IPSO 2021 Virtual Conference: July 19-21, 2021

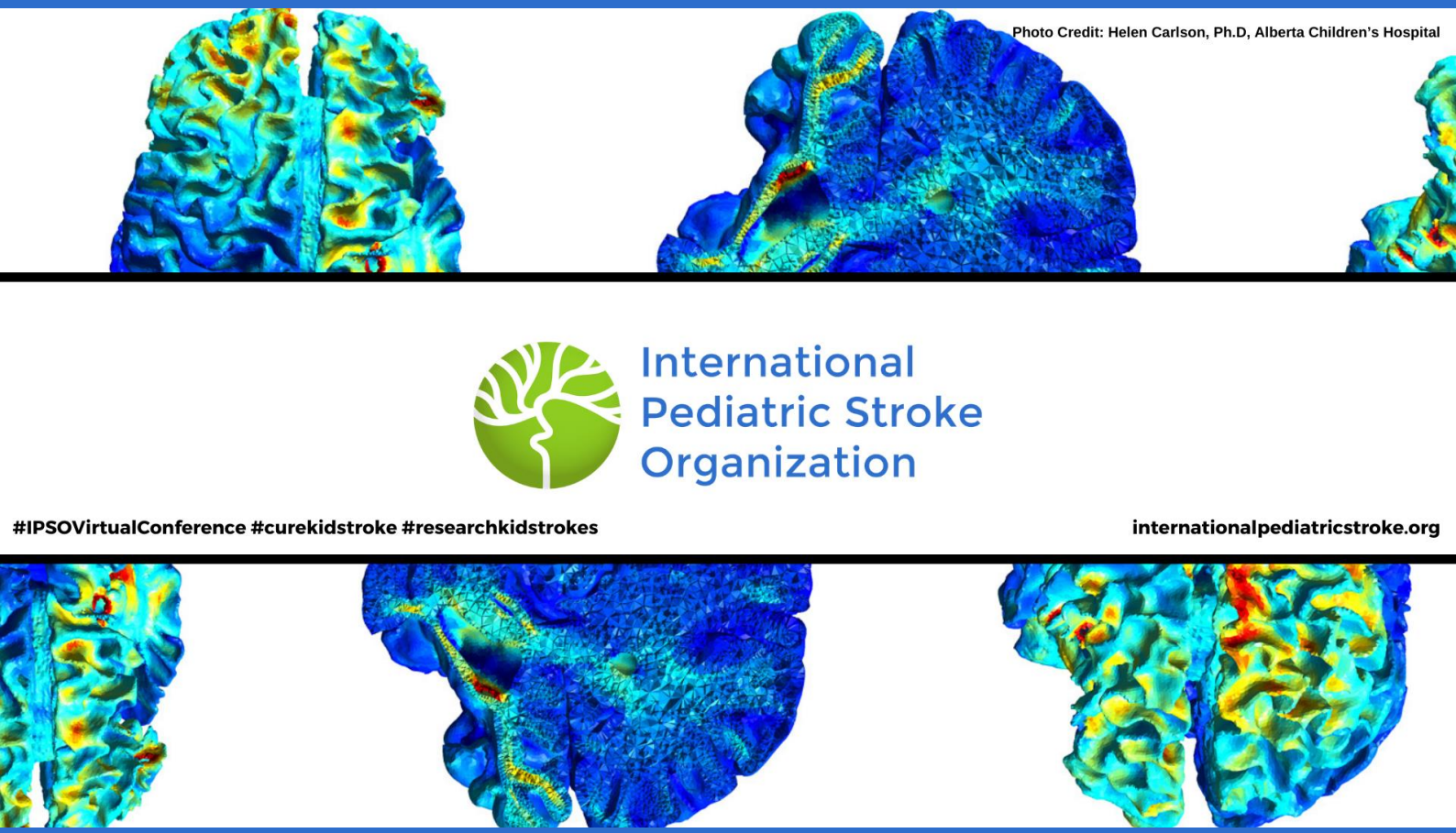

Photo Credit: Helen Carlson, Ph.D, Alberta Children's Hospital

International Pediatric Stroke Organization

#IPSOVirtualConference #curekidstroke #researchkidstrokes

internationalpediatricstroke.org

Poster Abstract Proceedings

IPSO Virtual Conference Abstract Proceedings

All posters are available on-demand throughout the virtual conference in both the Poster Section of the Underline platform, as well as on the social networking tool, Gather.Town.

Click on any of the categories below to review the poster abstracts.

Contents

A - Acute treatment

P1-A: Thrombolysis in children and adolescents with ischemic stroke. Experience of the Primary Pediatric Stroke Center in Moscow

Inna Shchederkina[1], Pavel Svirin[1], Ljubov Larina[1], Nina Maslova[1], Valery Kharkin[1], Matvey Livshitz[1], Elena Petrayakina[2]

[1]Morozov children city clinical hospital, [2]Morozov children city clinical hospital

Reperfusion therapy (thrombolysis) is widely used in adults, but the extrapolation of the method to children is impossible because of the age-related features of the hemostasis system and metabolism in children. Aim. Evaluate the efficacy and safety of thrombolysis in children with arterial ischemic stroke (AIS). Materials: From March 2018 to November 2019, ten children with stroke underwent thrombolysis. Criteria recommended by Thrombolysis in Pediatric Stroke were taken as criteria for selecting patients for thrombolysis. Ten children from 3.5 to 16 years old (average age -10.3g) were included. Results: Patients with stroke had: thrombophilia-2, autoimmune hemolytic anemia-1, polyangiitis-1, Epstein-Barr virus infection-1, transient nocturnal hemoglobinuria with aplastic anemia-1, arteropahty-2, Kavasaki disease -2. The severity of stroke was from 9 to 24 points by PedNIHSS (average -15.8b). The stroke was confirmed by MRI-angiography. The time of start of thrombolysis was from 4 to 10 hours (average 6.3 hours). Thrombolysis in all cases was carried out by the recombinant human tissue plasminogen activator - alteplase. None of our patients had any complications (hemorrhage). MRI control of the efficacy of thrombolysis was carried out after 6-8 hours. In 5 cases, the effect was pronounced: the change on the PedNIHSS scale: from 9 to 2b, from 22 to 8, from 9 to 4, from 10 to 2, from 15 to 6; in five - ineffective: 1- with the infection, two with progressive arteriopathy, one - with polyangiitis, one- transient nocturnal hemoglobinuria with aplastic anemia. A child with polyangiitis and AIS in the basilar artery was thromboextracted after ineffective thrombolysis. Conclusion: In our center, we considered as potential candidates for thrombolysis of all children with AIS who arrived at the clinic no later than 12 hours from the onset of the disease. In our observation, the effectiveness of thrombolysis depended on the etiology of AIS.

[PRINT ONLY] P2-A: Is Cupid at risk of Bowhunter's Syndrome? Vertebral artery compression with neck rotation in children with posterior circulation symptoms

Michael Dowling[1], Bruno Braga[1], Amy Hogge[1], Wilmot Bonnet[1], Rafael de Oliveiro Sillero[1], Nancy Rollins[1]

[1]University of Texas Southwestern Medical Center

We evaluated 22 children presenting with "posterior circulation" symptoms with provocative digital catheter angiography with neck rotation and observed "dynamic arterial compression" (DAC) involving the V3 segment where C1 rotates on C2 in 10 children. 17 (14M) presented with stroke, 12 had vertebral artery (VA) dissection, with 3 having "presumed dissection" as stroke mechanism. DAC of the VA was found in 10 (9M), all with observed VA dissection. These 10 were treated with surgical fusion at C1/C2. None had recurrent stroke post op. The 7 without DAC were not treated surgically. AIS etiologies: dissection 2, presumed dissection 3, basilar artery occlusion 1, cardioembolic 1. We presume that in 4 of these 7, supraphysiologic rotation from trauma led to VA dissection but they were not surgically treated as there was no DAC under "normal" degrees of rotation. In 3/7, one VA was either occluded or was unable to be catheterized so could not be assessed. 5 other children (4M) presented with posterior circulation symptoms but did not have AIS.

Back to the top

Provocative angiography and did not demonstrate DAC. Etiologies: post-concussion syndrome, opiate ingestion, MELAS, vertebral pseudoaneurysm, and psychogenic. Our observations, particularly of bilateral VA dissection at V3 in 3 children, with high incidence of DAC at this same region suggests that DAC may lead to VA dissection. Our observation of children without DAC demonstrates that such compression is not normal physiology. DAC may be an age or stage of growth-related RF for VA dissection. Other factors, such as arterial wall composition (collagen vascular genetic variants) and hypercoagulable states may also contribute. Our protocol for suspected posterior circulation stroke includes c-spine immobilization, evaluation by provocative digital catheter angiography, with strong consideration for surgical intervention in those demonstrating DAC.

[PRINT ONLY] P3-A: Electroencephalographic monitoring in pediatric arterial ischemic stroke - When, why, who

Michaela Waak[1], Andrea Andrade[2], Stephen Malone[1], Adriane Sinclair[1]

[1]Queensland Children's Hospital, [2]Schulich School of Medicine & Dentistry

Introduction Seizures are common in paediatric AIS, but there is limited knowledge regarding frequency and predictors of electrographic seizures. Without EEG monitoring these will go undetected resulting in lost opportunity for neuroprotection. Our aim was to determine the frequency and risk factors for clinical and electrographic seizures in paediatric AIS. Methods A prospective single centre study recruited patients 4 w - 18 yrs from July 2018 to January 2020. EEG monitoring commenced <10 days post stroke. The EEGs were reviewed by 2 independent neurologists/epileptologists. Acute assessments included PedNIHSS and modified ASPECTS, follow-up the PSOM. Results 16 patients were recruited with 17 strokes. Two patients were excluded due to being immediately post epilepsy surgery. Median age was 3.7 years (4 w -12 yrs). Seizures occurred in 6 (38%) patients. Of children <2yrs of age, 4/5 (80%) had seizures. Seizures occurred up to day 5. Median seizure burden was 5% (IQR 5-25%). The maximum number of seizures in 24 hrs ranged from 13-72 (median 30). Seizures were controlled within 48 hours in all children and patients required a median of 3 (1-5) AEDs. Vascular territory and PICU admission rate did not differ significantly between patient groups. On multivariate analysis, age was the only statistically significant variable with younger age predictive of clinical and electrographic seizures. EEG monitoring guided management through: Diagnosis of events, assessing seizure burden, guiding treatment with AEDs, guiding neuroimaging. Conclusions A high proportion of children with AIS had clinical and/or electrographic seizures, especially children <2yrs of age. Frequency and burden of seizures was high. Young age was the only statistically significant predictor of clinical and electrographic seizures. Further studies and larger cohorts are required to further study risk factors, assess impact on patient important outcomes and provide recommendations for intervention.

P4-A: Very-early craniectomy in an infant with a severe acute ischemic stroke

Rachel Reed[1], John Myseros[1], Dana Harrar[2], Yussef Kousa[1], Paola Pergami[3]

[1]CNMC, [2]Children's National Medical Center, [3]Georgetown University Medical Center/CNMC

Abstract We are presenting a very young pediatric case with extensive right MCA and PCA territories acute ischemic stroke (AIS) and severe midline shift who had an excellent outcome following an early decompressive craniectomy. Introduction Infarctions of the MCA and PCA usually lead to significant morbidity and mortality in patients1. Early surgical intervention with decompressive craniectomy in adults significantly improves survival rate and long-term functional outcome prognoses2,3. Despite this positive evidence,

craniectomies are rarely used in pediatric patients with AIS, particularly at a young age. We describe a 7-month-old female with large AIS with a positive outcome following an early decompressive craniectomy. Case report A 7 month-old previously healthy, fully vaccinated, twin girl presented acutely with seizures and was found to have an extensive frontal/parietal infarct. At the time of onset the patient was febrile, on amoxicillin for presumed otitis media. She presented with prolonged focal seizures requiring multiple medications leading to respiratory failure and intubation. An urgent MRI demonstrated extensive areas of infarction in the right MCA and PCA territories. MRA demonstrated severe stenosis of the right IC which was considered congenital in nature. No dissection or clots were identified. On hospital day 2, due to rapid development of brain edema with midline shift, the patient underwent a decompressive hemicraniectomy. After surgery she had an uneventful course and was successfully extubated and discharged home on ASA. The bone flap was successfully replaced 3 months later. At the most recent follow-up visit, the patient, now 11 months old, is walking with minimal leg involvement and decreased hand use. Discussion Evidence about craniectomy in pediatric AIS is still unclear. Some authors suggested that early craniectomy can result in excellent outcomes, particularly for MCA strokes, while others reported severe neurological deficits and/or high rates of surgical complications, particularly in neonates, following this procedure1,2,4,5,6. Although more data is necessary, decompressive craniectomy should be considered, when appropriate, for pediatric patients with malignant AIS; very young age is not necessarily an exclusion criterion for this procedure.

P5-A: Paediatric Arteriopathy Steroid Aspirin (PASTA) Trial

Maja Steinlin[1]

[1]University of Bern

Background: Focal cerebral arteriopathy (FCA) accounts for stroke in about one third of previously healthy children, 2/3 suffering long-term problems. FCA of inflammatory type (FCA-i) shows an inflammatory vessel wall infiltration, which encourages steroid treatment, despite lack of evidence. This trial is to show positive effect on stenosis, outcome and recurrence by treatment with steroids and standard of care (SC) compared to SC alone. Methods: Multicenter, parallel group, two-arm, randomized-controlled, clinical trial with blinded outcome comparing high dose course of steroids plus SC with SC alone in children with unilateral arteriopathy and acute ischemic stroke. Inclusion criteria: age 6 mo to 18 y; acute neurological focal deficits; confirmed ischemic infarction and unilateral cerebral arterial stenosis/irregularity on imaging. Exclusion criteria: Progressive CNS angiitis; syndromal diagnoses (as Moyamoya); genetic vasculopathies; sickle cell disease; arterial dissection. Participants will be randomized within 48 h (maximum 96 h after stroke onset) to SC alone (controls) or SC plus 3-days of 30mg/kg methylprednisolone IV followed by 4-week tapering of oral prednisolone (experimental group). Follow up will be at 1, 3, 6 and 12 with MRI/MRA at months 1, (3) and 6. Primary outcome will be change in FCA Severity Score (FCASS) from baseline to 1 mo, compared between 2 study arms. Secondary outcomes: neurological deficits over time (PSOM/RRQ), Vineland Adaptive Behavior Scale at 6 and 12 mo, neurocognition at 12 mo, recurrence-free survival, change in FCASS and residual stenosis at 6 mo. Safety outcomes: infections requiring hospitalization, serious adverse events, evolution to progressive arteriopathy. Sample size will be 70 children. Relevance of the study: The study will provide high-level evidence for most appropriate treatment for children with AIS due to FCA-i. Alignment of interventions and outcome with the FOCAS study in North America will allow pooled analyses.

B - Endovascular treatment

P6-B: Feasibility, safety, and outcome of endovascular recanalization in childhood stroke: The Save ChildS Study

Peter Sporns[1]

[1]University Hospital of Münster

IMPORTANCE: Randomized clinical trials have shown the efficacy of thrombectomy of large intracranial vessel occlusions in adults; however, any association of therapy with clinical outcomes in children is unknown. OBJECTIVE: To evaluate the use of endovascular recanalization in pediatric patients with arterial ischemic stroke. DESIGN: This retrospective, multicenter cohort study, conducted from January 1, 2000, to December 31, 2018, analyzed the databases from 27 stroke centers in Europe and the United States. Included were all pediatric patients (<18 years) with ischemic stroke who underwent endovascular recanalization. RESULTS: Seventy-three children from 27 participating stroke centers were included. Median age was 11.3 years (interquartile range [IQR], 7.0-15.0); 37 patients (51%) were boys, and 36 patients (49%) were girls. Sixty-three children (86%) received treatment for anterior circulation occlusion and 10 patients (14%) received treatment for posterior circulation occlusion; 16 patients (22%) received concomitant intravenous thrombolysis. Neurologic outcome improved from a median PedNIHSS score of 14.0 (IQR, 9.2-20.0) at admission to 4.0 (IQR, 2.0-7.3) at day 7. Median mRS score was 1.0 (IQR, 0-1.6) at 6 months and 1.0 (IQR, 0-1.0) at 24 months. One patient (1%) developed a postinterventional bleeding complication and 4 patients (5%) developed transient peri-interventional vasospasm. The proportion of symptomatic intracerebral hemorrhage events in the HERMES meta-analysis of trials with adults was 2.79 (95% CI, 0.42-6.66) and in Save ChildS was 1.37 (95% CI, 0.03-7.40). CONCLUSIONS: The results of this study suggest that the safety profile of thrombectomy in childhood stroke does not differ from the safety profile in randomized clinical trials for adults; most of the treated children had favorable neurologic outcomes. This study may support clinicians' practice of off-label thrombectomy in childhood stroke in the absence of high-level evidence.

P8-B: Venous sinus stenting for idiopathic intracranial hypertension

Matthew Amans[1]

[1]UCSF

Idiopathic intracranial hypertension (IIH) is a pathologic elevation of the intracranial pressure (ICP) that can lead to blindness, cognitive decline, debilitating headaches, and distressing pulsatile tinnitus (PT). IIH most commonly affects overweight women of childbearing age, and has an incidence of 20 in 100,000 in this patient cohort, and is becoming even more prevalent with the increasing obesity epidemic. We are also noticing a rise in childhood IIH as the childhood obesity epidemic similarly increases. MRI/MRV imaging demonstrates focal stenoses of the transverse dural venous sinus (TS) in up to 93% of patients with IIH versus 6.8% of asymptomatic control subjects. TS stenoses are critical to the pathophysiology of IIH. While the inciting event of IIH is unknown, it is apparent that TS stenoses are a critical part of a positive feedback loop, and disrupting TS stenoses with venous sinus stenting (VSS) can break the vicious cycle and cure IIH. The aim of this presentation is to review the pathophysiology underlying IIH and the role VSS can play in treating IIH patients. The indications of VSS, success rates, complications rates and revision surgery rates are reviewed in the context of other available surgical therapies including shunting, optic nerve sheath fenestration, and bariatric surgery.

