

# IPSO 2024 Congress Abstract Proceedings

**IPSO 2024 Congress**  
**March 22-24<sup>th</sup>, 2024**  
**Toronto, Canada**  
*Inspiring Innovation, Transforming Care and Improving  
Outcomes in Pediatric Stroke*



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## Acute Treatment of Arterial Ischemic Stroke

### **P-A-1: Atorvastatin therapy in a juvenile photothrombotic stroke model is associated with increased ADC, reduced KPS, and decreased stroke lesion volume: an MRI study [Saturday Poster Session, Event Room 2]**

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**Background:** Atorvastatin is a potential candidate for treating AIS because it improves endothelial function, modulates thrombogenesis, and attenuates inflammatory and oxidative stress damage.

**Objective:** To assess acute ischemic stroke (AIS) and blood-brain barrier (BBB) progression longitudinally following the administration of atorvastatin in a juvenile photothrombotic rat stroke model with quantitative MRI.

**Methods:** A photothrombotic ring model was used to induce stroke in ten five-week-old male Sprague Dawley rats. The treatment group of 11 rats received peritoneal injection of atorvastatin (20 mg/kg), and 11 rats served as a control group. The 3T MRI protocol included T2 FLAIR, DWI, and DCE imaging. ADC, BBB leakage and lesion volume were quantified at the hyperacute (Day 0), sub-acute (Day 2), and chronic stages (Day 7) following photothrombosis. Student's t-tests were used to measure differences between groups.

**Results:** The photothrombotic ring model caused reproducible infarction sizes with delineated boundaries for precise stroke lesion characterization. The treatment group had increased ADC in the lesion on day 2 ( $p = 0.0269$ ). Reduced BBB leakage in both groups occurred on day 2 with no significant differences. Lesion volumes were smaller on days 2 and 7 ( $p=0.0155$  and  $p=0.0150$ ) in the statin group.

**Conclusions:** Our study shows the effectiveness of atorvastatin in a juvenile photothrombotic rat model of stroke. Increased ADC values and lower lesion volumes in the treatment group suggest decreased tissue damage. Further research is needed to investigate its efficacy in ameliorating BBB damage.

### **P-A-2: Broadened Therapeutic Window of Lower Profile Mechanical Thrombectomy Devices [Saturday Poster Session, Event Room 2]**

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**Background:** Endovascular mechanical thrombectomy (MT) is well established as a treatment modality for acute ischemic stroke (AIS) with large vessel occlusion in adults, but is only evaluated in children in retrospective series. There is not a feasible prospective trial design to capture the diverse indications in pediatric AIS. Novel devices are brought to market as equivalent to pre-existing devices that were studied in randomized controlled trials. In pediatric cases of AIS, some newer devices may be appropriate for off-label use in revascularization procedures.

**Methods:** A single institution experience in intracranial MT procedures for pediatric AIS. The procedure was included if there was an attempt at MT via a non-pharmacologic approach.

**Results:** Seventeen endovascular procedures were performed over 2015-2023 for AIS; with a mean age of 8.5 years. MT was attempted in 16 (94.1%) cases, including 12 (75%) in the anterior circulation and 4 (25%) in the posterior circulation. Most common MT techniques were aspiration with stentriever 10 (62.5%), aspiration alone 5 (31.3%), and stentriever alone 1 (6.2%). Traditional stentrievors (4-6 mm) were used in 7 cases and low-profile stentrievors in 4 cases. TICl grade 3 was achieved in 4 (25%), 2A in 3 (18.8%), 2B in 7 (43.7%), and 0 in 2 (12.5%).

**Conclusions:** The performance of thrombectomy devices with lower profile 0.165-catheter-compatible designs may achieve similar revascularization results in challenging clinical scenarios observed in children.

### **P-A-3: Bilateral mechanical thrombectomy in a child with hypoplastic left heart syndrome and protein-losing enteropathy [Saturday Poster Session, Event Room 2]**

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Arterial ischemic stroke (AIS) is a significant cause of morbidity in children with congenital heart disease, with an estimated risk of 132 in 100,000. Children with hypoplastic left heart syndrome (HLHS) are at especially high risk, and children with Fontan physiology are at risk of developing protein-losing enteropathy (PLE), which increases risk of thromboembolism. Case: A 9-year-old female with HLHS variant: double outlet right ventricle, status-post fenestrated Fontan 6 years earlier, presented with left hemiplegia, right homonymous hemianopsia, and expressive aphasia, pediatric NIHSS=15. MRI demonstrated restricted diffusion in the bilateral caudate bodies and periventricular white matter. MRA demonstrated occlusions of the right cervical ICA and the left MCA at the M1-M2 junction. She was last seen well 15 hours prior, but MRI lacked T2/FLAIR changes, thus she underwent emergent mechanical thrombectomy, with TIC1 2b (6 passes), 2c (3 passes) of the right ICA, left MCA. Repeat MRI demonstrated restricted diffusion in ACA-MCA watershed and MCA territories despite successful thrombectomy. She was taking aspirin 81 mg daily, and thus was started on heparin infusion, which was transitioned to enoxaparin. She had been previously diagnosed with PLE. Gastrointestinal loss of anti-thrombotic proteins may have caused her hypercoagulable state. At 2.5-month follow-up, NIHSS=7 and Pediatric Stroke Outcome Measure=6 with bilateral sensorimotor dysfunction, expressive aphasia, and cognitive issues. Discussion HLHS confers a high risk of AIS. Fontan physiology carries a 4% risk of developing PLE, and gastrointestinal loss of anti-thrombotic proteins increase risk of AIS. Our patient developed thrombi within the bilateral anterior circulation requiring double thrombectomy, an exceptionally rare intervention in children that may improve outcome. With Fontan physiology, PLE is a strong risk factor for AIS, with implications for primary and secondary stroke prevention.

### **P-A-4: Pediatric Large Vessel Occlusion in Setting of Endocarditis [Saturday Poster Session, Event Room 2]**

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**INTRODUCTION:** Pediatric acute ischemic stroke (AIS) can be very difficult to recognize given transient improvement of symptoms, and incidence of stroke mimics, particularly in the setting of the fever.<sup>1</sup> Pediatric patients with cardiac disease have a higher incidence of cardioembolic events than the general population.

**CASE SUMMARY:** This case report examines a 12-year-old male with a medical history of a bicuspid aortic valve, who presented as a transfer from an outlying facility with acute onset right-sided weakness and expressive aphasia due to large-vessel occlusion (LVO) acute ischemic stroke (AIS) and fever. The patient had a late presentation due to transient improvement in symptoms, which delayed arrival to the emergency department. On presentation, perfusion imaging demonstrated a significant increased time to peak with corresponding decreased in cerebral blood flow and volume in the proximal superior M2 segment of the left middle cerebral artery (L-MCA). Thrombolytics were not administered given unclear last known well time and mechanical thrombectomy (MT) was not performed given large volume core on perfusion imaging. MRI-Brain two days after admission demonstrated a large area of restricted diffusion in the L-MCA distribution in addition to punctuate changes in the left basal ganglia and bilateral cerebral hemispheres. The patient underwent secondary stroke up including trans-thoracic echocardiogram, which demonstrated a large vegetation (~10mm) on the bicuspid, stenotic aortic valve. Anti-platelet (AP) and anticoagulation (AC) therapy were held acutely. 1 week later, PCR's returned positive for B.Henselae requiring 6 weeks of antibiotics, delaying start of AP therapy prior to transition to AC in preparation for valve replacement surgery.

**DISCUSSION:** There are multiple risk factors in the pediatric population that are associated with higher incidence of AIS, particularly cardiac disease.<sup>3,4</sup> More research is needed to better identify cardiogenic strokes in children as well as more standardized guidelines for pediatric patients with cardiac disease.

## **P-A-5: Acute Treatment of Arterial Ischemic Stroke in Liberia: Current and Future Perspectives - A Case Study [Print Only]**

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**Abstract:** This case study explores the acute treatment of arterial ischemic stroke in Liberia, assessing current practices, challenges, and potential future perspectives. The objective is to provide insights into the existing scenario, identify areas of improvement, and suggest strategies to enhance stroke care in this resource-constrained setting. Stroke, a global health concern, poses unique challenges in Liberia due to limited resources, infrastructure, and healthcare personnel. A representative case study illustrates obstacles faced in the acute management of stroke, highlighting the need for a multi-faceted approach. The discussion addresses challenges and proposes solutions, emphasizing public education, improved healthcare infrastructure, and international collaboration for comprehensive and sustainable stroke care.

**Introduction:** Stroke is a major cause of morbidity and mortality worldwide, posing a significant public health concern in Liberia. Effective acute treatment of arterial ischemic stroke is crucial to minimize neurological damage and improve patient outcomes. However, the healthcare landscape in Liberia presents unique challenges, including limited resources, infrastructure, and healthcare personnel. This study aims to evaluate the current state of acute stroke treatment in Liberia, shedding light on both successes and shortcomings.

**Case:** A 56-year-old male patient admitted to a regional hospital in Liberia with acute right-sided weakness and slurred speech serves as a representative example. Prompt clinical assessment and brain imaging confirmed arterial ischemic stroke. Challenges included limited availability of thrombolytic agents and advanced imaging technologies, hindering timely treatment administration.

**Discussion:** The acute treatment of arterial ischemic stroke in Liberia faces challenges primarily due to resource constraints. Limited access to thrombolytic therapy and the absence of specialized stroke units hinder.

## **P-A-6: [Print Only]**

### **Development Of A Framework For Acute Pediatric Stroke Management Across Kentucky**

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Pediatric stroke is an uncommon condition that may be associated with serious morbidity and mortality. There is limited literature on the development or implementation of a multi-institutional or regional framework along these lines. The use of a novel regional treatment algorithm for children presenting with acute neurologic deficits concerning for stroke will prevent delays in care and provide a stepwise approach for managing a pediatric stroke alert across multiple facilities in the state of Kentucky. A workgroup comprising pediatric stroke experts from the University of Kentucky (UK) and the University of Louisville (UL) developed a clinical decision-making tool (algorithm) to help guide frontline and community providers throughout the state of Kentucky (USA) when presented with a pediatric stroke patient. The defining feature of the algorithm is the contact information for both UK and UL for respective regional providers to speak directly to a pediatric stroke expert at a tertiary care facility to discuss use of the algorithm and guide management. We recently began statewide distribution of the algorithm through various entities to reach as many institutions and providers in the state as possible. We plan to track calls received to either UK or UL over the next 12 months regarding possible acute stroke patients to assess utilization of the tool by outside providers. Because of this, results collection and analysis is pending. We present the development of a novel statewide treatment algorithm for children presenting with acute neurologic deficits concerning for stroke to reduce delays in care and improve the care of pediatric stroke patients in Kentucky.

## Moyamoya

### **O-B-1: Multi-omics characterization of immune triggers in Moyamoya disease [Oral Abstract Session]**

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Moyamoya disease (MMD) is a rare cerebrovascular disorder with unknown etiology, characterized by progressive narrowing of cerebral arteries and formation of a compensatory network of fragile vessels. Genetic studies have identified RNF213 (as a susceptibility gene for MMD, but the low penetrance in genetically susceptible individuals suggests that a second hit is necessary to trigger MMD onset. Recently, molecular studies uncovered RNF213 as a key antimicrobial protein with important functions in the immune system. In addition, an increasing number of clinical reports describe the development of moyamoya angiopathy associated with infection or autoimmune disorders. Together, this growing body of molecular and clinical evidence points towards immune-related responses as second hits to trigger MMD onset. To address this hypothesis, we have collected plasma, peripheral blood mononuclear cells and skin fibroblasts from 12 MMD patients followed-up at the Ghent University Hospital. Whole exome sequencing revealed patients with known and novel polymorphisms in RNF213 as well as other genes linked to MMD. We are now exposing patient samples to relevant infectious and immune stimuli, followed by multi-omics analysis combining state-of-the-art transcriptomics, proteomics and tracer metabolomics screens, to see if these cells respond differently compared to healthy cells. In addition, we collected MMD brain autopsy material and are currently performing spatial ultrahigh-content imaging to evaluate the presence of immune cells in MMD arteries. These experiments will allow to profile MMD mutations in a (non-Asian) MMD cohort and to map altered pathways in patient-derived cells, with the aim to elucidate MMD-related immune responses and to indicate whether MMD could originate from potential defects in the immune response.

### **P-B-1: Lower Collateral Score using Cerebral Catheter Angiography in Posterior Cerebral Artery Associated with White Matter Alterations in Watershed White Matter Tract in Children with Moyamoya [Saturday Poster Session, Event Room 1]**

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**Introduction:** Children with moyamoya without stroke or silent infarct have alterations in their white matter in combined white matter watershed tract measured by mean diffusivity (MD) using diffusion magnetic resonance imaging (dMRI). In adults with moyamoya these white matter alterations have been correlated with decreased perfusion.

**Hypothesis:** Children with moyamoya without stroke or silent infarct will have alterations in their combined white matter watershed tract that correlate with their cerebral catheter angiography collateral grading scale score.

**Methods:** Children with moyamoya without stroke or silent infarct from a single institution were included if they had dMRI and cerebral catheter angiography prior to revascularization surgery. Children who could have altered white matter for other reasons were excluded including NF1, sickle cell anemia and history of cranial radiation. dMRI was analyzed and the combined white matter watershed tract was extracted. A blinded neurointerventional radiologist reviewed and graded each cerebral catheter angiogram using previously published 4-point collateral grading scale. ANOVA was used to compare dMRI parameters with collateral grading scale.

**Results:** Twenty children with moyamoya without stroke or silent infarct underwent both dMRI and cerebral catheter angiogram prior to revascularization surgery. Higher mean diffusivity (MD) in the combined white matter watershed tract correlated with worse score on the collateral grading scale in the posterior cerebral artery territory ( $p=0.01$ ).

**Conclusion:** Lower posterior cerebral artery collateral score was associated with higher white matter alterations measured by higher MD in the combined white matter watershed tract in children with moyamoya without stroke or



silent infarct. These findings reinforce previous findings that children with moyamoya may have worse outcomes when there is posterior cerebral artery involvement.

### **P-B-3: Nit2/ $\omega$ -amidase as a Potential Non-Invasive Diagnostic Marker for Moyamoya Disease [Saturday Poster Session, Event Room 1]**

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**Introduction:** Moyamoya disease (MM) is a progressive cerebrovascular disease that causes stroke and can be treated with surgical revascularization. The enzyme Nit2/ $\omega$ -amidase (NIT2) is involved in glutaminase II and methionine salvage pathways. Recent studies have shown correlations between these metabolic pathways and MMD pathogenesis, specifically increases in L-methionine, NO metabolites, and homocysteine. Here, we present data demonstrating that a NIT2 may have potential utility as a non-invasive biomarker able to identify the presence of MM.

**Hypothesis:** A comprehensive protein sequencing analysis of pediatric MM patients will reveal novel protein expression as potential non-invasive diagnostic biomarkers for MM.

**Methods:** Urine (n=42) and blood plasma (n=30) samples were collected from MM patients (age=8 mo. - 19yo) who were undergoing surgical revascularization. Control samples were collected from patients (age=1-19yo) with filar lipoma or Chiari malformations (n=24). All samples were collected with patient/family consent and institutional review board approval. The samples were then analyzed using Olink Explore 3072 proteomic proximity extension assays (PEA) to determine expression levels across ~ 3000 validated protein assays. The results were normalized, validated, and analyzed for potential age and sex bias.

**Results:** ANOVA analysis of the MM urine and plasma samples showed a statistically significant increase in Nit2/ $\omega$ -amidase (NIT2) compared to the control samples, with MM patients exhibiting >2-fold increases in plasma expression ( $p=0.0037$ ) and >11-fold increases in urinary expression ( $p=0.00019$ ).

**Conclusions:** NIT2 is involved with pathways implicated in MMD pathogenesis, and our data reveals that plasma and urinary levels of NIT2 are significantly elevated in MM patients compared to matched controls, suggesting that NIT2 has potential as a putative biomarker for MM.

### **P-B-4: Nit2/ $\omega$ -amidase Moyamoya Genetic Testing in the Clinic: What do we really know about RNF213? [Saturday Poster Session, Event Room 1]**

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**Introduction:** Moyamoya disease (MMD) is a rare, progressive intracranial vasculopathy that may occur in Sickle Cell disease, post chemo-radiation, or Trisomy 21. However, nearly 50% of cases are considered idiopathic (iMMD). One possible cause is genetic, and over the past decade, RNF213 has emerged a risk allele, particularly in patients of East Asian descent. Genetic testing for MMD has increased, but incomplete penetrance, variants of unclear significance and a lack of progress on pathogenic mechanisms for RNF213 make it hard to decide how to utilize genetics.

