP's is consistent with other studies and underlines the role of cerebellar efferent injury in PFS.

Keywords: medulloblastoma, posterior fossa syndrome, cerebellar mutism

FL-027

Oncology

Prognostic factors for cerebellar astrocytomas in children: a study of 135 cases.

Dattatraya Muzumdar¹; Ketan Desai¹; Trimurti Nadkarni¹; Atul Goel¹.

¹Department of Neurosurgery, Seth G.S.Medical College and King Edward VII Memorial Hospital, Mumbai.

Objective: To evaluate the prognostic factors influencing the length of survival of pediatric patients with cerebellar astrocytomas.

Methods: The clinical data of 135 patients under the age of 18 years with cerebellar astrocytomas were retrospectively analyzed. The radiological features, surgical findings, histology and adjuvant radiotherapy were reviewed. Patients were followed up on an outpatient basis.

Results: The age of the patients at presentation varied from 10 months to 12 years. The mean age at presentation was 7 years and 11 months. The average duration of symptoms was 5.8 months. The clinical features were predominantly related to intracranial hypertension and the location of the tumor. Forty-three tumors were located in the vermis and ninety-two in the cerebellar hemisphere. The brain stem was involved in 31 patients. All 135 patients had a preoperative contrast-enhanced CT scan. Midline vermian tumors were predominantly solid and enhancing, whilst the hemispheric tumors were cystic and nonenhancing. The tumors were graded into three subgroups based on histological characteristics: pilocytic astrocytoma, low-grade fibrillary astrocytoma and high-grade fibrillary astrocytoma. Total, radical and subtotal excision, as determined by the surgical impression, was performed in 109, 21 and 5 patients, respectively. Postoperative radiotherapy was administered to 18 patients. CSF diversion was carried out in 19 of 105 cases that had moderate to severe hydrocephalus.

Conclusion: Our study has shown that the location of the tumor (p < 0.05), histological grade (p < 0.001) and the extent of tumor resection (p < 0.001) have a significant and definitive relationship to the length of survival of pediatric patients with cerebellar astrocytomas. The patient's age (p > 0.05) does not influence the outcome.

Keywords: cerebellar, astrocytoma, pediatric

FL-028

Oncology

Cerebellar astrocytoma in a regional neuro-oncology centre; evaluating the need for post-surgical imaging surveillance

Hannah Brophy¹; Barry Pizer¹; Conor Mallucci¹; Shivaram Avula¹; Sue Hemsworth¹; Benedetta Pettorini¹; Dawn Williams¹; James Hayden¹.
¹Alder Hey, Liverpool.

Objective: Cerebellar astrocytomas constitute around 15 % of CNS neoplasms in childhood. Surgery aiming to complete resection, is the principal treatment modality, although need for and duration of post-surgical MRI surveillance is not well defined. In this respect, we conducted an evaluation of the outcome of cerebellar astrocytomas at our regional paediatric neuro-oncology centre.

Methods: Our tumour database was interrogated to identify all cases of cerebellar astrocytoma. Variables included patient age, gender, tumour grade, completeness of resection, duration of follow-up, remission status and adjuvant therapy.

Results: Between 1988 and 2007, 49 patients with cerebellar astrocytoma were reviewed - 43 pilocytic astrocytomas, 4 fibrillary astrocytomas, 1 ganglioglioma and 1 unspecified low-grade tumour. Thirty seven patients underwent complete resection (CR), none of whom relapsed. Of the 12 patients with incomplete resection, six patients progressed at between 4 months and 6 years. We thus changed the duration of MRI surveillance from 5 years to a maximum of 2.5 years (scans at 6, 18 and 30 months post surgery) in those patients who had undergone CR. Since this change in practise, a further 12 patients (all WHO grade 1) have been treated (data until December 2012-to allow > 2 years follow-up). Eleven tumours were completely resected and 1 had a near total resection. None of these 12 patients have relapsed (median follow-up - 4.9 years).

Conclusion: In this service evaluation, none of the 48 patients with completely resected cerebellar astrocytoma relapsed, supporting a change in practice to shorten the duration of post-operative surveillance. This has resulted in a time and cost saving to families and our health service. The data does, however, raise the question as to whether any surveillance imaging is required in completely resected pilocytic cerebellar astrocytoma. For incompletely resected tumours, a standard 5-year post-surgical imaging schema should be maintained.

Keywords: cerebellar astrocytoma, surgery, imaging, follow-up

FL-029

Posterior fossa tumor

Cerebellar pilocytic astrocytoma in childhood: investigating the long-term impact of surgery on cognitive performance and functional outcome

Thomas Pletschko¹; Anna Felnhofer¹; Doris Lamplmair¹; Christian Dorfer²; Thomas Czech²; Irene Slavc¹; Ulrike Leiss¹.

¹Department of Pediatrics and Adolescent Medicine, Medical University of Vienna, Vienna; ²Department of Neurosurgery, Medical University of Vienna, Vienna.

Objective: Prior results differ regarding the long-term effects of surgically removed paediatric cerebellar pilocytic astrocytomas (CPA). Thus, the aim of this study was to investigate the long-term impact on neurocognitive and functional outcome and to analyse age as an influencing factor.

Methods: Fourteen CPA patients were compared to the norm and to a group of fourteen high achieving peers regarding cognitive functioning, health related quality of life (HRQoL) and stress regulation. Mean follow-up time after diagnosis was 13.29 years (range: 3-21 years).

Results: Patients showed satisfactory academic achievement and did not differ from the norm except for the bodily dimension of HRQoL. However, there were marked differences in specific neurocognitive

functions between patients and high achievers. Age at diagnosis did not influence neurocognitive outcome.

Conclusion: CPA patients treated with surgery only seem to have a favourable long-term outcome, yet, in comparison with high achievers specific cognitive impairments become apparent.

Keywords: cerebellar pilocytic astrocytoma, neuropsychology, long-term effect, health related quality of life, cognitive performance.

FL-030

Oncology

Risk of malignant transformation of grade II gliomas in young adults and children- Leeds paediatric neurosurgery experience (2004-2014)

Simone Peraio¹; Senthil Selvanathan¹; Paul D. Chumas¹.

¹Department of Neurosurgery, Leeds General Infirmary, Leeds.

Objective: Grade II gliomas in adults are pre-malignant primary brain tumours and they are characterised by slow and continuous growth with 90% risk of malignant transformation. Despite the abundance of studies relating to low-grade glioma transformation in adults, evidence is lacking when it comes to describing the risk of malignant transformation (MT) in children and young adults.

Methods: The authors retrospectively reviewed their results in treating children and young adults (Aged 25 years and below) with grade II gliomas admitted to the Department of Neurosurgery, Leeds General Infirmary between 2004 and 2014. Those patients who had malignant transformation were identified.

Results: Of the 25 patients treated for grade II gliomas, two patients (Aged 12 and 16 years old) experienced MT despite initial surgery. The median latency of MT was 4.8 years. Both patients were females with the tumours being localised supratentorial. Initial diagnoses were grade II diffuse astrocytoma and grade II fibrillary astrocytoma. Histology for both patients following malignant transformation was anaplastic astrocytoma and glioblastoma respectively. Median survival post-transformation was 1.25 years despite second surgery. Ki67 showed a low proliferation index before the malignant transformation in both the patients and a high proliferation index after the malignant transformation. No evidence of 1p19q co-deletion was detected in both patients.

Conclusion: Our risk of malignant transformation in our series is 8%. Reports of MT in grade II gliomas are exceptionally rare with only one other publication reporting their results. Although this study revealed some aspects of MT in grade II glioma in children and young adults, our results were limited by the small number of affected patients. Therefore, a larger collaborative study of children with grade II gliomas with MT are warranted to identify molecular markers that could identify and ultimately risk stratify these patients.

Keywords: grade II gliomas, malignant transformation, young adults, children

FL-031

Oncology

Prognostic value of the extent of resection and early seeding metastases in pediatric glioblastoma

Sahin Hanaloglu¹; Burcak Bilginer¹; Firat Narin¹; Kader Karli Oguz²; Figen Soylemezoglu³; Nejat Akalan¹.

¹Hacettepe University Faculty of Medicine, Department of Neurosurgery, Ankara; ²Hacettepe University Faculty of Medicine, Department of Radiology, Ankara; ³Hacettepe University Faculty of Medicine, Department of Pathology, Ankara.

Objective: Less is known about pediatric glioblastoma (GBM) due to its rarity. The aim of this study was to explore clinical, radiological and pathological features, and treatment options of pediatric GBM and to investigate prognostic factors affecting the outcomes.

Methods: We retrospectively reviewed patient charts, imaging and follow-up data of 42 consecutive pediatric patients (age ≤ 18 years) with histologically proven GBM or its variants, treated at a single tertiary referral hospital.

Results: The mean age was 10.2 \pm 4.9 years. The most common tumor locations were lobar/hemispheric (68.3%) and thalamic (26.8%). 7.1% had seeding metastases at presentation, and additional 19% of patients developed CSF dissemination during the follow-up. Gross total resection (GTR) was achieved in 30.9%. Perioperative mortality and morbidity rates were 4.7% and 19%, respectively. Majority of patients received radiotherapy (78.6%) and chemotherapy (83.3%). Pathology confirmed glioblastoma or its variants in all patients (32 classical, 3 gliosarcoma, 7 giant cell GBM). Mean follow-up period was 18.1 months. The median PFS and OS were 7.0 (95% CI: 5.9–8.0) and 11.0 (95% CI: 8.9–13.1) months, respectively. Median survival after the seeding was 5 months. Age, sex, presence of preoperative seizures, deficits or hydrocephalus, the duration of symptoms, pathological variants have not been found as significant prognostic factors affecting the outcome. In the univariate analysis, the presence of postoperative deficits, tumor location, extent of resection and presence of early seeding metastases showed significance. However, in the multivariate analysis, only complete resection and the presence of early seeding metastases emerged as independent prognostic factors for survival.

Conclusion: Gross total resection should be safely attempted in pediatric GBM. A thorough and frequent radiological evaluation of the entire neuraxis for seeding metastases is also recommended as they adversely affect the outcomes.

Keywords: glioblastoma, malignant glioma, pediatric, children, prognostic factor

Tuesday, 6 October 2015
10:30 – 11:45

Hydrocephalus & Malformations Flash Parallel Presentations III: Shunts and complications

FL-032

Other

Role of primary Cilia in the developing chick ventricular system

Takayuki Inagaki¹.

¹Ibaraki Children's Hospital, Mito.

Objective: Impairment of Cilia function underlies a number of human diseases including hydrocephalus. However, the role of cilia in the developing embryo is not well understood. Chloral hydrate is known to have an adverse effect on the Cilia formation. In this paper, the possible role of cilia in developing chick embryos will be described mainly by focusing on the formation of the nervous system.

Methods: White leghorn chicken eggs were incubated until embryos reached hamburger and hamilton stages 4 to 10. Chick embryos were prepared for both in vitro culture and in ovo culture. Embryos were treated with a chloral hydrate solution for 20 minutes, and re-incubated for 24 hours. Embryos were collected from the incubator and examined morphologically after approximately 24 hours.

Results: In vitro: the embryos treated with chloral hydrate developed neural tube defects, reversed-sided heart looping, and an abnormally shaped primitive cerebral ventricle in a dose dependent manner. The embryos with neural tube defects developed a small primitive ventricular system. In ovo: with the same amount of chloral hydrate solution to the in vitro culture, embryos did not have anomaly described above. On the other hand, the embryos treated with higher dose solution showed more severe anomalies such as divided cardiac system and/or maldevelopment of rostral neural tube compared with the embryos cultured in vitro.

Conclusion: The importance of motile cilia in normal function of the cerebral ventricular system, including its role in circulation of cerebrospinal fluid, is widely recognized, but the role of primary cilia in early embryonic development is not well understood. In this study we found that embryos treated with chloral hydrate have many types of anomalies depending on the cultured system. Our results suggest that cilia also have an important role in early development of the ventricular system in addition to a role in axial development.

Keywords: primary cilia, neural tube, development, hydrocephalus, spina bifida

FL-033

Hydrocephalus

Trends in hospitalization of preterm infants with intraventricular hemorrhage and hydrocephalus in the United States, 2000–2010

Eisha A. Christian¹; Diana Jin²; Frank Attenello¹; Edward F. Melamed³; Timothy Wen²; Steven Cen⁴; William Mack¹; Mark D. Krieger³; J. Gordon McComb³.

¹Department of Neurosurgery, Keck School of Medicine, University of Southern California, Los Angeles; ²Keck School of Medicine, University of Southern California, Los Angeles; ³Division of Neurosurgery, Children's Hospital Los Angeles, Los Angeles; ⁴Department of Preventative Medicine, Keck School of Medicine, University of Southern California, Los Angeles.

Objective: Even with improved prenatal and neonatal care, intraventricular hemorrhage (IVH) occurs in approximately 25–30% of preterm infants, and a subset of these patients develop hydrocephalus. We aim to describe current trends in hospitalization of preterm infants with post-hemorrhagic hydrocephalus (PHH) using the Nationwide Inpatient Sample (NIS) and Kids Inpatient Database (KID).

Methods: The KID and NIS databases were combined to generate data for the years 2000–2010. All neonatal discharges with ICD9-CM codes for preterm birth with IVH alone or with IVH and hydrocephalus were included.

Results: There were 147,823 preterm neonates with IVH, and 9% of this group developed hydrocephalus during the same admission. Twenty-five percent and 28% of patients with Grades 3 and 4 IVH respectively developed hydrocephalus in comparison to 1% and 4% of patients with Grades 1 and 2 IVH. Thirty-eight percent of patients with PHH had permanent ventricular shunts inserted. Mortality rates were 4%, 10%, 18%, and 40% respectively for Grades 1–4 during initial hospitalization. Length of stay has been trending upward for both groups of IVH (49d in 2000, 56d in 2010) and PHH (59d in 2000, 70d in 2010). Average hospital cost per patient (inflation-adjusted) has also increased from \$201,578 to \$353,554 (IVH) and \$260,077 to \$495,697 (PHH) over 11 years.

Conclusion: For reasons undetermined, the number of admissions of neonates with IVH has increased despite a decrease in the number of preterm births. Rates of hydrocephalus and mortality correlated closely with IVH grade. Incidence of hydrocephalus in preterm infants with IVH remained stable between 8–10%. Over an 11-year period, there was a progressive increase in hospital cost and length of stay for preterm neonates with IVH and PHH.

Keywords: Nationwide Inpatient Sample, intraventricular hemorrhage, post-hemorrhagic hydrocephalus, preterm birth, hospital cost

FL-034

Hydrocephalus

Does a temporizing measure of cerebrospinal fluid drainage as the initial procedure alter the surgical outcome in premature infants with post-hemorrhagic hydrocephalus?

Eisha A. Christian¹; Edward F. Melamed²; Edwin Peck¹; Mark D. Krieger²; J. Gordon Mccomb².

¹Department of Neurosurgery, Keck School of Medicine, University of Southern California, Los Angeles; ²Division of Neurosurgery, Children's Hospital Los Angeles, Los Angeles.

Objective: It has been speculated whether the insertion of a temporary device to control hydrocephalus secondary to intraventricular hemorrhage (IVH) in the preterm neonate, with removal of the debris caused by such a hemorrhage, can reduce subsequent complications following insertion of a permanent cerebrospinal fluid (CSF) diverting shunt. This retrospective review is directed at examining this speculation.

Methods: A retrospective review of the medical records of all premature infants surgically treated for post-hemorrhagic hydrocephalus (PHH) between 1997 and 2012 at our institution was undertaken.

Results: Over 14 years, 91 preterm infants with PHH were identified. The initial procedure for 50 neonates was the insertion of a ventricular reservoir (VR) that was serially tapped for varying time periods. For the remaining 41 premature infants, a ventriculoperitoneal/atrial shunt (VS) was the first procedure. Patients with a VR as their initial procedure underwent CSF diversion significantly earlier in life than those who had VS as the initial procedure (29 vs. 56 days, $p < 0.01$). Of the infants with a VR as their initial procedure, 5/50 (10%) did not undergo a subsequent VS. The number of shunt revisions and the rates of loculated hydrocephalus and shunt infection did not statistically differ between the two groups.

Conclusion: Patients with initial VR insertion received a CSF diversion procedure at a significantly younger age than those who received a permanent shunt as their initial procedure. Otherwise, the outcomes with regards to shunt revisions, loculated hydrocephalus, and shunt infection were not different for the two groups.

Keywords: intraventricular hemorrhage, post-hemorrhagic hydrocephalus, preterm infant, ventricular reservoir, ventriculoperitoneal shunt

FL-035

Evidence-based practice

Surgical site infections in a paediatric neurosurgical unit - how change of practice has reduced infection rate

Dawn Williams¹; Linda Marshall¹; Lisa Wall¹; Stephane Paulus¹; Richard Cooke¹; Benedetta Pettorini¹.

¹Alder Hey, Liverpool.

Objective: Surgical site infections (SSI) represents a common healthcare associated infection and cause of morbidity. Despite multiple protocols in pre-operative preparation, theatre practices and microbiology techniques, only few available data has identified effective strategies to reduce SSI risk. Our department instituted a monitoring programme for SSI in 2011 and we introduced a care bundle in 2013. We present our SSI data after the introduction of the bundle.

Methods: The implemented bundle consists of pre-, peri- and post-operative interventions including MRSA and MSSA swabs, chlorhexidine wash, appropriate antibiotic prophylaxis, avoid unnecessary shaving,

limit theatre door openings, all staff to wear masks, double gloving at high risk procedures, keep wound covered for 48 hours. We have prospectively analysed 1 year of neurosurgical procedures in a single neurosurgical centre. We have prospectively collected data including type of procedure. Total number of infections by month, infection by category (implants - ITB pumps etc, CSF - including shunts/stents/reservoirs, Spinal, Cranial, Craniofacial), infection by SSI subgroup (superficial, deep, organ/space - no implant, organ/space - implant) and organisms identified. We used the CDC/NHSN surveillance definition of healthcare associated infection and criteria for specific type of infection in the acute care setting. All data is recorded on a database for analysis. All non implant surgery was followed up for 30 days, implants had a one year follow up according to NICE guidance.

Results: We have analysed data on 482 patients for 569 procedures. The data are summarised in the table below.

Conclusion: Our data demonstrates a significant reduction of SSI since the introduction of the bundle. The reduction of SSI from a peak of 8.1% to 4.0% is very encouraging and highlights the importance of a SSI monitoring system in place. There are still aspects which needs to be improved, such as SSI prevention in infants.

Keywords: surgical site infection, neurosurgery, care bundle, infection rate

FL-036

Evidence-based practice

The effect of magnetic resonance imaging on the neural tube of early chicken embryos

Emrah Kantarcioglu¹; Gokmen Kahilogullari¹; Murat Zaimoglu¹; Nazan Yildiz³; Ozlem Esin Atmis²; Elif Peker²; Zeynep Gunhan³; Deniz Billur⁴; Esra Erdemli⁴; Ilhan Erden³; Agahan Unlu¹.

¹Ankara University Faculty of Medicine, Department of Neurosurgery, Ankara; ²Ankara University Faculty of Medicine, Department of Otolaryngology, Ankara; ³Ankara University Faculty of Medicine, Department of Radiology, Ankara; ⁴Ankara University Faculty of Medicine, Department of Histology-Embriology, Ankara.

Objective: Neural Tube Defects (NTD) occur in the first weeks of embryonic life and develop due to incomplete closure of neural tube. Alongside their social consequences NTD are important because of their prevent abilities. In this study we aimed to research effect of MRI on midline closure of chick embryos and whether cause NTD or not.

Methods: We determined statistically significant neural tube closure defects and histologically growth retardation on chick embryos those exposed different Tesla powers of magnetic fields ($p < 0.05$).

Results: Striking feature of the study was the increment in the growth retardation was correlated with increasing Tesla.

Conclusion: With all these findings as a current diagnostic tool, MRI, can cause midline closure defects and growth retardation. From this aspect it is a possibility those effects can also occur in human embryos. This is the first laboratory report about the relation of NTD and MRI.

Keywords: Magnetic resonance imaging, chicken embryos, neural tube defects

FL-037

Hydrocephalus

Effect of EVD tunneling length on post operative CSF infection: a randomized controlled trial

Sara Hanaei¹; Zohreh Habibi¹; Farideh Nejat¹.

¹TUMS, Tehran.

Objective: External ventricular drainage (EVD) is a temporary device for decreasing intracranial pressure (ICP) caused by abnormal accumulation of CSF due to brain tumor, arachnoid cyst, head trauma, etc. As a foreign body, EVD increases probability of CSF infection and meningitis and it is hypothesized that increasing the distance between infection source (skin) and CSF may decrease CSF infection.

Methods: 48 patients were randomly allocated to one of 3 groups (A: tunneling=5 cm, B: 10 cm or C: >15 cm) according to a 6-block randomization. Postoperative prophylactic antibiotic (vancomycin) was used for all patients and the CSF was analysed for infection incidence every other day. Infection was defined by positive CSF culture and EVD would be discharged in the time of infection, no function or no further need.

Results: Patients were 1-168 months old (mean=45 months) with sex distribution of 60.4% male and 39.6% female. The background diseases were head trauma (6.3%), arachnoid cyst (14.5%), brain tumor (70.8%), post MMC hydrocephalus (6.3%) and other causes (8.3%). The mean duration of EVD usage was 6.9 days with range of 1-22 days. The infection rate were 2.1% in groups A and C and was 4.2% in group B which failed to reach significance. No significant association was detected between infection rate and EVD usage duration in this stage of study.

Conclusion: Although the results did not reach significance in this stage, the study is ongoing and completion of sample size is required for final decision. It is believed that increasing tunneling length to 10 cm can reduce infection rate but further increases may not affect the infection incidence. Besides, EVD usage duration could have influence on infection incidence as well.

Keywords: external ventricular drainage, tunneling, length, infection rate

FL-038

Hydrocephalus

Ventriculosubgaleal shunts: can a simple variation in technique prevent leaks and infection?

Bhaskar Thakur¹; Wisam Al-Faiadh¹; Airi Kugisaki¹; Angela Calado Bravo¹; Benedetta Pettorini²; Sanjeev Bassi¹; Chris Chris¹; Bassel Zebian¹.

¹Kings College Hospital, London; ²Alder Hey Children's Hospital, Liverpool.

Objective: Description of a new technique of ventriculosubgaleal shunt (VSGS) as a temporising measure for hydrocephalus in infants.

Methods: A curved skin incision was made at the junction of the fontanelle and coronal suture on the right side. In some patients a burr hole was performed anterior to the coronal suture. In others, access to the dura was through the coronal suture. A proximal catheter attached to a rickham reservoir was placed into the lateral ventricle. A 10cm distal catheter with additional holes was tunneled to the opposite side of the scalp after fashioning a subgaleal pocket using a 16F foley catheter.

Results: The procedure was performed on 4 patients. 1 patient had a CSF leak post op, due to the subgaleal pocket being too close to the skin incision. There was 0% blockage rate. None of the shunts required revision. 1 case where infection and haemorrhage were both present, weekly taps of the reservoir were carried out as head circumferences were increasing despite the VSGS. There was 0% infection/ secondary infection rate.

Conclusion: VSGS seems an effective temporising procedure in the management of hydrocephalus in infants. The new technique seems promising in reducing CSF leak and infection. More large scale studies are needed to validate the technique.

Keywords: VSGS, Ventriculo subgaleal shunt, Hydrocephalus

FL-039

Hydrocephalus

Does shunt selection affect the rate of early shunt complications in neonatal myelomeningocele associated hydrocephalus?

Volkan Etus¹; Gokmen Kahilogullari²; Hakan Karabagli³; Batihan Uye¹; Tugba Morali Guler⁴; Agahan Unlu².

¹Kocaeli University Faculty of Medicine, Department of Neurosurgery, Kocaeli; ²Ankara University Faculty of Medicine, Department of Neurosurgery, Ankara; ³Selcuklu University, Faculty of Medicine, Department of Neurosurgery, Konya; ⁴Karabuk University, Education and Research Hospital, Department of Neurosurgery, Karabuk.

Objective: The overall risk of shunt infection and malfunction is reported to be higher in patients with myelomeningocele. In this study, the effect of shunt selection on the rate of shunt revision due to early-shunt-complications in neonatal myelomeningocele-associated-hydrocephalus was evaluated.

Methods: The data of total 157 neonatal myelomeningocele cases belonging to three centers which, had undergone shunt surgery at the time of myelomeningocele repair between 2000 and 2014 were retrospectively analyzed. Patient records on clinical features, preferred shunt types and early-shunt-complications (within first 3 months) were reviewed. The patients who had been followed up for at least 1-year postoperatively were included in the study. Cases were grouped according to several features of the preferred shunt systems such as valve type, valve size/contour and catheter type.

Results: The analysis of the data revealed that 71 of the 157 patients (45.2%) have undergone early shunt-revision surgery due to various complications. Mechanical complications were the most frequent cause of shunt failure followed by infection. Regarding the several shunt complications (mechanical-dysfunction, exposed-shunt, infection and dynamic-malfunction) statistical analysis showed no significant difference among valve types (fixed-differential-pressure / anti-siphon or gravity-activated / flow-control / adjustable). Also no significant difference was shown between the catheter types (antimicrobial-impregnated or -coated vs standard). Only preferred valve contour/size (contoured regular /ultra-small / bur-hole / cylindrical / neonatal) seemed to significantly affect the rate of early-complications. The cases with neonatal-design valves or ultra-small valves seemed to have significantly less problems such as poor wound-healing, wound-dehiscence, CSF-leak or an exposed-shunt. Also infection rate due to such problems was found to be lower.

Conclusion: Neonatal myelomeningocele cases with prominent hydrocephalus frequently have friable-skin as a result of reduced subcutaneous tissue due to macrocrania. Our findings suggest that using small-sized (neonatal-design or ultra-small) valves may significantly reduce early-shunt-complication rate in those cases.

Keywords: hydrocephalus, shunt infection, shunt malfunction, myelomeningocele, neonate

FL-040

Infection

Institutional experience following hydrocephalus clinical research network standardized shunt surgery protocol to reduce cerebrospinal fluid infection

Eylem Ocal¹; Gregory W. Albert¹; Rongsheng Cai¹; Lea Woodrow¹; Mark O'Brien¹.

¹Department of Neurosurgery - University of Arkansas for Medical Sciences/ Arkansas Children's Hospital, Little Rock.

Objective: Prevention of shunt infection is a major concern for pediatric neurosurgeons. Approximately 8- 10 % of all shunts get infected, more commonly within the first few months of the surgery. The rate of infection is related to multiple variables. Additionally, the reported rate of infection varied widely across surgical centers in United States in studies. Recently, a

study was conducted by Hydrocephalus Clinical Research Network (HCRN) at 4 centers in US. Using a standardized protocol for shunt surgery, the infection rates decreased in participating centers. The same protocol was followed at Arkansas Children's Hospital (ACH) with the purpose of to validate the results and prove applicability of this protocol for individual centers to reduce shunt infection. We present our experience in this study.

Methods: The HCRN protocol was used by all the neurosurgeons in Arkansas Children's Hospital starting November 2011 for all shunt procedures where applicable. The results were collected prospectively as a part of a quality improvement project by the Infectious Disease Committee and Division of Pediatric Neurosurgery at Arkansas Children's Hospital. The institutional shunt infection rate prior to application of the standardized protocol and after application were compared.

Results: Our institutional results were in parallel with the HCRN study results. The infection rates were averaged over a 3 year period prior and after the application of the protocol and was found to be at 8.03%. The rate of shunt infection at ACH was decreased by almost half to 4.83% after initiation of the protocol.

Conclusion: Our institutional experience confirmed that using a standardized protocol is easily applicable and decreases shunt infection rates. Importantly, this in return increases patient's quality of life and will decrease complication related costs to the health care system. This protocol can be utilized across centers and amongst neurosurgeons conveniently without the need for costly resources.

Keywords: shunt, infection, standardized protocol, surgery

FL-041

Hydrocephalus

The management of paediatric ventriculoperitoneal shunt infections: results from a practice survey of British paediatric neurosurgeons

Matt Bailey¹; Chris Parks¹; Conor Mallucci¹.

¹Department Of Paediatric Neurosurgery, Alder Hey Children's Hospital, Liverpool.

Objective: When faced with an infected ventriculo-peritoneal (VP) shunt the majority of neurosurgeons would advocate removal of infected hardware and treatment with antibiotics. However there is little consensus in the literature on the choice of intravenous (IV) and/or intrathecal (IT) antibiotics, and for the duration of antibiotic therapy. We surveyed a cohort of British paediatric neurosurgeons to assess current practice.

Methods: British paediatric neurosurgeons were approached at a national paediatric neurosurgical meeting. Questionnaires were distributed. Two clinical scenarios were described: A) classic symptoms of shunt infection with organisms on VP shunt tap; B) pyrexia, headache and raised CSF cell counts but negative gram stain. Responses were collected on management of shunt hardware, and choice and duration of antibiotic treatment. Analysis of results was performed using Microsoft Excel.

Results: 21 completed questionnaires were received. 100% of responders would remove shunt hardware and insert an external ventricular drain (EVD) in scenario A and 85% in scenario B. The majority would use an antibiotic impregnated EVD (65%). IV antibiotics alone would be used by 72% in both scenarios. IT antibiotics alone would be used by 11% in scenario A and 6% in scenario B. Combined IV and IT antibiotics would be used by 17% in scenario A and 22% in scenario B. 11 different antibiotic regimens were suggested in scenario A and 13 different regimens in scenario B. Responders would treat with antibiotics for a periods ranging from 5 days to 28 days in both scenarios.

Conclusion: The majority of British paediatric neurosurgeons surveyed would remove an infected or potentially infected VP shunt and insert an EVD. IV antibiotics alone are used by the majority, although IT therapy and IV/IT combinations are used by some. There was little consensus on the specific antibiotic regimens for this common infection.

Keywords: shunt, infection, management, antibiotic

FL-042

Hydrocephalus

Pediatric ventriculoperitoneal shunt disconnection and fracture

Ash Singhal¹; D. Douglas Cochrane¹; Paul Steinbok¹; A. Ross Hengel¹.

¹University Of British Columbia and BC Children's Hospital, Vancouver.

Objective: Shunt disconnection and fracture (SDF) are well-recognized causes of ventriculoperitoneal shunt (VPS) dysfunction. No study to date has characterized the frequency of SDF individual component failure nor the clinical scenarios associated with these dysfunctions.

Methods: We conducted a retrospective review of VPS procedures (from 1982-2015) at BC Children's Hospital, and selected those patients with SDF. Operative and radiological records were reviewed to determine clinical presentation, location of SDF, and survival time of the individual component failure.

Results: In total, 95 patients were identified to have SDF (52 males). Twenty-four patients had multiple events, resulting in a total of 126 SDF reviewed. Average patient age at the time of component failure was 11.0 years (0.4 - 19.3 years). Peritoneal catheters broke in 91 cases (72%), with 43 cases disconnecting or fracturing at the distal end of the VPS valve, 45 in the neck or upper chest region, and 4 at the abdominal entry site. Ventricular catheters disconnected/ fractured in 25 cases (20%), valves broke in 9 cases (7%), and an antisiphon device detached from the valve in 1 case (1%). Average age of each broken shunt component was 8.2 years and there were no significant differences between the ages associated with the breakage sites/hardware. SDF was asymptomatic in 27 instances, while the other 99 were symptomatic. Of these (20/99) had pain or fluid collections along the shunt, and the remainder presented only with symptoms of general shunt dysfunction (headaches, nausea, lethargy, etc.).

Conclusion: SDF predominantly occurs in the peritoneal catheter. Shunt components/connections are prone to failure in the intermediate term, although there are instances of early failure. Symptoms are most commonly those of general shunt dysfunction, suggesting that routine radiographic assessment of the entirety of the shunt tubing is indicated with the symptomatic VPS patient.

Keywords: Hydrocephalus, VP Shunt, Fracture, Disconnection

FL-043

Hydrocephalus

Peritoneal failure is not uncommon in VP shunts in young infants.

Aly Khaled Bassim¹.

¹Cairo University, Cairo.

Objective: Ventriculoperitoneal shunt failure due to failure of the peritoneum to absorb CSF is underreported. It has often been ascribed to low grade infection. However, idiopathic peritoneal failure has not been considered. This is a prospective study, examining the incidence of such problem in infants below one year of age with recent shunt insertion.

Methods: 240 children with hydrocephalus of variable etiologies were followed after VP shunt insertion. Shunt failure was investigated by CT brain, shunt tapping and CSF analysis, abdominal ultrasound, as well as ESR and CRP to rule out infection. The diagnosis of peritoneal failure was concluded if the following criteria were fulfilled: Signs of shunt obstruction (enlarging head, CSF leak, wound swelling, progressive ventriculomegaly on CT or MRI of the brain) no signs of infection in CSF analysis as well as normal abdominal ultrasonography (i.e. no ascites or cysts formation). During shunt revision, a specimen of the peritoneum was sent for pathological verification.

Results: 240 children were operated upon by VP shunt insertion over one year period. 25 children (11%) had fulfilled the criteria of shunt obstruction due to peritoneal failure. No evidence of infection could be elicited in

them. Age was the main relevant factor as most kids were below three months of age. Pathological examination failed to show any signs of bacterial infection, rather non specific lymphocytic infiltrate.

Conclusion: Shunt obstruction due to peritoneal failure is more common than previously thought. This is mainly due to peritoneal immaturity rather than low grade infection. Shunt insertion in the first three months of life is particularly vulnerable to this complication.

Keywords: Hydrocephalus, Ventriculoperitoneal shunt, Shunt failure, Shunt infection

FL-044

Hydrocephalus

Trapped fourth ventricle; diagnosis and management

Chidambaram Balasubramaniam¹; Santosh Mohan Rao¹; K Subramaniam¹.

¹KKCTH, Chennai.

Objective: To analyse the problem of trapped fourth ventricle with respect to diagnosis and management. Trapped fourth ventricle was defined as the condition where in a child who has had a VP shunt, the fourth ventricle is dilated and is under raised pressure.

Methods: This is a retrospective chart review of 6 children who underwent surgery for trapped fourth ventricle. The criteria for diagnosis were analysed since not all dilated fourth ventricles are deemed trapped. Differentiation was made from Dandy Walker Malformation. The management issues were studied.

Results: Six children were identified as having trapped fourth ventricle. This was out of 1062 shunt procedures performed. The youngest was 3 months and the oldest 5 years, all had had a CSF diversion procedure and some had had multiple revisions. The chief presenting symptom was that of a posterior fossa mass. The commonest cause of trapped fourth ventricle was post infections/ inflammatory hydrocephalus. Imaging showed dilated fourth ventricle. Differentiation was made from Dandy Walker Malformation by the dysplasia/ absence of the vermis. A fourth ventricle peritoneal (pleural in one case) shunt was done by connecting a catheter placed in the trapped ventricle and connecting it distal to the valve of the VP shunt using a Y connector. All children improved. Even though the ventricle continued to be enlarged in some the pressure effects were not present.

Conclusion: Trapped fourth ventricle is an uncommon complication of hydrocephalus due to infection/ inflammation. It can also occur due to kinking of the aqueduct. If an endoscopic approach is possible the communication with the lateral and third ventricles can be reestablished. Deroofing fourth ventricle is an option but it may fail. A fourth ventricle shunt may be the treatment of choice in some.

Keywords: Trapped fourth ventricle, hydrocephalus, shunt

FL-045

Hydrocephalus

Changes in diffuse tensor imaging after VP shunt procedure on patients with severe congenital hydrocephalus

Wihasto Suryaningtyas¹; Dewa Putu Wisnu Wardhana¹; Sri Andreani Utomo²; Muhammad Arifin¹.

¹Department of Neurosurgery, Faculty of Medicine Universitas Airlangga, Soetomo General Hospital, Surabaya; ²Department of Radiology, Faculty of Medicine Universitas Airlangga, Soetomo General Hospital, Surabaya.

Objective: To determine the changes in Diffuse Tensor Imaging (DTI) on patients with severe congenital hydrocephalus that underwent VP shunt procedure.

Methods: A prospective observational study on congenital hydrocephalic patients was conducted on Soetomo Hospital Surabaya from February 1, 2013-January 31, 2014. Congenital hydrocephalus patients aged two years old or less and no other congenital craniofacial and brain abnormalities were eligible. Three months follow up were done. Variable observed included cerebral mantle thickness, ventricular index, and DTI parameters.

Results: Before surgery, 21 subjects with mean age 7.81 ± 7.95 months were included in this study for pre-operative analysis. Low Fractional Anisotropy (FA) values were found mostly on sCC (splenium Corpus Callosum) (61.9%) than on gCC (genu Corpus Callosum) (57.1%). There are 6 subjects with mean follow up 5.33 ± 3.83 months after surgery. FA value on bCC, and sCC value were decreased but it was increasing on gCC. Mean MD value was decreased on all corpus callosum parts. Mean FA value constantly decrease after surgery on ALIC and PLIC. Based on tractography results, the corpus callosum showed improvement after surgery. Ventricular index (Frontal horn ratio and Evans ratio) were all improved significantly ($p < 0.000$ and 0.002 , consecutively).

Conclusion: There are tendency for positive changes on DTI parameters after surgery. Association between DTI and hydrocephalus still need to be confirmed with a larger sample study.

Keywords: Severe Congenital Hydrocephalus, Diffusion Tensor Imaging, VP shunt

FL-046

Hydrocephalus

Adult long-term health-related quality of life of congenital hydrocephalus patients

Preuss Matthias¹.

¹University Leipzig, Leipzig.

Objective: Congenital hydrocephalus has a major impact on the life of patients and their relatives, long-term neurological development and social integration. Aim of the study was to assess the self-reported health-related quality of life (HRQoL) of patients after reaching adulthood.

Methods: 31 patients that required CSF-shunt therapy for congenital hydrocephalus within the first year of life between 1963 and 1987 agreed to undergo a structured SF-36 self-assessment. An age-matched German standard cohort was used as control. Additional parameters of surgical, social and global neurological outcome were analysed. Mean age of the patients was 35 years (range 26-51 years, 13 female and 18 male). Hydrocephalus etiologies were posthemorrhagic ($n=9$), postinfectious ($n=5$), aqueductal stenosis hydrocephalus ($n=10$), myelomeningocele ($n=2$) and unknown cause ($n=5$).

Results: Mean modified Rankin Scale score was 1.6 (range 0-4). Hydrocephalus patients achieved lower scores for SF-36 items physical functioning (70.5 vs. 93.5, $p < 0.05$), physical role functioning (74.2 vs. 88.3, $p < 0.05$) and general health perceptions (64.5 vs. 72.3, $p < 0.05$). Emotional, social role functioning and mental health items did not differ between both groups. Assessment of vitality and pain resulted in a trend to worse values. Whereas physical component score showed lower results (46.1 vs. 54.3, $p < 0.05$), mental component score was not significantly different (50.2 vs. 48.7, $p=0.3$). There was neither a statistical significant difference between subgroups of different etiologies nor an association with the number of subsequent hydrocephalus-related surgeries.

Conclusion: Adult HRQoL in congenital hydrocephalus appears to be similar compared to healthy controls for mental health and social functioning aspects. Physical impairment is a predominant factor of compromised quality of life.

Keywords: congenital hydrocephalus, long-term outcome, quality of life

Tuesday, 6 October 2015
10:30 – 11:45

Hydrocephalus & Malformations Flash Parallel Presentations IV: Endoscopy and malformations

FL-047

Hydrocephalus

Individualized entry points for endoscopic third ventriculostomy in pediatric cases

Zsolt Zador¹; David Coope²; Ian Kamaly³.

¹Institute of Cardiovascular Sciences, Centre for Vascular and Stroke Research University of Manchester, Manchester; ²Wolfson Molecular Imaging Centre, The University of Manchester, Manchester; ³Department of Pediatric Neurosurgery, Royal Manchester Children's Hospital, Manchester.

Objective: Endoscopic third ventriculostomy (ETV) is a minimally invasive method of CSF diversion for treating obstructive hydrocephalus. The procedure involves the transcortical passage of an endoscope to access the third ventricle floor for fenestration. Trauma to periventricular structures during this maneuver can result in morbidity or mortality, we therefore tested the tissue displacement associated with three frequently quoted (standard) ETV trajectories. We also developed a simple algorithm to simulate individualized ETV entry points for trajectories that do not cause any tissue displacement (“non-traumatic”) by incorporating ventricle dimension.

Methods: We created three-dimensional radioanatomical models from 30 pediatric patients treated for ventriculomegaly. Results were stratified based on ventricle size using the frontal horn ratio (FHR). The surface locations of the cranial ETV entry points were determined as Cartesian coordinates centered at Bregma. The distance from the midline in the coronal plane represented as “x” (A) and the distance from the coronal suture in the sagittal plane through Bregma represented as “y” (B). Correlation between the ventricle dimensions and the x, y coordinates were tested using linear regression models (C and D respectively).

Results: We found tissue displacement in 97–100% of cases for the three standard trajectories tested. Comparative analysis of these trajectories suggested a more posterior and lateral entry point would be less traumatic for larger ventricles. The coordinates for non-traumatic ETV entry points were then fitted along the following equations: $x=85.8a-13.3$ ($R^2=0.84$) and $y=69.6a-16.7$ ($R^2=0.83$). Where “a” represents the numeric value of the FHR.

Conclusion: The standard ETV trajectories are almost invariably associated with displacement to periventricular structures; therefore ETV trajectories should be tailored on a case-by-case basis. We provide the first model to simulate individualized entry points for ETV trajectories that avoid tissue displacement with incorporation of the ventricle size.

Keywords: ETV, hydrocephalus

FL-048

Hydrocephalus

Value of early postoperative MR in predicting failure of ETV

Suhas Udayakumaran¹; Varun Garg¹.

¹Division of Paediatric Neurosurgery, Amrita Institute of Medical Sciences and Research Centre, Kochi.

Objective: To look for the value of early post operative MR in predicting early Endoscopic third ventriculostomy (ETV) failure.

Methods: We analysed success of ETV in 30 consecutive ETV procedures in 30 children (below the age of 15years) between

March 2011 to march 2015. The demographics, aetiology, operative details, early clinicoradiological outcome was collected retrospectively. The early postoperative MR was analysed for flow void at the ETV stoma and correlated to the clinical and radiological success of ETV at the end of 6 months.

Results: Out of the 30 patients who underwent ETV based on standard indications and family preference with early postoperative MR, 7/11 patients did not show flow void in the postoperative MR and all of them had early failure of ETV. The sensitivity of using a postoperative MR to identify early ETV failure is 100% with a specificity of 82.6%.

Conclusion: We conclude that early postoperative MR can have 100% negative predictive value for the success of ETV and hence can be an effective guide during the postoperative decision making and to avoid patient morbidity.

Keywords: Endoscopic third ventriculostomy, MR Flow void, Hydrocephalus

FL-049

Hydrocephalus

Endoscopic third ventriculostomy for infants

Tuncer Turhan¹; Kadri Emre Çalışkan¹; Elif Bolat¹; Anar Fevziyev¹.

¹Ege University, Izmir.

Objective: Endoscopic third ventriculostomy (ETV) has been advocated as a treatment for patients with non-communicating hydrocephalus. Recent studies indicate that both age and cause of hydrocephalus determine the ETV outcome. In this study we collected the ETV patients data retrospectively and focused the patients operated under one year of age.

Methods: Between January 2010 and December 2014, 28 patients (14 girls and 14 boys) who under one year of age were underwent ETV procedure for hydrocephalus. For all of the patients ETV is the first procedure for hydrocephalus.

Results: All of the patients had obstructive hydrocephalus and in most of the patients obstruction site was aqueductus sylvius. Etiology of hydrocephalus were idiopathic aqueductal stenosis for 15 patients, intraventricular hematoma for 9 patients, CNS infection for 2 patients and chiari malformation type 2 for 2 patients. Most of the obstruction site were aqueductus sylvius in 25 patients and foramen magnum in 3 patients. Mean following time is 28.5 months (6 to 48 months) in this population. Hydrocephalus in ten patients (35.7 %) has been successfully treated by ETV procedure. On the other hand, ETV procedure failed in 18 patients (64.3 %) and CSF diversion procedures were used for treatment. Sixteen patients were operated under three months of age, and success rate of ETV is calculated as 18.75 % in this group. Success rate is clearly rising for the children older than 3 months and calculated as 58.3 %. Also this difference between the two age groups (0–3 months vs 3–12 months) is statistically significant (chi-square p: 0.039).

Conclusion: ETV is an option for children who have obstructive hydrocephalus under one year old. But the success rate of ETV is decreasing rapidly in children less than three months of age.

Keywords: Hydrocephalus, Endoscopic third ventriculostomy, infant

FL-050

Hydrocephalus

The challenges and controversies of neuroendoscopy in the first year of life: a serie of 22 cases.

Artur Henrique G B Da Cunha¹; Suzana Maria Bezerra Serra¹; Amanda De Oliveira Lopez¹.

¹Hospital Da Restauracao, Recife.

Objective: The management of hydrocephalus in the first year of life is still a challenge for the pediatric neurosurgeon. Regardless of the approach of choice the great vulnerability of the newborn compromise the results. An additional way, the predominance of the “minor CSF pathway” in this age group is one of the important reasons for the failure of the endoscopic third ventricle-cisternostomy (ETV). The shunt-implantation is still the most common treatment, despite the complications with serious consequences for the developing nervous system. The international literature has shown endoscopic approached good results in only 40%, with the best success in aqueduct stenosis cases (70%).

Methods: We reviewed the literature about neuroendoscopic management of hydrocephalus in the first year of life and present our experience of 22 cases operated on between the ages of 32 days and 10 months. Six patients were premature infants who developed hydrocephalus associated with intraventricular hemorrhage (HIV) (Grade III/IV). Fourteen patients presented stenosis of aqueduct (AE) and two more cases developed a unilateral ventricular hydrocephalus associated with obstruction of the foramen of Monro (OFM). In the pos HIV patients were held brainwashing, coagulation of the choroid plexus and the ETV. Only ETV were used in the aqueductal stenosis and septostomy and ETV in the OFM cases.

Results: The successfully results without any further surgery were 2/5 (40%) in the pos HIV patients, in 7/14 (50%) in the AE cases and in 50% in the OFM patients. The CSF-leakage and infection were the complication in four cases.

Conclusion: How and when to treat is a matter of good sense. Indication, methods and timing of treatment must be individually tailored each of cases. Because of the high-risk of malfunction and infectious complication related to shunt, primary neuroendoscopic management may be considered a treatment of choice to avoid or delay the shunt-implantation.

Keywords: neuroendoscopy, hydrocephalus, newborn, infants, premature

FL-051

Hydrocephalus

Endoscopic third ventriculostomy can be recommended in children under 1 year of age

Masashi Kitagawa¹; Ryuji Ishizaki¹; Yuzuru Tashiro¹; Takafumi Wataya¹.
¹Shizuoka Children's Hospital, Shizuoka.

Objective: Hydrocephalus is common disease among children. Although ventriculoperitoneal (VP) shunt is mainstay in hydrocephalus treatment, it is associated with high failure rates and complication. Endoscopic third ventriculostomy (ETV) with potentially lower complication rate could reduce risk of management of hydrocephalus. The aim of this study was to evaluate the efficacy of ETV in children ≤ 1 year and to find out factors that may be responsible for good outcome of ETV.

Methods: We reviewed relevant demographic and clinical data of all consecutive children ≤ 1 year of age who had ETV for hydrocephalus from April 1st, 2014 to March 31st, 2015.

Results: 9 patients are recruited (6 males 3 females). All patients had triventriculomegaly. Etiology of hydrocephalus is myelo meningocele (MMC) 4, Chiari type 1 malformation 3, congenital hydrocephalus 1, post intraventricular hemorrhage hydrocephalus 1. 2 procedures were failed. ETV success rate was 77.8%. 2 of 3 procedures (66.7%) of patients who had both pre ETV infection and pre ETV VP shunt were failed. Average of ETV success score were 45 in failure group and 54.3 in success group.

Conclusion: Outcome of ETV ≤ 1 years of age are good in this series. Pre ETV infection and pre ETV VP shunt increase risk of failure. ETV can be recommended in children ≤ 1 year of age.

Keywords: ETV, hydrocephalus, under 1 year of age

FL-052

Hydrocephalus

Role of endoscopic third ventriculostomy in the management of myelomeningocele related hydrocephalus

Pierre-Aurélien Beuriat¹; Alexandru Szathmari¹; Blandine Grassiot¹; Franck Plaisant²; Christophe Rouselle¹; Carmine Mottolese¹.

¹Department of Pediatric Neurosurgery, Neurological and Neurosurgical Hospital Pierre Wertheimer, Lyon; ²Department of Neonatal Intensive Care Unit, Mother and Children Hospital, Lyon.

Objective: Treatment of hydrocephalus related to myelomeningocele (MM) is debated. Endoscopic Third Ventriculostomy (ETV) has been proposed with contrasting results. We report our experience in the management of hydrocephalus related to MM and the advantages of ETV.

Methods: From 1994 to 2012, we treated 97 MM patients. Seventy developed a hydrocephalus needing a surgical treatment. Three types of procedure were used: ETV, ETV and a concomitant Ventriculo-peritoneal shunt (VPS) or a VPS.

Results: Our patients fell into 3 groups: 32 patients had an ETV, 20 had an ETV and a concomitant VPS and 18 had a VPS. No perioperative complications were reported. Fiftytwo patients underwent an ETV. Coagulation of the choroid plexus (CPC) was associated with the ETV. The overall success rate of ETV was 75%.