P9-B: Recanalization treatments in pediatric arterial ischemic stroke: preliminary results of the French KidClot study

Manoëlle Kossorotoff[1], Christian Denier[2], Kim Tran Dong[1], Celine Bellesme[2], Beatrice Husson[2], Olivier Naggara[1]

[1]Univ Hospital Necker-Enfants malades APHP, [2]Univ Hospital Bicetre APHP

Introduction: In adult stroke, the use of hyperacute recanalization treatments, i.e. IV thrombolysis (IV r-tPA) and endovascular thrombectomy (EVT), has dramatically increased over the recent years because of their good benefit-risk balance in selected indications. Recommendations in children are difficult to formulate as the occurrence of stroke is much lower in this population and randomized control trials nearly impossible to set up. It is thus important to collect and analyze clinical experience through regional or national databases. Methods: KidClot is an ongoing nationwide retrospective study collecting clinical, imaging, medical management, and outcomes data from children with arterial ischemic stroke treated by IV r-tPA and/or EVT in mainland France from January 1rst, 2015 to May, 31rst, 2018. To ensure maximal exhaustivity, KidClot is a multisource study: data are obtained from pediatric neurologists, vascular neurologists, and interventional radiologists in every academic hospital, and from the national multidisciplinary pediatric stroke conference. The main objective is to analyze feasibility and safety of such treatments. Secondary objectives are the following: to determine factors affecting treatment availability in pediatric stroke protocols, and prognostic factors for good outcome after recanalization treatments. Preliminary results: A first pre-inclusion screening retrieved at least 70 pediatric cases treated during the inclusion period in 30 academic centers. On January 31rst, 2020, 31 patients have been included from 15 centers, with a median age at stroke occurrence of 8.6 years old and a slight predominance of boys (sex ratio 1.2). 16 patients received IV r-tPA, 2 received intra-arterial r-tPA, and 14 had EVT. Combined treatment was given in 4 patients. Discussion: Further results will be presented in the Conference, as inclusions are still going on. Complete data should help answering safety questions and delineating prognostic subgroups.

C - Imaging

[PRINT ONLY] P10-C: Longitudinal imaging of GABA, glutamate/glutamine, and glutathione following pediatric stroke

Muhammad Saleh, Nicolaas A. Puts[1], Ryan Felling[2]

[1]Kings College, [2]Johns Hopkins Hospital

Introduction Pediatric stroke is a rare but important cause of neurologic disability during childhood and throughout life. We have a limited understanding of how age influences mechanisms and extent of recovery after stroke. Excitatory/inhibitory balance and reduction of oxidative stress may be important for synaptic plasticity and recovery after stroke. Hypothesis We hypothesize that we could use Hadamard-encoded multi-metabolite edited magnetic resonance spectroscopy (MRS) to simultaneously measure changes in the concentration of gamma-amino butyric acid (GABA) and glutamine/glutamate (Glx) and glutathione (GSH) during recovery after stroke. Methods We conducted edited MRS using HERMES during the acute stroke admission and at 3 months follow up. An isotropic 27mL voxel was placed overlapping the infarct and peri-infarct cortex. The same size reference voxel was placed in the corresponding contralesional hemisphere to serve as a control. The HERMES acquisition generated GABA-edited (with the 3 ppm GABA+ and Glx

(Glu+glutamine signals) and GSH-edited (with the 2.95 ppm GSH signal) spectra. Data were analyzed in Gannet. The GABA+ (GABA with macromolecules) and Glx signals were modeled between 2.79 to 4.1 ppm using a singlet Gaussian for GABA+, a doublet Gaussian for Glx, and a non-linear baseline. The GSH-edited spectrum was modeled between 2.25 and 3.5 ppm using a nonlinear baseline, a Gaussian for the GSH signal at 2.95 ppm through weighted nonlinear regression and four Gaussians to model the co-edited aspartyl signals. The 2.0 ppm Cr signal from OFF GABA/GSH step (no editing pulse applied) was modeled as a reference signal for calculating GABA+/Cr, Glx/Cr, and GSH/Cr. Results In the infarcted area, GABA+/Cr (and Glx/Cr) declined 0.0953 (and 0.0797) during the acute stroke period to 0.0725 (and 0.0675) at 3 months. Similarly GSH/Cr in the infarcted area declined from 0.0391 to 0.0256. Conclusions HERMES provides a powerful noninvasive tool to understand stroke recovery physiology. Simultaneous longitudinal modeling of GABA, Glu, and GSH may provide in vivo insight into changes in the excitatory/inhibitory balance and oxidative stress that mediate neural plasticity.

[PRINT ONL] P11-C: Imaging biomarker of recurrent stroke in children: Early radiographic recurrence or extension predicts late recurrence

Wilmot Bonnet[1], Joan Reisch[1], Tricia Plumb[1], Michael Dowling[1]

[1]University of Texas Southwestern Medical Center

Introduction: Pediatric AIS has a high recurrence rate of 10-25%. We observed a high rate of early recurrence or extension of stroke on repeat MRI performed within 2 weeks of index stroke and hypothesized that early radiographic extension of stroke or recurrent stroke would predict late clinical recurrence. Methods: IRB approved retrospective chart review of children ages 1 week to 18 years with AIS in our institutional pediatric stroke database who had repeat MRI within two weeks of initial acute presentation. Results: 67 patients had early repeat MRI. 24 (35.9%) had early extension or new stroke on repeat MRI. Of these, 6 (25%) had late clinical recurrence while only 1/47 (2.3%) who did not have early recurrence/extension went on to have late recurrence. 15/23 (66%) of patients with early change on MRI had early recurrence, 8/23 had extension. Neither was more predictive of late recurrence. 4/67 (5.9 %) had hemorrhagic conversion (one was symptomatic -headache). Radiographic extension or new stroke on early repeat MRI was significantly associated with late recurrence (p=0.0036). Using logistic regression, early extension/recurrence was found to be predictive of late recurrence, (OR 13.99, 95%CI 1.57-124). Conclusion: Children with AIS have a high rate of ongoing early, clinically silent, ischemic injury despite antithrombotic therapy, which is associated with late clinical recurrence. This early imaging finding may serve as a biomarker that might prove useful as an endpoint in clinical trials comparing early antithrombotic therapies or to identify patients at higher risk of subsequent recurrent overt stroke who might benefit from intensified therapy. Given the high rate of early and late recurrence and low hemorrhage rates from current antithrombotic therapies, more aggressive therapy with dual antithrombotic agents should be investigated.

D - Moyamoya

P12-D: Subclinical pediatric moyamoya disease preceded by non-specific brain-MRI signal changes, anxiety, fatigue and recurrent migraines

Amaar Marefi[1], Ahmed Sahly[1], Bradley Osterman[1]

[1]Montreal Children's Hospital, McGill University Health Center

Introduction: Moyamoya disease (MMD) in children commonly presents as cerebral ischemia (80%) rather than hemorrhage (20%) as well as headaches, seizures, involuntary movements and progressive decline in intellectual ability. We present a pediatric patient with MMD presenting with worsening migraines, anxiety, fatigue and nonspecific subcortical signal abnormalities on brain imaging. Case: A 7-year-old French-Canadian girl presented with a 2-year history of monthly, bifrontal headaches, associated with nausea, vomiting, photo- and phonophobia. An initial brain-MRI at 5 years of age showed non-specific subtle bifrontal subcortical millimetric white matter hyperintensities on T2/FLAIR, a finding often seen in pediatric migraine patients. She was treated with daily Vitamin B2 and NSAIDs as-needed, with a good initial response. At 7 years of age, her headaches occurred 3-4 times/month. She also complained of increasing anxiety, fatigue as well as 2-3 episodes of paroxysmal confusion, right-handed clumsiness, and expressive aphasia lasting minutes, triggered by stress or crying. The patient nevertheless continued to have a normal development and exam. A repeat MRI at 6-years-of-age showed mild progression of the bifrontal subcortical white matter hyperintensities. An MRI at 7-years-of-age showed further progression of the same signal changes, with prominent tortuous signal voids within the deep grey matter, mostly the basal ganglia and non-visibility of the M1 and M2 segments of the MCA bilaterally, confirming the diagnosis of MMD. Discussion: Approximately 90% of pediatric patients with MMD are diagnosed solely after a cerebral ischemic infarct. Early symptoms and neuroimaging can often be subtle or misleading. The worsening of migraine-like headaches, along with anxiety, fatigue, and paroxysmal self-resolving episodes of aphasia should prompt re-investigation. Conventional angiography is the gold-standard imaging modalities for the diagnosis of MMD. Conventional nonquantitative brain-MRI, while limited, may provide useful clues to obtain an early diagnosis in patients, such as in our case. It allows visualization of late sequelae of previous strokes, including silent strokes, appearing as T2/FLAIR hyperintensities, brain atrophy and ventriculomegaly. Abnormal vasculature can appear as multiple tortuous flow voids in deep structures on T1 and T2 sequences. The "ivy sign", due to leptomeningeal engorgement, can be seen on postcontrast-MRI or FLAIR-MRI.

P13-D: Discovery of novel genes predisposing to moyamoya disease

Amelie Pinard[1], Dongchuan Guo[2], Alana Cecchi[2], Stephanie Guey, Chaker Aloui, Michael Bamshad, Deborah Nickerson, Elisabeth Tournier-Lasserve, Dianna Milewicz[1]

[1]University of Texas Health Science Center at Houston, [2]The University of Texas Health Science C

Introduction: Moyamoya disease (MMD) is a cerebrovascular disease characterized by progressive bilateral occlusion of the distal internal carotid arteries and its branches, leading to a compensatory network of collateral vessels. Several genes harbor variants that increase the risk for MMD, but the genetic cause remain unidentified for many MMD patients of European descent. Hypothesis: MMD is associated with significant genetic heterogeneity and there are more genes yet to be identified. Methods: Exome sequencing was performed on DNA from 151 individuals (84 families with one or more affected members). Bioinformatics analysis filtered for damaging rare variants (CADD score >20 and minor allele frequency <0.0001 in gnomAD controls). Collaborative replication cohorts were interrogated for additional variants in novel genes for MMD. Results: Two large MMD families had the same rare variant in ANO1 segregating with disease and were subsequently determined to be distantly related; 5 additional rare variants were identified in MMD cases. ANO1 is a calcium-activated chloride channel and functional analyses determined that variants increase channel gating and calcium sensitivity, which is predicted to decrease smooth muscle cell contraction and increase proliferation. Trio analysis (affected child with two unaffected parents) revealed pathogenic de novo variants in CHD4, CNOT3, SETD5, three genes that encode proteins involved in chromatin remodeling and

Back to the top

previously identified to have de novo rare variants in children with developmental disorders. Taken together with our previously reported gene for MMD-like cerebrovascular occlusive disease (YY1AP1), these genes implicate disrupted chromatin remodeling as a molecular pathway predisposing to MMD. Finally, we defined a novel syndrome characterized by severe and progressive MMD before three years old, associated with occlusion of other arteries (abdominal aorta, renal, iliac, and femoral), that results from de novo RNF213 pathogenic variants in two discrete locations of the gene, distinct from location of the Asian RNF213 founder variant. Conclusions: Exome sequencing of MMD trios and families identified novel genes predisposing to MMD in unsolved patients and further confirmed genetic heterogeneity for the disorder. These data emphasize the need for exome data on MMD cases and family members and collaboration between research programs to identify all the genes predisposing to MMD.

P14-D: Characterization of vascular disease in Acta2 SMC-R179C/+ mice

Anita Kaw[1], Callie Kwartler[1], Abhijnan Chattopadhyay[1], Amelie Pinard[1], Dianna Milewicz[1]

[1]University of Texas Health Science Center at Houston

Introduction: Moyamoya disease (MMD) is a malignant cause of childhood stroke characterized by occlusion of the distal internal carotid arteries and branches by neointimal cells staining positive for smooth muscle cell (SMC) markers. ACTA2 encodes smooth muscle α-actin (α-SMA). A subset of ACTA2 mutations that predispose to thoracic aortic aneurysms and dissections including mutations altering arginine 179 are also associated with bilateral MMD-like occlusions. This study aims to elucidate the molecular pathogenesis of this disease to potentially inform the etiology of large vessel disease in stroke patients in the general population. Hypothesis We hypothesize that Acta2SMC-R179C/+ mice have disrupted differentiation of SMCs in cerebrovascular arteries, and thus the cells are primed to proliferate and migrate excessively, leading to MMD-like disease. Methods: We generated patient-derived ACTA2 R179C and control induced pluripotent stem cell (iPSC) lines, and the Acta2SMC-R179C/+ mouse model, which expresses the Acta2 R179C mutation in SMCs via the SM-22-Cre-Lox system. Phenotypic differences between mutant and WT brains are characterized using micro CT, non-invasive cuff blood pressure measurements, and echocardiography. SMC phenotype is evaluated by protein expression of contractile genes, BrdU ELISA proliferation assay, Transwell migration assay, and immunofluorescence. Results/Anticipated Results: Patient-derived ACTA2 R179C iPSC-derived SMCs from patients show de-differentiation, continued expression of stem cell markers, and increased proliferation and migration. rt-PCR and sequencing of genomic DNA performed in aortic tissue and explanted SMCs confirms the presence of the mutation in the mutant mice and absence in mice with only the floxed allele (WT). Two-dimensional gel confirms presence of mutant α-SMA. Acta2SMC-R179C/+ mice display straightened and narrowed cerebral arteries at 8 weeks of age and hypotension but no aortic disease between 2 and 12 months of age. Explanted SMCs from Acta2SMC-R179C/+ mice exhibit decreased levels of differentiation markers and increased proliferation and migration. Acta2SMC-R179C/+ SMCs also show less robust and disrupted α-SMA filaments. Conclusion: Acta2SMC-R179C/+ mice display MMD-like cerebrovascular disease and Acta2SMC-R179C/+ SMCs exhibit stem cell-like characteristics including de-differentiation, increased proliferation and migration.

P15-D: Arterial-spin labeling imaging for assessment of neoangiogenesis after pial synangiosis in pediatric moyamoya patients

ShihShan Lang[1], Rebecca Ichord[1], Meghan Connors[1], Hongyan Liu[1], Lusha Cao[1], Heather Griffis[1], Todd Kilbaugh[1], Neda Roman-Sedora[2], Hussein Nasser[2], Mougnyan Cox[2], Phillip Storm[1], Jimmy Huh[1]

[1]Children's Hospital of Philadelphia, [2]University of Pennsylvania

Introduction: Digital subtraction angiography (DSA) is commonly performed post pial synangiosis to establish the degree of neovascularization. However, angiography is invasive, and the risk of ionizing radiation is a concern in children. Our study aimed to identify whether arterial spin labelling (ASL) can predict post-operative angiogram grading. In addition, we aimed to identify whether patients who underwent ASL imaging without DSA had similar postoperative outcomes when compared to patients who received ASL imaging and post-operative DSA. Methods: The medical records of patients undergoing pial synangiosis at a single institution were reviewed during a 10-year period. Results: Twenty-two patients were identified. Twenty-one hemispheres of 13 patients with "ASL only" and 14 hemispheres of 9 patients with "ASL + DSA" were analyzed. Both groups had similar rates of MRI evidence of acute or chronic stroke preoperatively; 61.9% in the ASL only group and 64.3% in the ASL + DSA group. All symptoms improved in frequency after intervention except categories of hemorrhage and non-chronic headaches. We found a significant correlation between revascularization observed on the DSA and the ASL graded by 3 neuroradiologists. The ASL grades in patients with a postoperative Matsushima grade indicating robust neovascularization were significantly higher than the ASL grades of patients with a poor postoperative Matsushima grade. Conclusions: Non-invasive ASL perfusion imaging had an association with post-operative DSA neoangiogenesis following pial synangiosis surgery. There were no significant stroke-risk differences between ASL and ASL + DSA cohorts. Both cohorts show significant improvement in pre-operative symptoms after surgery. Providers may consider obtaining a postoperative ASL MRI imaging in lieu of a catheter angiogram.

P16-D: Stroke in children with moyamoya and down syndrome; 3 cases and review of the literature

Michelle Gratton[1], Bisan Shweiki[2], Sandrine de Ribaupierre[3], Michael Jurkiewicz[3], Andrea Andrade[3]

[1]Children's Hospital, London Health Sciences Centre, [2]University of Toronto, [3]Schulich School of Medicine & Dentistry

Background and Aims: There are few retrospective studies describing clinical-radiological features and outcomes of children with Down Syndrome (DS) and Moyamoya (MM). Our aims are to present three cases of DS/MM from our institution, perform a review of the literature to compile similar case reports, determine if there is a clinical-radiological syndrome specific to DS/MM, and determine if this association is related to different outcomes. Methods: Our clinical database was reviewed between 2017-2019 for patients with DS/MM and cross-referenced with our radiology PACS database. We then reviewed the clinical data, imaging, treatment, and outcomes of previously 38-reported cases in English world literature with DS/MM from 1977 to 2018 using PubMed, Ovid, Medline, and Embase. Results: Case 1: Six year-old girl with focal status epilepticus, left MCA infarction and bilateral MM. Case 2: Three year-old boy with right hemiparesis, left MCA infarction and bilateral MM. Case 3: Thirteen year-old boy with right leg monoparesis, MRI-A: left ACA infarction and left ICA-MCA incipient MM. Case 1 & 2: managed with multiple burr holes (MBH). Case 1: surgery was performed 5 days after initial stroke due to rapid progression. Case 3: treated conservatively. On follow-up, all children had minimal deficit and continued on ASA. We reviewed 38 DS/MM cases found in the literature: 30 children presented with hemiparesis and 12 were treated surgically. Conclusions: Our findings are suggestive that DS/MM presentation and severity is variable with relatively good outcomes post-surgery.

To our knowledge, Case 1 is the first reported child with DS/MM treated with urgent surgical MBH revascularization.

