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Abstract #1: Genetic Impact on Neurodevelopment in Non-syndromic Craniosynostosis

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Background/Purpose: Next generation sequencing technologies have permitted a molecular understanding of the patho-etiology of non-syndromic craniosynostosis (NSC), however the underpinnings of the subtle neurocognitive deficits observed in nearly half of all children with NSC remains largely unexplored. Using an integrative genomic approach, we sought to characterize how mutations identified in craniosynostosis might impart the neurodevelopmental phenotypes observed in children with NSC.

Methods/Description: We analyzed exome sequencing data from 500 individuals, including 404 case-parent trios, with craniosynostosis to identify novel genes and pathways implicated in the patho-etiology of NSC. Clinical data related to surgical intervention was available for each participant. Neurodevelopmental data from 256 cases with matched exome sequencing data was used to assess the potential contribution of high-risk genotypes to neurodevelopmental outcomes. Bulk RNA-sequence data were used to build a spatiotemporal expression map of the developing brain, and scRNA sequence data was used to identify cell types expressing genes of interest at critical neurodevelopmental timepoints. We assessed for enrichment of genes implicated in craniosynostosis in each spatiotemporal module of neurodevelopment.
Results:

We replicate a previous association of damaging de novo mutations in Wnt, BMP, and FGF/MAPK signaling to NSC, and identify novel mutations leading to epigenetic and transcriptional dysregulation. Patients with high-risk genotypes (damaging de novo and transmitted LOF mutations in intolerant genes; pLI>0.9) had significantly higher rates of neurodevelopmental delays than those who did not harbor these mutations. When stratifying patients based on the presence or absence of high-risk genotypes, there were no significant differences in neurocognitive outcomes between patients who had open vs. endoscopic surgical interventions. Transcriptomic analyses incorporating mutations identified via exome sequencing in NSC probands suggest that depletion of mid-fetal (post conception weeks 8-21)

stem cell populations affecting osteoblast differentiation, neural differentiation, and meningeal fibroblast proliferation underlie premature suture fusion and aberrant neurodevelopment.

Conclusions: Craniosynostosis is a neurodevelopmental disorder in which genetic perturbations dysregulate both neurogenesis and suture homeostasis. Exome sequencing, neurodevelopmental testing, and integrative transcriptomics provide multidimensional insight into the patho-etiology of both suture fusion and aberrant neurodevelopment. The results suggest that genetics may impact neurodevelopmental outcomes more than the type of surgical intervention performed and provide a framework for further large-scale investigation of genetic and neurodevelopmental anomalies in NSC.

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Abstract #2: Computer-assisted Virtual Surgical Planning and Robot Guided Laser Osteotomy for Frontoorbital Advancement – A Full Digital Workflow

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Raphael Guzman: none relevant to this study
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Marco Keller: none relevant to this study
Daniel Seiler: none relevant to this study
Christoph Kunz: none relevant to this study
Jörg Beinemann: none relevant to this study
Philipp Honigmann: none relevant to this study
Jehuda Soleman: none relevant to this study
Florian M. Thieringer: none relevant to this study

Objective: Robot-assisted surgery is leading a shift from traditional surgery to computer-assisted patient-specific interventions. This in-vitro lab study aims to investigate the potential of an established virtual surgical planning (VSP) workflow in combination with robot-guided laser osteotome system and medical three-dimensional (3D) printing. The goal was to analyse the feasibility to guide osteotomies and remodelling of the frontal bone in frontoorbital advancement (FOA) surgery. The process was simulated on patient specific 3D printed models.

Material and Methods: A computed tomography (CT) of a representative case of unilateral coronal synostosis was selected. The CT DICOM dataset was segmented, and VSP (MIMICS Innovation suite, Materialise, Leuven, Belgium) was performed using "mirroring" and superimposing an age-matched normative 3D skull model. A virtually predefined cutting plan with several non-linear freeform osteotomy paths (defined henceforth as "SMART" cuts) was transferred to the CARLO® system (Advanced Osteotomy Tools, Basel, Switzerland). 3D printed medical-grade bioresorbable thermoplastic biomaterial, RESOMER® Composite (Evonik Industry AG, Essen, Germany) was used for modelling. The printing time and accuracy

of the implants were noted. The implants and osteotomized models were digitized using an optical 3D scanning system (EinScan-SP, SHINING 3D Tech. Co., Ltd., Hangzhou, China) and compared with the virtual planning data.

Results: The CARLO® device performed the “SMART” cuts in 30 mins. The registration process had a root-mean-square error (RMSE) of 1.0 mm. There was close conformance between the planned and performed robot-guided FOA procedure in a lab setup. The implants had a good fit and smooth finish with an accuracy (RMSE) of 0.60 mm.

Conclusion: The digital workflow from virtual planning to laser osteotomy successfully demonstrate the integration of several novel technologies. In the future the system could perform "SMART" functional incisions and osteotomies in the frontal bone of babies undergoing FOA or ither cranial remodelling procedures with patient-specific bioimplants.

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Abstract #3: "How Low Should We Go? Safety and Craniometric Impact of the Low Occipital Osteotomy in Posterior Vault Remodeling"

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Background: A larger volume cranial vault expansion is likely facilitated by a low posterior cranial osteotomy beneath the torcula; however, this may impart an increased risk of venous bleeding. This study compares the safety infra-torcular to supra-torcular osteotomy in patients undergoing posterior vault reconstruction (PVR) or posterior vault distraction osteogenesis (PVDO). We also analyzed volumetric changes.

Methods: Patients undergoing initial PVR and PVDO between 2009-2021 at our institution were grouped by occipital osteotomy location, and retrospectively analyzed. Craniometric analysis was performed if patients had high-resolution CT scans available within 180 days pre-and post-operatively.
Results: A total of 187 patients were included: 106 (57%) who underwent PVDO, and 81 (43%) PVR. Infra-torcular osteotomy was more common in PVR (n=65, 80%) than PVDO (n=61, 58%; p<0.002.). Blood transfused was similar between low and high osteotomy cohorts in PVDO (p=0.285) and PVR (p=0.342). However, median transfused blood volume per kilogram of patient weight was greater in the low- vs high-osteotomy PVDO (p=0.010) and PVR (p=0.041) cohorts. Intra-operative venous sinus injury was rare. In the PVDO cohort, there was increased median intracranial volumetric gain in the low (263 mL) compared to high-osteotomy cohort (127 mL; p=0.043); however, when controlled for distraction distance only a trend was observed (p=0.221). Patients undergoing PVDO showed a larger median intracranial volume increase (168 mL) compared to those undergoing PVR (73 mL; p<0.001).

Conclusion: Infra-torcular osteotomy can be performed safely in most patients undergoing cranial vault remodeling, and does not appear associated with greater hemodynamic instability or sinus injury.

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Abstract #4: A comparison of neurodevelopmental outcomes in patients with who were treated with CVR vs endoscopic repair for sagittal craniosynostosis

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All authors have no relevant disclosures.

Introduction: Optimal surgical management of nonsyndromic sagittal craniosynostosis continues to be debated. Several studies have compared perioperative parameters and postoperative morphology of open cranial vault remodeling (CVR) to endoscopic-assisted strip craniectomy (ESC) for the treatment of sagittal craniosynostosis. However, data on neurodevelopmental outcomes comparing these techniques is lacking.

Methods: A multi-institutional retrospective neurodevelopmental study was conducted of school-age children (age 5-15 years) who had previously undergone CVR or ESC. Sites included Children's National Hospital (Washington, DC) and St. Louis Children's Hospital (St. Louis, MO). Patients diagnosed with syndromes affecting neurodevelopment were excluded. Neurodevelopment was measured using the General Cognitive Ability (GCA) score from the Differential Ability Scale-II (DAS-II).

Results: Sixty-four patients [17 CVR (5 Female:12 Male), 47 ESC (12 F:35 M)] were studied. Mean age at DAS-II testing was 8.2 ± 2.1 years. Mean GCA scores were within the normal range for both groups and group differences were not statistically significant (CVR 95.7 ± 16.1 vs ESC 102.7 ± 14.9; $p = 0.108$). A regression model including repair type, age at assessment, and Hollingshead total score (SES) as predictors of GCA had $R^2 = 0.32$. After adjusting for SES and age at DAS-II assessment, repair type was not significantly associated with GCA score ($p = 0.776$, $\beta = 1.14$, 95% CI [-6.84, 9.12]).