Conclusion: VPS in MM patient lead to complications. Two common arguments are reported against ETV. The first is the variability of the anatomy of the ventricle leading to possible operative complications. The second is the immaturity of the Pacchionis granulations. For us, the modified anatomy does not prevent from performing ETV. Concerning the issue of Cerebro Spinal Fluid (CSF) absorption in failed ETV, the adjunction of a shunt help to control hydrocephalus until the maturation of the Pacchionis system. Preoperative imaging helps the surgical decision to predict in which patients the ETV can be realized without risks. ETV in MM patients is a secure procedure with low rates of failure and no mortality, and reduces the rate of shunt implantation.

Keywords: endoscopic third ventriculostomy, hydrocephalus, myelomeningocele, outcome, ventriculo-peritoneal shunt

FL-053

Hydrocephalus

Predictors of outcome in cerebral aqueductoplasty: an individual participant data meta-analysis

Aria Fallah¹; Anthony Wang²; Alexander Weil³; George Ibrahim⁴; Alireza Mansouri⁴; Sanjiv Bhatia¹.

¹Miami Children's Hospital, Miami; ²University Of Miami, Miami; ³CHU Sainte-Justine, Montreal; ⁴University Of Toronto, Toronto.

Objective: The evidence supporting the efficacy and safety of Cerebral Aqueductoplasty (CA) are limited to small surgical series. We performed an individual participant data meta-analysis to determine the efficacy and safety of CA. Our secondary objective was to determine the effect of patients age, etiology, surgical approach and use of stent on success.

Methods: Electronic databases (MEDLINE, EMBASE, and CINAHL) were search with no language or date restrictions to identify cohort studies of consecutive participants undergoing CA (without concomitant Endoscopic Third Ventriculostomy or Cerebrospinal fluid shunt) that reported outcome. The outcome was defined as the time elapsed from the index operation until a second procedure was performed for CSF diversion.

Results: Of 146 citations, 14 articles reporting on 137 participants were eligible. One hundred and three (75%) participants did not require a second CSF diversion procedure. The mean duration till repeat CSF diversion procedure was 121.6 (95% CI=102.2-141.0) months. In multivariate analysis, older age at surgery (HR=0.43, 95% CI=0.21-0.88, p=0.020), congenital etiology (HR=0.18, 95% CI=0.04-0.85, p=0.030)

and use of stent (HR=0.30, 95%CI=0.13-0.70, p=0.006) were independent predictors of good outcome. Morbidity was experienced in 22% of participants mainly comprising of ophthalmoparesis and hemorrhage.

Conclusion: Small retrospective cohort studies are inherently prone to bias, some of which are overcome using individual participant data. The best available evidence suggests that CA is an effective procedure with a moderate morbidity profile. Older age at surgery, congenital etiology and use of stent predicts a good outcome with respect to delaying the requirement for a second CSF diversion procedure.

Keywords: Aqueductoplasty, Success, Predictors, Meta-Analysis

FL-054

Hydrocephalus

Neuroendoscopy interest in the treatment of complex hydrocephalus

Derradji Hakim¹.

¹Central Hospital of Army, Algiers.

Objective: The complex Hydrocephalus is a multi-compartmental hydrocephalus due to the existence of intraventricular partitions appeared after chronic meningitis episode or intraventricular hemorrhage. The evolution towards increased intracranial pressure requires treatment with bypass but the existence of several compartments forced to put more valves per patient. Neuroendoscopy helped overcome this constraint by unifying the compartments without requiring open surgery and consequently expose the child to several complications.

Methods: In the neurosurgery department of the Central Hospital of Army (Algiers), between March 2013 and March 2014, 10 patients were operated on using this technique

Results: Evolution for 08 patients was favorable clinically and radiologically. 01 patient had an infectious complication treated with antibiotics and received a bypass, and 01 patient died postoperatively without being able to identify the cause of death.

Conclusion: Neuroendoscopy enabled major advances in the treatment of complex hydrocephalus: first because it is a minimally invasive technique, and also because it avoids, or minimizes at least, the postoperative complications

Keywords: neuroendoscopy, complex hydrocephalus, increased intracranial pressure, valve

FL-055

Hydrocephalus

Surgical treatment of multiloculated hydrocephalus in children. Our experience

Sergei Kim¹; German Letyagin¹; Vasilij Danilin¹; Anna Sysoeva¹.

¹Federal Center of Neurosurgery, Novosibirsk.

Objective: Patients with different variants of multiloculated hydrocephalus are the most complex patient group faced by neurosurgeons. There is still no consensus which method of surgery should be preferred as the most effective and safe. Standard shunt implantation does not bring positive results. In our opinion, neuroendoscopic intervention in these patients is the most promising method of surgery.

Methods: 234 patients with various forms of hydrocephalus were treated in our hospital in 2013-2014 years. 19 (8%) of them had multiloculated hydrocephalus. 3 patients (16%) were treated using modified ventriculoperitoneal shunts. In 16 children (84%) we used neuroendoscopy. Nine interventions performed simultaneously with the shunt implantation. In 11 endoscopic procedures we used neuronavigation.

Results: A total of 28 procedures were performed in 19 children, including 11 navigated neuroendoscopic operations. There were 9 reoperations,

2 of them in group of navigated endoscopic surgery. All the children achieved clinical improvement. Follow-up included clinical examination and evaluation of MRI.

Conclusion: Endoscopic surgery is the treatment of choice in patients with multiloculated hydrocephalus. Application of navigated neuroendoscopy makes this kind of operations the most efficient and safe for the patient. The aim of operation is to drain maximum possible number of cavities using minimal amounts of the proximal catheters and shunting systems, and sometimes even prevent shunting.

Keywords: multiloculated, hydrocephalus, neuroendoscopy, navigated, endoscopy

FL-056

Hydrocephalus

Effectiveness of endoscopic third ventriculostomy combined with choroid plexus coagulation in the management of myelomeningocele associated hydrocephalus

¹Volkan Etus; ²Abidin Murat Geyik; Hakan Karabağlı³.

¹Kocaeli University Faculty of Medicine, Department of Neurosurgery, Kocaeli; ²Gaziantep University Faculty of Medicine, Department of Neurosurgery, Gaziantep; ³Selcuk University, Selcuklu Faculty of Medicine, Department of Neurosurgery, Konya.

Objective: The management of hydrocephalus is one of the most trying problems in children with myelomeningocele. In this study we compared the effectiveness of endoscopic third ventriculostomy (ETV) combined with choroid plexus coagulation (CPC) with ETV alone for the management of myelomeningocele associated hydrocephalus.

Methods: The data of total 87 myelomeningocele associated hydrocephalus cases belonging to two centers which, have been treated with ETV or ETV+CPC were retrospectively analyzed. The patients who had been followed up for at least one year postoperatively were included in the study. Of the cases 53 were treated with ETV alone (Group-A). ETV+CPC procedures were performed in 34 cases (Group-B).

Results: The success rate of ETV was 35.8% for Group-A. For Group-B the success rate was calculated as 44.1%. Complication rates and postoperative morbidity rates showed no significant difference when Group-A and Group-B were compared.

Conclusion: Our results seem to suggest that the success of ETV is relatively higher when combined with endoscopic CPC in MAH.

Keywords: choroid plexus coagulation, endoscopic third ventriculostomy, hydrocephalus, myelomeningocele

FL-057

Hydrocephalus

Comparison outcome of VP shunting versus endoscopic third ventriculostomy with choroid plexus cauterization in children less than one year with obstructive hydrocephalus

Amir Amini Navaei¹; Zohreh Habibi¹; Farideh Nejat¹; Ehsan Moradi¹.

¹TUMS, Tehran.

Objective: One of the main challenges in treating hydrocephalus is choosing ETV or VP_shunt, in spite of the other studies there is no definite guidelines yet, especially in the infants. The present study is going to investigate the result of ETV/CPC versus VP_shunt in children less than one year old with obstructive hydrocephalus.

Methods: The present randomized clinical trial was conducted on all obstructive hydrocephalus patients less than one year whom did not take any treatment before, written informed consent obtained. Outcome

evaluated based on clinical signs and imaging. Follow up was done at least for 6 months.

Results: The study performed on 30 participants (16male, 14 female). Aqueductal stenosis was most prevalent cause of hydrocephalus (19). 16 of them treated by VP_shunt and the rest 14 by ETV/CPC. Failure was found in 4 children in the ETV/CPC and 3 for VP_shunt but the difference was not significant.

Conclusion: Our primary results show both mentioned methods are the same, which is in concordance with other studies but this is an ongoing study and the final result will be available whenever the study will finish.

Keywords: ETV, CPC, shunt, obstructive hydrocephalus

FL-058

Infection

The use of neuroendoscopy in the treatment of refractory ventriculitis

Daniel Cardeal¹; Hamilton Matushita¹; Fernanda Andrade¹.

¹Neurosurgeon, Sao Paulo.

Objective: Ventriculoperitoneal shunt (VPS) is the main form of treatment of hydrocephalus in children. The VPS infections may be difficult to treat. Most neurosurgeons choose to remove the shunt and use systemic or intraventricular antibiotics and external ventricular shunt (EVS) before implanting a new VPS. In some cases, ventriculitis become refractory requiring long period of antibiotic treatment. We evaluated the use of neuroendoscopy in the treatment of refractory ventriculitis.

Methods: Nine children with repeated ventriculitis were submitted to neuroendoscopy with exhaustive “washing” of ventricular cavity and aspiration of pyogenic granuloma if present. We performed endoscopic fenestration in cases of multiple septations and, if possible, ventriculostomy with choroid plexus coagulation. We analyzed the cerebrospinal fluid and evolution of the infection after the procedure.

Results: Eight patients were under 15 months of age and one was 12 years old. Eight patients had ventriculitis related to VPS infection and one as a complication of meningitis. The etiologic agent was identified in 6 cases: 2 cases staphylococcus aureus, 2 cases staphylococcus epidermidis and 1 case pneumococcus, 1 case Enterococcus faecalis. Empirical antibiotics were the choice in cases of negative cultures. The cases were treated for about 3 weeks with EVS and systemic antibiotics without satisfactory improvement of the CSF. There were no procedure-related complications. The follow-up ranged from 4 months to 1 year. There was an improvement of the CSF after the procedure in 8 cases.

Conclusion: Neuroendoscopy is a safe procedure and helps the treatment of refractory ventriculitis decreasing hospitalization and antibiotic treatment.

Keywords: ventriculitis, hydrocephalus, neuroendoscopy

FL-061

Brain malformation

Occipital encephaloceles: a single center experience of 33 cases (2000-2014)

Selami Cagatay Onal¹; Yener Akyuva¹; Tuncay Ateş¹; Gökhan Reşitoğlu¹.

¹Inonu University School Of Medicine Department Of Neurosurgery, Malatya.

Objective: The aim of this presentation is the declaration of specific characteristics of thirty-three occipital encephalocele patients operated in a single institution within a period of fifteen years (2000-2014).

Methods: The charts of thirty-three occipital encephalocele patients were reviewed retrospectively. Clinical symptoms, surgical timing and strategy, infection rates, existence and progression of hydrocephalus, coincidental pathologies, prognosis and outcome are studied.

Results: Large masses are associated with neurological deficits such as cranial nerve abnormalities, poor feeding and sucking abilities, developmental delay, spasticity and blindness. Some patients present with large sacs and microcrania. Two of the patients had associated neural tube defects. One patient had concomitant cleft palate anomaly. Another had West Syndrome with intractable epileptic seizures. Concomitant or progressive hydrocephalus giving rise to shunt surgery was very common.

Conclusion: Prognosis and long term outcome are directly related to the amount of neural tissue involved in the sac and coexistence of other pathologies. Those patients who have CSF and very small volume of dysplastic neural tissue in the sac have a good chance of performing a normal or near-normal physical and neurological development. On the other hand, the more the neural involvement and the presence of associated neural abnormalities, the greater the rate of mental retardation and poor prognosis.

Keywords: Occipital encephalocele, hydrocephalus, Surgery, Outcome, Child

FL-062

Hydrocephalus

Incidence of secondary hydrocephalus after excision of huge encephaloceles in neonates - case study

Ehab El Refaee¹; Mohamed Refaat¹; Mohamed Reda¹.

¹Department Of Neurosurgery- Cairo University, Cairo.

Objective: Encephalocele represents as protrusion of cranial contents skull through a defect in the cranium, where the most common sites of occurrence are occipital and frontonasal regions. The surgical outcome is reported to be satisfactory. However, incidence of hydrocephalus in patients with encephaloceles is variable in the past literature. This study is aiming for detection of the relation between the size of the encephaloceles and the occurrence of hydrocephalus.

Methods: Data of all neonates with encephaloceles operated upon from September 2012 till September 2014 in our institute were collected. Surgery was performed during the first two weeks age in all cases. Data for the cases with encephaloceles larger than 10 cm in diameter were studied, where the clinical picture, surgical technique, pre and postoperative imaging and the follow up were analyzed.

Results: During the previously mentioned period 19 cases with encephaloceles were operated upon. The mean follow up period was 7months. 16 cases were occipital and 3 cases where frontal. In all cases the diameter of the cystic protrusion was larger than 10 cm. However, in 4 cases the largest diameter (measured on the MRI or CT images) was larger than 18 cm. All cases with frontobasal encephaloceles were smaller than 10 cm in diameter thus excluded from the study. In 7 cases, post operative ventriculomegaly occurred, of these 4 cases needed a permanent VP shunt implantation. Wound dehiscence occurred in 2 cases, that required secondary sutures with favorable outcome. One case died two weeks after the surgery due to poor general condition and wound infection.

Conclusion: Early surgical excision provides effective treatment in huge encephaloceles. 21% of cases could require CSF diversion afterwards depending on the associated anomalies. Despite of their size, giant encephaloceles can have excellent prognosis with no need for further treatment or CSF diversion.

Keywords: Encephalocele, hydrocephalus, congenital anomalies

Wednesday, 7 October 2015

14:00 – 14:50

Spine Flash Presentations V

FL-063

Craniovertebral Junction

Do we fix, halo or collar paediatric atlanto-axial rotatory fixation (AARF)? Is less more?!

Digant Kamdar¹; Guirish A. Solanki¹.

¹Departments of Paediatric Neurosurgery and Inborn Errors of Metabolism, Birmingham Children's Hospital NHS Foundation Trust, Birmingham

Objective: While torticollis is commonly seen, true AARF cock-robin deformity is an uncommon pediatric injury with potential for severe neurological deficit. Some experts use traction (up-to 72hrs) to determine treatment (orthosis or surgery). However very young age, learning disabilities, autistic spectrum behaviour or simply children too frightened to cooperate makes treatment fraught with difficulties. We report our institutional experience in the management of AARF. Patients: 32 children (19 girls, 13 boys) were treated between 2007 and 2015. F:M ratio:1.46. Median age: 6.4 years. Girls significantly older (8 1/2 vs 5 3/4) years.

Methods: Fieldings radiological classification was used. Only children with radiologically confirmed AARF were included. Children were treated with muscle relaxation, neck collar and manipulated under anaesthetic (MUA) at the earliest opportunity. Halo orthosis (Halo) used in late (>1month) presentation or failure to reduce. Surgery for recurrence despite Halo immobilisation.

Results: Falls and trauma accounted for 53% (37.5%, 15.6%) of cases respectively. 25% (8) were due to post-infectious syndrome (Grisels, TB osteomyelitis), 9% (3) in Downs, 6% (2) were iatrogenic following surgery, 6% (2) idiopathic cases. Post-Infectious cases were three times higher in girls. 62.5% (20) children were initially treated with a collar. 66% (21) ultimately required a halo orthosis. TB osteomyelitis required prolonged antibiotic treatment and halo immobilisation. However, only one child (3%) required a posterior C1-2 fusion for recurrent AARF. Complications included pin-site infection and halo slippage. There were no treatment-related cranial nerve or neurological deficits.

Conclusion: In children, AARF presenting with a painful torticollis often resolves with muscle relaxation and neck collar alone over a period of days. We propose that early presentation should be treated with muscle relaxants and a well-fitting collar initially ± MUA. Halo should be considered if MUA and collar treatment fail, or in overt instability. While rare, instability or recurrent AARF (despite Halo immobilisation over 3months) should be considered for surgery.

Keywords: Atlanto-Axial Rotatory Fixation, AARF, AARS, cervical, spine, cord compression

FL-064

Spine malformation

Does folic acid supplementation impact on the survival of myelomeningocele patients?

Jose Francisco M. Salomao¹; Renato M. Salomao²; Tatiana P. Cervante¹; Antonio R. Bellas¹; Soniza V. A. Leon².

¹Division Of Pediatric Neurosurgery National Institute Of Women, Children And Adolescents Health Fernandes Figueira (IFF), Rio De Janeiro; ²Center Of Biologic And Health Sciences. Federal University of The State Of Rio De Janeiro, Rio De Janeiro.

Objective: According some reports fortification of flours with folic acid may decrease the mortality of myelomeningocele (MMC) patients. The

aim of this study is to compare the outcome of two cohorts of MMC patients before (BAFF) and after (AFAF) fortification of flours became mandatory in Brazil in 2003.

Methods: Retrospective transversal study of 383 patients with MMC operated at our institution from January 1990 to December 2013. Patients not operated at our hospital and those not regularly followed up were excluded. The AF period was considered as beginning 36 months after issue of the law. There were 225 patients in Group 1 (BAFF) and 158 in Group 2 (AFAF). In both, neonatal deaths were studied separately and the groups were treated statistically.

Results: Overall 39 (10.1%) patients died: 16 (4.17%) during the neonatal period and 23 (6.0%) during a follow-up period ranging from 2-132 months. In the BFAF Group there were 31/225 (13.8%) deaths, being 11 (4.8%) in the neonatal period and 20 (8.9%) in older children. In the AFAF Group there were 8/158 (3.6%) being 5 (2.2%) neonatal and 3 (1.3%) in the follow-up group. There was a significant decrease in the overall mortality of the AFAF Group ($p<0,005$) as well as in the follow-up of this Group ($p<0,0040$). No statistical differences were found when comparing neonatal mortality in both groups $p=0,5680$).

Conclusion: Supplementation of flour with FA may impact on the survival of patients with MMC, especially during the follow-up period. The lack of impact during the neonatal period may be due to other congenital malformations, systemic complications or genetical influences on this group. Other contributory factors that may reduce mortality in the AFAF Group will be also considered.

Keywords: myelomeningocele, folic acid, neural tube defects, spina bifida

FL-065

Functional

Evaluation of tibial somatosensory evoked potentials in young children with tethered cord syndrome

Ezgi Erdogan¹; Huseyin Canaz¹; Ibrahim Alatas¹; Nursu Kara²; Kerem Ozel¹.

¹Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul.

Objective: The aim of this study is to investigate any deterioration in tibial somatosensory evoked potentials in correlation with lower extremity clinical findings in patients with tethered cord syndrome.

Methods: Sixteen patients age between 2.5-14 years, were taken to tibial SEP recording. Ten patients were being followed up for a tethered cord syndrome secondary to previous myelomeningocele surgery. Tibial SEPs were recorded from lumbar and cortical regions with the electrical stimulation of posterior tibial nerve without any need for sedation or anesthesia. Only cortical responses were evaluated because of the low reproducibility of lumbar responses due to muscle artifact. Lower extremity neurological findings were classified into 5 clinical groups (1-normal, 5-severe deficit) while changes in SEP responses classified into 3 groups (normal-mild, moderate, severe).

Results: In clinical group 5; there were 4 children. Two of them had severe, two had moderate changes in SEP. In clinical group 4 there was only 1 child who had severe changes in SEP responses. In clinical group 3, there were 3 children, one of them had severe, two had moderate changes in SEP. In clinical group 2; there were 6 children, 3 of them had normal-mild changes, 1 child had moderate changes and 2 children had severe changes in SEP. In clinical group 1 (no neurological deficit or complaints) there were 2 children both of them had normal SEP findings.

Conclusion: There is an agreement in the literature that tibial somatosensory evoked potential is the most sensitive electrophysiological evaluation for tethered cord syndrome. Our results are

compatible with literature. There is a correlation between the severity of clinical findings and deterioration in SEP responses. Since there is no normative data of SEP responses for young children, it will be valuable to include SEP recordings to follow up protocols of tethered cord syndrome.

Keywords: SEP, Tethered Cord Syndrome, Young children

FL-066

Spine malformation

The results of rectal manometry application in lipomyelomeningocele surgery

Kirill Sysoev¹.

¹Polenov Neurosurgical Institute, St. Petersburg.

Objective: The most common postoperative complication in lipomyelomeningocele surgery is considered to be bladder dysfunction. Because of this, the new methods of intraoperative monitoring of spinal cord autonomy functions need to be developed.

Methods: In 7 children aged between 2 mos - 2 years lipomyelomeningocele repair operation was carried out. Rectal manometry was performed intraoperatively. Balloon dilatation catheter with a pressure transducer was inserted into rectum and initial pressure was measured. At the same time electrostimulation mapping of the nerve roots, lower extremities and rectal external sphincter somatosensory and motor evoked potentials monitoring was carried out.

Results: During lipoma resection when bipolar electrocoating was performed intrarectal pressure decrease of 1-3 mm Hg was noted; during lipoma ultrasonic disintegration pressure transient increase of 1-2 mm Hg was registered. In these settings, the manipulations were stopped till the initial measurements recovery. Never intrarectal pressure changes matched with rectum external sphincter motor response, which is obtained by sacral nerve roots electrostimulation. The use of this method allowed us to avoid from postoperative pelvic disorders increasing. In one large transitional lipomyelomeningocele as pressure pronounced changes were found out, we decided only partially to remove the lipoma. In one patient with existing severe pelvic disorders rectal pressure changes during manipulation have not been identified.

Conclusion: Modern approach to lipomyelomeningocele surgery means complete removal of adipose tissue as much as possible on the border of the spinal cord - lipoma. This region corresponds to the sacral parasympathetic centers. In our opinion, rectal pressure changes can be estimated as a sign of the impact on the parasympathetic sacral centers. Application of this method seems to be justified in the surgical treatment of lipomyelomeningocele in children.

Keywords: lipomyelomeningocele, intraoperative monitoring, rectal manometry, tethered cord syndrome, surgery

FL-067

Spine malformation

The association of urodynamic findings and conus medullaris level detected by MRI and CT-myelography in scoliosis patients

Amir Azarhomayoun¹; Sara Hanaei¹; Kourosh Karimi Yarandi²; Abbas Amirjamshidi².

¹Tehran University Of Medical Science (TUMS), Tehran; ²Departments Of Neurosurgery, Sina Hospital, Tehran University Of Medical Sciences, Tehran.

Objective: Scoliosis is abnormal spine curvature as a result of abnormal vertebral shape or alignment. Consequently, the spinal cord will have abnormal alignment resulting in cord/root compression or low level conus

medullaris, which are associated with a variety of presentations. Although asymptomatic in some patients, low level conus medullaris can cause spectrum of urodynamic abnormalities. Hence, we have investigated the association of conus medullaris level and urodynamic findings in scoliosis patients.

Methods: A total of 65 patients with confirmed diagnosis of scoliosis underwent urodynamic study test and results were categorized into normal, mild, moderate or severe abnormality. Conus medullaris level was assessed by MRI and CT-myelography in applicable patients.

Results: The average conus level was L2. There was not significant association between conus medullaris level and urodynamic findings on MRI ($p > 0.05$). On the other hand, a significant association has been found between urodynamic findings and conus medullaris level detected by CT-myelography ($P < 0.05$). Patients with normal urodynamic findings tended to have higher average of conus medullaris level (upper than L3) compared to patients with severe urodynamic abnormality whose conus medullaris level were in lower position (mostly lower than L2-L3). Although association of UDS and scoliosis type was significant ($p < 0.05$), it was not the same for cobb's angle or KINGs classification.

Conclusion: The discrepancy between association of urodynamic findings and conus level in MRI or CT-myelogram might be explained as low quality and accuracy of MRI in finding the exact level of conus medullaris. Moreover, conus level was undetectable in some cases with MRI which could have possibly affected the results. Therefore, it seems that CT-myelogram is more accurate in detecting conus level in scoliotic patients. Besides, lower levels of conus medullaris is associated with severe urodynamic abnormalities while upper levels of conus is associated with normal urodynamic findings.

Keywords: Scoliosis, conus medullaris, CT-myelography, Urodynamic study

FL-068

Spine malformation

Split cord malformation in association with sacral extradural arachnoid cyst

Zohreh Habibi¹; Farideh Nejat¹; Sara Hanaei¹.

¹MD, Tehran.

Objective: Split cord malformation (SCM) may occur in association with open or closed spinal dysraphism, and other spinal abnormalities including kyphoscoliosis, hemivertebra, butterfly vertebra, and Chiari malformation. Few cases of concurrent intradural arachnoid cyst and SCM have been sporadically described; however, sacral extradural arachnoid cyst with SCM is very rarely reported.

Methods: Following institutional ethics committee approval, 7 patients with concomitant split cord malformation and sacral extradural arachnoid cyst were consecutively enrolled among 73 patients who were operated on for SCM type I or II between April 2008 and April 2014. Those with arachnoid cysts above sacral level or intradural cysts were not included.

Results: There were 7 patients with the age ranging from 18 to 119 months (mean 56.71 \pm 39.49). Considering preoperative images, SCM-I and SCM-II were detected in 6 and 1 cases, respectively. All cases harbored sacral extradural cysts containing fluid similar to cerebrospinal fluid and occasionally showing fistulous connection to subarachnoid space in MR myelograms. Sacral canal dilation with scalloped posterior elements was observed on CT in most cases. Tethered cord was detected in all 7 patients, and 6 ones had syrinx rostral to the level of SCM. There were 3 cases of scoliosis, and 4 patients suffered from lower extremities weakness. All patients had abnormal urodynamic study, mainly high pressure bladder and detrusor sphincter dyssynergia, among whom 2 ones had bladder wall thickness and 2 had post-voiding residue in ultrasound study.

Conclusion: It may not be possible to separate the symptoms of split cord malformation from those of extradural sacral arachnoid cyst in concurrent

occurrence. In symptomatic cases, it seems better to surgically manage both lesions in a single session. The most important advantage of handling both distinct lesions in one setting is a single exposure anesthesia which remains an important consideration in a child.

Keywords: SCM, sacral cyst, arachnoid cyst, extradural

FL-069

Spine malformation

Spinal congenital dermal sinuses King Hussein Medical Centre experience

Nidal Khasawneh¹.

¹Neurosurgery Department, King Hussein Medical Centre, Amman.

Objective: Spinal congenital dermal sinus tracts are a form of spinal dysraphism. Cause is attributed to a failure of dysjunction during fetal development. Presenting in childhood with skin stigmata, neurologic deficit, or infection. We reviewed our surgical experience, examining presenting symptomatology, operative findings, and patient outcomes.

Methods: We retrospectively analyzed 52 cases of dermal sinus tract operated over the last five years. All cases were operated on by the author from 2010 to 2015.

Results: Fifty two patients were identified; 32 female and 20 male. Nine cervical, 7 thoracic, 17 lumbar, and 19 lumbosacral tracts were explored. Thirty patients presented at <1 year of age, and 22 were >1 year. Reasons for referral included cutaneous findings (28), neurologic deficit (15), foot abnormalities (7), infection (6), pain (4), and scoliosis (1). Our initial examination revealed cutaneous findings (eg, sinus ostia, pigmentation changes, erythema, skin tags, subcutaneous masses) in 50 patients and neurologic deficit in 35. Age-related differences were apparent. Patients >1 year were more likely to have neurologic deficit (90%) as compared with those <1 year (53%). Bifid spinous processes were noted at dural tract entry in 32 patients, 40 tethered cords, 25 inclusion tumors, and 11 patients with evidence of arachnoiditis were found intraoperatively. Mean follow-up was 30 months. 20 (39%) remained neurologically intact, 22 (42%) improved, 4 (8%) were unchanged, and 6 (11%) were worse with 4 having decreased perianal sensation and 2 slightly worsened motor function postoperatively.

Conclusion: Patients were mostly referred for cutaneous stigmata evaluation. More than 50% had neurologic deficit, intradural tumors, or tethered cords. Skin stigmata should raise alarm and initiate prompt radiologic evaluation. Neurosurgical intervention with intradural exploration may preserve or improve neurologic function in these patients.

Keywords: Congenital Dermal Sinuses, spinal dysraphism, cutaneous stigmata

FL-070

Spine

Pediatric cervical kyphosis: review of literature and case series.

Mohammad Alfawareh¹; Abdulmajeed Alahmari¹.

¹Spine Department, King Fahad Medical City, Riyadh.

Objective: The purpose of this study is to review the etiology, presentation and management of this condition with report of our case series.

Methods: Pediatric cervical kyphosis is rare condition with heterogeneous etiologies that represent challenges in management. The unique characteristic of pediatric spine places them a greater risk with respect of deformity and rate of progression. The purpose of this study is to review the etiology, presentation and management of this condition with report of our case series. Methods: we performed Medline search of the literature using keywords: pediatric cervical kyphosis, children cervical kyphosis, and cervical deformity. Also the references in the obtained articles are reviewed. At

same time we reviewed our case series of pediatric cervical kyphosis presenting to our institution are presented with management of each.

Results: Cervical kyphosis can result from extensive laminectomies, trauma, neoplasms, metabolic disorders or congenital diseases. Well-defined indications for intervention not well-explored in the literature although presence of neurological deficit and pain are among the indication. Our series of six cases, one was caused by tumor, the second was caused by spine TB, the third case congenital and the last three were related to neurofibromatosis. Three of our cases were operated, with successful results, the fourth one was operated after she developed severe quadriplegia but couldn't be recovered and last two still under observation

Conclusion: Pediatric neck kyphosis is serious condition, it may need closed monitoring, diagnosis and management, progression of deformity or minor trauma can cause permanent neurological consequences. Early recognition and treatment is essential.

Keywords: neck kyphosis, congenital, neck deformity

FL-071

Spine malformation

Clinical and radiological evaluation of the spinal deformity and thoracic capacity in the patients with Jarcho-Levine syndrome

Burak Abay¹; Ibrahim Alatas²; Huseyin Canaz²; Isik Karalok³; Ezgi Erdogan²; Nursu Kara⁴; Kerem Ozel².

¹Department of Orthopedic Surgery, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ³Department of Radiology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ⁴Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul.

Objective: Jarcho-Levin syndrome (JLS) is a congenital disorder characterized by a variety of spinal and costal deformities.

Methods: In this study, we retrospectively evaluated the clinical examinations, plain X-rays and MRI scans of spinal deformities and thoracic capacity of 47 patients who have both spondylocostal dysostosis and neural tube defect, between 2010 and 2014.

Results: We evaluated the spinal deformities and thoracic capacity of 47 patients (19 male, 28 female) with Jarcho-Levin syndrome with mean age of 5,1 (1-18). The maximum Cobb angle was measured for scoliosis. The mean Cobb angle was 45,3° (9,7°-129,4°). We measured mean thoracic lordosis of 23,0° (2,2°-74,4°) in 31 patients. We evaluated mean lumbar kyphosis of 59,4 (7°-143,9°) in 29 patients. The mean space available for the lung (SAL) was calculated as 93,83% (63,55%-129,21%). Thoracic width was measured at the level of T6 vertebra or similar. Thoracic spine height was measured from the superior end plate of T1 to the inferior end plate of T12 in the anteroposterior X-ray. The mean ratio between the thoracic height and thoracic width was measured as 82,31% (53,53%-135%).

Conclusion: In this study, our aim was to survey the spinal and thoracic deformities of 47 patients with Jarcho-Levin syndrome. We observed increased prevalence of scoliosis, thoracic lordosis and lumbar kyphosis and decreased thoracic symmetry in this clinical series.

Keywords: Jarcho Levin Syndrome, Spina Bifida, Thoracic Capacity

FL-072

Spine

Spinal kyphotic deformity after laminoplasty in children

Christos Chamilos¹; George Papaevangelou¹; Spyros Sgouros¹.

¹"Mitera" Childrens Hospital, Athens.

Objective: Laminoplasty has emerged in recent years as deformity preserving alternative to laminotomy. The purpose of this study is to analyze the short-to-medium-term kyphotic deformity incidence in a small series of pediatric patients.

Methods: We analyzed retrospectively 10 patients operated for various pathologies (tumor, cysts, vascular) with laminoplasty in the last 7 years in a single institution. The laminoplasty was performed with craniotome and the laminas were attached back with sutures (cranially, caudally and on both sides laterally). We reviewed the incidence of subsequent kyphosis along with other factors (age, sex, pre-op deformity, levels of laminoplasty and pathology type). We measured kyphosis with the Cobb angle in the sagittal plane.

Results: The mean age was 9,8 years (range: 7 m – 18 y) and the mean duration of follow-up was 2,2 years (range 6 m -7 y). There were 6 tumors, 2 cysts (arachnoid and dermoid) and 2 AVM. Extent of laminoplasty was: 5 levels in 3 patients, 4 levels in 3 patients and 3 levels in 4 patients. The level laminoplasties were: 2 thoracolumbar, 3 thoracic, 1 cervical, 2 cervicothoracic and 2 lumbosacral. At latest follow up, only 2 patients (20%) developed mild kyphosis (1 irradiated for spinal cord glioblastoma, Kobb: 32o and 1 with vascular epidural malformation and adult body type, Kobb: 40o).

Conclusion: In our series there was no significant deformity after laminoplasty in the short to medium term follow up. There was no obvious correlation with the underlying pathology. The technique of laminoplasty appears safe with respect to subsequent risk of kyphosis. Longer follow-up is needed in order to determine the long-term deformity risk.

Keywords: laminoplasty, kyphosis, spinal deformity, spinal surgery

FL-073

Spine

Primary tumors of the mobile spine in children

David Pincus¹.

¹University Of Florida, Gainesville, FL.

Objective: Primary tumors of the spine are rare entities in children. The management of these lesions is complex and may involve excision, complex reconstruction, chemotherapy and radiation. We present an overview of spinal tumors in children and include case examples from our series. A treatment algorithm for the evaluation, surgical management and adjuvant therapies for these rare tumors will be provided.

Methods: The author's experience with evaluation and management of primary spinal column tumors in children was reviewed from 2000 to 2015. Medical records, pre- and postoperative imaging and types of adjuvant therapies were analyzed. A review of the literature was performed. Selected children underwent CT guided need biopsy followed by tailored treatment.

Results: Eight patients representing examples of aneurysmal bone cyst, giant cell tumor, chroma, osteoblastoma, chondrosarcoma, Ewing's sarcoma and osteoid osteoma were identified. Appropriately selected patients underwent CT guided need biopsy followed by tailored treatment. This strategy allows for tailored treatment including en bloc resection with spinal reconstruction, piecemeal resection with or without instrumentation followed by adjuvant therapy as indicated. Small, benign tumors may be resected and cured with few complications. In some cases, such as Ewing's sarcoma, no resection is needed and good outcomes are achieved with radiation and chemotherapy alone. Treatment of large, malignant tumors may involve spinal column resection and reconstruction and is associated with significant morbidity.

Conclusion: Spinal oncology is a rapidly developing field. For complex tumors, emergent CT guided needle biopsy is critical for treatment planning, even in the setting of neurological deficits. This strategy avoids contamination of the surgical field which may lead to improved outcomes with aggressive tumors. Recent advancements in surgical techniques,

chemotherapy and radiation appear to be improving outcomes. For aggressive and malignant tumors, the literature clearly demonstrates the benefit adherence to the Enneking principles of en bloc resection.

Keywords: spine, tumor, chordoma, giant cell tumor, chondrosarcoma

Wednesday, 7 October 2015

17:30 – 18:00

Spine Flash Presentations VI

FL-074

Trauma

Early diagnosis and treatment of growing skull fracture

Ishwar Singh¹; Saquib Azad Siddiqui¹; Seema Rohilla¹; Prashant Kumar¹.

¹Pt. B. D. Sharma University of Health Sciences, Rohtak, ROHTAK.

Objective: Growing skull fracture (GSF) is a rare complication of pediatric head trauma and causes delayed onset neurological deficit and cranial defect. GSF usually developed following the linear fracture with underlying dural tear resulting in herniation of brain. Early diagnosis and treatment are essential to avoid complication. However, there are no clear-cut guidelines for early diagnosis of GWF. Present study was conducted to laid down criteria for early diagnosis of GWF

Methods: : From 2010 to 2015 all pediatric patients of head trauma with linear fracture were evaluated. Patients of age < 5 year with cephalhematoma, bone diastasis of 4mm or more with underlying brain contusion were subjected for contrast MRI brain to find out the dural tear and herniation of brain matter. Patients with contrast MRI showing dural tear and herniation of brain matter were considered for high risk for development of GSF and treated surgically within one month of trauma. Patients with contrast MRI brain not showing dural tear and herniation of brain matter were regularly followed for any signs of GWF.

Results: Total 16 patients were evaluated and treated based on above four criteria. Four patients in which MRI did not show dural tear and herniation of brain matter were regularly followed –up and does not show any sign of GSF later on follow up.

Conclusion: Early diagnosis of GSF can be made based on the four criteria i.e. (1) age < 5 year with cephalhematoma, (2) bone diastasis 4mm or more (3) underlying brain contusion (4) contrast MRI showing dural tear and herniation of brain matter. Dural tear with herniation of brain matter is main etiopathogenic factor for development of GWF. Early diagnosis and treatment of GSF can yield good outcome

Keywords: growing skull fracture, pediatric, head injury, diagnosis and treatment

FL-075

Trauma

Determinants of blood loss in paediatric decompressive craniectomy following head injury

Edward Rice¹; Ian Anderson¹; Gnanamurthy Sivakumar¹; Paul Chumas¹; Atul Tyagi¹; John Goodden¹.

¹Paediatric Neurosurgery, Leeds Children's Hospital At The Leeds General Infirmary, Leeds.

Objective: Paediatric decompressive craniectomy following head injury remains a contentious topic, with recently published evidence suggesting a high risk of large volume intraoperative blood loss (IBL). This study aimed to corroborate or refute this suggestion and to identify potential risk factors.

Methods: 16 children underwent decompressive craniectomy following head injury during a 5-year study period. IBL and estimated blood

volume (EBV) were calculated and assessed for correlation with coagulopathy, polytrauma, age, grade of operating surgeon and surgeon-estimated blood loss (SEBL).

Results: Mean IBL was 1261 ± 1253 mLs. Median IBL/EBV was 0.34. Mean IBL/EBV was 0.45 ± 0.39 , the range being zero to 1.15. Six (38%) patients had IBL/EBL ≥ 0.5 , i.e. lost more than half of their EBL during the perioperative period. IBL did not correlate with coagulopathy, polytrauma or SEBL. A weak inverse correlation existed between IBL and patient age, and a trend approaching significance ($p=0.055$) suggested reduced IBL when surgery is performed by a neurosurgical consultant as opposed to a trainee.

Conclusion: This study confirms that there is a high risk of large volume blood loss in paediatric decompressive craniectomy following head injury, but no significant risk factors were identified. Larger, multi-centre studies are required to reliably identify risk factors for IBL, and should include grade of operating surgeon to further assess the apparent association between grade and IBL.

Keywords: traumatic brain injury, craniectomy, blood loss

FL-077

Trauma

Magnet-encephalography (MEG) studies in late accessory to suprascapular nerve transfer in obstetric brachial plexus lesions. Change of cortical motor programs

W.J.R. Van Ouwerkerk¹; A Hillebrand²; B Van Dijk².

¹Vrije Universiteit Medical Center, Dpt Neurosurgery, Amsterdam; ²Vrije Universiteit Medical Center, Dpt Neurophysiology, Amsterdam.

Objective: To investigate possible changes in central cortical representation a pilot study was performed in a case of a boy aged 15 years presenting with a late obstetric brachial plexus lesion (OBPL) with lack of active exorotation. A nerve transfer from the Accessory to Suprascapular Nerve was performed. Change of cortical representation for specific movements during follow up is studied using Magnet-encephalography (MEG).

Methods: Children with Group I OBPL of spinal roots C5 and C6 or the Superior Trunk may recover spontaneously except for active glenohumeral exorotation. All functions recover except for the function of the Suprascapular Nerve, which surprisingly is a branch of the otherwise recovered and functioning Superior Trunk. Contradictory observations are made in a series of these children. Electromyography showed voluntary muscle activity and MRI showed hardly any muscle atrophy or fatty degeneration of spinatus muscles. Intraoperatively the Suprascapular Nerve was reactive. The lack of active exorotation can be hypothesised to be caused by developmental apraxia. Central motor programs are developed between the 3rd and 7th month. The time frame where recovery of the OPBL also takes place. Other developing motor programs during recovery may irreversibly dominate over exorotation. In over 85 patients where only active exorotation did not recover an Accessory to Suprascapular Nerve Transfer was performed at varying ages, the majority over 12 months and even in much older children from 3 to 15 years of age. Over 90% of children reached.

Results: The cortical MEG-representation for exorotation of the affected arm changes from contralateral to bilateral and the representation of the non-affected arm changes from contralateral to ipsilateral representation.

Conclusion: Successful nerve transfer after an obstetric brachial plexus lesion seems not time dependent. In a pilot study MEG is a useful tool to study cortical representation of movement and to evaluate functional changes after nerve transfer.

Keywords: magnetencephalography, obstetric brachial plexus lesion, nerve transfer, apraxia

FL-078

Vascular

Risk factors associated with germinal matrix-intraventricular hemorrhage in preterm neonates

Dongwon Kim¹.

¹Department of Neurosurgery, Dongsan Medical Center, Daegu.

Objective: The purpose of this study is to identify the risk factors associated with the development of germinal matrix-intraventricular hemorrhage (GM-IVH) and the relationship of the severity of disease and prematurity.

Methods: A total of 168 premature neonates whose birth weight ≤ 1500 g or gestational age ≤ 34 weeks were examined by cranial ultrasound (CUS) for detection of GM-IVH among the babies admitted between January 2011 and December 2012 in our medical center neonatal intensive care unit. The babies were divided into two groups: GM-IVH and non-IVH. Clinical presentations, precipitating factors of the patients and maternal factors were analyzed.

Results: In univariate analysis, gestational age, birth weight, delivery method, presence of premature rupture of membrane (PROM) and level of sodium and glucose were statistically meaningful factors ($p < 0.05$). But only two factors, gestational age and presence of patent ductus arteriosus (PDA) were statistically meaningful in multivariate logistic regression ($p < 0.05$). Delivery method [normal vaginal delivery (NVD) to Caesarean section] was borderline significant ($p < 0.10$).

Conclusion: Presence of PDA and gestational age were the important risk factors associated with development of GM-IVH.

Keywords: Germinal matrix-intraventricular hemorrhage, Gestational age, Ductus arteriosus, patent

Wednesday, 7 October 2015

17:30 – 18:00

Spine Flash Presentations VII: Epilepsy and Functional FL-079

Epilepsy

Invasive monitoring with depth electrodes and electrophysiologically navigated resection of the epileptogenic focus in patients with pharmacoresistant seizures: a pediatric case series

Peraud Aurelia¹; Borggraefe Ingo²; Thorsteinsdottir Jun¹; Noachter Soheyl²; Tonn Joerg-Christian¹; Kreth Friedrich Wilhelm¹.

¹Department Of Neurosurgery, Klinikum Grosshadern, Ludwig-Maximilians-University Munich, Munich; ²Department of Pediatric Neurology, Ludwig-Maximilians-University Munich, Munich; ³Department of Neurology, Klinikum Grosshadern, Ludwig-Maximilians-University Munich, Munich.

Objective: In patients with pharmacoresistant seizures, presurgical workup can identify epilepsy surgery options when non-invasive imaging has failed. Early postoperative recurrences for epilepsy are often associated with suboptimal identification/resection of the epileptogenic focus. In the current pilot study a new approach was tested: the seizure onset zone was determined using invasive monitoring with stereotactically implanted depth electrodes; electrophysiologically navigated resection was done thereafter. We here report on the risks and the effectiveness of this approach in selected pediatric cases.

Methods: After preoperative evaluation using noninvasive techniques (scalp EEG monitoring, MR images) and review in the multidisciplinary epilepsy conference, decision of depth electrode implantation was made. All parents gave informed consent. Depth electrodes were stereotactically implanted. The exact seizure onset zone was determined by video-EEG-recordings and linked to the respective electrode positions in the post-implantation CT-scan (slice thickness: 1.25mm). After image fusion with the preoperative MRI and integration of these data into the navigation system, 3D reconstruction of the focus was possible and electrophysiologically guided resection was initiated.

Results: Out of the whole cohort of 45 patients four pediatric patients were selected for this case series (mean age: 8.7y, range: 3.4–18y). A frontal seizure focus was suspected in all cases (two left and two right hemispheric foci). For invasive monitoring 6–13 electrodes were implanted per patient without any morbidity. Histological examination revealed focal cortical dysplasia in three children and grey white matter differentiation disorder in one. Median follow-up was 13.5 months. Engel Class IA outcome was achieved in all 4 patients, No transient or permanent deficits were seen.

Conclusion: Stereotactic implantation of depth electrodes is safe. It enables precise identification of the epileptic focus even in pediatric patients thereby allowing electrophysiologically navigated focus resection with excellent clinical outcome.

Keywords: focal epilepsy, depth electrodes, invasive video EEG monitoring, electrophysiology, navigated resection

FL-080

Epilepsy

Tailored sus insular disconnection of the central areas: a new functional disconnecting technique for stroke-induced refractory epilepsy.

Scavarda Didier¹; Trebuchon Agnes²; Lepine Anne³; Milh Mathieu³; Pech-Gourg Gregoire¹; Villeneuve Nathalie³; Girard Nadine⁴; Bartolomei Fabrice².

¹Department of Pediatric Neurosurgery, CHU Timone Enfant, Marseille, France, Marseille; ²Department of Neurophysiology, CHU Timone, Marseille, France, Marseille; ³Department of Pediatric Neurology, CHU Timone Enfant, Marseille, France, Marseille; ⁴Department of Neuroradiology, CHU Timone, Marseille, France, Marseille.

Objective: Stroke in children is major cause of morbidity in children. 4 to 25% will develop a refractory epilepsy. Different surgical modalities have been reported such as: cyst fenestration, perilesional cortectomy, multilobar resections/disconnections, Hemispheric disconnection... We developed a more functional approach to treat stroke-induced epilepsy in children based on a sus-insular disconnection of the epileptogenic zone (EZ) tailored by Stereo-Electro-Encephalo-Graphy (SEEG). We report our preliminary results on 3 children with this real and new functional disconnecting technique.

Methods: Prospective study. 3 children who suffered from stroke induced refractory epilepsy. 2 boys, 1 girl. They benefited from: EEG, Video EEG, MRI, functional MRI, PET scan, MEG and visual evaluations. 2 children benefited from pre operative Wada test. They presented with daily seizures and frequent cranial traumatism. A frameless SEEG (Stealthstation Medtronic) was performed for each.

Results: SEEG allowed in each case to delineate the epileptogenic zone. EZ was centered in all cases by the ischemic area and surrounded the central sulcus. These studies showed that a Sus-insular disconnection would permit to protect the functional areas while disconnecting the EZ. Mean follow up is 13 months (24–3 months). All 3 patients are seizure free (Engel 1). The postoperative hemiplegia recovered within 3 weeks. The first 2 operated children improved their autonomy and were able to resume their schooling. For the latter the follow up is still too short.

Conclusion: Sus insular disconnection of the EZ tailored by SEEG allow disconnecting the large EZ while sparing the functional areas and boundaries. It looks as an alternative to hemispherotomy in these particular children and could allow a better cognitive outcome. Sparing temporal and occipital structures may improve the quality of life in these children. SEEG, allowing delineation of the EZ, must be considered as a regular part of the pre operative investigations.

Keywords: stroke, stroke induced epilepsy, frameless SEEG, tailored disconnection, sus insular disconnection

FL-081

Epilepsy

The surgical treatment of hypothalamic hamartoma in children

Jie Ma¹; Xiaozheng Ling¹; Feili Liu¹.

¹Xin Hua Hospital Affiliated To Shanghai Jiaotong University School Of Medicine, Shanghai.

Objective: To investigate the clinical characteristics of children with hypothalamic hamartoma, operation treatment and gelastic epilepsy electrophysiological mechanism.

Methods: 32 patients were collected in pediatric department of neurosurgery in Shanghai Xinhua Hospital from 2007 August to 2014 June, which included 21 males and 11 females. Average age was about 35.56 months old. The imaging manifestations of peri-operation period, clinical features, video EEG changes and therapeutic effect of operation were analyzed retrospectively and the combination of peri-operation period of EEG and the lesion tissue electrophysiological analysis on gelastic epilepsy electrophysiological mechanism.

Results: 9 cases present with simple precocious puberty, 9 cases with precocious puberty and epilepsy, 11 cases with simple epilepsy, 3 cases with asymptomatic, 1 case with giant arachnoid cyst, 1 case with Stunting and 3 cases with refractory epilepsy. There are operation complete resection in 8 cases, subtotal resection in 11 cases, and partially resection in 10 cases. Intraventricular mirror disarticulation in 2 cases, λ knife treatment after operation in 2 cases, drug treatment in 1 case. Operation approach include: pterional approach in 21 cases, right forehead pterion combined approach in 6 cases, right orbitozygomatic approach in 1 case, anterior interhemispheric approach in 1 case. Combined with intraoperative cortical electroencephalogram EEG and deep, the localization of the epileptogenic foci, precise excision. Postoperative follow-up of patients shows good prognosis, no death and disability, epilepsy and EEG were improved in different degree. After 3 cases of lesion tissue for electrophysiological studies, revealed the presence of abnormal discharge.

Conclusion: children with hypothalamic hamartoma was mainly manifested in precocious puberty and gelastic epilepsy. In the intraoperative cortical EEG and deep electroencephalogram guided resection of epileptic foci is the treatment of hypothalamic hamartoma is reliable, safe method. The lesion tissue cells of the abnormal electrophysiological activity may be the cause in the etiology of epilepsy.