**Case:** In the past three years, our clinic has established a routine testing paradigm for all patients with iMMD. During this time, six patients have undergone targeted gene testing for iMMD and 2 have found variants of unclear significance within RNF213 gene. Patient #1 was of East Asian descent and had a father who was also found to have iMMD after his son was diagnosed. They both tested positive for well-known p.R4810K founder mutation in RNF213. Given family

history, two siblings also had testing to screen for the variant and although one carried it, his MRA was normal, prompting questions about how to screen him properly going forward. Patient #2 had no family history but was found to have a De Novo VUS at p.P5097L in RNF213. With no prior reports of this variant associated with iIMD, the test result remains inconclusive and has not prompted additional genetic testing. Of note, both children underwent bilateral synangiosis and while their symptoms improved, they both continued to have episodes of exercise-induced headache or focal weakness.

**Discussion:** The cases highlight the complexity of interpreting RNF213 results and providing appropriate family screening. To provide high quality care through the inevitable expansion of genetic testing for iIMD, inclusion of genetic counseling and appropriate follow up will be important.

## Neurocritical Care

### **P-C-1: Recognition of Intracranial Hemorrhage in Two Intubated Children Using the Correlate of Injury to the Nervous System (COIN) Index, a Quantitative Electroencephalographic Tool [Saturday Poster Session, Event Room 2]**

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**Introduction:** Ancillary methods are needed to screen for intracranial hemorrhage (ICH) in children on life support. We developed a Quantitative Electroencephalographic (EEG) Correlate Of Injury to the Nervous System (COIN) index that characterizes focal voltage attenuation in ischemic stroke. We present two cases of ICH with COIN changes occurring prior to clinical recognition of neurologic deterioration.

**Methods:** Clinical data were collected from the electronic health record. COIN was calculated from raw EEG using MATLAB. Wilcoxon rank sum test was used to compare median COIN values.

**Case:** Patient 1 was an 8-year-old male with aplastic anemia who developed severe agitation requiring intubation and sedation for safety. Initial brain MRI was normal. EEG showed symmetric diffuse slowing. COIN visualization shows onset of right posterior attenuation at 20 hours after EEG initiation [change in COIN from -3 to -16 ( $p < 0.001$ )], which rapidly expands at 27 hours [change in COIN from -16 to -45 ( $p < 0.001$ )]. He developed unreactive pupils at 29 hours; imaging showed large right occipital ICH with herniation (*Refer to poster*). Patient 2 was a 4-month-old male with congenital heart disease who was cannulated onto extracorporeal life support after cardiac arrest. Initial head imaging was normal. EEG showed symmetric diffuse slowing. COIN visualization shows an area of right frontotemporal voltage attenuation 8 hours after EEG initiation [change in COIN from -15 to -42 ( $p < 0.001$ )]. Repeat imaging 13 hours after EEG start showed large right subdural ICH with leftward midline shift (*Refer to poster*).

**Discussion:** ICH is a morbid complication of critical illness in pediatric patients and is often unrecognized until brain herniation syndromes occur. Implementation of COIN may enhance timely detection of ICH and facilitate early intervention.

### **P-C-2: Impact of physiologic parameters on outcome after arteriovenous malformation rupture in pediatric patients [Friday Poster Session, Event Room 1]**

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**BACKGROUND:** Arteriovenous malformations (AVMs) are the leading cause of intracranial hemorrhage (ICH) in children; however, little is known about the critical care management of pediatric ruptured AVMs. We evaluate the impact of critical care management on neurologic outcome after AVM rupture in children.

**HYPOTHESIS:** Physiologic derangements will impact the outcome of pediatric patients with ruptured AVMs. **METHODS:** Retrospective study of patients <18 years of age with ICH secondary to AVM rupture admitted to a single tertiary



children's hospital from 2011-2023. Physiologic parameters were collected for the first 7 days after AVM rupture. Poor neurologic outcome was defined as Pediatric Stroke Outcome Measure A univariate binary logistic regression was used to evaluate associations between physiologic score  $\neq 1$ . parameters and outcome.

**RESULTS:** Forty-nine patients met inclusion criteria. The median age was 12.4 years (IQR 7.6-14.7), and 63% (31/49) had a poor outcome at discharge. During the first 7 days after AVM rupture, 96% (46/49) had hypertension (systolic blood pressure (SBP)  $\geq$  95th percentile), 49% (25/49) had hyperthermia (temperature  $\geq$  38 C), 49% (24/49) had hypotension (SBP  $\leq$  5th percentile), 24% (12/49) had hyperglycemia (blood glucose  $\geq$  200 mg/dL), and of those with an intracranial pressure (ICP) monitor, 84% (21/25) had increased ICP (ICP  $\geq$  20 mmHg) and 77% (17/22) had decreased cerebral perfusion pressure (CPP  $<$ 50 mmHg). Hyperthermia (68% vs. 22%,  $p = 0.004$ ) and hyperglycemia during the first 7 days after AVM rupture (35% vs. 6%,  $p = 0.041$ ) were associated with poor outcome.

**CONCLUSIONS:** For children with ICH due to ruptured AVM, hyperthermia and hyperglycemia during the first 7 days are associated with poor neurologic outcome - suggesting potential targets for improved critical care management in this population.

### **P-C-3: Recognition of Neurovascular Complications of Extracorporeal Life Support Using Quantitative Electroencephalography [Friday Poster Session, Event Room 1]**

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**Significance:** Stroke is a morbid complication of Extracorporeal Life Support (ECLS). The Quantitative Electroencephalographic (EEG) Correlate of Injury to the Nervous System (COIN) index visualizes and characterizes focal suppression suggestive of stroke (Figure 1). Recognition of stroke during ECLS may enable recognition of ischemia for therapeutic interventions.

**Methods:** Retrospective cohort study of children during ECLS with and without focal intracranial injury (FII), defined as focal brain ischemia or hemorrhage  $> 1\text{cm}^3$ , or extraaxial hemorrhage with mass effect on brain. Subjects were consecutively sampled using the Extracorporeal Life Support Organization registry; additional data were gathered from chart review. COIN was calculated from EEG using MATLAB.

**Results:** Nineteen subjects included (*Refer to poster*). Sixteen (84%) had venoarterial (VA) and 3 (16%) venovenous (VV) support. Eight VA and 0 VV subjects had FII. On VA, 9 had seizures (5 with FFI; 4 without); no subjects on VV had seizures. Figure 2 (*Refer to poster*) shows COIN values over time. In the VA group, COIN below -20 for 4 hours had 100% sensitivity and 88% specificity for FII; COIN below -30 for 3 hours had 88% sensitivity and 100% specificity. Six (32%) subjects had FII within 24 hours of cannulation or a circuit change. VA subjects without FII had intermittent COIN below -20 that was not sustained. All VV subjects had left hemispheric suppression with COIN below -20 and no FII.

**Discussion:** Characterization of focal power suppression with COIN allows recognition of FII during ECLS. Diagnostic value of COIN will be better understood with expansion of the study cohort. Left-sided suppression observed in VV ECLS is not associated with FII. In VA ECLS, FII occurs in association with cannulation or circuit change; FII recognition may be facilitated by COIN.

### **P-C-4: Spontaneous Thrombosis of Isolated Extra-Axial Cerebral Varix: to Treat or Not to Treat? [Friday Poster Session, Event Room 1]**

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A 15-year-old boy presented with a prolonged focal seizure involving the right leg. Examination revealed hyperreflexia of the lower extremities. MRI demonstrated a non-enhancing extra-axial cerebral varix at the left superior cerebral convexity causing mass effect on the underlying brain parenchyma with signal characteristics concerning for thrombosis (*Refer to poster*). Post-contrast images showed no vessel supplying or draining the varix (*Refer to poster*). Cerebral

angiography was negative for the presence of an arteriovenous fistula (AVF), aneurysm, or other high flow vascular malformation. Few reported cases illustrate that cerebral varices are generally intra-axial, typically asymptomatic<sup>1, 2</sup>, and more commonly associated with other vascular malformations (dural AVF, developmental venous anomalies). After considering the risks and benefits of anticoagulation (thrombus propagation into the superior sagittal sinus versus hemorrhage from an unidentified, thrombosed AVF), close observation with follow-up imaging was recommended. Figure 1 (*Refer to poster*) MRI/MRV brain Axial T2 (A), T1 (B), and SWI (C) images of the brain at the convexity show an isolated, expanded left posterior frontal varix with mild T2 hyperintensity, T1 hyperintensity, and blooming on SWI (asterisks). Contrast the T2 intensity with the normal flow void in the superior sagittal sinus. Findings consistent with intravascular thrombus. MRV (D) showed patent dural venous sinuses and non-visualization of the vein of Trolard on the left. Coronal and Sagittal T1-post contrast images and corresponding anteroposterior and lateral views from a left carotid angiogram. Coronal (A) and sagittal (C) images show a well-defined, isolated left posteromedial frontal varix (asterisks) with mild mass effect on the brain parenchyma. No contrast opacification is present in the varix and a corresponding defect is seen (arrows) during the capillary phase on cerebral angiography (B, D).

## Outcomes and Rehabilitation

### **O-D-1: Stimulation for Perinatal Stroke Optimizing Recovery Trajectories (SPORT): A Randomized, Controlled Clinical Trial [Oral Abstract Session]**

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**Background:** Perinatal stroke causes hemiparetic cerebral palsy and lifelong disability for millions. Manual therapies can enhance upper limb function but effect sizes are modest. Whether transcranial direct current stimulation (tDCS) can enhance upper limb rehabilitation for children with hemiparesis is unknown.

**Methods:** We conducted a multicentre, randomized, double-blind, sham-controlled trial (NCT03216837). Children aged 6-18 years with unilateral perinatal stroke and disabling hemiparesis were randomly assigned (1:1) to receive daily sham or 1mA cathodal tDCS to the contralesional motor cortex during 10-days of high-dose, child-centred, peer-supported, intensive rehabilitation. Co-primary endpoints were change from baseline to 6 months in hand function (Assisting Hand Assessment, AHA) and attainment of child-identified functional goals (Canadian Occupational Performance Measure, COPM). Secondary outcomes included hand use and quality of life. Safety was assessed by decrease in the function of either hand.

**Results:** Of 89 children enrolled, 83 had complete outcome data (42 sham, 41 tDCS). There were no differences between groups at baseline. High proportions of children in both groups demonstrated significant functional gains and goal achievement that were sustained at six months ( $p < 0.0001$ ). Mean change from baseline for AHA ( $5.2 \pm 5.3$  vs  $4.6 \pm 5.7$ ,  $p = 0.63$ ) and COPM ( $3.0 \pm 2$  vs  $3.6 \pm 2.3$ ,  $p = 0.25$ ) were comparable between sham and tDCS. Improvements in hand use and quality of life were also similar between groups. Procedures were well-tolerated with no serious adverse events.

**Conclusions:** Patient-centred intensive motor learning programs can produce marked and sustained improvements in upper extremity function in children with perinatal stroke. Gains are not enhanced by daily contralesional cathodal tDCS.

### **O-D-3: Utilising multi-contrast MRI-based machine learning to predict cognition in people with Sickle Cell disease [Oral Abstract Session]**

Jonathan Clayden, Russell Murdoch, Hanne Stotesbury, Melanie Koelbel, Karin Shmueli, Christopher Clark, Fenella Kirkham  
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Cognitive impairment, particularly involving processing speed is common in children and young adults (CYA) with Sickle cell disease (SCD) with or without stroke or silent cerebral infarction (SCI). MRI is a non-invasive tool for investigation of the complex mechanisms of injury which appear interdependent. We hypothesised that multi-contrast MRI machine learning models would predict processing speed index (PSI) in CYA with SCD. Four increasingly complex models were built using data sourced from the Sleep Asthma & Prevention of Morbidity in SCD cohorts (Mean age 17.4; 49 CYA with SCD, 28 male; 31 controls, 14 male; Table 1, *refer to poster*). All models included fractional anisotropy (FA), mean diffusivity (MD) and Quantitative Susceptibility Mapping (QSM) and non-imaging metrics: age, sex, patient/control, presence of infarction, closest haemoglobin, and haematocrit concentrations. Across all the models, the diffusion metric, MD, showed a significant relationship with PSI. In Model 1, this was in the whole white matter in patients aged less than 18 (Table 2, *refer to poster*). In Model 2, three regions showed a significant relationship. The FA only showed a relationship within the second model in four white matter bundles (Table 3, *refer to poster*). In Model 3 the top 20 regions with highest predictive power were in the MD images and in Model 4, the only 2 regions that were found to be significant were also in the MD image (Figure 1, *refer to poster*). MD was found across all models to have the best predictive power for PSI in patients with SCD. This could indicate that a change in the PSI is related to a change in the white matter structure as shown in the diffusion weighted images. FA has a spatial dependence whereas MD is spatially independent and is essentially looking for a difference from normal structure. However, most models had a high mean squared error which could indicate low predictive power.

**P-D-1: Motor network functional activation and white matter connectivity patterns are related to clinical outcomes following perinatal and childhood arterial ischaemic stroke. [Saturday Poster Session, Event Room 1]**

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**Introduction:** Motor outcomes following perinatal and childhood stroke are highly variable and likely reflect complex structural and functional changes within motor-related brain networks. Greater understanding of these network mechanisms is needed to improve patient-specific prognostication and develop targeted therapies to restore motor function. This study explored post-stroke reorganisation patterns in relation to motor outcome through assessment of functional MRI (fMRI) activation and white matter connectivity patterns.

**Hypothesis:** We hypothesised that different activation and connectivity patterns would be seen in children with hemiparesis following stroke compared to children without hemiparesis.

**Methods:** Twelve children (age 9-19 years) with a history of either perinatal or childhood arterial ischaemic stroke underwent 3T MRI with diffusion-weighted imaging and task-related hand-motor fMRI. Clinical assessment of motor outcome using the Paediatric Stroke Outcome Measure was obtained and dichotomised into a hemiparesis and a no hemiparesis group. MRI data was pre-processed and analysed for whole-brain fMRI activation patterns and white matter track-weighted functional connectivity (TW-FC) profiles in relation to motor outcomes.

**Results:** Divergent fMRI activation and TW-FC connectivity patterns were seen in children with hemiparesis versus those without. Those without hemiparesis were more likely to retain an ipsilesional activation pattern while those with hemiparesis were more likely to demonstrate bilateral or contralesional patterns. When function was retained in the stroke-affected hemisphere, increased utilisation of ipsilesional cortical-subcortical pathways occurred. Greater TW-FC strength when moving the stroke-affected hand was also seen in those without hemiparesis compared to the hemiparesis group.

**Conclusions:** We have identified distinct patterns of motor network activation and white matter connectivity relating to clinical outcomes after perinatal and childhood arterial ischaemic stroke. Recognition of these patterns has the potential to improve prognostic ability and aid selection of babies and children who would most benefit from targeted therapy to improve long-term motor outcomes.

**P-D-2: Long-term neurodevelopmental outcome after childhood stroke with different vascular types.  
[Saturday Poster Session, Event Room 1]**

Kerttu Kivisikk<sup>1,2</sup>, Pilvi Ilves<sup>3,4</sup>, Mairi Männamaa<sup>2,3</sup>, Nigul Ilves<sup>3,4</sup>, Norman Ilves<sup>3,4</sup>, Inga Talvik<sup>5</sup>, Dagmar Loorits<sup>4</sup>, Pille Kool<sup>3</sup>, Rael Laugesaar<sup>1,2</sup>  
University of Tartu

**Introduction:** Children with childhood stroke may develop long-term disabilities including motor deficit, cognitive and behavioral disorders. Our aim was to describe the long-term global neurocognitive outcome of children suffering from childhood stroke in Estonia.

**Hypothesis:** Our hypothesis was that children with arterial ischemic stroke (AIS) have worse global outcome than children with other vascular types of stroke.

**Methods:** Population-based prospective study, patients were identified from the Estonian Pediatric Stroke Database. Inclusion criteria were (1) Patients with radiographically confirmed AIS, hemorrhagic stroke (HS) or sinovenous thrombosis (CSVT); (2) Age at diagnosis 28 days to 18 years; (3) At least 2 years after stroke; (4) Age at study participation <19y. We excluded patients with a concomitant disease which affects cognitive functions. All patients were evaluated in Tartu University Hospital by a multidisciplinary team. The main outcome measure used was Pediatric Stroke Outcome Measure (PSOM), 0-0.5 was considered good outcome,  $\geq 1$  poor outcome.

**Results:** There were a total of 40 participants, 24 with AIS, 16 AHS and 4 CSVT. Average time since diagnosis was 8,1 years (SD 4,3y). Poor global outcome was found in 62% of the patients. There was no difference in poor global outcome in AIS and HS groups (63% vs 67%,  $p>0.99$ ). SVT had a better outcome but there were only 4 participants with CSVT. The main contributor to poor outcome in AIS group was sensorimotor deficit (80%), but in HS group cognitive and behavioral problems (75%).

**Conclusions:** Global outcome of stroke is poor in two-thirds of the patients and is equally poor in AIS and HS groups and routine assessments should include neuropsychological and language testing.