Conclusion: Interim analysis of the preliminary data suggests no significant association between type of surgical procedure and cognitive outcomes at school age in patients with sagittal craniosynostosis who had previously undergone CVR or ESC. As this is an interim analysis, conclusions are limited by the relatively small sample of patients who underwent CVR. Collection of data continues, with the goal to enroll 50 participants in each group.

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Abstract #5: Comparative Analysis of 2D and 3D Metrics for Evaluation of Postoperative Outcomes Following Endoscopic Suturectomy for Sagittal Craniosynostosis

Mauricio Medina MD, Eseosa Odigie, Michelle Buontempo NP, Jessica Feeney, Fiona Quinn, Caitlin Hoffman MD, and Thomas Imahiyerobo MD

Introduction: To assess whether 3D volumetrics can be used to track and evaluate postoperative course and outcomes of patients treated with endoscopic suturectomy for sagittal synostosis, we compared post-operative changes in 2D measurements with 3D volumetrics.

Methods: Patients treated with endoscopic suturectomy for sagittal synostosis from 2014 to 2021 were retrospectively reviewed. Head circumference (HC), cephalic index (CI), and total cranial volume (TCV) were measured using SmartSoc (Orthomerica) scans at three postoperative timepoints (mean + SEM): initial (25 $\pm$ 8 days), intermediate (122 $\pm$ 12 days), and final (198 $\pm$ 13 days). Differences in rate and total change in CI, HC, and TCV were performed.

Results: Forty-three patients underwent endoscopic suturectomy, 12 had at least 3 postoperative scans for helmeting. The mean CI, HC and TCV for the initial, intermediate, and final measurements were: CI = 70, 76 and 76.3; HC = 431, 455 and 469 cm; and TCV = 697, 838 and 929 ml, respectively. There was a direct correlation between CI and TCV ($r=0.67$, $p<0.001$), and HC and TCV ($r=0.78$, $p<0.001$). The mean total change was ΔCI = 6.18, ΔHC = 38 cm, and ΔTCV = 232 ml. All measurements showed significant differences between timepoints on the analysis of variance ($p<0.001$). Overall rate of change was higher for TCV (33.9%) than for CI (8.9%) and HC (8.8%) ($p<0.001$). TCV continued to demonstrate significant rate of change between intermediate and final timepoints (11.1%) compared to CI (0.3%) and HC (3.1%). There was a trend toward decreased rate of change in TCV for patients treated at an older age.

Conclusions: TCV demonstrates the highest rate of sustained change over the course of treatment compared to 2D metrics. Volumetrics may therefore be able to better define optimal timing of treatment and completion of helmet therapy in future and larger scale study.

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Abstract #6: Hemoglobin Disrupts Microglial IGF-1 Production in the Developing Brain

Brandon A. Miller, MD, PhD, FAANS

Intro: Intraventricular hemorrhage of prematurity (IVH) interrupts normal white matter development. During normal brain development, microglia play an important role in myelination by producing insulin-like growth factor 1 (IGF-1) which is necessary for oligodendrocyte progenitor cells (OPCs) to mature into myelinating oligodendrocytes. IGF-1 is produced by a subpopulation of microglia that express the antigen CD11c, and these microglia are enriched during early brain development.

Objective: To determine if hemoglobin altered the percentage of microglia that expressed CD11c and if hemoglobin reduced microglial IGF-1 production *in vitro* and *in vivo*.

Methods: A cell culture system of isolated microglia was used for *in vitro* studies. An established rat model of neonatal IVH was used for *in vivo* studies. Magnetic bead sorting was used to isolate $CD11c^+$ microglia. Real-time PCR was used to quantify IGF-1 expression. ELISA was used to measure IGF-1 and cytokine production. Immunohistochemistry was used to quantify white matter and label OPCs, microglia, and CD11c.

Result: *in vitro*, hemoglobin reduced the percentage of microglia that expressed CD11c, and reduced the IGF-1production measured by ELISA. In an animal model of neonatal IVH, where hemoglobin injection reduces white matter development but not OPC survival, $CD11c^+$ cells decreased their IGF-1 production as measured by real-time PCR.

Conclusion: Hemoglobin is a potent pro-inflammatory stimulus for microglia in the developing brain. Hemoglobin induces classically proinflammatory microglial activation, with production of cytokines and reduction of IGF-1 production. $CD11c^+$ microglia, the cells that provide the IGF-1 critical for myelination, reduce IGF-1 production *in vivo* when exposed to hemoglobin. This loss of microglial IGF-1 may be the reason OPCs fail to mature and myelinate effectively. Replacing IGF-1 maybe a therapeutic to protect white matter after neonatal IVH.

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Abstract #7: Younger Age at Spinal Cord Detethering is Potentially Associated with a Reduced Risk of Curve Progression in Children with Early Onset Scoliosis

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Introduction: In children with early onset scoliosis (EOS) who also have tethered spinal cord (TSC), spinal cord detethering procedures are commonly performed prior to spinal deformity surgery as curve stabilization or improvement may occur in some patients following the detethering procedure alone. However, there is a lack of data regarding the role of factors such as patient age or the degree of curve magnitude at the time of detethering, and their effect on subsequent scoliosis progression.

Aims/Objectives: The purpose of this study was to investigate whether EOS patient age or curve magnitude at the time of spinal cord detethering was associated with curve progression at a follow-up of at least 2 years. It was hypothesized that patients who undergo detethering at a younger age, or those with a smaller curve magnitude, would experience a reduced rate of curve progression compared to those who are older or with larger curves.

Methods: In this retrospective cohort study, patients with EOS who underwent spinal cord detethering at least 2 years prior to index surgical intervention for their spinal deformity were identified in a multicenter international registry. Radiographs were assessed just prior to the detethering procedure (pre-detether) and at the most recent visit prior to any spinal deformity surgery (most recent post-detether). Rates of curve progression >10°, stabilization within 10°, and improvement >10° were examined. Due to unequal follow-up in individual patients, Cox regression was utilized to investigate associations between primary variables (age and magnitude of major coronal curve) and rate of curve progression.

Results: 37 patients met inclusion criteria, and 18 (mean age: 3.7 ± 2.9 years, 66.7% female, mean follow-up: 3.4 ± 1.3 years) had radiographic data available for analysis. Pre-detether and most recent post-detether major coronal curves were 44.8 ± 18.5° and 47.6 ± 23.9°, respectively. Etiologies of scoliosis included 11 (61.1%) congenital, 3 (16.7%) idiopathic, 2 (11%) neuromuscular, 1 (5.6%) syndromic, and 1 (5.6%) unknown. 5 (27.8%) patients had curve progression >10° at a follow-up of 3.2 ± 1.2 years. 6 (33.3%) patients experienced stabilization within 10° at a follow-up of 2.9 ± 0.8 years. 7 (38.9%) patients experienced improvement >10° at a follow-up of 3.9 ± 1.6 years. Patients with progression >10° were older at the time of detethering when compared to those without (5.6 ± 2.8 vs. 3 ± 2.7 years, p=0.084) (Table). Regression analysis demonstrated that as age at detethering increased by 1 year, the rate of curve progression >10° increased by 28.6% [95% Confidence Interval (CI): 0.899; 1.839, p=0.169].

Conclusions: In a small multicenter cohort of EOS patients with concomitant TSC, younger age at the time of detethering was found to be associated with a lower rate of scoliosis progression. Each additional year at the time of detethering was associated with a nearly 30% increase in the rate of curve progression >10°. These results indicate a potential role for early spinal cord detethering in the EOS population but requires further investigation with a larger number of patients.

Table

	Progression >10°		
	Yes	No	p-value
Age at Detethering	5.6 ± 2.8	3 ± 2.7	0.084
Major Coronal Curve at Detethering	46.6 ± 15	44.1 ± 20.2	0.804
Follow-up	3.2 ± 1.2	3.4 ± 1.4	0.749

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Abstract #8: Clinical Features of Patients with Idiopathic Scoliosis and Tethered Cord to Prompt Consideration of Earlier MRI

Sayama CM, Larson E, Munger D

Background: Scoliosis has a known association with tethered cord. Early detection and treatment of tethered cord can mitigate curve progression, neurologic/urologic decline and pain. We hypothesize that patients with both scoliosis and tethered cord represent a unique clinical entity as they do not tend to present with the typical bowel/bladder dysfunction. Our goal was to describe the clinical characteristics of patients with both scoliosis and tight filum terminale in order to identify clinical factors that might aid in earlier identification, which could ultimately save children from larger fusion surgeries in the future.