Keywords: hypothalamic hamartoma, operation treatment, epilepsy

Thursday, 8 October 2015
09:10 – 09:40

Technology Flash Presentations VIII

FL-082

Education and Training

Alder Hey Index: in-house development of a software for paediatric neurosurgeons

Ganesalingam Narenthiran¹; Sandra Maria Tsoti¹; Ajay Sinha²; Conor Mallucci².

¹Department Of Neurosurgery, The Royal London Hospital, London; ²Royal Liverpool Children's 'Alder Hey' Hospital, Liverpool.

Objective: Developing software can be expensive and challenging for neurosurgeons. The aim of the project was to develop a neurosurgery software to aid paediatric neurosurgeons at clinics and MDT meetings,

without the input of professional software developers. The project utilised one of the new generation of user-friendly software development environments.

Methods: The software “Alder Hey Index” was developed with a graphical user interface (GUI) based software development environment, “Xojo 2015” from Xojo Inc on, MacBook Pro (13-inch; Early 2011); Memory 16 Gb 1333 MHz DDR3; 2.3 GHz intel Core i5 running Ox X 10.9.5. The software was designed and developed by neurosurgeons without input from professional software engineers. The codes were compiled into executive programmes that would run on Windows, Mac and Linux machines. Twenty neurosurgeons had volunteered to evaluate the usefulness and intuitiveness of the software.

Results: The ‘Alder Hey Index, the software that was developed with Xojo 2015, successfully ran on Windows, Mac and Linux machines. The software could perform the following calculations: ventricular indices (single or comparison), tumour volume ((single or comparison), cranio-facial ratios (single or comparison), Liverpool craniopharyngioma score, Endoscopic third ventriculostomy Success Score, Chicago Chiari Outcome Scale, ICP monitoring: bolus injection study and corrected age of premature infants.

Conclusion: We were able to develop a software for paediatric neurosurgeons using Xojo without input from professional software developers. Xojo and similar GUI based new generation of software development environments could be used by neurosurgeons to develop bespoke software for their use.

Keywords: Software, development, training, educations

FL-083

Technology

Low versus high-field iMRI: which one is better?

Robert Keating¹; Tiffani Defreitas¹; John Myseros¹; Suresh Magge¹; Chima Oluigbo¹.

¹Childrens National Medical Center, Washington.

Objective: Considerable experience has been gained with the use of iMRI in the pediatric neurosurgical arena, utilizing both low and high field applications. Recently we have had the opportunity to compare both systems within a singular institution and offer insight into the strengths and weaknesses presented by each system.

Methods: Over 52 months at CNMC, 60 patients, underwent iMRI surgery utilizing a low-field (0.15T) magnet for treatment of 51 brain tumors, 6 spinal cord tumors, 3 with hydrocephalus and one AVM. During a subsequent 30 months, with a new high-field 1.5T iMRI, 53 pts were treated (31 brain tumors, 19 functional and one spinal cord lipoma). Efficacy, complications, costs and technical considerations were significantly different between the two systems.

Results: Sixty patients treated with the low-field iMRI (32 M/28F), 10.5y (0.1-36) demonstrated a 63% surgical efficacy. There were 5.2 scans/pt (3-14) with an average additional time commitment of 1.4 hrs/case. Transient complications were seen in 3/60 (5%) with no long-term issues. High-field iMRI was performed on 53 patients, 23M/30F, 12.1y (5.1-35.4) demonstrating a 28.3% efficacy with 5.5% transient complications. Average time of imaging was 25.75 min (19-37) with the number of scans/series being 7 (4-14) adding 1.15 hours per case. Not surprisingly, imaging number and time decreased with experience.

Conclusion: In addition to cost, there are many other advantages/disadvantages between low and high field iMRI. Ultimately, both systems perform well with meaningful efficacy and low morbidity. However, the greatest difference may be related to inherent technical challenges for the surgeon depending upon which system is used.

Keywords: intraoperative MRI, low field iMRI, high field iMRI

FL-084

Technology

Practical utilities and limitations of VITOM® exoscope in child’s spinal lipoma surgery

Kenichi Nishiyama¹; Junichi Yoshimura¹; Yukihiro Fujii¹.

¹Brain Research Institute, University of Niigata, Niigata.

Objective: An exoscope, high-definition video telescope operating monitor system to perform microsurgery has recently been proposed an alternative to the operating microscope (OM). The aim of this study is to know the utilities and limitations of exoscope in comparison with OM, and to consider further refinements regarding image-based microsurgery with usage of exoscope system.

Methods: Weve undergone spinal lipoma surgery for children with VITOM® exoscope system and evaluated its availability and limitation. In addition, the specification of VITOM® was compared with other exoscope systems.

Results: It was possible to perform pediatric lipoma surgery under 2-dimensional (2D) motion images provided by VITOM® with a visual perception through dynamic cue and stereoscopically viewing corresponding to the motion parallax. The working distance of VITOM®: 250-600mm which was comparable to OM, provided abundant space to insert and manipulate the instruments. VITOM® provided the ability to maintain depth of field (DOF) of 35mm under field of view of 50mm. It minimized the need for repositioning and refocusing during the procedure. On the other hand, limitation of magnifying object and lack of 3-dimensional (3D) viewing made the surgeon unpleasant and prolonged the operating time. According to the literature, other exoscope systems are also available to provide large DOF. It should be emphasized as a strong point of exoscope. VITOM® can provides high resolution motion images, however, it is 2D. As further refinement for the suitable operative scope, digital zooming system with high resolution and 3D imaging would be a welcome. Moreover, functional and navigational images should be integrated on demand during surgery.

Conclusion: Large DOF is the advantage of exoscope. 3D imaging and digital zooming mechanism greatly desired for better clinical application.

Keywords: microsurgery, exoscope, microscope, spinal lipoma

FL-085

Technology

Silver nanoparticle-Rifampicin impregnated shunt catheters in the prevention of microbial biofilm formation - an in vitro laboratory study

Dhanalakshmi Jeyachandran¹; Suhas Udayakumaran²; G. S Gowd¹; Vivek Vinod¹; Raja Biswas¹; Manzoor Koyakutty¹.

¹Amrita Centre for Nanosciences, Kochi; ²Division of Paediatric Neurosurgery, Amrita Institute of Medical Sciences and Research Centre, Kochi.

Objective: Our work involved use of a combination of Silver nanoparticles - Rifampicin (SNR) impregnated shunt catheters. Rifampicin is a well-accepted broad spectrum antibiotic with known usage for a similar purpose. Additionally, combining with silver nanoparticle, we hypothesize that the silver nanoparticles, known for its antibacterial effect can facilitate the action of the antibiotics by generation of reactive oxygen species which damages the cell membrane by lipid peroxidation and thus improve penetration. To study the efficacy of silver nanoparticle - Rifampicin impregnated shunt catheters in preventing bacterial shunt infections in comparison with pure silver nanoparticle tubes (PSN) and pure rifampicin impregnated tubes (PR) in terms of spectrum, duration of antimicrobial action and reduction in development of resistance.

Methods: Biofilm assays like roll plate technique, transfer agar plate technique where in PR, PSN and co-loaded tubes (SNR) where compared for their biofilm resistance. This was confirmed by confocal microscopy. Presently, we plan to quantify the biofilm formation at different time point with crystal violet assay after 48 hours and weekly for a month.

Results: In roll plate technique all three tubes were effective against the bacterial biofilm formation in 48 hours. In transfer agar plate technique the SNR tubes showed greater inhibition than the PR and PSN tubes for a period of 20 days. This was further confirmed by Confocal Microscopy where the SNR tubes showed negligible bacterial adhesion compared to PSN and PR tubes which showed a little biofilm formation after 48 hours of incubation with Green Fluorescent Protein- expressing Staph. aureus. Further thorough investigations to evaluate the potential of the catheter is underway.

Conclusion: Our initial results demonstrated significant reduction in the bacterial biofilm formation in SNR tubes compared to PR and PSN. The spectrum of efficacy and duration of action is a work in progress

Keywords: Hydrocephalus, Shunt infection, antibiotic impregnated shunts

FL-087

Hydrocephalus

Current trend in the use of image guidance during insertion of VP shunt, ETV and EVD

Ganesalingam Narenthiran¹; Frederick Boop².

¹Department of Neurosurgery, The Royal London Hospital, London;

²Department of Neurosurgery University of Tennessee Health Science Center, Memphis St Jude Children's Research Hospital, Memphis.

Objective: The aim of this study was to ascertain the current utilization of image guidance for ventriculostomy during insertion of VP shunt (VPS), endoscopic third ventriculostomy (ETV), external ventricular drain (EVD) in severe head injury (sHI) in paediatric patients.

Methods: A questionnaire on the use of image guidance (frameless stereotaxy, FS; ultrasound, US; endoscope) and guidance device (Ghajar Guide) in ventriculostomy VPS, ETV and EVD in sHI, was designed in Google Form. An invitation to complete the questionnaire was posted on the following electronic mailing lists: Neurosurgery Research Listserv, AANS-Pediatric Neurosurgery Listserv, British Neurosurgery Research Group ML, British Neurosurgery Trainee ML. The entered data was automatically collected in the spread-sheet within Google Form. This was downloaded and converted to FileMaker 13 database. Analysis of the data was performed using the database and Excel pivot table. Statistical tests were performed using XLStat and GraphPad 6.

Results: There were 56 responses. The commonest approaches for inserting VP shunts in descending order were: frontal (41%), parietal (32%) and occipital (25%). Of those responded, 16%, 20%, 11% and 9% reported that they respectively either use FS, ultrasound, endoscope or Ghajar guide routinely or would like use them routinely. Twenty-two percent of those responded reported that they used FS routinely for insertion of endoscope into frontal horn during ETV. Only 4% of those responded reported that they used FS routinely when they inserted EVD in patients with severe head injury; however, further 13% reported that they would like to use FS routinely for this.

Conclusion: The study illustrates the adoption of frameless stereotaxy for routine use in ventriculostomy in different clinical scenarios. We will also present our full results of the survey, with reference to a similar previous study presented at ISPN congress in 2008 in Cape Town.

Keywords: hydrocephalus, ultrasound, Stereotaxy, frameless, shunts

FL-088

Evidence-based practice

A randomized trial to improve fundoscopy - video diversion

Michael Yang¹; A. Ross Hengel¹; J. Michael Kerr¹; Ash Singhal¹.

¹University Of British Columbia and BC Children's Hospital, Vancouver.

Objective: Fundoscopy is an important component of the neurological examination, but can be challenging in uncooperative children. Inability to assess for papilledema can lead to delay in diagnosis, referral to specialized ophthalmology clinics, and increased cost and patient inconvenience. This study investigated whether playing a distractor video during eye examination, improves the success, duration, and ease of pediatric fundoscopy.

Methods: We completed a prospective, multi-disciplinary, block-randomized trial. Patients aged 1-4 years were recruited in the emergency department, neurology, spinal cord and general pediatric clinic. Patients were randomized according to the order eyes were examined and which eye video was played for; non-video eye served as an internal control. Successful exams were defined as visualizing the fundus within 60 seconds. Time to visualize optic disc was recorded and difficulty of exam was examined using a 10-point Likert scale, for both physicians and parents.

Results: 101 subjects were recruited, with a mean age of 2.8 years. Overall, there was a 20% absolute improvement in the success rate of visualizing the optic disc in the video versus non-video group ($p < 0.01$). Fundoscopy exams were significantly more successful while utilizing video distraction for children 1-2 years of age ($p < 0.01$). This trend was not noted in children 3-4 years of age. Time to visualize optic disc was also improved ($\Delta 5.3s$, $p < 0.01$). Improvements in ease of examination with video were noted by caregivers and practitioners for all age groups ($p < 0.01$). Video distraction was significantly more successful in all age groups within the neurology and spinal cord clinics.

Conclusion: Video improved the ease, duration and most importantly the success of fundoscopy in younger children. This simple, inexpensive adjunct has great potential to improve the ease and efficacy of this important aspect of the neurological examination.

Keywords: video distraction, fundoscopy, randomized control trial

Thursday, 8 October 2015

12:20 – 13:45

Craniofacial Flash Presentations IX

FL-089

Craniofacial

Implications of uncorrected sagittal synostosis

Kamilah Dowling¹; James T. Goodrich¹.

¹The Children's Hospital at Montefiore, Bronx

Objective: To give audience a clear understanding of why sagittal synostosis is no longer just an aesthetic issue.

Methods: A retrospective chart analysis was conducted to identify children with a diagnosis of sagittal synostosis. Ten children who range in age from twenty-three months to eight years of age were identified. In these patients, families initially declined surgical intervention -based on the premise that sagittal synostosis was primarily an aesthetic issue. Families later returned for further consultation to discuss surgical interventions due to headaches and psycho social issues.

Results: Results indicate that a subgroup of children with untreated sagittal synostosis display symptomatology warranting surgical intervention. In this series of eight children all underwent cranial vault remodeling and respond well with resolution of headaches and issues with increased intracranial pressure. There was also a significant reduction in the

psycho-social issues dealing the skull deformities. Untreated sagittal craniosynostosis can, in some cases, lead to inhibition of brain growth, psycho-social issues, and an increase in intracranial pressure due to the premature synostosis.

Conclusion: The criteria of whether to treat sagittal synostosis should be re-considered. There is a clear and real group of children, that left untreated who will represent in later childhood with psychosocial issues from peer taunting and also significant clinical issues that included severe headaches as a result of the premature suture fusion and increased intracranial pressures.

Keywords: sagittal

FL-090

Craniofacial

A dual institutional cohort study of mild trigonocephaly with clinical symptoms. (An interim report)

Kazuaki Shimoji¹; Takaoki Kimura²; Takeyoshi Shimoji³; Masakazu Miyajima¹; Hajime Arai¹.

¹Department of Neurosurgery, Juntendo University School of Medicine, Tokyo; ²Department of Neurosurgery Juntendo Shizuoka Hospital, Shizuoka; ³Department of Neurosurgery, Okinawa Prefectural Southern Medical Center / Children's Medical Center, Okinawa

Objective: It is considered that it is rare to develop increased intracranial pressure (ICP) and developmental delay in single suture synostosis. However, reports has been published that increased ICP and developmental delay are seen in some degree in these pathologies. We have conducted a dual institutional cohort study to understand the role of decompressive cranioplasty for mild metopic suture synostosis associated with clinical symptoms.

Methods: 30 cases were registered from 2012. We report the first 20 cases, which finished the protocol. Subjects were children in the rage from 2–4 years old who has mild trigonocephaly showing clinical symptoms. 5 neurophysiological tests including Developmental Quotient (DQ) and Pervasive Developmental Disorders Autism Society Japan Rating Scale (PARS) a moderate test based on Childhood Autism Rating Scale (CARS) have been conducted in 4 time points (3months before surgery, just before surgery, 3 months and 6 months post-op). The primary end point was configured as improvement of DQ in 6 months after surgery and the secondary end point was configured as the improvement of other tests. Increase of 5 point was considered as an improvement.

Results: Half of the patients showed increase of DQ. In these cases, 3 children had a decline of the scores of DQ during 3months before surgery and just before surgery and the scores increased after surgery. In PARS, improvement was seen in 75% of the patients. In these cases some of them have shown improved PARS score without DQ improvement.

Conclusion: At this point, improvement of DQ was seen in 50% of the patients. This is an encouraging data that this surgery is playing a role in some degree to avoid disadvantages from continuous insult of increased ICP. There are cases that shows improvement in other tests even not seeing improvement of DQ, so that further analysis is necessary.

Keywords: single suture synostosis, intracranial pressure, developmental delay

FL-091

Craniofacial

Comparison of two treatment protocols in children with scaphocephaly

Chris Parks¹; Joern Wittig²; Ben Robertson²; David Richardson²; Sasha Burn¹; Matt Bailey¹; Christian Duncan²; Ajay Sinha¹.

¹Neurosurgery Department, Alder Hey Children's Hospital, Liverpool; ²Craniofacial Department, Alder Hey Children's Hospital, Liverpool

Objective: Different techniques are currently offered to correct scaphocephaly. We compare two procedures used in Alder Hey Childrens Hospital in Liverpool (UK) in different age groups.

Methods: The notes of 105 children born between 2005 and 2010 treated for scaphocephaly at Alder Hey Childrens Hospital were reviewed. 50 patients operated before the age of seven months were treated by strip craniectomy and micro barrel staving (group A). 55 patients older than six months at the operation date were treated by cranial vault remodelling (group B). Compared outcome measures were cephalic index (CI), complications, dural tear rate, reoperation rate, transfusion frequency and amount, duration of the procedure, hospital admission time and speech and language assessments.

Results

There was no significant difference in the last measured CI as aesthetic outcome measure (mean of 77% in group A vs 76% in group B). The only complication in group A was a patient with wound infection. In group B one patient had ischemic brain injury, in one the procedure had to be abandoned because of bleeding. There were significantly more dural tears in group B (1 vs 9). In group A there were six reoperations, three for unsatisfactory head shape, one for raised intracranial pressure, one to cover a soft spot with a split calvarial bone graft, one for wound infection. In group B there was only one reoperation for unsatisfying head shape. In Group B the transfusion rate was significantly higher (50% vs 87%) so was the duration of the procedure (mean theatre time 145 vs 331 min). No significant difference in hospital admission time and speech and language assessments was found.

Conclusion: In scaphocephalic patients referred before the age of seven months, a smaller passive operation can achieve satisfying results with the benefit of a less invasive operation but a higher reoperation rate.

Keywords: scaphocephaly, strip craniectomy, vault remodelling, cephalic index, baby

FL-092

Craniofacial

Improving the aesthetic outcome in scaphocephaly correction: hairline lowering during vault remodelling procedures

Calisto Amedeo¹; Joern Wittig²; Ben Robertson²; Sasha Burn¹; Ajay Sinha¹; David Richardson²; Christian Duncan²; Chris Parks¹.

¹Neurosurgery Department, Alder Hey Children's Hospital, Liverpool; ²Craniofacial Department, Alder Hey Children's Hospital, Liverpool

Objective: Scaphocephaly often presents with frontal bossing. A neglected consequence of this morphological feature has been an impact on the height of the hairline with consequences for postoperative outcome.

Methods: Sixteen patients with scaphocephaly and having a high hairline due to frontal bossing who underwent scaphocephaly correction by subtotal or total vault remodeling were analyzed. The median age at surgery was 18 months. The mean distance between the nasofrontal suture and the hairline (hairline height) was preoperatively 70mm (range 58 to 91mm).

Results: An obvious lowering of the hairline could be achieved in all 16 patients. The mean postoperative hairline height was 59mm (range 50mm to 73mm). There were no complications associated with the technique.

Conclusion: The hairline lowering technique is a useful addition to vault remodeling techniques and can improve the postoperative aesthetic appearance considerably. The authors recommend this technique in scaphocephaly patients, who present with a high hairline.

Keywords: scaphocephaly, hairline, bossing, vault remodelling, nasofrontal suture

FL-093

Craniofacial

The benefits and limitations of the use of springs in the correction of scaphocephaly: a contemporaneous audit

Will Rodgers¹; Graham Glass¹; David Dunaway¹; Arpan Tahim¹; Allan Ponniah¹; Silvia Schievano²; Alessandro Borghi²; Frieda Angullia¹; Owase Jeelani¹.

¹Great Ormond Street Hospital for Children, London; ²Institute of Child Health, London

Objective: Spring assisted correction of sagittal synostosis is a novel alternative to formal calvarial remodelling. Benefits include technical simplicity, favourable morbidity profile and reduced hospital stay however asymmetric results, incomplete correction, need for a second operation and lack of long term results are potential drawbacks. We present the results of a single institutions experience over 4 years using a novel spring design. We assess the outcome and discuss the benefits and limitations of the use of spring assisted correction of scaphocephaly secondary to sagittal synostosis.

Methods: All patients treated at a single unit between April 2010 and Oct 2014 were evaluated using retrospective review of our contemporaneous database. Patients with multisutural synostosis and those operated over 7 months of age were analysed separately. Data were collected for operative time, anaesthetic time, hospital stay, transfusion requirements and complications.

Results: 81 patients were included. Cephalic index was 68 pre-operatively, 71 at 1 day post-operatively and 73 at 3 weeks and 6 months post-operatively. Seven patients required transfusion, 5 patients had post operative infection requiring early spring removal, 5 patients required traditional calvarial remodelling surgery for incomplete correction and 9 patients required burring down of ridges, 1 patient sustained a venous infarct.

Conclusion: Our modified spring design and protocol represented an effective strategy in the management of sagittal synostosis in children under 6 months old. Improvement in scaphocephaly was achieved in all patients although some required further procedures. The morbidity and mortality profile is favourably comparable to more traditional techniques. In patients referred within the first 6 months of birth, this technique has become our procedure of choice. In phenotypically severe cases and the older age group the correction may require further remodelling surgery.

Keywords: craniosynostosis, spring, cranioplasty

FL-094

Craniofacial

Biomechanics of spring cranioplasty: a population study

Alessandro Borghi¹; Silvia Schievano¹; Allan Ponniah²; Will Rodgers¹; Freida Angullia²; David Dunaway²; Owase Jeelani².

¹Institute of Child Health, London; ²Great Ormond Street Hospital, London

Objective: Spring cranioplasty, performed for the treatment of sagittal craniosynostosis, has proven successful in widening the skull, shortening operating time and lowering blood loss; however, concerns remain with regards to unpredictable spring behavior and variable outcomes. The relationship between the spring biomechanics and their effect on the paediatric calvarium is hereby analysed by means of numerical modeling and retrospective analysis of patient imaging.

Methods: Data from 26 patients (age 3–6 months, 2 springs per patient) who underwent implantation of spring distractors for treatment of sagittal craniosynostosis were analysed. Spring opening was measured in situ at

the time of insertion and removal and via planar x-ray during follow up post-implantation. Spring behavior in terms of opening vs force exerted in the interaction with the skull was derived by applying an exponential model to force decay.

Results: Springs were in place for 123.2 ± 44.8 days (range: 35–205). Spring opening varied from 3.0 ± 0.6 cm (force = 26.83 ± 10.61 N) at implantation to 5.4 ± 0.5 cm (force = 5.05 ± 4.71 N) at time of removal. The force decay time constant τ was found equal to 1.13 ± 0.41 days.

Conclusion: This study suggests that the force exerted by the current springs decays by 90% within the first 10 days of implantation and a residual force equal to 18% of the insertion force is present at removal time. This mathematical analysis will be performed on a larger number of patients to better understand the spring/skull interaction, potentially adapt the spring implantation follow up protocol, and drive design a more effective spring distractors.

Keywords: spring cranioplasty, modeling, biomechanics

FL-095

Craniofacial

Evolution of facial shape change during frontofacial distraction surgery

Freida Angullia¹; Alessandro Borghi²; Silvia Schievano²; N U Owase Jeelani¹; David J Dunaway¹.

¹Great Ormond Street Hospital for Children, London; ²Institute of Child Health, UCL, London

Objective: Frontofacial distraction surgery corrects mid-face retrusion associated with syndromic craniosynostosis. Unpredictable shape changes to facial anatomy during distraction challenge judging outcomes of surgery. Correlation of facial surface change to distance of distraction quantifies shape changes observed during the distraction process.

Methods: Serial 3D facial surface scans (Rodin4D) were performed on 7 patients undergoing frontofacial surgery followed by RED frame facial distraction. Scans were performed pre-operatively, before frame, during distraction and after frame removal. Mean differences of surfaces were measured between time points and correlated with bony distraction distance. Whole face, forehead-brow, malar, nasal and lower-facial regions were assessed.

Results: There was significant on-table advancement of 11mm for the forehead-brow region. Fronto-orbital level distraction yielded 1mm malar and nasal advancement, with 1–2mm relative reduction in exorbitism. Maxillary level distraction produced 4–5mm of malar and nasal advancement, with 0.5–1mm of exorbitism correction. A reduction in facial advancement occurred between the end of distraction to frame removal. The soft tissue to bony movement ratio is approximately 0.01 for forehead-brow, 0.06 for orbits, 0.24 for malar, 0.29 for nasal, and 0.03 for lower facial regions.

Conclusion: This study quantifies fronto-maxillary advancement and exorbitism correction in frontofacial distraction surgery for multisutural craniosynostosis. Most fronto-orbital advancement occurs at osteotomy release whilst maxillary advancement occurs more at post-operative distraction. The overlying soft tissues move differentially from underlying bone making distraction difficult to predict probably due to device strain and skin elasticity. The ratios provide a guide to quantify soft tissue outcome from bony distraction. The process, however, remains unpredictable and at this stage still requires iterative judgement for optimal surgical outcome. Over-distraction may be required to compensate for post-distraction retraction of tissues.

Keywords: Syndromic, Craniosynostosis, Frontofacial, Distraction, skin:bone ratio

FL-096

Craniofacial

Evaluation of intracranial volume after expansion of the posterior cranial vault distraction osteogenesis (PVDO) in craniosynostosis patients

Takaoki Kimura¹; Osamu Akiyama²; Kazuaki Shimoji²; Masakazu Miyajima²; Hajime Arai².

¹Juntendo University Shizuoka Hospital, Shizuoka; ²Department of Neurosurgery Juntendo University, Tokyo

Objective: Recently, there are reports that posterior cranial vault distraction osteogenesis (PVDO) is useful for expanding the posterior area of the skull to gain intracranial volume in craniosynostosis patients. This method seems that it could obtain larger volume compared with fronto-orbital advancement (FOA). As a neurosurgical point of view, expanding this volume is important to control intracranial pressure. In our current study, we evaluated the pre- and post-operative intracranial volume of the craniosynostosis patients who underwent PVDO.

Methods: 7 children were treated in Juntendo University Hospital from 2011 to 2014. All cases had the expansion of PVDO and underwent 3DCT scan at pre- and post-operation. Expansion was performed 1mm/day over 2–4 weeks and the cranium was distracted posteriorly from 20 to 30 mm.

Results: Three cases were boys and four cases were girls. Two cases were diagnosed pancraniosynostosis and the others were Saetre-Chotzen, frontonasal dysplasia, Apert Syndrome, plagiocephaly and trigonocephaly. They had operation at 5 to 39 month, the mean age was 23 month. The mean distraction length was 31.05 mm, and the mean expansion of volume was 192 cm³. The mean post-operative change rate of intracranial volume was 121%. This increase was close to the volume expansion curve of normal children.

Conclusion: In our study the expanded volume and rate was higher than FOA. The expansion of PVDO will allow to obtain intracranial volume and increasing intracranial volume first may exclude the obstacle for the developing brain in craniosynostosis children. The expansion of PVDO before anterior lesion helps us to focus on reconstructing anterior lesion more cosmetically without considering the expansion of intracranial volume by expanding anteriorly.

Keywords: Craniosynostosis, PVDO, intracranial volume

FL-097

Craniofacial

Evaluation of posterior cranial vault distraction from the 3D photogrammetric imaging

Niina Salokorpi¹; Ville Vuollo²; Tuula Savolainen²; Juha-Jaakko Sinikumpu¹; George Sándor¹; Pertti Pirttiniemi²; Willy Serlo¹.

¹Oulu Craniofacial Centre, University of Oulu, Oulu; ²Department of Orthodontics, Oral Health Sciences, Faculty of Medicine, University of Oulu, Oulu

Objective: Purpose of the study was to evaluate intracranial volume increase after posterior cranial vault distraction using 3D imaging and compare it with the previously used evaluation method.

Methods: We evaluated 4 consecutive patients that were operated due to craniosynostosis. Two patients were previously operated due to scafocephaly and later developed re-stenosis. One patient had bilateral nonsyndromic coronal craniosynostosis and the last patient had Crouzon syndrome. Mean age at operation was 35.8 months. Posterior vault distraction with 4 distractors was performed in all patients. All patients had their 3D photographs taken the day before operation. Second imaging was performed in average 4.8 months after primary operation. 3D images were taken in Natural Head Posture with the 3dDM system (3dMD Cranial, Atlanta, GA, USA). Intracranial volume increase was calculated

from pre- and postoperative cephalograms, by calculating of standard hemispheroid volume increase with linear distraction according to the formula presented earlier (Serlo et al. 2010). The 3D images were analyzed using commercial software Rapidform2006 (INUS Technology, Inc., Seoul, Korea). For the analysis postoperative 3D images were overlaid over the preoperative images by superimposing facial areas. Position of the head was standardized by the means of the craniofacial landmarks. The lowest cut-off point for volume change calculations was the highest point of auricular tip. The change in the individual head volume was calculated for each patient.

Results: The mean advancement was 27 mm (from 23 to 31 mm). The mean increase in volume calculated as a geometric prediction from cephalograms was 22.4 % (from 20.2 to 25.7%). Accordingly increase in volume calculated from the 3D imaging was 17.7% (from 14.5 to 23.2%).

Conclusion: 3D photogrammetric imaging is a useful tool in assessment results of the posterior cranial vault distraction regarding the achieved increase of intracranial volume.

Keywords: Craniosynostosis, Posterior Distraction, Intracranial volume, 3D imaging

FL-098

Craniofacial

The importance of virtual surgical planning and computer assisted surgical correction of hypertelorism

Jane Skjoeth-Rasmussen.Dk¹; Lars Boegeskov¹; Thomas Kofod².

¹Neurosurgical Department, Rigshospitalet, Copenhagen; ²Department of Oral and Maxillofacial Surgery, Centre of Head and Orthopaedics, Copenhagen

Objective: The presentation will discuss the introduction of virtual surgical planning in patients with Hypertelorism.

Methods: Three patients aged 20, 11 and 6 years old diagnosed with Hypertelorism. All patient underwent diagnostic simulation based on CT data in TruMatch. One patient refused osseous corrections and was therefore planned for correction based on a patient specific implant (PSI) (PEEK by Synthes). Two patients were planned for surgical correction. In both cases the surgical simulation included both facial bipartition and box osteotomies for the correction of hypertelorism. Based on the simulation final surgical plan was accepted and performed. Two patients underwent surgical simulation in TruMatch which was transferred to the theatre based on surgical guides and pre- and outcome stereolithographic models.

Results: The clinical results are presented including superimposed radiographical data. The different surgical techniques and the necessary workflow is presented showing highly improved visualisation prior to surgical intervention, the techniques implemented in VSP. The three-dimensional scans were shown to be highly accurate in predicting the degree of deformity. There was a significant difference in the preoperative and postoperative interdacryon distance and midface height. The simulation correlated significantly with the postoperative result when interdacryon distance and midface height were analyzed.

Conclusion: Virtual surgical planning (VSP) and additive manufacturing offers important new tools adding better visualization, diagnostics for planning of surgical correction of complex craniofacial anomalies such as hypertelorism (facial bipartition, box osteotomies) or improving outcome by the use of patient-specific implants (PSI). VSP will also give improved methods for 3-D postsurgical evaluation leading long term to improved care for patients with hypertelorism and other craniofacial anomalies.

Keywords: hypertelorism, Computer-Assisted-Surgery

FL-099

Craniofacial

Complications of craniostenosis surgery: an analysis of 117 cases

Chidambaram Balasubramaniam¹; Santosh Mohan Rao¹; K Subramaniam¹.

¹KKCTH, Chennai

Objective: To analyse the complications of surgery carried out for craniostenosis.

Methods: This is a retrospective chart review of 117 children who underwent surgery for craniostenosis. The nature of complications were analysed. The complications were divided into mortality and morbidity. The outcome after surgery will be analysed separately. The distribution of the cases were as follows: anterior plagiocephaly 65 scaphocephaly 13; oxycephaly 12; trigonocephaly 10; Cruzon syndrome 6; Apert syndrome 5; Pfeiffer syndrome 3 (re operated twice); plagiocephaly with scaphocephaly 2; Clover leaf deformity 1 (Operated twice).

Results: MORTALITY Two children died during surgery - one each due to massive bleeding and air embolism. One child died in the immediate post operative period due to disseminated intravascular coagulation (resulting in a massive epidural hematoma). Delayed death was seen in a child with Cruzon Syndrome due to Obstructive Sleep Apnoea Syndrome. MORBIDITY These were minor and major MINOR: he minor complications were penetration of the scalp by the wires used for fixation. The wires had to be removed. Seizures were seen in one case which responded well to anti convulsants Superficial scalp infection was seen in two cases MAJOR: Wound dehiscence was seen in a case of Clover Leaf Skull and required plastic surgical correction. One child developed serious sepsis and required prolonged antibiotic therapy and irrigation with antibiotics and povidone iodine solution Reoperation was required in three one for pseudomeningocele one each for growing fracture and unacceptable cranial deformity. Ocular complications in the form of massive conjunctival and corneal edema needing tarsorrhaphy twice was seen in one and one child in spite of tarsorrhaphy developed corneal ulceration and perforation.

Conclusion: The complications following craniostenosis surgery can be serious they are not significant usually. Over the years the complication rate in our centre has shown a downward trend.

Keywords: Craniostenosis, complications, mortality, growing-fracture

FL-100

Craniofacial

Anterior skull base and forehead deformities in children – surgical treatment and results

Michal Tichy¹; Jiri Kozak¹; Miroslav Vaculik¹; Juraj Schwab¹; Petr Liby¹; Hana Krasnicanova¹.

¹University Hospital Motol, Prague

Objective: Anterior skull base and forehead deformities may lead not only to functional difficulties, but also cosmetic and psychological aspects are important factors for both children and parents. The correction of deformity is achieved by various surgical modalities and many novel techniques were described in past years with favorable outcomes. The precise evaluation is often difficult, there are no quantitative criteria generally accepted and the length of follow up for some surgical methods is not sufficient enough.

Methods: 150 children with nonsyndromic synostoses involving anterior skull base, treated at our centre for craniofacial and reconstructive surgery. The age varied from 2 months to 4 years. The indication for surgery in all of the cases was both cosmetic and preventive. The surgical procedure was based on complete remodelling of the anterior skull in

majority of cases, with the cranioplasty and bone fixation using resorbable meshes and pins.

Results: Early postoperative complications were recorded. There was no infection, bleeding or CSF leak requiring surgical intervention. Local hematomas and subcutaneous swelling were only temporary and resolved either spontaneously, or were evacuated by needle puncture. There was no postoperative mortality. The cosmetic results were excellent immediately after the surgery. Reoperation rate was very low: in trigonocephaly group 1 out of 74, plagiocephaly group 0 out of 47 and in brachycephaly group 3 out of 29.

Conclusion: Contemporary surgical methods often use only limited osteotomies of frontal bone, open or endoscopic, followed by long time molding of calvarium with helmets. The pathology arise mostly from skull base. Our philosophy and surgical procedures are based on one step correction using osteotomy and reconstruction of anterior base including supraorbital bar. Our study showed that this type of extensive surgery can have favourable outcome with no mortality, low complication rate and mostly permanent cosmetic results.

Keywords: Deformities, Anterior skull base, Reconstruction, Remodelation

FL-101

Craniofacial

Receptive language performance and phonological skills in children with surgically treated nonsyndromic craniosynostosis

Jasmin Alian¹; Willy Serlo²; Seppo Laukka³; Matti Lehtihalmes¹.

¹Faculty of Humanities, Logopedics, University of Oulu, Oulu;

²Department of Paediatrics And Adolescence, Division of Pediatric Surgery, Oulu University Hospital, Oulu; ³Faculty of Education, Learning Research Laboratory (Learnlab), University of Oulu, Oulu

Objective: Nonsyndromic craniosynostosis (NSC) has been associated with an increased risk to deviant language development. Our purpose was to compare receptive language performance and phonological skills of children with surgically treated NSC with case-matched controls without craniosynostosis.

Methods: The current study population includes 29 children with NSC. Their age varied from three to nine years at the time for evaluation. The patients had undergone surgery for NSC at the age of 2-27 months (mean 8.7±6.2). Data concerning children's early development and medical history were collected from parents' questionnaires and clinical reports. Receptive language skills were assessed with the Reynell Developmental Language Scales III (RDLS-III) in children under the age of seven and with the Illinois Test of Psycholinguistic Abilities (ITPA) in older children. Eye tracking procedure was used to evaluate real time speech perception events. Phonological skills were measured by the Finnish Test of Phonology. Controls will be matched to each case individually in relation to chronological age and sex.

Results: According to parents' questionnaires most of the children managed well in everyday life, one child has had slightly delayed early language development and four children had received speech therapy mainly for phonological difficulties. However, six of 21 children assessed with RDLS-III had receptive language scores lower than the normal range. Five of six children scored poorly in RDLS-III had birth weights (BW) <3000 g whereas almost all of the children scored within the normal range had BW >3000 g. Statistically significant correlation (p=0.015) was found. All eight children assessed with ITPA scored within the normal range. Results from other assessments as well as results from a control data group were also analyzed.

Conclusion: Children with NSC may experience difficulties in speech perception and phonological skills. Low BW may be considered as a predictive risk factor.

Keywords: nonsyndromic craniosynostosis, speech perception, phonological skills, eye tracking

FL-102

Craniofacial

Long-term surgical results of non-syndromic craniosynostosis

Mustafa Onoz¹; Gokalp Silav¹; Fatih Han Bolukbasi¹; Recep Basaran²; Nejat Isik²; Ilhan Elmaci¹.

¹Memorial Health Group, Department Of Neurosurgery, Istanbul; ²Medeniyet University Goztepe Education And Research Hospital, Department Of Neurosurgery, Istanbul

Objective: Craniosynostosis refers to early fusion of one or more cranial sutures. Often it leads to abnormal head shape. The aim of this study was to evaluate long-term surgical results as radiological and clinical of non-syndromic craniosynostosis treated in our clinics.

Methods: 70 patients who were operated in our clinics with diagnosis of craniosynostosis between 1995 and 2015. Birth date of the patient, maternal age, age at surgery, gender, duration of follow-up, type of craniosynostosis, complaints, neurological examination at admission, surgical technique, preoperative and postoperative radiologic imaging were recorded.

Results: 21 female, 49 male patients were included to the study. The mean age at surgery was 9,42 months. The mean age of mothers was 24,98 years. Follow-up was ranged between 1 and 20 years, mean follow-up was 9,05 years. According to types of craniosynostosis there were 27 (38,5%) of trigonocephaly, 21 (30%) of scaphocephaly, 2 (2,8%) of plagiocephaly, 2 (2,8%) of brachicephaly. Also, 18 (25,7%) of multiple synostosis were found. Long-term radiological results showed that craniofacial remodeling with pushing forward of orbital bone had good outcome of expanding of brain parenchyma and cosmetics.

Conclusion: surgical techniques must be chosen according to cranium features of patients. Resynostosis is one of the major problems of this pathology. Cranial remodeling with pushing forward of orbital bone had good outcome of expanding of brain parenchyma and cosmetics and low rate of resynostosis.

Keywords: craniosynostosis, surgery, remodeling, follow-up

FL-103

Oncology

Primary skull lesions in pediatric population: KFSH experience

Anwar Ul Haq¹; Essam Alshail¹; Hindi Al Hindi¹; Anas Dababo¹; Amane Al Kofide¹.

¹King Faisal Specialist Hospital and Research Centre, Riyadh, Riyadh

Objective: Primary skull lesions are rare in pediatric patients. Differential Diagnosis of these lesions is broad and includes congenital, inflammatory, traumatic, vascular and neoplastic lesions. To present a review of clinicopathological features, surgical management and outcome of primary skull lesions in pediatric patients at a tertiary care hospital.

Methods: It is retrospective review of clinic pathological features, diagnosis, management and outcome of primary skull lesions in children carried out at Department of Neurosurgery, King Faisal Specialist Hospital and Research Centre, Riyadh, over past 5 years.

Results: We identified 25 cases of primary skull lesions in children. Age range was 1 month-14 years. Majority of the lesions were benign. The most common presenting symptom was painless mass. The lesions identified included Dermoid/epidermoid cyst, Encaphalocoele, Leptomeningal cyst, Infantile hemangioma, Infantile myofibroma, Cavernoma, Neurofibroma, Osteoid osteoma, Osteoblastoma, Fibrous Dysplasia,

Benign Ossifying fibroma, Langerhans cells histiocytosis, Ewing Sarcoma and Rhabdomyosarcoma. Majority of lesions underwent gross total resection and primary reconstruction of skull defect. Preoperative angiography and embolization was carried out in two patients because of high vascularity of tumor to decrease intraoperative blood loss. Clinical outcome was excellent and recurrence was rare in benign lesions.

Conclusion: Majority of the primary skull lesions in children are benign. Gross total resection is the treatment of choice and recurrence is uncommon after gross total resection.

Keywords: Primary skull tumors, Fibrous dysplasia, Ewing sarcoma

FL-104

Craniofacial

Osteolytic lesions of skull: differentiation of eosinophilic granuloma and fibrous dysplasia

Recep Basaran¹; Mustafa Onoz²; Fatih Han Bolukbasi¹; Mustafa Efendioğlu¹; Basak Caner¹; Nejat Isik¹; Ilhan Elmaci².

¹Medeniyet University Goztepe Education and Research Hospital, Department of Neurosurgery, Istanbul; ²Memorial Health Group, Department of Neurosurgery, Istanbul

Objective: Eosinophilic granuloma (EG) is a variety of histiocytosis-X with unknown etiology. EG is characterized by single or multiple skeletal lesions, and predominantly affects children, adolescents, and young adults. One of the more common sites is the skull. Symptoms include localized pain, tenderness and swelling. Another osteolytic lesion of skull similar to EG is fibrous dysplasia (FD). FD is characterized by slow, progressive replacement of a localized area of bone by an abnormal proliferation of isomorphic fibrous tissue. Especially, cystic subtype is similar to EG. We aimed to differentiate two osteolytic skull lesions (OSL) according to radiological and histopathological features.

Methods: We evaluated patients who operated with complaint of OSL. We noted sex, age at operation, neurological examination, localization of lesion, radiological and histopathological features of lesions.

Results: A total of 6 patients were identified with EG and FD. Average age at operation was 9,16 years (range, 4-15 years). There were 3 females and 3 males. Four patient were classified as monostotic cystic FD and 2 as EG. Anatomically more involved areas of skull were frontal (50%) and parietal (33,3%) bones. All had normal neurological examination. Radiological features were similar. In EG, A well-defined lytic lesion was seen in skull bones. It involved both the inner and outer table. In FD, ground-glass opacities, cystic and well-defined borders, expansion of the bone, with intact overlying bone were seen. Histopathologically, there were many Langerhans cells, eosinophils, neutrophils and lymphocytes in EG. In FD, there was large fibrous matrix with scattered curvilinear trabeculae of woven bone with no surrounding osteoblastic rimming.

Conclusion: EG and FD are similar lesion according to clinical presentation and radiological features. Both of them are benign pathologies affecting skull bones. But they have different histopathological features.

Keywords: eosinophilic granuloma, fibrous dysplasia, radiology, histopathology, osteolytic lesion

Poster Presentations

P-001

Oncology

Langerhans cell histiocytosis (LGH) of the sacrum: report of two cases

¹Mohammad Alfawareh; Anwar Mohammad¹.

¹Spine Department, King Fahad Medical City, Riyadh

Objective: To report two cases of isolated LCH sacral lesion in pediatric patients who managed conservatively.

Methods: Introduction: Langerhans cell histiocytosis (LCH), also called eosinophilic granuloma of bone, is a relatively rare disorder of unknown etiology, probably arising from circulating myeloid dendritic cells. It is most common in children 5 to 10 years of age. LCH can involve any of body tissues. The occurrence of LCH in the sacrum is extremely rare. Purpose: To report two cases of isolated LCH sacral lesion in pediatric patients who managed conservatively. Materials and methods: Two cases review.

Results: Two cases of sacral LGH were reported, diagnosed by biopsy, managed conservatively, and doing well at the final follow up.

Conclusion: Both of them are less than 5 years old, both were managed conservatively. LCH location and number of lesions determine its mortality and morbidity potentials.

Keywords: Histiocytosis, LHG, pediatric tumor

P-002

Oncology

Endoscopic ultrasonic aspiration for cystic intraventricular craniopharyngioma

Oscar García-González¹; Paola Hernandez-Ponce².

¹Hospital Regional De Alta Especialidad Del Bajío, León; ²Universidad Autonoma De Guadalajara, Guadalajara

Objective: We report our experience in purely endoscopic excision of intraventricular craniopharyngiomas in pediatric patients with the minimally invasive ultrasonic aspirator Soring-Sonoca /GAAB neuroendoscope system.

Methods: Five patients with intraventricular craniopharyngioma were operated by a purely endoscopic approach using a GAAB rigid endoscope and the Söring-Sonoca ultrasonic aspiration system.

Results: One patient was operated 3 times and another patient two times and also an Ommaya reservoir was placed into de cystic remaining cavity as well in the another five patients. The tumor size varied between 1.9 and 5.5 cm in the largest diameter. The cystic content of the tumor could be aspirated completely in all five patients. There were no long-term complications.

Conclusion: Ultrasonic aspiration can be applied safely and successfully in selected endoscopic procedures. The use of this technique could expand the indications for endoscopic approaches to include also solid intraventricular lesions. The Söring-Sonoca ultrasonic aspirator is a highly effective tumor decompression system that can be effectively used in a purely endoscopic approach to intraventricular craniopharyngiomas.

Keywords: Craniopharyngioma, Neuroendoscopy, Ultrasonic aspiration

P-003

Oncology

Choroid plexus papilloma in children: an experience with 15 cases

Amir Abbas Ghasemi¹; Soudabeh Ashoori¹.

¹Urmia University Of Medical Sciences, Urmia

Objective: Choroid plexus papillomas (CPP) are rare epithelial brain tumors that knowledge regarding their behavior and treatment is limited and challenging. In this study we reviewed our experience with 15 cases of CPPs in Children.

Methods: In this retrospective study we analyzed 15 cases of CPP operated at our institution since December 2001. The variables included in the

study were as follows: clinical presentation, age, sex, imaging characteristics, degree of resection and outcome. Glasgow outcome scale was used to assess outcome.

Results: Among 15 cases of study group; 10 (67%) cases were male and 5(33%) cases were female. The mean age at presentation was 3.5 ± 2.7 years (2 months to 12 years). The most common presentation was raised intracranial pressure (91%). Locations of the tumors were as follows: lateral ventricle (73%), 4th ventricle (13%), 3rd ventricle (7%) and CP angle (7%). Tumors appearance in plain CT.Scan were as: hyperdense in 8, isodense in 6 and hypodense in 1. All the tumors were hypointense in T1W images and hyperintense in T2W images. All the lesions were uniformly enhanced both on CT. Scan and MRI.complete excision was performed in all cases. Postoperative complications included: pneumocephalus (53%), subdural effusion (33%), neurologic deficit (13%). 3 (20%) cases required shunt placement. The mean follow up period was 6.4 years (1 to 11 years). all the patients were completely well at final follow up (Glasgow outcome scale of 5).

Conclusion: CPP are rare intracranial tumors that complete surgical resection is usually achievable with the least possible complications and offers an excellent prognosis.

Keywords: papilloma, choroid plexus, brain tumor

P-004

Oncology

Mixed germ cell tumors in septum pellucidum after radiochemotherapy of suprasellar germinoma: de novo metachronous or recurrent neoplasms?

Ki Seong Eom¹.

¹Wonkwang University Hospital, Iksan

Objective: Mixed germ cell tumors (GCTs) consisting of a germinoma and a mature teratoma in the septum pellucidum have never been described previously; the patient we present here is the first reported example.

Methods: This case involves an 18-year-old man who presented with abnormal behavior, impairment in recent memory, and emotional change. Five years ago, he received five cycles of chemotherapy using cisplatin and ectoposide and 24G of local radiotherapy for clinical diagnosis of suprasellar germinoma in another hospital. The tumor was then completely resected. Magnetic resonance imaging in our hospital revealed a large fatty mass located primarily in the septum pellucidum and some portions of the corpus callosum; a heterogeneous enhancing tumor was observed in the surrounding area. The second tumor was completely removed. The histological diagnosis was mixed GCTs containing the component of a germinoma and a mature teratoma.

Results: This report is worthwhile, considering previous reports in relation to our case-even though the first tumor was not histologically diagnosed. This is the first case of mixed GCTs in septum pellucidum consisting of a germinoma and a mature teratoma. Neurosurgeons should always pay attention to the pathological variety and dynamic pathological changes in GCTs once they start to examine and treat them.

Conclusion: This case is characterized by a second GCT occurring at a different site and with a different histological type, long after complete resolution of suprasellar germinoma. Although it is very difficult to draw conclusions about the correlation between the first and second GCTs, neurosurgeons should always pay attention to the pathological variety and dynamic pathological changes in GCTs once they start to examine and treat them.

Keywords: Germcell tumor, Germinoma, Teratoma, Septum pellucidum, Metasynchronous

P-005

Oncology

Isolated skull metastasis of ewing's sarcoma in a child

Patricia Puerta¹; Antonio Guillén¹; Jaume Mora²; Mariona Suñol³; Santiago Candela¹; Enrique Ferrer¹.

¹Department of Pediatric Neurosurgery. Hospital Sant Joan De Déu, Barcelona; ²Department of Oncology. Hospital Sant Joan De Déu, Barcelona; ³Department of Pathology. Hospital Sant Joan De Déu, Barcelona

Objective: Ewings sarcoma (ES) was described as a “diffuse hemangioendothelioma of bone”. It affects long bones and pelvis. The predominant sites of metastases include lung, bone and bone marrow. Metastasis of ES to the central nervous system (CNS) is uncommon. We present an extremely rare case of skull metastasis of ES.

Methods: We present a child affected of a metastatic ES of the skull and we describe our management based on a literature review through Pubmed Medline.

Results: A 10-year-old male underwent resection of an ES in the left 5th rib 15 months previously, followed by chemotherapy and radiotherapy. He had a painless, progressive swelling over his right parietal area. CT showed an isodense parietal tumour which was homogeneously enhanced by contrast medium. It was partly extracranial and partly intracranial, with bone destruction. Gross total removal of the tumour was performed and the infiltrated dura was excised. We performed a duraplasty and a titanium mesh cranioplasty. The histology was identical to the specimens from the left rib tumour. The diagnosis was skull metastasis of ES. Adjuvant treatment was performed.