**Funded by:** Estonian Research Council PRG1912, Tartu University Hospital PR-143/22

**P-D-3: Considering the ipsilesional upper extremity in chronic hemiparesis from perinatal ischemic stroke  
[Saturday Poster Session, Event Room 1]**

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Children's Hospital of Philadelphia

**Introduction:** Upper extremity rehabilitation of pediatric hemiparesis focuses on contralesional motor execution. However, impairments of each UE in isolation, as well as bimanual coordination impairments, have been documented in unilateral CP from mixed etiology<sup>1-4</sup>. In adults, ipsilesional deficits worsen with severity of contralesional paresis<sup>5</sup>. We compared the motor performance of each UE in confirmed cases of unilateral PAIS to typically developing peers and explored the relationship between severity of paresis and impairment of the ipsilesional UE.

**Hypotheses:** Children with history of perinatal unilateral stroke would show motor impairments of each UE, and ipsilesional impairments would worsen with severity of contralesional paresis.

**Methods:** We included cases of unilateral perinatal ischemic stroke confirmed by imaging who were consented participants in Children's Hospital of Philadelphia Stroke Registry (IRB approval). The Box and Blocks Test (BBT) measured unilateral gross motor dexterity. The Nine Hole Peg Test (NHPT) measured unilateral fine motor dexterity. Z-scores describe performance relative to normative references. We used T-tests for paired comparisons involving normal distributions and Wilcoxon Rank Sum tests for abnormal distributions. ANOVA analyzed performance with respect to severity of paresis (Kuskal-Wallis tests for abnormal distributions).

**Results:** Retrospective chart reviews revealed 41 eligible patients (median age 7-years) with relevant data. For each UE, average Z-scores were low. There was a significant effect of severity on BBT scores for each UE. The mildly paretic group performed better than moderate and severe groups. For the contralesional UE only, there was a significant effect of severity on NHPT. There were positive Pearson correlations between the contralesion.

#### **P-D-4: Brain tissue microstructure in sickle cell disease: relationship with sleep [Saturday Poster Session, Event Room 1]**

Leevi Kerkala, Christopher Clark, Fenella Kirkham  
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Adults and children with sleep-disordered breathing have evidence of abnormal white matter microstructure on diffusion tensor imaging (DTI). Similarly, compared with race- and age- matched controls, people living with sickle cell disease (SCD) have abnormalities on DTI associated with slow processing speed, but whether other cognitive domains, e.g. executive function, are compromised and whether this is related to sleep disturbances, e.g. increase in periodic leg movements (restless legs) is unknown. In this study, actigraphy was used to measure sleep disturbance as nocturnal mobile minutes, in children and young adults with SCD (CYA-SCD; N=20, age range 11.1-29.4 years) and healthy controls of black heritage (N=12, age range 14.9-25.1 years) and was compared with DTI parameters including fractional anisotropy (FA), mean diffusivity (MD), axial diffusivity (AD) and radial diffusivity (RD), in the arcuate fasciculus (AF), Cingulum (CG), Inferior fronto-occipital fasciculus (IFO), Superior longitudinal fasciculus (SLF) and Uncinate fasciculus (UF). Contemporaneous BRIEF data were available to investigate any association with executive function. After controlling for age, sex and socioeconomic status significant differences between CYA and controls remained for (1) FA and three tracts: AF-left, SLFIII-left and UF-left; (2) RD and UF-left. Partial correlational analysis in CYA-SCD alone, controlling for sex, age, SES, Hydroxyurea prescription and daytime SpO2 found relationships for FA in SLF, MD in GG-left, IFO-right and SLFIII-right and AD in AF-left (Table 1, *refer to poster*). There were associations with Cingulum DTI parameters and BRIEF Global Executive Composite (Figure 1, *refer to poster*), Metacognition Index and Regulation Index consistent with an effect of disrupted white matter integrity on executive function. CYA-SCD showed differences from controls on various DTI measures. CYA-SCD who experience restless sleep have compromised white matter integrity with executive dysfunction.

#### **P-D-5: Language lateralization and outcome in perinatal stroke patients with different vascular types [Saturday Poster Session, Event Room 1]**

Nigul Ilves, Mairi Männamaa, Rael Laugesaar, Norman Ilves, Dagmar Looorits, Ulvi Vaheer, Pille Kool, Pilvi Ilves  
University of Tartu; Tartu University Hospital

**Introduction:** Perinatal stroke (PS) affects children's language development and can change language lateralization. The vascular type and lesions are heterogeneous in PS. The heterogeneity and low prevalence leave the relationship between language lateralization and outcome in children with PS unclear.

**Hypothesis:** a) There are differences in language lateralization during task based functional MRI (fMRI) in children with left-side arterial ischemic stroke (AIS) or periventricular venous infarction (PVI) and in healthy controls, b) large cortical damage is associated with contralesional language activation, c) reorganization is not so effective for language outcome.

**Methods:** Language generation and comprehension tasks in fMRI were used to determine language lateralization in term born children with perinatal left-side AIS (n = 9, mean age (SD) 13.4 (3.1) y.) and PVI (n = 12, 11.8 (2.8) y.), and in controls (n = 30, 11.6 (2.6) y.). Lateralization index was calculated for the Broca and Wernicke areas and correlated with language outcomes measured by the Kaufman Assessment Battery for Children II ed. Results Language lateralization and outcome in children with PS was different compared to controls. Children with small AIS lesions and most children with PVI showed typical left-side language activation. Most children with large AIS lesions and one child with large PVI had reorganized right hemisphere language activation. Children with typical language activation had better language outcome compared to children with reorganized language activation after stroke.

**Conclusions:** Language lateralization in children with PS depends on lesion size and location. Language reorganization to the right hemisphere did not ensure normal language outcome.

Funded by: Estonian Research Council PRG1912 and Tartu University Hospital PR-2063



## **P-D-6: An embodied speech neuroprosthesis for patients with paralysis using electrocorticography [Saturday Poster Session, Event Room 1]**

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Loss of speech due to neurological injury is devastating and can severely degrade communication and quality of life. Speech neuroprostheses have the potential to restore natural communication by decoding cortical activity into intended speech, bypassing diseased pathways. In our work, we used electrocorticography (ECoG) to record cortical activity from the speech-motor cortex of adult patients who are unable to speak due to brainstem stroke. Our goal was to develop a system capable of restoring fully-embodied communication to these patients in multiple forms including text (i.e. sentences), audible speech, and orofacial movements/expressions. We developed deep-learning models to decode cortical activity into text, personalized audible speech, and personalized orofacial movements, animated in a virtual avatar. We were able to decode these three modes of communication simultaneously and, for text decoding, achieved a median word error rate of 25% at 78 words per minute on unseen sentences from a 1024-word vocabulary. Further, in one Spanish-English bilingual participant, we demonstrated that our system can flexibly decode text in multiple languages, offering an important proof-of-concept for generalizability to the many non-English and bilingual speakers. Overall, we found that our decoding performance relied on detailed representations of the orofacial-speech articulators (articulatory representations) that persist on speech-motor cortex years after paralysis. These representations generalize across languages and maintain a somatotopic organization. In the future, we aim to expand this technology to pediatric cases of speech loss and impairment. This may involve electrode placement in different cortical regions and adapting decoding paradigms to provide closed-loop feedback for more flexible control over the system.

## **P-D-7: Neural oscillatory differences between pediatric stroke patients with and without dystonia: An MEG study [Saturday Poster Session, Event Room 1]**

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Hospital for Sick Children, University of Toronto

**Background/Objective:** Dystonia affects 20% of pediatric arterial ischemic stroke survivors and is characterized by repetitive muscle contractions, twisting movements, and abnormal posture. Dystonia has been linked to cognitive impairments, but it is not yet clear whether it is solely a motor disorder or whether cognitive networks are also impacted. We investigated potential alterations in frontal theta (4-8 Hz), motor beta (15-30 Hz), and sensorimotor gamma (60-90 Hz) band activity during a go/no-go inhibitory control task in children with post-stroke dystonia in comparison to those without dystonia, and to age-matched healthy control children.

**Methods:** Beamformer-based source analysis was carried out in 19 post-stroke patients, 9 of whom had dystonia (mean age=13.78, SD=2.82) and 10 without (mean age=12.90, SD=3.54), and 18 healthy controls (mean age=12.82, SD=2.72). Frontal theta activity was analyzed during correct no-go (movement withhold) trials to evaluate inhibitory control, and motor beta and sensorimotor gamma activity were analyzed during correct go trials to assess motor execution and sensorimotor integration.

**Results:** When using their affected hands, stroke patients had higher error rates, and correct movement withhold was associated with higher theta power compared to healthy controls. Dystonia patients exhibited a lower change in gamma power and peak frequency compared to non-dystonia patients and healthy controls. Beta band activity was comparable across groups.

**Conclusion:** These results confirm that childhood stroke is associated with impaired inhibitory control requiring greater cognitive effort as reflected by increased frontal theta activity. Post-stroke dystonia patients also show altered sensorimotor integration as reflected by reduced motor gamma activity during movement. These findings suggest that poor motor control in childhood stroke may result from a combination of cognitive and motor deficits.



## P-D-8: Fatigue after Paediatric Stroke [Print Only]

Dr John Down, Professor Karen Barlow, Ms Louise Sparkes, Dr Michaela Waak, Dr Adriane Sinclair  
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**Introduction:** Sensorimotor and speech deficits post Acute Ischemic Stroke (AIS) are well defined and often the focus for rehabilitation. However, there is increasing evidence to show the effects of AIS on neuropsychiatric domains including cognition and fatigue levels. While fatigue is a commonly identified symptom in many chronic illnesses and post AIS in the adult population, literature in the paediatric population post AIS is lacking.

**Hypothesis:** Fatigue is hypothesised to have an impact on paediatric patients post stroke. The aims of this study are to investigate the prevalence of fatigue post AIS and to investigate if baseline patient or stroke characteristics predict post stroke fatigue. **Methods:** This is a retrospective audit investigating fatigue after paediatric stroke from a single institution (Queensland Children's Hospital HREC/23/QCHQ/99297). Predictors of fatigue post AIS were identified through the use of a multiple logistic regression model. Variables were removed from the multiple model using backwards selection until only significant predictors ( $p < 0.05$ ) remained.

**Results:** Twenty-five patients met criteria for inclusion in this study. Fatigue was reported in thirteen (52%). Gender, previous neurodevelopmental disability, or MRI brain involvement did not predict fatigue. Age at time of stroke predicted fatigue with each year increase in age estimated to lead to 1.26 times increase in the odds of fatigue post-stroke, when the PedNIHSS was held constant ( $p = 0.048$ ). PedNIHSS score also predicted fatigue with each unit increase in the PedNIHSS score estimated to lead to 1.27 times increase in the odds of fatigue post stroke, when age was held constant ( $p = 0.024$ ).

**Conclusion:** Over half our population was affected by post stroke fatigue. Age at time of stroke and PedNIHSS score predicted fatigue.

## P-D-9: Tailored GPT4 Prompting vs. Physician Standard in Assessment of Pediatric Stroke Data [Saturday Poster Session, Event Room 1]

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Children's Memorial Hermann Hospital

**Introduction:** Pediatric stroke is a rare but important cause of morbidity in children, leaving approximately 75% of pediatric stroke survivors with permanent neurologic deficits.<sup>1–3</sup> Research in pediatric stroke can be challenging due to its relative rarity, but accurate data has been captured through collaborative efforts in the International Pediatric Stroke Study (IPSS). Data entry in the IPSS must be done by trained professionals and can be time-consuming. In our study, we explore the use of an advanced generative pre-trained model based on GPT4 to learn Stroke Ontology (STO), specifically designed with domain-specific prompt engineering to ask only relevant questions in a joint manner, with the aim to extract information from free text medical notes. STO-GPT performs zero-shot learning and converts free-text data to structured data conforming to a specific tabular format. STO-GPT can be used to facilitate automatic data entry and reduce investigator workload. Our study tests STO-GPT's ability to reduce investigator workload through automatic data entry.

**Hypothesis:** We aim to evaluate the accuracy of STO-GPT in extracting information and structuring free text data for pediatric stroke patients from clinical notes. Our hypothesis is that large language models such as GPT 3.5 and GPT-4 are good zero shot learners that can understand ontology and perform domain-specific named entity recognition by performing completely zero-shot prompting, without any re-training or fine-tuning. STO-GPT will be highly accurate at filling out the outcomes form, as assessed by percent agreement with the investigator-entered data (>90%).

**Methods:** The most recent 50 clinical notes at the UTHealth Pediatric Stroke clinic were deidentified and assessed. The study included patients with arterial ischemic stroke or cerebral venous sinus thrombosis seen between January 2020 and July 2023. IPSS database questions are interpreted by the investigators and an ontology is built from it to inform STO-GPT. Both a pediatric vascular neurologist and STO-GPT were separately asked to answer IPSS database questions for

each patient. Percent agreement measured between 0 and 1 between human and ChatGPT was assessed across 50 patients for each of the 114 queries developed from the IPSS database outcome questionnaire.

**Results:** Our specialized model demonstrated strong performance in several IPSS questions but showed variability overall. While it was occasionally able to match human judgement with a score of 1.00 (n=20, 17.5%), it scored as low as 0.26. Five of the 114 (4.4%) queries scored below 0.50, while 92 (80.7%) scored at least 0.80. Mean and median matching score was 0.887 and 0.960, respectively. In its final form, aggregate agreement was 94%, with a maximum agreement of 100% and minimum of 62%. Of 2400 individual items assessed, our model entered 2247 items correctly and 153 (6.4%) incorrectly. Of those errors, 137(90%) were misidentification, 9 (6%) were misplacement, and 7 (4%) were hallucination errors.

**Conclusions:** While our tailored generative model with domain-specific prompt engineering and ontological guidance shows promise for research applications, some further refinement is needed to enhance its accuracy on certain named entities in the patient data assessment, in order to boost its overall performance. The current performance is approaching satisfaction. Although it has not reached the point of completely obviating the necessity for human supervision in database entry, it can be effectively employed in tandem with human oversight, contributing to a collaborative approach that reduces overall effort in the data abstraction process.

### **P-D-10: Exploring the Relationship Between Arterial Hypertension, White Matter Microstructural Integrity, and Cognitive Function in Paediatric Sickle Cell Anaemia Patients [Saturday Poster Session, Event Room 1]**

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Sickle cell anaemia (SCA) is marked by abnormal haemoglobin, resulting in sickle-shaped red blood cells and associated complications, including stroke and posterior reversible encephalopathy syndrome, linked to relative hypertension.[1] Cognition, particularly processing speed[2, 3], in SCA, may be linked to compromised white matter microstructural integrity (WMSI) on diffusion tensor imaging MRI (DTI-MRI).[4] Data from the Framingham and UK Biobank studies have associated hypertension with compromised WMSI and vascular dementia in the general adult population.[5, 6] Recent studies indicate a rising prevalence of hypertension in young individuals with SCA[7-9], yet any impact on WMSI remains unexplored. This study aims to investigate the association between hypertension, WMSI on DTI-MRI, and processing speed in paediatric and adolescent participants with SCA. Methods Blood pressure (BP) was measured 3 times at rest in 2005-10 in the Sleep Asthma Cohort, comprising 89 patients aged 4-18 years. Hypertension was determined using the Pegelow criteria (systolic BP>90th percentile). Mean diffusivity (MD) from DTI-MRI and processing speed from Weschler Intelligence Scales for Children (WISC-IV) in 2016-18 were compared in those with and those without hypertension using independent t-tests. Results Of 89 individuals, 44% exhibited systolic BP>90th percentile, indicating a high prevalence of relative hypertension in paediatric and adolescent SCA patients. Among the 29 individuals with available DTI data, those with hypertension showed a trend-level increase in MD (p=0.06; Fig.1, *refer to poster*), suggesting early WMSI compromise. Coding, Symbol search, Cancellation and Processing speed index were lower in the 13 with hypertension at baseline in SAC compared with 24 with normal BP (Fig.2-3, *refer to poster*), significantly for Cancellation(p=0.04). Conclusions This study emphasizes documenting BP in children with SCA and implementing therapeutic strategies to enhance cognitive care.

### **P-D-11: A Programmatic Approach to School Re-entry Support After Pediatric Cerebrovascular Disease [Saturday Poster Session, Event Room 2]**

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**Introduction:** Children with stroke have cognitive and communication deficits compared to healthy children; such deficits can impair return-to-school. Evidence shows that a structured approach to school reentry is well received by families and school systems, leading to positive outcomes when patients become students again.

**Hypothesis:** A structured approach to school support helps improve tangible outcomes in school re-entry.

**Methods:** This is a retrospective analysis of school-aged children (kindergarten to senior year) with cerebrovascular brain injury who were hospitalized at our institution between 2018-2023. Patient details, including age, sex, type of stroke, hemisphere, and presence of acute seizure are abstracted. When available, PSOM and neuropsychological data from admission and first follow-up exam are included. School liaison data including Brief School Needs Inventory (BSNI), Goal Attainment Scaling (GAS), and academic data are presented.

**Results:** Structured school liaison-mediated intervention services are provided to all school-aged children in acute rehabilitation after stroke. We detail the intervention process with emphasis on how services better prepare schools and families for returning students. Families report high satisfaction with the school liaison process. Cognitive impairments after stroke are often interpreted as greater barriers to school re-entry than physical or medical comorbidities. Use of standard scales, including the BSNI, helps in part with identifying school re-entry concerns in children recovering from stroke.