Methods: We conducted a retrospective review of patients with scoliosis who underwent filum lysis at our institution from 2015-2021. Demographic data, presenting symptoms, imaging characteristics, pre- and post-operative Cobb angle, surgical management, and outcomes were all collected and tabulated.

Results: We identified 26 patients previously deemed to have idiopathic scoliosis who underwent tethered cord release (TCR). Mean age was 11 years and mean follow-up 8.1 months. The most common presenting symptoms included back pain (53%), leg pain (31%) and leg length discrepancy/asymmetry (38%). A syrinx was identified in 38% of patients and conus terminating at or below L2 in 69%. Urine/bowel dysfunction only present in 23%. Eighty-six percent experienced improvement/resolution of their back pain after TCR. The mean Cobb angle was 34.4 preoperatively and 37.5 postoperatively. 9 patients had stable curves and 2 had improved curves. Ultimately 8 patients had fusion surgery: 4 that worsened and 4 underwent pre-planned fusion. 7 had insufficient post op imaging.

Conclusions: Patients with idiopathic scoliosis do not appear to present with typical tethering symptoms and almost 60% present with back pain, which 86% improved after TCR. More than 1/3 present with either leg pains or LLD. Back/leg pain or asymmetry should prompt earlier MRI imaging in this population. Notably, half of patients ultimately requiring fusion were referred too late. Even mild back pain should prompt consideration of MRI as timelier treatment with TCR tends to stabilize curves from worsening.

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Abstract #9: Diagnosis of Single-Suture Craniosynostosis with Cranial Ultrasound: A Single Institution Experience

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Introduction: Minimally invasive endoscopic-guided surgery followed by cranial orthosis therapy is an option for patients who are diagnosed with craniosynostosis at less than 4-6 months of age. As a result, early diagnosis is encouraged. However, the optimal protocol for the diagnostic workup of craniosynostosis remains controversial. Physical examination is often sufficient for diagnosing single-suture craniosynostosis in many cases, but sometimes imaging is necessary when the physical exam is equivocal. While head CT with 3D reconstruction is considered the gold standard, its routine use is controversial due to the long-term risks of ionizing radiation in pediatric patients. Ultrasound represents another option for visualization of the cranial sutures without the need for ionizing radiation or sedation. Here, we present our institutional experience with the use of ultrasound for the diagnosis of craniosynostosis.

Methods: All patients at our institution who underwent an ultrasound of the cranial sutures and were diagnosed with craniosynostosis between September 2019 and September 2021 were retrospectively reviewed. Demographic, clinical, and radiological data were collected.

Results: Forty-five patients with craniosynostosis were treated during the study period. Of these, 22 (49%) were male. Six patients (13%) had multi-suture involvement. Of the remaining 39 patients, 22 (56%) were diagnosed with sagittal craniosynostosis, 9 (23%) with metopic craniosynostosis, and 8 (21%) with coronal craniosynostosis. X-rays were performed in 3 cases (8%), a head CT was obtained in 6 cases (15%), and ultrasound was performed in 16 cases (41%). There was 1 case of sagittal craniosynostosis in which the suture was interpreted as patent on ultrasound, but CT confirmed craniosynostosis.

Conclusions: Ultrasound of the cranial sutures is an option for the diagnosis of craniosynostosis. Ultrasound provides effective visualization of the sutures without the use of ionizing radiation or sedation, but technical aspects of ultrasonography and its interpretation have an associated learning curve.

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Abstract #11: Relationship Between Neurosurgical Experience and Complications in Pediatric Brain Tumor surgery

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Objective: Neurosurgical complications in pediatric brain tumor surgery have been reported in prospective and retrospective case series. This study evaluates surgical morbidity and mortality based on the primary surgeons' experience over a 4 year period. Namely, surgical complications were compared between a recent graduate of pediatric neurosurgery fellowship in the initial 4 years of practice and the senior authors of the neurosurgical group.

Methods: The authors retrospectively identified all pediatric brain tumor patients that underwent surgical intervention over a 4-year period at a single institution. Operative morbidity and mortality was determined using the Drake classification. Unacceptable outcomes were graded using the NHS Greater Glasgow and Clyde impact/severity descriptors of 4-5 (Major and Extreme) and motor/sensory/cranial nerve neurological deficits lasting greater than 30 days.

Results: 220 procedures in 194 patients were identified. In 58 cases (26.4%) a complication attributable to surgery was identified. Within 30 days of surgery, one patient died because of irreversible brain stem injury due to a hemorrhagic pineal tumor despite surgical intervention (overall group mortality 0.005%). The overall complication rate was 23.6% and 28.2% for the recent graduate and senior authors respectively (p=0.5333). Major and Extreme complications according to the NHS Greater Glasgow and Clyde descriptors was 11.2% to 20.6% (p=0.0974), and deficits lasting greater than 30 days were observed in 10.1% versus 18.3% (junior vs senior surgeons respectively; p=0.1235).

Conclusions: Surgical experience was not found to be a significant factor in pediatric brain tumor surgery in overall complication rates and severity grading of morbidity. With appropriate training, recent pediatric neurosurgery fellows and training programs should rest assured of the competency of recent graduates in the field of pediatric brain tumor surgery.

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Abstract #12: A Case of a Stereotactic Magnet and the Need to Think Outside the Box

McClung Smith, Catherine, MD[1]; Giakas, Alec, BS[2]; Skarli, Stanley, MD[3]

Introduction: Air pellet guns are a relatively common cause of injury in children and can result in severe injury including intracranial injury and intracranial pellet retention. In cases where projectiles are retained the decision for or against removal often comes down to surgical accessibility and risk of infection. In cases where a projectile is in deep cerebral tissue the retrieval becomes much more difficult. While cases in the literature describe projectile retrieval with endoscopic and stereotactic assistance there are no cases in which a ferromagnetic pellet is retrieved with the assistance of a magnet, nor does the literature describe cases where an instrument is created specific to the projectile retrieval.

Methods: The authors present a review of the literature and an illustrative case in which a ferromagnetic projectile was retrieved with the assistance of a stereotactic magnetic instrument that was designed specifically for the surgery.

Results: Long term follow-up exam and imaging of the patient shows that the projectile was able to be safely retrieved with the instrument that was specifically designed for the procedure.

Conclusions: This review of the literature and illustrative case show that deep intracranial projectiles may be safely removed, though, the surgeon may need to be creative in the mode of retrieval. The illustrative case also shows that ferromagnetic projectiles may be safely removed with the assistance of stereotactic guidance and a magnetic instrument. Surgeons should be willing to think outside the box for surgical planning and consider designing instruments when the needs of the surgery demand it. In addition, cases where a ferromagnetic projectile needs to be retrieved the authors experience shows that retrieval may be easier when performed early.

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Abstract #13: Vertebral Lesions in Pediatric Chronic Recurrent Multifocal Osteomyelitis: A Case Series and Recommendations for Neurosurgical Management

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Objective: Chronic recurrent multifocal osteomyelitis (CRMO) is a rare pediatric autoinflammatory disorder associated with a high risk of pathologic vertebral body fracture that may require neurosurgical intervention. However, there are no current guidelines for the role of pediatric neurosurgeons in management of this condition. Based on our institution's experience with these patients, we demonstrate the importance of close and early involvement of neurosurgeons in caring for patients with CRMO with vertebral involvement.

Methods: Retrospective chart review was performed to identify pediatric patients under age 18 with a clinical diagnosis of CRMO treated at our institution between 2014 and 2021. Patients were included if they had radiographic evidence of at least one vertebral lesion. Clinical, radiographic, laboratory, and histopathological data were reviewed. All patients were evaluated via Jansson and Bristol CRMO diagnostic criteria. Medical and surgical management was evaluated based on clinic and operative notes.

Results: Sixty patients were identified with a clinical diagnosis of CRMO, of which six had radiographic evidence of vertebral involvement (10%). Five of six patients had multifocal involvement (83.3%). Three patients had multiple vertebrae affected (50%). All patients were treated medically, while four were assessed for surgical intervention (66.7%). One patient (16.7%) required intervention due to vertebra plana leading to a progressive kyphotic deformity and significant spinal canal stenosis.

Conclusions: In conjunction with management by the primary pediatric rheumatology team using NSAIDs, DMARDs, immunotherapies, and bisphosphonates, given the risk of pathologic fractures and potential resulting long-term neurologic deficits, we recommend close monitoring and management by pediatric spine surgeons for any CRMO patient with vertebral lesions.