Conclusion: ES is the second most common malignant bone tumour. It usually occurs in patients younger than 20 years of age and it's more frequent in males than females. Primary ES most often originates in long bone shafts, pelvic bones, ribs and vertebrae. This tumour frequently develops metastases. The principal sites of metastases are lung, bone and the bone marrow. Meningeal invasion and spinal cord compression are the best known forms of CNS involvement. The frequency of skull metastases of ES in children is unknown but, to our knowledge, there are only three cases previously reported. The multimodal treatment for ES is considered to be the optimal treatment method. Solitary skull metastasis should be removed totally, followed by systemic chemotherapy.

Keywords: Ewing's sarcoma, Skull metastasis, CNS involvement

P-007

Oncology

Establishment of a rapid and efficient genetic analysis system of pediatric glioma

Jun-Ichi Adachi¹; Kozue Watanabe¹; Tomonari Suzuki¹; Joji Ishida¹; Kazuhiko Mishima¹; Ryo Nishikawa¹.

¹Department of Neurosurgery/Neurooncology, Saitama Medical University International Medical Center, Hidaka

Objective: Of the genetic mutations seen in pediatric gliomas, IDH1/2 mutations, chromosome 1p/19q loss, MGMT promoter methylation, and mutations of TERT (telomerase reverse transcriptase), BRAF, and H3F3A, have become increasingly important as biomarkers for the diagnosis, prognosis, and treatment response. Here we report on the establishment of a system, within our institution, of the efficient and rapid detection of these genetic mutations.

Methods: Tumor-derived DNA and RNA extracted from either frozen or paraffin-embedded histological sections were used as specimens. For the point mutation assay, the following mutations of: IDH1/2, H3F3A, BRAF codon 600, and the TERT promoter were screened using High Resolution Melting (HRM) method, and

identified by DNA sequencing. For the MGMT promoter methylation, a quantitative analysis was performed using the methylation-specific HRM method. Real-time PCR was used for determining the presence or absence of the BRAF-KIAA fusion gene, and FISH was used for detecting chromosome 1p/19q loss. Further, immunostaining was performed to analyze mutant TP53, high expression levels of EGFR and loss of ATRX (alpha-thalassemia/mental-retardation syndrome-X-linked) expression.

Results: Screening of each of the genetic mutations by the point mutation assay using the HRM method was completed in one run of approximately 70 minutes. Furthermore, all genetic mutation results were obtained within 5~72 hours after nucleic acid extraction using our assay system.

Conclusion: For clinics that specialize in diagnosis and treatment of pediatric brain tumors, the genetic biomarker information mentioned above will become indispensable in the future. Outsourcing this testing has issues relating to cost and time to obtaining results, and thus testing in-house is desirable. Utilizing PCR primer settings that allow the screening of multiple genetic mutations in one run, and simple and sensitive/specific assays such as the HRM method, allows efficient and rapid analysis.

Keywords: glioma, biomarker, pediatric

P-008

Oncology

Pilomyxoid astrocytoma presenting with central precocious puberty
Ruslan Yunusov¹; Şahin Hanalioğlu¹; Kader Karlı Oğuz²; Figen Söylemezoğlu³; Burçak Bilginer¹.

¹Hacettepe University Faculty of Medicine, Department of Neurosurgery, Ankara; ²Hacettepe University Faculty of Medicine, Department of Radiology, Ankara; ³Hacettepe University Faculty of Medicine, Department of Pathology, Ankara

Objective: Pilomyxoid astrocytoma (PMA) is a rare form of glioma seen usually in children and is regarded as a variant of pilocytic astrocytoma. Pilomyxoid astrocytoma is characterized by monotonous, bipolar spindle cells with an angiocentric arrangement within myxoid background in contrast to typical biphasic histology of pilocytic astrocytoma. The most common location of PMA is hypothalamus and optic chiasm.

Methods: Here, we report an 8-year old girl with a diagnosis of PMA.

Results: An 8-year old girl presented to our clinic with visual disturbance and precocious puberty for one year. Cranial MRI showed a 4x4x5 cm suprasellar mass. The tumor involved optic chiasm and right optic nerve extending to right middle cranial fossa and rhomboid fossa. The patient underwent subtotal excision via anterior interhemispheric transcallosal approach. Histopathological examination revealed a neoplasm with medium cellularity in a loose myxoid background. Neoplastic cells had oval nuclei with open chromatin and narrow spindle-shaped cytoplasm showing perivascular arrangement. Rosenthal fibrils were absent. Immunohistochemistry studies showed GFAP positivity and IDH-1 negativity in neoplastic cells. Ki-67 proliferation index was below 1%. These findings suggested the diagnosis of pilomyxoid astrocytoma.

Conclusion: Pilomyxoid astrocytoma was defined as a subgroup of pilocytic astrocytoma in 1999. It usually arises in hypothalamus and optic chiasm during infancy or childhood. Mean age at presentation varies between 10 and 18 months. Pilomyxoid astrocytoma shows unique histological characteristics distinct from pilocytic astrocytoma. The most important histological feature of PMA is monotonous bipolar spindle cells with angiocentric

arrangement in myxoid background. Their natural history is more aggressive than pilocytic astrocytoma. Local recurrence rates vary between 55% and 76% after the surgery. 11% to 14% of PMA patients had dissemination via cerebrospinal fluid. Despite no consensus regarding the optimal treatment, radiotherapy following surgery is usually preferred to control the residual disease.

Keywords: Pilocytic astrocytoma, Pilocytic astrocytoma, Precocious puberty

P-009

Oncology

Neuro oncology at a large regional paediatric hospital: an analysis of practice in 2014

Catherine Pringle¹; Ian Kamaly-Asl¹; John-Paul Kilday¹.

¹Royal Manchester Children's Hospital, Manchester

Objective: Analysis of neuro-oncology practice in 2014 at a large regional children's hospital, aiming to identify areas of practice which could be improved.

Methods: Retrospective analysis of case notes, electronic patient records, imaging and histology of all patients presenting to the paediatric neuro oncology services in 2014.

Results: 28 patients presented to Neuro oncology services at our unit in 2014. Of these, 26 were new referrals and 2 patients previously known to us. 13 patients presented directly to our unit, 14 patients were referred from hospitals within our catchment area and one patient was referred from outside the United Kingdom. Average age was 9.1 years (range 2-16 years). Average duration of symptoms prior to presentation was 74 days (range 0-365 days). Headache and seizure were the most frequent presenting symptom (8 and 7 cases respectively). 29 tumour procedures were carried out in total and 2 patients were non-operatively managed. 3 patients underwent emergency tumour surgery (<24 hours), 20 patients were operated on within a week of presentation, and 3 patients had elective surgery. 3 patients had redo surgery on the same tumour within the 2014 period. Post-operative MRI scans were carried out within 48 hours in 19/26 patients. 15 additional procedures were carried out for CSF diversion and wound management. Average length of stay was 15.7 days (range 2-57 days). Post-operative complications included pseudomeningoceles (2) and hydrocephalus requiring CSF diversion (4).

Conclusion: We are a busy paediatric neuro oncology centre, providing high level of care for our patients. We aim to improve our practice through prospective morbidity and mortality and tumour resection data collection. Public awareness of symptoms of brain tumours in young children also needs to be addressed to reduce time to diagnosis.

Keywords: neuro oncology, paediatrics, United Kingdom

P-010

Oncology

Novel therapeutic target for medulloblastoma therapy: using mtor related signaling pathways.

Paul Lee¹; Arthur Wang¹; Avinash Mohan¹; Michael Tobias¹; Raj Murali¹; Meena Jhanwar-Uniyal¹.

¹New York Medical College, Valhalla

Objective: Medulloblastoma (MB) is the most common malignant brain tumor in children. Recent genetic analyses have suggested that MB comprises of four distinct molecular variants: WNT,

SHH, Group 3, and Group 4. The mTOR is a central regulator of metabolism in malignant cells. HIPPO pathway intersects with PI3K and SHH pathways to modulate mTOR activity, an important regulator in cell growth and dissemination. We utilized HIPPO and mTOR pathways and used arsenic trioxide (ATO), which interacts with both mTOR and SHH pathways for treatment of MB.

Methods: Immunohistochemistry analysis was done in MB tumors (achieved under approved IRB) to establish the activation of AKT/mTOR and HIPPO pathways. Functional analysis was performed for cell migration, cell cycle and proliferation in MB cell following treatment with inhibitors and ATO and PI3K/mTOR inhibitors. YAP siRNA treatment was used to inhibit HIPPO pathway. YAP-regulated miRNA-29, which mediates mTOR by modulating PTEN, was investigated.

Results: We observed a correlation between the expression of activated pAKT and pmTOR and YAP (the downstream effector of HIPPO pathway). Treatment with inhibitors of PI3K/AKT (LY294002) and mTOR (rapamycin), given with EGF or SHH inhibitor Cyclopamine or ATO reduced cellular motility. Inhibition of HIPPO pathway suppressed cell proliferation as established by using 3 unique 27mer duplexes of YAP1 siRNA. Combined inhibition of SHH with either LY294002 or rapamycin reduced cell proliferation. PI3K inhibition caused translocation of YAP to nucleus as demonstrated by immunofluorescence.

Conclusion: Inhibiting these key pathways with molecularly targeted therapies represents an important approach to improving MB outcomes. We demonstrated that combined targeting of mTOR/HIPPO pathway with SHH provides an alternative strategy for treatment of MB.

Keywords: SHH, mTOR, PI-3K, Arsenic Trioxide

P-012

Oncology

From the globe to the chiasm - combined intraorbital and intracranial resection of optic gliomas in children with NF1

¹Schuhmann Martin; Spiriev Toma¹; Besch Dorothea¹.

¹Eberhard Karls University of Tübingen, Tübingen

Objective: Optic gliomas are for the vast majority of cases no surgical tumors in NF1 due to their diffuse nature and involvement of optic nerve, chiasm and optic tract/hypothalamus. An exception are unilateral intraorbital to pre-chiasmatic tumors in case of blindness associated with increasing exophthalmus and orbital pain.

Methods: We report on two girls aged 3 years 1 month and 4 years 11 months with unilateral large optic gliomas extending from the globe of the eye almost to the chiasm. Both exhibited the combination of blindness, increasing exophthalmus and in one case excruciating pain.

Results: In both girls an extradural approach to the orbit was performed. The superior orbital fissure and the optic canal were extradurally exposed. The intraorbital tumor was resected, followed by intradural resection of the remaining tumor. The optic canal was sealed. The orbital and its roof were fully reconstructed. Pain and exophthalmus were relieved. In both children the ophthalmic vessels and the function of all orbital muscles was preserved with normal ocular movements after a recovery period. One girl developed after 18 months a recurrence at the exit site of the optic nerve from the globe. A spread of the disease to the chiasma was prevented.

Conclusion: A combined microsurgical intra- and extradural approach to unilateral optic nerve gliomas extending from the globe to the chiasm is possible when the combination of blindness, exophthalmus and pain indicates treatment. This can be achieved with preservation of all orbital function present at time of surgery and relieves exophthalmus and pain. Alternative treatment options would have been chemotherapy over 12-18 months which

does not address exophthalmus and pain directly. Since resection at the optic nerve exit site of the globe has to be conservative to preserve the integrity of the eye ball, a local recurrence at this transition zone is possible

Keywords: optic nerve glioma, skull base surgery, neurofibromatosis type 1

P-013

Oncology

Cystic craniopharyngioma in a 12 year old girl.

Nurullah Yuceer¹; Gonul Guvenç¹; Ceren Kızmazoglu¹; Ilker Cingoz¹; Gokhan Gurkan¹

¹Izmir Katip Celebi University, Izmir

Objective: Craniopharyngiomas are intra-cranial tumours, relatively frequent in children. The aim of this study is to analyze the clinical, radiological and operative features of cystic craniopharyngioma in a pediatric case.

Methods: A 12-year-old girl who presented with headache, nausea and vomiting. A CT scan and MRI that disclosed a solid plus cystic lesion associated with calcification extending to third ventricle, anterior cranial fossa and middle cranial fossa from suprasellar area. Neurological examination demonstrated optic atrophy in the left side, and papill edema in the right side. There was vision loss in the left side. The patient underwent a gross total tumour excision by left pterional craniotomy plus subfrontal approach. Postoperative stage was uneventful. Pathological examination confirmed a craniopharyngioma.

Results: Cystic changes and calcification are the pathological features of pediatric craniopharyngiomas.

Conclusion: Surgical removal of cystic plus solid craniopharyngiomas in children is associated with significant operative morbidity and recurrence rates.

Keywords: craniopharyngioma, child, magnetic resonance imaging, computerized tomography

P-014

Oncology

Pilomyxoid astrocytoma; case report

Utkan Topçu¹; Emre Özkara¹; Zühtü Özbek¹; Funda Canaz²; Murat Vural¹; Ali Arslantaş¹

¹Eskişehir Osmangazi University Neurosurgery Department, Eskişehir;

²Eskişehir Osmangazi University Pathology Department, Eskişehir

Objective: Pilomyxoid astrocytoma (PMA) is a recently defined pediatric brain tumor; PMAs were previously classified within the pilocyticastrocytoma (PA) category. Nevertheless, PMA has different histological features and has been shown to behave more aggressively than PA.

Methods: A 15-year-old girl presented with epileptic seizures and headaches for one month. On neurological examination she had left hemi paralysis (4/5). Magnetic resonance imaging (MRI) indicated a well-defined, cystic tumor 4*5*5 cm size associated with a small nodule in the right talamic region (fig 1, fig 2). The midline shift was also noted. She underwent nearly totally resection of the tumor.

Results: Histo-pathological pattern of removed tumoral mass was in a myxoid matrix and some areas anjio-centric monomorphous tumour cells. Tumour cells are having elongated and fibrillar roces, mild nuclear pleomorphism. There was seen any Rosenthal fibers and eosinophilic granular bodies. Some focal areas; necrosis and glomeruloid artery structure was seen. Immunohistochemical study shown that S100 and GFAP (+), EMA and P53 focal weak positive, proliferation index of Ki-67 was %1-2. Patient diagnosis was reported Pilomyxoidastrocytoma. PMA is a subtype of PA, which has been included as grade 2 astrocytoma in WHO tumor classification system. Studies reports that PMA has more aggressive

nature when compared with typical PA. Therefore, its necessary to be differentiated other low grade gliomas.

Conclusion: Pilomyxoid astrocytoma is a recently defined pediatric brain tumor. And is a grade 2 brain tumours. This tumours different from Pilocytic astrocytomas at histological features and clinical course.

Keywords: pilomyxoid, astrocytomas, tumor, pathology, pediatric

P-015

Oncology

Efficiency of 5-aminolevulinic acid fluorescence guidance in the treatment of children with recurrent neuroepithelial tumor

Aleksandr Kim¹; William Khachatryan¹

¹Polenov Neurosurgical Institute - Federal North-West Medical Research Centre, St.Petersburg

Objective: Evaluation of fluorescence-guided resection of pediatric recurrent neuroepithelial tumors.

Methods: 24 children aged from 3 to 18 were retrospectively analyzed. Comparison of the results of two fluorescence guidance resections were made in three patients twice operated during the period in question. For 5-ALA administration the Alasens preparation was used (20 mg per kg body weight). The solution was ingested 4 hours before the planned blastomatous tissue resection. The Alasens preparation was used only in the absence of contraindications, with the consent from the parents and approval of the Neurosurgery Institute Committee on Ethics. All the patients underwent, pre- and post-operationally, a complete clinical MRI examination, with a histological re-examination of the tumor.

Results: Operations were repeated during the period of from 3 months to 12 years. Fluorescence was absent in four in cases (two of those, with neuroepithelial tumor Gr. I, 1 – Gr. III, 1 – Gr. IV); and was negligible in seven cases (4 – Gr. I, 2 – Gr. II, 1 – Gr. IV). In the other 13 cases, the use of fluorescence guidance increased the reliability of verification of residual blastomatous tissue, and provided for the latter's differentiation from postradiation and cicatricial changes. Gross-total or near-total tumor removal was achieved in 19 (79.2%) out of 24 cases, with 92,3% in the group with efficient fluorescence. No clinically relevant side effects were recorded in the patients involved. Three patients showed transitory post-operative ALT and AST increase. Two girls, aged 7 and 11 years, showed hypersensitivity to light 2 hours after the operation, decreasing 3 days thereafter.

Conclusion: The use of 5-ALA fluorescence guidance proved to be a safe method in pediatric patients, which can improve intra-operative recurrent neuroepithelial tumor detection and help distinguish it from nonneoplastic pathological tissues. This may increase of the totality of blastomatous tissue removal.

Keywords: 5-aminolevulinic acid, fluorescence-guided resection, gliomas, recurrent operations, children

P-016

Oncology

Sinonasal adenocarcinoma involving the anterior skull base: an exceptional entity in the pediatric population

Patricia Puerta¹; Antonio Guillén¹; Andrés Morales²; Santiago Candela¹; Mariona Suñol³; Enrique Ferrer¹

¹Department of Pediatric Neurosurgery. Hospital Sant Joan De Déu, Barcelona; ²Department of Oncology. Hospital Sant Joan De Déu, Barcelona; ³Department of Pathology. Hospital Sant Joan De Déu, Barcelona

Objective: Sinonasal adenocarcinoma is an uncommon and aggressive malignancy of the nasal cavity and paranasal sinuses associated with poor prognosis. These tumors are divided into salivary-type adenocarcinomas and non-salivary adenocarcinomas. Non-salivary adenocarcinomas are divided into intestinal and non-intestinal types. We present a case of a young child with a non-intestinal adenocarcinoma, a previously unpublished clinical condition.

Methods: We present a pediatric patient with a non-intestinal adenocarcinoma involving the anterior skull base and we describe our management.

Results: A 9-year-old male presented with a one month history of severe frontal headaches and vomiting. Family members noted disinhibition of speech and atypical behavior. His neurological examination revealed a left hemiparesis. CT and MRI showed a 5.9 x 4.3 x 5.7 cm right sided mass arising from the nasal cavity. It eroded through the cribriform plate into the anterior cranial fossa. A combined endonasal endoscopic and bifrontal craniotomy surgical plan was devised to treat this lesion. Postoperative MRI demonstrated gross total tumour resection. After histologic examination, a diagnosis of non-intestinal adenocarcinoma was made. Hybridization for Epstein Barr Virus was negative. Postoperatively, he experienced significant reduction in headaches and improvement of his hemiparesis. He received adjuvant chemotherapy and radiotherapy.

Conclusion: We present the first pediatric case of a non-intestinal adenocarcinoma of the nasal cavity and intracranial involvement. To our knowledge, there aren't pediatric cases of sinonasal adenocarcinoma reported in the English literature. The treatment of these tumors remains a challenge due to the rarity of diagnosis. We treat our patient including radical surgical resection accompanied by adjuvant chemotherapy and radiotherapy. The rarity of this tumor in children means that multicentre effort will be required to determine the optimal treatment.

Keywords: sinonasal adenocarcinoma, skull base, children

P-017

Oncology

Clinical outcome of ependymoma on pediatric and adult patients: a single institutional experience

Tae-Young Jung¹; Shin Jung¹; In-Young Kim¹; Woo-Youl Jang¹; Kyung-Sub Moon¹; Kyung-Hwa Lee²; Seul-Kee Kim³; Jung-Kil Lee¹.

¹Department of Neurosurgery, Chonnam National University Hwasun Hospital, Gwangju; ²Department of Pathology, Chonnam National University Hwasun Hospital, Gwangju; ³Department of Radiology, Chonnam National University Hwasun Hospital, Gwangju

Objective: The purpose of this study was to evaluate the clinical outcome of ependymoma to select the subsequent management plan regarding pediatric and adult cases.

Methods: Between 1997 and 2013, thirty four patients with 23 ependymoma and 11 anaplastic ependymoma were pathologically diagnosed. Six were pediatric patients (mean age, 5.1 years, range, 1.3-11) with a male-to-female ratio of 2:4, while 18 were adults (mean age, 50 years, range, 19-70) with a male-to-female ratio of 13:15. We analyzed the tumor location, extent of tumor resection, adjuvant treatment and the clinical prognostic factors related to progression-free survival (PFS) and overall survival (OS).

Results: In 6 pediatric patients, the tumor showed the infratentorial location in five and supratentorial in one. The mass was totally and subtotally resected in 3 and 3, respectively. Pathologically, ependymoma was diagnosed in 4, and anaplastic ependymoma in 2. The adjuvant treatment was carried out in 5 patients. PFS was 64.1±24.6 months and OS was 73.4±22.2 months. In 28 adult patients, the tumor showed the infratentorial location in 13, supratentorial in 8 and spinal cord in 7. Ependymoma was

diagnosed in 19 and anaplastic ependymoma in 9. The mass was totally removed in 16 and subtotally in 12. Adjuvant treatment was done in 10 patients. The PFS was 143.6.1±14.3 months and the OS was 156.0±12.6 months. The clinical factor of age showed the statistical significance related with OS ($p=0.027$). In adult ependymoma, pathology was related with OS ($p=0.047$) and the 12 totally resected ependymoma without adjuvant treatment showed one recurrence during follow-up.

Conclusion: The pediatric ependymoma was found to have worse survival than adult and the tumor histology was more influential in adult. The totally resected ependymoma without anaplastic pathology could be observed without any adjuvant treatment in adult patients.

Keywords: ependymoma, pediatric, adult, prognosis, survival

P-018

Oncology

Optic nerve glioma in two sisters with family history of M. Recklinghausen

Jivko Surchev¹; Lyudmila Todorova²; Kiril Georgiev¹.

¹Medical University of Sofia, Sofia; ²Institute of Biophysics and Biomedical Engineering, Bulgarian Academy of Sciences, Sofia

Objective: We present two sisters with optic nerve glioma from a mother with skin manifestations of M. Recklinghausen.

Methods: A 4 years older sister, getting exophthalmus because of retrobulbar tumor of the left optic nerve. Due to an intact vision, a biopsy is made and decompression of the orbit roof. Histological results: pilocytic astrocytoma. After a loss of vision and increased exophthalmus, 2 years later the optic nerve was cut at the prechiasmatic level to prevent the engagement of the chiasm and preserving the other optic nerve. There were no tumor cells in the resection line. Three years later, keeping in mind the skin stigmas for M. Recklinghausen of the mother and the older sister, we checked the second daughter 5 years old. We found a right optic nerve retrobulbar tumor, lowering her vision. After 5 months became blind with existing exophthalmus. The affected nerve was cut at the prechiasmatic level (there were no tumor cells in the resection line), decompression of the orbital roof and tumor excision. Histological result: pilocytic astrocytoma.

Results: The follow-up of the two sisters, respectively – 12 and 11 years, using ophthalmological and MRI examination, we found that there is no enlargement of the retrobulbar part of the tumor and respectively the chiasm, secondly the healthy optic nerve are preserved intact. Neurofibromatosis is found in approximately 10% of the nerve pathways glioma cases. In this family the carrier of NF1 is the mother with skin manifestations, and the two daughters - besides the “cafe au lait” spots, the optic nerves are also affected by pilocytic astrocytomas

Conclusion: Optic nerve glioma is a rare tumor with low mortality, but extremely high frequency of losing vision. Prevention of the vision of the healthy eye must be leading in the surgical behavior.

Keywords: Optic nerve glioma, M. Recklinghausen, Exophthalmus, Family history

P-019

Oncology

Papillary glioneuronal tumor in a 15 year old boy: a case report

Nurullah Yüceer¹; Ceren Kızmazoğlu¹; Gonul Guvenç¹; Ismail Kaya¹.

¹Katip Çelebi University Atatürk Trainig and Research Hospital Department of Neurosurgery, Izmir

Objective: Mixed glioneuronal tumour of the central nervous system was called papillary glioneuronal tumor. WHO in 2007 classified the papillary glioneuronal tumor as a grade I neuronal-glioma because of its

indolent clinical course. Only approximately 70 cases have been reported in the last ten years. We report a rare case of papillary glioneuronal tumor.

Methods: A 15 year old boy presented with headache and had no neurological deficits on physical examination. MRI revealed a cystic lesion located in the subcortical white matter of the left frontal lobe to lateral wall of the left frontal horn. The lesion demonstrated peripheral and basal nodular enhancement

Results: The patient was taken to the operating room and underwent a left frontal craniotomy, with the total resection of the mass. The patient post-operatively had no neurological deficits.

Conclusion: Control CT scan demonstrated total resection of the tumor after the operation. The diagnosis of papillary glioneuronal tumor (WHO Grade I) was confirmed by pathology. Papillary glioneuronal tumour should be considered in the differential diagnosis of frontal lobe lesions in the childhood

Keywords: Papillary Glioneuronal Tumor, childhood, WHO Grade I

P-020

Oncology

Malignant transformation in a desmoplastic non-infantile ganglioglioma

Natalie Smith¹; Bhaskar Thakur¹; Miren Aizpurua¹; Angela Calado Bravo¹; Safa Al-Sarraj¹; Sanjeev Bassi¹; Chris Chandler¹; Bassel Zebian¹.

¹Kings College Hospital, London

Objective: Desmoplastic infantile gangliogliomas (DIG) are rare, supratentorial glioneuronal tumours of infants. They are histologically benign and considered as WHO grade I tumours. When presenting in children over the age of 1, they are termed desmoplastic non-infantile ganglioglioma (DNIG) and are more rare. They are not usually associated with malignant behaviour. We present a rare case of recurrent DNIG with malignant transformation.

Methods: A case report and review of the literature.

Results: A four year-old girl presented with a four-month history of worsening headaches, aggressive behaviour and left facial droop. MRI revealed a large right parieto-occipital lesion with mass effect. The lesion was subtotally resected. Histology was consistent with desmoplastic infantile ganglioglioma, WHO Grade I. Follow up MRIs demonstrated aggressive growth requiring further debulking at 7 months and 10 months. The subsequent biopsies showed similar features; however increasing mitotic activity in the predominant desmin positive astrocytic cells. The proliferation index (Ki67) was focally up to 4% in the first biopsy; increasing to 8% and 12% in the subsequent biopsies. Histology from the third resection was consistent with anaplastic ganglioglioma, WHO Grade III.

Conclusion: This case of DNIG demonstrated aggressive growth despite multiple operative interventions, and histology strongly suggesting malignant transformation. Histology has shown malignant transformation, which has only been reported in a handful of cases in the literature. This has important implication for the follow up and management of these lesions.

Keywords: DNIG, Ganglioglioma, malignant

P-022

Oncology

Primary embryonal tumor of spine in young children: report of 2 case

Hsin-Hung Chen¹; Tai-Tong Wong²; Hsiu-Ju Yen¹; Tsui-Fen Yang¹.

¹Taipei Veterans General Hospital, Taipei; ²National Yang-Ming University, Taipei

Objective: Embryonal tumor in the central nervous system is highly malignant neoplasm usually seen in young children and infants. Prognosis is usually very poor. It most commonly occurs intracranially, primary location in the spine is rare and prognosis is even poorer.

Methods: We report 2 cases of primary embryonal tumor of spine in young children. They were a boy and a girl with age younger than 2 years old. They presented with rapidly progressing paraplegia and urine/stool incontinence. Magnetic resonance image of the spinal cord revealed an intradural extra medullary mass occupying T-L spine with extra-spinal extension. Emergent surgery for removal of tumor were performed.

Results: The pathology is atypical teratoid/rhabdoid tumor and medulloblastoma, respectively. Both of them received high-dose chemotherapy with autologous peripheral blood stem cell rescue for 2 to 3 courses. The radiotherapy was postponed until they were 3 years old. The neurological recovery was good and one patient is survived for 2 years but the other was dead with recurrence (multiple leptomeningeal dissemination and lung metastasis) 1 year post-op.

Conclusion: We review the literature on spinal malignant embryonal tumors and discuss the pathology, treatment, and outcome of these rare neoplasms.

Keywords: central nervous system embryonal tumor, spinal tumor, pediatric neurosurgery, chemotherapy

P-023

Oncology

Surgical experience with lateral ventricular choroid plexus papilloma in children

Ehab M Eissa¹.

¹Neurosurgical Department-Kasr Alainy Medical School -Cairo University, Cairo

Objective: surgical experience with lateral ventricular choroid plexus papilloma in children introduction choroid plexus papilloma is the most common lateral ventricular tumor in the children with frequent location in the atrium surgical access to the lateral ventricle remains one of the challenging procedures in the neurosurgery because of the deep location and relation to vital structures especially eloquent area of the brain. In this study, the transcortical microscopic surgery was used to reach the tumor and identify its vascular and structural relation with minimal cortical incision. Endoscopic assisted microscopic surgery was done in the last two cases.

Methods: the seven cases of trigonal location were operated using posterior parietal lobule incision in the dominant side or small tumors in nondominant side and parietotemporal junction incision in large tumors stretching the cortex in nondominant side. The case located in the frontal horn of lateral ventricle was operated by transcortical frontal incision Endoscopic access to the ventricle and navigation of the tumor were done through small incision which was followed by microscopic removal were done in the last two cases.

Results: total removal was done in six cases and in the remaining two another surgical setting were needed to achieve total removal two cases had postoperative visual deficit and one case transient motor deficit and one case had transient sensory aphasia. Endoscopic assisted approach provide short and safe way to the atrium with excellent identification of tumor extension, vascular pedicle and relation and also can help and check total removal

Conclusion: endoscopic assisted access to the lateral ventricular choroid plexus papilloma enhance safe and total microscopic removal of this vascular tumor through small cortical incision

Keywords: choroid plexus papilloma, lateral ventricular tumors, atrium of the lateral ventricle

P-024

Oncology

Choroid plexus papilloma: a report of three cases

Recep Basaran¹; Nejat Isik¹; Naci Balak¹; Basak Caner¹; Sahin Aslan¹; Mustafa Efendioglu¹; Ferruh Gezen¹.

¹1- Medeniyet University Goztepe Education And Research Hospital, Department Of Neurosurgery, Istanbul

Objective: Choroid plexus papilloma (CPP) is rare intracranial neoplasm of ventricles. Approximately 85% of all CPP occur in children under the age of 5 years. CPP accounts for 1% of all brain tumors and 2-6% of all pediatric brain tumors. Clinical presentation of significant hydrocephalus is very common due to increased production of cerebrospinal fluid (CSF). Localizations of CPP are the lateral ventricles in children and the fourth ventricle in adult. Radiologically, CPP is well-defined lobulated masses, associated with hydrocephalus and has marked homogenous contrast enhancement. In this report, we aimed to present three cases of CPP located in posterior lateral ventricle.

Methods: We evaluated cases operated in diagnosis of choroid plexus papilloma retrospectively.

Results: Three cases are presented in this report. Ages of patients at operation were 10, 6 and 5 months. First clinical presentation was vomiting at admission to emergency room. However, one of them was admitted with enlargement of head circumference. Neurological examinations were normal usually but there was bilateral pupillary stasis in the cases aged of 10 months. Common origin of the tumors was left posterior horn of lateral ventricle in two of them. These tumors were enlarging posterolaterally. They compressed third ventricle and also foramen monro and caused hydrocephalus including lateral ventricles. Origin of other tumor was third ventricle. This tumor enlarged superiorly. It invaded bilateral lateral ventricles and caused three ventricular enlargement and hydrocephalus. All cases were operated. Two of them were healed completely but one of them was died at second day of operation. In long term follow-up of the patient, there is no recurrence.

Conclusion: as a rare intraventricular neoplasm of pediatrics, CPP has good prognosis by the total excision of lesion. CSF seeding is uncommon but imaging of entire central nervous system axis is required.

Keywords: Choroid plexus, papilloma, pediatric, surgery

P-025

Oncology

Diffuse astrocytoma mimicking ganglioglioma in a 2 years old girl: a case report

Nurullah Yüceer¹; Ceren Kizmazoglu¹; Gonül Güvenç¹; Ismail Kaya¹.

¹Katip Çelebi University Atatürk Trainig and Research Hospital Department of Neurosurgery, Izmir

Objective: Central nervous system tumors are the most common solid tumors and second most common malignancies of childhood. Signs and symptoms of childhood brain tumors may include headache, nausea, and abnormal coordination in up to one-third of patients at presentation. Here we report the case of a patient at which diffuse astrocytoma appeared like ganglioglioma on MRI.

Methods: Two years old girl had seizures for 8 months and she has used antiepileptic drugs. The patient had no nausea and vomiting. Upon physical examination, the patient was awake, alert and had no neurological deficits. Magnetic resonance imaging (MRI) showed low- enhancing, 3cm, alternating T1 hypointense and T2 hypointense striated appearance of the lesion in the left temporal lobe. The lesion had necrosis in the center appeared as ganglioglioma at the prediagnosis.

Results: The patient was taken to the operating room and underwent a left frontotemporal craniotomy, with the total resection of the mass. The

patient postoperatively had no neurological deficits. Control CT scan demonstrated no residual tumor after the operation.

Conclusion: The diagnosis of diffuse astrocytoma (WHO Grade II) was confirmed by pathology. Diffuse astrocytoma should be considered in the differential diagnosis of temporal lobe lesions in the childhood.

Keywords: Diffuse Astrocytoma, Ganglioglioma, Temporal lobe

P-026

Oncology

Pediatric glioblastoma multiforme: clinical experience of two institutions

Recep Basaran¹; Mustafa Onoz²; Ramazan Sari²; Nejat Isik¹; Basak Caner¹; Ferruh Gezen¹; Ilhan Elmaci².

¹1- Medeniyet University Goztepe Education and Research Hospital, Department of Neurosurgery, Istanbul; ²Memorial Health Group, Department of Neurosurgery, Istanbul

Objective: Glioblastoma multiforme (GBM) is the most common astrocytoma in adults but rare in children. Despite the proven benefit of surgical resection and treatment with chemotherapy and radiotherapy, it has very poor prognosis with a median survival of 12 months. We report our experience about glioblastoma multiforme on children population in two institutions.

Methods: we evaluated seven patients with GBM who were treated surgically in our institutions between 1990 and 2010. We documented age on operation, complaints and neurological examination at admission to hospital, localization of the tumors. We followed-up the patients with radiological and neurological evaluations.

Results: The mean age was 10.28 years, and the majority of them (5/7) were female. Clinical presentations were usually headache and hemiparesis. Four of them had radiotherapy and chemotherapy after aggressive surgical resection. The mean survival was 13.6 months. Five cases survived more than 12 months.

Conclusion: GBM in children are very rare pathology. These aggressive tumors rapidly infiltrate adjacent brain tissue and, they are difficult to treat totally with surgery, radiotherapy and chemotherapy. As results, prognosis was unfavorable.

Keywords: glioblastoma, pediatric, surgery, high grade, survival

P-027

Oncology

Surgery of pineal region tumour using infratentorial supracerebellar approach in semi-sitting position in paediatric patients: is it more advantageous?

Boon Seng Liew¹; Azmin Kass Rosman¹; Gurmit Singh²; Johari Siregar Adnan³.

¹Department of Neurosurgery, Hospital Sungai Buloh, Sungai Buloh;

²Department of Neurosurgery, Hospital Kuala Lumpur, Kuala Lumpur;

³Department of Neurosurgery, Hospital Sultanah Aminah Johor Bahru, Johor Bahru

Objective: Surgery of pineal region tumour is usually challenging due to its location and its proximity to important structures. Few surgical approaches have been described in the literatures. The objective of this paper is to describe the surgical technique and outcomes in five paediatric patients diagnosed with pineal region tumours who were operated using an infratentorial supracerebellar approach in semi-sitting position at Hospital Sungai Buloh, Malaysia from 2010 to 2013.

Methods: Technique: The head end of the table is elevated to an angle of 50 to 60 degree from the floor. The operating table is

positioned to maintain the patient in a semi-sitting position with pelvic support angled at 20 to 30 degrees counter clockwise. This will keep the hip flexed at maximum 90 degree. Midline vertical skin incision, craniotomy exposing the transverse sinus and dura opening are performed. The cisternal magna is opened to drain the cerebrospinal fluid. Some of the bridging veins are cauterized and divided to free the superior surface of the cerebellum. Next, arachnoid dissection is made to expose the quadrigeminal cistern and tumour surface. Care be is taken during removal of tumour to avoid injury to neural and vascular structures.

Results: Complete tumour removal was achieved in one case (20%), subtotal removal in two cases (40%) and partial removal in the remaining two cases (40%). Post-operative pneumocranium were observed in all cases. Frontal burr hole was made to treat pneumocranium in one case. Pneumocranium resolved spontaneously in the remaining cases. No neurological sequelae was noted post-operatively. Neuropathological examination revealed diagnosis of mature teratoma in two cases, germinoma in two cases and pilocytic astrocytoma in one case.

Conclusion: Infratentorial supracerebellar approach in semi-sitting position is a practical surgical approach for pineal region tumour in paediatric patients. More research is recommended to understand safety concerns.

Keywords: Pineal Region Tumour, Infratentorial supracerebellar approach, Paediatrics, Outcomes, Surgical Technique

P-029

Oncology

A case of pediatric hemangiopericytoma located in sellar-suprasellar region

Emre Sağlam¹; Dicle Karakaya¹; Şahin Hanalioğlu¹; Kader Karlı Oğuz²; Figen Söylemezoğlu³; Burçak Biginer¹.

¹Hacettepe University Faculty of Medicine, Department of Neurosurgery, Ankara; ²Hacettepe University Faculty of Medicine, Department of Radiology, Ankara; ³Hacettepe University Faculty of Medicine, Department of Pathology, Ankara

Objective: Hemangiopericytoma (HPC) derived from pericytes, the cells surrounding capillaries and postcapillary venules and is highly vascular and malignant tumors. Pediatric intracranial HPC is so rare that accounting for less than 1% of all central nervous system (CNS) tumors. It is most commonly localized in suprasellar region.

Methods: We represent a rare case of pediatric sellar-suprasellar with clinical and radiological findings.

Results: A 14-year old boy presented with one-year history of headache to our clinic. Neurological examination on admission was normal. Laboratory studies revealed central hypothyroidism and central adrenal insufficiency. The mass lesion consisting of solid and cystic components showed iso-intensity on T1-weighted MRI images, iso-intensity, including partial high intensity on T2-weighted MRI images and strong contrast enhancement. In addition, it destructed skull base and infiltrated left cavernous sinus. The patient underwent subtotal excision and histopathologic diagnosis was hemangiopericytoma. Adjuvant radiotherapy was given.

Conclusion: HPC is a malignant tumor because of local recurrence and high metastasis rates in CNS. They are mostly localised at parasagittal and falx region. Sellar-suprasellar HPC is very rare. HPC is most common in adults, the adult to child ratio is approximately 9:1. The mean age at diagnosis is 40. The clinical presentation of HPC usually depends on the anatomical location. The tumor is usually seen iso-intense on T1-weighted MR images and hyper-intense on T2-weighted MRI, may show contrast enhancement. Dural tail sign can be seen, less common than in meningiomas. In this case, meningiomas could be considered as primary differential diagnosis as well as craniopharyngiomas, optic chiasmatic gliomas, suprasellar germinomas. Furthermore,

sellar localized hemangiopericytomas can mimic pituitary adenomas by causing endocrine dysfunction. Because of high vascularity it can cause surgical problems at endoscopic endonasal transfenoidal approaches. The most appropriate treatment is a combination of radical surgery and adjuvant radiotherapy.

Keywords: Hemangiopericytoma, sellar, suprasellar, pediatric

P-030

Oncology

Primary intracranial low-grade fibromyxoid sarcoma in a pediatric patient

Burcu Göker¹; Mehmet Osman Akçakaya²; Sadık Server³; Mustafa Kemal Hamamcıoğlu¹; Talat Kırış⁴.

¹Department of Neurosurgery, Istanbul Bilim University, Şişli Florence Nightingale Hospital, Istanbul; ²Department of Neurosurgery, Liv Hospital, Istanbul, Istanbul; ³Department of Radiology, Istanbul Bilim University, Şişli Florence Nightingale Hospital, Istanbul; ⁴Department of Neurosurgery, Istanbul Florence Nightingale Hospital, Istanbul

Objective: Low-grade fibromyxoid sarcoma (LGFS) is a rare type of soft tissue tumors, which was first described by Evans in 1987. Since then, about only a 100 cases; most commonly located in the deep soft tissue of the lower extremities or trunk, have been reported in the literature. Primary intracranial involvement is extremely rare with only four reported cases by far.

Methods: Herein we present an additional pediatric patient with a primary intracranial LGFS. To our knowledge this is the first report of a pediatric patient with primary intracranial LGFS.

Results: A four-year old boy admitted with headache, nausea and vomiting. His neurological examination revealed no abnormalities. Cranial computed tomography (CT) scan revealed a right frontal 7x7x4 cm sized, extraaxial mass lesion. The magnetic resonance imaging (MRI) scan showed heterogeneous hypointensity on T1-weighted and isohyperintensity on T2-weighted sequences. The lesion showed diffuse heterogenous contrast enhancement. The patient underwent surgery. With a right frontal craniotomy the mass lesion was microscopic totally en-bloc removed. The pathological findings were in consistence with a low-grade fibromyxoid sarcoma: A fibrotic center surrounded by lobules of myxoid components. The mitotic activity and cellularity were low, with a Ki-67 proliferation index of less than 1 %. The tumor showed positive staining for mason, and negative staining for GFAP, reticulin, synaptophysin, EMA and S-100. The postoperative period was uneventful and the patient was discharged from the hospital without any neurological deficits or complications. There was no evidence of recurrence during the 18-month follow-up period.

Conclusion: The present case showed clinical and radiological similarities to intracranial extraaxial lesions. Myxoid and chordoid meningiomas were also listed in the pathological differential diagnosis of LGFS. Although rare, LGFSs should be kept in mind in the differential diagnosis of extraaxial mass lesions also in pediatric age group.

Keywords: low-grade fibromyxoid sarcoma, brain tumor, mesenchymal tumor, pediatric

P-031

Oncology

Primary spinal primitive neuroectodermal tumor: a unique case presented with subarachnoid hemorrhage one year prior to the diagnosis

Thomas W. Morris¹; Raghu H. Ramakrishnaiah²; Murat Gokden³; Eylem Ocal¹.

¹Department of Neurosurgery - University of Arkansas for Medical Sciences, Arkansas Children's Hospital, Little Rock; ²Department of Radiology- University of Arkansas for Medical Sciences, Arkansas Children's Hospital, Little Rock; ³Department of Pathology- University of Arkansas for Medical Sciences, Arkansas Children's Hospital, Little Rock

Objective: Spinal primitive neuroectodermal tumor is a rare aggressive tumor that typically occurs in children and young adults. They can present in a variety of ways, of which subarachnoid hemorrhage is a rare one but not in isolation. The authors present a unique case of a spinal primitive neuroectodermal tumor initially heralded by subarachnoid hemorrhage and hydrocephalus approximately a year prior to the diagnosis, the etiology of which was not discovered despite thorough diagnostic evaluation. **Methods:** We report a case of a 17 year old male who initially presented with signs of high intracranial pressure and hydrocephalus after acute onset of headache. He was noted to have subarachnoid hemorrhage with xanthochromic CSF on lumbar puncture. He underwent extensive diagnostic lab testing and imaging, including diagnostic cerebral angiography, spinal MRI /MRA, CSF pathology and cytology, all of which were inconclusive. A tiny focal enhancement on the dorsal aspect of the spinal cord at T6 vertebral level was noted. The MRA and a repeat high resolution MRI did not yield further imaging findings. The study was concluded as nonspecific focal leptomeningeal enhancement warranting follow-up. Patient remained neurologically intact but eventually required shunt placement for hydrocephalus. He was followed closely as an outpatient with repeat imaging studies of spine and brain. Patient re-presented one year later with acute back pain, and paraparesis. Repeat imaging showed extensive meningeal enhancement. The case was discussed with its rare presentation; imaging findings and the relevant current literature were reviewed.

Results: Patient underwent surgery followed quickly by chemotherapy and radiation. He continued to have worsening neurological function despite appropriate therapy and surgery.

Conclusion: This was an unusual case in terms of its rare presentation, imaging findings and delayed diagnosis. We aim to share our experience and provide a discussion about this rare pathology, its variable presentation and potential pitfalls in management.

Keywords: spine, primitive neuroectodermal, subarachnoid hemorrhage, tumor, surgery

P-032

Oncology

Neurofibromatosis presenting in the first year of life: case report and review of the literature

Mehmood Albarazi¹; Bhaskar Thakur¹; Neda Barzegar - Befroei¹; Angela Calado Bravo¹; Rosalie Ferner¹; Nick Thomas¹; Bassel Zebian¹. ¹Kings College Hospital, London

Objective: Neurofibromatosis (1 or 2) with symptoms or signs of neurological deficit presenting in the first year of life is very rare. Prognosis in this subgroup is difficult to predict and management is challenging.

Methods: Case report and literature review. A 3-month-old girl born at term who presented with difficulty swallowing since birth and bilateral mild weakness of wrist extension. Imaging demonstrated a plexiform neurofibroma in the upper cervical spine extending to the retropharyngeal space. She was initially referred as a suspected retropharyngeal abscess. Examination revealed that she had multiple Café au Lait spots and the diagnosis of neurofibromatosis type 1 was thus made.

Results: In total there are 21 patients reported to have had a diagnosis of neurofibromatosis (1 or 2) made in the first year of life. The clinical presentation in this subgroup shows great variability. Due to the paucity

of reports it is difficult to draw conclusions regarding prognosis and best management. Our patient was managed conservatively and we discuss our reasoning.

Conclusion: It is vital especially in this subgroup of patients that, once the diagnosis is suspected, prompt referral to supraregional centres is made both to ensure that the experience is concentrated in

Keywords: Neurofibromatosis, NF1, infant, plexiform neurofibroma

P-033

Oncology

Pediatric CNS tumors: result of a reference center in middle Anatolia Abdulfettah Tümtürk¹; Ekrem Ünal²; Hüseyin Per³; Selim Doğanay⁴; Oğuz Galip Yıldız⁵; Mehmet Akif Özdemir²; Halil Ulutabanca¹; Olgun Konaş⁶; Abdulhakim Coşkun⁴; Ali Kurtsoy¹.

¹Erciyes University, Faculty of Medicine Department of Neurosurgery, Kayseri; ²Pediatric Hematology and Oncology, Erciyes University, Faculty of Medicine, Department of Pediatrics, Division of Pediatric Hematology & Oncology, Kayseri; ³Erciyes University, Faculty of Medicine, Department of Pediatrics, Division of Pediatric Neurology, Kayseri; ⁴Erciyes University, Faculty of Medicine, Department of Radiology, Division of Pediatric Radiology, Kayseri; ⁵Erciyes University, Faculty of Medicine, Department of Radiation Oncology, Kayseri; ⁶Erciyes University, Faculty of Medicine, Department of Pathology, Kayseri

Objective: CNS tumors are the leading cause of cancer related deaths in childhood. Although CNS tumors are reported to occur with almost similar incidence in the same age range in different developed countries, the incidence of pediatric CNS tumors in developing countries is not well known. We aimed both to provide epidemiologic features of primary malignant CNS tumors in children who were admitted to our center and to shed light on the epidemiological features of the CNS tumors in Turkish children 0–17 years of age.

Methods: Medical records of the patients with CNS tumors managed at Erciyes University over a 15-year period (2000–2015) were analyzed retrospectively.

Results: There were 268 (157 boys and 111 girls) children eligible for the study. The median age was 8 years, ranging 1 month to 17 years. The most common complaints in our study were headache, vomiting, seizure and torticollis. The majority of 268 tumor cases were located supratentorial (n=139, 51,86%), and the rest were infratentorial (n=123, 45,89%) and spinal (n=6, 2,23%). Males were more likely to develop CNS tumors. Glial tumors, embryonal tumors, ependymal tumors were the most common tumors.

Conclusion: Pediatric brain tumors present with various initial symptoms and signs. Despite headache and vomiting were the most common complaints of those patients, interestingly, torticollis was another common complaints of the patients of our series and supratentorial localization was higher than Infratentorial ones. The current study is a single institution study and needs cautious interpretation.

Keywords: Childhood, Brain Tumors, Spinal tumors, epidemiology

P-034

Oncology

Management of CNS atypical teratoid/rhabdoid tumors in pediatric age group: CCHE experience in 16 cases.

Mohamed A. El Beltagy¹; Mohamed Reda²; Mohamed Saad Zaghoul³; Hala Taha⁴; Nada El Khateeb⁵; Madeeha Awad⁶.

¹Chief Neurosurgeon, Children's Cancer Hospital Egypt (CCHE, 57357), Consultant Neurosurgeon and Professor, Neurosurgery Department, Kasr Al-Ainy School of Medicine, Cairo University, Egypt, Cairo; ²Consultant Neurosurgeon, Children's Cancer Hospital Egypt (CCHE, 57357). Lecturer Of Neurosurgery, Kasr Al-Ainy School of Medicine, Cairo University, Egypt, Cairo; ³Consultant And Head of Radiation Therapy Department, Children's Cancer Hospital Egypt (CCHE, 57357), Cairo; ⁴Consultant And Head of Surgical Pathology Department. Children's Cancer Hospital Egypt (CCHE, 57357), Cairo; ⁵Research Department, Children's Cancer Hospital Egypt (CCHE, 57357), Cairo; ⁶Consultant of Pediatric Neurooncology, Children's Cancer Hospital Egypt (CCHE, 57357, Cairo

Objective: Atypical Teratoid/Rhabdoid Tumor (AT/RT) is a highly malignant embryonic tumor in the Central Nervous System (CNS) with a very poor prognosis that mainly affects children of a young age. We will discuss our experience in management, and outcomes of AT/RT patients diagnosed and treated at Children's Cancer Hospital Egypt (CCHE-57357).