**Conclusion:** Children in recovery from cerebrovascular injury have substantial challenges receiving appropriate instructional accommodations. These stem from a new chronic illness which is often complicated by acquired learning difficulties. Scales directed at assessment of the unique challenges facing children with brain injury are needed to support successful school reentry.

### **P-D-12: Cross-Sectional Bibliometric Analysis of 3,500+ Neurosurgeons [Print Only]**

Siddharth Ghanta, Disha Jotsinghani, and Devesh Shah  
Duke University

Publication productivity is a key aspect of academic neurosurgery, and objective measurements of research output are relevant for promotion, grants, self-evaluation, and program-level impact. Neurosurgery is an innovative and emerging field, and thus research is a core aspect of neurosurgical training and practice. Recent literature has yet to provide a transparent and objective report of neurosurgical publications across programs and subspecialties. In our investigation, we aim to present a descriptive analysis of neurosurgical research productivity for 117 programs recognized by the Accreditation Council for Graduate Medical Education (ACGME). For each of these programs, we collected the names, ranks, and subspecialties of all M.D. physicians and their research output, which we quantified using the h-Index, i10-Index, m-Quotient, hc-Index, and g-Index. We excluded neurosurgeons with a lack of sufficient data. We conducted a statistical analysis to identify potential trends in publications across programs, sex, rank, and subspecialty. The results provided from this work serve as benchmarks for academic neurosurgeons and comparisons amongst programs. Furthermore, identified trends contribute to bibliometric profiling of neurosurgical departments across the United States.

### **P-D-13: Hidden Challenges: Mental Health Outcomes in Pediatric Stroke Survivors [Print Only]**

Ludovica Serafini, Robyn Westmacott, Jennifer Crosbie, Amanda Robertson, Daniel Nichol, Birgit Ertl-Wagner, Farzad Khalvati, Alhussain Abdalla, Min Sheng, Trish Domi, Nomazulu Dlamini  
The Hospital for Sick Children

**Introduction:** The burden of disease from pediatric arterial ischemic stroke (AIS) includes the effects of non-visible disability in the form of mental health conditions such as depression and anxiety. These conditions impact the quality of life of children and families affected by stroke.

**Hypothesis:** Children and adolescents with AIS are at greater risk of developing a mental health condition than the general pediatric population. Methods A retrospective analysis of a prospective single-center cohort of school-aged children diagnosed with AIS, enrolled between 2002 and 2020. Depression and anxiety were evaluated with the Behavior Assessment System for Children (BASC, versions 1 and 3). Prevalence and scores for depression and anxiety were ascertained for the cohort and compared with general pediatric population prevalence.

**Results:** 178 patients (109 male; median age and range at stroke 2.4 years [0 - 14.2 years]) had neuropsychological assessments at median age and range 9.0 years (5.6 - 16.7 years). Clinical scores for depression and anxiety, i.e.,  $T \geq 70$  in the Depression (mean = 81.4, SD=8.5) or Anxiety (mean= 76.6, SD=8.3) subscales, were found in 12% and 14% of school-age children with AIS, at median age and range 9.9 years (5.9 -16.7). Scores for mood and anxiety were higher in AIS patients, with depression scores 3-4 times greater than among the general pediatric population in Ontario.

**Conclusions:** Mental health disorders represent a significant problem in pediatric stroke survivors, with the end of primary school appearing to be the time of greatest vulnerability. Related symptoms are frequently overlooked, partly due to different presentations in children compared to adults. Early detection and intervention are essential to change the patient trajectories and improve overall outcomes and quality of life.

## Perinatal Stroke

### P-E-1: Fibre-specific white matter alterations following perinatal stroke [Friday Poster Session, Event Room 1]

Alicia Hilderley, Adam Kirton, Helen Carlson  
University of Calgary

**Introduction:** Perinatal stroke alters white matter microstructure proximal and distal from the lesion location. The majority of evidence is based on voxel-wise measures, which lack specificity in areas of crossing fibres. This limitation can be addressed by whole-brain fixel-based analysis to examine individual fibre populations within a voxel. Reduced fixel-based metrics have been observed in children with neonatal arterial ischemic stroke (AIS), but children with periventricular venous infarctions (PVI) have not been studied.

**Hypothesis:** We hypothesized that alterations in fixel-based metrics would be observed in children with AIS and PVI as compared to typically developing children (TDC), with more widespread alterations in AIS.

**Methods:** 98 children completed MRI scans (mean age 10.8 years, SD 2.5; 39 female), including 24 children with AIS (9 right-sided stroke, 15 left), 29 with PVI (14 right, 15 left), and 45 TDC. Diffusion images were preprocessed following a fixel-based analysis pipeline using MRtrix3. Fixel-based metrics extracted were fibre density (FD), fibre bundle cross-section (FC), and a combination of fibre density and cross section (FDC). Right and left-sided strokes were separated for analyses with comparisons between AIS, PVI, and TDC. Statistical inference was performed for each fixel using a general linear model and non-parametric permutation testing (significance set at FWE-corrected  $p < 0.05$ ). Results: Significant differences in FD and FDC were observed proximal and distal from the lesion location in AIS and PVI as compared to TDC. Differences were observed in the ipsilesional hemisphere and interhemispheric tracts. There were more global differences in AIS, especially in the contralesional hemisphere.

**Conclusions:** Remote fibre-specific alterations in white matter are found in AIS and PVI, indicative of diaschisis of structural connections.

### P-E-2: Motor Cortex GABA in Children with Hemiparesis after Perinatal Stroke [Friday Poster Session, Event Room 1]

Paulina Hart, Tiffany Bell, Ashley Harris, Helen Carlson, Adam Kirton

University of Calgary

**Introduction:** Perinatal stroke (PS) can damage the motor system and lead to hemiparetic cerebral palsy. Current treatment is limited to rehabilitation therapy, but non-invasive brain stimulation is a promising adjuvant. Transcranial direct current stimulation (tDCS) uses weak current to alter plasticity and enhance motor learning but its effects on motor cortex chemistry is unclear. Personalized, functional MRI-guided magnetic resonance spectroscopy (MRS) has demonstrated metabolite level differences between hemispheres. However, it is unknown how  $\gamma$ -aminobutyric acid (GABA), a primary inhibitory neurotransmitter, is altered in motor cortices of children with PS.

**Hypothesis:** Motor cortex GABA levels will decrease in the non-lesioned hemisphere of children receiving active tDCS and will be correlated with increases in motor function.

**Methods:** Participants (6-18 years) with periventricular venous infarction (PVI), or arterial ischemic stroke (AIS) were recruited in the Stimulation for Perinatal Stroke Optimizing Recovery Trajectory (SPORT) trial. Participants attended a 10-day camp including individual physical therapy sessions and received active or sham tDCS. At baseline, post-camp, and 6-month follow-up, MRIs were performed, and motor function was assessed using the Assisting Hand Assessment (AHA), and Box and Blocks Test (affected hand, BBTA). Task-fMRI was used to localize the motor cortex for MRS voxel placement. GABA-edited MEGA-PRESS was used to measure GABA.

**Results:** Fifty-two participants (AIS  $n=25$ , PVI  $n=27$ ; mean age=10.6 years) were included. Using linear mixed models, the non-lesioned hemisphere showed a significant decrease in GABA across time ( $p=0.048$ ) regardless of tDCS. Motor function significantly increased ( $p<0.001$ ) but was not associated with changes in GABA in either hemisphere ( $p>0.05$ ).

**Conclusions:** Decreased GABA after intervention warrants further research on the role of inhibitory neurotransmission in motor learning in children after PS.

### **P-E-3: Top Risk Factors for Perinatal Stroke: A Comprehensive List [Friday Poster Session, Event Room 1]**

Bithi Roy; Annabel Webb; Karen Walkerb; Catherine Morgana; Nadia Badawi; Iona Novak  
The University of Sydney

**Background:** This comprehensive study provides an in-depth look at the risk factors associated with perinatal stroke and helps you make informed decisions about infants at risk. This study investigated risk factors for term infants (>37 weeks of gestation).

**Methods:** 1) Comparison of prospective population-based perinatal stroke data with the Australian general population data obtained from the Australian Institute of Health and Welfare (AIHW) Australia's Mothers and Babies reports. 2) Case-control study. A multivariate logistic regression model with univariate odds ratios, associated 95% confidence intervals, and stepwise backward variable selection was used.

**Results:** Between 2017 and 2019, 60 perinatal strokes occurred between 37 and 42 weeks of gestation; birth weight 2114-4470 g; 58% male infants, included 95% (57/60) with multiple risk factors. Significant risk factors were smoking during pregnancy (OR 1.48; 95% CI 1.09, 1.99), cesarean section ( $p=0.04$ ), neonatal resuscitation ( $p<0.01$ ), abnormal cord blood gas ( $p<0.01$ ), infection/sepsis ( $p<0.01$ ), congenital heart disease ( $p<0.01$ ), hypoglycemia ( $p<0.01$ ), 1-minute Apgar score <7 (OR 1.54; 95% CI 1.15, 2.08), 10-minute Apgar score <7 (OR 1.26; 95% CI 1.02, 1.54), and hypoglycemia (OR 1.49; 95% CI: 1.07, 2.06).

**Conclusions:** Perinatal stroke is associated with multiple risk factors. Exposure to smoking, 10-minute Apgar score <7, neonatal infection and hypoglycemia were independent risk factors. Emergency cesarean section, resuscitation at birth and abnormal cord blood gas were additional risk factors.

### **P-E-4: PERINATAL STROKE-A CASE STUDY ON STROKE IN UNBORN AND NEWBORN BABIES (PERINATAL STROKE) IN GHANA [Friday Poster Session, Event Room 1]**

Philip Addai, MD  
Ejisu Government Hospital



**Abstract:** Stroke in unborn and newborn babies, also known as perinatal stroke, is a significant concern in Ghana, West Africa, where limited research and awareness exist on this topic. This abstract outlines a study aimed at investigating the prevalence, risk factors, and outcomes of perinatal stroke in Ghana.

**Introduction:** Perinatal stroke refers to strokes occurring between the 20th week of gestation and the 28th day after birth. It can result in long-term disabilities and cognitive impairments if not promptly diagnosed and managed. In Ghana, there is a lack of comprehensive data and research on this condition, hindering early detection and intervention.

**Hypothesis:** We hypothesize that perinatal stroke is an underrecognized issue in Ghana, with potentially significant consequences for affected infants. The study aims to determine the prevalence and identify potential risk factors associated with perinatal stroke in Ghana.

**Methods:** This study will utilize a retrospective cohort design, analyzing medical records and conducting interviews with parents or caregivers of infants diagnosed with perinatal stroke in selected healthcare facilities across Ghana. Data collected will include demographic information, medical history, pregnancy complications, and delivery-related factors. Neuroimaging studies will also be reviewed to confirm stroke diagnoses. **Results:** Preliminary findings suggest that perinatal stroke is more prevalent in Ghana than previously documented, with potential risk factors including maternal hypertension, preeclampsia, and complications during delivery. Infants with perinatal stroke face an increased risk of developmental delays and neurologic deficits.

**Conclusions:** Perinatal stroke in Ghana is a pressing issue that requires increased attention from healthcare providers, policymakers, and researchers.

### **P-E-5: High prevalence of collagenopathies in preterm and term born children with periventricular venous hemorrhagic infarction [Friday Poster Session, Event Room 1]**

Norman Ilves, Sander Pajusalu, Tiina Kahre, Rael Laugesaar; Ustina Samarina, Dagmar Loorits; Pille Kool, Pilvi Ilves  
The University of Tartu, Tartu University Hospital

**Background:** Children with antenatal porencephaly have been shown to have high incidence of pathogenic changes in COL4A1/A2 genes. The etiology of presumed antenatal periventricular venous infarction (PVI) which can also lead to porencephaly is not clear.

**Hypothesis:** The hypothesis was that children with presumed PVI show high prevalence of collagenopathies, as has been found earlier in fetuses and in children with porencephaly.

**Methods:** Genetic analysis and MRI were performed in 85 children: term born children ( $\geq 36$  GW) with antenatal PVI ( $n=6$ ) or presumed antenatal ( $n=40$ ) PVI and preterm children ( $<36$  GW) with periventricular hemorrhagic infarction (PVHI) ( $n=39$ ). Genetic testing was performed using exome or large gene panel ( $n=6700$  genes) sequencing.

**Results:** Pathogenic variants associated with stroke were found in 11/85 (12.9%) children with PVHI/PVI. Among the pathogenic variants, COL4A1/A2 and COL5A1 variants were found in 7/11 (63%) children, two children had pathogenic variants associated with coagulopathy and two other children had other variants associated with stroke. Children with collagenopathies had significantly more often bilateral multifocal stroke with severe white matter loss and diffuse hyperintensities in the white matter and moderate to severe hydrocephalus compared to children with PVHI/PVI without genetic changes in the studied genes ( $p \leq 0.01$ ). Severe motor deficit and epilepsy developed more often in children with collagenopathies compared to children without genetic variants ( $p=0.0013$  and  $p=0.025$ ), respectively.

**Conclusions:** Children with PVHI/PVI have high prevalence of pathogenic variants in collagen genes (COL4A1/A2 and COL5A1). Genetic testing should be considered for all children with PVHI/PVI.

Funded by: Estonian Research Council PRG1912 and the Tartu University Hospital PR-143/22

### **P-E-6: Maternal pyelonephritis as a potential cause of perinatal periventricular venous infarction in term born children [Friday Poster Session, Event Room 1]**

Norman Ilves, Rael Laugesaar; Kristiina Rull, Tuuli Metsvaht; Mare Lintrop, Maris Laan, Dagmar Loorits, Pille Kool, Pilvi Ilves  
The University of Tartu



**Objective:** Pregnancy related risk factors for presumed perinatal periventricular venous infarction (PVI) diagnosed after the neonatal period in term born children are unclear.

**Hypothesis:** Pregnancy related risk factors and their onset time are similar in term born children with presumed periventricular venous infarction and in preterm born children with periventricular hemorrhagic infarction (PVHI).

**Study design:** Antenatal records and pregnancy outcome data were retrospectively assessed in children with presumed PVI (n=43, born <36 gestational weeks, GW) or neonatal PVHI (n=86, born <36 GW) and compared to a matched control group (n=2168, ≥36 GW) from a prospective study.

**Results:** Children with presumed PVI had significantly more frequently maternal bacterial infections compared to the control group (47% vs. 20%, respectively, odds ratio (OR), 3.44 [95% confidence interval (CI), 1.87-63], P<0.0001), whereas no difference was found compared to the neonatal PVHI group (49%, OR 0.91; [95% CI, 0.44- 1.9], P=0.80). Mothers with bacterial infection in the presumed PVI group had significantly more often pyelonephritis compared to the control group (50% vs. 3.4%, respectively, OR, 28 [95% CI, 10-78], P<0.0001). The median gestational age at the diagnosis of maternal infection did not differ between the presumed PVI and neonatal PVHI groups [median (IQR) 26.5 (21-31) vs. 23.5 (21.5-27) GW; P=0.24], but women of the control group had bacterial infection earlier in pregnancy [16.4; 12.4-26.1 GW; P=0.0013].

**Conclusions:** Our data suggest the potential of prompt antibacterial treatment in the case of maternal bacterial infections, especially between GW 21 and 31, for prevention of presumed PVI.

Funded by: Estonian Research Council PRG1912 and the Tartu University Hospital PR-143/22.

### **P-E-7: Necroptosis Blockade Potentiates the Neuroprotective Effect of Hypothermia in Neonatal Arterial Ischemic Stroke [Friday Poster Session, Event Room 1]**

Mathilde Chevin (1), Stéphane Chabrier (2,3), and Guillaume Sébire (1)  
McGill University

NAIS is the most common form of pediatric stroke, imposing a heavy burden on life-long motor, cognitive, and behavioural disabilities. The pathophysiology of NAIS remains largely unknown; hence, there is no evidence-based preventive or curative neuroprotective strategy available for patients affected by NAIS. Our research has revealed that hypothermia (HT) prevented 44% of NAIS occurrences, reduced stroke volume by 37%, and improved motor behaviour in rats. In addition, despite growing awareness of the potential implication of necroptosis (i.e. programmed cell death) in various diseases, its role in NAIS remains elusive. Few preclinical studies have uncovered relevant findings to reinforce the role of necroptosis in NAIS. However, such early mechanisms of cell death are likely to be at the origin of NAIS-induced brain injuries. There is limited understanding of the interplay between necroptotic blockade and HT in this pathology. Altogether, these findings prompted us to focus our research on the unexplored role of necroptosis in NAIS; and more specifically, the combined effect of necroptotic blockade with hypothermia in treating NAIS. Using a preclinical Lewis rat model of term human NAIS induced by unilateral carotid ligation and ischemia, we demonstrated a neuroprotective effect of Necrostatin-1 (Nec-1: a compound blocking necroptosis) in combination with HT. The beneficial effect of Nec-1 combined with HT against brain injuries was observed at the mechanistic level, on both pMLKL and TNF- $\alpha$  and the anatomical level, on brain volume loss visualized by magnetic resonance imaging (MRI). HT alone showed no effect on activated necroptotic effectors. This study opens new avenues for understanding the specific cell death mechanisms in NAIS-induced brain injuries and the potential use of novel therapeutics.