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Abstract #15: Hypothermia in Cranial Vault Reconstruction Patients

Katrina Ducis, Sam Stegelmann, Laurie Ackerman

Objective: Hypothermia in adult surgical patients has been correlated with an increase in the occurrence of surgical site wound infections, increased bleeding, slower recovery from anesthetics, prolonged hospitalization, and increased healthcare costs. Pediatric surgical patients are at potentially increased risk for hypothermia because of their smaller body size, limited stores of subcutaneous fat, and less effective thermoregulatory capacity. This risk is exacerbated during pediatric craniofacial surgery by increased surface exposure to cold during induction, line placement and positioning, and prolonged surgical procedure times. The purpose of this prospective study was to identify rates and recovery of hypothermia in patients undergoing maximal warming efforts.

Methods: Demographic and clinical data were collected on 69 patients who underwent cranial vault surgery between April 2018, and February 2021. An active warming protocol, including increased ambient room temperature, forced warming blanket and warming light, was in place during this entire time period. Data points included patient age, gender, diagnosis, surgical procedure, positioning, weight, estimated blood loss for the case, transfusion volume and temperature readings throughout different phases of perioperative care. Temperatures were obtained upon arrival to the day of surgery, upon presentation to the operating room, during positioning, at incision, at the end of the procedure and recovery arrival.

Results: Of the 413 time points collected, patients were hypothermic 20% of the time which occurred in 48 separate patients. Hypothermia was most likely at positioning (65%). Of the patients who were hypothermic at positioning, unsurprisingly, 90% remained hypothermic at incision. The cohort of patients who were hypothermic at incision, 26.7% did not recover to normothermia (as defined by 36°C or greater) by the end of the operation. Prone patients had higher rates of hypothermia.

Conclusions: Patients undergoing cranial vault operations frequently experience hypothermia during the perioperative period despite high compliance with active warming protocols. Further efforts are needed to mitigate this issue to prevent further downstream issues including insufficient coagulation, blood loss, and recovery from anesthesia.

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Abstract #16: Open Retroperitoneal Inferior Vena Cava Cannulation for Distal Ventriculoatrial Shunt Catheter Placement

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Keywords: hydrocephalus, ventriculoperitoneal shunt, ventriculoatrial shunt,

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Multiple alternative sites for distal ventriculoperitoneal shunts have been described including pleural, atrial, uretral, fallopian, and gallbladder placement.7,15-18 In medically complex patients the sites for CSF diversion can be exhausted. We present a case where open retroperitoneal inferior vena cava cannulation was used for successful atrial catheter placement in a 17-month-old female. The patient had a complex abdominal, pulmonary, and vascular history precluding placement of the distal catheter in other sites or atrial placement through more peripheral venous cannulation. The patient underwent uncomplicated open retroperitoneal exposure of her IVC with cannulation and placement of atrial catheter under fluoroscopic guidance. At 5 months after surgery the patient has not required revision with appropriate placement of the distal atrial catheter.

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Abstract #17: Intrathecal Morphine Administration in Pediatric Patients Undergoing Selective Dorsal Rhizotomy: A Pilot Study

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Keywords: cerebral palsy, selective dorsal rhizotomy, pain control, intrathecal morphine

Introduction: Several studies have reported that intrathecal morphine administration improves postoperative pain outcomes, lower oral opioid use, duration of hospital stays, and recurrent visits for pain control compared to the standard regimen. The purpose of this study was to evaluate the effectiveness of intrathecal morphine (for post-operative pain control) in pediatric patients diagnosed with cerebral palsy (CP) following selective dorsal rhizotomy.

Methods: This was a retrospective, cohort analysis over the course of 4 years. The analysis consisted of a treatment group which received intrathecal morphine (5 mcg/kg) injection and a control group that did not receive the injection prior to dural closure. All patients underwent multilevel laminectomies for SDR at Akron Children's Hospital. The effectiveness of the treatment was measured by total dose of hydromorphone administered on PCA, number of days on oral narcotics, and cumulative dose of oral narcotics (oxycodone).

Results: Of the analyzed 15 pediatric patients, 7 patients received intrathecal morphine injection while the other 8 did not receive the treatment prior to dural closure. There was a difference of 1135 mcg in total PCA dose between the study group (3243mcg) and the control group (4378mcg). The total PCA dose based on weight was lower in the study group (163mcg/kg) vs. in the control group (171mcg/kg). The mean total number of days on oral narcotics was greater in the study group (2.6) vs. the control group (1.8). The total oral narcotic dose was greater in the control group (17.9mg) compared to the study group (14.1mg).

Conclusion: Based on our findings, the administration of intrathecal morphine clinically reduces the opiate need in the first 96 hours post-operatively. The use of intrathecal morphine allows CP patients to more quickly regain their functional status, for which the SDR procedure was originally performed.

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Abstract #18: I Can See Clearly Now: The Use of Augmented Reality in Pediatric Craniofacial Surgery

Joanna E. Gernsback MD, Mohammad Abdul-Mukit MS, Christian El Amm MD

Background: The use of augmented reality in surgery is the superposition of 2-D imaging data on the 3-D anatomy of a patient using semi-transparent glasses. There has been limited analysis of the impact of augmented reality in pediatric craniofacial surgery on patient outcomes and resident education.

Methods: We performed a retrospective chart review of patients evaluated in the Oklahoma Children's Hospital multidisciplinary craniofacial program who underwent augmented reality-assisted surgery. We used the HoloLens goggles (Microsoft, Redmond, Washington) from March 2021 to September 2021 on an array of craniofacial anomalies.

Results: An analysis of 14 cases demonstrated promising results. There were 9 boys and 5 girls. Age at the time of surgery was 5.8 years (range, 0.6-17.4 years). Length of surgery and EBL were 2 hours, 39 minutes and 263 cc, respectively. Most notably, there were no injuries to the dural venous sinus with the use of augmented reality in performing our osteotomies, although there was one anterior superior sagittal sinus tear while removing a bone flap, due to the dura being stuck to the bone.

Conclusions: Augmented reality may represent a useful adjunct to craniofacial surgeons. It assists in the intraoperative planning of osteotomies in and around dural venous sinuses. Future directions of our research may include the effect of augmented reality on surgical education.

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Abstract #19: Asymmetry of the Occipital Condyle-C1 Interval in Children with Atlanto-Occipital Dislocation

Sudhakar Vadivelu

Objective: Failure to diagnose atlanto-occipital dislocation (AOD) can be a significant cause of morbidity and mortality in pediatric trauma. Assessment and measurement of the occipital condyle-C1 interval (CCI) is useful in diagnosis. We have previously identified age-dependent changes in normal occipital condylar-C1 joint morphology. The extent to which right – left asymmetry of the CCI interval changes during development and how this may be used to detect AOD is unknown and is the basis for this study.

Methods*:* Normative data was obtained from a series of 124 children with CT imaging for reasons unrelated to trauma or developmental abnormality and 16 children with AOD. Right - Left CCI asymmetry was evaluated using 4 equidistant measurements across the sagittal or coronal occipital condylar-C1 joint space to determine if age specific asymmetry values exist and significance subsequently determined from the newly identified asymmetry metric.

Results: A total of 248 joint spaces were measured in normal children (age range 2 days to 22 years). The right-left CCI asymmetry did not vary substantially by age, in either the sagittal (p=0.096) or coronal (p=0.198) planes. With all age groups combined from the normative control group, the best fit cut off for both sagittal (p=0.0001) and coronal (0.0003) asymmetry interval measures is 0.30. Specificity for this asymmetry metric is 67 – 100%, 2SD and 3SD respectively, and 69% sensitivity for both sagittal and coronal planes.

Conclusions: This is the first large study to directly examine age related maturation of right – left symmetry in the occipital condyle – C1 joint space. Symmetry between the right and left joint space is preserved during development. Identifying greater than 0.3mm asymmetry of the CCI may aid in CT detection of AOD in all clinically suspicious patients, but 69% sensitivity and relatively high false positive rate make MR imaging necessary for definitive diagnosis.