Methods: Charts of patients with pathologically confirmed AT/RT diagnosed and treated at CCHE in the period between July 2007 and December 2013 were retrospectively reviewed. Demographic data, clinical treatment and outcome were collected and analyzed using IBM SPSS version 20. As per CCHE standard treatment guidelines for AT/RT, patients are subjected to surgery followed by pre-irradiation chemotherapy, radiation therapy, and maintenance chemotherapy.

Results: Sixteen patients (9 males/ 7 females) with mean age 2.1 years (range 0.5-4.5) were identified. Patients presented with lobar (n=6), posterior fossa (n=4), lateral ventricles (n=2), pineal body (n=2), thalamic (n=1) and spinal lesions (n=1). All patients were subjected to surgery with gross total resection (GTR) (n=5), Subtotal resection (STR) (n=8) and biopsy (n=3). Fifteen patients had smooth postoperative recovery. There was one case of delayed postoperative mortality after 50 days in intensive care unit. One year overall survival was 41.5 % with median survival of 9 months. One year event-free survival was 34.7% with median survival of 5.3 months. The age at diagnosis, gender, tumor location and extent of resection had no significant impact on overall and event-free survival.

Conclusion: AT/RT is a very aggressive tumor with dismal prognosis. Total surgical resection of the tumor did not improve overall or event free survival. Studying of larger number of patients over a longer period of time is recommended.

Keywords: at/rt, teratoid, rhabdoid, brain tumors, pediatric

P-035

Oncology

Multisurgical spinal PNET after radiation and chemotherapy: case report

Zhi-Hua Chen¹; Hu-Fei Qian¹; Jia Wei¹; Qi-Jia Zhan¹; Ruo-Ping Chen¹.

¹Children's Hospital Of Shanghai, Shanghai

Objective: Primitive neuroectodermal tumor (PNET) is high grade malignant, spinal PNET (sPNET) is extremely rare. We evaluated the effectiveness of comprehensive treatment to a 13-years-old boy.

Methods: The patient underwent four times T8-9 intraspinal surgeries with neuroelectrophysiological monitoring, additionally for pedicle screw fixation. The spinal radiation therapy was utilized, and chemotherapy is given during and following radiation. Every operation, the tumor tissue was harvested for molecular test to guide the different regimens.

Results: From the first operation at 2013 September to now, during 20 months the tumor reoccurred three times even after the radiation and chemotherapy. The extent of resection were total at first time and nearly total at second, but it was sub-totally resection at third and fourth time because the tumor was aggressive into chest. The patients double lower

limbs strength level was VI grade after first surgery, but gradually decreased to zero every recurrence and Babinskis sign was positive when he discharged. Chemotherapy regimens were changing according to molecular diagnosis. MGMT was methylation firstly but unmethylation secondly. The second molecular result showed ERCC1 high expression meant Cisplatin was ineffective, but TOP2A high expression indicated Etoposide was the good choice. The third time, molecular test showed Tubulin B3 and PRM1 was low expression so that Taxol associated with Irinotecan was used. Unfortunately, the tumor reoccurred fourth time, molecular test suggested BRAF V600E, EGFR, PTEN and VEGFR-2 would be effective. Up to now, the condition was stable with application of Bevacizumab. Additionally, the pathological immunohistochemical results were variation.

Conclusion: sPNET tumor always evolved its biological hallmarks to survival even experienced radio-chemotherapy, which may be related to cancer stem cells. Clinically, active multi-resection is benefit to intraspinal decompression and harvest tissue for molecular test to select chemotherapy which can improve the postoperative outcome.

Keywords: spinal, Primitive neuroectodermal tumor, comprehensive treatment, molecular genetic test

P-036

Oncology

Pilomyxoid astrocytoma of spinal cord: case report and literature review

Anwar Ul Haq Dr¹; Essam Al Shail Dr¹.

¹King Faisal Specialist Hospital and Research Centre, Riyadh

Objective: Pilomyxoid Astrocytoma is a rare and new clinicopathological entity. It has different histopathological features and clinical behavior as compared to pilocytic astrocytoma. It is WHO grade II tumor. The most common location of Pilomyxoid astrocytoma is chiasmatic/hypothalamic region. Pilomyxoid Astrocytoma of Spinal Cord is extremely rare. Aim and Objective is to described a case of extensive Pilomyxoid Astrocytoma of Spinal Cord presenting as holocord (Syringomyelia and Syringobulbia) in a two years girl and a review of cases of Pilomyxoid Astrocytoma of Spinal Cord reported in literature.

Methods: 2 years old girl presented with 4 months history of neck pain, torticollis and mild scoliosis. Her neurological examination revealed right upper limb weakness with power 4+/ 5. Power in rest of the limbs was 5/ 5. MRI spine showed extensive syringomyelia and syringobulbia extending from medulla oblongata to D10 level. Gadolinium enhanced MRI showed heterogeneously enhancing intramedullary spinal tumor in the lower cervical and upper thoracic region. The syrinx was also enhancing from medulla to lower thoracic region.

Results: Patient underwent C7-T2 laminotomy, biopsy of the tumor, drainage of syrinx and laminoplasty under neurophysiological monitoring. Histopathology was consistent with Pilomyxoid astrocytoma. Patient was referred to Pediatric hematology oncology for chemotherapy. She received chemotherapy with baby brain protocol consisting of Vincristine and Cytosar. Her tumor responded to chemotherapy nicely. Her follow up MRI at six months showed significant reduction in tumor and syrinx.

Conclusion: Pilomyxoid Astrocytoma of Spinal Cord is extremely rare. Only 10 cases of Pilomyxoid Astrocytoma of Spinal Cord has been described in literature and our is 11th case. This is a first case of Pilomyxoid Astrocytoma presenting as holocord. Pilomyxoid Astrocytoma should be considered in the differential diagnosis of Intramedullary Spinal Tumors. The treatment consist of surgical debulking followed by adjuvant therapy with chemotherapy and/or radiotherapy depending upon the age of patient

Keywords: Pilomyxoid Astrocytoma, Spinal Cord Pilomyxoid Astrocytoma, holocord tumor

P-037

Oncology**Intracranial undifferentiated malign neuroglial tumor in Smith-Lemli-Opitz Syndrome: a possible predisposing factor for primary brain tumors**Ayfer Aslan¹; Alp Özgün Börcek¹; M. Kemali Baykaner¹.¹Division of Pediatric Neurosurgery, Gazi University Faculty of Medicine, Ankara

Objective: Smith-Lemli-Opitz Syndrome (SLOS) is a rare hereditary autosomal recessive disorder of cholesterol synthesis because of 7-dehydrocholesterol reductase deficiency. The characteristic features are growth abnormalities like microcephaly, ptosis, micrognathia and multisystemic structural malformations like heart, lungs, kidneys, gastrointestinal tract and genitalia. Here we aimed to present our case and attract the attentions to any possible relationship between SLOS and intracranial undifferentiated neuroglial tumor development.

Methods: 10-year-old male patient who had had diagnosis of SLOS for infant age and had some progressive thickening and lumps on his head since 6 months. After some varifications it was understood that they are due to a huge intracranial lytic mass lesion. The skull was microcephalic as a component of syndrome and was mostly destroyed by the mass (Figure 1). We operated him in our clinic, and performed debulking of the mass (Figure 2).

Results: After his diagnosis of brain tumor, he was operated in our clinic and he was survived. According to histopathologic results as undifferentiated malignant neuroglial WHO grade 4 tumor, adjuvant radiotherapy and chemotherapy was performed after surgery.

Conclusion: Today it is known the cholesterol of which synthesis is abnormal in SLOS, takes place in the Hedgehog signaling pathway which leads the transcription of genes that regulate cell proliferation and differentiation. It's possible to make a hypothesis that there is a strongly relationship with SLOS and intracranial undifferentiate malign tumors via impairment of Hedgehog proteins activity and cell differentiation. This probable relationship is an unworked field which should be paid more attention on. In the literature there is only one case report that mentioned co-existing of intracranial malignant germ-cell tumor and SLOS.

Keywords: Smith-Lemli-Opitz Syndrome, Sonic hedgehog

P-038

Oncology**Abdominal neuroblastoma with solitary presentation as increasing head circumference: case report and literature review**Anwar Ul Haq Dr¹; Essam Al Shail Dr¹; Mohammad Anas Dababo Dr¹; Amani Alkofide Dr¹.¹King Faisal Specialist Hospital And Research Centre, Riyadh, Riyadh

Objective: Neuroblastoma is the most common solid tumor in infants and the second most common solid tumor in children. Most common presentation of abdominal neuroblastoma is abdominal mass, abdominal distension, constipation or pain. Abdominal neuroblastoma primarily presenting as increasing head circumference is rarely reported in literature. We report a case of one year old male child with abdominal neuroblastoma who presented with history of increasing head circumference without any symptoms related to abdominal tumor. Abdominal neuroblastoma was discovered on imaging studies.

Methods: One year old male child presented with history of increasing head size from the age of 8 months. He did not has any symptoms of raised intracranial pressure or any symptom related to abdominal tumor. His head circumference was 57 cm. CT brain and skull showed diffuse thickening of calvarium and facial bones. Thickness of calvarium was 4-5

cm. There were expansile masses at bilateral parietal areas. The calvarial involvement had typical sunburst appearance/ hair on end appearance. Abdominal X-Ray showed retroperitoneal calcifications. CT chest and abdomen showed retroperitoneal mass with calcification and metastasis to ribs and lungs. MIBG scan was positive for retroperitoneal tumor with diffuse skull, ribs, pelvis and appendicular metastasis.

Results: He underwent open biopsy from the left parietal skull mass. Biopsy report was consistent with poorly differentiated neuroblastoma. MYCN oncogene amplification was positive. He was referred to pediatric Hematology and Oncology for chemotherapy. Currently he is receiving chemotherapy consisting of vincristine, cisplatin, doxorubicin, cyclophosphamide and etoposide.

Conclusion: Metastatic neuroblastoma is an uncommon cause of increasing head circumference and is not commonly encountered in neurosurgical practice. It should be considered in the differential diagnosis of increasing head circumference in young child. Solitary presentation as increasing head circumference due to skull metastasis indicates aggressive nature of the primary abdominal neuroblastoma which was verified by MYCN oncogene amplification.

Keywords: Neuroblastoma, metastatic neuroblastoma, skull metastasis

P-039

Oncology**Intracavitary interferon alpha treatment in cystic craniopharyngiomas**Mehmet Saim Kazan¹; Ethem Taner Göksu¹; Nurşah Eker².¹Department of Neurosurgery, Akdeniz University Faculty of Medicine, Antalya; ²Department of Pediatrics, The Division Of Pediatric Hematology And Oncology, Antalya

Objective: Craniopharyngiomas are challenging lesions to resect completely and safely due to neighbouring on the critical neurovascular structures. Intracystic interferon alpha (IFN α) chemotherapy has been used as an alternative treatment for predominantly cystic craniopharyngiomas.

Methods: We have used this modality in four children. A silicone catheter was inserted to the cyst cavity by direct puncture or microsurgical access and connected to an Ommaya reservoir. In every application, one ml IFN α 2A was injected into the Ommaya reservoir. Each application consisted of an injection of 3,000,000 units of IFN α 2A, totaling 36,000,000 units, which was considered one treatment cycle. Prior to application, cyst fluid was aspirated until the flow discontinued or the patient began to complain of a headache. Magnetic resonance imaging (MRI) was obtained at one, three and six months after the initial application.

Results: The ages of children were 6, 9, 11, 14 and follow-up times were 37, 24, 33 and 12 months respectively. Catheter insertions was carried out with direct frontal puncture in two, subfrontal microsurgical access in one and interhemispheric transcallosal transventricular route in one case. In one child, fever and fatigue were observed at the beginning of applications but they did not required the discontinuation of treatment. The number of treatment cycle was three in one case and one in other three subjects. Following the treatment, cyst volume decreased more than 75% in all patients. In three children who were admitted with hydrocephalus, ventricular system became in normal size and no one required shunt application.

Conclusion: Intracystic IFN α treatment has been considered as an efficacious, safe, inexpensive and less invasive modality for predominantly cystic craniopharyngiomas. Our early results were highly satisfied and drug was well - tolerated without any significant side effects. Further series and long-term follow-up could reveal whether this therapeutic option provides a definitive tumor control.

Keywords: Craniopharyngioma, intracystic treatment, interferon alpha

P-040

Oncology

Retrospective analysis of brainstem tumor in children: 60 cases report from the Department of Pediatric Neurosurgery, Xinhua HospitalZixuan Wang¹; Jie Ma¹¹Shanghai Xinhua Hospital, Shanghai

Objective: According to the 2014 Childhood Cancer Statistics, brain and CNS cancer has the second highest incidence rate and the highest mortality rates. Brainstem Tumor (BST) is always considered to be the most risky pediatric tumor as brainstem plays an important role in the regulation of cardiac and respiratory function. Years ago, brainstem remains an inoperable region. In this report, we'd like to talk about our surgical management of BST by presenting 60 cases we collected.

Methods: The data of clinical course were obtained and carefully reviewed from 60 cases of brainstem tumor patients, which were accepted in our department from Jan.1st 2006 to April.30 2015. We conclude the clinical characteristics of these cases: sex, age, tumor location, clinical features, operative approach route, resection degree, prognosis, and the effect of radiation and chemotherapy, etc.

Results: Among all 60 cases, average sex is about 5.6 years old. There are 43 male and 17 female. Common location includes medulla oblongata, pons, midbrain, cervicomedullary junction, cerebellar-pontine angle (CPA). Tumors can present to be intrinsic or exophytic. The tumors were approached mainly by several routes, posterior midline, retro sigmoid and poppen approach. We did total resection in 5 cases, subtotal resection in 6 cases, partial resection in 19 cases, and biopsy in 20 cases. 2-year progression-free survival rate is 90%. 11 patients accepted radiation or chemo-therapy, 4 patients cannot be contacted.

Conclusion: Brainstem tumor is not an inoperable disease with detailed pre-operation examination and exquisite operating. Exophytic and low grade tumors are usually easy to be removed and have better prognosis. The most common approach routes are through posterior midline approach, retrosigmoid approach and poppen route. The most common pathologic type is Pilocytic Astrocytoma (WHO II). 2-year event-free survival was 90%. 11 patients accepted radiation or chemo-therapy and have a relatively optimistic prognosis.

Keywords: brainstem tumor, pediatric, surgery, children, case report

P-041

Oncology

The management of posterior fossa tumor in children: a report of 200 cases from ChinaJie Ma¹; Baocheng Wang¹¹Xin Hua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai

Objective: This review was to investigate treatment outcome and prognostic factors after treatment of posterior fossa tumors.

Methods: A total of 200 patients with resection for posterior fossa tumor cases treated at our institute between 2006 and 2014 were included in the study. Demographic variables, clinical variables, radiological findings and treatment details with respect to age, sex, signs and symptoms, location of tumor, extent of surgical resection, histopathology diagnosis, follow up period and outcomes were recorded.

Results: 165 (82.5%) tumors locate in the midline and 12 (6%) lesions occur in the CPA zone and 23 (11.5%) tumors were at the cerebrum. 7 children with metastasis at the primary diagnosis cover the embryonal tumor(AT/RT 3 cases, MB 2 cases), ENP 1 case and metastatic tumor 1

case. The histopathology confirmed 135 (67.5%) lesions were high malignant tumor such as MB 32%, AT/RT 6%, glioma 12%, EPN 16% and others. In the pediatric population, posterior fossa masses display the risk of low degree glioma, accounting for 32.5% in this study. Tumor resection was performed in all patients with gross total resection in 46 (23%) patients, near total resection in 98 (49%) patients and subtotal resection in 23 (11.5%) patients. All older children (>3 years) underwent postoperative craniospinal irradiation (CSI) delivering a median craniospinal dose of 36 Gy with additional boosts to the posterior fossa up to 54 Gy. Median overall survival was 37 months for the total group. Low grade glioma showed an improved outcome compared with other histology with a median survival of 78 months compared with that of high malignant histology being a median survival of 34 only.

Conclusion: Treatment of posterior fossa tumor with surgery and CSI yields long survival rates in children. Histology was associated with a different outcome.

Keywords: posterior fossa, tumor, treatment

P-042

Oncology

Optic pathway gliomas one national centre's experience and literature reviewFeili Liu¹; Jie Ma¹¹Xin Hua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai

Objective: Optic pathway gliomas (OPGs) are generally low-grade gliomas, and pilocytic astrocytoma and diffuse astrocytoma are the most common pathology types, which can occur anywhere along the optic pathway including the optic nerves, chiasm, and tracts. It occurs most often under the age of 10 and frequent in children with neurofibromatosis type 1 (NF1), and account for 3%~5% of all childhood CNS tumors. The treatment and management of OPGs is remains challenging.

Methods: Retrospectively study of the 40 cases experience of the pediatric department of Neurosurgery Xinhua Hospital and literature review.

Results: This review discuss the current understanding and advances of the OPGs about the genetic condition, evaluation, examining tumor progression and Optimal management based upon visual symptoms, tumor location, patient age, growth rate and the social and economic factors.

Conclusion: Multi-disciplinary team (MDT) including neurosurgery, ophthalmology, radiation oncology, neuro-oncology, neuroradiology is a good choice for the OPGs patients treatment. And both retrospective and prospective study is needed to explore new therapy, particularly the targeted therapies.

Keywords: Optic pathway glioma, treatment, children

P-043

Oncology

Extraneural metastasis of glioblastoma multiformeMario Garcia-Conde¹; Vanessa Hernandez-Hernandez¹; Liberto Brage-Martin¹; Pablo Febles-Garcia¹; Julio Plata-Bello¹; Ayoze Doniz-Hernandez¹¹Hospital Universitario De Canarias, La Laguna

Objective: Glioblastoma multiforme (GBM) is the most common primary malignant e of the central nervous system in adults with a mean age at diagnosis of 64 years. In the paediatric population accounts for 3% of all CNS tumors. They are generally confined

to the intracranial compartment, with a less than 2% having extraneural dissemination.

Methods: We present the case of a 15 year old male with a right occipital lesion. The procedure is performed with subtotal resection and with left homonymous hemianopia as the only postoperative complication. Post-Surgical pathology reports resulted in a grade IV astrocytoma for which he received adjuvant radio-chemotherapy. His post-treatment Karnofsky index was 90%. Combined treatment ended 3 months after initial entry. Temozolomide was used for his maintenance and palliative treatment. An MRI one month after completing cancer treatment showed local tumor progression. 5 months later, the patient was admitted with a fever, flank pain, and dyspnea. Radiological studies showed a right pleural effusion. Drainage was performed and samples for cytological and biochemical analysis were collected and resulted in atypical cells that corresponded with pleural metastasis of GBM. In addition, the patient started having abdominal pain and radiological studies showed the presence of free peritoneal fluid, and acalculous cholecystitis versus liver metastases.

Results: The patient progressed unfavourably with increased work of breathing and analgesia requirement due to abdominal pain, and died 24 hours after this last diagnosis.

Conclusion: Glioblastoma multiforme in childhood and the development of extraneural metastatic disease is a rare event as this entity usually presents in adults and is confined to the CNS.

Keywords: glioblastoma multiforme, extraneural metastases, children

P-044

Oncology

Neurosurgical management of CNS germ cell tumour: the safe and invasiveness

Xianlun Zhu¹.

¹Prince of Wales Hospital Chinese University of Hong Kong, Hong Kong

Objective: Surgical intervention for germ cell tumour (GCT) varies in the indication and modality where invasiveness and safety are the main concern. The objectives of the report are [1] the success rate of obtaining pathological diagnosis and tumour excision; [2] Surgical complication and invasiveness.

Methods: All CNS germ cell tumour (GCT) received neurosurgical intervention in our institute up to 2014.

Results: Total 32 cases from 1987 to 2014 (may not be complete before 2000); 1987-2005: 7 cases; 2006-2014: 25 cases. Gender ratio: Female to male 10: 22; Age at diagnosis: mean 13 (SD 7.4 years), median 11 (range 10 months -14 years). Follow-up period: 9 months to 27 years. Overall outcome: 4 cases passed away (diagnosed in 84, 99, 01, 05). Pathology (cases): Germinoma 22, mixed tumour 3, malignant teratoma 2, matured teratoma 2, choriocarcinoma 2, embryonic carcinoma 1. Surgical modality for tumour intervention: [1] Craniotomy for biopsy / decompression / excision. [2] Burr hole stereotactic needle biopsy. [3] Endoscopic tumour biopsy + Endoscopic 3rd ventriculostomy. [4] Transphenoidal biopsy. The successful rate for pathological diagnosis was 92.5%. Surgical complication rate was 0.5%. With the development of stereotactic / endoscopic and transphenoidal biopsy minimally invasive techniques, primary craniotomy biopsy-guided resection has been much less used after 2006. For endoscopic biopsy through the 3rd ventricle for pineal region tumour, the chance of negative or incomplete sampling rate is high among other biopsy methods in our series.

Conclusion: Neurosurgical intervention for CNS germ cell tumour is safe and effective. The choice of different surgical modality should be tailored to individual case. Primary craniotomy for biopsy-guided resection has been much less used with the availability of minimally invasive surgical modalities.

Keywords: Germ cell tumour, CNS, Neurosurgery

P-045

Oncology

Molecular characterization of a pleomorphic xanthoastrocytoma cell line isolated from a 14-year-old male

Emin Umit Bagriacik¹; Kemali Baykaner²; Melek Yaman¹; Nihan Örüklü¹; Alp Börcek²; Sanem Gökçen¹; Hakan Emmez³

¹Department of Immunology, Gazi University, Faculty of Medicine, Ankara; ²Department of Pediatric Neurosurgery, Gazi University, Faculty of Medicine, Ankara; ³Department of Neurosurgery, Gazi University, Faculty of Medicine, Ankara

Objective: Previously we isolated a primary anaplastic pleomorphic xanthoastrocytoma cells from resected tumor tissue of a 14-year old male. The primary cells were cultured passage by passage over one year to establish a stable cell line. The purpose of this study was to characterize genes that were related to development of carcinogenesis and malignancy in the established stable cell line.

Methods: PXA cells were grown in DMEM cell culture medium supplemented with 10% FBS, 2mM L-glutamine and antibiotics. Total RNA was isolated using a total RNA isolation kit. Gene expression was assayed by a real time PCR array kit.

Results: Gene expressions for 84 genes were tested. We detected important increases and decreases in expression of several genes related to cancer pathways. ACSL4, SLC2A1, SNAI3 are among those genes.

Conclusion: In conclusion, those genes would be associated with development of carcinogenesis in glial tumors.

Keywords: pleomorphic xanthoastrocytoma, PCR Array, Gene expression

P-046

Oncology

The relationship between apoptosis and STMN1 regulating the chemosensitivity of glioma cells to TMZ

Jie Ma¹; Feili Liu¹.

¹Xin Hua Hospital Affiliated to Shanghai Jiaotong University School of Medicine, Shanghai

Objective: To investigate the role of autophagy in the process of STMN1 regulating glioma chemosensitivity to TMZ.

Methods: Using TEM observe autophagy ultrastructure—autophagolysosome after TMZ induction in U251-MG (RNAi-STMN1). Western-blot was used to detect protein expression. To observe the TMZ-induced autophagic vacuoles in U251-MG through GFP-LC3B tracer method under laser scanning confocal microscope (RNAi-STMN1), as well as fluorescence intensity changes after the suppression of autophagy by 3-MA or bafilomycin A1. And qRT-PCR and western-blot are used to detect gene and protein expression levels. Finally, the sensitivity to TMZ of U251-MG (RNAi-STMN1) is assayed by CCK-8 kits after the inhibition of autophagy by 3-MA and/or bafilomycin A1 early and late.

Results: After TMZ induction, autophagic vacuoles emerge in cytoplasm of U251-MG. For U251-MG (Si-STMN1) after TMZ induction, The degree of cell damage is worse than U251-MG and U251-MG (Scramble), and autophagic vacuoles in the cytoplasm increase. In U251-MG and A172 cell lines, LC3, Beclin1 and ATG5 mRNA and protein expression level significantly increase ($P < 0.05$) after TMZ induction, U251-MG (RNAi-STMN1) group reach statistical highly significance ($P < 0.01$), as well as Beclin1 in A172 (RNAi-STMN1). Comparing U251-MG (RNAi-STMN1)+TMZ+3-MA/Baf group and U251-MG(RNAi-STMN1)+TMZ group, the number of autophagic vacuoles reduce, achieving a statistical significant difference ($P < 0.05$). After the suppression of the early or late, autophagy induced by TMZ is

still enhanced. The expression level of autophagy-related genes / proteins (LC3, Beclin1 and ATG5) was significantly increased ($P < 0.05$). The cell viability U251-MG (RNAi-STMN1)+TMZ+3-MA+Baf group is higher than U251-MG (RNAi-STMN1)+TMZ group statistically ($P < 0.05$); separated early or late suppression of autophagy alone, comparing U251-MG (RNAi-STMN1)+TMZ+3-MA/Baf group and U251-MG (RNAi-STMN1)+TMZ group, the cell viability increase does not reach the statistical significant difference ($P > 0.05$).

Conclusion: TMZ can induce autophagy in U251-MG and U251-MG (Si-STMN1), autophagy-related genes / proteins significantly increase after TMZ induction.

Keywords: glioma, TMZ, STMN1, autophagy, chemosensitivity

P-047

Posterior fossa tumor

Cerebellar teratoma in infant

Yasou Yamanouchi¹; Hideyuki Ohshige²; Takahiro Yamahara²; Kunikazu Yoshimur¹; Masahiro Nonaka²; Akio Asai².

¹Tsurumiryokuchi Hospital, Moriguchi; ²Kansai Medical University, Hirakata

Objective: Intracranial teratomas are rare and 1.2% of all pediatric brain tumors. The majority of intracranial teratoma are located in pineal and suprasellar regions. We experienced two rare cases of cerebellar teratoma in female infant.

Methods: Case 1: 4 month old female infant admitted to our hospital because of poor milk intake and vomiting. Neuroradiological examination revealed large tumor with calcified spots and cysts in the posterior fossa. Hydrocephalus was apparent. Baby showed sunset phenomenon and opisthotonic posture. Serum AFP was 1149ng/mL and hCG β subunit was under 0.1m IU. Suboccipital craniotomy was performed on the day following admission. Pathological diagnosis was immature teratoma and potoperative chemotherapy was performed. The infant was discharged without any problem and follow ups were performed for 9 years. No recurrence of neoplasm are noted at present. Case 2: 1 year 4 month old female infant was admitted to our hospital. She started to vomit frequently at 10 month of age and enlargement of head circumference. MRI showed large cystic tumor with hydrocephalus in the posterior fossa. Suboccipital craniotomy was performed and the tumor was removed totally. The patient showed transient swallowing problem. Pathological diagnosis was mature teratoma. She is 4 years old now and doing well.

Results: We reported two rare cases of cerebellar teratoma in female infant. One was mature teratoma and the other was immature teratoma.

Conclusion: Strategy for total removal is essential. Chemotherapy is required for the case with immature teratoma.

Keywords: teratoma, cerebellum, infant, female

P-048

Posterior fossa tumor

Acquired torticollis related to nervous system pathologies in childhood.

Abdulfettah Tümtürk¹; Ekrem Ünal²; Selim Doğanay³; Ahmet Küçük¹; Hüseyin Per⁴; Sefer Kumandaş⁴; Ali Kurtsoy¹.

¹Erciyes University, Faculty of Medicine Department of Neurosurgery, Kayseri; ²Pediatric Hematology and Oncology, Erciyes University, Faculty of Medicine, Department of Pediatrics, Division of Pediatric Hematology & Oncology, Kayseri; ³Erciyes University, Faculty of Medicine, Department of Radiology, Division of Pediatric Radiology,

Kayseri; ⁴University, Faculty of Medicine, Department of Pediatrics, Division of Pediatric Neurology, Kayseri

Objective: Torticollis may be the first symptom of the diseases of posterior cranial fossa and cervical spinal cord. The aim of this study is to call attention to the underlying CNS disease of the childhood in the children presented with torticollis.

Methods: We aimed to evaluate the clinical features of the children presented with torticollis in the last ten years. Clinical records of 27 cases both diagnosed or treated for CNS disease between 2006- 2015 were retrospectively reviewed.

Results: We report 27 cases of pediatric CNS diseases presented with torticollis. (12 girls and 15 boys with the median age of 8 years, ranging 3 months to 12 years). The identified etiologies of the enrolled children were cerebellar tumors in seven, pontine glioma in one, brain stem gliomas in five, an astrocytoma of the spinal cord located between C1 and C6 cervical vertebrae in one, acute disseminated encephalomyelitis in one, spontaneous spinal epidural hematoma in one, cervical osteoblastomas in two, aneurysm of the anterior communicating artery in one, arteriovenous malformation in one, eosinophilic granulomas in two, aneurysmal bone cysts in one, neuroenteric cyst of the spinal cord in one, Chiari type 3 malformation in one, arachnoid cysts causing brainstem compression in two, cerebellar empyema in one. Twenty-one of the patients underwent surgery. The parents of three patients refused surgery. Medical treatment was performed in other three patients.

Conclusion: Acquired torticollis is not a diagnosis but rather a sign of an underlying disorder. Various underlying disorders from relatively benign to life-threatening conditions may present with torticollis. CNS diseases have an important place among them. The first step should always be a careful and complete physical examination, which must include all the systems. Imaging must be performed, in particular, if neurological symptoms exist. Early diagnosis of these disorders will reduce mortality and morbidity.

Keywords: Acquired torticollis, Posterior fossa tumors, spinal tumors, Children, differential diagnosis

P-049

Posterior fossa tumor

Unusual spread of a medulloblastoma

Kate Poulgrain¹; Andrew Hunn¹.

¹Royal Hobart Hospital, Hobart

Objective: Medulloblastomas are the most common malignant brain tumour in the paediatric population. They represent the most common embryonal tumour and account for 20% of all paediatric CNS tumours. Treatment centres around surgical resection followed by adjuvant chemoradiotherapy. Recurrences are more likely to happen within the first 2 years, and commonly are associated with dissemination through the CSF. Described sites of systemic spread are bone / bone marrow, lymph nodes, lungs /pleura and liver. We present a case of a rare presentation of spread of a medulloblastoma to the skin.

Methods: A literature review of spread of medulloblastoma to the skin yielded one case report of medulloblastoma metastasizing to the cheek.

Results: Medulloblastoma disseminates frequently in the cerebrospinal fluid and associated areas. There are also several published reports of spread to bone. Spread to the skin however is a rare occurrence. Given this mode of dissemination, consideration may need to be given to surgical removal, similar to the principles that govern bony sarcoma resection.

Conclusion: Skin metastasis of medulloblastoma is a rare occurrence and should prompt consideration of surgical resection technique.

Keywords: Medulloblastoma, Dissemination, Metastasis

P-050

Posterior fossa tumor

Management of MB in children: 85 cases report from the Department of Pediatric Neurosurgery, Xinhua Hospital
Zixuan Wang¹; Jie Ma¹.¹Shanghai Xinhua Hospital, Shanghai

Objective: According to the 2014 Childhood Cancer Statistics, brain and CNS cancer has the second highest incidence rate and the highest mortality rates. Medulloblastoma, the most common CNS Embryonal tumor, is also the most common malignant solid tumor in children. In this report, we'd like to talk about the management of MB by presenting 85 cases we collected.

Methods: The data of clinical course were obtained and carefully reviewed from 85 cases of MB, which accepted operation procedure from Jan. 1st 2006 to Mar. 18 2015. We conclude the clinical characteristics of these cases: sex, age, tumor location, clinical features, operative approach route, resection degree, prognosis, and the effect of radiation and chemo-therapy, etc.

Results: Among all 85 cases, average sex is about 4.3 years old. There are 61 male and 24 female. Almost all tumors locate at IV ventricle or cerebellum. Most tumor present to be solid. 5 out of 85 cases have metastasis. The tumors were approached mainly by 2 routes, which will be presented in detail in the paper. In 70 cases, the tumors were totally removed. Overall survival was 56%, which is 48 out of 85 patients, 22 patients accepted radiation or chemo-therapy, 9 patients cannot be contacted.

Conclusion: MB is a malignant, high invasive embryonal CNS tumor with low average age of onset, mostly affecting male children. MB is most solid and locate in IV ventricle or cerebellum. The most common approach routes are through vermis cerebelli and cerebellar medullary fissure. Five-year event-free survival was 43%, overall survival was 56%. Most patients accepted radiation or chemo-therapy and have a relatively optimistic prognosis, 18 out of 22 experienced 5-year event-free survival.

Keywords: medulloblastoma, CNS tumor, surgery, children, pediatric

P-051

Posterior fossa tumor

Goldenhar Syndrome and medulloblastomaCristian Bravo Garcia¹; Santiago Candela Canto¹; Antonio Guillen Quesada¹; Yislenz Narvaez Martínez¹; Elena D'avella¹; Enrique Ferrer Rodriguez¹.¹Hospital Sant Joan De Déu, Barcelona

Objective: We describe the presentation of medulloblastoma in a patient with Goldenhar Syndrome (GS) and the likely association between them.

Methods: We aim to describe the second case reported in literature. Because this is a rare association we also provide a brief literature review to supplement this case report.

Results: A 3-year-old boy diagnosed with GS was admitted to the hospital with 1 month history of cerebellar ataxia and vomiting with no other neurological symptoms complaints. A CT scan showed a heterogeneous tumor located in the 4th ventricle with acute onset hydrocephalus which required an urgent external ventricular drain placement. The MRI showed a contrast enhancing 3.2 cm diameter solid tumor, located on the floor of the 4th ventricle with spinal leptomeningeal dissemination. A transvermian approach was performed on the next day with subtotal resection following a ventriculo-peritoneal shunt placement. Histopathologically the tumor was diagnosed as classic medulloblastoma. He is awaiting for oncologic treatment. The incidence of GS is between 1:35000 and 1:56000. It involves the first and second branchial arches. Most of the cases are sporadic and the exact etiology is yet unknown. Autosomal dominant and autosomal recessive modes of inheritance have

been suggested. The classic features of this syndrome include craniofacial, renal, cardiac and vertebral abnormalities and it can also be associated with other tumors such as the medulloblastoma described previously by Aizenbud et al. The identification of cancer stem cell populations, termed brain tumor-initiating cells in medulloblastoma, has provided novel cellular targets for the study of aberrantly activated signaling pathways such as Shh and Wnt which are also implicated in the development of the first and second branchial arches.

Conclusion: There is a key point association between GS and medulloblastoma. This association depends on the molecular basis of these two pathologies through the Shh and the Wnt pathways.

Keywords: goldenhar, medulloblastoma, sonic hedgehog

P-052

Hydrocephalus

Alternate physiological CSF outflow in premature newborns hydrocephalusVolkodav Oleg¹; Zinchenko Sveta¹.¹Crimean Medical University, Simferopol

Objective: Pediatric neurosurgery actual task is the CSF outflow correction due to secondary post-hemorrhagic hydrocephalus combined non-traumatic (spontaneous intraventricular-subarachnoid hemorrhage) and traumatic genesis.

Methods: Premature newborns post-hemorrhagic hydrocephalus neurosurgical treatment method was suggested and special ventricular-subarachnoid stent (VSS) was invented and proved (Certificate in Copyright Law, pattern № 45865 from 02.10.2012). Its algorithm including subgaleal ventricular-subarachnoid drainage with following shunting via VSS to restore the physiological liquor circulation.

Results: Twenty-four VSS operations to low-birth-weight newborns with post-hemorrhagic hydrocephalus were performed in two modifications. Invented VSS we are used in premature newborns with minimal body mass 1180gr (28 weeks of gestation). In deep premature newborns with extreme low weight (minimal newborn body mass on the moment of operation consist of 520gr and 25 weeks of gestation) we are used the modify silicon perforate tube from VSS. Post-operative quantity criteria include: neurological anamnesis, ophthalmoscopy, neuro-imaging (neurosonography, brain CT-scan and MRI). Subgaleal ventricular-subarachnoid drainage (Certificate in Copyright Law, pattern № 34523 from 11.08.2010) via VSS supported primary effective liquor purification from blood (simultaneous ventricles and subarachnoid space) and intracranial pressure correction. Following CSF ventricular-subarachnoid shunting (Certificate in Copyright Law, pattern № 38061, 20.04.2011) support effective liquor outflow from lateral ventricles to subarachnoid space for it physiological absorption.

Conclusion: VSS let to restore the physiological CSF outflow alternative to ventricular-peritoneal shunting with shunt-depend condition and high risk of it dysfunction and decrease time of recovery treatment.

Keywords: premature newborns, hydrocephalus, ventricular-subarachnoid stent

P-053

Hydrocephalus

Alternate physiological CSF outflow in neonatal intra-cerebral hemorrhageVolkodav Oleg¹; Zinchenko Sveta¹.¹Crimean Medical University, Simferopol

Objective: Neonatal intra-cerebral hemorrhage high fibrinolytic activity with altered spontaneous hemostasis in the hematoma region lead sometime to it invasion with lateral ventricle and subarachnoid space communication (i.e. porencephalus).

Methods: Neonatal intra-cerebral hematoma surgical treatment method was suggested and proved (Certificate in Copyright Law, patent 38063, 20.04.2011). Its algorithm including porencephalus model after hematoma removal with simultaneous effective purification from blood ventricles and subarachnoid space and further physiological CSF outflow via invented ventricular-subarachnoid stent (VSS), Certificate in Copyright Law, N 45865 from 02.10.2012.

Results: Twelve emergency neurosurgical operations VSS were performed to the low-birth-weight newborns with intra-cerebral hematomas (minimal newborn body mass on the moment of operation consist of 1460gr and 29 weeks of gestation). Indications to the surgery are: rough hemisphere compression by the hematoma with midbrain dislocation more than 5mm, basal cistern deformation, homo-lateral ventricle compression with contra-lateral hydrocephalus. Neuro-imaging: neurosonography, brain CT-scan and MRI. After adequate intra-cerebral hematoma decompression, special drainage from VSS input to hematoma cavity for external CSF purification hematoma remnant and it communication with ventricles and subarachnoid space.

Conclusion: Following VSS with physiological CSF outflow from the hematoma cavity after external drainage removal (pure liquor) let to reduce the multifocal occlusive hydrocephalus risk and improve newborns rehabilitation.

Keywords: neonatal intra-cerebral hemorrhage, ventricular-subarachnoid stent

P-055

Hydrocephalus

Distal shunt failure due to obesity: a rare cause of shunt malfunction Vaner Köksal¹; Tugba Morali Guler².

¹Recep Tayyip Erdogan University Medical School, Rize; ²Karabuk Government Hospital, Karabuk

Objective: Shunt obstruction is one of the mechanical problems of shunts and it can occur in any part of the shunt system. The ventricular part of the catheter and the valve of shunt are the most common obstructed sides. Obstruction of the distal part of the shunt catheter is rarely seen. It can be obstructed by strangulation of catheter or peritoneal adhesions. Catheter obstruction with peritoneal fat tissue especially in the use of distal slit catheters is also reported. But there is no catheter obstruction reported for the distal catheters with multiple pores which are used nowadays in the literature. A distal shunt malfunction due to obstruction by adipose tissue in an obese patient is discussed in this case.

Methods: A 55-year old female 116 kg in weight and 150 cm in height admitted to the hospital with hydrocephalus.

Results: The patient went to revision for six times after the first shunt operation in 5 years time. The clinical findings while admitting to the hospital with shunt malfunction were always visual disturbance (double vision) and headache related to the intracranial hypertension. Generalized epileptic seizures were also occurred. In the last attack of shunt malfunction clinical deterioration was so fast and the patient was admitted to the hospital with transtentorial herniation. Emergent surgery was done and when the elements of shunt system was checked, it was seen that the obstruction site was again the distal catheter with omental fat tissue obstructing it. The distal catheter was made to work by replacing it in a different place in periton.

Conclusion: Shunt failure due to proximal catheter obstruction is a frequently seen complication. However distal shunt catheter obstruction with fat tissue is a rarely seen situation it can cause life threatening clinical results such as herniation and progressive clinical deterioration.

Keywords: distal shunt obstruction, hydrocephalus, obesity, shunt malfunction

P-056

Hydrocephalus

Endoscopic treatment of a pediatric colloid cyst

Assen Bussarsky¹; Kiril Georgiev¹.

¹Dept. of Neurosurgery, Univ. Hospital "St. Ivan Rilski", Medical Univeristy - Sofia, Sofia

Objective: Colloid cysts in children below 10 years are extremely rare and present both diagnostic and therapeutic challenge. Most reported cases underwent open microsurgical excision. We present a case of endoscopic removal of a pediatric colloid cyst

Methods: A 8 years old girl presented with a history of intermittent headaches with a duration of 1.5 years. In the last week before admission the symptoms worsened progressively with constant headache, vomiting and blurred vision. After CT and MR the patient was referred to our department with the diagnosis of a "glial tumor" in the third ventricle. On imaging studies biventricular hydrocephalus due to a colloid cyst in the anterior third ventricle was present.

Results: The child underwent endoscopic resection of the lesion with complete resolution of symptoms and without newly acquired deficit. Follow-up MRI showed complete removal of the lesion.

Conclusion: Endoscopic resection of colloid cysts is a viable option even in the pediatric patients

Keywords: colloid cyst, endoscopy, pediatric

P-057

Hydrocephalus

Retroperitoneal transluteal migration of distal lumboperitoneal shunt catheter: an unusual complication of cerebrospinal fluid shunt.

Case report

Jivko Surchev¹; Kiril Georgiev¹; Alaa William¹.

¹Medical University of Sofia, Sofia

Objective: We present an unusually rare complication – migration of the distal lumboperitoneal shunt catheter through the retroperitoneum and the gluteal muscles to the hypoderma where it imitates hypodermal abscess.

Methods: A female child, shunted at the age of 4 years and 3 months with lumboperitoneal shunt. A total of 30 months after shunt implantation, a swelling in the left gluteus occurred, 4 cm aside of the middle line of the os coccygis projection level. After incising a doubted subcutaneous abscess, at the bottom of which found the tip of the distal catheter. No fluid was flowing through it, which is a sign of dysfunctioning valve. The abdominal x-ray and CT showed correct positioning of spinal catheter and valve. The peritoneal catheter passes through Foramen ishiadicum majus enters the retroperitoneal area and through the gluteal muscles reaches the hypodermal area.

Results: We present pictures of the physical examination and the x-ray and CT, which determined the diagnosis (the way in which the catheter's tip traverses from the peritoneal cavity to the hypoderma under gluteal area). Among the peritoneal CSF shunts the perforations with migration of the tip of the catheter are relatively rare complications. Perforations to hollow organs with protrusion through anus, vagina, scrotum, urethra, vessels, thoracic cage are most common. Only one similar case of penetration through the muscles and the tip of the catheter reaching the hypoderma area at the level of lower thoracic vertebrae paraspinal is described (Greene Jr., C. Valerie. *Pediatr Neurosurg* 2013; 49:86-88).

Conclusion: Despite the continuous improvement of the material of flexible silicone catheters, the injury of hollow organs with subsequent migration to unusual areas still occurs. Our case is the only one in which the catheter makes its way through the retroperitoneum and the gluteal muscles.

Keywords: CSF shunt, transgluteal migration, catheter migration, complications

P-058

Hydrocephalus

Macrocephaly-capillary malformation (M-CM) syndrome. Clinical management in our institution.

Alain Flor-Goikoetxea Gamo¹; Mariana Alamar Abril²; Santiago Candela Canto²; Patricia Puerta Roldan²; Antonio Guillén Quesada²; Enrique Ferrer Rodriguez².

¹Hospital Clínico Universitario Lozano Blesa, Zaragoza; ²Hospital Sant Joan De Déu, Barcelona

Objective: Macrocephaly-capillary malformation síndrome (M-CM) is rare genetic malformation syndrome, with about 150 cases published in the literature. The disorder is characterized by abnormal body and head overgrowth and cutaneous, vascular, neurologic, and limb abnormalities. The neurologic abnormalities can be congenital, such as cortical dysplasia or polymicrogyria; or aquired, such as ventriculomegaly/hydrocephalus or tonsillar herniation. The objective of this work is to assess the clinical management of this patients in our institution.

Methods: Since 2001, 4 patients with M-CM associated hydrocephalus have been operated. All of them diagnosed within the ages from 6 to 8 months by neuroimaging (CT scan, MRI and transfontanelar ultrasound). All of them with tonsillar herniation. Two of them were treated with ventricle peritoneal shunt, and the other two by endoscopic third ventriculostomy (ETV).

Results: One of the shunted patients died two years after diagnosis of postoperative complications after a shunt revision. The other shunted patient is controlled in the outpatient clinic (fundoscopy and neuroimaging). The ETV cases are followed with MRI and fundoscopy every 6 months. The tonsillar herniation has not progressed in any case.

Conclusion: ETV is the elective treatment for M-CM associated hydrocephalus, provided that serial MRI could be performed during the follow up, to assess its performance.

Keywords: M-CM, Hydrocephalus, ETV

P-059

Hydrocephalus

A rare cause of late shunt dysfunction due to ripping of proximal ventricular catheter

Burak Kinali¹; Volkan Murat Ünal¹; Senem Alkan Özdemir²; Ali Karadağ¹; Nail Özdemir¹.

¹Tepecik Research and Education Hospital, Neurosurgery, Izmir; ²Dr. Behçet Uz Children's Hospital, Neonatology, Izmir

Objective: Despite the progress in shunt technology there is no significant change in complication rates, especially in pediatric patients. These complications occurs either by corruption of entirety of the system or by obstruction of the catheters. In our case we encountered a late complication due to a small cut on the proximal (ventricular) catheter causing patient to have clinical symptoms with slit ventricle.

Methods: A 6 year old female patient who had ventriculoperitoneal shunt operation admitted to our hospital with headache and vomiting. There were no abnormal neurological findings except lower extremity palsy

due to meningomyelocele history. In the Computer Tomography scans there were no progressive ventricular dilation or splitting of catheters from the pump. After observing patient with CT scan and clinical follow up, on the second admission with the same symptoms we decided to operate the patient for a VP shunt revision as the pump doesn't function and the symptoms persist.

Results: During the operation we saw that there is a cut on the proximal part of ventricular catheter that probably occurred during the connection between proximal catheter and the pump in the first operation. We decided to replace both the pump and proximal part. Patient's symptoms resolved with the renewal. There were no complication during the 2 month follow-up.

Conclusion: We believe that small cut that occurred on the junction of ventricular catheter with the pump caused the shunt function improperly causing slit ventricle in the chronic phase with persisting headache and vomiting.

Keywords: slit ventricle, shunt dysfunction, catheter, hydrocephalus

P-060

Hydrocephalus

Advanced shunting in complex hydrocephalus patients. Rambam experience.

Sergey Abeshaus¹; Leon Levi¹; Joseph Guilburd¹; Menashe Zaaroor¹.

¹Rambam Health Care Campus, Haifa

Objective: Ventricular shunt is the most common neurosurgical procedure. In most of the cases it is a straight forward and rather simple for any experienced neurosurgeon. We found in our series that less than 5% of cases are complex and require a special experience and sophisticated armamentarium to ensure successful outcome for a patient. We would like to share our experience in management of complex hydrocephalus cases.

Methods: All patients were treated at Rambam HCC. Preoperative and post operative imaging obtained. AXIEM Medtronic neuronavigation was used in all cases. Codman antibiotic impregnated tubing and Medtronic Strata adjustable valves were utilized for shunting procedure. Senior pediatric neurosurgeon took a part in preoperative planning and during procedure in all presented cases. All patients had an inpatient and outpatient clinic follow up with necessary surveillance imaging.

Results: Our series shows comparable with uncomplicated CSF diversion procedures shunt survival rate.

Conclusion: Both neurosurgical literature reports and our institution experience support a better shunt survival with a sophisticated approach to complex hydrocephalus patients.

Keywords: complex hydrocephalus, neuronavigation

P-061

Hydrocephalus

The under drainage problems of low-pressure hydrocephalus patients: a clinical study

Vaner Köksal¹.

¹Recep Tayyip Erdogan University Medical School, Rize

Objective: Normal pressure hydrocephalus (NPH) is one of the few causes of reversible dementia. In this study, the low-pressure hydrocephalus cases with shunt systems working improperly or problems about the pressure settings of the valves are mentioned.

Methods: Patients presenting with the clinical signs and symptoms of spontaneous normal-pressure hydrocephalus (NPH) were selected between the years 2008-2015. There was no other history of cerebral pathology in these cases. Two times lumbar puncture (LP) for CSF drainage was performed firstly and then CSF pressures of the cases were measured

during LPs. CSF drainage amount of approximately 40–50 milliliter was provided. Patients who benefit from after LP were diagnosed as NPH and also according to CSF pressures required shunt valve was decided. Cases were divided into two main groups according to the CSF pressure: CSF pressure is higher than and lower than 100 mm H₂O pressure.

Results: The VP shunt surgery was performed to 28 patients with NPH. 17 of the patients were male and 11 were female. The ages of the patients 65 and 81. The mean age was 74.6'i. The standard medium pressure reservoir valve was the first choice in 21 patients. The adjustable valves were preferred in 7 patients. The mean follow-up time was 2 years. Standard medium pressure valve insufficiency occurred in 3 patients during follow-up, they were replaced with the adjustable valves. Also insufficient drainage was occurred in two cases with adjustable valves despite excessive reduction of pressure settings of the valves.

Conclusion: The decreasing elasticity of the brain especially after seventh decade makes the neural tissue easily affected even from minimal pressure increments and as a result classical triad findings occur. Gravity helps to remove the excess CSF from the intracranial compartment. Therefore, shunt is good choice.

Keywords: hydrocephalus, low pressure, malfunction, under drainage

P-063

Hydrocephalus

Dandy-Walker Variant posterior cyst fenestration using a flexible endoscope: technical nuances.