### **P-E-8: Fingerprinting individual differences in lesion impact through imaging: The FIDELITI Dashboard, a patient-centered dashboard of brain health [Friday Poster Session, Event Room 1]**

Helen Carlson, Jordan D. Hassett, Brandon T. Craig, Alicia J. Hilderley, Keith O. Yeates, Melanie Noel, Jillian Miller, Frank P. MacMaster, Signe Bray, Karen Barlow, Brian L. Brooks, Catherine Lebel, Nils Forkert, Adam Kirton  
University of Calgary

**Introduction:** Childhood and adolescence are periods of massive developmental change continuing into early adulthood. Developmental growth charts traditionally measuring age-related trajectories for height and weight have provided insights into normal variation around reference data. Open-source neuroimaging databases have recently afforded development of similar trajectories in brain biomarkers. We introduce the FIDELITI Dashboard (Fingerprinting Individual Differences in Lesion Impact Through Imaging), a patient-centered dashboard that visualizes brain neuroimaging biomarkers at-a-glance. We illustrate the clinical utility of the dashboard by capturing personalized neuroimaging profiles of children with perinatal stroke. Perinatal stroke occurs before the 28th day of life and is the leading cause of hemiparetic cerebral palsy. Deficits in attention, executive function, language, and vision may also occur and early intervention is key.

**Hypothesis:** The FIDELITI Dashboard can identify brain biomarkers deviating from typical trajectories.

**Methods:** The reference cohort was 828 typically developing volunteers aged between 6.5-24.0 years (mean age (SD) = 14.50 (3.75) years) from the Human Connectome Project (n=609) or the Alberta Children's Hospital collaboration (n=219). To illustrate clinical utility, scans of six children with perinatal stroke (mean age (SD) = 11.2 (2.4) years) were additionally processed using the FIDELITI Dashboard. Functionally relevant biomarkers including cortical thickness, region volumes, functional connectivity, and white matter microstructure were extracted.

**Results:** For children with stroke, deviations from the reference cohort (Figure 1, *refer to poster*) were seen for cortical thickness and volumes of lesioned-hemisphere motor cortex, and functional connectivity between motor cortices. Non-lesioned hemisphere metrics often fell within the normal variation of the reference cohort. Non-motor domains also showed deviations, such as functional connectivity for language and executive function networks identifying areas that could potentially be treated with intensive cognitive therapy.

**Conclusions:** The individualized, patient-centered FIDELITI Dashboard can identify deviations from reference trajectories and has potential applications for other neurodevelopmental conditions.

### **P-E-9: The thalamus and basal ganglia are smaller in children with epilepsy following perinatal stroke [Friday Poster Session, Event Room 1]**

Ulvi Vaher<sup>1,2</sup>, Norman Ilves<sup>1,3</sup>, Nigul Ilves<sup>1,3</sup>, Rael Laugesaar<sup>2,4</sup>, Mairi Männamaa<sup>1,2</sup>, Dagmar Loorits<sup>3</sup>, Pille Kool<sup>1</sup>, Pilvi Ilves<sup>1,3</sup>  
University of Tartu

**Objective:** It is still unclear which children develop epilepsy after perinatal stroke. The aim: to evaluate the volume of the thalamus and the basal ganglia in children after perinatal stroke in relation to poststroke epilepsy.

**Methods:** The study included 29 children with perinatal arterial ischemic stroke (AIS), 33 children with presumed periventricular venous infarction (PVI) and 46 controls. MRI was performed at the age of 4-18 years and the volume of the thalamus, caudate nucleus, putamen, globus pallidus, hippocampus, amygdala and nucleus accumbens was evaluated.

**Results:** During a median follow-up time of 12.8 years (IQR: 10.8-17.3) in the AIS group and 12.5 years (IQR: 9.3-14.8) in the PVI group epilepsy developed in 10 children (34.5%) with AIS and in 4 (12.1%) children with PVI,  $p=0.036$  (OR=3.8, 95% CI: 1.04-14). Epilepsy and interictal epileptiform discharges (IED) without clinical seizures were more expressed in children with AIS (n=16, 55%) than in children with PVI (n=7, 21.2%),  $p=0.0057$  (OR=3.8 95%CI: 1.04-14). In the AIS group ipsi- and contralesional thalamus, ipsilesional caudate nucleus and nucleus accumbens were significantly smaller in children with epilepsy compared to children without epilepsy. In the PVI group the ipsilesional thalamus, caudate nucleus, globus pallidus, and nucleus accumbens were smaller in the group of epilepsy plus IED alone, compared to children without epilepsy.

**Conclusions:** In children with AIS, epilepsy or IED occurred more often compared to children with PVI. Patients with AIS and PVI with severe damage to the basal ganglia and the thalamus are at a higher risk for development of epilepsy and should be monitored.

Funding: Estonian Research Council PRG1912, Tartu University Hospital PR-2063, University of Tartu \*Vaher et al. Front Neurol. 2023 Sep 28;14:125247

### **P-E-10: Assessment of perceptual abilities in children with arterial ischemic stroke compared to their typically developing peers [Friday Poster Session, Event Room 1]**

Maiani, M., Carlson, H., Kirton, A.  
University of Calgary

**Introduction:** Perinatal stroke leads to lifelong challenges with daily functioning due to motor, cognitive and visual impairments. Cerebral visual impairment (CVI) is common after arterial ischemic stroke (AIS), but prevalence is largely unknown and screening is not consistently part of standard care pathways. Perceptual skills are vitally important for participation in daily activities, contributing to healthy development through play, school and socializing.

**Hypothesis:** Children with AIS are more likely to score below age-related norms on commonly used perceptual screening tools than typically developing children (TDCs).

**Methods:** 26 children with AIS and 27 TDCs, ages 7-18, were assessed using the Apples test, Motor-Free Visual Perception Test 3rd edition (MVPT-3) and the Jerry Johns Basic Reading Inventory. Parents were given two CVI questionnaires; the CVI teach and the Visual Skills inventory (VSI).

**Results:** Children with AIS had lower percentile ranking on the MVPT-3 compared to TDCs. There was no difference in performance on the Apples test, however children with AIS spent more time scanning for stimuli ( $p=0.002$ ). Conversely, children with AIS were more likely to read below their grade level compared to TDCs ( $p=0.005$ ) with no significant difference in reading speed ( $p=0.25$ ). There was no correlation between higher scores on the CVI teach and perceptual or reading assessment results. Significant differences were present on half of the subcategories of the VSI ( $p<0.03$ ), with parents of children with AIS stating they were more likely to show CVI-like behaviours “sometimes” or “often”, with TDCs showing these behaviours “never”.

**Conclusions:** Children with AIS have lower average performance on standardized tasks of perceptual skills and reading, highlighting the importance of early perceptual screening.

### **P-E-11: Perinatal arterial ischemic stroke: how informative is the placenta? [Friday Poster Session, Event Room 1]**

Joel Fluss, Jessica Hirschel, Francisca Barcos, Anne-Laure Rougemont  
Pediatric Neurology Unit, University Children's Hospital, Geneva

**Introduction:** Neuroplacentology is an expanding field of interest that addresses the placental influence on fetal and neonatal brain lesions, and on further neurodevelopment. The objective of this study was to clarify the link between placental pathology and perinatal arterial ischemic stroke (PAIS). Prior publications have reported different types of perinatal stroke with diverse methodologies precluding firm conclusions and its relevance in clinical practice.

**Methods:** We report here the histological placental findings in a series of 16 neonates with radiologically confirmed PAIS. Findings were grouped into 3 categories of lesions: 1) Inflammation, 2) Placental and fetal hypoxic lesions, and 3) Placentas with a high birthweight/placenta weight ratio. Matched control placentas were compared to the pathological placentas when feasible. The eight term singleton placentas were compared to a series of 20 placentas from a highly controlled amniotic membrane donation program; in three twin pregnancies, the placental portions from the affected twin and unaffected co-twin were compared.

**Results:** Slightly more than half (9/16, 56%) had histopathological features belonging to more than one category, a feature shared by the singleton control placentas (13/20, 65%). More severe and extensive lesions were however observed in the pathological placentas. One case occurring in the context of SARS-CoV-2 placentitis further expands the spectrum of Covid-related perinatal disease.

**Conclusion:** Our study supports the assumption that PAIS can result from various combinations and interplay of maternal and fetal factors and thus confirms the value of placenta examination. Yet, placental findings must be interpreted with caution given their prevalence in well-designed controls.

## P-E-12: Deep medullary vein thrombosis in neonates [Friday Poster Session, Event Room 2]

Fiza Laheji MD, Wilmot Bonnet, MD, Michael Dowling, MD PhD  
UT Southwestern Medical Center

**Introduction:** Deep medullary vein thrombosis(DMV) is a rare cause of neurological damage noted in both term and preterm infants. No clinical trials or definite treatment algorithms are available for DMV thrombosis in neonates. The choice of antithrombotic therapy is therefore based on the experience of the single centers or on indications extrapolated from available guidelines on CSVT.

**Aims:** Recently published systematic review on DMV thrombosis in the neonates found a total of only 75 cases in the literature. In none of these cases was treatment options were described. We aim to present a institute experience describing 3 cases of neonatal DMV thrombosis and different management methods.

**Methods and findings:** Between July -August 2023, we prospectively studied 3 neonates with deep medullary vein thrombosis as evidenced by characteristic hemorrhage noted on MRI. 1st patient- no anticoagulation was initially started given co-morbid conditions and critical illness. However, repeat imaging showed worsening of hemorrhage, thereby indicating progression of thrombosis resulting in venous hemorrhage. Pt was therefore started on anticoagulation with LMWH. No worsening hemorrhage was noted after. 2nd patient- Anticoagulation was not started, and no clot progression was noted. 3rd patient- Anticoagulation was started from a cardiac standpoint. No worsening in hemorrhage was noted.

**Observation:** Anticoagulation in patient with progression of DMV thrombosis did not worsen existing hemorrhage and prevented further propagation. In patient with no worsening thrombosis, absence of anticoagulation did not result in worsening of thrombosis and likely self-resolved. Anticoagulation used for a different indication did not worsen intraparenchymal hemorrhage.

**Next steps:** To conduct a retrospective chart review on a larger cohort of patient to analyze further the use on anticoagulation for DMV thrombosis.

## Risk Factors and Prevention

### O-F-2: Pediatric Craniocervical Arterial Dissection: Clinical Presentation, Risk Factors, Neuroimaging and Pediatric NIH Stroke Scale in a Series of Chilean Patients [Oral Abstract Session]

Fernanda Balut, María José Hidalgo, Mario Matamala, Daniela Muñoz, Susana Lara, Diane Vergara, Eduardo Bravo, Mónica Troncoso  
Hospital Clinico San Borja Arriaran, Universidad de Chile

**Introduction:** Craniocervical arterial dissection (CCAD) is among the most reported cerebral arteriopathies associated with pediatric arterial ischemic stroke, with variable clinical presentation depending on the location and the extent of the lesion. Neurovascular imaging is necessary to confirm the diagnosis of CCAD.

**Objectives:** To describe clinical presentation, risk factors, Pediatric NIH Stroke Scale (PedNIHSS) and neuroimaging in a series of pediatric cases with stroke due to arterial dissection.

**Methods:** Retrospective, descriptive study. Children with diagnosis of arterial dissection, aged between 29 days and 18 years were studied from 2001 to 2018. Clinical characteristics, risk factors and neuroimaging were described. PedNIHSS severity score was estimated for patients older than 4 months of age.

**Results:** Eight patients, 6 male. Median age at stroke was 5.3 years old. Most frequent onset symptoms were headache (6/8), focal motor deficit (4/8) and ataxia (4/8). Stroke was more frequent in posterior circulation (5/8). All patients had at least one risk factor, and history of trauma was present in 5 cases. Two patients had an abnormal thrombophilia study. All patients received either heparin or AAS. Median PedNIHSS was 4.5. MRI showed ischemic stroke in all patients. Vascular imaging demonstrated narrowing or occlusion of the arterial lumen. In 3 patients Vessel Wall Study

demonstrated hyperintense lesion of the vessel wall with eccentric contrast enhancement. Conventional angiography findings included vessel occlusion, stenosis, intimal flap and pseudoaneurysm.

**Conclusion:** Our series demonstrated a male predominance, with posterior circulation more frequently compromised. Headache is a frequent symptom of onset, and history of trauma is usually present. CCAD should be suspected when a patient with these characteristics presents to the ED, and vascular studies must be performed to confirm the diagnosis.

### **P-F-1: Connective Tissue Disease and Dissection: A Rare Case of Pediatric Stroke in Beals Syndrome [Saturday Poster Session, Event Room 1]**

Carolina Salazar, Megan Barry, Angeliq Mercier, Joshua Baker, Carlos Prada, Vamshi Rao, Divakar S Mithal  
Lurie Children's Hospital of Chicago

**Introduction:** Dissecting aneurysms of the vertebral or cervical artery cause approximately 2% of all strokes but represent nearly 20% of strokes in younger patients. Trauma is thought to cause dissection, but in children, atraumatic dissection is also common. The etiology of idiopathic or spontaneous dissection remains unclear, but one leading risk factor is thought to be underlying connective tissue disease (CTD). CTD are heterogeneous in clinical presentation and while Vascular Ehlers-Danlos and Marfan Syndrome are the most common disorders associated with dissections, there are other CTDs that may lead to stroke.

**Case:** We present a case of a seven-year-old female who was admitted with headache, altered mental status and focal left sided weakness. On MR imaging, she was found to have diffusion restriction of the right pons as well as occlusion of the basilar artery. Subsequent vessel wall imaging demonstrated enhancement of the right vertebral artery, initially concerning for atypical focal cerebral arteriopathy vs dissection. A broad stroke workup did not identify a definitive etiology. During follow up, she was noted to have residual mild hemiparesis with notable joint hyperlaxity and had a history of congenital cataracts. A CTD gene panel was sent that revealed a paternally inherited variant of unknown significance in FBN2. FBN2 is known to cause the autosomal dominant CTD Beals syndrome, however, the patient did not have the typical congenital contractures and her father was unaffected. Despite only rare reports of dissection with FBN2 variants, the patient subsequently developed contractures consistent with a mild form of Beals syndrome suggesting it contributed to her dissection and stroke.

**Discussion:** The case highlights the complexity in diagnosing CTDs associated with stroke, even when the suspicion is high.

### **P-F-2: Introduction to a Customized Pediatric Neurovascular Disease and Stroke Gene Panel: A Practical Molecular Genetics Approach for Clinicians [Saturday Poster Session, Event Room 2]**

Elissa Engel MD, J Michael Taylor MD, Yaning Wu, Sudhakar Vadivelu, Wenying Zhang PhD  
Cincinnati Children's Hospital Medical Center

**Introduction:** The underpinning molecular mechanisms of pediatric stroke and vascular malformations are complex. As diagnostic whole exome sequencing become more readily available, affordable, and covered by third party payers, the opportunity to employ this technology in routine diagnostic evaluation is emerging. We detail the clinical disorders, techniques, and decision support available for a new customized pediatric gene panel.

**Methods:** A whole exome panel with 80 genes selected for pediatric disorders causing vascular malformations, ischemic stroke, intracranial hemorrhage, vasculopathy, and hemiplegic migraine was developed in house. Analysis is completed using Human Comprehensive Exome kit from Twist Bioscience capturing exonic regions of genes from the genomic DNA extracted from the patient. Variants within exons and flanking sequences are identified and evaluated by a validated in-house pipeline including consultation with a stroke specialist and geneticist.

**Results:** We will detail the patient demographics, clinical indications, and results for panels completed in 18 months after test launch in July 2022. Results are classified as pathogenic/likely pathogenic or variants of uncertain significance based on available published literature, genetic databases (ClinVar), and in-silico modeling by following the 2015 ACMG



recommended variant classification guideline. The majority of exams identify variants of unknown significance for further clinical correlation.

**Conclusions:** A pediatric neurovascular disease and stroke gene panel can provide valuable information to clinicians. Identification of a causative genetic etiology can allow individualization of treatment and counseling as it relates to disease progression, risk of recurrence, and screening for other syndromic associations.

### **P-F-3: Infection and pediatric Arterial Ischemic Stroke with Focal Cerebral Arteriopathy: data from the Covid-19 pandemic [Saturday Poster Session, Event Room 2]**

Raluca Tudorache, Manon Jaboyedoff, Aurelie Gabet, Valerie Olié, Francois Angoulvant, Philippe Tuppin, Thomas Lesuffleur, Manoelle Kossorotoff  
University Hospital Necker-Enfants Malades

**Introduction and Hypothesis:** Infection may trigger pediatric arterial ischemic stroke (AIS) related to Focal Cerebral Arteriopathy (FCA). Sanitary measures (non-pharmaceutical interventions NPIs) during the Covid-19 pandemic resulted in a major decrease in pediatric viral infections. We explored the consequences on the incidence of pediatric AIS with FCA.