P17-D: Distinct clinical and radiographic presentation and evolution in childhood moyamoya syndrome

Matsnga Leyila Kaseka[1], Mahmoud Slim[1], Prakash Muthusami[1], Peter Dirks[1], Elizabeth Pulcine[1], Mahendranath Moharir[1], Gabrielle deVeber[1], Daune MacGregor[1], Nomazulu Dlamini[1]

[1]The Hospital for Sick Children

Introduction: Moyamoya is a progressive steno-occlusive arteriopathy affecting the distal internal carotid artery and its branches. Associated to an underlying condition such as neurofibromatosis type 1 (MMS-NF1) or sickle cell disease (MMS-SCD), it is called moyamoya syndrome (MMS) while an isolated arteriopathy is referred to as moyamoya disease (MMD). It remains unknown whether the associated condition impacts its evolution. Objective: We hypothesize that MMD presents and evolves differently compared to MMS. Methods: We performed a single-center chart review of patients with moyamoya followed between 2003 and 2019. Clinical and radiographic features at diagnosis were obtained. Subsequent stroke or neurosurgery clinic visits were reviewed to identify ischemic event occurrence. Evidence of disease progression or new infarction was sought on follow up MRI studies. Group comparisons between children with MMD and MMS and between MMD and MMS subgroups (MMS-NF1 and MMS-SCD) were conducted using Pearson's Chi-squared, Fisher's Exact, Mann-Whitney U, or t test, as appropriate. Survival analysis by cerebral hemisphere using Cox regression were conducted to assess ischemic event occurrence, disease progression and silent infarction at follow-up. A false discovery rate (FDR) p-value adjustment for multiple comparisons with a q-value< 0.05 was applied. Results: One hundred and eleven patients were identified (MMD=49; MMS-NF=23; MMS-SCD=21). MMD presented more with TIA than MMS (22.45% vs MMS 4.84%, q=.02), while MMS-NF1 presented less with a stroke compared to the other groups (MMD 46.94% vs MMS-NF1 8.7% vs MMS-SCD 42.86%; q=0.0032). MMS-NF1 had more frequently unilateral disease (MMD 26.53% vs MMS-NF1 78.26% vs MMS-SCD 33.33%; q=.0003) and less infarcts (MMD 67.35% vs MMS-NF 26.09% vs MMS-SCD 80.95%; q=.0008) at diagnosis, while MMS-SCD were less likely to have an ivy sign (MMD67.35% vs MMS-NF1 52.17% vs MMS-SCD 9.52%, q=.0003). At follow-up, MMD patients were more likely to suffer another ischemic event compared to MMS patients (OR 1.92, p=.019), these events were mainly TIA (OR 2.5, p=.004). However, compared to the other groups, MMS-NF1 were less likely to have another TIA (OR 0.43, p=.045). Conclusion: MMS has varying phenotypes. MMS-NF1 tends to be unilateral with a lower burden of infarction at presentation, while an ivy sign is not a common feature of MM-SCD. Compared to MMS-SCD and MMD, MMS-NF1 is less likely to have further ischemic events.

E - Outcomes

P18-E: Neuropsychological function across time in a child with NOTCH3 mutation and CADASIL

Carla Morton[1], Jo Tilley[1], Fernando Acosta[1], Marcela Torres[1]

[1]Cook Children's Medical Center

The symptoms of Cerebral Autosomal Disorder Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (CADASIL) do not typically manifest until midlife, but it has been diagnosed in a few children. Although she appeared healthy as a young child, BV had a heterozygous NOTCH3 gene mutation, and she experienced previously undetected strokes in the left frontal area. At 4 years 9 months, BV exhibited sudden onset aphasia and left hemiparesis, and she was found to have had acute ischemic strokes involving the right anterior cerebral artery (ACA) and left middle cerebral artery (MCA) territories. Extensive vascular disease involving the internal carotid arteries, right ACA, and left MCA was also observed. Within days, BV experienced new strokes in the right centrum semiovale, left interior frontal white matter, and right parietal lobe. She underwent encephaloduroarteriosynangiosis (EDAS) procedures in the left brain hemisphere at 5 years 0 months and in the right brain hemisphere at 5 years 3 months. BV experienced brain edema following the second procedure, and she was diagnosed with an acute ischemic stroke in the right MCA area affecting the majority of the temporal, parietal, and occipital lobes approximately 1 week following that procedure. BV had neuropsychological evaluations at 5 years 1 month, 5 years 10 months, and 7 years 1 month. At the time of her first evaluation, BV's visuospatial processing was average, but her verbal abilities were generally low average. She also exhibited severe inattention and impulsivity. After revascularization surgery and additional strokes, BV's language processing was stable, but her visuospatial processing had declined and was generally low average to mildly impaired. BV's cognitive functioning had remained generally stable with some areas of modest development at the time of her third neuropsychological evaluation at 7 years of age.

P19-E: The relationship between selected risk factors and the pathogenesis and consequences of arterial ischemic stroke in children

Beata Sarecka-Hujar[1], Ilona Kopyta[1]

[1]Medical University of Silesia in Katowice

Background: The incidence of arterial ischemic stroke (AIS) is relatively low in children in contrast to adults. In pediatric patients however, risk factors for AIS are still not well known and understood. In many pediatric patients more than one risk factor can be identified however, according to various data, about half of all stroke cases in developmental age are cryptogenic. Genetic risk factors may be primarily involved in its aetiology, as indicated by the age of patients. Aim: Our previous studies aimed to analyse the relations between selected polymorphisms in the MTHFR, E-selectin and prothrombin genes, some non-genetic factors (e.g. sex, age, the presence of FCA arteriopathy, the presence of heart disease or the type of stroke) and the pathogenesis and neurological consequences of AIS in children. Methods: Analyses of the relationships between genetic polymorphisms and AIS were carried out in children with AIS, their parents as well as in healthy children. The lipid and coagulation profiles were analyzed retrospectively. The data were analysed statistically with STATISTICA 12.0. Results: We observed that the carrier state of the T allele of the 677C>T polymorphism, in contrast to 1298A>C polymorphism, in the MTHFR gene may be considered a risk factor for AIS in children, especially in boys. The 98G>T polymorphism in the selectin-E gene is not associated with the risk of AIS in children, although there is a tendency for more frequent occurrence of T allele and T allele carriers in healthy children than in children with AIS. The carrier state of the A allele of the 20210G>A polymorphism in the FII gene is a risk factor for ischemic stroke, both in children and young adults. In addition, the occurrence and number of post-stroke neurological consequences depend on the type of AIS. Children with at least one deficit after stroke were younger at the time of AIS than children without any deficits. Arteriopathy is associated with the type of AIS as well as the presence of post-stroke consequences. The lipid profile and coagulation parameters do not, however, differ between stroke children with FCA from stroke children in whom FCA does not occur. Conclusions: The occurrence of AIS in children may result from the presence of

many risk factors, the analysis of which may allow an in-depth understanding of the pathogenesis of the disease in developmental age.

P20-E: Post-stroke epilepsy in Polish pediatric patients- own research results and up-to-date literature review

Ilona Kopyta[1], Beata Sarecka-Hujar[1]

[1]Medical University of Silesia in Katowice

Background: Arterial ischemic stroke is a set of rapidly developing clinical symptoms characterized by a sudden, focal or generalized brain disorder resulting from the dysfunction of brain circulation, lasting 24 hours or longer or leading to death. One of the most severe post-stroke outcome in children are seizures and epilepsy however the exact cut off and the definition of late post-stroke seizures (LPSS) and epilepsy (PSE) should be distinguished and defined. In newborns, which are mostly excluded from the group of pediatric stroke and considered separately, and young infants seizures are one of the most typical AIS presentation. Incidence of seizures in AIS children is several times higher when compared to adult AIS patients; according to some research results seizures may be observed in nearly 40% of AIS children; in about 20% of the post-stroke epilepsy is diagnosed later on. Authors present the own results on risk factors and course of post-stroke epilepsy in children as well as up-to-date literature review on this field. Methods: In own research on post-stroke epilepsy in children the examined group of patients consisted of 78 AIS children at the age of 1month to 18 year at the age of stroke; 13 of them presented early seizures within 7 days after stroke onset, and 10 out of 78 met the criteria of post-stroke epilepsy diagnosis. The data obtained in my research were analyzed statistically using the following software: STATISTICA 12.0.Then the up-to-date literature data were evaluated on predictors, clinical course and treatment of PSE. Results: In our research post-stroke epilepsy occurred in 10 children; participants affected by LPSS were younger than children without seizures in follow-up. The frequencies of TACI(Total anterior circulation infarct and focal cerebral arteriopathy of childhood(FCA) were significantly higher in the late seizure subgroup than in the group without seizures (71% vs 26%, p=0.014, OR 7.17, and 100% vs 51%, p=0.015 respectively). Multivariable Cox analysis showed that age at time of stroke (p=0.027), FCA(p=0.010), Conclusion: In own data the predictors of PSE were: age at the stroke onset, number of ischemic foci as well as FCA. The latest up-to-date data show that the first EEG asymmetry can independently predict unprovoked seizures or epilepsy in the year following anterior circulation ischemic stroke. Epilepsy in AIS pediatric patients has a great impact on everyday activity and school achievements.

P21-E: The impact of age at pediatric stroke on long-term cognitive outcome

Stephanie Abgottspon[1], Thaqi Qendresa, Leonie Steiner, Nedelina Slavova[1], Sebastian Grunt[1], Maja Steinlin[1], Regula Everts[1]

[1]University Hospital Bern, Switzerland

Introduction and Aim: Pediatric arterial ischemic stroke is a rare event but is accompanied by an increased risk for cognitive sequelae. The association between age at stroke and long-term cognitive outcome remains however unclear. Here, we investigated the impact of age at pediatric arterial ischemic stroke on long-term cognitive outcome in order to identify patients particularly at risk for the development of long-term cognitive sequelae.

Methods: This cross-sectional study included patients in the chronic phase of stroke (> 2 years after stroke) previously diagnosed with neonatal or childhood arterial ischemic stroke and a control group. Several cognitive domains, including intelligence, executive functions (working memory, inhibition, and cognitive flexibility), processing speed, memory, letter fluency, and visual-motor skills were assessed with neuropsychological tests. Cognitive long-term outcome was compared across patients after neonatal stroke (0 – 28 days), early childhood stroke (29 days – 5 years) and late childhood stroke (6 – 16 years).

Results: 52 patients after neonatal or childhood arterial ischemic stroke (mean age: 14.82 years, SD = 4.53) and 49 healthy controls (mean age: 14.28 years, SD = 5.36) met the inclusion criteria. Cognitive outcome was significantly worse in the pediatric stroke group compared to the control group. A non-linear effect of age at stroke (irrespective of lesion size and lesion location) was found for working memory, cognitive flexibility, processing speed,

and verbal learning with early childhood stroke (29 days – 5 years) showing significantly worse cognitive outcome compared to neonatal or late childhood stroke.

Conclusion: Age at stroke is an important factor for post-stroke recovery and modulates long-term cognitive outcome irrespective of lesion size and lesion location. Children after early childhood stroke are at particular risk for alterations of long-term cognitive functions.

P22-E: Back 2 Class: A review and survey of school inclusion after acquired brain injury

Bethany Johnson Kerner[1], Nathan Evanson[2], Jennifer Frey[3], Patricia Dillhoff[2], Jonathan Kurz[4], J Michael Taylor[2]

[1]University of California, San Francisco, [2]Cincinnati Children's Hospital, University of Cincinnati, [3]The George Washington University, [4]Ann and Robert H. Lurie Children's Hospital, Northwestern University

Introduction: After sustaining brain injury, children have unique challenges to their activities of daily living compared to their healthy peers. These barriers are especially notable in the classroom, where a combination of physical, emotional, social, and cognitive skills is required to allow optimal performance. Medical staff and educational professionals share a common mandate to facilitate a child's best future. When injury interferes with a child's function, the achievement potential is threatened. Methods:This presentation is a review of return to school initiatives in place for children in the USA and medical provider utilization of these systems. We will present comparative models of educational inclusion services, identifying strengths and opportunities for expansion. Preliminary results from a survey of pediatric brain injury care providers will detail utilization of these services. Results: We describe a continuum of services divided into 4 discrete paradigms: brain injury-specific classroom, transitional school model, day rehabilitation program, and school outreach/community-based services. Programs vary dramatically in cost, payer, service coverage, and geographic availability. There is minimal data on efficacy and little agreement on endpoints to demonstrate successful intervention. A wide variation in access to and utilization of services is reflected in healthcare provider experience. Conclusions: There is a clear need to develop robust, available, and successful programs to return children to their education after brain injury.

[PRINT ONLY] P23-E: Neurocognitive outcome after childhood stroke: a pilot study

Kerttu Kivisikk[1], Pilvi Ilves[2], Mairi Männamaa[3], Eve Õiglane- Šlik[1], Inga Talvik[4], Tiina Talvik[5], Rael Laugesaar[1]

[1]Tartu University, [2]Radiology Clinic of Tartu University Hospital, [3]Tartu University Hospital, [4]Tallinn Children's Hospital, [5]Department of Pediatrics, University of Tartu

Introduction: Children with paediatric stroke may develop long-term disabilities including motor deficit, epilepsy, cognitive, behavioural and social disorders. Our aim was to describe the neurocognitive outcome of children following stroke, and to find out how the follow-up of the patients has been organized in Estonia. Hypothesis: After stroke children are followed-up regularly and they have a relatively good outcome. Methods: Patients were derived from Estonian Pediatric Stroke Database from January 1995 to October 2018. Only children treated or consulted in Tartu University Hospital were included. Results: In October 2018 there were a total of 82 patients with childhood stroke in the database, among them 46 (56%) were treated in our hospital. After excluding dead patients (4 patients, 9 %), the final group consisted of 42 patients. Twenty-two patients (52%) had visited (pediatric)neurologist within last 2.5 years at the median age of 11 years 8 months (IQR 7y10mo to 15y8mo). Moderate-severe sensorimotor defcit was described in 17 (41%) children. Epilepsy, speech impairment, or cognitive deficit was described in 15 (35%) patients. Data about diferent subgroups are shown in Table 1 (see appendix). We also found that information about the use of rehabilitation services is often missed in the medical records. Conclusions: Only half of the patients with pediatric stroke had been regularly followed-up by a (child)neurologist. About half of the pediatric stroke patients have a significant motor deficit, epilepsy, speech and/or cognition problems. The results may be biased as patients with more significant neurologic deficit tend to visit doctors more often. The existing follow-up system is not well organized and needs to be updated so that patient follow-up would be more complete and thorough.

P24-E: Dysphagia incidence following arterial ischemic stroke or cerebral sinovenous thrombosis in children

Victoria Sherman[1], Mahendra Moharir[1], Ishvinder Bhathal[1], Kevin Thorpe[2], Rosemary Martino[2]

[1]The Hospital for Sick Children, [2]University of Toronto

Objective Stroke occurs across the lifespan and is becoming increasingly recognized in childhood. Dysphagia is common after stroke in adults; however, few studies have explored dysphagia in children post stroke. This study assessed incidence and predictors of dysphagia in children following stroke. Methods The Canadian Pediatric Ischemic Stroke Registry for a tertiary hospital was used to identify consecutive patients aged 1mo-18yrs, admitted with arterial ischemic stroke (AIS) or cerebral sinovenous thrombosis (CSVT) over five years (Jan/13-Nov/18), and included demographic and stroke details. Two independent raters reviewed medical charts to capture in-hospital feeding and swallowing impairment. Bivariate analysis and simple logistic regression derived incidence and odds ratios with respect to select variables such as type, severity and laterality of stroke. Results There were 106 children: 73(68.9%) AIS, 35(33.0%) CSVT; 57(53.8%) male; and, median age 5.5 years(1.3-11.2). There were 26(25.0%) with severe neurological deficit at stroke presentation, 12(11.3%) at discharge. Hemiparesis was present in 52(49.1%), 26(50.0%) left sided. Across the entire sample, the incidence of feeding impairment was 20(18.9%), and swallowing impairment was 38(35.8%). Of those with feeding and swallowing impairment, the majority were AIS (17, 85.0% and 33, 86.8%) and had a right sided or bilateral neurological deficit, 12(60.0%) and 27(71.1%) respectively. Of those with swallowing impairment, 11(28.9%) had severe neurological deficit. There were 43(40.6%) children who had either feeding and/or swallowing impairments. Conclusion These findings suggest that dysphagia is a consequence of stroke in children, with up to 40.6% affected. Due to study design limitations, these estimates are likely conservative leaving the true incidence unknown. Prospective work is needed to better understand the incidence and characteristics of dysphagia in this unique population.

P25-E: Care and outcomes of pediatric patients with in-hospital stroke versus community onset stroke

Elissa Ortolani[1], Dana Cummings[1]

[1]UPMC

In-hospital stroke refer to stroke occurring in a patient hospitalized for another condition. Unlike stroke occurring in the community, there is a potential for rapid diagnosis and treatment given access to medical resources. Existing research in adult population however suggest higher rates of morbidity and mortality in patients with in-hospital strokes compared to patients with community onset stroke. Worse outcomes may be multifactorial, including risk factors inherent to the inpatient status, and relative contraindication for acute therapy. Pediatrics studies have begun to evaluate the implementation of in-hospital pediatric stroke alert activation, however little is known regarding the care and outcomes of the subgroup of pediatric patients. Our goal is to evaluate the stroke care and outcomes of pediatric patients with in-hospital and community onset stroke over a 10 year period at the Children's Hospital of Pittsburgh through an IRB approved, retrospective chart review. Inclusion criteria includes children age ≤18 years with diagnosis of acute stroke by clinical and radiographic criteria. We have screened a total of 785 Patients, resulting in 188 community strokes and 78 in-hospital strokes.Our primary outcome is the time from stroke symptoms to the first neuroimaging. Secondary outcomes include presenting symptoms, stroke risk factors, neuroimaging preformed, treatment, length of stay, , and discharge destination. We concluded, In-House pediatric strokes occur in younger children with the largest risk factor of congenital cardiac disease vs. Community Strokes occurring in older children with the most common risk factor being vasculopathy and infection. Despite quick access to resources, there was no statically significant difference in time to obtain imaging between groups. Reflecting the delays in recognizing pediatric stroke in both settings. In-Hospital pediatric strokes, similar to the adult population have longer hospital admissions, higher percentage of rehab needs on discharge and higher mortality. Our study is limited in it is a retrospective historical analysis and more research is needed in this area.