John Emelifeonwu¹; Drahoslav Sokol¹; Chandrasekaran Kaliaperumal¹; Pasquale Gallo¹.

¹Royal Hospital for Sick Children, Edinburgh

Objective: Hydrocephalus occurs in approximately 25% of Dandy-Walker Variant (DWV) patients and the optimal surgical management remains contentious. However, when associated with obstruction of the Sylvian aqueduct, the conventional surgical principle is that of supratentorial hydrocephalus treatment with an endoscopic third ventriculostomy (ETV) or with a ventriculo-peritoneal shunt (VPS) insertion plus posterior fossa drainage by a cysto-peritoneal shunt. The aim of this case presentation is to highlight an alternative treatment option of posterior fossa endoscopic cyst fenestration using the flexible endoscope.

Methods: A two-month old boy presented with macrocephaly and signs of raised intracranial pressure. An MRI scan revealed a DWV posterior fossa cyst with effacement of the Sylvian aqueduct, hypoplasia of the vermis and cerebellar hemispheres and associated hydrocephalus. The hydrocephalus was initially managed with an ETV, which failed, prompting insertion of a VPS. This was followed by a staged endoscopic fenestration of the posterior fossa cyst using a flexible endoscope through a single right occipital burr-hole. Technical nuances of the endoscopic technique are highlighted.

Results: The post-operative MRI scan demonstrated a reduction in sizes of the cyst and a remarkable decrease in the mass effect on the brainstem with a reopened Sylvian aqueduct. The patient made an excellent and quick recovery after the endoscopic procedure.

Conclusion: The endoscopic cyst fenestration using a flexible endoscope through a posterior fossa approach is an effective minimally invasive technique in the treatment of a DWV cyst with aqueductal stenosis that negates the risks and morbidities associated with cysto-peritoneal shunts in this patients group. The maneuverability of the flexible endoscope permits a clear visualization of the cyst and peri-cyst anatomy, therefore allowing for a safe and adequate fenestration.

Keywords: Dandy Walker Malformation, Hydrocephalus, Neuroendoscopy, Surgical Technique

P-064

Hydrocephalus

Hydrocephalus in aqueductal stenosis – a retrospective analysis and proposal of subtype classification

Preuss Matthias¹.

¹University Leipzig, Leipzig

Objective: Treatment of aqueductal stenosis (AQS) has underwent several paradigm shifts during the past decades. Currently endoscopic ventriculostomy (ETV) is recommended as treatment of choice. Several authors have addressed the issue of variable ETV success rates depending on age and pathogenetic factors. Aim of the study was a retrospective analysis of potential types of AQS and comparison of the treatment response after ETV or VP-Shunt.

Methods: 100 patients (median age 8.0 years, range 0–79 years) have been treated for MRI-proven aqueductal stenosis. Initial signs & symptoms, type of surgery, complications and follow-up of the patients were analyzed. Treatment success was defined by neurological improvement, head circumference in childhood and radiological response by Evans ratio.

Results: Four Types of AQS can be defined with distinct age ranges and typical signs & symptoms: congenital Type-I, juvenile chronic (tectal tumor-like) Type-II, adult acute Type-III, adult chronic (normal pressure hydrocephalus-like) Type-IV. Type I (<2 years of age) is more successfully treated with VP-Shunts than with ETV (87% vs. 50%), adult acute Type III responded excellent to ETV. Treatment of chronic-juvenile Type II by ETV resulted in similar neurological success in 81.9% but showed radiological improvement in only 19% compared to 64% after VP-Shunt. There has been no influence of persistent ventriculomegaly in Type II after ETV in contrast to VP-Shunt therapy during short-term follow-up of five years. Type IV patients responded neurologically in 70% after ETV, but radiological response was low (5%).

Conclusion: AQS can be divided into four distinct age groups and types in regards of clinical course and symptomatology. Depending on the type, ETV cannot be unequivocally recommended. Congenital Type-I AQS seems to have better outcome with VP-Shunts whereas acute adult Type-III offers excellent ETV results. Juvenile chronic Type II still requires prospective investigation of long-term ETV outcome, especially when ventriculomegaly persists.

Keywords: aqueductal stenosis, ETV, VP-shunt

P-065

Hydrocephalus

Magnetic resonance elastography in the evaluation of patients with slit ventricle syndrome

Adam Sandler¹; Kristy Tan¹; Alex Krolick¹; Cecile Yama¹; Rick Abbott¹; James Goodrich¹; Avital Meiri¹; Pat Mcallister²; David Limbrick²; Philip Bayly²; Mark Wagshul¹.

¹Albert Einstein College of Medicine/ Montefiore Medical Cent, Bronx;

²Washington University in St. Louis School of Medicine, St. Louis

Objective: Alterations in the compliance of the brain may account for the variety of symptoms experienced by chronically-shunted hydrocephalic patients with small ventricles and functioning shunts. In addition, changes in peri-ventricular compliance have been proposed to explain the observed failure of ventricles of some shunted patients to enlarge during shunt malfunction. Clinical evaluation and management of severe headache disorders in shunted patients with small ventricles remains a challenge, as routine imaging techniques neither provide treatment nor elucidate the underlying compliance-related mechanisms of headache in these patients. Magnetic Resonance Elastography (MRE), a non-invasive imaging of intracranial compliance, is a new method which can provide much needed progress in treating these patients.

Methods: Shunt-dependent patients shunted in infancy for hydrocephalus, who suffer from chronic debilitating headaches and, possess small/slit ventricles, were selected for brain MRI including magnetic resonance elastography (MRE). Healthy controls had no history of hydrocephalus or other neurological disorders. MRE images were used to calculate the regional compliance by inducing a very-low amplitude (~100 microns) external mechanical wave transmitted through the zygomatic arches and the wave propagation speed used to calculate stiffness.

Results: Figure 1 show MRE results from a typical patient dataset. Figure 1(b) shows the compliance or stiffness map and shows good anatomical correlation with the ventricular structure as seen in the high-resolution anatomical image (Figure 1(a)). Motion maps show good mechanical wave penetration throughout the brain

Conclusion: In its capacity to measure changes in cerebral compliance in shunted patients, and thereby to test the accuracy of long-standing theories of shunt-related disorders related to compliance, Magnetic Resonance Elastography shows great promise to assess the long-term effects of shunting on the biomechanical properties of the human brain.

Keywords: hydrocephalus, slit ventricle syndrome, compliance, elastography, headache

P-066

Hydrocephalus

Does time matter in VPS after germinal matrix hemorrhage in premature infants

Basak Caner Topkoru¹; Nejat Isik¹; Recep Basaran¹; Caglar Bozdogan¹; Ilke Mungan Akin²; Sibel Sevuk Ozumut²; Derya Buyukkayhan².

¹Istanbul Medeniyet University Goztepe Education and Research Hospital Neurosurgery Department, Istanbul; ²Medeniyet University Goztepe Educational and Research Hospital Pediatric Department, Istanbul

Objective: The risk of intraventricular hemorrhage is high in premature infants with low birth weight because of the insufficient germinal matrix tissue. In some of those infants hydrocephalus can be encountered because of the permanent or temporary impairment in the CSF circulation due to blood products. In this study we aimed to evaluate our posthemorrhagic hydrocephalus cases where we had to insert ventriculoperitoneal shunt (VPS).

Methods: We have retrospectively reviewed the cases who were admitted to the Newborn ICU and operated by the Neurosurgery Clinic in Istanbul Medeniyet University Goztepe Education and Research Hospital between January 2010 and January 2015.

Results: There were 10 premature babies (2 girls, 8 boys), where VPS was inserted due to posthemorrhagic hydrocephalus. Their mean birth weeks and birth weights were $30 \pm 1,8$ weeks and $1372,5 \pm 357,7$ gr respectively. 7 (70%) of these patients (Group 1) were born and followed in the Newborn ICU in various other hospitals in Istanbul and sent to our Newborn Clinic at average 55th $\pm 29,1$ days postnatally. There were no statistically significant difference between two groups regarding the gestational weeks and birth weeks. But there were significant difference between the groups regarding the average hospital stay and time from birth to VPS insertion time ($p < 0,05$). Concerning the associated prematurity complications (bronchopulmonary dysplasia (BPD), retinopathy of prematurity (ROP)) there were no Grade 2 and higher ROP or medium-severe BPD in the second Group. In Group 1 3 patients were discharged with O2 support and 2 patients required laser photocoagulation. There was no mortality.

Conclusion: Premature newborns under 32 weeks with hemodynamic instability are under risk of severe germinal matrix hemorrhage. Close follow-up of these patients is necessary because early external ventricular drainage or VPS insertion can improve neuromotor evolution. These patients should be closely monitored for the prematurity complications.

Keywords: hydrocephalus, ventriculoperitoneal shunt, germinal matrix hemorrhage

P-067

Hydrocephalus

Long tunnelled external ventricular drain as long-term treatment of hydrocephalus in a child with recurrent shunt malfunctions

John Emelifeonwu¹; Drahoslav Sokol¹; Pasquale Gallo¹; Jothy Kandasmy¹; Chandrasekaran Kaliaperumal¹.

¹Royal Hospital for Sick Children, Edinburgh

Objective: Hydrocephalus is commonly associated with intracranial tumours and is due to disruptions in the CSF pathways. Internalised CSF diversion shunts are often required but are prone to malfunction especially when CSF protein is high. EVDs are considered short-term alternatives particularly because of the high rates of infection associated with long-term use. We report a case of long-term, long-tunneled EVD as treatment of hydrocephalus in a child with an astrocytoma and high protein containing CSF.

Methods: A four-month old boy was admitted to hospital with failure-to-thrive and disconjugate eye movements. An MRI scan revealed a hypothalamic tumour infiltrating the third ventricle with associated hydrocephalus. A biopsy confirmed a WHO grade 1 pilocytic astrocytoma and his parents declined resection because of the risks. Hydrocephalus was initially managed with a VP shunt which required multiple revisions. It was converted to long-term, antibiotic-impregnated long-tunneled EVD (LTEVD) with parental education about management.

Results: 14 internalised shunt revisions (11 VP and 3 VA) were performed in 3 years. His CSF protein-content was persistently high during revisions. LTEVD has been used for 10 months and managed almost exclusively by his parents in the community. No infections have occurred during this period. On several occasions LTEVD blockages were resolved by aspirating the catheter with a syringe. This has more frequently been performed by parents. Surgical revision has been needed on only 3 occasions. The longest duration of an LTEVD was 6 months.

Conclusion: With parental motivation and education, antibiotic-impregnated LTEVD is a long-term management option of hydrocephalus following multiple shunt malfunctions. LTEVD have lower rates of complications such as displacement and infection when compared to the standard short-tunneled EVDs. LTEVD malfunctions may be rectifiable by interrogating the externalised catheter, thus negating the risks and morbidities associated with general anaesthetic and invasive surgical exploration of internalised shunts.

Keywords: Hydrocephalus, intracranial tumours, long-tunneled EVD

P-068

Hydrocephalus

Congenital hydrocephalus: results of a consecutive series

Recep Basaran¹; Basak Caner Topkoru¹; Nejat Isik¹; Naci Balak¹; Sibel Sevuk Ozumut²; Ilke Mungan Akin².

¹Istanbul Medeniyet University Goztepe Education and Research Hospital Department of Neurosurgery, Istanbul; ²Istanbul Medeniyet University Goztepe Education and Research Hospital Department of Pediatrics, Istanbul

Objective: Hydrocephalus is a complicated disorder due to impaired CSF circulation. Among the most encountered reasons of neonatal hydrocephalus are congenital hydrocephalus (CH) (aqueduct stenosis, Dandy Walker malformation, Arnold Chiari malformation), spinal disraphism, meningitis, ventriculitis, newborn sepsis, intracranial hemorrhages. The incidence of real congenital hydrocephalus is 1-1.5 in 1000 live births. We aimed to assess the cases with congenital hydrocephalus.

Methods: We have retrospectively reviewed the cases with congenital hydrocephalus who were admitted to the Newborn ICU and operated by the Neurosurgery Clinic in Istanbul Medeniyet University Goztepe

Education and Research Hospital between January 2010 and January 2015.

Results: There were 14 patients (7 girls, 7 boys) in a 5 year period. All cases were diagnosed in the antenatal period and abortion was denied. The average gestation weeks, birth weight and head circumferences were $37,8 \pm 1.6$ weeks, $3544,1 \pm 1242,3$ gr and 47 ± 4 cm, respectively. The median operation day was 4 and the average hospital stay was 29 ± 9 days. In two patients VPS had to be renewed due to shunt infections. All the patients were consulted by Pediatric Cardiology, Pediatric Neurology and Genetic Departments. 4 of the patients were syndromic. Two of the patients died.

Conclusion: The morbidity and mortality of the CH cases is high and the hospital stay is long despite the advanced diagnostic and treatment techniques. Infection is the most important factor affecting the long term survival.

Keywords: congenital hydrocephalus, ventriculoperitoneal shunting

P-069

Hydrocephalus

Factor X deficiency and hydrocephalus

Nesrin Akkoyun Kayran¹; İbrahim Alataş¹; Hüseyin Canaz¹.

¹Neurosurgeon, Istanbul

Objective: Background: Congenital factor x deficiency is the rare disorder among the coagulation disorders. Symptom severity varies in the affected people. If the factor x level is under 10%, it is categorized as severe. The disorder can cause hydrocephaly because of ventricular bleeding.

Methods: Case: The girl was diagnosed in the perinatal period as hydrocephalus and delivered by cesarean section following a full term pregnancy. Coagulation studies revealed severe factor x deficiency. After replacement therapy ventriculoperitoneal shunt was inserted. Computerized tomography (CT) scan of brain showed bilateral subdural hemorrhage after one month of operation. CT scan showed recovery after shunt revision. Then, because of recurrent obstruction, ventriculoperitoneal shunt revised five times more. The shunt had been obstructed related to the high cerebrospinal fluid protein levels. Following ventriculoperitoneal shunt disfunctions, endoscopic third ventriculostomy was performed and the signs and symptoms improved. Patient was died because of pneumonia.

Results: Discussion: Severe hemorrhagic problems can cause cerebral bleedings and assays reveal high cerebrospinal fluid protein levels. Although our patient suffered communicated hydrocephalus, she improved by endoscopic third ventriculostomy. We suggest endoscopic third ventriculostomy operation for the patients with high shunt disfunction probability. There is one case report on factor x deficiency with hydrocephalus. In that case the patient had not suffered with shunt disfunction. Ours is the first case who suffer recurrent ventricular bleeding and shunt disfunction.

Conclusion: We suggest initially endoscopic third ventriculostomy operation for the patients with sever factor x deficiency.

Keywords: factor x deficiency, hydrocephalus, endoskopik ventriculostomy

P-070

Hydrocephalus

The ShuntScope - new technique for catheter placement in complex cases of hydrocephalus in pediatric neurosurgery

Stefan Linsler¹; Sebastian Antes¹; Mohammed Salah¹; Joachim Oertel¹.

¹Saarland University Hospital, Department Of Neurosurgery, Homburg

Objective: The long-term preserving of ventriculoperitoneal shunt function depends on the correct placement of the ventricle catheter. Nearly 4% of ventricular catheters are misplaced. There are already many tools to optimize the insertion, such as neuronavigation or stereotactic techniques.

Methods: We used the new semirigid ShuntScope (Karl Storz, Germany) for the catheter placement in cases of slit ventricles, pseudotumor cerebri, cystic lesions and in cases of aqueductoplasty. The study was carried on 33 patients (19 males, 14 females), mean age 40 (5 to 78 years, 7 children). 24 patients presented with slit ventricles or difficult anatomic ventricular configurations. The ShuntScope was used diagnostic and therapeutic. The small outer diameter of 1mm allows to pass through the catheter under directly endoscopic view with a very good image resolution of 10 000 Pixels.

Results: The main value of this new technique was the ability to place the catheter tip accurately within distorted or small ventricles. Even controlled and guided placements in the fourth ventricle were possible in cases of aqueductoplasty. The semirigid endoscope allowed a safe intraoperative correction of the catheter placement due to a cover sheet function of the ventricular catheter. The postoperative neuroradiological imaging revealed the catheter tip placement analogous to the intraoperative findings and video recording analysis. No bleeding complications or infections occurred.

Conclusion: The ShuntScope is very useful for safe catheter placement in complex cases of hydrocephalus. With this new technique misplacements of shunt catheters are completely avoidable. The ShuntScope technique should be implemented in Pediatric Neurosurgery.

Keywords: shuntscope, hydrocephalus, vp shuntplacement

P-071

Hydrocephalus

Is radionuclide technique effective for assessment of ventriculo-peritoneal shunt patency?

Özgür Demir¹; Fatih Ersay Deniz¹; Erol Öksüz¹; Serdar Savaş Gül².

¹Gaziosmanpaşa University, Department of Neurosurgery, Tokat;

²Gaziosmanpaşa University, Department of Nuclear Medicine, Tokat

Objective: Ventriculo-peritoneal shunts are inserted to treat the symptoms of hydrocephalus. Hydrocephalus-related signs and symptoms that worsen or unimproved after shunt placement may indicate a malfunctioning or obstructed shunt. In this presented case we assessed the ventriculo-peritoneal shunt patency with radionuclide technique and showed the efficiency.

Methods: A newborn male baby presented with tense anterior fontanelle. Head circumference was 53 cm. Computed tomography (CT) showed large dilation of lateral ventricles with decreased parenchymal tissue (Figure 1). Surgical procedures were performed for treatment of hydrocephalus and meningocele in the same séance. Ventriculo-peritoneal shunt was inserted for the treatment of hydrocephalus. After the operation tense anterior fontanelle remained the same. Control CT revealed dilation of lateral ventricles with ventricular catheter inside (Figure 2).

Results: We used radionuclide technique to show shunt patency. The outer side of the reservoir was covered by a pliable material that could be depressed manually to pump the reservoir and punctured with a needle. Dietilentriaminpentaasetikası (Tc-99m DTPA), 0.5mCi (37MBq) was injected via reservoir by manually. 30-minute, 1 and 2-hours images were taken. 30-minute images showed the passage of radionuclide to the ventricular system via ventricular catheter (Figure 3). 1-hour images showed the passage of radionuclide to the peritoneal catheter (Figure 4). 2-hour images showed the passage of radionuclide to the peritoneal cavity via peritoneal catheter (Figure 5). The half-life of the radionuclide was measured as 5 minutes. The patency of the shunt system was found as intact.

Conclusion: We found that the shunt system of the patient was working. So the patient didn't undergo unnecessary reoperation. Radionuclide CSF shunt imaging can be used to determine whether the shunt is patent and to exclude shunt obstruction. This technique can also show the localization of shunt obstruction. We recommend this technique in the case of suspecting malfunctioning ventriculo-peritoneal shunt.

Keywords: Hydrocephalus, nuclear medicine

P-072

Hydrocephalus

Management of trapped fourth ventricle in a patient with chronic hydrocephalus and myelomeningocele.

Vanessa Hernandez-Hernandez¹; Mario García-Conde¹; Liberto Brage-Martin¹; Pablo Febles-García¹; Hector Roldan-Delgado¹; Victor Garcia-Marin¹.

¹Hospital Universitario De Canarias, La Laguna

Objective: Chronic hydrocephalus in children is a common entity. Those patients eventually develop an alteration of normal intraventricular anatomy leading to the occurrence of trapped ventricles secondary to the neof ormation of neomembranes or alteration in the cerebrospinal fluid flow.

Methods: We report the case of a 21 year old female with a surgical history of open myelomeningocele repair at birth. She suffered abrupt episodes of insidious and progressive headaches at the nuchal region that significantly limited her quality of life. CT of the brain showed tetraventricular dilatation with a strikingly large IV ventricle. An endoscopic procedure associated with neuronavigation and valvular review in the same surgical procedure was performed. We accessed the right frontal pole and a third ventriculostomy was performed. Nevertheless, no evidence of flow through the stoma was noticed. Following that, we explored the Sylvius Aqueduct observing a thin membrane that blocked it. We perforated this membrane and then put a multi-perforated catheter through lateral, third, and fourth ventricles and connected it with the previous shunt.

Results: 6 months post-operation, the patient was asymptomatic and the radiological control showed no hydrocephalus.

Conclusion: Endoscopic techniques allow an assessment of the intraventricular anatomy and they can alleviate an obstruction that is not due to a valve malfunction, with optimal results in patients with complex hydrocephalus associated with trapped ventricular cavities.

Keywords: hydrocephalus, myelomeningocele, trapped four ventricle, shunt failure, neuroendoscopy

P-073

Hydrocephalus

Acute intracranial hypertension caused by a choroid plexus cyst: case report and review of the literature

Gonzalo Lepe¹; Pieropaolo Frassanito²; Luca Massimi²; Massimo Caldarelli²; Giampero Tamburrini².

¹Hospital Universitario Lucus Augusti, Lugo; ²Institute of Neurosurgery, Catholic University Medical School, Rome

Objective: Choroid plexus cyst (CPC) are mostly occasionally discovered during routine prenatal ultrasound investigations, the majority of them gradually disappearing during pregnancy or the first year of life. Symptomatic cases usually manifest with chronic intermittent signs of increased intracranial pressure; an acute clinical onset has been only occasionally described.

Methods: We present the case of a 4-month-old boy who acutely presented the onset of drowsiness and irritability, with a tense anterior fontanelle.

Neuroradiological investigations (US, CT, MR) disclosed an intraventricular cyst obstructing both Monro foramina (sx>dx), with its major extension inside the third ventricle. At surgery (left frontal endoscopic approach) the cyst was found to arise from the left lateral ventricle choroid plexus; it was shrunk with the help of a thulium laser opening the access to the third ventricle. At the end of the procedure a preventive third-ventriculostomy was performed. Neurological conditions promptly improved postoperatively. A control US confirmed the resolution of the obstructive hydrocephalus.

Results: Choroid plexus cysts (CPC) usually are discovered during prenatal assessment. The CPC normally disappears, but when not occurs they don't usually produce clinical problems. Acute intracranial hypertension is a rare clinical presentation described few times in the literature. Endoscopic approach is a surgery election for the good results.

Conclusion: Acute hydrocephalus related to a CPC is a rare occurrence, the higher risk being related to anteriorly located lesions. 3D MR sequences can be helpful to establish the correct diagnosis and therefore the correct surgical management strategy.

Keywords: choroid plexus cyst, acute intracranial hypertension, hydrocephalus, endoscopic approach

P-075

Brain malformation

Endoscopic coagulation of choroid plexus in infants with hydranencephaly: safety and feasibility

Oscar García-González¹; Paola Hernandez-Ponce².

¹Hospital Regional De Alta Especialidad Del Bajío, León; ²Universidad Autonoma De Guadalajara, Guadalajara

Objective: This study explored the use of endoscopic coagulation of the choroid plexus (ECCP) for the treatment of hydranencephaly in two children

Methods: We retrospectively reviewed patients with hydranencephaly (n = 4) who underwent endoscopic choroid plexus coagulation. The four children underwent computed tomography or magnetic resonance imaging to confirm the presence and to measure the size of the choroid plexus before surgery. ECCP was performed through a right frontal burr hole. A rigid neuroendoscope and monopolar coagulator were used to achieve choroid plexus coagulation. Endoscopic management was considered successful if a shunt was not subsequently required.

Results: Endoscopic management was successful in the 4 patients (100%) who did not require a shunt over a median follow-up period of 12 months (range 6-18). There were no additional complications noted after endoscopic surgeries.

Conclusion: Endoscopic choroid plexus coagulation can enable some infants with low progressive and hopeless forms of hydranencephaly to avoid a ventriculoperitoneal shunt. Long-term watchful follow-up is mandatory after surgery.

Keywords: Hydranencephaly, Neuroendoscopy, Choroid Plexus, Electrofulguration

P-076

Brain malformation

Early surgical reparation of congenital aplasia cutis with autologous cranial vault graft in a child with adams oliver syndrome. Case report and literature review.

Santiago Candela¹; Rousinelle Da Silva¹; Mariana Alamar¹; Patricia Puerta¹; Gemma García-Fructuoso¹; Enrique Ferrer¹.

¹Department of Pediatric Neurosurgery. Hospital Sant Joan De Déu Barcelona, Barcelona

Objective: Adams Oliver Syndrome associates aplasia cutis and transverse defects in extremities. Aplasia cutis is a rare condition consisting in skin and sometimes periosteum, bone and dural defects usually in cranial vertex. Its management is controversial. The aim of this work is to report our experience with a single case.

Methods: We present a child affected of Adams Oliver Syndrome and we describe our management based on a literature review through Pubmed Medline.

Results: At birth it was observed a 6 x 3 cm skin and cranial defect with underlying thin duramater. She associated symmetrical distal phalanx hypoplasia in both feet and telangiectatical cutis marmorata. She also associated a contralateral nevus sebaceus. She was administered antibiotic profilaxis and the defect was covered with wet sterile gauze dressing till she was operated in the 6th day of life. The surgery consisted in closing the skin defect after covering the bone defect with an autologous bone graft obtained from the temporal region. There were no postoperative complications. The CT performed after one year from the surgery showed complete ossification of the cranial defect and of the donor site. Now there is a good cosmetic result with hair in all the surgical site. The literature review reveals that patients treated conservatively have increased risk of infection and hemorrhage and a longer hospital stay, while our experience with our patient coincides with the good results of the reports advocating early surgical reconstruction.

Conclusion: Early surgical reparation of congenital aplasia cutis is a good option in patients with extensive cranial defects because, compared to conservative treatment, it shortens hospital stay, reduces the risk of bleeding and infection and a better cosmetic result is obtained. Autologous cranial vault graft allows a quick and adequate ossification of the cranial defect.

Keywords: Aplasia cutis, Adams Oliver Syndrome, Cranial vault defect, Surgical reparation, Children

P-077

Brain malformation

Congenital left temporal large arachnoid cyst causing intraorbital optic nerve damage in the second decade of life

Cahit Kural¹; Marcel Kullmann¹; Martin U. Martin U.¹

¹Neurosurgery, Tübingen

Objective: Intracranial arachnoid cysts are congenital malformations which are frequently found in the middle fossa. They are generally asymptomatic lesions. We present a case of left temporal arachnoid cyst causing visual loss by intraorbital compression of the optic nerve

Methods: A girl was diagnosed with an arachnoid cyst at the age of nine months. The symptomatic cyst was observed over the years, last MRI was done at age 14y. She presented with loss of vision of the left eye and strong headaches at age of 16y. An ophthalmological examination revealed anopia of the upper quadrants of the left eye. Repeated examinations showed deterioration of field defects, so she was finally transferred to our clinic. MRI and CT showed complete erosion of the lateral orbital wall by the cyst with compression of the optic nerve at its entrance into the orbital canal. Microsurgical fenestration of the arachnoid cyst to the basal cisterns and exploration of the eroded orbital wall to rule out remaining bony laminae was done over temporal mini-craniotomy. CSF flow between the basal cisterns and arachnoid cyst was observed during the surgery

Results: Considerable improvement of the vision with decrease of visual field defects was observed in the early postoperative period. Headache was also relieved. Five months after the surgery, further visual improvement to complete restitution of optic nerve function had occurred. On MRI size of the cyst was unchanged as expected. However, an increase on the fatty tissue surrounding the optic nerve within the orbit and better delineation of the nerve at its entrance in the optical canal was noted

Conclusion: Even Long-standing asymptomatic arachnoid cysts may progress rapidly and cause neurological deficits by orbital erosion and

intraorbital optic nerve compression. These deficits may improve in a short time period after immediate surgical treatment

Keywords: Arachnoid cyst, Surgery, Fenestration, Loss of vision

P-078

Brain malformation

Acquired Type I Chiari malformation in 16-year-old girl

Sung-Won Jin¹; Jong-Il Choi¹; Sang-Dae Kim¹.

¹Korea University Ansan Hospital, Ansan

Objective: Chiari malformation first described two specific types of hind-brain deformities. Type I Chiari malformations are characterized by the presence of cerebellar tonsils in the upper cervical canal while the fourth ventricle remains above the foramen magnum. Type I malformations possibly arise during embryological development but have less certain predispositions than Type II malformations, which are associated with spina bifida. Previous reports have shown the presence of acquired Chiari malformation. They indicate a mechanism that remains unproved. We present a case of acquired Chiari malformation from head trauma after ten years without history of ventricular and lumbar shunting procedures.

Methods: A 16-year-old female patient was referred from department of ophthalmology with the papilledema in both eyes. It is considered raised intracranial pressure (ICP). She did not complain of headache. A brain magnetic resonance image (MRI) showed a Chiari I malformation with 9 mm of tonsillar herniation below the foramen magnum and hydrocephalus. Review of the past history revealed that she has small size epidural hematoma at posterior fossa that occurred after traffic accident, ten years ago. At that time, there was no evidence of Chiari I malformation in a brain MRI.

Results: The patient therefore underwent a suboccipital decompression and endoscopic third ventriculostomy (ETV). And then she has improved papilledema without any postoperative complications from the surgical procedure

Conclusion: In our case, we suggest that acquired Chiari malformation arose from downward displacement of the cerebellar structures was due to post-traumatic hydrocephalus and disorder of CSF circulation from posterior fossa caused herniation of cerebellar structures by epidural hematoma. Consequently, secondary hydrocephalus occurred.

Keywords: acquired chiari malformation, trauma, hydrocephalus

P-079

Brain malformation

Phace syndrome: a Korean case.

Ki Bum Sim¹; Chang Sub Lee¹; Sukh Que Park².

¹Jeju National University Hospital, Jeju; ²Soonchunhyang University Seoul Hospital, Seoul

Objective: The PHACE syndrome is a neurocutaneous syndrome that includes the following primary features: posterior fossa malformations of the brain, large facial hemangiomas, arterial anomalies, cardiac anomalies and aortic coarctation, and eye abnormalities. We document intracranial nonvascular and vascular abnormalities with large cervicofacial hemangioma to provide evidence that this patient fall within the spectrum of PHACE syndrome. Awareness of the relationship between these conditions is essential to ensure their proper management and to improve the survival rate of the patients with those anomalies.

Methods: A 23-month-old female patient was presented with giant cervicofacial hemangiomas which involved the right side of the head and neck and extended to the chest.

Results: Magnetic resonance imaging showed Dandy-Walker malformation, hydrocephalus, a right persistent trigeminal artery, aneurysmal dilatation in the right internal carotid artery (ICA), and both ICA hypoplasia.

Conclusion: On the basis of our experience with this patient and with others previously reported, we stress the importance of a multidisciplinary approach for all patients with large cervicofacial hemangiomas.

Keywords: PHACE syndrome, Infantile hemangioma

P-082

Brain malformation

Endoscopic fenestration strategy of bilateral temporal arachnoid cyst

Keun Soo Lee¹; Gi Chang Lee¹.

¹Paik Hospital, Busan

Objective: Endoscopic fenestration of arachnoid cyst is one of the best treatment options. It is a minimally invasive and effective. We report one side approach to treat bilateral temporal arachnoid cyst and discuss for treatment strategies.

Methods: After a mild head trauma, 10 year-old boy visit our hospital for headache. In the CT scan, we found the incidental finding of bilateral arachnoid cyst. After follow-up of 2 months, headache was not improved, so his parents want to treat for arachnoid cysts. We planned an endoscopic fenestration of arachnoid cyst to basal cistern and approached to the left side that was larger than right. We used 7mm diameter rigid endoscope. Through the basal cistern, we could enter the right arachnoid cyst. So both side arachnoid cysts were able to connect to the basal cistern in a single operation.

Results: We planned an endoscopic fenestration of arachnoid cyst to basal cistern and approached to the left side that was larger than right. We used 7mm diameter rigid endoscope. Through the basal cistern, we could enter the right arachnoid cyst. So both side arachnoid cysts were able to connect to the basal cistern in a single operation.

Conclusion: Through the basal cistern, we can approach to the bilateral MCA bifurcation area. So bilateral ICA aneurysms such as mirror aneurysms can clip in a single operation. Bilateral temporal arachnoid cyst could fenestrate to basal cistern in single operation in some cases.

Keywords: endoscope, bitemporal arachnoid cyst, basal cistern

P-083

Brain malformation

Alobar holoprosencephaly: an unusual case report and review of literature regarding the survival

Ömer Can Yıldız¹; Dieter Class¹; Raimund Firsching¹.

¹Universitätsklinikum Magdeburg, Klinik Für Neurochirurgie, Magdeburg

Objective: Holoprosencephaly (HPE) is a complex structural anomaly of the brain resulting from failed or incomplete cleavage of the forebrain into right and left hemisphere at the early gestation with a prevalence of 1/16000 live births and an incidence as high as 1 fetus affected in a number of 250. A classification is made based on the level and degree of failure of cleavage: Lobar HPE, semilobar HPE and alobar HPE where a single brain ventricle and no interhemispheric fissure can be seen. Alobar HPE is associated with a very poor prognosis with a 6 month survival rate being under 50% and a 1 year survival rate as low as 20%. However there are also some rare cases of alobar HPE with a prolonged survival documented. Here we present a patient, meanwhile 5 years old and review of the literature with analysis of survival rates of alobar holoprosencephaly.

Methods: The newborn was examined after birth in regular intervals and his records have been investigated thoroughly. The family has also been questioned. A systematic search was done in MEDLINE, Web of Knowledge, and Scopus databases for publications regarding the survival rates of the patients with alobar HPE.

Results: The boy was born 2010 with a prenatally diagnosed alobar HPE. Agenesis of the nose, one-sided anophthalmia and double-sided cheilognathopalatoschisis were seen. On 11.08.2010 a ventriculoperitoneal shunt was placed due to progressive hydrocephalus. Epileptic seizures were observed. Currently the child is living with a foster family and has been stable over a year with good seizure control under antiepileptic drug therapy. The review of literature confirmed the very poor prognosis of this kind of CNS malformation.

Conclusion: Although alobar HPE is associated with a very poor prognosis, every patient should be assessed individually, as a prolonged survival might be possible.

Keywords: alobar holoprosencephaly, survival, case report

P-084

Brain malformation

Third ventricle floor variations and anomalies in myelomeningocele related hydrocephalus: experience in 300 endoscopic third ventriculostomy procedures

Volkan Etus¹; Murat Geyik²; Umit Celakil¹; Aykut Gokbel¹; Melih Caklili¹; Atakan Emengen¹.

¹Kocaeli University Faculty of Medicine Department of Neurosurgery, Kocaeli; ²Gaziantep University Faculty of Medicine, Department of Neurosurgery, Gaziantep

Objective: Anatomical variations and abnormalities of ventricular system are of great importance in endoscopic third ventriculostomy (ETV) procedure, since ETV may become technically demanding and full of risks in such conditions. The floor of the third ventricle with a confusing topography is particularly problematic in ETV procedure.

Methods: A retrospective review was performed on our 300 myelomeningocele related hydrocephalus cases that have been treated with ETV at our neuroendoscopy unit between 2005 and 2015. The series consisted the pediatric cases that have been initially treated with ETV for myelomeningocele related hydrocephalus and also the myelomeningocele cases that have been treated with ETV for management of CSF-shunt dysfunction. Reviewing the ETV video recordings, we determined the encountered variations and anomalies of the third ventricle floor in cases with myelomeningocele related hydrocephalus.

Results: The third ventricle floor showed significant anatomical variations and anomalies in 197 cases. The mostly encountered anatomical features were as follows: thick and prominent massa intermedia; parenchymatous, opaque and/or thick floor of the third ventricle; narrow tuber cinereum; hollow, steep or vascular floor of the third ventricle; small anterior chamber of the third ventricle, existence of interhypothalamic adhesions and existence of adhesions or bridges between mamillary bodies. The analysis of our data revealed that, the ratio of the cases with variations and anomalies of the third ventricle floor was 65,6% in our series.

Conclusion: The influence of various anatomical situations on operational strategy of ETV procedure in myelomeningocele related hydrocephalus cases is discussed over examples of mostly encountered anatomical variations and abnormalities of the third ventricle floor.

Keywords: hydrocephalus, myelomeningocele, third ventricle, variation

P-085

Brain malformation**A giant occipital encephalocele SAC of a newborn with almost whole cranial contents**Özgür Demir¹; Fatih Ersay Deniz¹; Erol Öksüz¹.¹Gaziosmanpaşa University, Department of Neurosurgery, Tokat

Objective: Encephaloceles account for 10 to 20% of all craniospinal dysraphisms. The large sized swellings may have significant brain herniation, abnormality of the underlying brain, microcephaly and ventriculomegaly. Such patients usually have poor prognosis. The contents of encephalocele may be occipital lobe, cerebellum or brainstem. However, they are extremely rare to have both cerebral hemispheres. We presented a microcephalic newborn baby with occipital encephalocele including almost whole cranial contents.

Methods: A newborn female baby presented with swelling over the occipital region. The baby was born at 32 weeks of gestation by caesarean section with a birth weight of 2100 gms. Head circumference was 21 cm with a closed anterior fontanelle. There was a large occipital swelling which was tense, non-cystic measuring 13 cm (Figure 1). There was no additional abnormality on physical examination except for a large mass in the occipital region.

Results: There was even no response to pain and no spontaneous respiration on her neurological examination. She was intubated and connected to mechanical ventilator. Both of pupils were dilated and response to light was decreased. Cardiac support was given due to hypotension. A computerized tomography scan (CT) demonstrated the encephalocele with evidence of herniation of posterior fossa contents and almost whole cerebral hemispheres. CT images also revealed a significant defect of the occipital bone (Figure 2, 3). Surgical procedure was not performed because of her neurological and cardiac condition. The patient died one week after the birth.

Conclusion: Patients with giant encephalocele and large amount of brain tissue in the sac usually die either shortly after birth. Presented case is extremely rare because the encephalocele sac had almost whole cranial contents. Our patient had little amount of neural tissue in the skull. Our patient also died one week after the birth.

Keywords: Encephalocele

P-086

Craniovertebral Junction**Stock car racing related cranio-cervical junction injury presenting with bilateral hypoglossal and abducens nerve palsy**Himanshu Shekhar¹; Jothy Kandasamy¹; Drahoslav Sokol¹; Pasquale Gallo¹; Chandrasekaran Kaliaperumal¹.¹Royal Hospital for Sick Children, Edinburgh

Objective: We report a rare case of post-traumatic bilateral hypoglossal and abducens nerve palsy in an eleven year old boy.

Methods: Clinical case review

Results: An eleven year old boy presented to us following a sport related injury. He was the driver of an old “Mini” in a stock car race and was wearing a harness and helmet when he crashed, hitting firstly another car, and then a wall at about 50 mph. CT Brain and Cervical spine showed acute subdural haemorrhage anterior to the brainstem and a left sided occipital condyle fracture. Clinically he was alert and following commands, his symptoms included dysphasia, dysarthria and horizontal diplopia. On examination he had severe bilateral hypoglossal and abducens nerve palsy. A subsequent MR cerebral angiogram excluded vertebral and basilar arterial injury. MR Cervical spine showed pre-vertebral soft tissue swelling and high T2/STIR signal in the posterior neck soft tissues, mainly between the occiput and C2. Immediate management included hard

collar and a short course of Dexamethasone. Two weeks later he experienced slight improvement in his speech and swallowing but the abducens palsy persisted. A follow-up MR was obtained in the second week noted persistent pre-pontine subdural haemorrhage. Further multi-disciplinary management included collar care, speech therapy and ophthalmology follow-up. He underwent bilateral medial rectus Botulinum toxin injections, with some improvement in bilateral lateral rectus function. Follow up cervical spine imaging (CT, MR Cervical Spine & X-ray Flexion-Extension) at three months from trauma confirmed spinal stability and collar was removed. Both cranial nerve palsies had resolved in his most recent follow-up (12 months from injury).

Conclusion: This case highlights the management of post-traumatic bilateral abducens & hypoglossal nerve palsy. Hard cervical collar can be considered adequate in selected cases with a low index of suspicion for cranio-cervical instability.

Keywords: occipital condyle fracture, hypoglossal nerve palsy, abducens nerve palsy, cranio-cervical injury

P-087

Craniovertebral Junction**Skeletal deformity complicating halo orthosis following cervical fixation in MPS: report on the use of a customised non-invasive brace**Guirish A. Solanki¹; Suresh Vijay¹; Saikat Santra¹.¹Departments Of Paediatric Neurosurgery And Inborn Errors Of Metabolism, Birmingham Children’S Hospital NHS Foundation Trust, Birmingham

Objective: Cervical spine instability(CSI) and cord compression(CCC) occur frequently in the mucopolysaccharidoses, especially type VI (Maroteaux-Lamy Disease, MPSVI) and type IV (Morquio Disease, MPSIV). Standard surgical treatment involves cervical decompression and/or stabilization with a period of external fixation with a Halo external orthosis (HO). Concomitant skeletal deformity can make the application of a HO difficult in some young children with MPSIV and MPSVI. We report our experience of performing combined cervical spinal decompression and stabilization using a custom-made brace for immobilization in three children with MPS disorders.

Methods: Two children with MPSIV (aged 7 and 10) and one with MPSVI (aged 3) were identified as requiring surgery for CCC with CSI based on clinical and radiological findings. Skeletal deformity, including pectus carinatum, precluded the use of a HO and the patients were individually measured for custom-made spinal brace jackets to provide postoperative immobilization. Recombinant human bone morphogenetic protein was also used in the two MPSIV patients to aid bone fusion.

Results: Preoperative brace size measurements proved inaccurate in the immediate postoperative period due to surgical delay and postoperative swelling. Temporary immobilization with plaster braces was required until postoperative swelling had settled and patients were re-measured for individual plastic braces. Radiological studies (CT) confirmed satisfactory postoperative alignment was maintained in all patients. Bone fusion was demonstrated within 8 weeks and was satisfactory by 12 weeks with no complications. The brace was well-tolerated by the patients, the main concern being with interference with mouth opening, requiring re-fashioning of the chin section.

Conclusion: Where skeletal deformity precludes the application of a HO, noninvasive immobilization using a spinal brace jacket is an acceptable alternative. Ensure accurate measurements are taken to ensure a good fit. Temporary plaster bracing may be needed in the immediate postoperative period.

Keywords: cervical, spine, cord compression, Mucopolysaccharidosis, MPS IVA

P-088

Spine

Segmental spinal dysgenesis: a report of early surgical intervention and outcome in a 14 month-old child and reviewSean Morell¹; Richard E. Mccarthy¹; Eylem Ocal².¹Department of Orthopedics, University of Arkansas for Medical Sciences, Arkansas Children's Hospital, Little Rock; ² Department of Neurosurgery - University of Arkansas for Medical Sciences, Arkansas Children's Hospital, Little Rock**Objective:** Segmental spinal dysgenesis is a rare congenital deformity resulting in spondyloptosis and subsequent neurological dysfunction. It is usually at the thoracolumbar junction. There is little known concerning surgical intervention and its timing in these patients. The goal of this report is to present and review this rare entity, to discuss treatment options, timing of surgery and subsequent follow-up.**Methods:** Segmental spinal dysgenesis was originally described by Winter et al. It is an uncommon congenital spinal defect involving localized agenesis in the upper lumbar or thoracolumbar spine. It is associated with focal canal stenosis, hypoplastic or absent vertebrae, instability, and subluxation of the spine with worsening kyphosis that leads to progressive neurological deficit. It has been associated with maternal diabetes, various drugs, and toxins. We present a current case involving an infant diagnosed with segmental spinal dysgenesis at the thoraco-lumbar junction soon after birth, who had surgery at 14-months of age, and his subsequent follow-up. We also review treatment and surgical approaches to two other previous similar cases.**Results:** The patient was diagnosed with segmental spinal dysgenesis following birth. He had good neurological function with full knee flexion and extension, weak dorsi- and plantar flexion; intact sensation to pain and touch down to the ankles. He was treated with bracing until 14-months of age. Surgery was then performed when bone quality was deemed adequate for instrumentation. The patient underwent L1 hemivertebra excision and L2 vertebral resection (VCR) with fibular strut grafting from T12-L3 with posterior instrumentation (pedicle screws and rods). His neurological outcome was better when compared to previous cases who presented and were operated at later ages.**Conclusion:** There are few reported cases in the literature concerning the treatment of segmental spinal dysgenesis, timing for surgical intervention and techniques for stabilization. We present our experience and provide a discussion about this rare pathology.**Keywords:** spinal dysgenesis, spondyloptosis, surgery, congenital

P-089

Spine

Usage of minimally invasive spine surgery techniques on pediatric patients with lumbar disc herniationMichal Tichy¹; Jiri Steindler¹.¹University Hospital Motol, Prague**Objective:** Out of the total quantity of all surgical interventions for lumbar disc herniation is less than one percent done on patients between ages 11 to 20 years old.. Back pains occurring to population under 11 years old are much more likely to be a manifestation of other diseases (tumours, trauma, congenital anomalies, inflammations). Using the considerate of minimally invasive spine surgery (MISS) techniques enables faster recovery to normal patient's life activity.**Methods:** The MISS techniques are practiced on child and adult patients at our department since the year 2004. Microdiscectomy falls into our most frequently applied type of surgery. It is done by using METRx system (Minimal Exposure Tubular Retractor). Due to low incidence of

child patients with this particular diagnosis we have accomplished so far 16 clearly miniinvasive operations. The average age of our patients has been 16 years (span from 14 to 19), the sex ratio is 12 males: 4 females.

Results: We did not record neither peroperative nor postoperative complications, there was no necessity to reoperate any of these patients (the follow-up spans from 1 to 9 years, average 4,5 years), practically all operated patients are completely satisfied without any complications in ordinary life activities.**Conclusion:** Minimally invasive spine surgery is according to our experience more considerate to young patients than older more extensively used methods. We endorse that qualitative and quantitative development of these modern techniques in spinal surgery will continue in the years to come.**Keywords:** Pediatric spine, Minimally invasive, Disc herniation

P-090

Spine

Wide multilevel costotransversectomy; as an alternative to combine approach to achieve spinal fusion of 360 degrees in correction for a case of severe congenital kyphosisİhsan Doğan¹; Murat Zaimoğlu¹; Gökmen Kahiloğulları¹.¹Ankara University School of Medicine İbni Sina Hospital Department of Neurosurgery, Ankara**Objective:** Extended costotransversectomy is an option to reach vertebral body at thoracic levels and establishing anterior column fusion without to be exposed to risks of approach related morbidities in anterior surgery. We aimed to share our experience about efficacy of costotransversectomy in revision surgery for a case of congenital kyphotic deformity.**Methods:** 14 year old female was admitted for congenital kyphosis. The curve was measured as 101° from T3 to T12 by Cobb's method. Unsegmented block vertebrae was observed from T7 to T12. Apex was T9. Total laminectomy was performed into the levels which belong to block vertebrae. Corrective wedge osteotomy was performed into the apex of the curve. Pedicle screw fixation was carried out three levels above and below the osteotomy. In first postoperative day, loss of correction and grade-2 retrolisthesis at osteotomy line was observed on imaging studies. Revision was decided due to lack of stability. One each level pedicle screw placement was done. Anterior column fixation was performed at the osteomy site by using Z plate and screws to achieve circumferential fusion via extended costotransversectomy approach by using the same incision from previous surgery.**Results:** Postoperative curve magnitude was 62°. There was no listhesis. The patient was neurologically intact. She mobilized on the first day of revision surgery. Using brace was not recommended postoperatively.**Conclusion:** Extended costotransversectomy approach is an option to achieve ventral spinal fusion. Combining with posterior approach by using the same incision facilitates to accomplish circumferential fusion via one portal and in one session. In our case, we have managed a postoperative lack of stability without taking approach related risks of an additional anterior procedure by using extended costotransversectomy. It provided safe access to the surgical field without making a new incision.**Keywords:** costotransversectomy, congenital kyphosis

P-091

Spine malformation

Tethered cord syndrome and occult spinal dysraphism. Clinical features and surgical results.Oscar García-González¹; Paola Hernandez-Ponce².

¹Hospital Regional De Alta Especialidad Del Bajío, León; ²Universidad Autonoma De Guadalajara, Guadalajara

Objective: The clinical features and outcome in 24 pediatric patients with TCS treated surgically at the Hospital Regional de Alta Especialidad del Bajío between June 2009 – February 2014 was studied retrospectively.

Methods: 17 children with spinal lipomas and closed spinal dysraphism, 4 children with dermal sinus, one of them infected, 2 children with myelocystocele, and 1 child with tight filum terminale were included. **Results:** During the period in 10 boys and 14 girls aged from 2 months to 11 years (mean age 12.9 months) detethering procedures were performed. Most of the patients were asymptomatic prior to surgery and 31.8% of the patients presented with neuro-orthopedic syndrome and urinary dysfunction. CSF collection, occurred in 4 cases and infection in 2 cases; none of these complications caused permanent morbidity. Of all signs and symptoms, muscle weakness and bladder dysfunction responded most favorably to surgical treatment.

Conclusion: Prophylactic detethering appears to be effective among asymptomatic patients. Early surgical detethering seems to be more effective in improving neurological symptoms. Once symptomatic, the surgical detethering in TCS patients tend to stop further progression of symptoms and even improved.

Keywords: Tethered Cord

P-092

Spine malformation

Open spina bifida in premature newborns

Volkodav Oleg¹; Zinchenko Sveta¹.

¹Crimean Medical University, Simferopol

Objective: The actual task of neonatal neurosurgery is the open spina bifida (OSB) treatment in premature newborns (PN). They are presented: myelomeningocele and rachischisis.

Methods: 25 urgent neurosurgical operations were performed to PN (newborns minimal body mass consist of 860gramme and 26 weeks of gestation); in one PN rachischisis Th5-L3. OSB plastic art method in PN was proposed (Copyright №17679, 21.08.2006), ensures minimal invasiveness, plasticity, functionality and efficiency principles. OSB plastic art peculiarities in PN presumes extensive vertebral canal posterior wall defect and soft tissues hypotrophy, rough anatomical failure, skin changes and CSF leakage.