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Abstract #20: Current Spread from Bugbee Wire During Monopolar Cautery Application Within a Ventricular Catheter. Technical Note

Daniel Hansen, John Honeycutt, David Donahue

Cook Children's Medical Center

Boop et al. previously demonstrated the spread of monopolar current in a proteinaceous solution using a metal stylet inserted within a ventricular catheter. In our practice we use a flexible insulated wire (Bugbee wire) with a 2mm exposed tip in place of a rigid metal stylet to free up ventricular catheters during proximal shunt revision. In theory this limits the spread of current to only a small area surrounding the wire end. This technical note extends on that prior work by demonstrating not only the ease of replicating the described testing setup, but also the difference in thermal spread when using an insulated wire. We additionally were able to capture the spread of the electrical current using high-resolution photography giving us a more precise understanding of the coagulation pattern using an insulated wire. Testing ensues at various powers showing extent of spread beyond the catheter.
This report validates the previous work of Boop et al. and shows that use of the Bugbee wire allows for more controlled application of cautery through a ventricular catheter. Appropriate power settings should be individualized based on the particular power delivery source.

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Abstract #21: Preclinical Treatment of Existing Posthemorrhagic Hydrocephalus with a Novel Drug Cocktail

Shenandoah Robinson, MD, and Lauren Jantzie, PhD

Introduction: Treatment of existing posthemorrhagic hydrocephalus of prematurity remains challenging. In a subset of patients with intraventricular hemorrhage, spontaneous recovery of CSF dynamics occurs, suggesting potential for restoration. We hypothesize PHHP persists due to a self-propagating chronic inflammation and that neuro-immunomodulatory treatment could eliminate elevated intracranial pressure (ICP). Melatonin may potentiate Roxadustat, a member of a new drug class of prolyl hydroxylase domain inhibitors, via Sirtuin-1. We tested an infantile cocktail regimen of Roxadustat plus Melatonin in an established rat PHHP model.

Methods: PHHP was induced using bilateral intracerebroventricular injection of littermate lysed red blood cells on postnatal day 1 (P1) in rats of both sexes exposed to chorioamnionitis. On P10 PHHP rats were randomly allocated to receive 10 days of ROX+MLT or vehicle intraperitoneally. Observers were masked to treatment. Intra-aural distance, a surrogate for head circumference, was measured. Cisterna magna opening pressure (OP) was measured at P21, (human toddler), or P30 (young adult). Normality was tested with Shapiro-Wilk test, and differences between groups were compared using appropriate parametric or nonparametric tests with post-hoc corrections and $p<0.05$ considered significant.

Results: Macrocephaly and elevated ICP are hallmarks of PHHP. At P21, PHHP rats had larger heads ($n=9$-12, $p=0.0017$) and elevated OP ($p<0.0001$). In another cohort, at P30 vehicle-treated PHHP rats ($n=13$) had larger heads compared to shams ($n=22$) and higher OP (both $p<0.0001$). Notably, ROX+MLT-treated PHHP rats ($n=11$) had smaller heads compared to vehicle-treated PHHP rats ($p=0.02$) and lower ICP ($p=0.04$). Head size and ICP for ROX+MLT-treated PHHP rats did not differ from shams. Additional imaging, histological and biochemical analyses are underway.

Conclusions: CSF dynamics reflect choroid plexus secretion, ependymal motile cilia propulsion and glymphatic system reabsorption of CSF- components all vulnerable to chronic inflammation. These early results suggest an oral drug cocktail may effectively treat existing PHHP, potentially reducing lifetime shunt-dependence.

The authors declare they have no conflicts of interest.

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Abstract #22: The Construction of a Composite CSF Diversion Surgery Index (CDSI) for Decision Making in CSF diversion Surgery for Pediatric Patients Following Prenatal MMC Repair – Initial Experience

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Introduction: The MOMS trial initial results and primary outcome related to shunt placement demonstrated a statistically significant difference between the prenatal and postnatal repair groups (40% versus 82%, P<0.001). The decision to insert a VP shunt was made by an independent committee of neurosurgeons who utilized pre-determined clinical and radiographic data to ascertain the need for shunting in each patient. The criteria were not universally agreed upon or applied, as evidenced by the data which showed that while 64 of 91 children (70%) in the prenatal cohort met criteria, only 40 of the 91 children (44%) received a shunt. The objective of our study was to create a comprehensive index that could be used as a prediction tool to improve decision making in diagnosing progressive hydrocephalus in patients following prenatal MMC repair.

Materials and Methods: Thirty-three patients participated in the study. Fourteen patients with prenatal repair and DTI with shunt surgery and 19 patients with prenatal repair and DTI who did not require a shunt were included. Logistic regression analysis was performed using SPSS Statistics software to assess the MOMS index, DTI measures, FOHR, and sex in predicting patients who would require shunting.

Results: Based on our data analysis, the CDSI demonstrated a sensitivity of 78.6% and a specificity of 86.5%, with an overall accuracy of 84.8%. This rate is better than the performance of all of the individual indices based on sensitivity and overall accuracy, and also better than most of the individual indices based on the specificity alone.

Conclusion: Our CDSI consists of the MOMS criteria, MOMS "Tulipan" revised criteria, and additional conventional clinical measures, as well as objective non-invasive neuroimaging biomarkers based on DTI. These initial overall findings may allow for group-based statistical analysis to be transitioned to individualized decision-making for diagnosis of progressive hydrocephalus in patients following fetal MMC repair.

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Abstract #23: Ventriculopleural Shunts in a Pediatric Population: A Review of 170 Consecutive Patients

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Objective: The authors sought to determine the outcome of using the pleural space as the terminus for ventricular CSF-diverting shunts in a pediatric population.

Methods: All ventriculopleural (VPl) shunt insertions or revisions done between 1978 and 2018 in patients were identified. Data recorded for analysis were: age, sex, weight, etiology of hydrocephalus, previous shunt history, reason for VPl shunt insertion or conversion from ventriculoperitoneal (VP) or ventriculoatrial (VA) shunt, valve type, nature of malfunction, presence of shunt infection or pleural effusion, and conversion to a different distal site.

Results: A total of 170 patients (mean age 14 ± 4 years) with a VPl shunt were followed up for a mean of 57 ± 53 months. The reasons for conversion to a VPl shunt for 167 patients were previous shunt infection in 57 (34%), multiple abdominal procedures in 44 (26%), inadequate absorption of CSF in 34 (20%), abdominal pseudocyst in 25 (15%), and obesity in 7 (4%). No VPl revisions were required in 97 (57%) patients. Of the 73 (43%) patients who did require revision, the most common reason was proximal obstruction in 32 (44%). The next most frequent complication was pleural effusion in 22 (30%) and included 3 patients with shunt infection. All 22 patients with a clinically significant pleural effusion required changing the distal end of the shunt from the pleural space. Pleural effusion was more likely to occur in VPl shunts without an antisiphon valve. Of the 29 children < 10 years old, 7 (24%) developed a pleural effusion requiring a revision of the distal catheter to outside the pleural space compared with 15 (11%) who were older (p=0.049). There were 14 shunt infections with a rate of 4.2% per procedure and 8.2% per patient.

Conclusions: VPl shunts in children under 10 years of age have a significantly higher rate of symptomatic pleural effusion, requiring revision of the shunt's terminus to a different location. VPl shunt complication rates are similar to those of VP shunts. The technical difficulty of inserting a VPl shunt is comparable to that of a VP shunt. In a patient older than 10 years, all else being equal, the authors recommend the distal end of a shunt be placed into the pleural space rather than the right atrium if the peritoneal cavity is not suitable.

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Abstract #24: A Comprehensive Protocol to Reduce Gram-Negative Rod Shunt Infection: Results of a Quality Improvement Project

Abstract Topic: Quality Improvement

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Introduction: Facing a rise in gram-negative rod (GNR) shunt infections, specifically Serratia marcescens, we added steps to our shunt infection prevention protocol to address this epidemic.

Methods: The protocol was developed by adding steps to our existing protocol to address GNR infections. New components of the protocol included pre-operative bath and hair wash; no foley catheter placement; single dose pre-operative intravenous gentamycin; no gel to comb and part hair; changing outer gloves after draping; no touch technique; skin preparation with alcohol, chlorohexidine scrub before chlorohexidine prep; vancomycin-gentamycin irrigation; vancomycin powder in all incisions at closure; and dressing incisions before drape removal. Shunt infections were defined by positive CSF gram stain or culture, positive shunt wound culture, shunt erosion through skin, or abdominal pseudocyst within 6 months of shunt surgery. The protocol was implemented January 2020.