P26-E: Predictors of neurological outcome of arterial ischemic stroke in children

Tatia Aprasidze[1], Nana Tatishvili[1], Teona Shatirishvili[1], Giorgi Lomidze[2]

[1]M. Iashvili Children Central Hospital, David Tvildiani Medical University, [2]Institute of Neurology and Neuropsychology

Background:Stroke is an important cause of mortality and morbidity in children. Aim:The aim of the study was to evaluate long-term neurological outcome in children with arterial ischemic stroke (AIS) and explore predictive factors that affect poor outcome. Patients:56 patients aged between one month and 17 years who were treated at M.Iashvili Children Central Hospital, Tbilisi, Georgia, with an onset of stroke from 2007 to 2017 were included. Methods:To explore predictive factors of outcome, the following data were collected: demographic characteristics, risk factors, the presenting signs, radiological features, and presence of stroke recurrence. Neurological status at discharge and long-term neurological outcome at least 1 year after stroke was evaluated according to Pediatric Stroke Outcome Measure (PSOM) subscale. Results: The reported outcome after childhood stroke was variable with long-term neurological deficits in one-third of patients (30.4%). The neurological outcome was worse in males, in patients with multiple stroke episodes, and in those

with infractions involving a combination of cortical and subcortical areas. Conclusion:Pediatric AIS carries the risk of long-term morbidity, and neuroimaging has a predictive influence on outcome.

F - Perinatal stroke

P27-F: A service evaluation of the University Hospital Southampton neurodevelopmental protocol to follow up and identify attention outcomes in children with perinatal stroke

Erin Bannister[1], Fenella Kirkham[1]

[1]University of Southampton

Background: Perinatal stroke occurs secondary to a focal disruption of blood flow in the brain occurring between 20 weeks' gestation and the 28th day of life. Current literature has associated perinatal stroke with adverse attention outcomes. Despite this, when parents of children with perinatal stroke were surveyed, very few recalled conversations with their clinician regarding their child's attentional outcome. There is therefore a need to evaluate the University Hospital Southampton (UHS) neurodevelopmental protocol to follow up attention outcomes in perinatal stroke. Aims: Firstly to evaluate adherence to the UHS neurodevelopmental protocol. Secondly, to evaluate the use of neuroimaging data and clinical data regarding neonatal seizures and post-natal epilepsy to identify children at risk of attention difficulties. Methodology: We conducted a service evaluation to determine whether the attention problem score from the Child Behavior Checklist (CBCL), was available from the parent reported questionnaire administered at the 2-year follow-up appointment. Neuroimaging data were analysed to identify cortical, white matter, basal ganglia, and thalamic involvement and to quantify infarct size. Presence of neonatal seizures and post-natal epilepsy were extracted from clinical notes. Chi-square was used to compare proportions. Results: CBCL data were only available for 35/63 patients. Reasons for missing CBCL data included patients not attending follow up, not returning questionnaires and inadequate data handling meaning data were not stored correctly. 13/35 children had attention scores in the borderline or clinical range. Basal ganglia and thalamic involvement (p=0.015), neonatal seizures (p=0.028) and post-natal epilepsy (p=0.042) were associated with borderline or clinically significant attention scores. Post-natal epilepsy predicted the highest relative risk of adverse attention outcomes. Conclusion: These clinical predictors could be used to identify children at high risk for attention difficulties to encourage parental adherence to the neurodevelopmental protocol, ensuring a complete clinical dataset and providing information to enable appropriate therapy and education.

[PRINT ONLY] P30-F: Upper extremity motor timing and coordination in infants with perinatal stroke: a longitudinal kinematic assessment

Jill Heathcock[1], Julia Mazzarella[1]

[1]The Ohio State University

Background and Objectives: Perinatal stroke (PS) is bleeding or interrupted blood flow to the brain around the time of birth. PS is a leading cause of hemiplegic cerebral palsy (CP), which typically causes the greatest impairment to the upper extremity (UE). In this study, we measured UE kinematics longitudinally in infants with PS, PS with later diagnosis of CP (PS + CP) and TD. Study Participants & Setting: N=21 full-term infants participated in this longitudinal study; 11 infants with PS (4 PS + CP) and 10 infants without PS. Infants were

evaluated in a laboratory setting. Materials/Methods: Infants were evaluated monthly from 2-6 months of age. Infants were place in an infant chair and reflective markers were placed on the infants' hands. A toy was presented at midline shoulder height to stimulate reaching. Movements were recorded at 120Hz for 3 x 30-second trials using a 10-camera Vicon motion capture system. Common spatial-temporal variables important for timing and coordination were calculated: movement length (mm) and speed (mm/s), number of reaches, and straightness ratio (total hand path length/straight line distance). A linear regression analysis was used to determine correlation of age and diagnosis with these variables. Results: Results of the linear regression showed that age and diagnosis were both significantly correlated with many of the variables tested. For movement length, there was an average increase of 3.5mm per additional month of age (95%CI [.07, 7.0], p=.046), and an average difference of -7.2mm per level of diagnosis with TD > PS > PS + CP where the poorest performance was observed in infants with PS + CP, 95%CI[-12.5, -2.0], p=.008. Other variables followed a similar pattern. Peak velocity of a movement had an average difference of 90.3mm/s per LOD (95%CI[-178, -2.7], p=.044). Number of reaches had an average increase of 1.23 per additional month of age, (95%CI[.46, 2.0],p=.002), and an average difference of -2.0 with LOD, (95%CI[-3.2, -.84], p=.001). Last, straightness ratio had an average difference of .19 with LOD, (95%CI[.072, .31], p=.002). Conclusions /Significance: These results demonstrate significant differences in spatial-temporal variables of reaching between infants with TD, PS and PS + CP, as well as with age. These preliminary findings suggest the possibility of using kinematic assessment to identify infants with PS who will later develop hemiparetic CP.

P31-F: Gadd45b modulates neurogenesis following perinatal hypoxic-ischemic brain injury

Thomas Robinson[1], Ji Young Park[1], Kevin Silva[1], Ryan Felling[2]

[1]Johns Hopkins University, [2]Johns Hopkins Hospital

Introduction Neurogenesis occurs after perinatal brain injury as a result of gene expression changes. DNA methylation and demethylation is one mechanism by which gene expression is regulated. Recent work has elucidated a role for Gadd45 proteins in mediating gene demethylation. Hypothesis We hypothesize that Gadd45 is responsible for active demethylation after perinatal hypoxia/ischemia and promotes neurogenesis as part of the regenerative response to injury. Methods We induced perinatal HI in 10 day old C57BL/6 pups by ligation of the right common carotid artery followed by exposure to hypoxia (8% O2/92% N2) for 45 minutes. The pups were sacrificed at 4 or 24 hours for molecular biology or immunohistochemistry. We used quantitative RT-PCR to study gene expression of Gadd45 family and TET family proteins. We evaluated targeted gene methylation status with methylation specific PCR and in situ DNA modifications using immonhistochemistry. Neurogenesis was quantified by 5-ethinyl-2'-deoxyuridine (EdU) incorporation in wild type and Gadd45b knockout mice. Results Gadd45 family mRNA is rapidly upregulated by 14-fold in the subventricular zone (SVZ) by 4 hours following perinatal HI, and remains upregulated at 24 hours. Small transient increases in expression were seen for Gadd45a and Gadd45g at 4 hours. There is no significant change in the expression of TET mRNA. Preliminary suggest specific demethylation of the BDNF promoter by 24 hours. Immunohistochemistry revealed increased expression of 5-hydroxymethylcytosine specifically within the SVZ progenitor cells, and neurogenesis was reduced in Gadd45-/- mice. Conclusion Perinatal HI induces upregulation of Gadd45b, which may contribute to demethylation of growth promoting genes in the neurogenic niche, driving the neurogenic response to perinatal hypoxic-ischemic brain injury.

P32-F: A new kind of bleed: Subpial hemorrhages in the neonatal intensive care unit

Ania Dabrowski[1], Melisa Carrasco McCaul[2], Andre RF Barreto[2], Aylin Tekes[2], Lisa Sun[2]

[1]physician, [2]Johns Hopkins School of Medicine

Background and Aims: Subpial hemorrhages (sPH) were first described in infants in 1972, but they remain underrecognized on neuroimaging and the clinical implications of sPH are poorly understood. The aim of this study is to describe the etiologies, risk factors, clinical presentations, and imaging findings of sPH in neonates. Methods: Institutional Neonatal Intensive Care Unit database was queried for presence of sPH. All infants recorded to have intracranial hemorrhage on brain MRI were reviewed by a pediatric neuroradiologist. Through retrospective chart review, data on pregnancy/birth history, comorbidities, and clinical concerns that led to imaging were collected. Results: Twenty-six infants with sPH were identified: 13 (50%) were preterm, 11 (42%) had low birthweight. Comorbidities included hypoxic-ischemic encephalopathy (n=1, 42%), birth trauma (n=6, 23%), cardiopulmonary disease (n=6, 23%), coagulopathy (n=6, 23%), ventricular shunt (n=3, 12%), severe congenital brain anomalies (n=1, 4%), and chromosomal anomaly (n=1, 4%). MRI acquisition was at a median of 5 days of life (range 1 to 41 days). sPH frequently accompanied intraparenchymal (n=17, 65%), subdural (n=16, 62%), intraventricular (n=13, 50%), and subarachnoid hemorrhage (n=8, 31%). In addition, 7 children (27%) had ischemic stroke. All children with sPH had either another form of hemorrhage, ischemic stroke, or both. In addition, 9 patients (34%) had follow-up MRIs. Development of adjacent encephalomalacia was consistently demonstrated in children with large subpial hemorrhages. Sixteen infants with sPH (62%) had clinical and/or electrographic seizures. Conclusions: sPH are associated with multiple comorbid medical conditions, other types of intracranial hemorrhage, and ischemic strokes. Further studies to evaluate long-term clinical implications of subpial hemorrhages are needed.

P33-F: Cognitive outcomes in preschoolers with presumed vs. acute perinatal stroke

Christine Mrakotsky[1], Julie Swanson[2], Michael Rivkin[2]

[1]Boston Children's Hospital/Harvard Medical School, [2]Boston Children's Hospital

Introduction: In children, perinatal stroke is the most common type. Long-term morbidities can be high, and include motor, cognitive and behavioral domains. While some studies point to poorer global, motor and adaptive outcomes in presumed perinatal compared to acute perinatal stroke, few have studied specific cognitive functions in a developmentally homogenous sample. We aimed to assess cognitive outcomes in a homogenous sample regarding age, time since injury, and development. We predicted that delay in diagnosis and hence intervention may have an impact on long-term cognitive outcome. Hypothesis: Preschoolers with presumed perinatal stroke demonstrate poorer cognitive function than those with acute neonatal stroke. Methods: Children age 4;0-5;11 years (n=63, Acute Perinatal=39, Presumed Perinatal=24) underwent comprehensive neuropsychological assessment as part of standard care. Cognitive function was assessed with age-appropriate standardized tests of general cognitive ability, language, visual-spatial, memory, and motor functions. Parents completed ratings of executive and behavioral function. Data from initial evaluations at a single time point were analyzed retrospectively. Cognitive and behavioral function of the overall sample were compared to population means, as well as between acute and presumed perinatal subgroups. Results: Compared to population means, the perinatal group demonstrated poorer processing speed, auditory memory, visual-spatial and visual-motor skills (pFDR<.01-.05), and increased problems with executive functions (pFDR<.01; flexibility, emerging metacognition) and adaptive skills reported by parents/teachers. Further, the presumed perinatal stroke group showed reduced verbal ability (knowledge, reasoning, naming), and processing speed than the acute perinatal group (ps<.01-.05). Stroke location and maternal education were significant contributors. Conclusions: Perinatal stroke causes diminished cognitive and behavioral function in preschoolers and kindergartners. We found cognitive differences (verbal, processing speed) in children with presumed versus acute perinatal stroke. Findings will be further explored for predictors of

outcome in an ongoing study; they underscore the importance of early diagnosis and intervention to improve long-term cognitive and psychosocial function.

P34-F: Whole-exome sequencing analysis in antenatal and perinatal intracerebral hemorrhage

Moran Hausman-Kedem[1], Gad Malinger[2], Shelly Shiran[2], Jonathan Roth[1], Shlomi Constantini[1], Aviva Fattal-Valevski[1], Shay Ben Shachar[1]

[1]Dana-Dewk Children's Hospital, Tel Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel Av, [2]Tel Aviv Sourasky Medical Center, Sackler Faculty of Medicine, Tel Aviv University

Objective Perinatal intracranial hemorrhage (pICH) is a rare event that occurs during the fetal/neonatal period with potentially devastating neurological outcome. However, the etiology of pICH is frequently hard to depict. We investigated the role of rare genetic variations in unexplained cases of pICH. Methods We performed whole-exome sequencing (WES) in fetuses and term neonates with otherwise unexplained pICH and their parents. Variant causality was determined according to the American College of Medical Genetics and Genomics (ACMG) criteria, consistency between suggested genes and phenotypes, and mode of inheritance. Results Twenty-six probands (25 families) were included in the study (9 with a prenatal diagnosis and 17 with a postnatal diagnosis). Intraventricular hemorrhage (IVH) was the most common type of hemorrhage (n = 16, 62%), followed by subpial (n = 4, 15%), subdural (n = 4, 15%), and parenchymal (n = 2, 8%) hemorrhage. Causative/likely causative variants were found in 4 subjects from 3 of the 25 families (12%) involving genes related to the brain microenvironment (COL4A1, COL4A2, and TREX-1). Additionally, potentially causative variants were detected in genes related to coagulation (GP1BA, F11, Von Willebrand factor [VWF], FGA, and F7; n = 4, 16%). A potential candidate gene for phenotypic expansion related to microtubular function (DNAH5) was identified in 1 case (4%). Fifty-five percent of the variants were inherited from an asymptomatic parent. Overall, these findings showed a monogenic cause for pICH in 12% to 32% of the families. Interpretation Our findings reveal a clinically significant diagnostic yield of WES in apparently idiopathic pICH and support the use of WES in the evaluation of these cases

G - Stroke etiologies

P35-G: Prevalence of SARS-CoV-2 in childhood arterial ischemic stroke during the COVID-19 Pandemic: A multicenter experience

Lauren Beslow[1], Dipak Ram[2], Dana Harrar[3], Jonathan Santoro[4], Brian Appavu[5], Stuart Fraser[6], Thomas Rossor[7], Marcela Torres[8], Manoëlle Kossorotoff[9], Yenny Zuñiga Zambrano Zuñiga Zambrano[10], Marta Hernández-Chávez[11], Sahar Hassanein[12], Dimitrios Zafeiriou[13], Jenny Wilson[14], Michael Dowling[15], Ilona Kopyta[16], Timothy Bernard[17], Nomazulu Dlamini[18]

[1]Children's Hospital of Philadelphia, Perelman School of Medicine at the University of Pennsylvania, [2]Royal Manchester, [3]Children's National Medical Center, [4]Children's Hospital Los Angeles, [5]Barrow Neurological Institute at Phoenix Children's Hospital, [6]Children's Memorial Hermann Hospital, [7]Evelina London Children's Hospital, [8]Cook Children's Medical Center, [9]Univ Hospital Necker-Enfants malades APHP, [10]HOMI Fundación Hospital Pediátrico la Misericordia, [11]Pontificia Universidad Católica de Chile, [12]Ain Shams University, [13]Hippokratio General Hospital, [14]Oregon Health and Science University, [15]University of Texas Southwestern

Medical Center, [16]Medical University of Silesia in Katowice, [17]Children's Hospital Colorado, [18]The Hospital for Sick Children

Introduction In a meta-analysis of 108,571 adult patients with COVID-19, 1.4% had acute cerebrovascular diseases. Data collected March-May 2020 from 61 international centers showed that 0.82% of pediatric patients hospitalized with SARS-CoV-2 had acute ischemic strokes and that 4.7% of pediatric ischemic stroke patients tested had SARS-CoV-2. Hypotheses: We hypothesized that SARS-CoV-2 testing for children with acute arterial ischemic stroke (AIS) increased in June-December 2020 compared to during the early pandemic. We also hypothesized that the risk of acute AIS among pediatric SARS-CoV-2 hospitalizations is even lower than the early estimate. Methods: We surveyed international sites with pediatric stroke expertise. Survey questions included: numbers of hospitalized SARS-CoV-2 patients <18 years; numbers of incident childhood (29 days-18th birthday) AIS cases; frequency of SARS-CoV-2 testing for children with AIS; and numbers of childhood AIS cases positive for SARS-CoV-2 June 1-December 31, 2020. Results: Sixty-one centers from 21 countries provided childhood AIS data. Forty-seven centers (77%) provided SARS-CoV-2 hospitalization data. SARS-CoV-2 testing was performed in 335/373 acute AIS cases (89.8%) compared with 99/166 (59.6%) in March-May 2020, P=0.0000. Twenty-three of 335 AIS cases tested (6.9%) were positive for SARS-CoV-2 compared with 6/99 tested (6.1%) in March-May 2020, P=0.78. Of 23 AIS cases with SARS-CoV-2 June-December 2020, SARS-CoV-2 was the main stroke risk factor in 7, a contributory factor in 13, and incidental in 3. From centers with SARS-CoV-2 hospitalization data, of 6,693 hospitalized SARS-CoV-2 pediatric patients, 23 had AIS (0.34%) compared with 6/971 (0.62%) from March-May 2020, P=0.19. Conclusions: SARS-CoV-2 testing increased among children with AIS since the early pandemic. Risk of AIS is even lower than previously reported among children hospitalized with SARS-CoV-2. Over 6% of incident childhood AIS cases were positive for SARS-CoV-2, but SARS-CoV-2 was the main stroke risk factor in only 30% of strokes positive for SARS-CoV-2.