Results: OSB plastic method includes careful medullar plate selection by the arachnoid crease with the sac contents inspection, spinal cord untethered, skin-fascia flap formation. Subsequent is the stored hernia sac immersion, medullar plate neurulation, vertebral canal posterior wall reconstruction. Pia-arachnoid membrane fold is formed in the vertebral canal lateral pockets to CSF absorption, spinal blood supply preserve through the pia mater vessels network. Skin-fascia flap have used for shelter - sealing of the hernia gate and vertebral canal, providing plastic reliability and CSF leakage prevention.

Conclusion: This method allows to the OSB plastic in PN any forms and sizes with vertebral canal posterior wall reconstruction, minimum risk of bleeding, reliable sealing in conditions of the soft tissue malnutrition, a reduction of the time of surgery and anesthesia.

Keywords: premature newborn, open spina bifida, plastic art

P-093

Spine malformation

Klippel-Feil syndrome and interruption of aortic arch

Mehmet Şah Ipek¹; Nilüfer Okur¹; Osman Akdeniz¹.

¹Maternity and Children Hospital, Diyarbakır

Objective: Klippel-Feil syndrome (KFS) is a complex syndrome of osseous and visceral anomalies that include the classical clinical triad of short neck, limitation of head and neck movements and low posterior hairline. It may also be associated with anomalies of the genitourinary, musculoskeletal, neurologic and cardiac systems.

Methods: We report a case of a newborn infant having KFS associated with interruption of aortic arch.

Results: A female neonate with a gestational age of 38 weeks and weight of 2400 g who was born through C-section was transferred to the neonatal intensive care unit due to meconium stained amniotic fluid and respiratory distress. At the delivery room, the infant demonstrated decreased tone and poor respiratory effort requiring intubation and aspiration of meconium. On physical examination of the patient were found short neck, low hairline and limitation of neck motions, low-set ears, in addition to cyanosis, tachypnea of more than 70 breaths per minute, and chest retraction. Chest X-ray showed bilaterally diffuse patchy infiltration and congenital high scapula (Sprengels deformity). Lateral cervical radiograph revealed fused cervical segments (Figure 1). Echocardiogram demonstrated ventricular septal defect, interruption of the aortic arch (the interruption occurs proximal to the origin of the left common carotid artery) and aortic valve stenosis. Ultrasonography revealed the presence of a bilateral pleural effusion and ascites, and mildly dilated renal collecting system and increased echogenicity of the renal parenchyma. The patient died while waiting for emergency heart surgery on day 7.

Conclusion: A variety of cardiovascular anomalies have been reported in patients with KFS. This is in our best knowledge the first report of interrupted aortic arch and KFS. As seen in the presented case, the association of cardiac defects with KFS may be more serious in early life, and comprehensive evaluation of patients with KFS is required, and may be life-saving.

Keywords: Klippel-Feil syndrome, interrupted aortic arch, cardiovascular anomalies

P-095

Spine malformation

Missed limited dorsal myeloschisis: an unfortunate cause for recurrent tethered cord syndrome

Sandip Chatterjee¹.

¹Park Clinic Kolkata, Kolkata

Objective: To highlight the recurrent tethered cord syndrome in relation to the relatively new pathological entity of “limited dorsal myeloschisis” and to mention the difficulties faced by the paediatric neurosurgeon in developing countries with reference to LDM which was not recognized at initial presentation.

Methods: Four cases of recurrent tethered cord syndrome who were operated early in life (not by paediatric neurosurgeons) as “meningocoeles” and who were then subsequently referred to the senior author as they presented with clinical signs of re-tethering of the cord.

Results: The first child of 1 year age represented with a cervicodorsal swelling 10 months after primary surgery for a cervical “meningomyelocele” done elsewhere on the second day of life. The second was a girl of three who was operated at birth and presented with severe brachialgia and neck pain after 2 years. The third was a 19 year old girl operated at birth presenting with a spastic paraparesis and also hand weakness associated with kyphosis. All three patients improved symptomatically and neurologically after redo surgery.

Conclusion: The diagnosis of LDMS, especially in developing countries, is frequently missed. This needs to be tackled addressed so that children with this subset of neural tube defects, who otherwise would normally

have a very good outcome, may not be blighted and left scarred for life at an early age.

Keywords: limited dorsal myelosischisis

P-096

Spine malformation

Primary tethered cord

Alina Khodorovskaya¹; Wiliam Khachatryan¹.

¹Russian Neurosurgical Association, Sankt-Petersburg

Objective: Tethered cord syndrome is the most widespread disease in the pediatric spinal neurosurgery, there are no conventional tethered cord (TC) criteria or indications for surgical treatment of TCS in case of spinal disorders beyond the scope “elongated spinal cord – thickened terminal filament”.

Methods: Data on 64 patients with congenital malformations of the lumbosacral spine at the age from 1 month to 17 years who underwent examination and surgical treatment in Polenov Russian research neurosurgical institute in the period from 2002 to 2014 were analyzed. The patients were included if they met the following criteria: diagnosed or possible tethered cord syndrome and no prior operations associated with spinal dysraphia.

Results: Results: According to our data, primary TC was found in 34.4% of patients with various types of spinal dysraphia including 12% of patients with the split cord; 6% of patients with lipomas; 4.5% of patients with intradural arachnoid cysts; 11.9% of patients with lipomyelocele. In patients with myelocele without a lipomatous component no primary TC was found. On the basis of revealed clinical, morphological and introsopic relations the types of TC course were determined.

Conclusion: The elongated spinal cord is not necessarily the sign of TC in patients with split cord and lipo (meningo) myelocele. In case of myelo (menigo) cele, the elongated spinal cord is a compensatory mechanism that prevents the prenatal development of TCS. In case of lumbosacral lipomas? the rapid enlargement of a subcutaneous lipoma is a criterion of TC and an indication for surgical treatment. In case of split cord the presence of a syringomyelia cavity above the level of the diastema is a criterion of TC and an indication for surgical treatment. In case of lipo (meningo) myelocele an asymmetric neurologic deficit is a criterion of TC and an indication for surgical treatment.

Keywords: Primary tethered cord

P-097

Spine malformation

High incidence of lower extremity deformities in patients with jarcho-levin syndrome

Burak Abay¹; Ibrahim Alatas²; Huseyin Canaz²; Isik Karalok³; Ezgi Erdogan²; Nursu Kara⁴; Kerem Ozel².

¹Department of Orthopedic Surgery, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul;

³Department of Radiology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ⁴Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul

Objective: Neurological inability of lower extremities secondary to spina bifida commonly presents as lower extremity deformities in the patients with Jarcho-Levin syndrome (JLS). Children with JLS develop a wide variety of congenital and acquired hip joint deformities, knee joint and foot deformities.

Methods: We examined orthopedically 34 patients (15 male, 19 female) with JLS. Same orthopedic surgeon evaluated the clinical examinations and plain X-rays of lower extremity deformities of the 34 patients with JLS. **Results:** 24 (18 unilateral, 6 bilateral) of 34 patients (70,5%) present as hip dislocation. 8 of 34 patients (23,5%) had bilateral hip flexion contractures. 11 of 34 patients (32,3%) had knee flexion contractures. 2 of 11 patients (18,2%) with knee flexion contracture presented as congenital patellar dislocation which is a severe and rare form of knee deformity. 16 (11 bilateral, 5 unilateral) of 34 patients (47,0%) present as foot deformities. 15 of 16 patients (93,7%) had equinus foot deformity. 12 of 15 patients (80%) with equinus foot deformity had recurrence after serial casting for Pes equinus varus (PEV).

Conclusion: There is a high incidence of lower extremity deformities in patients with JLS who have no motor activity in the lower extremity. These deformities of lower extremity diminish the ambulation of the patients with JLS. Recurrence of surgically treated lower extremity deformities are high because of lack of physical therapy and poor communication with the family and orthopedic surgeon.

Keywords: Jarcho Levin Syndrome, Spina Bifida, Lower extremity anomalies

P-098

Spine malformation

Myeloschisis- a pathology of transition - spina bifida intermedia

Suhas Udayakumar¹; Chiazor Onyia².

¹Division of Paediatric Neurosurgery, Amrita Institute of Medical Sciences and Research Centre, Kochi; ²Neurosurgery Division, Department of Surgery, Obafemi Awolowo University Teaching Hospitals Complex, Ileife

Objective: Myeloschisis although an open defect, has a distinct difference with myelomeningocele. The literature is sparse on this issue.

Methods: We report 3 neonates with myeloschisis. All the three at surgery showed split cord, dermoid and one of them had a lipoma.

Results: Split occurs in third week and nonclosure of neuropore occurs in fourth week of gestation. Clearly non-closure of posterior neuropore is a part of the pathology. We propose additionally, it involves defective secondary neurulation and the key to the defective transition may be an abnormal axially condensed mesenchyme.

Conclusion: We propose the term spina bifida intermedia be used to describe myeloschisis as it represents a pathology of the junctional neural cord and on the timeline occurs between primary and secondary neurulation.

Keywords: spina bifida, myeloschisis

P-099

Spine malformation

Evaluation of quality of life in mothers of children with meningomyelocele

Senem Alkan Özdemir¹; Nail Özdemir²; Volkan Murat Ünal²; Esra Arun Özer¹.

¹Tepecik Research and Training Hospital, Neonatology, İzmir; ²Tepecik Research and Training Hospital, Neurosurgery, İzmir

Objective: To assess the quality of life in mothers of children with meningomyelocele and to correlate them with the clinical examination findings of the childrens.

Methods: During 2 years period, 50 infants with meningomyelocele underwent surgical repair. We also investigated the relationship between childrens disability and mothers physical and psychological dimensions of

quality of life. Mothers were interviewed for quality of life using the World Health Organization quality of Life (WHOQOL-BREF) questionnaire.

Results: There was no significant difference in demographic characteristics and the mean total score of quality of life between groups.

Conclusion: We can increase mothers health and quality of life and the rate of antenatal diagnosis with wide planning and supportive intervention by the families and society.

Keywords: meningomyelocele, quality of life, WHOQOL-BREF questionnaire

P-100

Spine malformation

Cutaneous lesions in occult spinal dysraphisms

Ibrahim Alatas¹; Huseyin Canaz¹; Isik Karalok²; Nursu Kara³; Ezgi Erdogan¹; Kerem Ozel¹.

¹Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Department of Radiology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ³Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul

Objective: Congenital midline paraspinal lesions, mostly localized in the lumbosacral area, are widely recognized as markers of occult spinal dysraphism (OSD).

Methods: We retrospectively reviewed the clinical records of 69 pediatric patients with OSD who presented with congenital midline lumbosacral skin lesions in our center. Cutaneous lesions and associated spinal defects were determined. Patients were grouped as hypertrichosis, port wine stain, sacral dimple and lipoma according to their cutaneous lesions and their MR images were reviewed.

Results: There were 18 patients with hypertrichosis. 5 patients had isolated tethered cord (TC), 9 patients had TC and split cord malformation (SCM), 2 patients had lipomyelomeningocele with TC, 2 patients had isolated SCM. There were 10 patients in port wine stain group. 4 patients had isolated TC, 3 patients had TC and SCM, 3 patients had isolated posterior fusion defect (PFD). There were 21 patients with sacral dimple. 14 patients had PFD, 4 patients had syringomyelia, 3 patients had TC. There were 20 patients with lipoma. 16 patients had lipomyelomeningocele with TC, 3 patients had lipomyelomeningocele and SCM, 1 patient had isolated TC.

Conclusion: OSD is frequently associated with a combination of 2 or more different congenital midline lumbosacral lesions. It is important to detect these lesions before the occurrence of neurologic or orthopedic manifestations. Magnetic resonance imaging is the best radiologic imaging method. However, USD may be used in some cases.

Keywords: Hypertrichosis, sacral dimple, port wine stain, lipoma, occult spinal dysraphism

P-101

Spine malformation

Idiopathic spinal epidural arachnoid cyst: a case report

Minichini Viviana¹; Nebbio Andrea²; Fricx Christophe³; Lolli Valentina⁴; Fontange Quitterie⁵; De Witte Olivier¹.

¹Department of Neurosurgery, Erasme Hospital (ULB), Brussels;

²Department of Pediatrics, Reine Fabiola Children Hospital, Brussels;

³Department of Pediatric Surgery, Erasme Hospital (ULB), Brussels;

⁴Department of Neuroradiology, Erasme Hospital (ULB), Brussels;

⁵Department of Pathological Anatomy, Erasme Hospital (ULB), Brussels

Objective: Idiopathic spinal epidural arachnoid cysts (iSEACs) are rare lesions which mainly affect the thoracic spine in children and the lumbar in adults. Often they result in progressive spastic paraparesis and

radiculopathy. Many surgical treatments have been reported including simple osseous decompression, marsupialisation of the cyst, and total cyst resection.

Methods: We report a case of a 8-year-old female child who has presented with progressive walking troubles since three months due to a right foot drop accompanying with hypoesthesia in both legs without any sphincter deficiency. MRI revealed a Th11-L3 epidural arachnoid cyst of 11.0 x 2.0 x 3.5 cm with mass effect on the anterior spinal cord and the cauda equine. A preoperative CT myelography suggested a cyst-subarachnoid space communication. A Th11-L4 laminoplasty was performed with a total excision of the cyst. Contrary to the results of the CT myelography, intraoperatively, there was no cyst-subarachnoid space fistula found.

Results: No recurrence has been observed during the follow-up period until now. A good outcome has been achieved.

Conclusion: SEAC are rare findings. The pathogenesis remain unknown and congenital neural tube defect is supposed to play an important role. Nevertheless, acquired origin such as spinal surgery, trauma, chronic arachnoiditis have also been reported. Early surgical intervention is suggested for progressive symptomatic patients in order to improve any neurological deficit. Communication with the subarachnoid space should be closed during the surgery, whenever possible. Furthermore, the exposure should be wide enough to allow harmless dissection of the cyst and to provide complete resection.

Keywords: Spinal Epidural Arachnoid Cyst

P-102

Spine malformation

Outcomes of neonates with meningomyelocele: single institute experience

Senem Alkan Özdemir¹; Nail Özdemir²; Serpil Özkoç Erof³; Volkan Murat Ünal²; Esra Arun Özer¹.

¹Tepecik Research and Training Hospital, Neonatology, Izmir; ²Tepecik Research and Training Hospital, Neurosurgery, Izmir; ³Tepecik Research and Training Hospital, Pediatric Development, Izmir

Objective: In this report, the authors focus on the management of meningomyelocele and its associated conditions.

Methods: During 2-years period, 50 neonates with meningomyelocele underwent surgical repair. All children were evaluated with the Denver Developmental Screening Test in addition to neurological examination.

Results: Overall, all neonates were discharged from hospital. There was a 35 neonates (70%) of hydrocephalus. Ventriculoperitoneal shunt performed in an all hydrocephalic patients. There was a 24 neonates (48%) Chiari type 2 malformation. Posterior fossa decompression with duraplasty performed in a 7 patients for Chiari type 2 malformation. The incidence of pes equinovarus deformity was 44% (22 neonates). There was a kyphosis in 16 neonates (32%). Kyphectomy performed in a 6 neonates during meningomyelocele repair. Paraplegia was noted in 38 neonates (76%) after birth. During the follow-up (range 5-20 months, mean 10,3±4,2 months), 7 (20%) neonates with shunt infections (35 of shunts) and 5 neonates (10%) with wound problems.

Conclusion: Despite a national universal health care system; self care may not be possible in a large percentage of cases. There are many associated neurological disabilities that have to be faced, and a coordinated network of care still remains an important issue throughout the patients life. Additionally, these infants reach adulthood and social continence, choice of proper techniques and so many peculiarities that indications for treatment are uncertain.

Keywords: meningomyelocele, Denver Developmental Screening Test

P-103

Spine malformation

Our 5 year experience of spina bifida cases: report of 82 cases

Basak Caner Topkoru¹; Nejat Isik¹; Recep Basaran¹; Ilke Mungan Akin²; Sibel Sevuk Ozumut²; Derya Buyukkayhan²; Ferruh Gezen¹.

¹Istanbul Medeniyet University Goztepe Education and Research Hospital Department of Neurosurgery, Istanbul; ²Istanbul Medeniyet University Goztepe Education and Research Hospital Department of Pediatrics, Istanbul

Objective: Neural tube defects are the most encountered congenital malformation in the neonatal period. We aimed to review our spina bifida cases over a 5 year period.

Methods: We have retrospectively reviewed the cases with spina bifida who were admitted to the Newborn ICU and operated by the Neurosurgery Clinic in Istanbul Medeniyet University Goztepe Education and Research Hospital between January 2010 and January 2015.

Results: There were 82 patients. 3 of them were lost due to associated problems before they got operated. 76 of them were operated in the early period. Defect localizations were as follows: lumbar (44.7%), lumbosacral (27,6 %), toracolomber (13,2 %), toracal (5,3 %), sacral (5,3 %) and cervical (2,6 %). One patient had lipomeningomyelocel and 8 patients had meningocel. The rest had meningomyelocel. 55 of the patients (72.3%) required ventriculoperitoneal shunting. In 9 of these cases VPS was performed after an average of 13 days in a second operation. The mean hospital stay of these patients was significantly longer than those who were shunted in the same operation with the defect closure. Pes equino varus, Chiari Type 2, corpus callosum agenesis, hydronephrosis, hypotiroidism were the most associated pathologies.

Conclusion: Spina bifida is a complicated disorder requiring multidisciplinary approach and long term follow-up.

Keywords: spina bifida, neural tube defects, hydrocephalus

P-104

Spine malformation

Algorithm for the diagnosis and surgical treatment of tethered cord syndrome (TCS) in children.

Vasilii Danilin¹; German Letyagin¹; Sergei Kim¹; Anna Sysoeva¹.

¹Federal Neurosurgical Center, Novosibirsk

Objective: The purpose of this study was determining algorithm diagnostic and surgical treatment of tethered cord syndrome in children.

Methods: The work is based on analysis of 57 children was operated in Federal Neurosurgical Center. The algorithm of preoperative diagnosis in these children included: MRI, MDCT with 3D volume rendering of the spine area concerned, MDCT of the brain (in children with spinal hernia), physical examination (to identify spinal malformation stigma) and clinical and neurological examination.

Results: Assotiation MRI and MDCT was helped us get a complete picture of the pathological process that enabled us to plan the surgery (considering both soft tissue and skeletal malformations and pathological processes). All surgeries were performed with neurophysiological monitoring. Deterioration of neurological deficit not received all patients.

Conclusion: Diagnostic procedure in children with tethered cord syndrome must necessarily include not only the conduct of the MRI area concerned, but also MDCT with 3D volume rendering, helping to plan the operation very carefully. Surgery must be performed with neurophysiological monitoring, which greatly reduces the risk of postoperative deterioration of neurological deficit.

Keywords: spinal malformation, tethered cord syndrome

P-105

Spine malformation

Congenital cervical kyphosis in two siblings: case presentations

Mehmet Alptekin¹; Abidin Murat Geyik¹; İbrahim Erku¹; Mert Nazik¹.

¹Gaziantep University Faculty of Medicine, Department of Neurosurgery, Gaziantep

Objective: Congenital Cervical Kyphosis is rarely seen. Many syndromes belonging to different genes which causes segmentation defect in the vertebra is listed. Among these, the Klippel-Feil syndrome, Larsen Syndrome and Dwarfism are associated with cervical vertebral malformation. In our presentation, our approach to such cases will be revealed through 2 siblings with cervical kyphosis operated in our clinics.

Methods: Case 1: A three-year-old child, who is inflicted with the disability of walking without support and has urinary incontinence, regressed in the last one month before the admission. In the examination of the patient, there existed pathologically paraparesis, DTRs hyperactive, bilateral Babinski reflex positivity and clubfoot deformity in both feet. In the radiological imaging, there existed kyphosis in the cervical area, spinal cord injury and syringomyelia appearance. Anterior releasing is applied in the patient's treatment as a first step. After ligament release, C3 and C4 corpectomy, distractable cage placement and C2-5 anterior fixation is applied in to operations to the patient. In the postop period, the patient uses SOMI brace. Case 2: On detecting in his brother cervical kyphosis, the 14-month patient who was called to our clinic is hospitalized in our clinic. Explicit pathology in the neurological examination is not detected. Anterior releasing is applied in the first stage. After that, C4 corpectomy, otogen grafting and C3-5 anterior fixation is applied to the patient. In the check-ups performed in the postop period, it is seen that the grafting of the patient is fusion and the kyphotic deformity got better.

Results: In the rarely seen cervical kyphosities cases, surgery option is especially young ages is limited. In both our cases, the combined use of the anterior releasing and anterior fusion techniques is beneficial for our patients.

Conclusion: The congenital cervical kyphosis is seen in both siblings brings to mind the hereditary transit.

Keywords: Congenital Cervical Kyphosis, anterior releasing, corpectomy

P-106

Spine malformation

Ultra-early surgery and outcome in neonatal myelomeningocele cases

Volkan Etus¹; Hakan Karabagli²; Murat Geyik³; Umit Celakil¹; Aykut Gokbel¹; Melih Caklili¹; Atakan Emengen¹.

¹Kocaeli University Faculty of Medicine Department of Neurosurgery, Kocaeli; ²Selcuk University Selcuklu Faculty of Medicine Department of Neurosurgery, Konya; ³Gaziantep University Faculty of Medicine, Department of Neurosurgery, Gaziantep

Objective: The effect of ultra-early surgery on morbidity and neurological outcome in neonatal myelomeningocele cases was evaluated.

Methods: The data of total 345 myelomeningocele cases which, have been operated between 2000 and 2014 were retrospectively analyzed. Patient records on clinical features and operation timing were reviewed. The patients who had been followed up for at least 1-year postoperatively were included in the study.

Results: The analysis of the data revealed that 97 of the 345 patients have undergone ultra-early surgery (within 8 hours after birth). This group was compared with the remaining 248 cases by means of surgical morbidity and neurological outcome. In the ultra-early surgery group, 7 cases (7,

2%) had wound problems at myelomeningocele repair site. The number of the cases with postoperative wound problems at myelomeningocele repair site was found to be 41 (16.5%) for the rest of the cases. According to the records of their neurological status and Spina Bifida Neurological Scale (SBNS) scores, ultra-early surgery group showed better results during the follow-up period.

Conclusion: The ultra-early surgical intervention performed within 8 hours after birth showed benefits regarding a lower incidence of postoperative wound problems and dehiscence as well as a better neurological outcome.

Keywords: myelomeningocele, outcome, timing

P-107

Spine malformation

A new case of lumbosacral tail

Mario Garcia-Conde¹; Vanessa Hernandez-Hernandez¹; Liberto Brage-Martin¹; Pablo Febles-Garcia¹; Luis Perez-Orribo¹; Victor Garcia-Marin¹.
¹Hospital Universitario De Canarias, La Laguna

Objective: Although this is a rare condition, there are numerous reported cases of appendices or sacral "tails". It is a congenital malformation that is usually associated with other spinal disorders.

Methods: We report the case of 2 year old male who at birth presented with a skin appendage in the sacral region. He was neurodevelopmentally intact, presenting only with the aesthetic defect. To rule out any underlying spinal abnormalities, we performed a MRI in which a filum terminale lipoma at L5-S1 level was observed including the sacrum appendix and resulting in a tethered cord. In order to prevent the onset of neurological symptoms and to correct the aesthetic defect which produced the skin lesion, he was taken to the operating room where we proceeded to cut the terminal filum at L5 - S1 under neurophysiological monitoring and removed the sacral tail without exceeding the muscular plane.

Results: The child was discharged after 5 days without any neurological deficit and with good wound healing. During his outpatient follow up visits, he was noted to have excellent aesthetic results with normal ambulation and sphincter control. Post-procedure MRI showed ascent of the spinal cord, indicating a successful surgery. The treatment of these injuries must have two fundamental principles: 1). To rule out the presence of any underlying pathology (tethered cord), assess the treatment to prevent neurological impairment; and 2) to correct the aesthetic defect.

Conclusion: Prior to diagnosing a child with an appendix or lumbosacral tail, we must conduct proper clinical and radiological studies, allowing appropriate treatment and monitoring of the case. In addition, the involvement of a multidisciplinary team can prevent the occurrence of progressive neurological deficits using the correct diagnosis and surgical treatment.

Keywords: human tail, caudal appendage, spinal malformations, dysraphism, tethered cord

P-108

Spine malformation

Long-term outcome in congenital spinal dysraphisms: a single center observational study of adult cohorts

Raghav Singla¹; Deepak Gupta¹; Shashank Sharad Kale¹; Bhawani S Sharma¹.

¹Department of Neurosurgery, All India Institute of Medical Sciences, New Delhi

Objective: Children born with spinal dysraphism are often found to have significant neurological and urological sequelae and often succumb to life threatening end stage renal disease secondary to neurogenic bladder.

Methods: This is a single center study of 75 patients aged > 16 years (42 males, 33 females) of spinal dysraphism at our center from 2002-2013. Mean age of patients in the study group is 25.1 years. Pain was noted in 55 cases (73.3%), motor symptoms in 49 cases (65.3%), bladder bowel involvement in 38 cases (50.6%) and prophylactic surgery before scoliosis correction was noted in 24 cases. Radiologically, split cord malformations were noted in 20 cases, lipomas in 27 and neuroenteric cysts noted in 3 cases. Previous history of surgery for spinal dysraphism in the form of repair or detethering was noted in 17 cases. Mean age for patients with bladder/bowel involvement was 26.9 years and lipomas were noted in 19 (50%) of these 38 patients with pre-existing neurogenic bladder.

Results: Mild worsening in motor symptoms was noted in 5/75 cases while worsening of sensory symptoms occurred in 3/75 patients. Transient worsening of urological symptoms was observed in 8 patients. Pain improved significantly in adult onset group as compared to childhood onset group (p=0.017). Improvement in motor and urological symptoms did not show a significant difference.

Conclusion: Decreasing incidence of spinal dysraphism patients presenting at our hospital has been noted in the past decade. Close follow up and appropriate intervention have improved outcome in such cases. However, deterioration of renal functions remains a cause of worry over long term.

Keywords: long term outcome, tethered cord, spinal dysraphism

P-110

Trauma

Highly selective repeated brain ct for pediatric traumatic brain injury

Won-Hyung Kim¹; Sung-Won Jin¹; Sang-Dae Kim¹; Jong-Il Choi¹.
¹Korea University Ansan Hospital, Ansan

Objective: Repeated computed tomography (CT) follow up for traumatic brain injury patients is often performed. But it may expose the patients to unnecessary harmful radiation exposure especially in children. Furthermore, there is debate the indication and timing for repeated CT scans. In our study, patients were classified into several small groups and identified the specific conditions that influenced on the therapeutic plans or outcomes.

Methods: All patients of age 0 to 18 years that had come and admitted to our hospital for head trauma were enrolled. Patients were classified into 9 subgroups according to trauma mechanisms. Type and amount of hemorrhage and changes in the amount of hemorrhage on repeat CT were analyzed as well as initial Glasgow coma scale, Glasgow outcome scale, need for surgical intervention.

Results: Between March, 2007 and December, 2013, 269 pediatric patients had admitted to our hospital for head trauma. All patients received repeat CT before discharge and 166 patients received repeat CT within 24 hours after initial CT scan. There were progressions in the amount of hematoma in 48 (28.9%) patients. Among subgroups classified by trauma mechanism, there was no significant difference. But among various hemorrhage types, epidural hemorrhage more than 10cc measured in initial CT was found to be at risk of progression to need for surgical intervention significantly on routine repeated CT without neurological deterioration than other types of hemorrhage. Based on initial GCS, severe head trauma group (GCS 3-8) was at high risk of progression on routine CT without change of clinical neurologic status.

Conclusion: We suggest that the patients with EDH more than 10cc or GCS below 9 should be received repeat CT within 6 hours even though absence of significant clinical deterioration.

Keywords: traumatic brain injury, repeated brain CT, trauma mechanism, epidural hematoma, GCS

P-111

Trauma

Acute cortical blindness due to depressed occipital skull fracture: a pediatric caseVaner Köksal¹; Tugba Morali Guler².¹Recep Tayyip Erdogan University Medical School, Rize; ²Karabuk Government Hospital, Karabuk

Objective: Although linear skull fractures are common in children, depressed fractures constitute 25% of cases. A depressed skull fracture is defined as the displacement of bony fragments below the level of the whole thickness of skull through the dura or also the brain parenchyma. The depressed skull fractures are classified as open fractures and closed fractures according to the integrity of the overlying skin. Neurological findings according to depressed skull fractures are related with the severity of the trauma, compression of the broken bone to the parenchyma, the presence of contamination in open depressed fractures and the dural damage. Also the functions of the cortical area under compression affects the kinds of neurological deficits. The development of acute blindness is a very rare complication of depressed skull fractures reported in the literature. The depression of the bony island into the sagittal sinus also makes it difficult to repair for this case.

Methods: 6-year old male admitted to the emergency department with acute blindness after car accident. CT scan showed a depressed skull fracture of closed type in the occipital region. Emergent decompressive surgery was done because of the presence of acute blindness during admission to the hospital. The superior sagittal sinus preserved well during surgery.

Results: It was obtained during admission that a bony island was depressed over the superior sagittal sinus causing 1-1.5 cm compression to the parenchyma related with visual field. Because of acute visual loss emergent surgery was applied. Visual loss was totally improved by the second day postoperatively.

Conclusion: Types of depression fracture, neurological status of the patient and presence of neurological findings can affect necessity and also the time of surgery. In the presence of acute blindness emergent surgery should be done also for closed type depressed skull fractures.

Keywords: blindness, depressed skull fracture, occipital skull fracture, trauma, sagittal sinus

P-112

Trauma

Subdural hematoma mimicking subdural empiema in a Shaken Baby.Santiago Candela¹; Mariana Alamar¹; Patricia Puerta¹; Mariona Suñol²; Antonio Guillén¹; Enrique Ferrer¹.¹Department of Pediatric Neurosurgery. Hospital Sant Joan De Déu Barcelona, Barcelona; ²Department of Pathology. Hospital Sant Joan De Déu Barcelona, Barcelona

Objective: Shaken Baby Syndrome (SBS) is still a source of medical controversy. Diagnosis is difficult, not only for the clinic, but also because of the legal implications.

Methods: We present a child known to be affected of SBS brought by Social Services for fever and hemiparesis. We describe our initial management and final diagnosis.

Results: A 9-month infant (protected by the Social Services for polytrauma with subdural hygromas, retinal hemorrhages and femur fracture secondary to a possible abuse at the age of 6 months) was brought for persistent fever despite antibiotic treatment administered to an alleged

pneumonia. Since the child had a right hemiparesis, cranial CT scan was performed showing an extra-axial collection of 6 cm thick that enhanced intensely after contrast administration suggesting an empiema. He required two surgeries (first through a burr-hole and later a craniotomy) and intravenous broad spectrum antibiotic coverage. All microbiological cultures were negative and so was panbacterial PCR. Histopathology showed fibrous tissue with signs of bleeding. He evolved favourably and was discharged after 6 weeks of antibiotic treatment. Literature review reveals many papers about SBS. They talk about prevention, pathogenesis, clinical manifestations, difficulties in diagnosis, treatment and legal and social implications. Many is told about blood appearance in CT scan depending on time elapsed between injury and onset of symptoms but none about contrast enhancement in child abuse. Anyway it is known that chronic subdural hematomas can enhance with contrast.

Conclusion: Diagnosis of SBS is challenging for clinicians. In this case, as we never had microbiological nor histopathological evidence of infection, we think that fever was due to the pneumonia and we attribute the contrast enhancement to inflammatory phenomena within the hematoma. We may attribute these changes to an unfavourable evolution of the initial abuse.

Keywords: Shaken Baby Syndrome, Child abuse, Subdural hematoma, Subdural empiema, retinal hemorrhages

P-113

Trauma

Rapid spontaneous resolution of acute epidural hematoma underlying frontal sinus fractureBurak Kinali¹; Volkan Murat Ünal¹; Senem Alkan Özdemir²; Ali Karadağ¹; Nail Özdemir¹.¹Tepecik Research And Education Hospital, Neurosurgery, İZMİR; ²Dr. Behçet Uz Children's Hospital, Neonatology, İZMİR

Objective: Acute epidural hematomas plays a major role in mortality and morbidity due to head traumas. Deciding the treatment option mostly depends on the patients neurological status and radiological findings. Eventhough there is mass effect of such patients, in the last years there are several writings about rapid resolution of the hematoma and related mechanism had been published. In our case, we will report a young patient who had a frontal epidural hematoma with rapidly resolution underlying the frontal fracture.

Methods: A 15 year old boy had admitted to our hospital due to a head trauma occured by falling from his bicycle and hitting his forehead on the asphalt road. His glasgow coma scale was 14 and there were no other neurological deficit. The initial CT scan showed up a left frontal epidural hematoma measured as 15 mm x 50 mm underlying a frontal sinus fracture. Due to patients somnolance and epidural hematoma with the maximal thickness of 15 mm we took the patient to operating room within 2 hours and we did the craniotomy in half an hour.

Results: After the craniotomy, we expected to see a grown extradural hematoma whereas there were no blood in the epidural space when we raise the bone flap. Dura was completely intact and was white in colour.

Conclusion: Eventhough there are more papers about rapidly remission of epidural hematomas in the last years by different kind of mechanisms, surgical treatment is still the gold standart. Just like in our case, we should keep in mind that patients with epidural hematoma related to frontal sinus fracture has a potential of rapid remission from the frontal sinus.

Keywords: epidural hematoma, trauma, rapid resolution, frontal sinus fracture

P-114

Trauma

Cranio-cerebral birth injuries in term newborn infants: a retrospective series

Nachtergaele Pieter¹; Van Calenberg Frank¹; Lagae Lieven².

¹Department of Neurosurgery, University Hospital of Leuven, Leuven; ²Department of Pediatric Neurology, University Hospital of Leuven, Leuven

Objective: In an attempt to further define the spectrum of cranial birth injuries, we analyzed 21 consecutive cranial birth injuries seen at our institution.

Methods: We performed a retrospective chart review from the medical records of our department from 1994 to 2015. We included 21 infants of 36 weeks gestational age or older with a diagnosis of cranial birth injury (severe cephalhematoma, skull fracture including growing fracture, epidural, subarachnoid and subdural hemorrhage, intracerebral and intraventricular hemorrhage and other types of brain injury). Types of injuries and their locations, presenting signs and symptoms and their timing, and what treatment(s), if any, were required, were recorded. Various maternal and neonatal factors (Apgar scores at 1 and 5 minutes, gestational age, birth weight, birth length, head circumference and parity) and the mode of delivery (spontaneous vaginal delivery, elective C-section, urgent C-section, use of forceps or vacuum extraction) were recorded.

Results: The most common initial presentations were swelling of the skull (43% of cases) and seizures (19% of cases). Average Apgar scores were 6.45 at 1 min and 8.4 at 5 min; 48% of children had abnormally low Apgar scores at 1 min. The most common intracranial lesion were skull fractures (33%). Neurosurgical treatment was required in 11 infants (52%). One infant in our series died. Assisted mechanical delivery by either forceps and/or vacuum extraction occurred in 43% of infants. In comparison, in the year 2013 only 13.97% of deliveries at our institution were mechanically assisted.

Conclusion: Although this series is too small to make firm conclusions, it is remarkable that the rates of assisted mechanical deliveries in our series far exceeded the rates at our institution in the year 2013.

Keywords: Cranio-cerebral birth injuries, term newborn infants, neurosurgery

P-115

Trauma

Demonstration of Traumatic Subarachnoid Hemorrhage from the Cisternal Segment of the Anterior Choroidal Artery: A Case Report

Ki Bum Sim¹; Chang Sub Lee¹; Tae Ki Yang¹; Sukh Que Park².

¹Jeju National University Hospital, Jeju; ²Soonchunhyang University Seoul Hospital, Seoul

Objective: We present a case of angiographically confirmed transection of the cisternal segment of the anterior choroidal artery (AChA) associated with a severe head trauma in a 15-year old boy.

Methods: The initial brain computed tomography scan revealed a diffuse subarachnoid hemorrhage (SAH) and pneumocephalus with multiple skull fractures. Subsequent cerebral angiography clearly demonstrated a complete transection of the AChA at its origin with a massive extravasation of contrast medium as a jet trajectory creating a plume.

Results: We speculate that severe blunt traumatic force stretched and tore the left AChA between the internal carotid artery and the optic tract. In a simulation of the patients brain using a fresh-frozen male cadaver, the AChA is shown to be vulnerable to stretching injury as the ipsilateral optic tract is retracted.

Conclusion: We conclude that the arterial injury like an AChA rupture should be considered in the differential diagnosis of severe traumatic SAH.

Keywords: Traumatic subarachnoid hemorrhage, Anterior choroidal artery, Transection, Angiography, Cadaver

P-116

Trauma

Children with fatality injures–assessment of violence and abuse in island: focusing head injury

You-Nam Chung¹; Ki Bum Sim¹; Hyun Wook Kang².

¹Department of Neurosurgery, Scholl of Medicine, Jeju National University, Jeju City; ²Department of Forensic Medicine, Scholl of Medicine, Jeju National University, Jeju City

Objective: Head trauma from child abuse is a devastating and potentially lethal form of infant and child physical abuse first recognized in the early 1970s. And recently, child abuse reported cases more increased death in infant and child.

Methods: In Jeju Island, 8 different child abuse autopsy cases are collected from the year 2000 to 2014 and analyzed on the basis of age, gender, offender, injury pattern, and cause of death. Autopsy was performed by a medical examiner by Pathologist.

Results: Among the 8 victims, the male to female ratio was 3 to 5, and their ages varied from 1 month to 9 years old. In terms of offenders, 6 out of 8 cases are involved with biologic parents, and one case was committed by stepmother; in addition, there was one unnatural death case. The causes of death were brain injury, hemorrhagic shock, and asphyxias, strangulation. It is necessary to make an anticipatory guide for the child abuse. The perpetrators are socio-economic hierarchy in the age of 25-38 years old were included low-income, less educated were nervous and mental disorders as schizophrenia, the two cases is the case with uicheojeung. Before family members were killed, including 2 cases of suicide by the father, by selecting the pessimism in a difficult economic situation.

Conclusion: Clinicians should be able to recognize suspicious injuries, perform a comprehensive examination and auxiliary tests, detect injuries, report child abuse, and document injuries for legal use. Data from 8 autopsy cases were compared to report on child abuse by the Ministry of Health and Welfare, published in 2013.

Keywords: Child abuse, abusive head trauma, autopsy

P-117

Trauma

Severe dural tear and brain injury after vacuum extraction birth

Sebastian Senger¹; Joachim Oertel¹; Stefan Linsler¹.

¹Saarland University Hospital, Department of Neurosurgery, Homburg

Objective: Traumatic brain lesions after vacuum extraction delivery are rare. Most common complication after operative vaginal deliveries is subgaleal hematoma applied by trained obstetricians.

Methods: There are only single case reports about major complications after vacuum extraction delivery in the literature available. The authors present a case of a severe brain damage and dura tear after vacuum extraction delivery.

Results: In the presented case surgical treatment of the dural tear was required. The child recovered completely in follow up.

Conclusion: Long-term follow-up of babies born via vacuum extraction who experienced brain damage usually shows complete resolution of hemorrhage and show a regular neurological development. Surgery is only required in rare cases of severe dural ruptures preventing persisting CSF fistulas.

Keywords: vacuum extraction, birth, brain injury, dural tear

P-119**Trauma****Skull base sport injuries in young athletes**

Nikolaos Syrmos¹; Argyrios Mylonas²; Charalampos Iliadis¹; Georgios Gavridakis³; Vasileios Valadakis¹; Kostantinos Grigoriou¹; Dimitrios Arvanitakis¹.

¹Neurosurgery Department, Venizeleio General Hospital, Heraklion, Crete; ²Department of Anatomy, School of Sports Science, Aristotle University of Thessaloniki, Macedonia; ³Ct-Scan Department, Venizeleio General Hospital, Heraklion, Crete

Objective: Aim of this study was to review cases of skull base sport injuries in young athletes.

Methods: During a 14 year period (2000-2014) 10 young individuals (<30 years) with skull-base injuries- fractures during sports activity were admitted to our hospital. The injuries resulted mainly from falls during sea related sports and motor-cycling activity, ball sports and contact sports activity.

Results: Emergency CT-scan was performed in all 10 cases (100 %). Surgery was required in 3 cases of severe injuries. Other injuries (leg and arm) were presented in 5 cases (50 %).

Conclusion: Accurate initial support and primary health aid care for and skull base injury young patients appears to be necessary.

Keywords: skull base, injuries, athletes, trauma

P-120**Trauma****Traumatic athletic brain injuries in rural health**

Nikolaos Syrmos¹; Argyrios Mylonas²; Andreas Televantos¹; Nikolaos Kapoutzis¹.

¹Neurosurgery Department, Venizeleio General Hospital, Heraklion, Crete; ²Department of Anatomy, School of Sports Science, Aristotle University of Thessaloniki, Macedonia

Objective: Aim of this study was to review cases of brain injuries during sports activity in children and in young adults (<30 years old)

Methods: During a 10 year period (2000-2010), 26 individuals with traumatic brain injuries during sports activity were admitted to our hospital. The injuries resulted mainly from falls, motor-cycling activity, ball sports and contact sports activity

Results: Neurological examination and radiological evaluation was performed in all 26 cases (100 %). Further investigation and transportation was required in 8 cases. Other injuries (leg and arm) were presented in 13 cases.

Conclusion: Accurate initial support and primary health aid care for patients with appears to be necessary for patients with traumatic brain injuries during sports activity.

Keywords: brain injuries, athletes, trauma, rural health

P-121**Trauma****Management of combined brain and spine sport injuries**

Nikolaos Syrmos¹; Argyrios Mylonas²; Charalampos Iliadis¹; Georgios Gavridakis³; Vasileios Valadakis¹; Kostantinos Grigoriou¹; Dimitrios Arvanitakis¹.

¹Neurosurgery Department, Venizeleio General Hospital, Heraklion, Crete; ²Department of Anatomy, School of Sports Science, Aristotle

University of Thessaloniki, Macedonia; ³Ct-Scan Department, Venizeleio General Hospital, Heraklion, Crete

Objective: Aim of this study was to review cases of combined brain and spine during sports activity.

Methods: During a 6 year period (2004-2010), 16 young individuals (<20 years) with combined brain and spine injuries during sports activity were admitted to our hospital. The injuries resulted mainly from falls during sea related sports, motor-cycling activity, ball sports and contact sports activity.

Results: Emergency CT-scan was performed in all 16 cases (100 %). Surgery was required in 2 cases of severe brain injuries. Other injuries (leg and arm) were presented in 11 cases.

Conclusion: Accurate initial support and primary health aid care for patients with appears to be necessary for patients with combined brain and spine injuries.

Keywords: brain injuries, athletes, trauma, spine injuries

P-122**Vascular****Encephaloduroarteriosynangiosis for cerebral proliferative angiopathy in a child**

Patricia Puerta¹; Antonio Guillén¹; Jordi Muchart²; Verónica González³; Santiago Candela¹; Enrique Ferrer¹.

¹Department of Pediatric Neurosurgery. Hospital Sant Joan De Déu, Barcelona; ²Department of Radiology. Hospital Sant Joan De Déu, Barcelona; ³Department of Neurology. Hospital Sant Joan De Déu, Barcelona

Objective: Cerebral proliferative angiopathy (CPA) is a rare vascular disease considered as separate from classical brain AVMs. It usually occurs in adolescent and middle-aged females. There are few case reports in the pediatric population. We present a case of a young child with papilledema in association with CPA, a previously unpublished clinical condition.

Methods: We present a child affected of CPA and we describe our management.

Results: An 8-year-old boy presented to the emergency room for acute left hemiparesis and headache which lasted two hours. Fundoscopy demonstrated bilateral papilledema. MRI revealed a diffuse network of densely enhancing vascular spaces over the right cerebral hemisphere and left basal ganglia. Angiography showed a widespread angiopathy fed by multiple arteries, suggestive of CPA. A lumbar puncture was performed and was noted to have an elevated CSF pressure. He underwent a lumbar-peritoneal shunt. One month later, papilledema disappeared. However, his symptoms worsened over the next months with disabling headache and becoming more frequent and lasting TIAs. We decided to operate him because of the possibility of a progressive ischemic course in the future. An encephaloduroarteriosynangiosis (EDAS) and encephalomyosinangiosis (EMAS) were performed. Clinical evolution was satisfactory during one year of follow-up.

Conclusion: CPA is a rare entity occurring in 3.4% of patients with AVMs. Patients usually present with headaches, seizures and progressive neurological deficits due to ischemia with vascular steal. Lasjaunias et al have defined the angioarchitectural features unique to CPA malformations: large-sized, nidus which has a classical appearance with scattered “puddling” of contrast, and absence of dominant arterial feeders. Treatment indications should be established very strictly and confined to hemorrhage, fragile angioarchitecture, uncontrollable seizures, and disabling headaches. Because the main mechanism of this disease is ischemia, therapies that increase cortical blood supply, like synangiogenesis or calvarial burr holes, can be indicated.

Keywords: Cerebral Proliferative Angiopathy, Children, Encephaloduroarteriosynangiosis

P-123

Vascular

Choroid plexus arteriovenous malformation in a preterm neonate
Volkan Murat Ünal¹; Nail Özdemir¹; Senem Alkan Özdemir²; Özkan İlhan³.

¹Tepecik Research and Education Hospital, Neurosurgery, Izmir; ²Dr. Behçet Uz Children's Hospital, Neonatology, Izmir; ³Tepecik Research and Education Hospital, Neonatology, Izmir

Objective: Arteriovenous malformations (AVM) of the choroid plexus are rare. This pathology mostly manifest as intraventricular hemorrhage. An extensive review of the literature revealed that only seven other cases have been reported. We report the eighth case of choroid plexus AVM presenting with intraventricular hemorrhage. Moreover, this is the first report of choroid plexus AVM in preterm neonate. The aim of this article is to report our case and discuss the diagnosis, surgery, treatment and prognosis.

Methods: The patient was a 1610-grams female preterm of 33 weeks' gestation delivered caesarean section. She needed mechanic ventilation for one month. She had an increased head circumferences after ten days of the birth. Computed tomography (CT) of the brain demonstrated intraventricular hemorrhage (IVH) in the third ventricle and hydrocephalus. A third ventriculostomy was planned when her weight was 2500 grams.

Results: A preterm, small for gestational age neonate infant with hydrocephalus was underwent third ventriculostomy. During the surgery, the lesion was diffuse, large, reddish and vascular. The lesion closing the entrance of foramen Monroe was coagulated and taken biopsy via endoscopic approach. Post operative period was uneventfully. The histological diagnosis was AVM of the choroid plexus. She was doing well six months after surgery. Postmenstrual age was seven months and her weight was six kilograms. There was no neurological deficit.

Conclusion: To our knowledge, a preterm neonate with a choroid plexus AVM has not been reported in the literature. Choroid plexus AVM presenting with IVH is rare, since the diagnosis of this pathology is difficult and may be mistaken. Endoscopic approach is appropriate for a clinically stable patient with an IVH if hydrocephalus is present.

Keywords: AVM, choroid plexus, hydrocephalus, IVH, preterm

P-124

Vascular

Extracranial internal carotid artery pseudo aneurysm presenting as life threatening hematemesis: a case report and literature review.
Rakesh Ranjan¹; Shambhaji Pawal¹.

¹Aditya Birla Memorial Hospital, Pune

Objective: To describe an extremely rare case of extra cranial internal carotid artery (ICA) pseudo aneurysm presenting as massive hematemesis in a pediatric patient and discuss about the management and outcome.

Methods: A 12 years old male child was referred to the gastroenterology department following two episodes of hematemesis. The episodes were followed after lifting a heavy bag on neck and shoulder 10 days ago. There was no other significant history. An upper GI endoscopy was done which showed telangiectasis in the stomach. A bronchoscopy was done which was normal. A contract CT scan of abdomen was normal. Within a day of admission, he had massive hematemesis leading to hemorrhagic shock. He was intubated, resuscitated with blood transfusion and put on inotropic support. Fresh bleeding was noted from the oral cavity. He became unconscious with left hemiparesis and developed anisocoria. A contrast CT scan of neck revealed a large pseudo aneurysm of the right ICA (2.2 cm diameter) at the level of C2 vertebrae with a daughter sac. CT scan of the brain in the same setting revealed evolving right ICA infarct with hemiaton. Options of stenting versus open control of pseudo aneurysm was considered. In view of high location and lesser morbidity, a

cerebral DSA was done and sapphire stent was placed in the ICA across the neck. The child underwent right decompressive craniectomy and intracranial pressure monitoring. Within few hours, the hypotension was corrected, inotropes were withdrawn. He showed progressive recovery and was conscious.

Results: The child made a slow but gradual recovery with residual hemiparesis of left side.

Conclusion: Pseudo aneurysm of extra cranial ICA is a rare clinical condition. Presentation as massive hematemesis is extremely rare and potentially fatal. A timely and accurate diagnosis and endovascular control of the aneurysm with aggressive control of ICP is key to recovery.

Keywords: Pseudo aneurysm, Internal carotid artery, Hematemesis, Carotid stenting

P-125

Vascular

Moya-Moya disease in children

Federico Bianchi¹; Luca Massimi¹; Paolo Frassanito¹; Gianpiero Tamburrini¹; Concezio Di Rocco²; Massimo Caldarelli¹.

¹Pediatric Neurosurgery, Catholic University Medical School, Rome; ²INI, Hannover

Objective: Moya-Moya (MM) is a rare disease characterized by progressive occlusion of the anterior Willis' circle. Direct and indirect revascularization techniques are available to treat this condition. The effectiveness of indirect revascularization by means of encephalomyosynangiosis (EMS) with/without accessory burr holes is here assessed.