**Methods:** Using national public health databases, we identified children hospitalized between 2015 and 2022, with AIS presumably related to FCA using age proxy (29 days-7 years) and excluding other stroke causes, including cardiac and hematological conditions. Considering the delay between infection and AIS, we compared a pre-pandemic reference period, a period with NPIs, and a post-NPIs period.

**Results:** Interrupted time-series analyses of the monthly incidence of pediatric AIS due to FCA showed a significant decrease in the NPIs period compared to the reference period: -33.5%, 95%CI [-55.2%;-1.3%], p=0.043.

**Conclusions:** These data support the association between infection and FCA in pediatric AIS.

### **P-F-4: Fibrocartilagenous Embolism Causing Cervical Spinal Cord Infarction with Supratentorial Involvement in an Adolescent Male [Saturday Poster Session, Event Room 2]**

Barbra Giourgas MD, Cyril Lukianov, MD, Thomas Koch, MD  
Medical University of South Carolina

**Introduction:** Spinal cord infarcts (SCI) are an extremely rare cause of myelopathy in children, with few cases reported in the literature. Etiology is often unknown, and fibrocartilagenous embolism (FCE) is one unusual mechanism. FCE is thought to be herniation of the nucleus pulposus of vertebral discs, which induces prolapse of cartilagenous material into neighboring vasculature. We present a case of an adolescent male with cervical SCI with multifocal supratentorial strokes, presumed to be due to FCE after minor trauma. To our knowledge, there are no cases of a child with cervical SCI and supratentorial strokes due to FCE.

**Case:** A previously healthy 15-year-old male presented with right neck pain, emesis, dizziness, and bilateral upper extremity (UE) numbness and weakness after playing football. Physical exam revealed 3/5 strength in the proximal left UE, 0/5 in the proximal right UE, with 4/5 distal strength bilaterally, diminished touch and temperate sensation and absent reflexes bilaterally. Lower extremities were not involved. MRI brain and spine showed multifocal infarcts within the right cerebellum, bilateral parietal and occipital lobes and left thalamus, and a SCI at C2-C4. Vascular imaging was negative. Disc irregularity was seen from C3-C6 suggesting paravertebral cartilagenous embolism as a source. ASA 81 mg was started, and sensation returned to normal with slight improvement in strength.

**Discussion:** While SCI represent only 1% of adult strokes, their occurrence in children, especially with supratentorial involvement, is even rarer. Given intervertebral disc abnormalities, infarct characteristics and absence of vascular irregularities, this was thought to be due to a thromboembolic phenomenon from a FCE. To our knowledge, there are less than 10 reports of pediatric SCI due to this mechanism and none with cerebral involvement. SCI due to FCE is likely under-recognized in pediatrics, but timely recognition and management is imperative to improve outcome.



### **P-F-5: Ischemic Stroke Reduction in Children with Sickle Cell Disease and Moyamoya Syndrome Treated with Acetylsalicylic Acid: Subgroup Analysis of the Stroke in Sickle Cell Revascularization Surgery (SiSCRS) Study [Saturday Poster Session, Event Room 2]**

Philipp R Aldana, P. David Adelson, Shih-Shan Lang, Paul Grabb, Stephanie Greene, James Johnston, Jeffrey Leonard, Suresh N Magge, Neena Marupudi, Joseph Piatt, Rafael de Oliveira Sillero, Edward R Smith, Jodi Smith, Jennifer Strahle, Sudhakar Vadivelu, John C Wellons III, David Wrubel, Aleksandra S Dain, Lauren A Beslow, Asmaa Hatem, Salvador Gutierrez, Otavio de Toledo, Nicklaus Millican, John M Pederson, Ricardo A Hanel  
Wolfson Children's Hospital/University of Florida Jacksonville

**Objective:** To evaluate the effect of acetylsalicylic acid (ASA) therapy on risk of cerebrovascular events (CVEs) in pediatric patients with sickle cell disease and moyamoya syndrome (SCD-MMS).

**Methods:** This is a multicenter, retrospective study of children with SCD-MMS treated with routine care (chronic blood transfusion and/or hydroxyurea) and ASA therapy during periods both routine care alone (RC) and with the addition of antiplatelet therapy (RC+A). We monitored CVEs rates—a composite of strokes and transient ischemic attacks (TIAs). Andersen-Gill extended Cox models were used to compare cumulative incidence of CVEs, accounting for variation in treatment over time, age at treatment onset, time of moyamoya diagnosis, and CVE history. Additional analyses were performed by CVE type, including ischemic strokes and TIAs. Subgroup analyses were performed among patients receiving RC that did not undergo revascularization surgery (Non-Surgical group or RCNS) and patients identified as surgical candidates during their pre-surgical phase (Pre-Surgical group or RCPS).

**Results:** 90 patients treated with ASA were included. Between treatment regimens, there was no significant difference in CVEs of any type (RC=14.4 vs. RC+A=11.6 events per 100 patient-years) or TIAs (RC=2.6 vs. RC+A=3.1 events/100 patient-years). However, RC+A treatment periods were associated with significantly lower ischemic strokes compared to RC alone (RC=10.1 vs. RC+A=5.8 events/100 patient-years), with an instantaneous risk of ischemic stroke roughly half that compared to treatment periods consisting of conservative management alone (HR=0.49, 95% CI: 0.26-0.91; p=0.025). Within the Non-Surgical subgroup treatment periods with antiplatelets were also associated with reduced ischemic strokes (RCNS=9.5 vs. RCNS+A=2.4 events per 100 patient-years), with an instantaneous risk of ischemic stroke roughly 4-fold lower than compared to treatment periods consisting of conservative management alone (HR=0.27, 95% CI: 0.09-0.79; p=0.018). Like the primary analysis, there were no significant differences in incidence of overall CVEs or TIAs. Conversely, within the Pre-Surgical subgroup receiving routine care, treatment regimens had a similar incidence of overall CVEs (RCPS=18.0 vs. RCPS+A=20.2), ischemic strokes (RCPS=10.8 vs. RCPS+A=10.1), and TIAs (RCPS=4.8 vs. RCPS+A=5.0 events/100 patient years). Despite differences in treatment effects on CVEs between the two patient subgroups, interaction tests did not yield any statistically significant interactions between treatment and patient subgroup for any of the CVE types (any CVE: p=0.104, ischemic stroke: p=0.072, TIA: p=0.577), possibly indicating an underpowered analysis or that the disparities in treatment responses are influenced by other unaccounted variables that differ between subgroups.

**Conclusion:** Our analysis supports the efficacy of acetylsalicylic acid therapy in reducing ischemic strokes among children with SCD-MMS. While our findings suggest a potentially heightened effectiveness of antiplatelet therapy in mitigating stroke risk among less severe cases of SCD-MMS where revascularization surgery is not warranted, evidence is limited. Larger cohorts, ideally with prospective and randomized controlled designs, are warranted to further elucidate these findings.

### **P-F-6: Acute Stroke Care in Nigerian Children with Sickle Cell Disease: Training Curriculum for HealthCare Professionals in Low-Income Settings [Saturday Poster Session, Event Room 2]**

Djamila L. Ghafuri, Halima Bello-Manga,<sup>2</sup> Richard Carozza, Fenella J. Kirkham, Mariana Ciobanu, Edwin Trevathan, Michael R. DeBaun, and Lori C. Jordan  
Vanderbilt University Medical Center

**Objective:** Nigeria has the highest proportion of children with sickle cell anemia (SCA) globally; an estimated 150,000 infants with SCA are born annually. Without primary prevention, 10% will have a stroke in childhood. Knowledge gaps exist in recognizing pediatric acute strokes in low- and middle-income countries without robust emergency medical systems, limited neurologists and neuroimaging modalities. As part of NIH-funded SPRING and SPRINT trials,<sup>1,2</sup> a sustainable Sickle Cell Disease Stroke Prevention Teams program<sup>3</sup> was established to address the need for stroke care in northern Nigeria. We describe the health professional stroke training curriculum and specific application in low-resource settings.

**Methods:** Children aged 2-16 years with SCA (HbSS or Hb S $\beta$ O thalassemia) were enrolled in SCD primary and secondary stroke prevention trials in northern Nigeria (SPRING and SPRINT trials, respectively)<sup>1,2</sup> and were found to have clinical strokes during study follow-up. As neuroimaging was not available in this region until just as the trials were nearing completion, diagnosis of clinical stroke depended on WHO criteria. Physicians and nurses were trained in-person and via video lectures in performing neurological exams using the adapted PedNIHSS (Figure 1, *refer to poster*).<sup>4</sup> We collected and reviewed the study case report forms and recorded videos of the neurological examinations.

**Results:** A total of 6 general medical officers or pediatricians completed the curriculum successfully. A total of 8 acute primary strokes and 11 acute-on-chronic strokes were identified in SPRING (N=220) and SPRINT (N=101) trials, respectively (Table 1, *refer to poster*) by our trained study personnel. Challenges identified included distinguishing acute from chronic stroke, stroke mimics, and modesty concerns during the neuro exam with clothing obscuring the exam in female children.

**Conclusions:** We developed a curriculum to train healthcare providers in pediatric acute stroke recognition and care in a low-resource setting, initially training physicians with translatable, context-appropriate interventions. Work led by Nigerian physicians is ongoing to train nurses and community health workers to recognize acute stroke in children with SCA.

### **P-F-7: Epidemiology of cerebral venous thrombosis in children with acute bacterial intracranial infections: a single institution retrospective study [Friday Poster Session, Event Room 2]**

Nehali Mehta, MD<sup>1</sup>; Sanjeev Swami, MD<sup>2</sup>; Mark Halverson, MD, FRCPC<sup>3</sup>; Leah Loerinc, MD<sup>1</sup>; Claudia Gambrah-Lyles, MD<sup>1</sup>; Jennifer McGuire, MD, MSCE<sup>1</sup>  
Children's Hospital of Philadelphia

**Introduction:** Cerebral sinovenous thrombosis (CSVT) is a known complication of acute bacterial intracranial infection.

**Objectives:** To determine the annual proportion of, risk factors for, and treatment patterns for CSVT in a cohort of children with acute bacterial intracranial infections.

**Methods:** Retrospective single-center cohort study of children age 1-18 years hospitalized at a tertiary care children's hospital for acute bacterial intracranial infection between 01/01/2015 and 02/01/2023. Cases were identified by discharge ICD-10 codes for "meningitis," "meningoencephalitis," "subdural empyema," "epidural abscess," and "brain abscess." Medical charts were manually screened to ascertain the diagnosis and to abstract relevant clinical data.

**Results:** One hundred and three patients were included in this study. Median age of the cohort was 10.2[5.9,12.7] years. 31/103 (30%) had CSVT. Annual proportion of CSVT did not change during the study period; however, the total number of cases is rising. Need for neurosurgical intervention(aOR=11.6, p=0.027), need for ENT intervention(aOR=6.1, p=0.013), temporal location of infection(aOR=7.9, p=0.004), and concurrent mastoiditis(aOR=29.5, p=0.004) were associated with development of CSVT in this cohort. The sigmoid sinus(55%, 17/31), transverse sinus(42%, 13/31), jugular vein(45%, 14/31), and superior sagittal sinus(45%, 14/31) were the most common locations for CSVT. Two patients(6%) had evidence of venous infarction; one(3%) had evidence of venous hemorrhage. 81%(25/31) of patients with CSVT received anticoagulation for a median of 90[56,106] days. Lack of thrombus resolution was associated with occlusive thrombus(p=0.003), number of sinuses involved(p=0.006), transverse sinus thrombosis(p=0.021), and jugular vein thrombosis(p=0.031). All children with CSVT underwent follow-up imaging, last performed at a median of 43[25,95] days following thrombus detection. 65%(20/21) children demonstrated thrombus resolution at a median of 36.5[22,50] days.

**Conclusion:** CSVT is common in acute bacterial intracranial infection. Consider empiric surveillance in children with acute bacterial intracranial infection with mastoiditis, temporal location of infection, and/or those who require surgical intervention. Optimal duration of anticoagulation in this population should be further explored.

### **P-F-8: Cerebrovascular Dysfunction in Children with Congenital Heart Disease - a plausible mechanism of brain injury [Saturday Poster Session, Event Room 2]**

Amanda Robertson, David Mikulis, Brian McCrindle, Trish Domi, Nomazulu Dlamini  
Hospital for Sick Children

**Introduction:** Previous studies have shown widespread abnormal perfusion injury in children with congenital heart disease (CHD) in the absence of stroke. However, the mechanism causing these global changes is unknown.

**Hypothesis:** In this study, we hypothesized that children with CHD would have abnormal cerebrovascular reactivity (CVR) compared to healthy controls as indicated by fractional negativity (fneg) a quantitative indicator of CVR.

**Methods:** Ten healthy controls and five CHD/no stroke patients underwent CVR BOLD-MRI. fneg values were obtained in the left and right hemisphere for gray matter, white matter, and whole brain (combined white and gray matter) tissue regions, and compared between groups. The non-parametric Mann-Whitney U test was used to determine statistically significant group differences ( $p < 0.05$ ).

**Results:** Of the patients studied, 60% had moderate/complex congenital heart lesions and lesions involving cyanosis. Both gray and white matter fneg were greater in the CHD group compared to healthy controls and all hemispheric comparisons for gray matter yielded statistically significant between group differences. Left sided gray matter was greater ( $p = 0.008$ ) in the CHD group ( $0.016i, \pm 0.0069$ ) compared to healthy controls ( $0.0071i, \pm 0.0034$ ), and right sided gray matter was also greater ( $p = 0.028$ ) in the CHD group ( $0.015i, \pm 0.0074$ ) compared to healthy controls ( $0.0068i, \pm 0.0042$ ). Statistically significant hemispheric differences in fneg ( $p < 0.05$ ) were also found for the white matter and whole brain tissue (combination of white and gray matter), between groups.

**Conclusions:** This data suggests that children with CHD/no stroke, have abnormal CVR compared to neurotypical controls. We suggest that these abnormal cerebral hemodynamics contribute to stroke risk and cognitive outcome in CHD.

### **P-F-9: Diffusion Weighted Imaging Characteristics in Deep White and Gray Matter Structures in Pediatric Patients with Moyamoya Disease [Saturday Poster Session, Event Room 2]**

Ahmed Mohamed, Loxlan Kasa, Alhusain Abdullah, Amanda Robertson, Liza Pulcine, Mahendranath Moharir, Peter Dirks, Pradeep Krishnan, Prakash Muthusami, Nomazulu Dlamini

**Background:** Moyamoya Disease (MMD) is a rare steno-occlusive arteriopathy associated with transient ischemic attacks (TIA), stroke, and cognitive impairments. In adult MMD, normal-appearing deep gray and white matter structures (NADWM) show diffusion and volume changes that are associated with cognitive impairments. Apparent diffusion coefficient [ADC] measures the mean diffusion and is sensitive to these changes. The association of ADC and volume changes in NADWM in pediatric MMD is not clear. Our objective is to determine whether ADC is elevated and volume is reduced in NADWM in pediatric MMD patients.

**Hypothesis:** ADC values are higher and volume is lower in NADWM of MMD patients compared to controls.

**Methods:** Twenty-nine MMD patients with no stroke and seven healthy controls (HC) were retrospectively analyzed from a single center. Diffusion and T1-weighted images were collected and co-registered. A script implementing a convolutional neural network (CNN) based approach was used to segment and binarize cortical regions of interest (ROI) to allow extraction of ADC values (units =  $[x10^{-6} \text{ mm}^2/\text{s}]$ ). Volumes were calculated in these ROIs including: 1) all white matter (WM), and NADWM, including: 2) thalamus, 3) caudate, 4) putamen, 5) globus pallidus, 6) hippocampus. Mean ADC values and volumes were compared for: 1) all ROIs between MMD and controls and 2) all ROIs in the MMD affected and the contralateral hemispheres within the MMD group with unilateral disease.

**Results:** Patients were, on average, 7.7±5.4 years at MMD diagnosis. In all WM, MMD patients had significantly higher ADC values than controls in the left (mean= 865.7±101.6 versus 762.4±21.6 [ $\times 10^{-6}$  mm<sup>2</sup>/s], p=0.01) and right hemispheres (mean= 869.9±104.9 versus 761.0±21.3 [ $\times 10^{-6}$  mm<sup>2</sup>/s], p=.01), respectively. In normal appearing deep gray matter NADGM, no significant differences were found between the MMD and control groups in any of the ROIs, including the thalamus, caudate, putamen, globus pallidus and hippocampus. In the NADGM of MMD patients, higher ADC values were found in the putamen (mean= 778.2±46.0) $\times 10^{-6}$  mm<sup>2</sup>/s], p=0.041), hippocampus (mean=1009.5±58.6 [ $\times 10^{-6}$  mm<sup>2</sup>/s], p=0.005) and all NAWM (mean= 860.3±57.0 [ $\times 10^{-6}$  mm<sup>2</sup>/s], p<0.0001) in the MMD affected compared to the contra-lesional hemisphere. In MMD patients, volumes were significantly higher in cerebral white matter volume in the unilateral (232423.00±/-34925.75mm<sup>3</sup>) compared to bilateral (194293.41±/-37592.73 mm<sup>3</sup>, p=.01) MMD.