P36-G: The Brain-Immune Connection: ADA2 deficiency is a preventable cause of pediatric stroke

Barbra Giourgas[1], Sonal Bhatia[1]

[1]Medical University of South Carolina

INTRODUCTION: Adenosine deaminase 2 (ADA2) deficiency is a rare genetic cause of pediatric cerebrovascular disease typically associated with polyarteritis nodosa-like multi-systemic vasculitis. Patients usually present with strokes around age 2, after presenting with a livedo reticularis rash, systemic signs of inflammation and immunodeficiency. We present a now 2-year-old female, who initially presented at 5 months of age, then showed recurrent cerebral infarcts at 8 months of age, after which she was found to have a compound heterozygous pathogenic mutation involving the ADA2 gene. We highlight the importance of identifying ADA2 mutations in recurrent idiopathic strokes even in infancy and despite lack of accompanying systemic features, given available preventative immunotherapies. CASE: A previously healthy 5-month-old female initially presented with altered mental status without focal deficits and was found to have bi-thalamic infarcts with a non-yielding work up (vessel imaging, hypercoagulable studies, echocardiogram). She re-presented at 8 months of age with sudden left sided weakness with acute infarcts of the right corona radiata and internal capsule; early chronic infarcts of the right basal ganglia and the deep right periatrial white matter, likely representing a third ischemic event. Cerebral angiography was without signs of vasculitis. Whole exome sequencing sent given recurrent idiopathic strokes, identified a compound heterozygous pathogenic mutation in the ADA2 gene. She is maintained on etanercept with no recurrent infarcts but with residual mild left hemiparesis. DISCUSSION: ADA2 deficiency represents a rare cause for pediatric stroke, but one that is

easily identifiable with genetic testing and preventable with immunosuppression. With less than 75 reported cases, our patient is one of the youngest, and to our knowledge only to have ischemic strokes in the absence of systemic symptoms thus far. Consider early genetic testing for ADA2 mutations in infantile stoke, despite lack of systemic manifestations.

P37-G: Progressive multifocal leukoencephalopathy in a child with juvenile dermatomyositis presented as stroke

Ramit Singla[1], Norimitsu Kuwabara[2]

[1]Resident, [2]Central Michigan University

Case description 16 years old female with JD was admitted for stroke with symptoms of sudden onset right-sided hemiparesis, diplopia, dysarthria & difficulty word finding. On examination,was alert, oriented & neurologically intact except dysarthric speech, ataxic gait, bilateral dysmetria & dysdiadokinesia. MRI brain showed patchy asymmetric areas of T2/FLAIR hyperintensities without diffusion restriction involving pons, bilateral middle cerebellar peduncles & hemispheres. Initially started on immunosuppressive therapy (Steroids, Methotrexate & Cellcept) with concerns for cerebral vasculitis but due to worsening symptoms, imaging findings further investigation revealed positive JC Virus in her CSF fluid. Thus diagnosed with PML. Subsequently was treated with plasmapheresis. After 6 months of treatment & rehab she continues to show improvement. She is now able to sit without support & her speech is more clear. Recent JC virus results show 333 copies/ml (down from 2,900 at time of diagnosis). Recent MRI shows some improvement as well. Discussion: Active replication of JC virus in the brain leads to the development of PML, characterized by focal areas of demyelination and necrosis of glial cells(3). Diagnosis of PML is based clinically, neuroimaging & detection of JC virus in CSF. Most commonly reported neurological features are mono- and hemiparesis (50%), apathy (46%), confusion (38%), visual disturbance (29%), and seizures (21%)(2). In the pediatric population less than 50 cases of PML have been reported related to AIDS & less than 10 cases with inherited immunodeficiency. To our knowledge this is the first PML case reported in a pediatric patient with Juvenile Dermatomyositis (JD) and subsequent immunosuppression(1). References: 1.ReAngelini L, et al. Progressive multifocal leukoencephalopathy in a child with hyperimmunoglobulin E recurrent infection syndrome and review of the literature. Neuropediatrics. 2001;32(5):250-255. doi:10.1055/s-2001-19119 2.Shitrit D, et al. Progressive multifocal leukoencephalopathy in transplant recipients. Transpl Int. 2005;17(11):658-665. doi:10.1007/s00147-004-0779-3 3.Burke MT, et al. Progressive multifocal leukoencephalopathy with gastrointestinal disease in a pediatric kidney transplant recipient. Pediatr Transplant. 2013;17(5):E119-E124. doi:10.1111/petr.12107 Images: Asymmetric T2/FLAIR hyperintense signal within the right cerebellar peduncle the extensive cerebral hemisphere without evidence of diffusion restriction

P38-G: A cohort study of clinical and etiological profile of arterial ischemic stroke among Indian pediatric population: A single center experience

Ashwini B S[1], Minal Kekatpure[1], Vikram Huded[1], Rajiv Aggarwal[1], Tarishi Nemani[1], Anil Sapare[1], Sagar Hiremath[1]

[1]Narayana Health City

Introduction: Pediatric stroke is one of the leading causes of morbidity and mortality worldwide. Given the paucity of literature on etiology and presentation of childhood stroke in India, the clinico-etiological profile

study would give better insights into understanding, treating and preventing childhood stroke among the Indian population. Methods: This is a Retro-prospective cohort study conducted at Narayana health city, Bangalore over a 6months period. 76 children in the age group of 0-18yrs diagnosed as arterial ischemic stroke clinically and radiologically, were included. Data was collected from hospital database and at OPD visits, analyzed and compared with available Indian and international data. Results: Our study found that most common causes for pediatric stroke were: cardio-embolic (28%), cryptogenic (27%), arteriopathy (17%) and infections (15%). There was a definite male preponderance (66%) and most of them presented at 0-5yrs age (52%). Clinically, common presenting features were hemiparesis (63%), seizures (53%), facial paresis (26%), encephalopathy (22%), fever (17%), headache (9%). 57% of patients had single cerebral hemispheric involvement. Multiple vascular territories were involved in 30%. Nine children had recurrence (11%) of which, four had Moya Moya Disease, two had cardioembolic stroke, one had vertebral artery dissection, one had propionic aciduria and one Prothrombin gene mutation. Most of them were managed conservatively (95%). Four children (7%) underwent interventions: STA-MCA bypass with EDAMS, vertebral artery coiling and 2 mechanical thrombectomy. Conclusions: Our study found the cardio-embolic phenomena as the commonest cause of AIS in India. Rarer causes of stroke were also identified. 27% remained cryptogenic despite exhaustive evaluation. A definite male preponderance was noted. Further multi-centric studies with larger sample size would confirm these findings and shed light on common causes, modes of diagnosis, preventive and curative treatment specific to Indian pediatric population.

P39-G: Intrathecal chemotherapy-associated vasospasm in children with cancer

Lisa Sun[1], Wendy Ziai[2], Pat Brown[2], Rebecca Gottesman[2], Ryan Felling[3]

[1]Johns Hopkins School of Medicine, [2]The Johns Hopkins School of Medicine, [3]Johns Hopkins Hospital

INTRODUCTION: Mechanisms of chemotherapy-associated neurotoxicity are poorly understood. We hypothesized that a subgroup of children receiving intrathecal (IT) cytarabine develop subclinical vasospasm, which may contribute to long-term neurocognitive sequelae of cancer. METHODS: In this prospective self-controlled case series, we used transcranial Doppler ultrasound to serially evaluate cerebral blood flow velocities (CBFV) in participants less than 26 years old receiving IT cytarabine for new-onset cancer. We evaluated the frequency of cerebral vasospasm and compared demographics, laboratory data, baseline CBFV, and incidence of neurologic symptoms at 1-month follow up between children who did and did not develop cerebral vasospasm during induction. RESULTS: All 18 subjects had elevated anterior circulation CBFV at baseline when compared to age-based normative data. Four of 18 participants (22%) met criteria for vasospasm within 4 days of IT cytarabine administration. There was a non-significant trend toward younger children (6.2 versus 10.9 years, p=0.19) and more females (75% versus 36%, p=0.27) in the vasospasm group. The distribution of oncologic diagnoses differed between vasospasm and non-vasospasm groups (p=0.02), with acute myeloid leukemia identified as a potential risk factor for vasospasm. Children with vasospasm were more likely to have received intravenous cytarabine (75% versus 0%, p=0.01) and less likely to have received steroids (25% versus 100%, p=0.01). There were no differences in IT cytarabine initial or cumulative dose, baseline CBFV, hematocrit, or change in hematocrit from baseline between children with and without vasospasm. No serious neurologic complications occurred. Headache occurred with similar frequency in children with and without vasospasm (25% versus 21%, p=0.9). CONCLUSIONS: A subpopulation of children with hematologic malignancies, particularly AML, develop subclinical vasospasm after IT cytarabine. Future research is needed to determine if subclinical vasospasm in children receiving chemotherapy may be a harbinger of neurologic or cognitive sequelae, and if so, to focus on prevention of these complications in cases of cerebral vasospasm.

P40-G: Transient cerebral arteriopathy in children and adolescents, features of clinical manifestations and course

Alexander Kessel[1], Inna Shchederkina[1], Alexandra Kuznetsova[1], Yury Khachaturov[1], Elena Kuzmina[1]

[1]Morozov children city clinical hospital

The problem of a pediatric stroke has become relevant in recent decades. More than 50% of children with ischemic stroke are diagnosed with arteriopathy. Aim: assessment of the clinical features and course of arteriopathy in children diagnosed with arterial ischemic stroke (AIS). Materials: 72 patients with AIS, who were treated in Moscow primary pediatric stroke center from 2013 to 2018. Boys vs girls - 42 vs 30 (58,4%vs41,6%), the average age 7.6 years. The observation period was from 16 months to 5 years. Results: in 43 children out of 72 were diagnosed the arteriopathy. 30/43 showed signs of unilateral focal stenosis of the distal internal carotid artery (ICA) and / or the middle cerebral artery (CMA) and / or the anterior cerebral artery (PMA) with / without occlusion, The severity of stroke by PedNIHSS in average 7,3. CT was made: CT angiography 28 patients (from 7 days to 4 months from the onset of stroke (1 patient twice), 8 - direct catheter angiography. Arteriopathies in patients with focal cerebral artheropathy were divided: vasculitis- 3, progressive arteriopathy -4, dissection-3, transient cerebral arteriopathy (TCA)- 20. Dynamic MRI study (from 3 to 6 months) showed: a complete restoration of blood flow-5; increase of stenosis -4; decrease of stenosis - 11; without dynamics relatively acute period -3. MR-angiography after 9-12 months: full restoration of blood flow-9; reduction of stenosis (incomplete recovery) -7. Relapse of AIS was observed in 6 patients (11.6%)with artheriopathy during the follow-up period. Conclusion: Modern methods of neuroimaging have significantly improved the diagnosis of childhood stroke, clarifying the nature and extent of cerebral vascular lesions, including artheriopathy, which is very important in determining the management of children and adolescents.

P41-G: Cancer-associated stroke in children: Results from the International Pediatric Stroke Study (IPSS)

Lisa Sun[1], Alexandra Linds[2], Mukta Sharma[3], Mubeen Rafay[4], Sudhakar Vadivelu[5], Sarah Lee[6], Leonardo Brandão[2], Brian Appavu[7], Jeremie Estepp[8], Juliette Hukin[9], Sahar MA Hassanein[10], Anthony Chan[11], Lauren Beslow[12]

[1]Johns Hopkins School of Medicine, [2]The Hospital for Sick Children, [3]Children's Mercy, University of Missouri Kansas City, [4]University of Manitoba, Children's Hospital Research Institute of Manitob, [5]University of Cincinnati College of Medicine, [6]Stanford University School of Medicine, [7]Barrow Neurological Institute at Phoenix Children's Hospital, [8]St. Jude Children?s Research Hospital, [9]Children?s and Women?s Health Centre British Columbia, [10]Ain Shams University, [11]McMaster University, [12]Children's Hospital of Philadelphia, Perelman School of Medicine at the University of Pennsylvania

OBJECTIVES: The prevalence of cancer among children with ischemic stroke is unknown. This study sought to evaluate cancer-associated childhood ischemic stroke in a multinational pediatric stroke registry. METHODS: Children between 29 days and 19 years of age with arterial ischemic stroke (AIS) and/or cerebral sinovenous thrombosis (CSVT) enrolled in the International Pediatric Stroke Study between January 2003 and June 2019 were included. Data including types of cancer, stroke etiology, and stroke treatment were compared between subjects with and without cancer using Wilcoxon rank sum and chi-square tests. RESULTS: Remote or active cancer was present in 99 of 2968 children (3.3%) with AIS and 64 of 596 children (10.7%) with CSVT. Children with cancer were older than children without cancer in both the AIS (9.5 versus

5.7 years, p=0.0001) and CSVT groups (8.2 versus 6.3 years, p=0.014). Among children in whom cancer type was identified, 42 of 88 (48%) AIS cases had brain tumors and 35 (40%) had hematologic malignancies; 45 of 58 (78%) CSVT cases had hematologic malignancies and 8 (14%) had brain tumors. Primary causes most commonly reported in children with cancer-associated AIS included: arteriopathy (n=34, 62%), cardioembolic (n=9, 16%), prothrombotic state (n=4, 7%), and infection (n=4, 7%). The most frequently reported primary cause of CSVT in children with cancer was prothrombotic state (n=38, 83%). Children with cancer were less likely to receive any antithrombotic therapy for AIS (58 % v. 80%, p=0.007) and anticoagulation for CSVT (71% v. 87%, p=0.046), compared with children without cancer. Recurrent AIS (5% v. 2%, p=0.04) and CSVT (5% v. 1%, p=0.006) were more common among children with cancer. CONCLUSION: Cancer is an important risk factor for incident and recurrent childhood stroke. Although multiple risk factors for stroke were noted in children with cancer, presence of cerebral arteriopathy in children with AIS and prothrombotic state in children with CSVT appear to be the most significant underlying mechanisms contributing to increased risk for stroke in this population. Future research investigating the role of stroke prophylaxis in the high-risk subpopulations identified in this study are warranted.

P42-G: Ischemic stroke and immune encephalitis: Which came first?

Maria Rios Pistoia[1], Laura De Rosa[1], Maria Vaccarezza[1], Clarisa Maxit[1], Guilllermo Agosta[1]

[1]Hospital Italiano de Buenos Aires

Introduction: Immunomediated encephalitis has been studied with great interest in recent years. So far, etiology is not known, but it could be stowed by viral infections. Most cases have normal neuroimaging. Our patient presented an associated ischemic stroke. Since our patient presented the two pathologies, related in time, we wondered what the relationship between them might be. Clinical case: Patient of 2 years, without perinatological history to highlight, that after a febrile picture presents clonic focal crises of upper right limb, continuous, difficult to manage and ipsilateral hemiparesis. MRI shows ischemic stroke in the territory of the middle cerebral artery and left anterior choroid artery. A week it presents hemorrhagic reformation, with good evolution.At 30 days from the onset of symptoms, it presents sensory alteration and adds abnormal movements. Anti-NMDA encephalitis is confirmed: it had qualitative antibodies in Cerebrospinal Fluid and plasma. MRI chronic evolution of known injury. Electroencephalogram with typical pattern of anti-NMDA encephalitis. It required first- and second-line treatment. After 6 months, there is clinical improvement. At the moment the child is 7ys and go to regular school, he has a mild hemiparesis. Discussion: We wondered if there was any relationship between these two pathologies. Is it an ischemic stroke that has evolved into an NMDA anti-NMDA encephalitis or an anti-NMDA encephalitis that has had an associated ischemic brain injury? And although they seemed far away, they have several pathophysiological mechanisms that relate them. In both pathologies, the brain receptor involved in cell damage is GluN1 NMDA. We can't confirm theories with any complete tests.

H - Stroke readiness & guidelines

P43-H: Creation of a Suspected Stroke Protocol: Streamlining evaluation and diagnosis of children with acute focal neurological deficit

Marcie Files[1], Jennifer Flint[1], Timothy Zinkus[1], Valorie Mc Dougall Kestner[1], Celeste Tarantino[1], Roha Khalid[1], Mukta Sharma[1]

[1]Children's Mercy, University of Missouri Kansas City

Objectives: 13/100,000 children suffer from an acute ischemic stroke per year, resulting in one of the top 10 causes of death in children. Literature shows that two-thirds of children with an acute stroke present to a hospital within 3-6 hours of symptom onset yet there remains a delay in diagnosis due to lack of awareness and lack of protocols for neuroimaging and treatment. Our goal was to improve the care of pediatric patients with symptoms of an acute ischemic stroke by streamlining early recognition of symptoms and diagnosis. Methods: We created a stroke protocol for our hospital that allows for rapid neuroimaging of patients with an acute neurological deficit who are eligible for thrombolysis or clot retrieval. Results: Before development of the stroke protocol at our hospital, a 2 year old patient presented to the Emergency Department 1.5 hours after acute onset of left sided weakness. He was admitted to the Neurology service and MRI performed 22 hours after the onset of symptoms demonstrated an acute infarct. After development of the stroke protocol, a 7 year old female developed acute onset of left sided weakness while inpatient. The stroke protocol was initiated and an acute ischemic stroke was identified on MRI within 4 hours. She was able to receive intravenous tPA as well as undergo thrombectomy. Conclusions: The development of an acute stroke protocol at our hospital has improved the evaluation and diagnosis of children with focal neurological deficits and allows for rapid diagnoses and treatment of patients with acute ischemic strokes.

P44-H: Stroke post congenital cardiac surgery - Adherence to national guidelines and reasons for non-adherence

Alexander Simpson[1], Isobel Eckersall[1], Nicole Perry[1], Katharine Forrest[1]

[1]Royal Hospital for Children, Glasgow

Background Stroke is a recognised complication following congenital cardiac surgery. In 2017, RCPCH published a guideline outlining investigations to be performed in children with evidence of stroke. Objectives To determine adherence to the RCPCH Stroke Guideline in patients with evidence of stroke who had undergone congenital cardiac surgery. To perform a structured survey to explore reasons for non-adherence. Methods Records of all paediatric cardiac patients undergoing surgery between January 2014 and July 2020 were cross-referenced with patients recorded as having ischaemia on brain imaging. 34 patients were identified. 16 patients who died during admission were excluded. Records of the remaining 18 patients were examined to ascertain rates of adherence to the RCPCH's recommended investigations. Subsequently, a structured survey was conducted amongst paediatric intensive care doctors to explore reasons for non-adherence. Results Table 1 documents adherence to investigations recommended by the RCPCH. Of note, complete adherence was not achieved in any patients. Of 33 doctors surveyed, 10 (30%) were aware of the guideline. When asked how likely they were to use the term 'stroke' to describe focal ischaemia on imaging, 16 (48%) respondents stated they were likely/very likely, with 9 (27%) stating they would be unlikely/very unlikely. When asked about which laboratory investigations they would perform, various tests referenced in the guideline were mentioned, but no respondents mentioned iron, total iron binding capacity, plasma homocysteine, lipoprotein, 2GP1 or alpha galactosidase. Discussion Cardiac investigations and routine blood tests were performed as per guidelines on congenital cardiac surgery. Neurological imaging was generally performed in keeping with RCPCH guidelines. However, other investigations were performed on an ad-hoc basis. The survey results suggest that this may be due to a lack of awareness of both the stroke guideline and the significance of the laboratory tests. Furthermore, disparities in terminology may contribute to a lack of

consideration of the stroke guideline. As a result of this audit, the RCPCH Stroke Guideline will be available through the NHS GG&C guidelines application. Education will be delivered to raise awareness, and a stroke checklist will be introduced.