Methods: During the 2002-2015 period, 28 children (15M + 13F, 5.5 years mean age with a 3 months-15 years range) have been consecutively operated on at our Institution. MM was associated to other syndromes in 7 cases. Seizures (35,7%), transient ischemic attack (28,6%) and focal neurological deficits (25%) were the main onset symptoms; the diagnosis was incidental in 3 cases (10,7%). Bilateral carotid occlusion was found in 22 cases and an unilateral MM in the remaining 6. The posterior Willis' circle was involved in no patients. Overall, 46 surgical procedures were performed: EMS with accessory burr holes in 41 cases, EMS alone in 4 cases, burr holes alone in one case.

Results: All patients presented a postoperative improvement of their preoperative symptoms after a mean 6-years follow-up. Such an improvement was detected starting from the first months after surgery. Post-operative brain arteriography demonstrated a good new arterial network in all cases in spite of the progression of the carotid occlusion. Postoperative MRI showed the occurrence of new ischemic areas in 1/3 of cases without clinical sequelae. There were no postoperative complications except for one case of contralateral subdural hematoma.

Conclusion: EMS, especially if completed by accessory burr holes, represents a safe and effective treatment for MM in the pediatric age.

Keywords: moya moya disease, encephalomyosynangiosis, burr holes, carotid artery

P-127

Vascular

Arteriovenous fistula causing hydrocephalus by venous hypertension: case report

Mario Garcia-Conde¹; Vanessa Hernandez-Hernandez¹; Liberto Brage-Martin¹; Pablo Febles-Garcia¹; Julio Plata-Bello¹; Ayoze Doniz-Hernandez¹.

¹Hospital Universitario De Canarias, La Laguna

Objective: We present a 3 year old male with a rare case of hydrocephalus secondary to an arterio-venous fistula which drained directly to superior sagittal sinus. This case clearly shows the relationship between venous congestion and hydrocephalus.

Methods: A 3 year old child with no past or family medical history was sent from the paediatrics service for progressive macrocephaly and moderate psychomotor retardation. The physical examination revealed positional plagiocephaly and a head circumference of 57 cm with abundant venous vasculature in frontal region noticeable by just clinical observation. An MRI was performed and showed the presence of hydrocephalus and an extensive arteriovenous fistula whose main contributions were the anterior and middle cerebral arteries from the left side draining into the superior sagittal sinus, having a varicose appearance (up to 3.15 cm in diameter). A Chiari Type I malformation with tonsillar descent to C2-C3 space was also noted. Cerebral angiography established the existence of a fistula between the left anterior and middle cerebral arteries and superior longitudinal sinus validating previous findings seen on MRI.

Results: The patient was treated with coil embolisation in two sessions in order to diminish the risk of complications due to the sudden change of the vascular flow. The left anterior cerebral artery was occluded first followed by the middle cerebral artery. The procedure was conducted without complications and the patient was discharged without associated problems. The post-procedure MRI showed significant changes, with improvement of hydrocephalus and Chiari malformation. The cognitive and psychomotor development of the patient also improved.

Conclusion: Hyperaemic hydrocephalus due to arteriovenous fistula is a very rare condition in children. Its treatment is crucial because those children become dramatically affected by intracranial hypertension. The endovascular approach seems to be the most secure and effective to close the fistula and to reduce the hydrocephalus and Chiari malformation.

Keywords: hydrocephalus, arteriovenous fistula, chiari malformation, endovascular treatment, venous congestion

P-129

Epilepsy

Long-term efficacy of vagal nerve stimulation in children with epilepsy; a single institute experience

Mahmoud Taha¹; Raidah Baradi¹; Tarek Jallul¹.

¹King Fahad Specialist Hospital - Dammam, Dammam

Objective: Vagal nerve stimulation (VNS) is an adjunctive therapy for improving seizure control in patients with epilepsy. Our objective is to present the long term outcome of this device in our institute. This report describes the efficacy of VNS therapy in a series of intractable patients with partial and primarily generalized epilepsy.

Methods: Entrance criteria included video/EEG proof of seizure type, MRI and seizure frequency documentation for at least 3 months prior to implant, follow-up data available for at least 18 months post implant. Seizure frequency, side effects, and VNS settings were collected at each visit. Using a retrospective chart review, 15 patients were evaluated for intractable epilepsy that were in our care from 2010 to 2012 at KFSHD. The following information was collected from past neurology chart notes: demographics, type of pre-implantation evaluation, VNS parameters, VNS side effects, diagnosis, seizure types, number of seizure and seizure frequency reduction prior to and after VNS therapy, and number of AEDs tried prior to and after the implantation. Patients were considered to be responders if they had a fifty-percent reduction in seizures. Our pre-implantation evaluation consisted of continuous video-EEG monitoring, neuropsychometric testing, and an MRI with epilepsy protocol.

Results: 3 patients (20%) were seizure free at the last follow up after VNS implantation, (n=3, 20%) had 50-90% seizure reduction, (n = 4, 26.7%) had 25-50% seizure reduction, and (n = 4, 26.7%) did not improve. 2 patients had normal EEG at the last follow up after VNS implantation, and (n = 3, 26.7%) improved by 25%. No significant adverse event was reported.

Conclusion: VNS is a safe and effective treatment for children with pharmacoresistant epilepsy who are not candidates for epilepsy surgery. The VNS dose needs to be individualized.

Keywords: vagal nerve stimulation, epilepsy, refractory seizures

P-130

Epilepsy

Seizures associated with brainstem ganglioglioma: a review

William Gump¹.

¹Kosair Children's Hospital, Louisville

Objective: Gangliogliomas are known to be highly epileptogenic tumors, and most commonly occur in the temporal lobe. A small percentage arise as intrinsic brainstem tumors. The brainstem is not typically considered an epileptogenic zone.

Methods: A previously healthy 9 year old boy with several months of progressive nausea/vomiting and hand paresthesias was found to have a large dorsally exophytic intrinsic brainstem tumor. Neurologic exam was normal. There was no personal or family history of seizures. He was taken to surgery for tumor debulking, relief of obstructive hydrocephalus, and tissue identification.

Results: Radiographic gross total resection of the enhancing and cystic portions of the tumor was achieved. Pathologic diagnosis was WHO grade 1 ganglioglioma. Postoperatively, the patient remained intubated due to intermittent apneic episodes which seemed to be associated with staring spells, and was noted to have frequent jerking movements involving his upper and/or lower extremities independently. Prolonged video EEG monitoring demonstrated abnormal bifrontal rhythmic delta and theta activity. Levetiracetam therapy was initiated, which abolished the jerking movements and apnea. The postoperative course was otherwise complicated by a right vocal cord paresis. Gangliogliomas are known to be strongly associated with epilepsy, but very rarely arise intrinsic to the brainstem. Prior reports have not described seizures as presenting symptom or postoperative complication from brainstem ganglioglioma. Postoperative seizures from other dorsally exophytic brainstem tumors are rarely reported. Gross total resection can be curative of epilepsy with supratentorial ganglioglioma, and improves overall and progression-free survival. The epileptogenic potential of this tumor in the infratentorial compartment is not well characterized.

Conclusion: The epileptogenic potential of ganglioglioma may not be limited based on tumor location. Seizure prophylaxis should be considered with this diagnosis, independent of anatomic location. Overall long term prognosis is favorable.

Keywords: epilepsy, ganglioglioma, brainstem tumor

P-131

Epilepsy

Clinical characteristics, surgical and neuropsychological outcomes in drug resistant tumoral temporal lobe epilepsy in children: an observational study

Dattatraya Muzumdar¹; Sangeeta Ravat²; Vivek Iyer²; Yogesh Ghodge²; Urvashi Shah²; Neena Sawant³.

¹Department of Neurosurgery, Seth G.S.Medical College and King Edward VII Memorial Hospital, Mumbai; ²Department of Neurology, Seth G.S.Medical College and King Edward VII Memorial Hospital, Mumbai; ³Department of Psychiatry, Seth G.S.Medical College and King Edward VII Memorial Hospital, Mumbai

Objective: Glioneuronal tumors are found in nearly one third patients who undergo surgery for pharmacoresistant epilepsy with temporal lobe being the most common location. Few studies, however have concentrated on the neurological and neuropsychological outcomes after surgery, hitherto none from India.

Methods: We studied 34 patients with temporal lobe tumors and drug resistant epilepsy. These patients underwent anterior temporal lobectomy or lesionectomy based on the involvement of the hippocampus and mesial temporal structures. The clinical history, EEG, neuropsychology profile and MRI were compared. Seizure outcome was categorized using Engels classification.

Results: The mean age at surgery was 20 years. At a mean follow up of 62 months, 85.29% of the patients were seizure free (Engels Class I). All 8 patients with intraoperative electrocorticography (ECoG) guided resection were seizure free. Presence of a residual lesion was significantly associated with persistence of seizures post surgery ($p=0.002$). Group analysis revealed no significant shifts in IQ and memory scores postoperatively. There was a significant improvement in the quality of life scores (total and across all subdomains) in all patients ($p<0.001$). Postoperative EEG abnormalities predicted unfavourable seizure outcome

Conclusion: Surgery for temporal lobe tumors and refractory epilepsy offers complete seizure freedom in majority. Complete surgical excision of the epileptogenic zone is of paramount importance in achieving seizure freedom. Intraoperative electrocorticography (ECoG) is a useful adjunct to ensure complete removal of epileptogenic zone, thus achieving optimal seizure freedom. There is a significant improvement in the quality of life scores ($p<0.001$) with no negative impact of surgery on memory and intelligence. Even the patients who are not seizure free can attain a worthwhile improvement post surgery.

Keywords: Temporal lobe tumors, Neuropsychological outcomes

P-132

Epilepsy

An evolutionary approach to febrile seizures and thermoregulation Alexandra Kunz¹

¹Harvard University (Extension), Cambridge

Objective: Febrile seizures (FS) are always a relevant topic; thermoregulation and febrile responses, complex processes, are important aspects of the unsolved puzzle.

Methods: Here, FS are explored from comparative “evolutionary pressure” data-sets for insights/contributing factors to age-dependent vulnerability.

Results: Thermoregulatory responses evolutionary quest is for maximal performance at optimal temperature, experimentally shown for insects/viruses population growth, not performance. Relying on external heat sources, ectotherms narrow range of performance thermal-sensitivities is explained by natural selection, not thermodynamics; endotherms, birds/mammals, thermally constrained set-points evolved promoting heat loss, not enhancing performance. Mammalian brains selective brain cooling (SBC) is a special evolutionary case within the thermal core because hyperthermia, causing febrile seizures, limits performance; SBC separates brain temperature (T) regulation independently from the body to keep $T_{\text{brain}} < T_{\text{trunk}}$, $p<0.01$. Species-specific SBC mechanisms during hyperthermia promote reversing normal blood flow, from brain→skin to skin→brain, to cool/maintain constant cerebral metabolism. A 4-part venous pathway connects extracranial diploic/emissary veins with intracranial meningeal veins/sinuses; the richly vascularized/complex

human diploe has an age dependent developmental pattern, fully established, age 5, large variations at each age. Primate emissary veins respond immediately to hyperthermia; their parietal/mastoid/condyloid/post-glenoid foramina prominence shifts in an evolutionary pattern: Tarsius 0%, 0%, 0%, 100%; Lemurs 0%, 74.4%, 0%, 99%; orangutan 3%, 81.6%, 1%, 2%; chimpanzee 8.7%, 14%, 16.5%, 0%; human 60.5%, 68%, 77%, 0.6%. Furthermore, intrinsic brain geometry plays an important evolutionary role in thermoregulatory patterns/heat distribution. Notably, perinatal discontinuity of ontological size/shape changes in chimps/humans at 2–4 months, $p<0.0044$, produces topographical changes in vascular system; an expanded human frontoparietal volume, now globular, with highest concentration of diploic/emissary veins, richly anastomosed/reticulated, affects heat dissipation. Brain surface: volume ratio values for chimps/humans heat loading, 1.59 vs 0.91, respectively, confirms globular shape decreases thermic values in heat transfer.

Conclusion: In light of evolution, human ontological variations offer an option to FS unsolved puzzle.

Keywords: febrile seizures, thromoregulation, evolution, brain geometry, globular shape

P-133

Functional

Intraoperative neuromonitoring during tethered cord operations in young children

Ezgi Erdogan¹; Huseyin Canaz¹; Ibrahim Alatas¹; Nursu Kara²; Kerem Ozel¹

¹Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul

Objective: Intraoperative neuromonitoring is an invaluable and efficient method to increase safety of surgery. Operations of children with spina bifida are now routinely being monitored with motor evoked potentials, electromyography and mapping techniques in our institution. The results of sixteen monitoring of tethered cord syndrome were presented in this paper.

Methods: Sixteen children aged between 1.5 month - 8 years were monitored during tethered cord surgery in Spina Bifida Center. Motor evoked potentials (MEP), continuous electromyography (EMG) and mapping with direct stimulation were used to intraoperative neuromonitoring. For MEP, transcranial stimulation was applied at C3/C4 and lower extremity muscles (quadriceps, tibialis anterior, gastrocnemius and anal sphincter) were monitored. EMG were recorded continuously from same muscles. Any repeated EMG activity were reported to neurosurgeon. Monopolar probe was used to stimulate any tissue to identify functional nerves and find out possible functional nerve fiber in filum. Since the current spreads to close nerve tissue above 5 mA, we preferred to increase the stimulation intensity not more than 5 mA.

Results: In 15 patients MEP recorded and in 13 of them we could elicited response at least one myotome successfully. In two infants that we couldnt get MEP response, were 1.5 months and 3.5 months aged. In 8 patient MEP responses from anal sphincter were elicited. Since the responses were fluctuating in amplitudes during the surgery, we preferred to use all or nothing criteria as our warning thresholds. No MEP change was observed and no new or worsen postoperative neurological deficit was occurred.

Conclusion: As we know from the literature, MEP has a low success in very young children due to incomplete neural maturation. According to our experience, MEP can be recorded safely and successfully in children older than 6 months. Mapping with direct stimulation assists surgeon for safer detethering.

Keywords: Intraoperative Neuromonitoring, Tethered Cord Syndrome, Young children

P-134

Functional

Intraoperative neurophysiological evaluation of roots and neural placode during myelomeningocele repair

Ezgi Erdogan¹; Huseyin Canaz¹; Ibrahim Alatas¹; Nursu Kara²; Kerem Ozel¹.

¹Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Department Of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul

Objective: The aim of our study was to evaluate the functions of roots and neural placode by intraoperative direct electrical stimulation during myelomeningocele repair and provide information to neurosurgeon for a safer surgery.

Methods: Roots and neural placode were stimulated in 10 infants with myelomeningocele (6 lumbosacral, 4 thoracolumbar levels). Continuous electromyography and triggered electromyography were performed throughout the operation.

Results: Muscle responses were elicited with monopolar root stimulation, intensities between 0.2 mA -2 mA for placode stimulation intensities between 0.6-4 mA. In 2 infants with distal muscle weakness, both proximal (quadriceps, tibialis) and distal (gastrocnemius, anal sphincter) muscle responses were obtained with stimulation of roots and placode. In two other infants with severe weakness at proximal muscles and plegia at distal muscles, proximal and distal muscle responses elicited with stimulation of roots but no response was obtained from distal muscles with stimulation of placode. In one of the neurologically intact infants we couldnt get any response from placode and in the other we could get a response with high intensity (4 mA) due to the non-functional tissue covering the placode. In 1 infant, stimulation of right distal region of placode induced right muscle responses however when we moved the probe proximally on placode we observed left side muscle responses.

Conclusion: In all patients we could obtain responses with stimulation of roots which shows that roots were functional in all patients. Stimulation of placode is a more difficult method because of the non-functional tissue blocking the current to pass to neural tissue. Infants with MMC may have a different spinal neuroanatomy due to neural migration defect and this is crucial for surgical manipulations during operation. Intraoperative mapping can guide neurosurgeon, increases the safety of surgery and contribute to the pathophysiology of neurological defects in those infants.

Keywords: Myelomeningocele, Intraoperative neurophysiology, root stimulation

P-135

Functional

Short-term results of selective dorsal rhizotomy for spastic cerebral palsy in GMFCS levels IV-V

Arcisse Comfort¹.

¹Clinical and Research Institute of Emergency Children's Surgery and Trauma, Moscow

Objective: Selective dorsal rhizotomy (SDR) is a well accepted neurosurgical option for the relief of spasticity interfering with motor function in children with spastic cerebral palsy (CP) in GMFCS levels I-III. However, little is known about efficacy of SDR in GMFCS levels IV-V. The object of this study was to evaluate the short-term (1 year) effects of SDR for children with CP in GMFCS levels IV-V.

Methods: This study group consisted of 40 children with CP in GMFCS levels IV-V, consecutively operated. Mean age was 4,75 years (range 2,5-7,0). They were all evaluated by a multidisciplinary team preoperatively and at 1, 2, 6 and 12 months postoperatively. These evaluations included quantitative, standardized assessment of lower-limb tone (Ashworth Scale), Gross Motor Function Measure (GMFM-66), and performance of skills and activities and amount of caregiver assistance by the Pediatric Evaluation of Disability Inventory (PEDI).

Results: After SDR statistically significant durable improvement in lower-limb muscle tone, gross motor function, and performance of skills and activities were found. In addition to the impact of SDR on the lower limbs non-statistically significant suprasegmental improvements were also found as well as improvements in speech and cognitive function.

Conclusion: SDR is a safe and effective procedure for reducing spasticity in children with CP in GMFCS levels IV-V. Based on improvement of muscle tone, gross motor function, and performance of skills and activities it creates the optimal condition for further rehabilitation for these patients.

Keywords: Selective Dorsal Rhizotomy, Cerebral Palsy, Spasticity

P-137

Evidence-based practice

Chronic lymphocytic inflammation with pontine perivascular enhancement responsive to steroids: a case report

Minichini Viviana¹; Detemmerman Dominique²; Nebbioso Andrea³; Loli Valentina⁴; Fontange Quitterie⁵; De Witte Olivier¹.

¹Department of Neurosurgery, Erasme Hospital (ULB), Brussels;

²Department of Pediatric Surgery, Erasme Hospital (ULB), Brussels;

³Department of Pediatrics, Reine Fabiola Children Hospital, Brussels;

⁴Department of Neuroradiology, Erasme Hospital (ULB), Brussels;

⁵Department of Pathological Anatomy, Erasme Hospital (ULB), Brussels

Objective: CLIPPERS is a recently defined inflammatory central nervous system (CNS) disorder of unknown aetiology, reported only in 60 cases so far. CLIPPERS typically presents with subacute progressive symptomatology related to brainstem, cranial nerve and/or cerebellar involvement. Characteristically, MRI consist of punctuate pattern of gadolinium enhancement in the pons, brainstem, cerebellum and sometimes of the spinal cord, extending variably into the supratentorial CNS structures. Laboratory tests may reveal non-specific abnormalities of the CSF, elevated ANA, histological features of vasculitis or a perivascular lymphohistiocytic infiltration. Differential diagnosis includes neurosarcoidosis, CNS lymphoma, granulomatosis, CNS vasculitis and others.

Methods: We report a case of a 13-year-old female child who had presented during the last 4 years three episodes of sudden progressive walking troubles, left seven nerve palsy and diplopia. MRI revealed multiples hyperintensive lesions of the white matter in the supra- and infratentorial CNS structures typical for CLIPPERS. Serial CSF analyses showed only moderate increase of proteins without any signs of infection, neoplastic cells or oligoclonal bands. The blood test was positive for ANA. Right frontal neuronavigation guided biopsy of the deep white matter was performed that revealed an inflammatory lymphocytic T-predominant perivascular infiltration.

Results: A good outcome has been achieved following an intravenous glucocorticosteroids treatment.

Conclusion: CLIPPERS is a rare neuro-inflammatory entity with unknown pathogenesis. Histopathological features combined with clinical and radiological response to glucocorticosteroids suggest an autoimmune-mediated process. Final diagnosis of CLIPPERS is challenging and requires from physicians careful exclusion of alternative ones. Brain biopsy should be considered in patients when alternative diagnoses remain probable.

Keywords: CLIPPERS, Steroids, Encephalitis

P-138

Hydatid disease

Lift suboccipital retrosegmental craniectomy for removal of lift cerebellar hydatid cyst & treatment of hydrocephalus with out CSF shunting. A case reportAl-Mutasim Bellah Etaiwi¹.¹Iraqi Board for Medical Specialization, Baghdad**Objective:** To remove hydatid cyst from posterior fossa without rupture & treatment of Hydrocephalus with out CSF shunting.**Methods:** A 14-year-old boy presented with headache & ataxia secondary to hydatid cyst involving the lift cerebellar hemisphere. He underwent lift suboccipital retrosegmental craniectomy for removal of hydatid cyst. The underlying hydrocephalus resolved after surgery with out need for CSF shunting.**Results:** Headache, ataxia, lift cerebellar signs, hydrocephalus resolved after surgery. No complications.**Conclusion:** We can attack the primary cerebellar lesion directly & hydrocephalus resolved without need for CSF shunting.**Keywords:** Hydatid cyst, posterior fossa, hydrocephalus, CSF shunting

P-139

Infection

Spontaneous perforation of intestine by peritoneal catheter, 16 months after ventriculoperitoneal shunt replacementGültekin Baş¹; Emre Özkara¹; Zühtü Özbek¹; Umut Alici²; Murat Vural¹.¹Eskişehir Osmangazi University Neurosurgery Department, Eskişehir;²Eskişehir Osmangazi University Pediatric Surgery Department, Eskişehir**Objective:** The peritoneal end of the ventriculoperitoneal (VP) shunt has been associated with complications such as pseudocyst formation, perforations of hollow viscus, penetration into solid organs and abdominal wall and protrusion outside body. Unfortunately, the shunt devices have a high incidence of malfunction mainly due to catheter obstruction or infection and are associated with various complications, 25% of which are abdominal. We report a case of small intestine perforation by VP shunt after 16 months of operation.**Methods:** A 16-month-old boy who had undergone VP shunt placement for obstructed hydrocephalus 15 month ago was admitted to emergency department by vomiting and breaking down of general condition. On examination, he was hypotonic, confused and his axillary temperature was 38,3°C. There was no signs of meningeal irritation like neck rigidity, and abdominal examination was normal.**Results:** The child was operated immediately and we saw that shunt shunt pump was filled with pus. We pulled ventricular catheter and it sent for microbiological examination. CSF was xanthochromic. Peritoneal catheter was pulled and we saw the end of peritoneal catheter was fecal contamination. We placed external ventricular drainage system and. Emergency laparotomy was done by pediatric surgeon and it was shown that catheter of VP shunt was perforated the small intestine from the terminal ileum (Fig 2). They repaired small intestine and after the operation child went to the critical care unit. Prophylactic antibiotics started. Escherichia coli was produced on his CSF microbiological examination. 9 days after operation he was died.**Conclusion:** It is evident that when the bowel perforation is detected and corrected at an early and asymptomatic stage, the prognosis is excellent. On the other hand this complication can turn to be fatal when infectious complications develop.**Keywords:** shunt, infection, complication, intestine, perforation

P-141

Infection

Foreign body granulomatous inflammation when using haemostatic materialsAleksandr Kim¹; Ivanov Vadim¹; Oleg Don¹; Kirill Burnin¹; William Khachatryan¹.¹Polenov Neurosurgical Institute - Federal North-West Medical Research Centre, St.Petersburg**Objective:** In case of reparative processes abnormality in a wound, contemporary haemostatic medications based on adsorbed gelatin, microfibrillar collagen or oxidized cellulose may cause nonspecific inflammation accompanied by formation of a foreign body granulomas. Sometimes these granulomas may be treated as recrudescence of tumor or an apostema, which complicates the choice of patients surveillance.**Methods:** The study describes 2 cases of formation of foreign body granulomas after usage of gelatin-based foam haemostatic agent combined with thrombin**Results:** The first patient, 4 years old, was operated for cavernoma in the left parietal lobe. The cavernoma was removed. When performing follow-up MRT 4 months after the operation an oval-shaped space-occupying mass was detected at the place of the removed cavernoma. A reoperation for removal of the space-occupying mass was executed. Histological and histochemical examinations showed a foreign body granuloma. The second patient, 16 years old, was operated for fibrillary astrocytoma of the right temporal lobe. When performing follow-up MRT 4 months after the operation a round-shaped space-occupying mass was detected. However, on completion of positron-emission tomography and magnetic resonance spectroscopy no data on blastomatous process were obtained. Glucose hypermetabolism was observed in the area of interest. Taking into account asymptomatic development of the expansive process, no reoperation has been executed. For six months the patient has been in stable condition. Histological examination in the first case showed that brain wound healing was accompanied by an aseptic inflammation with formation of a foreign body granuloma, with manifestations of focal destructive vasculitis, inhibition of granulation tissue maturation and abnormality of organization and encapsulation processes.**Conclusion:** When choosing treatment for patients with foreign body granulomas after neurosurgical operations, consideration should be given to stationary or regressive character of morphological changes attributable to them. Differential diagnosis with recurrence of tumor also remains relevant.**Keywords:** granulema, textiloma, gelatin-based foam, inflammation, foreign body

P-142

Craniofacial

Analysis of craniostylosis cases operated by Multi-directional Cranial Distraction Osteogenesis (MCDO) procedure: from the point of expansion rate of the skull volumeMasahiro Kameda¹; Shigeki Ono²; Kiyoshi Yamada³; Eijiro Tokuyama³; Isao Date¹.¹Department of Neurological Surgery, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama;²Department of Neurosurgery, Kawasaki Medical School, Okayama;³Department of Plastic and Reconstructive Surgery, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences, Okayama

Objective: In our hospital, neurosurgeons and plastic surgeons perform collaborative operations for treating patients with craniofacial disorders. Previously, we treated craniosynostosis patients by conventional cranioplasty and distraction osteogenesis. We have recently started Multi-directional Cranial Distraction Osteogenesis (MCDO) procedure in addition to conventional methods for craniosynostosis patients. We will show the result of craniosynostosis cases operated by MCDO procedure in this presentation.

Methods: Three craniosynostosis patients operated by MCDO procedure were included in this analysis. We analyzed shape of the skull, amount of distraction length and amount of the skull volume.

Results: One boy and two girls were operated (age of the operation: 1 to 5 year old, median 2 year). One of these cases was multiple-suture craniosynostosis. After the operation, the amount of distraction length was tailored piece by piece to obtain better shape of the skull (average of total amount of distraction: 13.7mm) One month after the distraction, MCDO system was removed. Compared to the preoperative state, 14% of skull volume was expanded after MCDO procedure. This expansion rate of skull volume was similar to the Apert patient case which we performed anterior expansion by conventional distraction osteogenesis method (expansion rate of skull volume 13%, amount of distraction length 22.2mm). The longest follow up period is 16 months. In this patient, expanded skull volume is still preserved, but, the osteotomy line is blurred gradually.

Conclusion: Compared to the conventional distraction osteogenesis, MCDO procedure is useful to get better shape of the skull, and moreover, this procedure is more cost-effective because it enables us to get similar expansion rate of the skull volume with the half amount of distraction length in the shortened hospital stay.

Keywords: craniosynostosis, Multi-directional Cranial Distraction Osteogenesis

P-143

Craniofacial

Secondary shunt-induced craniosynostosis

Kiril Georgiev¹; Assen Bussarsky¹; Jivko Surchev¹; Dilyan Ferdinandov¹.

¹Dept. of Neurosurgery, Univ. Hospital "St. Ivan Rilski", Medical Univeristy - Sofia, Sofia

Objective: One of the less common complications of CSF shunting in infancy is secondary craniosynostosis. It presents a significant diagnostic and therapeutic challenge. The authors review their experience with the treatment of this condition.

Methods: The records of 718 pediatric patients operated on during a 30 year period (1984 – 2014) were reviewed. 30 cases with secondary craniosynostosis were identified. Details on etiology of hydrocephalus, age at shunting, and shunt type were noted along with age at the diagnosis of craniosynostosis. Mean follow-up period of operated patients was 10.2 years (range 1.5 – 20 years).

Results: Mean age at shunting of the cases with secondary craniosynostosis was 3.52 months (range 1 – 9 months). Mean time to diagnosis of craniosynostosis was 7.1 months after initial shunting. Etiology of hydrocephalus and type of shunt did not influence the incidence of secondary craniosynostosis. 19 out of 30 patients were operated on. In 3 cases linear craniectomies were employed. In all other cases extensive calvarial remodeling using authors technique was used.

Conclusion: Shunt induced craniosynostosis is rare. Prompt diagnosis and appropriate treatment give acceptable cosmetic results

Keywords: secondary craniosynostosis, hydrocephalus, complications

P-146

Craniofacial

Cranial vault expansion with the split bone flap technique in shunt related craniosynostosis: our experience in Lyon

Pierre-Aurélien Beuriat¹; Christian Paulus²; Blandine Grassiot¹; Alexandru Szathmari¹; Carmine Mottolese¹.

¹Department of Pediatric Neurosurgery, Neurological and Neurosurgical Hospital Pierre Wertheimer, Lyon; ²Department of Oral and Maxillofacial Surgery, Mother and Children Hospital, Lyon

Objective: Shunt-related craniosynostosis causing craniocerebral disproportion represents a particular complication of the treatment of hydrocephalus. When the modification of the shunt opening pressure, does not improve the symptomatology, surgery for correction of craniocerebral disproportion is indicated. We present the results and advantages of the split bi-frontal bone technique that is a modification of the previous used frontal bone advancement technique

Methods: We retrospectively reviewed all the patients operated for a craniosynostosis in our institution from 1995 to 2012. Among those, we report a series of 5 patient treated with this technique that consists in a splitting bi-frontal bone flap that is left floating posteriorly to increase the cranial volume.

Results: All patients whom benefited of this procedure were cured. This technique increases the volume of the skull favoring the expansion of the brain and allows good cosmetic results. No peri-operative complications were reported

Conclusion: The fact that the brain has a more appropriate volume allows the disappearance of the clinical symptomatology related to the split ventricle syndrome. The advantage is the respect of the fronto-orbital shape in children and adults with the same results of the classic frontal advancement in term of volume expansion. The study of cerebral blood flow confirms the increase of the cerebral blood perfusion and the improvement of the regional cerebral blood flow in borderline regions. We preconize, for the treatment of slit ventricle syndrome, the bi-frontal split technique in children and adults for its efficacy, the low rate of complications and the respect of aesthetical constraints.

Keywords: craniocerebral disproportion, slit ventricle syndrome, hydrocephalus, iatrogenic craniosynostosis

P-147

Craniofacial

The management of chronic headaches in a patient with proteus syndrome.

Almas Ahmed¹; Chris Uff¹.

¹The Royal London Hospital, Barts Health, London

Objective: This patient presented as a 19 female with a chronic worsening headaches and nausea and lethargy. She was diagnosed with Proteus syndrome that had a non-communicating hydrocephalus and was treated with a left trans-parietal VPS at the age of 6 months. She presented to the emergency department with worsening headache and dizziness but no focal neurology.

Methods: After taking a history and examination the patient had a CT head and X-ray shunt series. The CT head showed left sided hyper density around the proximal catheter, no hydrocephalus, and the X-ray was suspicious of a disconnected VPS.

Results: Patient had a right sided VPS inserted 2 days after admission. She was clinically well after the operation however postop was complaining of headaches again. The pressure setting of her VPS has been changed multiple times and the headaches are now subsiding.

Conclusion: The patient suffered from a congenital hydrocephalus. Patient with Proteus syndrome have been reported to having periventricular cysts, seizures and mental retardation. This patient had no such cysts but had multiple meningioma's no cause was found for the hydrocephalus however a result of a non-functioning VPS caused increasing headache even though her CT head showed no evidence of a dilated ventricular system.

Keywords: Hydrocephalus, VPS, congenital

P-148

Craniofacial

3D numerical modeling of spring cranioplasty: a tool for predicting outcomes and help surgical planning.

Alessandro Borghi¹; Silvia Schievano¹; Richard Hayward²; David Dunaway²; Owase Jeelani².

¹Institute of Child Health, London; ²Great Ormond Street Hospital, London

Objective: Spring Cranioplasty for the treatment of sagittal craniosynostosis is a valid alternative to the traditional total calvarial remodeling. This procedure has proved functionally and aesthetically effective in correcting skull deformities; however, the final outcomes remain moderately unpredictable due to an incomplete understanding of normal craniofacial mechanics and unsophisticated distractor design. In this work, a computational model of spring cranioplasty is presented and validated: the results show that spring expansion is predictable by means of computational modelling.

Methods: A 5 month old boy diagnosed with scaphocephaly was surgically treated with springs in March 2014, following CT assessment. During surgery, local skull thickness, position and dimension of the osteotomies, type and model of the springs were recorded. CT data were processed in order to create a 3D model of the patient which was imported into software for numerical modeling (Ansys Mechanical - Ansys Inc.). Predefined linear spring connection conditions were used to simulate the effect of spring expansion: the cranioplasty spring loading curves were retrieved using an analytical method previously developed.

Results: The opening of the anterior and posterior spring were calculated and compared with the measurements 3% and 16% differences between predicted and on-table measurement of spring opening were found, for anterior and posterior spring respectively. Good quantitative and qualitative agreement between the simulations, on-table measurements and calvarial 3D scanning performed after surgery was found. The use of a viscoelastic model allowed prediction of the time dependent adaptation of the skull to the spring forces, with good agreement with spring measurements performed on planar x-rays.

Conclusion: The model accurately predicted the on-table spring opening as well as the spring kinematics. It will be used as tool for improving surgical planning as well as for designing novel, more sophisticated spring distractors.

Keywords: spring cranioplasty, finite element modeling, segmentation

P-149

Craniofacial

Comparative analysis of a group of craniosynostosis cases, with patients following of 7 years.

Ingrid Trujillo¹; Sebastián Viguera¹; Ruben Muñoz¹; Fernando Perez¹.

¹Hospital Regional De Concepcion, Concepcion

Objective: The main goal of this study is to analyze 45 patients of craniosynostosis in the clinical hospital of Concepción, Chile. All patients operated from 2008 to 2015 were children ranging in age from 6 months to two years, followed for 1-7 years after surgery.

Methods: We select for this work 13 (31%) scaphocephaly, 11(26%) trigonocephaly, 10 (24%) plagiocephaly and 7 (17%) braquicefalías. were performed 37 (88%) cranial remodeling, 2 (5%) unilateral frontorobitarios progress 27 (64%) bilateral frontoorbitarios progress, 8 (19%) scaphocephaly remodeling and 5 (12%) scaphocephaly wires for the treatment of the reported cases.

Results: For bone fixation and stabilization we used titanium plates and resorbable material, achieving a greater comfort for patients the resorbable plates. The aesthetic results were tabulated in good, fair and bad responding to the harmony and symmetry of temporoparietal cranial contour, frontorobitaria and frontal boundary. Where good achieve all symmetries of contours, regular does not achieve all the symmetry of contours, but does not require a second surgery and the bad, a second surgery is required. Main results of all procedures was aesthetically good (93%). We did not get the same results with springer, where a good temporoparietal contour was not achieve in 2 (5%) cases, the same applies a with the unilateral progress bar frontoorbitario plagiocephaly 1 (2%), this last one having a regular result, which was not achieve a good frontoorbitario contour.

Conclusion: Among the complications, we had only one serious (2%) complication of stroke. In conclusion, based on our experience, independent of the technique used and age of the patient, cranial remodeling is aesthetically effective and predictable in their long-term results.

Keywords: craniosynostosis

P-150

Craniofacial

Eosinophilic granuloma of a child

Özgür Demir¹; Erol Öksüz¹; Fatih Ersay Deniz¹.

¹Gaziosmanpaşa University, Department of Neurosurgery, Tokat

Objective: Eosinophilic granuloma is part of Langerhans cell histiocytosis. It is a rare, benign bone tumor, often involving the head bones. Most patients present between 5 and 15 years of age. Lesions of eosinophilic granuloma may disappear spontaneously. Painful lesions can be treated with radiotherapy, chemotherapy, surgical curettage of the tumor or local infusion of cortisone.

Methods: A six year old girl presented to our clinic with severe painful swelling on the right parietal region with a gradual increase in size and frequent episodes of headache for 2 months. Radiological findings demonstrated right parietal cranial lytic lesion with intra and extracranial extension (Figure 1, 2). Whole body bone scintigraphy showed no bone lesion except right parietal region (Figure 3).

Results: The patient was treated with gross total excision of tumor and cranioplasty without any adjuvant therapy. Histopathology of the tumor showed cellular components of Langerhan cells admixed with chronic cellular infiltrate and eosinophils. Findings were consistent with eosinophilic granuloma. The patient recovered well with no known recurrence till date (Figure 4).

Conclusion: There is no definitive treatment option for this disorder. Treatment option may be changed depending on the extent of the disease and the symptoms. We recommend surgical excision in the presence of intracranial extension of painful lesion.

Keywords: Eosinophilic granuloma

P-151

Craniofacial

Experience of applying minimally invasive methods of correction of children's craniosynostosis

Aleksandr Kim¹; Vadim Ivanov¹; Konstantin Samochernykh¹; William Khachatryan¹.

¹Polenov Neurosurgical Institute - Federal North-West Medical Research Centre, St. Petersburg

Objective: The problem of early surgical treatment of children with congenital malformations is one of the most important in children's neurosurgery. Early diagnosis and minimally invasive surgical correction of children with craniosynostosis allow delivering the best cosmetic results, promoting patients adaptation and avoiding development of complications typical for this pathology, including cranio-cerebral disproportion.

Methods: We carried out a comparative study of surgical treatment outcomes among children from 3 months to 5 years old with craniosynostosis. The selection covers the period from 2012-2014 and includes 57 patients. Children younger than 5 months constituted the 1st group, children older than 5 months – the 2nd group. The first group includes 13 patients; the second one includes 44 patients. 11 patients from the first group went through endoscopic cranioplasty which allowed shortening the duration of surgical interference, minimizing intraoperative blood loss and thus improving the postoperative course substantially. The patients from the second group went through open reparative surgery using titan and biodegradable implants.

Results: Significant reduction of rehabilitation period was observed in the group of patients operated through endoscopic assistance, which is connected primarily with minimal invasiveness of the method and minimization of intraoperative blood loss. It should be also mentioned that the children from the 1st group had better cosmetic results of treatment, which fact is connected with original absence of severe skull deformation appearing with older children.

Conclusion: Endoscopic cranioplasty belongs to minimally invasive methods of treating patients with craniosynostosis, allows improving the postoperative course and preventing development of a wide range of complications. Early diagnosis allows extending the range of application of the given method in treating patients with craniosynostosis. Studying biomechanical properties of the cranio-cerebral system, CSF circulation and cerebrovascular responsiveness allows prognosticating the possibility of development of cranio-cerebral disproportion after minimally invasive endoscopic cranioplasty.

Keywords: craniosynostosis, endoscopic, endoscopic cranioplasty, minimally invasive methods, craniostenosis

P-152

Craniofacial

Long term intracranial pressure monitoring during gradual distraction osteogenesis for craniosynostosis: a case of apert syndrome

Jun Kurihara¹; Yuu Kageyama¹; Atsuo Yoshino².

¹Children's Medical Center, Department of Neurosurgery, Saitama;

²Nihon University School of Medicine, Department of Neurosurgery, Tokyo

Objective: Cranial expansion using distraction osteogenesis is one of the useful surgical methods for craniosynostosis. But the change of intracranial pressure (ICP) during gradual cranial expansion is unknown and the goal of cranial expansion also is not setting. We report on a case of long term ICP monitoring using telemetric system during gradual cranial expansion for craniosynostosis.

Methods: A 1-year-old girl suffered from Apert syndrome, diagnosed by craniofacial deformity and polysyndactyly. Computed tomography

showed bilateral coronal suture synostosis. She underwent fronto-orbital advancement by distraction method and implantation of telemetric ICP sensor. ICP measurement was carried on during surgery and after surgery.

Results: At the results, Intraoperative ICP was decreased in association with cranial expansion, but ICP after surgery gradually increased over 20mmHg. During gradual distraction osteogenesis ICP did not change associated with cranial volume over 20% and after cranial expansion ICP was gradually decrease over a month.

Conclusion: This is the first report of long term ICP monitoring during gradual cranial expansion for craniosynostosis. Intraoperative ICP reflected the cranial expansion but ICP right after surgery did not reflect the effect of cranial expansion. During distraction osteogenesis ICP was not necessarily correspond to cranial volume. Long term ICP monitoring is necessary for the evaluation of cranial expansion by gradual distraction osteogenesis and telemetric ICP monitoring is the best way of long term ICP monitoring.

Keywords: craniosynostosis, distraction osteogenesis, long term ICP monitoring, telemetric system

P-153

Craniofacial

Scaphocephaly treatment by limited invasive scalp approach.

Vanessa Hernandez-Hernandez¹; Mario García-Conde¹; Lucia González-Bautista¹; Liberto Brage-Martin¹; Pablo Febles-García¹; Luis Perez-Orribo¹.

¹Hospital Universitario De Canarias, La Laguna

Objective: A retrospective study of cases treated on our service was performed to compare two surgical techniques for the treatment of scaphocephaly.

Methods: Data of 24 patients treated in the last 8 years on our service with scaphocephaly have been included for the study. Group 1 is composed of 12 patients who underwent bitemporoparietal remodelling with osteofixation and Group 2 patients treated using minimally invasive approach. The following variables were compared: age of intervention, duration of the surgical procedure, transfusion requirements, days of hospitalisation, presence of complications, time tracking, preoperative and postoperative cranial index, reoperation, use of cranial orthosis after surgery and satisfaction with the operation.

Results: No significant differences between the two groups were found in terms of satisfaction with the operation. However the surgical time and hospital stay was clearly lower in Group 2.

Conclusion: Although both techniques are valid for the treatment of scaphocephaly, treatment by a less invasive technique offers good results with a significant decrease in operative time and postoperative stay which in a greater number of cases may correspond to a lower incidence of adverse events.

Keywords: Scaphocephaly, surgical treatment, craniosynostosis, learning curve, minimally invasive techniques

P-154

Other

The anxiety and depression levels related with somatization with parents who have children with spina bifida

Ibrahim Alatas¹; Gizem Tarhan²; Kerem Ozel¹; Ezgi Erdogan¹; Nursu Kara³; Huseyin Canaz¹.

¹Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Department of Psychology, Istanbul Bilim University, Istanbul; ³Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul

Objective: The purpose of this study was to determine depression, anxiety, the level of hopelessness and the relative of these levels with somatization, obsession, anxiety, interpersonal sensitivity, psychotics, paranoid, phobia, fury, sleeping and eating disorders of parents.

Methods: At Şişli Florarance Nightingale Hospital department of Spina Bifida between 10 of March and 3 of April 2014 Beck Depression Inventory, Beck Anxiety Inventory, Beck Hopeless Inventory and SCL-90-R test was applied to who appealed for 82 cases (60 women -22 men).

Results: On 82 cases who applied, On Beck Depression Inventory 21 people of 82 cases was measured with medium level, 4 people was measured with heavy level of depressive symptoms. Incidences who has measured with medium and heavy levels, related to SCL-90-R test was appeared high obsession symptoms on these 21 people. On 82 cases who applied for Beck Hopelessness Inventory, on 16 cases was appeared medium level hopelessness level. These cases which related with SCL-90-R test was appeared more. On 82 cases who applied, was appeared on 26 people were medium level, 1 person was heavy level anxiety. The cases who has medium and heavy level anxiety related with SCL-90 test was appeared high interpersonal sensitivity on 25 people.

Conclusion: In our country, these symptoms shows the parents who has children with Spina Bifida, have high possibility of obsessive compulsive diagnostic who show depression symptom. The parents with medium level hopelessness diagnostic show frequently depressive symptoms. The symptoms affirms hopelessness, depression, and anxiety related with positive way of obsessive depression and interpersonal sensitivity factors. These 3 factors should be recognized by the health care personnels and the people who show these symptoms should get help by psychologic or psychiatric.

Keywords: anxiety, depression, hopelessness, parents, spina bifida

P-155

Other

Giant intracranial arachnoid cyst: a neglected case despite parents' concerns

Dimitris Kombogiorgas¹.

¹Metropolitan Hospital, Athens

Objective: The current main surgical options in management of intracranial arachnoid cysts are microsurgical excision, neuroendoscopic fenestration and insertion of CSF shunt. However, there is no consensus on the therapeutic approach of choice in symptomatic patients. A neglected symptomatic patient with massive intracranial arachnoid cyst is presented aiming to emphasize the need for individualization of management of those patients and development of a network to ensure proper follow up of those patients.

Methods: A 4 months old baby boy was seen with one month history of bilateral converging quint. He was diagnosed with a left temporal cyst, prenatally. MRI brain at 2 days of age showed left temporal arachnoid cyst Type I, agenesis of corpus callosum and an intrahemispheric cyst. Parents did not follow the advice of treating pediatrician who consulted them to ask for neurosurgical consultation. Family lives in Middle East and is Arabic in origin. Based on their family and social culture, patient's parents followed the grandfathers view (father's side), who strongly advised them not to ask for neurosurgical consultation regarding his grandsons health problem. Urgent follow up MRI brain scan was done which showed massive enlargement of arachnoid cyst with associated non-communicating hydrocephalus. Patient was operated on for insertion of Cysto-Peritoneal shunt. He had an uncomplicated operation and uneventful postoperative course.

Results: Patient had been improved neurologically and the size of his arachnoid cyst was massively reduced, postoperatively.

Conclusion: Despite the modern tendency of endoscopic surgical treatment of arachnoid cysts, individualization of surgical management is recommended. Also, development of a network to follow up the progress

of cases by use of information technology should be considered as the above case was neglected due to cultural issues.

Keywords: Arachnoid cyst, Hydrocephalus, Shunt, Squint

P-156

Other

Using free osirix software in the surgical planning of endoscopic third ventricle approaches in pediatric patients

İhsan Doğan¹; Murat Zaimoğlu¹; Gökmen Kahiloğulları¹; Ağahan Ünlü¹.

¹Ankara University School of Medicine İbni Sina Hospital Department of Neurosurgery, Ankara

Objective: Determination of appropriate localization and size of burr hole for endoscopic approaches to the third ventricle is the initial and most important step for the purpose of reaching the pathology, gaining a wide surgical freedom zone and minimalizing the cerebral retraction. The objective of this study is to find a practical, effective and inexpensive method alternative to neuronavigation systems in planning the surgical strategy for endoscopic approaches to the third ventricle pathologies.

Methods: Radiological examination and measurements were studied on 5 pediatric patients. All patients were undergone thin section contrast enhanced magnetic rezonans imaging (MRI). MRI compatible markers were placed on midline and midpupillary line over frontal bone. All data of these patients were processed by using free Osirix Software and cranial 3-D view of these patients were reconstructed. The pathology or region to be reached was detected. An imaginary line starting from the pathology and passing through the foramen Monro was extended to the calvarium. This region was chosen as the localization of the burr hole. The exact localization of this region on patients were measured according to the previously placed markers.

Results: In all cases, previously determined burr hole localization using free Osirix Software allowed entrance to the third ventricle via foramen Monro and access to the surgical destination.

Conclusion: A 3-D reconstruction of the third ventricle, third ventricular pathologies and surrounding brain structures can be reconstructed with free OsiriX software. With this method, endoscopic entrance region and endoscopic trajectory line passing through the foramen Monro can be defined and preoperatively examined easily.

Keywords: endoscopy, third ventricle approaches, surgical strategy, free Osirix Software

P-157

Other

Determination of depression, and anxiety levels of the parents of the children who were diagnosed as spina bifida

Ibrahim Alatas¹; Burcu Turk Lal²; Kerem Ozel¹; Mehmet Tokmak³; Ezgi Erdogan¹; Nursu Kara⁴; Huseyin Canaz¹.

¹Spina Bifida Research Center, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul; ²Department of Psychology, Kanuni Sultan Suleyman Hospital, Istanbul; ³Department of Neurosurgery, Istanbul Medipol University, Istanbul; ⁴Department of Neonatology, Florence Nightingale Hospital, Istanbul Bilim University, Istanbul

Objective: Neural tube defect (NTD) which is the most frequently encountered congenital disorders of the central nervous system is a condition which emerges as a result of failed closure of brain, spinal cord, and spinal column during the first weeks of the embryonal life. Failure of closure along the spinal extension of the neural tube, affecting especially lumbosacral region is called "spina bifida". Spina bifida means "clef" or

“open” spinal cord in Latin. As reported in the literature, parents having mentally or physically disabled children are most frequently under stress with higher anxiety level when compared with those without. Psychological disorders are more frequently seen in mothers of disabled children relative to those without. This study was planned to determine sociodemographic characteristics of the children with spina bifida, anxiety, and depression levels of the parents.

Methods: This survey study was performed on parents of the children diagnosed as spina bifida in Neurosurgery Department of our hospital. Data were collected by Interview Form prepared by the researchers, Becks Anxiety Inventory (BAI), and Becks Depression Inventory (BDI

Results: Fifty-two parents participated in our study. Mean total Becks Depression Inventory (BDI) scores of mothers, and fathers were 15.4 ± 2.82 , and 11.1 ± 2.12 points, respectively. Mean total Becks Anxiety Inventory (BAI) scores of mothers, and fathers who volunteered to participate in the study were 17.2 ± 12.02 , and 10.2 ± 5.65 points, respectively.

Conclusion: In this study anxiety, and depression levels of parents with children diagnosed as spina bifida. In our study depression, and anxiety levels of mothers were higher than those of the fathers. Counselling centers which will inform parents about what will be done about the child's disease after establishment of the diagnosis of spina bifida.

Keywords: anxiety, depression, spina bifida