**Conclusions:** In pediatric patients with MMD, we found elevated ADC values in deep gray and white matter structures in the diseased hemisphere. We also found higher volumes in cerebral white matter of unilateral compared to bilateral disease patients. Our ongoing analysis integrates hemodynamic profiles, cognitive outcomes, and the associations with diffusion characteristics in this MMD cohort.

### **P-F-10: Atlantoaxial instability, Arterial Dissection, and Ischemic Stroke in Down Syndrome: A Silent Cascade? [Saturday Poster Session, Event Room 2]**

Abhijit Das, MD; Karla Salazar, MD; Manasa Sudheendra, MD; Daniel Davila-Williams, MD  
Baylor College of Medicine

**Introduction:** Down syndrome (DS) is associated with various stroke risk factors, including congenital heart disease, moyamoya vasculopathy, malignancy, hypothyroidism, and obstructive sleep apnea. While rare, craniocervical arterial dissection is a key consideration in this patient population given their predisposition to atlantoaxial instability (AAI). In this case report, we discuss a pediatric patient with DS presenting with multiple ischemic strokes secondary to vertebral artery dissection (VAD).

**Case Presentation:** An 11-year-old girl with trisomy 21, closed atrial septal defect and history of right-sided Bell's palsy presented with acute difficulty walking, horizontal nystagmus and left-sided weakness. Initial NIHSS was 6. Brain MRI revealed evidence of acute ischemic stroke of the right pons and left cerebellum. CTA revealed vessel wall irregularity of the left vertebral artery at C1-C2, suggestive of VAD. Hypercoagulability workup was unrevealing. She was started on enoxaparin and transferred to inpatient rehabilitation before being discharged home. Though clinically stable, a repeat brain MRI 4 months later showed new areas of ischemia involving the left pons with dissecting aneurysm of the left vertebral artery at the V3 segment on CTA. Neck CT noted mildly dysmorphic C1 and C2 vertebrae with a mildly hypoplastic posterior C1 arch. On cerebral angiography, vasculopathy was further characterized as eccentric fusiform dilation of the distal left vertebral artery. Aspirin was then started in addition to enoxaparin, which she continues presently

**Discussion:** Though the exact mechanism of VAD secondary to AAI is unknown, it is proposed that increased stress on the arterial wall caused by repeated motion, bending, and stretching of the vertebral artery secondary to C1-C2 instability may increase the chance of dissection. Further work on determining causality could be beneficial, particularly among patients with DS given their baseline predisposition to AAI.

### **P-F-11: Idiopathic Intracranial Hypertension (IIH) and Management/Mismanagement of Associated triggers [Friday Poster Session, Event Room 2]**

Moyenda Joseph ,Fenella Kirkham  
University College London

In idiopathic intracranial hypertension (IIH) there are signs and symptoms consistent with raised intracranial pressure despite normal brain parenchyma. Underlying causes, e.g. malignancy or ventriculomegaly must have been excluded. The key aim of our Service Evaluation, approved by University Hospital Southampton (UHS) Child Health, was to

determine the extent to which triggers such as high body mass index (BMI) or iron deficiency anaemia (IDA) had been excluded in IIH patients and, if present, whether appropriately managed. The (UHS) CHARTS computerised system was used to retrospectively obtain documentation relating to paediatric patients with IIH between 01/01/2000 and 31/12/2019. The Data retrieved included Lumbar puncture opening pressure, Body Mass Index (BMI), full blood count (FBC), indices of Iron Deficiency Anaemia, Acetazolamide or steroid used was gathered as well as previous thrombotic events. The data were analyzed using SPSS compared to the Friedman's revised criteria and National Institute for Clinical Excellence (NICE) guidelines for treatment. Of 52 Patients BMI was recorded in 46 (median age=12.9 (M), 13.3 (F) with a median BMI of 25 (range 14.4-53.4). There was an average of 0.5 BMI increase from initial presentation to final discharge from Neurology (Figure 1, *refer to poster*). Of the 22 overweight/obese patients, 10 were prescribed Topiramate, 7 of whom had relief of symptoms. Of the 22 patients with low indices (Table 1, *refer to poster*), 10 were prescribed iron and there was relief in IIH symptoms for 7. Key limitations include missing data and/or limited follow up information. Patients with FBC with indices consistent with IDA (low haematocrit or mean cell volume) should have ferritin and transferrin measured and should be considered for iron supplementation or a change in diet. More emphasis should be on promoting weight loss as this intervention was poorly documented amongst our patients. Topiramate might be combined with Acetazolamide.

### **P-F-12: Stroke in Pediatric-onset Systemic Lupus Erythematosus: An Atypical Case [Friday Poster Session, Event Room 2]**

Michael Matt MD<sup>1</sup>, J. Michael Taylor MD, <sup>1,2</sup>, Minal Aundhia MD<sup>1</sup>, Cheryl Bayart MD<sup>1,2</sup>, Marguerite M. Care MD<sup>1,2</sup>, Lara Berklite MD<sup>1,2</sup>, Sarah Kitts MD<sup>3</sup>, Oscar F. Lopez-Nunez MD<sup>1,2</sup>, Sudhakar Vadivelu DO<sup>1,2</sup>, Ekemini A. Ogbu MD MSc <sup>1,2,4</sup>

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**Introduction:** Stroke is a cause of major morbidity and mortality in pediatric-onset systemic lupus erythematosus (pSLE). The presence of anti-phospholipid antibodies and hypertension are known risk factors for stroke events in pSLE. We present a patient with pSLE who initially presented with overall lower SLE disease activity (SLEDAI-2K) and lacking these known risk factors that developed concurrent hemorrhagic stroke and cerebral venous sinus thrombosis.

**Case:** A 16-year-old, previously healthy female presented with 6-months of unintentional weight loss and tender, subcutaneous nodules on both legs. Labs showed ANA titer 1:1280, positive Ro, Smith, and RNP antibodies, and mildly low C4 level. Antiphospholipid antibodies and lupus anti-coagulant were negative. Her SLEDAI-2K was 6 (active disease range is 6 - 105). She was diagnosed with pSLE, placed on hydroxychloroquine, mycophenolate mofetil and oral steroids. Nodule punch biopsy showed inflammatory changes (Figure 1). Weeks later, she developed sudden, severe headache from an acute hemorrhagic stroke with intraventricular extension (Figure 2). Cerebral angiography showed segmental narrowing and dilation of all medium-sized anterior and posterior circulation arteries suggesting vasculopathy (Figure 3). However, brain and superficial temporal artery biopsies did not show vasculitis. During recovery, she developed extensive thrombosis of the left transverse, sigmoid sinus, and proximal internal jugular vein. She improved on treatment with cyclophosphamide and rituximab infusions. At 8-month follow-up, she reports mild left-sided weakness but no interruption in school and work. In pSLE, vasculitis in superficial, peripheral, and CNS vessels can occur concurrently. CNS vasculitis can occur with relatively lower overall SLE disease activity without antiphospholipid antibodies. Further exploration of biomarkers demonstrating active cerebrovascular disease are needed in pSLE.

### **P-F-13: A case of Post-Herpes Encephalitis Vasculopathy Causing Right MCA and PCA Stroke in a 13-month-old with RNF213 mutation [Print Only]**

Rachel Pearson MD; Rutu Dave MD, Janetta Arellano MD, Charles Grose MD



Herpes viruses are a known risk factor for arteriopathy and stroke in pediatric patients. Many genetic risk factors have also been identified that may predispose certain children to stroke. We describe a case of a patient who developed a right middle cerebral artery and posterior cerebral artery stroke with a history neonatal HSV-1 encephalitis (HSE) and a pathogenic RNF213 mutation. She was initially diagnosed with HSE at 2 weeks of age. During her first year of life, her sister was diagnosed with moyamoya disease with a pathogenic RNF213 mutation. Our patient and other family members were found to have the same mutation through familial genetic testing. When the patient was 13 months old, one month after discontinuing acyclovir prophylaxis, she had recurrence of HSE with associated vasculopathy and stroke. She later also developed post-HSE anti-N-methyl-D-aspartate receptor encephalitis. Neonatal HSV-1 infection, as compared to HSV-2, is less severe, and it is rare to have relapse after discontinuation of antiviral prophylaxis. Immunosuppression, however, may increase the risk of HSE recurrence. Recent studies also suggest a potential role of single gene mutations in increasing individual's susceptibility even in the absence of immunosuppression. RNF213 has been implicated in multiple pathways of innate immunity, inflammation, and angiogenesis. Our patient's vascular imaging did not demonstrate features consistent with moyamoya disease, however, we hypothesize that the RNF213 mutation still may have played a role in her the pathogenesis of her stroke by virtue of diminished immunocompetence. This is the first case, to our knowledge, of a child presenting with stroke secondary to herpes vasculopathy and a known carrier for the RNF213 mutation.

#### **P-F-14: Monogenic causes of Arteriopathies and Arterial Ischemic Stroke in the Pediatric Population [Print Only]**

Vivek Pai, Manohar Shroff, Carmen Parra-Farinas, Prakash Muthusami, Pradeep Krishnan  
The Hospital for Sick Children

**Introduction:** Ischemic stroke in children has an estimated incidence 1 per 100,000 individuals. In contradistinction to their adult counterparts, genetic causes of stroke form a large subgroup of children sufferings from arterial narrowing and stroke. Late recognition of stroke or an underlying predisposing condition may limit access to critical interventions and preventive therapies. These patients are also prone to increased psycho-functional disabilities and long-term seizures. The purpose of this poster is to provide an exhaustive imaging review of monogenic causes of large and small vessel arteriopathies and arterial ischemic stroke in the pediatric population.

**Methods:** A generic literature review was performed. CT and MRI studies performed for evaluation of patients with suspected and confirmed arteriopathies and /or arterial ischemic stroke were reviewed. Only patients with confirmed genetic mutations were included in this review.

**Findings:** The following conditions will be discussed in this poster: A. Large and / or Medium Vessel Steno-occlusions: 1. Moya-Moya Disease (RNF213 and BRCC3 gene) 2. Moya-Moya like disease: Sickle Cell Disease; Neurofibromatosis type 1; Morning Glory Syndrome; Down Syndrome 3. PHACES 4. Grange syndrome 5. Alagille Syndrome 6. ACTA-2 Mutation 7. Generalized Arterial Calcification of Infancy (GACI) B. Small Vessel pathologies 1. COL4A mutations 2. DAD2 mutation 3. Aicardi-Goutieres Syndrome

**Conclusion:** Early recognition of genetic etiologies of stroke may facilitate prompt testing and initiation of specific preventive measures while directing further evaluation of affected, carriers and at-risk individuals in families.

#### **P-F-15: Recurrent Posterior Circulation Strokes in a 15 year-old Male: Atypical Focal Cerebral Arteriopathy or Mechanical Arteriopathy? [Print Only]**

Michael Kung, MD, Maximillian Halabi, Rachel Pearson, MD

Focal cerebral arteriopathy (FCA) is a common etiology for pediatric arterial ischemic stroke (AIS). While classically characterized by unilateral, anterior circulation stenosis, posterior circulation FCA has been described. Rotational vertebral arteriopathy is also associated with posterior circulation AIS, particularly in young males. We describe a 15-

year-old male with recurrent posterior circulation strokes. He presented with acute left-sided numbness. Imaging showed bilateral posterior circulation infarcts with right posterior cerebral artery (PCA) stenosis and distal occlusion. Aside from recent cough, history was unremarkable. Labs showed: PCR with (+) Coronavirus-229E, CRP 20.5 mg/L, ESR 17 mm/hr, ANA+. Other labs, echocardiogram, and CSF were unrevealing. He was discharged on aspirin. He returned after 1 week for right upper quadrantanopia, correlating with new bilateral PCA-territory infarcts. Angiography showed bilateral PCA occlusions, concerning for possible vasculitis. Despite high-dose steroids, symptoms worsened, and new strokes developed. Given pure posterior circulation involvement, rotational vertebral arteriopathy was considered and c-collar placed. Rotational angiography failed to demonstrate dynamic stenosis or occlusion. Cervical X-rays and whole genome sequencing were unrevealing. Treatment was escalated to dual antiplatelet therapy (DAPT). He clinically improved with stable imaging and was discharged on DAPT and steroid taper. 6 weeks later, he returned with dysarthria, left limb ataxia and weakness. Imaging showed a new left cerebellar infarct with vessel wall thickening in the left vertebral artery (VA). Anticoagulation was added (replaced aspirin). 4-month follow-up imaging showed persistent left VA irregularity/occlusion but was otherwise stable. This case demonstrates the importance of maintaining a broad differential in children with posterior circulation AIS, as correct diagnosis informs approach to treatment and secondary prevention.

## Stroke Readiness and Guidelines

### **P-G-1: Designing the Pediatric Stroke Standard Operating Procedure (SOP): a Necessary Destination Despite a Challenging Journey [Saturday Poster Session, Event Room 2]**

Kriti Bhayana MD, Sumit Parikh MD, Ravi Talati, MD  
Cleveland Clinic

**Introduction:** Stroke has been recognized as an adult health problem, but it is also an important cause of acquired brain injury in neonates and children. Despite national guidelines, pediatric centers, have traditionally lacked the infrastructure to triage, diagnose, and treat childhood arterial ischemic strokes quickly. Writing pathways and order-sets can help ensure that the pediatric stroke codes are effective.

Often, when there is an acute stroke alert in a pediatric unit at an integrated hospital system, it has been unclear in terms of which team needs to be activated (pediatric neurology versus adult acute stroke service) resulting in delays in initial treatment. Thus, there was an urgent need for a new pediatric stroke Standard Operating Procedure (SOP).

**Hypothesis:** An SOP will streamline acute as well as long-term management of pediatric stroke patients.

**Methods:** A team comprising of Pediatric Neurology, Hematology, Pediatric ICU, Cardiology and Cerebrovascular (CV) Center was setup. Latest national guidelines, prior in-house pediatric stroke care-path, and current best practices in various pediatric hospital systems were reviewed. An elaborate Pediatric Stroke SOP was thus created.

**Results:** The SOP includes recognition of stroke symptoms (in pediatric and neonatal population), appropriate imaging and labs, acute treatment, detailed diagnostic tests for determining stroke etiology, long-term management, nursing protocols for monitoring in the ICU (PICU and CVICU) and pharmacy protocols for anticoagulant reversal. The SOP is currently under modification after initial review by the above-mentioned teams, but available to the pediatric neurology trainees and staff as a reference tool. With this document, the trainees endorse a higher level of confidence managing these patients, particularly in the acute setting.

**Discussion:** This SOP will facilitate the evaluation, management, and treatment of an acute pediatric stroke, as well as guide clinicians in long-term care of these patients. It will also help in appropriate utilization of resources and limit redundancy of testing in light of new research.

After modification, the project is intended for pre-implementation simulation testing, and it will also undergo quality control prior to Enterprise-wide circulation.

### **P-G-2: Ischemic Stroke in Pediatric patient following Chiropractor Manipulation [Friday Poster Session, Event Room 2]**

Jonathan Arnold MD, Kyle Ortiz MD, Rabia Qaiser MD  
Indiana University

Pediatric ischemic events following Chiropractor manipulation poorly understood and under documented phenomenon. Here we present a case of a 20-month-old male with a past medical history of asthma and poor weight gain that developed to the hospital with intense vomiting, lethargy, and hypoxia following neck manipulation by a chiropractor the day prior. A computed tomography scan at presentation showed crowding of the basal cisterns, sulcal crowding, low lying tonsils, and hypoattenuation of the right posterior occipital lobe. Magnetic resonance imaging showed right posterior cerebral artery territory diffusion restriction with smaller foci of restricted diffusion in the left occipital lobe, left basal ganglia, cerebellar hemispheres, and left anterior temporal lobe. Magnetic resonance angiography and venogram of the head showed no arterial large vessel occlusion or venous thrombi. However magnetic resonance angiography of the neck demonstrated significant tortuosity of the right vertebral artery. Echocardiogram was negative for vegetations or emboli. During hospital stay also found to be febrile and viral work up showed patient to be positive for Coronavirus HUI1 and Rhinovirus/Enterovirus. Patient was treated with dual antiplatelet therapy. After an extended hospital course patient was able to discharge and subsequent repeat magnetic resonance scans showed no worsening of ischemic territories and subsequently downgraded to aspirin alone. This report illustrates how chiropractor manipulations can lead to ischemic events.

### **P-G-3: Dynamic susceptibility contrast perfusion imaging using a novel contrast agent: Transient hypoxia-induced deoxyhemoglobin [Friday Poster Session, Event Room 2]**

Ece Su Sayin, Olivia Sobczyk, James Duffin, Julien Poublanc, David J. Mikulis & Joseph A. Fisher  
University of Toronto

**Background:** Dynamic susceptibility contrast for cerebral perfusion imaging requires the intravascular injection of paramagnetic agents such as gadolinium (Gd). Here we describe the use of transient hypoxia-induced changes in deoxyhemoglobin (dOHb) as a paramagnetic MRI contrast and compare the perfusion metrics obtained to those using the clinical standard, Gd.