P46-H: Early diagnostics of a stroke in children. Experience of Moscow primary pediatric stroke center and Moscow Ambulance

Inna Shchederkina[1], Yury Khachaturov[1], Nikolay Plavunov[2], Andrey Sidorov[2], Elena Petryaykina[1], Valery Kadyshev[2], Alexander Kessel[1]

[1]Morozov children city clinical hospital, [2]First aid station

Last years there has been a tendency to increase the incidence of stroke in children, which may be associated with an improvement in the diagnosis of this pathology in the pediatric population. Aim. Analysis of the clinical synmptoms and the conformity of the diagnoses: guides (according to the ambulance) and the discharge one. Materials. From October 2018 to December 2019, 502 hospitalization in the Primary pediatric stroke center of Morozov Hospital by the Moscow Ambulance with diagnoses of arterial ischemic stroke (AIS), hemorrage stroke (HI) and transit ischemic attack (TIA) were analyzed. 48.8% boys and 51.2% girls were hospitalized. Results. Of 502 admissions, the directing diagnosis was TIA - 38.6%, AIS - 52,6%, HI - 8.8%. The symptoms were: headache -38.5%; dizziness - 25.5%, ataxia - 21.1%; sensory impairment - 10.6%, paresis - in 10%, speech impairment and visual impairment were detected in 9.6% and 5.4%, facial asymmetry - 6.2%, and seizures - 4%. In 30.7% patients, at the time of admission to the Center, there was no a neurological deficit. The average time from the onset of the first symptoms to hospitalization in the hospital was more 4 hours. Neuroimaging was performed in 54.8%. Computed tomography(CT)- 41.4%, magnetic resonance imaging (MRI) - 20.7% cases, in 7.9% both CT and MRI. AIS was confirmed in 11,2%, HI -9.1%. In confirmed AIS, the common symptoms were: paresis- 84.6%, ataxia- 84.6%; facial asymmetry- 61.5%; speech impairment and impaired sensitivity - 38.5%, respectively; visual impairment - 15.4%; nausea or vomiting and convulsions - 7.7%. In 84% patients, stroke was not confirmed. Conclusion Based on the analysis of the most frequent symptoms and "mistakes", the "Attention, stroke!" Scale was developed for ambulance doctors, which has been practiced since January 2020 in the Moscow Ambulance.

P47-H: Pediatric code stroke: a single center experience

Stuart Fraser[1], Nicole Gonzales[2], Mary Koenig[2]

[1]University of Texas McGovern Medical School, [2]Dr.

Introduction: Pediatric acute stroke is often misdiagnosed in the acute setting. Computed tomgraphy imaging has a low sensitivity and specificity for diagnoses in pediatric patients presenting with acute focal neurologic deficit. We established a collaborative protocol involving both the vascular neurology and pediatric neurology divisions in our institution for pediatric code stroke. Methods: Multidisciplinary meetings were held with the division of vascular neurology, endovascular surgery, child neurology, intensive care and emergency medicine to update our pediatric acute stroke protocol to prioritize magnetic resonance imaging over computed tomography. Educational lectures about pediatric acute stroke were given to the department of emergency medicine, division of vascular neurology, division of child neurology, and division of pediatric intensive care. Results: The protocol has been active for 18 months. There have been 20 pediatric code strokes. Magnetic resonance imaging was obtained first in 14 of 20 cases. The average time from code stroke

to imaging in the emergency department was 60 minutes. The average time in the inpatient setting was 115 minutes. Final diagnosis was acute ischemic stroke in 3 patients. The other diagnoses were; RCVS (1), Methotrexate toxicity (1), Seizure (10), Metabolic (2), Migraine (1), Conversion (1), Bell's Palsy (1). 1 patient recieved tissue plasminogen activator and mechanical thrombectomy for basilar artery occlusion (Figures 1 and 2). Conclusions: Magnetic resonance imaging is a sensitive and specific first-line imaging choice in pediatric patients presenting with acute focal neurologic deficit. Multidisciplinary collaboration including both vascular and pediatric neurologists may improve diagnosis time for patients presenting with acute focal neurologic defict.

[PRINT ONLY] P48-H: Developing algorithm for early evaluation and management for stroke in sickle cell disease in pediatric emergency department

Nazia Tabassum[1], Jay Rilinger[1], Celeste Tarantino[1], Valerie McDougall Kestner[1], Timothy Zinkus[1], Mukta Sharma[1]

[1]Children's Mercy, University of Missouri Kansas City

Background: Sickle cell Disease (SCD) is most common autosomal recessive disease associated with vasculopathy leading to neurological complications like overt stroke (ischemic, hemorrhagic), TIA, CSVT, seizure, hemiplegic migraine and PRES . Early evaluation for neurological complications is important in management of Stroke for patient presenting with neurological complaints. This entails multidisciplinary team approach for prompt neuroimaging and expedited care with IV line placement, obtaining labs and timely intervention with either simple or exchange transfusion. Below is the algorithm developed by multidisciplinary team- Pediatric Hematology, Intensivist, Neurology, Emergency Medicine, Transfusion medicine specialist and Neuroradiology. Objectives: Ealry identification and implementation via stroke alert in ED, inpatient or during transporation Early notification to multidisciplinary team to initiate treatment To establish sickle cell stroke algorithm and powerplan to ease the anxiety of providers in placement of orders Monthly Stroke Meeting updates: to identify gaps in knowledge, reasons for delayed activation and notifications, and new sickle cell research updates. Conclusion: Appropriate early imaging and early intervention is ideal for management of stroke in pediatric SCD to minimize morbidity and mortality. Development of algorithm and power plan has led to early initiation of treatment.

P49-H: Pediatric acute stroke response protocols

Dana Harrar[1], Giulia Benedetti[2], Anuj Jayakar[3], Jessica Carpenter[4], Tara Mangum[5], Melissa Chung[6], Brian Appavu[7]

[1]Children's National Medical Center, [2]Seattle Children's Hospital; University of Washington, [3]Nicklaus Children's Hospital, [4]Children's National Hospital; George Washington University, [5]Phoenix Children's Hospital, [6]Nationwide Children's Hospital, [7]Barrow Neurological Institute at Phoenix Children's Hospital

Background and purpose: The implementation of acute stroke response protocols has resulted in a decrease in the time to diagnosis of acute stroke in children at select institutions, and this is essential for expanding access to hyperacute therapies. We aim to describe existing pediatric acute stroke response protocols in the United States and Canada to better understand how pediatric centers might implement such protocols within the context of institution-specific structures. Methods: We performed a survey-based study of pediatric stroke specialists focused on institutional acute stroke protocols. The survey queried: hospital demographics; child

neurology and pediatric stroke demographics; acute stroke response; imaging; and hyperacute treatment. Results: Forty-seven surveys were analyzed. Most respondents practice at large, free-standing children's hospitals with moderate-sized neurology departments and at least one neurologist with expertise in pediatric stroke. The majority of institutions have an acute stroke pathway or protocol, and all but one of these include the activation of a stroke alert page. Most institutions cite magnetic resonance imaging as the preferred imaging modality and employ abbreviated magnetic resonance imaging protocols for acute stroke imaging. Most institutions also have either computed tomography- or magnetic resonance-based perfusion imaging available. At least one patient had been treated with intravenous tissue plasminogen activator and/or mechanical thrombectomy at the majority of institutions in the year prior to our survey. Conclusions: An acute stroke response protocol is utilized in at least 41 pediatric centers in the United States and Canada. Most acute stroke response teams are multidisciplinary, have annual experience providing intravenous tissue plasminogen activator and/or mechanical thrombectomy, and prefer abbreviated magnetic resonance imaging over computed tomography for stroke diagnosis. Further studies are needed to standardize practices of pediatric acute stroke diagnosis and hyperacute management.

P50-H: Is stroke on your differential? Acute stroke alert implementation in a pediatric ER

Rebecca Kahler[1], Joy Bartholomew[2], Mukta Sharma[3]

[1]Children's Mercy, [2]Children's Mercry, [3]Children's Mercy, University of Missouri Kansas City

Background: Twenty to forty percent of children who suffer a stroke will die. Of those who do live, 50-80% will have permanent neurologic deficits. Early identification and intervention increases survival and decreases sequelae yet recognition of a stroke is often missed. With an incidence of only 6.4/100.000 in children aged 0-15, the rarity, the atypical presentations compared to adults and potential confounding stroke mimickers often impede prompt diagnosis. Objective: Improve recognition that children do have strokes and expedite early assessment and diagnosis that is imperative to good outcomes. This presentation will advance nursing skills in the recognition of stroke types and symptoms to familiarize them with the tools needed, such as the Pediatric National Institute of Health Stroke Scale (PedNIHSS), to aid timely diagnoses. Design/Methods Retrospective institutional data review of children presenting with strokes between 07/2017-03/2018 was compared to the nationally recognized predisposing conditions. The prevalence of risk factors in this review exceeded what is commonly known and has been instrumental in refining our screening process, development of nurse assessment tools and a stroke alert protocol. Results: Seventy-five incidences of stroke were identified during an 8 month period, the majority were through the Emergency department with initial triage by a nurse. The causes and risk factors were categorized and 11 diagnostic groups were documented. There were 7 outliers to the commonly recognized associated conditions. All children who met assessment criteria for suspected stroke, average two -three per month, received the PedNIHSS evaluation. This screening process also identified stroke mimickers who did not require additional evaluations such as MRI. Those who were positive for PedNIHSS, defined as a score greater than 4, received timely intervention which has shown to improve long term outcomes. An acute stroke alert protocol that includes an electronic order set was developed and its efficacy continues to be evaluated at quarterly multidisciplinary meetings. Conclusion: Pediatric stroke is a reality and as pediatric providers we need to be knowledgeable and attentive to presenting symptoms. Stroke needs to be in our differential, we need to be familiar with the PedNIHSS and we need to intervene promptly for the best possible outcomes.

P51-H: Developing a pediatric ischemic stroke code protocol at a comprehensive stroke center: The Georgetown experience

Jacqueline Leutbecker[1], Ashley Carlson-Chalifoux[1], Kelsey Monacci[1], Paola Pergami[1]

[1]Georgetown University Medical Center/CNMC

Background: Epidemiologically pediatric strokes are rare; therefore, index of suspicion is low and many cases are not recognized early enough to qualify for acute treatment. With this small patient population, many regions have yet to develop protocols and policies for acute pediatric stroke response. In the District of Columbia, MedStar Georgetown University Hospital (MGUH) is the only Joint Commission Accredited Comprehensive Stroke Center with a pediatric intensive care unit. Purpose: The purpose of our program was to develop institutional protocols for pediatric stroke patients who are candidates for hyper-acute treatment. Protocols are focused on pediatric patients who present through our emergency department or are transferred from other facilities within the time-window for acute treatment. The protocols are established to ensure a clear process for physicians and staff to follow. Methods: An interdisciplinary group met to discuss modification of our current Adult Ischemic Stroke Code Protocol needed for a coordinated approach to pediatric stroke patients. The team members included the Stroke Coordinator, Stroke Nurse Navigator, Stroke Nurse Practitioner, Pediatric Neurologist, Medical Director of the pediatric ICU, Manager of the Pediatric ICU, Pediatric Anesthesia, Medical Director of the stroke program, and the Emergency department team. Results: The interdisciplinary team was able to adapt a protocol using the Adult Ischemic Stroke Code Protocol. Key differences between the adult and pediatric stroke code work-flow consisted of including pediatric neurologists early in the decision process, pediatric nurses to assist with monitoring children, pediatric anesthesia for assistance with imaging and/or endovascular intervention. The order sets were adjusted to include weigh-based calculation for medications, age-based monitoring parameter and specific pediatric needs. Conclusion: It is possible to develop a Pediatric Stroke Code Protocol based on the Adult Ischemic Stroke Code Protocol to include all modification appropriate for pediatric care and still maintain the rapid workflow that everyone is familiar with (from the adult stroke protocol). It is essential to include all key stakeholders including the pediatric and vascular neurology team members to ensure a smooth and safe process.

[PRINT ONLY] P52-H: Early experience of the first paediatric code stroke at an Australian Children's Hospital

Michaela Waak[1], Natalie Deuble[1], Adriane Sinclair[1]

[1]Queensland Children's Hospital

Introduction Arterial ischaemic stroke (AIS) is a time critical neurological emergency. In-hospital delay to diagnosis in children is common. Methods A code stroke was developed aiming to improve time to diagnosis and standardisation of management. Bundle elements incorporated were timely recognition (pre-hospital and ED), system to decrease in hospital delay (rapid access to stroke neurologist, rapid sequence MRI), guideline including save administration of reperfusion therapies, neuroprotective care and secondary prevention. We describe the first 12 months of code stroke experience with comparison to 24 months pre-implementation (Code stroke and AIS data from January 2019 to January 2020 were prospectively collected and compared to AIS patients between January 2017 and December 2018). Results 17 code strokes were activated. Four patients (24%) had a diagnosis of AIS, 8 had migraine, 3 had infection/autoimmune encephalitis, 1 had a functional neurological disorder and 1 had Moya-Moya. Four additional patients were diagnosed with AIS but

did not meet criteria for code activation (presentation > 24 hours post symptom onset, age). One patient presented to a mixed ED with an adult code stroke system which was activated before patient transfer to our center. All code patients were triaged in <10 minutes and were assessed by an emergency senior medical officer within 20 minutes. Time to magnetic resonance imaging (MRI) was 20 - 180 minutes (mean 56 min). One patient received rTPA and improved clinically (PedNIHSS 9 on D0, 4 on D1). In all patients MRI was diagnostic and impacted management decisions. in the 2 years pre-code 16 patients were diagnosed with AIS state-wide, however only 3 patients would have activated a stroke code on arrival to our institution. Time to diagnostic neuroimaging ranged from 1 hour to 5 days. Two patients received reperfusion therapies. Conclusions Code stroke implementation resulted in rapid access to medical assessment, diagnostic imaging and treatment for AIS. Comparison to pre-code AIS data suggests that code implementation has also improved state-wide access to comprehensive stroke care. Ref: Elbers, J. The pediatric stroke code: Early management of the child with stroke. J. Pediatr. 167, 19-24.e4 (2015); Kassardjian, C. In-Patient Code Stroke: A Quality Improvement Strategy to Overcome Knowledge-to-Action Gaps in Response Time. Stroke 48, 2176-2183 (2017),Mallick, A. Diagnostic delays in paediatric stroke. JNPP 2015

I - Stroke rehabilitation

P53-I: The relationship between resting-state functional connectivity and motor outcome after pediatric arterial ischemic stroke

Leonie Steiner[1], Stephanie Winkelbeiner[1], Regula Everts[1], Andrea Federspiel[1], Sandeep Kamal[1], Juan Antonio Delgado Rodriguez[1], Salome Kornfeld[1], Nedelina Slavova[1], Roland Wiest[1], Alain Kaelin-Lang[1], Maja Steinlin[1], Sebastian Grunt[1]

[1]University of Bern

Introduction: Studies investigating pediatric patients after arterial ischemic stroke (AIS) with hemiparesis compared with patients with good clinical outcome and typically developing peers (TDP) are scarce. The comparison of different motor outcome after AIS and the underlying functional connectivity in the motor network could provide valuable insights into adaptive vs. maladaptive reorganization. Hypothesis: We hypothesized that patients with hemiparesis show lower interhemispheric and intrahemispheric functional connectivity than patients with good clinical outcome and TDP. Secondly, we hypothesized that asymmetry of upper limb function is related to functional connectivity of the motor network. Methods: We recruited patients after AIS in the chronic phase (2 or more years after diagnosis; diagnosed before 16 years). Nine patients with hemiparesis, nine patients with good clinical outcome and 11 TDP underwent a standardized motor assessment, single-pulse transcranial magnetic stimulation (TMS), and resting-state functional magnetic resonance imaging (rs-fMRI). TMS was performed to determine the type of corticospinal tract wiring and rs-fMRI to investigate network connectivity within the motor network. Results: All participants showed a contralateral corticospinal tract wiring pattern, as expected in a typically developing brain. Patients with hemiparesis showed lower intra- and interhemispheric connectivity compared with patients with good clinical outcome. In contrast, patients with good clinical outcome showed considerably higher intrahemispheric connectivity compared with TDP. Furthermore, lower intrahemispheric connectivity was related to greater asymmetry of upper limb function. Conclusion: These findings contribute to a better understanding of neuroplasticity, motor recovery, and how targeted therapies after AIS could affect network changes. Future studies should clarify whether timing or type of rehabilitation influence connectivity.