Results: 196 shunt operations were performed in 2020 after implementation of the shunt infection protocol, while 268 shunt operations were performed in 2019. Two shunt infections occurred in 2020 and one was positive for GNRs, while 21 shunt infections occurred in 2019 and 5 were positive for GNRs. The overall shunt infection rate for 2020 after implementation of the protocol was 1.02%, down from 7.84% in 2019 ($p = 0.0009$, ARR 6.8%, RRR 87%). From 2010 to 2019, our average shunt infection rate was 5.19% (range 2.07% - 10.26%). No significant deviations in the protocol were noted for the 2 infections in 2020. Adherence to the protocol in 2020 was >95% in all categories, except "no touch

technique" (54%), skin prep using all 3 modalities (84%), and shampoo/bath prior to surgery (34%). No complications occurred secondary to the new protocol.

Conclusions: Implementation of a shunt infection protocol to combat an increase in GNR infections resulted in a significant decrease in our overall shunt infection rate.

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Abstract #25: Reconsidering Hydrocephalus in the Setting of Achondroplasia

Dale Swift, MD

Obective: Ventriculo-peritoneal shunts (VPS) for hydrocephalus in patients with achondroplasia are known to have a higher failure rate than in other hydrocephalus populations. However, the etiology of hydrocephalus in this group is considered "communicating" and therefore not amenable to endoscopic third ventriculostomy (ETV). We studied the long-term results of ETV in this population. In addition, the development of hydrocephalus in these patients was investigated with particular attention to the potential contribution of cerebellar tonsil herniation (CTH) and foramen magnum decompression (FMD) when performed.

Methods*:* Medical records and neuroimages of patients with achondroplasia who had undergone ETV or VPS were reviewed. Ventricular volumes and frontal and occipital horn ratios (FOHR) in ETV patients were measured pre- and post-operatively. Rates of reoperation were also compared. Posterior fossa anatomy was assessed to determine the presence of CTH and its temporal relation to FMD.

Results*:* Nineteen patients with achondroplasia were treated for hydrocephalus; 10 underwent ETV only, 7 underwent VPS only, and 2 patients had VPS followed by ETV. There was a significant difference in repeat cerebrospinal fluid surgery between ETV and VPS. In ETV patients, ventricular volume and FOHR ratios were generally normal if measured at birth and increased significantly until the time of ETV. After ETV all patients demonstrated significant and sustained decreases in these ventricular measurements up to 15 years. All ETV patients demonstrated low lying or herniated cerebellar tonsils and in some patients, tonsillar herniation appeared to be precipitated by FMD.

Conclusions: ETV is efficacious, safe and durable in the treatment of hydrocephalus in patients with achondroplasia. Although many studies indicate that hydrocephalus in these patients is "communicating", a subset may develop an "obstructive" component that is progressive and responsive to ETV. The obstruction in this setting may be due to CTH and paradoxically worsened by FMD.

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Abstract #26: A Novel Peptide Vaccine Directed to CMV pp65 for Treatment of Recurrent Malignant Glioma and Medulloblastoma in Children and Young Adults: Preliminary Results of a Phase I Trial

Eric M. Thompson, Daniel Landi, Gary Archer, Eric Lipp, Ashley Walter, Bridget Archambault, Bea Balajonda, Charlene Flahiff, Denise Jaggers, James Herndon, Evan Buckley, Kristin Schroeder, Dina Randazzo, Annick Desjardins, Margaret Johnson, Katherine Peters, Mustafa Khasraw, Michael Malinzak, Duane Mitchell, David Ashley, and John Sampson

Introduction: The cytomegalovirus (CMV) antigen, pp65, is ubiquitously expressed in malignant glioma and medulloblastoma but not in healthy brain. The objective of this Phase I trial (NCT03299309) was to assess the safety and feasibility of a novel pp65 peptide vaccine (PEP-CMV) in children and young adults with recurrent medulloblastoma and malignant glioma.

Methods: Vaccines contain a synthetic long peptide (SLP) of 26 amino acids encoding multiple potential class I, class II, and antibody epitopes of CMV pp65 across several haplotypes. This SLP is administered as an emulsion in Montanide ISA 51. Patients receive a single course of temozolomide to induce lymphopenia, tetanus/diphtheria toxoid site preconditioning, then vaccines administered intradermally every two weeks for 3 doses, then monthly.

Results: To date, 22 patients have been enrolled. Diagnoses include medulloblastoma (n=2), glioblastoma (n=12), anaplastic oligodendroglioma (n=2), anaplastic astrocytoma (n=3), and malignant glioma NOS (n=3). Mean number of prior treatment regimens is 4.9 (range 1-12). Mean age is 22yo (range 6-35) and 45% of patients are male. The median KPS is 80. The median number of vaccines given at time of analysis is 3.3 (range 1-12). There have been no ≥ 3 Grade toxicities related to the vaccine. One patient developed nausea, vomiting, palpitations, and tachycardia after vaccination and had elevated inflammatory cytokines consistent with cytokine release syndrome. Median PFS is 2.5 months (95% CI: 1.7,4.5) and median OS is 6.5 months (95% CI 3.3, 7.9). Immune response to pp65 as determined by ELISpot was found in 75% of patients. On MRI, 6 of the 11 evaluable patients have had at least stable disease with three of those having a partial response.

Conclusions: Preliminary results demonstrate that PEP-CMV is well-tolerated and elicits an immune response in heavily pretreated, multiply recurrent patients. A multi-institutional Phase II trial is scheduled to open spring 2022.

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Abstract #27: A small molecule RELA nuclear localization inhibitor increases survival in ZFTA-RELA fusion ependymoma

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Summary: ZFTA-RELA fusions drive the most common class of supratentorial ependymoma. Given the lack of other recurrent mutations in these aggressive and frequently lethal tumors, drugging this fusion represents a major therapeutic opportunity. However, this prospect is complicated by the fact that both proteins are transcription factors, which limits targeting mechanisms. Members of our group recently described a novel approach to wild type RELA inhibition through the inhibition of nuclear translocation of RELA caused by the blocking of conformational changes leading to the exposure of the nuclear localization sequence by a small molecule called CRL1101. Here, using a novel autochthonous model of ZFTA-RELA ependymoma that recapitulates the transcriptomic and pathological features of this disease, we demonstrate that CRL1101 crosses the blood-brain barrier and significantly improves survival. The poor prognosis of patients with recurrent ZFTA-RELA ependymoma and the demonstrated pre-clinical efficacy of CRL1101 in vivo strongly warrants further development of this therapeutic.

KEYWORDS p65, NLS, anaplastic, epigenetics, gliogenesis, NF-kB, AAVS1 locus, Ependymal cell, Nfkb1, scRNA-seq, scATAC-seq, p50, p105, C11orf95.

DISCLOSURES
Cedars-Sinai and the senior authors have filed for patent protection for CRL1101 and potential clinical uses. CRL1101 has been licensed for non-ependymoma indications.

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Abstract #28: Three-dimensional Organoid Culture Unveils Resistance to Clinical Therapies in Adult and Pediatric Glioblastoma

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Background: Glioblastoma (GBM) is the most common primary brain tumor with a dismal prognosis. The inherent cellular diversity and interactions within tumor microenvironments represent a significant challenge to effective treatment. Traditional culture methods may mask the complexity of such interactions while three-dimensional (3D) organoid culture systems derived from patient cancer stem cells (CSCs) can preserve cellular complexity and microenvironments. Our objective was to determine whether organoid cultures show increased patterns of resistance to potential clinical therapies compared to traditional sphere cultures.

Methods: Adult and pediatric surgical specimens were collected and established as 3D organoids. We created organoid microarrays and visualized bulk and spatially defined differences in cell proliferation using immunohistochemistry (IHC) staining, as well as cell cycle analysis by flow cytometry with 3D regional labeling. We tested the response of CSCs grown in each culture method to temozolomide, ibrutinib, lomustine, ruxolitinib, and radiotherapy using proliferative and viability assays.

Results: Compared to sphere cultures from the same patient, organoids showed diverse proliferative cell populations and broad resistance to all therapies tested, albeit with both intraspecimen and interspecimen variability in the extent of resistance. Organoid specimens demonstrated a blunt response to current GBM standard of care therapy (combination temozolomide and radiotherapy) and maintained both cellular proliferation in their outer rim and overall structure and viability compared to the matched sphere specimens.

Conclusions: Our results suggest that growth of tumor specimens as organoid cultures may better reflect the cellular diversity and clinical reality of GBM therapeutic response. Patient-derived GBM organoids offer a valuable complement to traditional culture methods and may have powerful predictive capability of personalized drug sensitivities and therapeutic resistance.

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Abstract #29: Toward Understanding the Relationship Between Ventriculoperitoneal Shunt Placement and Hearing Loss

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University of Minnesota Departments of Neurosurgery[1], Otolaryngology[2], Speech-Launguage-Hearing[3], and Biostatistics[4;] and University of Iowa Departments of Neurosurgery[5] and Otolaryngology[6].