**Methods:** We studied 8 healthy controls, 8 patients with steno-occlusive disease, and 8 patients with low-grade glioma using a 3-Tesla MRI scanner running standard BOLD sequences. Transient hypoxia was induced via an automated gas blender running a feed-forward gas algorithm targeting 2 reductions of pulmonary PO<sub>2</sub> from 95 mmHg to 40 mmHg, followed by full reoxygenation within a single inhalation. A second BOLD sequence was acquired following an intravenous injection of 5 ml of Gd. Resting perfusion metrics were calculated using a standard tracer kinetic model using deconvolution of an arterial input function measured from the middle cerebral artery.

**Results:** All patients showed abnormal patterns of resting perfusion metrics with the same individual regional variations when receiving dOHb as with Gd. In patient populations, both techniques allow for the distinction between healthy and affected hemispheres consistent with the patient's clinical notes. There was no significant difference between resting perfusion measures (MTT, rCBF and rCBV) obtained using transient hypoxia induced dOHb and GBCA (one-way ANOVA  $P > 0.05$ ).

**Conclusions:** The resting perfusion measures obtained from brief transient hypoxia are spatially and quantitatively comparable to those obtained using Gd in the same healthy controls and patients. The advantages of transient hypoxia as a contrast agent include elimination of injections (non-invasive), no risk of allergic reaction, no risk of tissue toxicity in patients with renal insufficiency, and no environmental accumulation of potentially toxic Gd into the environment.

### **P-G-4: EARLY BRAIN INJURY IN GHANA: A COMPREHENSIVE CASE STUDY [Friday Poster Session, Event Room 2]**

Elizabeth Oti

Korle Bu Teaching Hospital

This paper delves into the critical issue of early brain injury in Ghana, presenting a comprehensive case study that investigates the prevalence, contributing factors, and the socio-economic impact of this public health concern. The study employs a mixed-methods approach, combining quantitative and qualitative data collection methods to provide a nuanced understanding of the complexities surrounding early brain injury in the Ghanaian context. The results reveal the multifaceted nature of this problem, shedding light on the challenges faced by affected individuals and their families. The paper concludes with insights into potential interventions and policy recommendations aimed at mitigating the adverse effects of early brain injury and improving overall public health in Ghana.

## Vascular Malformations and Hemorrhagic Stroke

### **O-H-1: Endoluminal sampling in situ enables genetic characterization of vein of Galen Malformation [Oral Abstract Session]**

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**Introduction:** Vein of Galen malformation (VOGM) is the result of aberrant arteriovenous shunting between choroidal/subependymal arteries and the embryologic prosencephalic vein of Markowski and is amongst the most severe cerebrovascular disorders of childhood. While endovascular treatments have improved outcomes, morbidity and mortality remain high.

**Objective:** We hypothesized that in situ analysis of the VOGM lesion using endoluminal tissue sampling (ETS) would enable identification of somatic mutations and transcriptional programs, expanding our understanding of VOGM genetics, pathogenesis, and maintenance.

**Method:** We utilized a Mendelian, trio-based study design, collecting germline DNA (cheek swab) from patients and their families. Flow cytometry was used to isolate endothelial cells (EC), defined as CD31+ and CD45-, obtained from explanted coils during ETS in conjunction with routine endovascular treatment. Whole-exome sequencing (500x coverage) of germline and EC (somatic) DNA was performed to identify inherited, de novo, and somatic mutations. We also form the collaborative Vein of Galen Malformation Genetics Research Consortium (VOGM-GRC) to aggregate VOGM cases for large-scale genetics analysis.

**Result:** Our cohort contains 24 patients (9 with VOGM, 6 of whom are African American), including 6 complete trios, where ETS was performed 11 times in 6 patients from both arterial and venous embolizations. Yield from ETS varied from 3 to 254 experimental cells. We identified de novo and inherited mutations in at least 1 patient in 572 and 189 genes, respectively, including novel and previously implicated genes in cerebrovascular biology. Pathway analysis identified regulation of the extracellular matrix as a potential shared mechanism. We also identified a mutation in Ephb6 in two separate families (inherited and de novo) which is predicted to disrupt Ras/MEK/ERK signal transduction.

**Conclusion:** Genetic analysis of VOGM identifies potentially clinically actionable genetic alterations. Efforts to optimize WES methodology from low-input DNA content obtained through ETS are needed. ETS, when safe to undertake, may add to the armamentarium of pediatric cerebrovascular providers and may inform development of a molecular-genetic taxonomic classification enabling rationale selection of personalized pharmacotherapeutics.

### **P-H-1: Cerebral Proliferative Angiopathy - Literature Review and Single Institute Experience [Friday Poster Session, Event Room 2]**

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**Objective:** We aim to share a case series of 5 patients with Cerebral Proliferative Angiopathy(CPA) and different management methods used for these refractory cases.

**Background:** Cerebral proliferative angiopathy is a rare phenomenon. There is limited data in the literature on the management and natural history of this condition. Formerly known as the “diffuse nidus” or “holohemispheric giant cerebral arteriovenous malformation (AVM),” it is morphologically considered to be a congenital vascular anomaly. According to the Stroke paper published in 2008, CPA was defined as an atypical entity that appears to differ significantly from normal brain AVMs. Until 2016 per review published in "Interventional Neuroradiology", only a total number of 74 cases had been reported thus far in the literature.

**Design/Methods:** We conducted a retrospective chart review at our institution and reviewed patients with a diagnosis of cerebral proliferative angiopathy, confirmed on neuroimaging. We reviewed different parameters such as : age of diagnosis, history of infarct or bleed, treatments used, repeat imaging to evaluate for progression of disease, development of seizures, latest PSOM score.

**Results:** All 4 of the patients in our cohort were Male and were noted to have refractory disease resulting in ischemic strokes and intracerebral hemorrhages. They were treated with gama knife Vascular embolization and also the novel use of rapamycin and sirolimus in one of the cases. All 4 of the patients were on AEDs for seizure management.

**Conclusions:** CPA is a rare disease with limited data in the literature on treatment guidelines and the progression of disease. By highlighting our single institute experience of 4 cases of pediatric CPA, and the novel use of sirolimus, rapamycin we aim to shed light on this condition and help develop treatment guidelines in the future.

#### **P-H-4: Complex pediatric cerebral arterial venous malformation treated with trametinib [Friday Poster Session, Event Room 2]**

Wilson, J.L., Selden, N.R., Collins, K.L., Murphy, B., Nesbit, G.M., Cho, Y.J.  
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**Introduction:** High grade pediatric cerebral arterial venous malformations (AVM) can be challenging to treat. With the discovery that AVMs result from somatic mutations in the RAS/RAF/MAPK signaling pathway, there have been reports of successful treatment of extra-CNS and spinal cord AVMs with the MEK inhibitor, trametinib.

**Case:** This male patient was diagnosed in-utero with an inoperable deep cerebral AVM. At age 6 years, he underwent radiation with 18 Gy. At age 9, he developed a hemorrhage resulting in right hemiplegia. He had a subacute neurological decline beginning around 13 years of age, prompting return to care at age 15. Neurological examination showed fluctuating alertness, hemiplegia, dystonia, and parkinsonism. His imaging showed progression of his Spetzler-Martin grade IV AVM and cerebral atrophy. Chronic ischemic steal and venous hypertension likely caused his decline. The interdisciplinary neuro-vascular team judged that he was not a candidate for surgical or interventional therapy and that radiotherapy was high-risk. In April 2022 he started trametinib 1 mg daily which he tolerated well except for an acneiform rash. He remained clinically stable with no change on annual MRI and MRA until he developed a non-catastrophic cerebral hemorrhage in June 2023, likely due to a venous thrombosis as the location of the bleed was remote from the nidus (figure). Compared to imaging prior to trametinib treatment, the post-hemorrhage angiogram showed slightly diminished flow through the AVM. He continues on trametinib as further radiation treatment is considered.

**Discussion:** High grade cerebral AVMs pose significant treatment challenges. Targeted medical therapy with MEK inhibitors is being explored for systemic and spinal AVMs. The current patient tolerated trametinib well with possible mild improvement in angiographic characteristics of his cerebral AVM. Further study is needed to elucidate the role of MEK inhibitors in the treatment of cerebral AVMs.

#### **P-H-5: Four Complex Aneurysms in the first four years of life [Friday Poster Session, Event Room 2]**

Flavio Requejo



**Introduction:** Aneurysms in children are very infrequent in the first years of life. Most of them are dissecting in nature. Endovascular treatment is a minimally invasive option to treat these patients. Localization and age of the patient (growth of the cerebral arteries) are relevant to determine the best treatment option.

**Cases:** Two male children had dissecting lesions in the M1 segment of the MCA (1 year and 4 years old. Fig 1 and 2). One male had a post-surgical aneurysm in the M4 segment of MCA (5 months of age, Fig 3). One female (18 months old. Fig 4) had a top basilar dissecting aneurysm. Intracranial bleeding was the presentation in three and mass effect in one patient with aneurysm in the M4 segment of MCA (Fig 1) Flow Diverter stent was used in the 4-year-old child with M1 dissecting aneurysm. Occlusion of the parent artery and the aneurysms was undertaken in the other three children. Total aneurysm exclusion from circulation was achieved in all the patients. The follow-up showed a very good clinical outcome.

**Discussion:** Cerebral Aneurysms in little children are rare and frequently complex. A common etiology is dissection making treatment difficult because many times a normal cerebral artery involved in the lesion must be occluded. Flow diverter stents are an option after 4 years of age because of the growth of cerebral arteries.

**Objective:** To point out that endovascular treatment is a safe and effective technique but must be tailored in every patient taking into account localization of the lesion and the age of patients.

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### **P-H-6: “Infantile dural arteriovenous fistula” - a reconsideration [Friday Poster Session, Event Room 2]**

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**Background:** “Infantile type” dural arteriovenous shunts (IDAVS) are rare and heterogeneous vascular lesions, complicating their classification and management. The current tripartite classification of pediatric dural arteriovenous shunts (DAVS) into dural sinus malformation (DSM), “infantile type,” and “adult type,” does not stand up to scrutiny, given the variable presentations of the latter two types in children.

**Objective:** To estimate the prevalence of IDAVS and evaluate the long-term outcomes post-endovascular treatment (EVT).

**Methods:** A retrospective review of a pediatric cerebrovascular database between 2006-2023 was conducted. Patient data was analyzed to evaluate the presentation and long-term outcomes of IDAVS.

**Results:** IDAVS were identified in 8(0.5%) of 1463 patients, with mean age at diagnosis of 34.7 months; males comprised 62.5%. Most common clinical presentations included macrocephaly (37.5%), seizures (25%), and dilated scalp veins (25%). EVT was performed in 87.5% of cases, averaging 5.8 procedures per patient. Radiographic obliteration was observed in 28.6%. Good clinical outcomes (mRS  $\leq$  2) were achieved in 85.7%. Our findings showcased discrepancies and limitations in the current classification of pediatric DAVS, prompting a re-evaluation.

**Conclusion:** IDAVS accounted for a small proportion of pediatric cerebrovascular pathologies, with markedly heterogeneous presentations. Stepwise selective embolization was associated with favorable outcomes (in a higher proportion than reported in other published cohorts) and is recommended over an aggressive approach aimed at complete angiographic obliteration. Our proposed revised classification system bifurcates pediatric DAVS into DSM and all other DAVS that are manifest in children, thereby enhancing diagnostic clarity and therapeutic approaches.

### **P-H-7: Lectin-type Oxidized LDL Receptor-1 as a Potential Biomarker in Cerebral Cavernous Malformations [Friday Poster Session, Event Room 2]**

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Boston Children's Hospital

**Background:** Cerebral cavernous malformations (CCMs) are clusters of thin-walled capillaries with mutational profiles that promote abnormal proliferation, angiogenesis, and bleeding and which can present with seizure, headache, or neurological deficits. LOX-1 (lectin-type oxidized LDL receptor 1) is a 50kDa transmembrane protein that has been implicated in endothelial cell dysfunction and ischemia. Here, we present data demonstrating that LOX-1 may have potential utility as a non-invasive biomarker able to identify the presence of CCMs.

**Methods:** CCM plasma (n=10) and urine samples (n=23) were collected from pediatric CCM patients (age=1-24yo). Matched healthy controls (n=24) were collected from pediatric patients (age=1-19yo) with Chiari I malformation or fatty filum, and otherwise normal findings with full imaging of the brain and spine. All samples were collected with patient/family consent and institutional review board approval. Samples were analyzed with Olink Proteomic proximity extension assay (PEA). The differences in expression for over 2,900 unique proteins were quantified between healthy control and CCM plasma and urine samples. The results were normalized, validated, and analyzed for potential age and sex bias.

**Results:** ANOVA analysis of the CCM plasma and urine samples showed a statistically significant increase in LOX-1 compared to the control samples, with CCM patients exhibiting >2-fold increases in plasma expression (p<0.0034) and >5-fold increases in urinary expression (p<0.0012).

**Conclusions:** LOX-1 is involved with pathways implicated in CCM pathogenesis and our data reveals that plasma and urinary levels of LOX-1 are significantly elevated in CCM patients as compared to matched controls. These data suggest that LOX-1 has potential as a putative biomarker for CCM and may merit further investigation related to its potential mechanistic impact on CCM pathogenesis.

### **P-H-8: Endoscopic Third Ventriculostomy with possible Cauterization is Safe and Effective for treatment of Hydrocephalus in previously embolized Vein of Galen Malformations [Friday Poster Session, Event Room 2]**

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Boston Children's Hospital

**Introduction:** Vein of Galen Malformations (VOGM) are arteriovenous malformations in young children and neonates. Patients can present with communicating hydrocephalus due to underlying venous hypertension. Rarely, patients present

with obstructive hydrocephalus due to direct mass effect of the malformation on the cerebral aqueduct. Given the limited literature in management of embolized VOGM patients with persistent hydrocephalus, we evaluate the safety and efficacy of endoscopic third ventriculostomy (ETV) in this patient population.

**Methods:** We conducted a single institution retrospective review of all VOGM patients who presented with worsening hydrocephalus despite treated malformation via embolization and underwent ETV from 2022 - 2023.

**Results:** 4 patients met inclusion criteria. Average age at the time of ETV was 5 months (2-11 months). Two patients (50%) had obstruction of the aqueduct on preoperative imaging. All patients underwent prior endovascular embolization, average number of embolizations 2.75 (range 2 to 5 embolizations) with no further targetable sites and minimal flow within the malformation at final embolization. All patients had evidence of worsening hydrocephalus based on preoperative FOHR trends (average 0.63 +- st.dev 0.07) and Evans' Index (0.62 +- 0.1). One patient underwent cauterization at time of ETV without adverse events. There were no complications after ETV, with an average of 5% (1.5%-15%) reduction in FOHR at the most recent followup, average 2.6 months after ETV (0-7.7 months). Our ETV success rate is 75%. One patient with minimal reduction (1.5%) in FOHR is under consideration for permanent CSF diversion.

**Conclusion:** We demonstrate safety and efficacy of ETV in VOGM patients, with improvement in hydrocephalus in the majority of patients.

### **P-H-9: Not just a port wine stain birthmark: PHACE syndrome presenting in a premature neonate [Saturday Poster Session, Event Room 2]**

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**Introduction:** PHACE (posterior fossa anomalies, hemangioma, arterial anomalies, cardiac anomalies and eye anomalies) syndrome is a rare complex of intra/extracranial vascular anomalies with an undetermined genetic basis. In this case, we present a premature infant with a facial cutaneous lesion initially concerning for Sturge-Weber syndrome (SWS) but later found to have extensive vascular anomalies confirming a diagnosis of PHACE syndrome.

**Case Presentation:** A premature infant delivered via C-section at 27 weeks GA due to preterm labor, placental abruption and breech presentation was noted to have a facial cutaneous lesion involving a right V1 distribution. Neurology is consulted for a concern for SWS with initial workup revealing a normal head ultrasound and EEG. Genetic testing is negative for GNAQ variants. At 2 months of life (35 weeks GA), the patient's cutaneous lesion evolves and grows with ulceration at the inferior margin, suggestive of a facial hemangioma and prompting concern for PHACE syndrome given the presence of supraumbilical raphe and a sternal cleft on exam. MRI brain and vessel imaging revealed a small right cerebellar hemisphere, right orbital hemangioma, right cavernous sinus hemangioma, dural arteriovenous fistula draining into bilateral transverse sinuses, absent left internal carotid artery, as well as restricted diffusion along the frontal and peritrial medullary veins. At 3 months of age, she exhibits mild gross motor delay.

**Discussion:** This case demonstrates a rare presentation of PHACE syndrome in a premature infant with extensive intracranial vascular anomalies. While current guidelines shed light on the use of anti-platelet agents for primary arterial ischemic stroke prevention, further investigation is needed in the management of dural AV fistulas and venous malformations in this age group. It additionally highlights the need to maintain a broad differential in the evaluation of facial cutaneous lesions in neonates.