P54-I: Multi-component occupational therapy intervention in a pediatric constraint induced movement therapy camp setting

Shelley Dean[1]

[1]Crossway Pediatric Therapy

Introduction: Occupational therapy is one of the rehabilitative professions that treats children with hemiplegia secondary to perinatal and pediatric stroke (Rowland, Cooke, & Gustafsson, 2008). There is limited research on the assessment and rehabilitation of pediatric hemiplegia secondary to stroke (Marcroft, Tsutsumi, Pearse, Dulson, Embelton, & Basu, 2018). Aims: The goal is to demonstrate the effectiveness of combining pediatric constraint induced movement therapy (CIMT) with a collection of evidenced based rehabilitation interventions for the treatment of hemiplegia secondary to pediatric and perinatal stroke in occupational therapy. Hypothesis:The combined approach will yield increased quality of movement, more efficient motor patterns and spontaneous use of the affected side for participation in meaningful bilateral and unilateral tasks. Methods: This quasi-experimental, case series study examined children ages 7 to 13 years in a theme-based CIMT camp for 10 or 15 days for two consecutive or alternate years. Each of the six participants received multiple components of intervention combined with CIMT. Pre and post-test measures were taken to examine outcomes using the Abilhand-Kids and Goal Attainment Scaling to compare unilateral and bilateral function from the first round of camp were compared to the second round of intensive camp. Results: The analyses determined that the dosage of CIMT camp for pediatrics can be effective for 5 hours a day for 10 or 15 days especially for first time attendees. Additionally, there is significant sustained benefit to attending CIMT camp the following year or the alternate year. There is value in combining evidenced based practices with CIMT. These participants demonstrated an increase in mean difference for functional participation, range of motion and automaticity of use of the hemiparetic hand for meaningful activities after an intensive combined approach of CIMT, and collective evidence-based rehabilitation. Conclusion: Essential components of evidenced based rehabilitative interventions combined with CIMT can facilitate improvement of motor control for the pediatric stroke population. This work is the first to demonstrate an effective framework for the unilateral treatment of pediatric hemiplegia combining the use of essential multi-components. This report can be used as a guideline for pediatric stroke treatment in the field of occupational therapy for the purpose of knowledge translation.

[PRINT ONLY] P55-I: A comparison of high intensity periodic rehabilitation to usual weekly in children with cerebral palsy

Jill Heathcock[1], Jeff Pan[1], Rachel Ferrante[2], Nathalie Maitre[2], Kelly Tanner[2], The ACHIEVE Stakeholders[1]

[1]The Ohio State University, [2]Nationwide Children's Hospital

Background: Cerebral palsy (CP) is the most common childhood motor disability. CP affects the development of a child's movement, balance, and posture. Pediatric rehabilitation can help children with CP move and be more functionally independent. The research team compared giving children with CP either consistent weekly physical therapy sessions or daily high intensity 2-week periodic bursts followed by 18-week breaks. Importantly, the total number of therapy hours and the length of the treatment periods were identical which was identified as a missing gap in the current literature. The objectives were to (1) compare the short-term and explore the long-term effects of 2 service delivery models: 1 hour per day, 1 x per week for 40 weeks (usual weekly); and 2 hours every weekday for two 10-consecutive-weekdays (total 4 weeks), for a repeated

"periodic" bout (high intense periodic) of outpatient physical therapy (PT); Methods: Using a randomized controlled trial (RCT) with a patient-centered self-selection group option, N=100 children with CP ages 2-8 years and GMFCS levels I-V participated in this study (treatment groups described above). Results: A Linear mixed model demonstrated a significant improvement at the end of 9 month treatment period for the primary outcome measure the GMFM-66 (Gross Motor Function Measure). The difference between the daily and weekly schedule treatment was not statistically significant (-0.95, 95% CI: -2.96, 1.05; P=0.35), where weekly has non-significant higher improvement than high-intensity periodic. Statistical analysis for secondary outcome measures including Goal Attainment scaling and the Bayley Scales of Infant Development followed a similar result with significant main effects of time and no main effect of group or interaction of group and time. Conclusions: Motor skills, measured by clinical outcome measures, goal attainment scaling, and parent surveys consistently demonstrated patient-centered improvements following high intensity period and usual weekly physical therapy but not difference between groups. Often, the weekly group showed a larger non-significant change score which was opposite to our hypothesis. Younger children and those in lower GMFMCS levels (better function) showed more positive changes. Parent satisfaction was related to better improvements in motor function.

P56-I: Participation and manual ability in children diagnosed with arterial ischemic stroke: Do parents of more affected children expect more from their children?

Sebastian Grunt[1]

[1]Inselspital, University Childrens Hospital

Introduction Childhood arterial ischemic stroke (AIS) is rare but associated with significant morbidity. Approximately 30% of new-borns and 50% of children diagnosed with AIS later develop hemiparesis. The relationship between their manual ability and participation at home, school and in the community has not been investigated. Patients and Methods Fourteen children with AIS (diagnosis at least 2 years ago, age 5 to 17 years) and 21 typically developing peers were examined. Participants were contacted by mail and asked to fill in questionnaires regarding their manual ability (ABILHAND-Kids) and participation (Participation and Environment Measure - Children and Youth [PEM-CY]). A Mann-Whitney-U test was used to calculate group differences, correlations were calculated using a Spearman Rank Correlation coefficient. Results There was a significant difference in manual ability between stroke patients and typically developing peers ($p<0.001$). While there were no significant group differences in participation frequency and involvement at home, school and in the community, parents of AIS patients desired significantly higher participation at school ($p=0.022$). There was a significant correlation between manual ability and the desired change in participation at school ($p=0.026$). Conclusion Although children after AIS seem to participate equally often and be equally well involved in participation at home, school and in the community, parents of children diagnosed with AIS appear to desire a higher level of participation at school - especially when manual ability is reduced. Future studies must investigate whether increased parental participation desires in this vulnerable patient group are associated with increased pressure to perform at school.

P57-I: Dr. Mom: Neuropsychological treatment from the perspective of a parent of a perinatal stroke survivor

Carla Morton[1]

[1]Cook Children's Medical Center

Neuropsychologists play an integral role in a comprehensive pediatric stroke team, and raising a perinatal stroke survivor provides unique insights when working with patients and their families. First, neuropsychologists can help parents understand their child's current and potential cognitive and behavioral outcomes as well as coping with uncertainty regarding their child's future. In addition, depending upon medical factors surrounding their child's stroke, parents may need help coping with their own psychological trauma and feelings of guilt. Parents may also benefit from guidance on how to manage changes in relationships with extended family and friends, and they should be prepared to model responses to questions from others regarding their child. In addition to developmental deficits, pediatric stroke survivors are at increased risk for behavioral dysregulation, and parents may need coaching in behavioral strategies or referrals to psychiatric treatment resources. As a child becomes more self-aware, parents can also benefit from support as they help their child understand his or her medical condition and grieve any newly appreciated losses that their child may experience. Because school is a child's job, parents may need to become their child's academic advocate, and they may be expected to educate school personnel about childhood stroke. In addition to a child's current challenges, parents should also help teachers understand that some stroke-related cognitive deficits may not become apparent until the child is older, and that his or her need for academic support may increase over time. Providing top-quality stroke care does not end with medical stabilization. Stroke survivors and their families should be treated from a lifespan perspective with an understanding of the far-reaching effects that a child's stroke can have upon the entire family system.

P58-I: Intensive bimanual therapy for pediatric chronic hemiparesis: implementing research and optimizing caregiver and patient-reported outcomes

Todd Levy[1]

[1]Children's Hospital Philadelphia

Background And Aims Existing research supports intensive therapy involving goal-directed training and practice of skills. Such programs are challenging to deliver in the ambulatory setting due to cost and logistics. We aimed to implement a Hand-Arm Bimanual Intensive Therapy (HABIT) program, measure and improve participant-reported outcomes, and expand program size while maintaining quality. Methods HABIT invovled a camp-like environment for 6 hours/day X 10 days, plus home programs. Fourteen campers (ages 6-11) completed the program in 3 groups. Caregiver education improvements were based on a pilot program (2017). Assisting Hand Assessment (AHA) hierarchy guided each camper's treatment (2018-2019). Program capacity increased from 4 to 6 campers. The role of psychology was was intensified yearly. Outcome assessments included AHA, Canadian Occupational Performance Measure (COPM), PROMIS Parent Proxy Upper Extremity Questionnaire, The Pediatric Outcomes Data Collection Instrument (PODCI), and participant surveys. Results: -Group AHA scores improved at program completion (P=.0006) and remained statistically significant at 30 days (P=.0007) despite a decreasing trend. Pattern consistent with literature. -8 of 13 achieved at least the smallest detectable difference (SDD), consistent with literature -Group exceeded criteria for clinically meaningful change -9 of 13 campers achieved clinically meaningful changes in performance and satisfaction -Improvements for 2018, sustained for 2019 Surveys show improved parent and patient-reported outcomes in 2018, sustained for 2019 Conclusions We implemented HABIT in our ambulatory care center with results comparable to the research literature. AHA score improvements were sustained at 30 days in an expanded program that was enhanced based on QI initiatives, with high satisfaction levels reported by participants.

J - Vascular malformations & ICH

P59-J: Neurosurgical service in the primary pediatric stroke center in Moscow

Zokir Zohidov[1], Matvey Livshitz[1], Inna Shchederkina[1], Alexander Levov[1], Gennady Chmutin[2]

[1]Morozov children city clinical hospital, [2]Russian University of Friendship's

Hemorrhage (HI) accounts for about half of all strokes in children. In total, vascular brain abnormalities accounted for 43.5% of all hemorrhages. Aim: to evaluate the incidence of HI in the primary pediatric stroke center of the Morozov Children City Clinical Hospital; revealed cerebral vascular anomalies without rupture, neurosurgical treatment options. Material: children who were treated in the primary pediatric stroke center from 2017 to 2019, with cerebrovascular diseases, and are included in the Moscow Pediatric Stroke Register. Results: 142 children were enrolled in the register in 2017: 62.2% of ischemic stroke and transient ischemic attacks (IS and TIA), 8.1% of sinus thrombosis (ST) and 29.7% of HI; in 2018, 282 children - 58% IS + TIA, 5.6% ST, 36.4% HI (7% cerebral vessel anomalies without rupture); in 2019, 235 children - 52.7% of IS and TIA, 12.8% of ST, 34.5% of HI (12.8% cerebral vessel anomalies without rupture). Cerebral angiography was performed in 90 patients, of which 45 (50%) patients did not have vascular pathology, 45 (50%) -pathology of cerebral vessels, of which: AVM-in 28 (62.2%) patients, arterial aneurysm - 6 (13.3%), venous angioma i-4 (8.8%), arteriovenous fistula - 3 (6.6%) patients, ST - 2 (4.4%), Moya-Moya-2 (4.4%). Neurosurgical treatment of 14 patients (2017-2018): 11 (78.6%) with arteriovenous malformations, 2 (14.2%) with cavernomas, 1 (7.1%) aneurysm. Age ranged from 7 to 17 years. Ten of them (71.43%) were operated on: microsurgical -5 (50%), endovascular5 (50%). Conclusion: The neurosurgical service is an important part of the work of primary pediatric stroke centers. Improving diagnostic methods has improved the early detection of cerebral vascular anomalies with the optimal treatment tactics. According to the Moscow pediatric stroke Center, annually the number of first diagnosed cerebral vascular anomalies increases by 30-40%, with the predominance of AVM.

P60-J: Brainstem arteriovenous malformation in two adolescent Filipinos

Elbert John Layug[1], Minette Krisel Manalo[1], Jerickson Abbie Flores[2], Marilyn Tan[1]

[1]University of the Philippines - Philippine General Hospital, [2]Jose Reyes Memorial Medical Center

Introduction Brainstem location of arteriovenous malformation occurs in 2-6% of all intracranial AVMs and more commonly presents as hemorrhage in adults with mean age of 25-38.5 years. This is the first local report of two cases presenting during the adolescent period with headaches. Case and Discussion The first case is a 13-year old male who had a one-year history of occipital headache eventually developing left hemiparesis, slurred speech and right-sided ataxia (mRS 3). Brain MRI revealed brainstem AVM extending to the right cerebellum (Figure 1) confirmed by angiography (Figure 2). After instituting medical supportive management, patient was sent home. Gradual improvement of the neurologic deficits (mRS 2) were noted three months after discharge. The second is an 18-year old female taking risperidone for Major Depressive Disorder and Post-Traumatic Stress Disorder who had recurrent episodes of migrainous-like headaches but consulted for galactorrhea. Cranial CT scan showed a midbrain AVM (Figure 3) confirmed by angiography (Figure 4). Endocrine work-up revealed risperidone-induced galactorrhea which resolved after drug withdrawal. Patient was discharged while waiting for definitive management. However, her AVM ruptured four months later manifesting as Weber syndrome (mRS 3). Patient was stabilized and eventually sent home. There is gradual

improvement of the left hemiparesis (mRS 2) five months from the hemorrhagic stroke. Considering the poor prognosis of brainstem hemorrhage, these two cases illustrate the need for high-index of suspicion in seemingly benign headaches as early detection and management of this rare condition could spell the difference in their outcomes. Currently, treatment option for these patients is stereotactic radiosurgery. Keywords: pediatric hemorrhagic stroke, brainstem AVM

P61-J: Multimodal assessment of cerebral hemodynamics after pediatric hemorrhagic stroke secondary to cerebral arteriovenous malformation rupture

Brian Appavu[1], Stephen Foldes[1], Brian Burrows[1], P. David Adelson[1]

[1]Barrow Neurological Institute at Phoenix Children's Hospital

Introduction: Management after hemorrhagic stroke from cerebral arteriovenous malformation (AVM) rupture remains controversial due to strategies to prevent hemorrhagic expansion while maintaining cerebral autoregulation (CA). We investigated associations of hemodynamics on outcomes after pediatric AVM rupture. Hypotheses: Measures of CA, baroreflex sensitivity (BRs) and heart rate variability (HRV) are associated with outcome at 12 months after pediatric cerebral AVM rupture. Methods: Children (<18 years old) with ruptured cerebral AVMs at a single critical care unit over 5 years were retrospectively analyzed. Continuous measurements of data including arterial blood pressure (ABP), intracranial pressure (ICP), cerebral perfusion pressure (CPP) and heart rate (HR) were evaluated. CA measures include pressure reactivity index (PRx), weighted pressure-reactivity index (wPRx), pulse-amplitude index (PAx), and correlation coefficient between ICP pulse amplitude (AMP) and CPP (RAC). Model-based indices of HRV, BRs, CA and estimations of optimal CPP (CPP-Opt) were computed along with percent of time below lower limits of autoregulation (LLA). Outcomes were determined using Pediatric Glasgow Outcome Score - Extended (GOSE-Peds) at 12 months. Patients with scores below 5 were grouped as "unfavorable outcomes". Association of biomarkers with outcome was determined with Wilcoxon singed rank test. Results: 14 children who underwent multimodal monitoring (6 female; mean [IQR] age, 10.57 [1.5]) were included. 8/14 children underwent intraparenchymal ICP monitoring from which CA was measured. Unfavorable outcomes were associated with lower median BRs values as compared to favorable outcomes (p=0.001) and lower HRV as compared to favorable outcome (p=0.026). Favorable outcome was consistent with decreased percent time below LLA based upon PAx and RAC, although this study was not powered for statistical significance (p=0.057). Conclusion: Decreased BRs and HRV were associated with unfavorable outcomes in children after hemorrhagic stroke from AVM rupture. Prospective studies are warranted to understand CA and its impact on outcomes.

P62-J: Etiology of intracerebral hemorrhage in children: cohort study, systematic review & meta-analysis

Gregoire Boulouis[1], Sarah Stricker[1], Sandro Benichi[1], Jean François Hak[1], Manoëlle Kossorotoff[2], Nicolas Garcelon[1], Rosella Mancusi[3], Nathalie Boddaert[1], Thomas Blauwblomme[1], Olivier Naggara[2]

[1]Hopital Necker Enfant Malades, [2]Univ Hospital Necker-Enfants malades APHP, [3]GHU – Paris

Introduction Understanding non-traumatic pediatric intracerebral hemorrhage (pICH) etiological spectrum is key to diagnostic work-up and care pathway orientation. We aimed to evaluate the etiologic spectrum of diseases underlying pICH. Patients and Methods Children treated at our institution for a pICH were included

in an inception cohort initiated in 2008, retrospectively inclusive to 2000 and analyzed in October 2019. We then conducted a systematic review of relevant articles published between 1990 and 2019, identifying cohorts with pICH patient. Identified populations, and patients from our cohort were pooled in multi category diagnosis meta-analysis. Results A total of 243 children with pICH were analyzed in the cohort study. The final primary diagnosis was an intracranial vascular lesion in 190 patients (78.2%), a complication of a cardiac disease in 17 (7.0%), a coagulation disorder in 14 (5.8%). Hematologic and cardiologic etiologies were disproportionately more frequent in children younger than 2 years old (p<0.001). The systematic review identified 1309 children in 23 relevant records, pooled in meta-analysis. Overall there was important heterogeneity. The dominant etiology were vascular lesions, with an aggregate prevalence of 0.59 [95% confidence interval: 0.45-0.64, p=0,001 Q:302.8, I^2: 92%]. In eighteen studies reporting detailed etiological spectrum, arterio-venous malformation represented the dominant etiology (68.3% of all vascular causes, [95%CI 64.2 - 70.9%), followed by cavernoma (15.7% [95%CI 13.0 - 18.2%]). Conclusion The most frequent etiology of pICH are brain arteriovenous malformations. The probability of an underlying vascular etiology increases with age, and conversely hematologic and cardiac causes are dominant causes in children younger than 2 years old.

P63-J: Mortality and functional outcome after pediatric intracerebral hemorrhage: a cohort study, systematic review and meta-analysis

Gregoire Boulouis[1], Sarah Stricker[1], Sandro Benichi[1], Jean François Hak[1], Manoëlle Kossorotoff[2], Nicolas Garcelon[1], Rossela Mancusi[3], Nathalie Boddaert[1], Thomas Blauwblomme[1], Olivier Naggara[2]

[1]Hopital Necker Enfant Malades, [2]Univ Hospital Necker-Enfants malades APHP, [3]GHU Paris

Introduction The clinical outcome of pediatric intracerebral hemorrhage (pICH) is scantly reported in a comprehensive way. In this cohort study of patients with pICH, systematic review and meta-analysis, we aimed at describing the basic clinical outcomes after pICH. Methods Children treated at our institution for a pICH were included in an inception cohort initiated in 2008, retrospectively inclusive to 2000 and analyzed in October 2019. We then conducted a systematic review of relevant articles published since 1990, Identified populations, and patients from our cohort were pooled in multi category diagnosis meta-analysis, and analyzed with regards to clinical outcomes. Results Amongst 243 children screened for inclusion, 231 patients were included and analysed. Median age at ictus was 9.6 years [interquartile range: 4.6 -12.5], and 128 (53%) patients were male. After a median of 33 months of follow-up (interquartile range 13-63), 132 patients (57.4%) had a favourable clinical outcome, of which 58 (44%) had no residual symptom. Amongst 2622 records screened, we retained 24 studies with available data for the final analyses. Overall, the aggregate proportion of children with complete recovery was 26% [95% confidence interval: 16.4-34.2%, Q:170, I^2: 89%], and of those with residual deficits 52.7% [95% confidence interval: 39.7-60.3%, Q:170, I^2: 89%]. When pooled with our cohort, the aggregate case fatality at last follow up was 15.3% [95% confidence interval: 11.2-19.9%, Q:191, I^2: 87%]. Conclusion Here we showed that mortality affects 1 in 6 children after pICH, and that the majority of children have residual neurological deficits at latest follow up. Complementary data from the cohort study are to be presented.