Background: Hearing loss (HL) has been reported after ventriculoperitoneal shunt (VPS) placement in 38% to 83% of children. Because existing studies are retrospective in selected populations, the incidence is unknown, and the mechanism unclear. Because CSF communicates with perilymph through the cochlear aqueduct (CA), it is hypothesized that CSF drainage results in perilymph drainage, endolymphatic expansion, cochlear dysfunction, and HL. Supporting this hypothesis are altered electrocochleography (ECOG) and distortion product otoacoustic emission (DPOAE) recordings post VPS. This abstract aims to review prior, and describe current proposed studies to: (1) determine the incidence, magnitude, and recoverability of post-VPS HL in children and (2) test the hypothesis that post-VPS HL is due to an altered perilymph/endolymph gradient.

Methods: We hypothesize that an association between VPS and HL exists and that 60% of children undergoing VPS placement will develop HL (defined as ≥ 20 dB worsening at any frequency or ≥ 10 dB worsening at two contiguous frequencies) after VPS. Ninety-four children will have baseline and post-VPS audiologic testing with repeat studies at 2 and 12 weeks to assess recovery and possible delayed HL. For mechanistic hypothesis testing, the relationships between CSF drainage, endolymph expansion, and HL will be evaluated in sedated children undergoing VPS. Pre- and post-operative ECOG, DPOAEs, ABRs and CSF pressure measurements will be obtained. We hypothesize: (1) CSF drainage is associated with increased ECOG summating potential to action potential ratios; increased DPOAE magnitude and decreased DPOAE phase angles; (2) post-VPS HL is associated with larger ECOG changes; decreased or absent DPOAEs; decreased amplitude and increased latency of ABR wave I; and (3) larger CSF pressure changes will be associated with larger ECOG, DPOAE and ABR changes.

Conclusions: Completing this proposed study will yield essential information about an under-recognized complication in treating hydrocephalus in children.

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Abstract #30: Treatment of Cervical Stenosis in Infants and Toddlers with Skeletal Dysplasia

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While foramen magnum (FM) stenosis is ubiquitous in infants with achondroplasia, some of the less common skeletal dysplasias are associated with cervical stenosis. When this creates symptomatic cord compression, cervical decompression may be indicated.

We reviewed patients seen in our skeletal dysplasia program over the past 15 years and identified 16 patients with skeletal dysplasias other than achondroplasia under 36 months who underwent surgical treatment for cervical stenosis without instability or kyphosis. Cervical stenosis without instability is very common in children with thanatophoric dysplasia and rhizomelic chondrodysplasia punctata (RCDP), and occasionally seen in Ellis van Creveld (EVC). Stenosis at C1 with or without instability is common in metatropic dysplasia. The three thanatophoric infants we treated had remarkable stenosis at the FM and C1 with one tight to C3. They all did well with surgery and one is a long-term survivor. The seven children with RCDP typically had diffuse cervical stenosis and early in the series underwent multilevel laminectomies with onlay cervical graft and a halo. More recently they have been treated with laminoplasties without fusion. Four metatropic dysplasia patients had C1 stenosis without C1-2 instability and underwent a single level laminectomy although one developed instability postoperatively. Eight additional children with metatropic dysplasia had C1 stenosis associated with C1-2 instability and underwent decompression and fusion in the same procedure. Two infants with EVC had cervical stenosis, one just at C1 and the other diffusely throughout the cervical spine. This child was managed with laminoplasties without fusion.

Cervical decompressions for critical stenosis seen in certain skeletal dysplasias can be performed safely with little morbidity and good clinical outcomes. FM and C1 decompression have little risk of post-laminectomy kyphosis. Multilevel cervical decompressions should either seek to maintain the posterior tension band or undergo prophylactic fusion at the time of the decompression.

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Abstract #31: Surgical Revascularization Decreases Stroke and TIA Risk in Children with Sickle Cell Disease and Moyamoya Syndrome: Results of the Stroke in Sickle Cell Revascularization Study (SiSCRS)

Philipp Aldana, MD, Joseph Piatt, MD, Ricardo Hanel, MD PhD, Sabrina Han, MS, Corinna Schultz, MD and Manisha Bansal, MD for the SiSCRS Investigators

Background: Recent studies have suggested that surgical revascularization may be a safe and effective therapy to reduce risk of cerebrovascular complications in patients with sickle cell disease and moyamoya syndrome (SCD-MMS). These studies have been limited by small sample size and lack of a control group for comparison.

Objective: To investigate whether revascularization surgery reduces the risk of cerebrovascular events (CVEs) in comparison to conservative management alone in a retrospective cohort of children with SCD-MMS.

Methods: A retrospective review of data from 14 major U.S. pediatric neurosurgery centers of SCD-MMS patients (≤18 y.o.) was performed. Detailed information on sickle cell disease, stroke and surgical histories were extracted. The incidence of CVEs (stroke and TIAs) between patients treated with surgical revascularization was compared to those with conservative management alone. Multivariate regression models were generated and logistic regression analyses were performed.

Results: A total of 141 patients with SCD-MMS were studied. 78 (55.3%) were treated with conservative management and revascularization surgery (Surgery group) and 63 (44.7%) were treated with conservative management alone (Conservative group). Patients in the Surgery group had reduced odds of developing a CVE over the duration of their risk period (odds ratio = 0.27, 95% CI: 0.08-0.94, P = .040). Furthermore, when comparing patients in the Surgery group during their pre-surgical periods and post-surgical periods, patients had markedly reduced odds of developing a CVE after surgery (odds ratio = 0.22, 95% CI = 0.08-0.58, P = .002). Postoperatively, 5 patients had CVEs and 5 had non CVE complications.

Conclusion: Our retrospective study provides strong evidence that revascularization surgery can be performed safely and reduce risk of CVEs in patients with SCD-MM. This is the largest study of its kind to date. A prospective study will be needed to validate our findings and study designs will be discussed at the meeting.

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Abstract #32: The Use of Responsive Neuro Stimulation (RNS) In Adult and Pediatric Patients with Diffuse And Non-Localizable Seizure Foci/Networks.

P. David Adelson MD, Jorge Arango MD, Nicole Falls, BS, Angus Wilfong MD, Korwyn Williams MD, Randa Jarrar MD, Matthew Troester DO, John Kerrigan MD, Jordana Fox Kensicki DO, Brian Appavu MD, Varina Boerwinkle MD

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Objectives: Medically refractory epilepsy arising from diffuse or non-localizing seizure foci remain a therapeutic challenge, as resective surgery is often not an option and indirect neurostimulation may have not been efficacious. In these instances, responsive neurostimulation (RNS) has been utilized in adults with good outcomes in the majority of patients. The utility of RNS in children has not been systematically explored. In this study, the authors present a single institution's experience with RNS in pediatric patients.

Methods: A single-center retrospective review of patients who underwent RNS implantation at Phoenix Children's Hospital during a 4-year period.

Results: Twenty-two patients underwent RNS implantation utilizing different anatomical targets, Male 55%. The mean patient age at treatment was 16.6 years, (range 6.4-23.7 years), and the average follow-up of up to 1.8 years (range 3 months to 3.4 years). All patients had preoperative noninvasive evaluation that included resting state functional MRI (rsfMRI) as well as invasive monitoring with stereoelectroencephalography (SEEG) to help determine the RNS targets. All patients had variable positive responses with reduction of seizure frequency and or intensity. Overall, seizure frequency reduction of > 50% was seen in the majority (77%) of patients without serious adverse events.

Conclusions: The use of RNS in medically intractable patients provided improvements in seizure control after implantation with decreases in seizure frequency > 50% from baseline as compared to preoperatively in the majority of patients. Preliminarily, early findings indicate that rsfMRI and SEEG, as part of the pre-implantation evaluation, were helpful for RNS targeting and that RNS can be used safely even in young children.

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Abstract #33: Responsive Neurostimulation in Pediatric Drug-Resistant Epilepsy: Initial Center Experience

Robert J. Bollo, MD; Carey A. Wilson, MD; Chris Espinoza, MD; Kimberly Orton, RN; Matthew T. Sweney, MD

Introduction: Drug-resistant epilepsy (DRE) impacts one-third of children with seizures, and surgical treatment is strongly recommended in this population. While resection of the epileptogenic zone confers the highest chance of durable seizure freedom, this may be challenging or not possible when seizure onset overlaps eloquent cortex, is multifocal, or in primary generalized epilepsy. Further, only approximately 1% of children with DRE undergo surgical treatment, and surgery for DRE remains among the most under-utilized evidence-based treatments in medicine. Responsive neurostimulation (RNS) is a potential solution to these important challenges, but there is a critical need for data in pediatric patients. We present the initial experience with RNS in children with DRE at a single NAEC level 4 epilepsy center.