P64-J: Hemorrhage expansion after pediatric intracerebral hemorrhage

Jean François Hak[1], Baslie Kerleroux[1], Sandro Benichi[1], Sarah Stricker[1], Philippe Meyer[1], Manoëlle Kossorotoff[2], Nathalie Boddaert[1], Andrea Morotti[3], Thomas Blauwblomme[1], Olivier Naggara[2]

[1]Hopital Necker Enfant Malades, [2]Univ Hospital Necker-Enfants malades APHP, [3]IRCCS Mondino

Introduction Hemorrhage expansion (HE) is a known predictor of clinical deterioration, and poor clinical outcome following admission after an intracerebral hemorrhage (ICH) in adults, but remains poorly reported in children. In a large inception cohort of children with ICH, we aimed to explore the prevalence of HE, its associations with clinical outcomes and its clinical-imaging predictors. Patients and Methods The report is based on an inception registry of children presenting with an ICH at a single quaternary university pediatric hospital (Necker Enfants Malades, Paris, France), retrospectively inclusive to January 2000. The study sample was restricted to children with two CT scans within 72 hours of symptoms onset available for re-asessement, and a clinical follow-up duration of more than 12 months. Hemorrhage expansion was defined as being clinically significant if the difference between follow-up and baseline ICH volumes exceeded 12.5cc or 33% of the baseline volume. Results Fifty two children met inclusion criteria, amongst which 8 (15%) demonstrated hemorrhage expansion. Children with hemorrhage expansion had more frequent coagulation disorders (25,0% vs 2,3%, p=0,022), focal deficits at presentation (75,0% vs 43,2%, p=0,08) and infra-tentorial ICH (25,0% vs 9,1%, p=0,01). After adjustment for variables associated with HE in univariable analyses at a p<0.1, only the presence of coagulation disorders at baseline remained independently associated with HE (adjusted OR 14,4 95%CI [1,04-217], p=0,048). HE was independently associated with poor (KOSCHI < 5A, adjusted OR 5,77 95%CI [1,01-38,95], p=0,043) as well as severe (KOSCHI < 4A, aOR 20,22 95%CI [3,06-133,68], p= 0,002) clinical outcomes Conclusion Hemorrhage expansion frequency is unexpectedly high after a pediatric ICH, affecting over 15% of children in our sample, and was found to be strongly associated with poorer clinical outcome, regardless of its definition.

P65-J: Incidence rates and predictors of seizures in children with familial cerebral cavernous malformations

Christine Fox[1]

[1]UCSF

Introduction: Familial cerebral cavernous malformation (FCCM), an autosomal dominant disease caused by mutations in CCM1, CCM2 or CCM3, often presents with childhood seizure. We sought to determine seizure incidence rate and factors associated with seizures in FCCM. Methods: Seizure data were collected from FCCM cases enrolled in the Brain Vascular Malformation Consortium. We estimated the incidence rate of childhood (<19 years) seizure onset using interval-censored survival analysis. We tested whether CCM genotype and large (>5 mm) brain lesion count on imaging at enrollment were associated with earlier onset of seizures. Results: The study included 479 FCCM cases (61% female), of whom 188 (39%) had a seizure prior to enrollment. Median age at enrollment was 42 years (IQR: 22-55) and 102 (21% of the cohort) were <19 years old. Among 393 patients with genetic testing, 88% had the Common Hispanic Mutation in CCM1, 5% had a different CCM1 mutation, 5% had CCM2 mutations, and 2% had CCM3 mutations. Median large lesion count was 3 (IQR: 1-5). Individuals with higher than expected large lesion counts for their age and sex were at increased risk of first seizure (HR=1.4 per SD unit increase, p=0.001). Those with a CCM3 mutation were also at increased seizure risk compared to others (HR=3.1, p=0.026). The incidence rate of a seizure in childhood was 21.2% (95% CI: 17.9-24.3). The 43 children with seizure history at enrollment reported a median frequency of 1 seizure in the prior year (IQR: 0-1, with 2 patients reporting >100). Eleven with prior seizures were not on anticonvulsant medication. Of the 20 children who reported an anticonvulsant medication, four reported poor seizure control. Conclusions: In the largest study of FCCM to date, children had a high prevalence of seizures and medically refractory epilepsy. Those with higher large lesion counts or CCM3 genotype had a higher seizure risk.

K - Venous sinus thrombosis

P66-K: Risk factors for pediatric cerebral sinus vein thrombosis

Marcela Torres, MD[1], Tyler Hamby, PhD[1], Sarah Philip, MS[2], Jo Ann Tilley, DNP, APRN, CPNP[1]

[1]Cook Children's Medical Center, [2]Texas College of Osteopathic Medicine, UNT Health Science Center

Intro Cerebral sinus vein thrombosis (CSVT) is a type of stroke consisting in thrombosis of the dural sinus and/or cerebral veins. The estimated incidence in children is 0.6 per 100,000 children per year however the symptoms and presentations are variable among all age ranges raising the concern for missed diagnosis. Despite new progress on pediatric stroke, CSVT is under-recognized and its risk factors are not well established. Hypothesis To describe risk factors of pediatric CSVT together with its signs and symptoms at presentation. Methods A single-center review of pediatric patients (<18 years) with radiographically confirmed CSVT between 2008 and 2018, seen by the Hematology service at Cook Children's Medical Center were identified through the Cook Children's database. Results There were 87 patients identified. Symptoms at diagnosis were nausea/vomiting (33%), headache (32%), fever (29%) and neurologic deficits (25%). The most common neurologic deficits reported were altered mental status (9%) and increased intracranial pressure (9%) followed by focal weakness/hemiparesis (5%), cranial nerve palsy (5%), diplopia (4%), papilledema (4%) and ataxia (2%) (Table 1). Comorbidities included mastoiditis (26%), otitis media (24%), Lemierre's syndrome (10%), retropharyngeal abscess (5%) and severe anemia (hemoglobin < 7 g/dl; 5%) (Table 2). Results of thrombophilia work up and sinuses involved are included in Figure 1 and Figure 2, respectively. Conclusions Infectious causes such as otitis media, mastoiditis and Lemierre's syndrome can be common contributors of pediatric CSVT. Anemia is a little known independent risk factor for thrombosis, especially CSVT, and presented in 5% of our patients. Not all patients with CVST underwent a complete thrombophilia evaluation, but thrombophilia was still rare finding. We suggest that if these infections are diagnosed, there should be a high index of suspicion for CSVT even in the absence of neurological deficits/symptoms.

[PRINT ONLY] P67-K: Unusual presentation of CSVT in children - case studies

Vesna Brankovic-Sreckovic[1], Ivan Vukasinovic[2], Galina Stevanovic[1], Dusan Damjanovic[2]

[1]Clinic for Child Neurology and Psychiatry, [2]Clinical Center Serbia

Introduction: Cerebral SVT, a potentially treatable disorder, is often overlooked or misdiagnosed as its symptoms and signs can be unspecific. Therefore, appropriate diagnostic and etiologic works up are necessary in suspicious cases. Case Studies: Case#1 is a 15 yr old- girl who presented with unilateral posterior headache, visual impairment, chronic papilledema and signs of ICH, after a history of chronic headaches in the last 2 years. The investigations revealed stasis of optic disc of 4dpt bilaterally, without significantly raised open pressure on LP and normal CSF profile. DSAV and MRI with MRV showed multiple obstructions of venous structures on the left side, both superficial and deep venous structures, giving the appearance of SAH in the left temporal region. The treatment with acetazolamide and anticoagulation therapy lead to lowering of ICP, partial recanalization of venous occlusion and subsequent secondary optic atrophy. The history of persistently elevated platelet count during childhood and follow-up, lead to a diagnosis of essential thrombocytosis. Case#2: is a 15 yr-old-girl who presented with eclampsia and epileptic seizure in

37GW of pregnancy. 8h after cesarean section, she developed signs of raised ICP, altered consciousness and life-threatening condition. CTV revealed SSS thrombosis, ICH in the frontal parasagittal regions and subsequent SAH. The patient was treated rigorously with antiedematous and anticoagulation therapy, which lead to clinical and neuroradiological improvement and favourable outcome without neurological sequelae. Laboratory investigation was unspecific, except homozygous state for PAI-1 and MTHFR mutation. Discussion: Those two cases indicate that the underlying cause of CSVT can predict initial and chronic neurological symptoms. Cryptogenic and chronic CSVT in pt#1 lead to the underlying diagnosis of essential thrombocytosis. Acute CSVT in pt#2 after delivery is due to a hypercoagulable state and/or hypovolemia. Both patients received prolonged therapeutic anticoagulation (OAT) and no recurrence after 6m/12m follow-up.

P68-K: Cerebral venous sinus thrombosis in infants after surgery for congenital heart disease

Dana Harrar[1], Margaret Goss[2], Mary Donofrio[3], Jonathan Murnick[3], Yaser Diab[3], Jennifer Meldau[3], Mark Connelly[3], Anqing Zhang[4], Pranava Sinha[3], Can Yerebakan[3], Jessica Carpenter[3]

[1]Children's National Medical Center, [2]Dell Children's Hospital, [3]Children's National Hospital; George Washington University, [4]Children's National Hospital

Introduction: Children with congenital heart disease (CHD) may be at increased risk for cerebral sinus venous thrombosis (CSVT). The incidence of CSVT in neonates undergoing repair of congenital heart disease has been reported to be as high as 28%. Hypothesis: We hypothesize that the incidence of CSVT in this population is lower than previously described. Methods: Neonates who had CHD repair with cardiopulmonary bypass (CPB) in the first 30 days of life between 2013 and 2019 were identified from a prospective cardiac surgery database. Patients who had a post-operative brain MRI were identified from a prospective institutional database. Demographic, clinical, and surgical data were obtained from the cardiac surgery database and the medical record. Imaging was reviewed by a pediatric neuroradiologist. Results: One hundred eighty-four neonates who had repair of CHD using CPB between 2013 and 2019 had a post-operative brain MRI. CSVT was identified in 4.3% (n=8) and only in those with transposition of the great arteries with an intact ventricular septum (TGA-IVS), interrupted aortic arch (IAA), or coarctation of the aorta (CoA). TGA-IVS (p=0.007) and IAA (p=0.023) were associated with an increased risk for CSVT. Other risk factors for CSVT included longer cross-clamp time (98 minutes [interquartile range (IQR) 77.5-120] v. 67 minutes [IQR 44-102], p=0.03) and shorter time between surgery and MRI (10 days [IQR 7-12.5] v. 20 days [IQR 12-35], p=0.004). CSVT involved the transverse sinus in all patients. Five patients (62.5%) were treated with anticoagulation; four (80%) had complete resolution of their thrombus and one (20%) had partial resolution. Three patients were not treated, all of whom had at least partial resolution of their thrombus. No patient had complications of their thrombus or of anticoagulation. Conclusion: We identified a low incidence (4.3%) of CSVT in neonates with CHD undergoing surgery with CPB in the first 30 days of life. All CSVTs were asymptomatic and discovered on routine post-operative imaging. CSVT was found only in patients with TGA, IAA, or CoA and was associated with a shorter time between surgery and imaging. At least partial recanalization was universal, regardless of anticoagulation. Further studies are needed to establish best practices for surveillance, prevention, and treatment of CSVT in neonates undergoing CPB for repair of CHD.

P69-K: Successful treatment of pediatric cerebral venous thrombosis stroke with Bivalirudin

Oluwaseun Olaiya[1], Sana Farooki[2], Mukta Sharma[3]

[1]Children's Mercy, Univ of Missouri Kansas City School of Medicine, [2]Children's Mercy, [3]Children's Mercy, University of Missouri Kansas City

Introduction Cerebral venous thrombosis (CVT) is an uncommon cause of stroke in children, presentation is variable but can include seizures and other neurologic symptoms. Anticoagulation is the cornerstone of treatment and endovascular interventions can also be used for management. Bivalirudin is a direct thrombin inhibitor used for the treatment of thrombosis in children especially as an alternative to heparin in heparin induced thrombocytopenia (HIT). It has not been well studied in children with CVT. Objectives: To report a case of successful treatment of CVT stroke in a pediatric patient with bivalirudin Case: 16-year-old female with polycystic ovarian syndrome on oral contraceptives as well as obesity with BMI of 43 who presented to the pediatric intensive care unit as a transfer from a local hospital. She presented there with altered mental status, seizures and acute respiratory failure. She arrived at our ICU intubated and sedated. MRI/MRA Brain/Neck were obtained that showed complete thrombosis of the left transverse sinus, sigmoid sinus and superior internal jugular vein down to the C1 level with partial thrombosis extending down to the C1-C2 level. She was started on bivalirudin after discussion with the Neurology, coagulation and neuro radiology teams. Neurologic exam after extubation was within normal limits. She continued bivalirudin for 6 days and was transitioned to rivaroxaban at discharge. She had no adverse effects while on anticoagulation. Thrombophilia work up revealed she is heterozygous for factor V Leiden. Discussion This case highlights the successful use of bivalirudin for treatment of CVT in a pediatric patient. In addition to its use for treatment of HIT, bivalirudin has been used in pediatric patients on extracorporeal membrane oxygenation (ECMO). However, this is the first documented case report of treatment of a pediatric patient with CVT with bivalirudin. Further prospective studies are needed to clarify its utilization in pediatrics.

P70-K: Apixiban therapy of adolescent cerebral sinus venous thrombosis

Jenny Wilson[1], Michael Recht[1], Trisha Wong[1]

[1]Oregon Health and Science University

Introduction: Direct oral anticoagulants (DOACs) are becoming an acceptable alternative to vitamin K oral antagonists and heparin derivatives in the treatment of thrombosis in adults. However, the recommended treatment for cerebral sinus venous thrombosis (CVT) remains coumadin or low molecular-weight heparin.1 Though there are no reports of DOAC use in pediatric CVT, they are appealing as drug levels do not have to be followed, food and medication interactions are minimal, and they are taken orally. We report two adolescents with CVT treated with apixaban. Case 1: A 16 year-old girl presented with three days of headache after an increase in estrogen content of oral contraceptive pills. Her head CT and CT angiogram were consistent with CVT of the majority of the superior sagittal sinus without venous infarct. She was started on enoxaparin before transitioning to apixaban 5 mg twice daily the next day. She tolerated the medication well. She had resolution of her CVT on MRA after 3 months, so her apixaban was discontinued. Case 2: An 18 year-old man with trisomy 21 and acute lymphocytic leukemia presented with headache and left-sided weakness 3 weeks after his 2nd PEG-asparaginase dose. Imaging showed a right parenchymal hemorrhage CVT of the left sigmoid, transverse and superior sagittal sinuses. He was initially treated with a heparin drip for 24 hours, then switched to apixaban 10 mg twice daily for 6 days, then 5 mg twice daily, which he has tolerated well. There was no progression of CVT after 6 months of treatment. Due to lack of recanalization and need for further PEG-aspariginase, he will remain on apixaban for one year. Discussion: We report two adolescents with CVT treated with apixaban, one with recanalization and the other with symptoms resolution

and lack of clot progression. Apixaban was well-tolerated without bleeding. Prospective data are needed comparing heparin-based and oral vitamin K antagonist treatments to DOACs in this population. 1. Ferro JM, Bousser MG, Canhao P, et al. European Stroke Organization guideline for the diagnosis and treatment of cerebral venous thrombosis - endorsed by the European Academy of Neurology. European journal of neurology. 2017;24(10):1203-1213.

P71-K: Cohort of patients with cerebral venous thrombosis in neonates and children: treatment and follow-up of 14 patients

Maria Fernanda Rios Pistoia[1], Guillermo Agosta[1], Clarisa Maxit[1]

[1]Hospital Italiano de Buenos Aires

Introduction: Cerebral venous thrombosis is a rare cause of stroke in the neonatal and paediatric population. In the last 2 decades there has been significant growth in the study and diagnosis of this rare disease in pediatrics. We are interested in sharing our experience in the management of this pathology in order to contribute to the evidence of the effectiveness and safety of the treatment. Objective: to share our experience in the management of cerebral venous thrombosis in pediatrics. Method: A descriptive longitudinal, prospective and retrospective study was conducted from January 2015 to DEcember 2020 evaluating patients with cerebrovascular disease. Of 88 follow-up patients with cerebrovascular disease 14 patients had cerebral venous thrombosis. Children from 0 days of life to 18 years of life were included who had acute neurological signs (altered consciousness, headache, seizures or neurological focal signs) with radiological confirmation with resonance or tomography venography, where thrombus or interruption of venous flow was evident, or not by associating infarction or bleeding. Results: 14 patients were evaluated, 9 of whom were evaluated from baseline and 3 were evaluated after the acute period. The majority were male(8/ 61.5%). More than 50% occurred in the neonatal period. Of the predisposing factors found: 2 patients associated with head and neck infections; 1 meningitis; 1 sepsis. 6 had heart disease, 1 ischemic hypoxic encephalopathy; 1 required respiratory ECMO. Most common clinical presentation were: impaired consciousness, headache, seizures, apnea. One patient was detected by prenatal eco Received treatment. 11/13. None had post-anticoagulant bleeding. 8 patients had treatment during 6 weeks or 3 month according to age; 2 followed was lost. 3 to die before assessing whether they managed to resolve thrombosis.; 1 required prolonging the anticoagulation time according to their age. Conclusions: so far there are few series of patients published with this pathology. It is important to communicate the experience in order to obtain strong information about the efficacy and safety of the treatment.