Methods: We reviewed the charts to collect demographic, clinical, and radiographic data for patients with DRE treated with RNS at our center, beginning with the first patient treated with RNS in 2017. Particular scrutiny was applied to seizure type, duration of epilepsy, non-invasive evaluation, invasive EEG, previous therapeutic surgical treatment, resection at the time of RNS placement, RNS strategy including unilateral or bilateral electrodes, cortical or thalamic stimulation, surgical complications, duration of follow-up, and seizure outcome. The clinical decision to pursue RNS therapy was made by consensus of our multidisciplinary team.

Results: Twenty-five RNS devices were implanted in 22 patients. The mean age at surgery was 13.9 years (range, 5-21 years). Eleven patients (50%) had bilateral electrode implants, and 9/11 (82%) of those with unilateral electrodes were implanted on the left (dominant) hemisphere. Three patients with Lennox-Gastaut Syndrome (LGS) or LGS-like epileptic encephalopathy underwent thalamocortical electrode placement with bilateral thalamic stimulation targeting the centromedian nucleus. The mean follow-up is 17.3 months (range, 1-47 months). Two patients (9%) experienced complications: one patient developed an infection requiring explantation, and one patient a post-operative acute epidural hematoma requiring emergency evacuation. Twelve patients (55%) have at least 1-year follow-up, and all have experienced a significant reduction in clinical seizures.

Conclusions: Initial experience with RNS in pediatric patients with DRE demonstrates safety and efficacy comparable to that seen in adult populations with DRE. Ongoing prospective multicenter data collection, including in children undergoing thalamic stimulation, is critical to identify the best pediatric candidates for treatment with RNS, optimize surgical targeting, and establish the efficacy and complication rate in children.

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Abstract #34: Responsive Neural Stimulation of Bilateral Centromedian (CM) Nucleus of the Thalamus in Drug Resistant Pediatric Epilepsy: Preliminary Experience at UCLA Mattel Children's Hospital

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Introduction: Responsive neural stimulation (RNS) is an FDA-approved and only closed-loop treatment option for uni- or bi-focal drug resistant epilepsy in adults. Recently in a multicenter pediatric study, its safety and efficacy were found to be similar to the landmark adult studies. There is increasing interest in the using this technology for closed-loop thalamic stimulation. The goal of this study was to report our single-center experience of bilateral centromedian (CM)-thalamic stimulation in a consecutive cohort of children with drug-resistant epilepsy.

Methods: Consecutive pediatric patients with drug resistant epilepsy who underwent responsive neural stimulation of the CM nucleus of the thalamus at UCLA Mattel Children's Hospital between 2020 and 2021 were included in this study. Demographic data, clinical variables, stimulation settings, complications and latest seizure outcome data was captured for all patients.

Results: Seven patients aged 3-22 y (Median 13y) were included in this study. Three (42%) of these patients had an existing Vagus Nerve Stimulator implant at the time of bilateral CM RNS surgery. Six (86%) of the patients had severe epilepsy consisting of multiple daily seizures with 2 (28%) patients having Lennox-Gastout Syndrome. Two (28%) patients developed a delayed superficial site infection requiring device explant. Ictal recordings were recorded in all patients enabling closed-loop stimulation. All patients with greater than 3-month follow-up from the date of implantation have experienced greater than 50% seizure reduction.

Conclusion: Our preliminary experience shows that bilateral CM nucleus RNS may be a feasible option in multifocal or generalized severe epilepsy in pediatric patients. Multicenter collaborative clinical research with longer follow-up durations is required to assess safety and efficacy.

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Abstract #35: Minimally-Invasive Interlaminar Approach for Infra-conus Selective Dorsal Rhizotomy

Neena I. Marupudi, MD, MS[1, 3]; Michael H. Lawless, DO[1]; Erika Erlandson, MD[2,3]; Sandeep Sood, MD[1, 3]; Steven Ham, DO[1, 3]

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Introduction: Selective dorsal rhizotomy (SDR) is an effective, well-validated surgical approach for treating spasticity in children with cerebral palsy. Conventional techniques include multi-level laminectomy, single-level laminectomy at the conus, and infra-conus single-level laminectomy. We present a novel modification of the infra-conus technique: an interlaminar approach for minimally-invasive SDR with limited or no bone removal. This technique minimizes pain and deformity and allows for earlier post-surgical mobility and participation in therapy.

Methods: Patients undergoing SDR with an interlaminar approach at Children's Hospital of Michigan (2018-2021) were included in the study with spastic diplegia, hemiplegia, or quadriplegia. Outcome measures included assessment of spasticity, pain, spinal deformity/kyphosis, bowel/bladder dysfunction, infection, CSF leak, and if functionally applicable, Gross Motor Function Measure (GMFM), strength, and endurance.

Surgical technique entails a 3-cm midline incision spanning the interlaminar junction below the conus. Ligamentum flavum is removed in the interlaminar space, and if necessary, a limited mini-laminotomy is performed. The thecal sac is opened, and using intraoperative electromyography, motor and sensory nerve roots are distinguished. Abnormal sensory nerve rootlets are partially cut. Target muscle groups are determined based on preoperative assessments.

Results: Postoperatively, none of the patients developed a CSF leak, intractable pain, infection, spinal deformity/kyphosis, or bowel/bladder dysfunction (n=38, follow-up: 3-months to 3.5-years). Spasticity decreased by a Modified Ashworth Score of ≥ 2 in all affected muscle groups. In patients with spastic diplegia meeting criteria for optimal functional benefits, the mean postoperative rate of improvement in GMFM score was 2.03 per month ($p<0.0001$), exceeding preoperative baseline between 3 and 6-months after surgery. The mean postoperative rate of improvement in a 6-minute walk test was 62 steps per month ($p=0.0006$), exceeding preoperative baseline testing at 12 months after surgery.

Conclusion: This interlaminar infraconus surgical technique for SDR is feasible, safe, and produces expected clinical improvement in this patient population. The technique minimizes risk of spinal deformity/kyphosis and intractable pain, while optimizing early participation in inpatient rehabilitation programs.

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Abstract #36: Human Brain Growth: Avoiding the Mismeasure of Man

Steven J. Schiff, MD, PhD

Introduction: The study of brain size and growth has a long and contentious history. The normal brain growth and CSF accumulation relationship is critical to characterize because it is impacted in numerous conditions of early childhood where brain growth and fluid accumulation are affected such as infection, hemorrhage, hydrocephalus, and a broad range of congenital disorders.

Objective: This study describes normal brain volume growth with respect to age, sex, and cerebrospinal accumulation.

Methods: We analyzed 1067 magnetic resonance imaging (MRI) scans from 505 healthy pediatric subjects from birth to age 18. The volume trajectories were compared between using Smoothing Spline ANOVA. Population growth curves were developed using Generalized Additive Models for Location, Scale, and Shape.

Results: Brain volume peaked at 10-12 years of age. Males exhibited larger age-adjusted total brain volumes than females, and body size normalization procedures did not eliminate this difference. We unexpectedly discovered that the ratio of brain to CSF volume exhibits a novel universal age-dependent relationship independent of sex or body size (Peterson et al, J Neurosurg Ped, published online 7/9/2021). Strong volumetric laterality was shown in normal temporal lobe and hippocampal growth. Additional applications of these normal growth curves are shown to the 2-year follow-up of 100 patients with postinfectious hydrocephalus (Schiff et al, J Neurosurg Ped, 2021, published online 7/9/2021), as well as the predictive risk analysis of the propensity to develop subdural fluid accumulations (Lane et al, J Neurosurg Ped, 2021, published online 10/1/2021).

Conclusions: These findings enable the application of normative growth curves in managing a broad range of childhood disease where cognitive development, brain growth, and fluid accumulation are interrelated. Normative asymmetry opens new avenues in the evaluation of epilepsy. With the increasing availability of volumetric MRI software, and new inexpensive commercial low-field MRI, the prospect of incorporating normative brain growth into the management of many neurosurgical conditions of childhood is now widely feasible.

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Abstract #38: Invasive and Non-invasive Brain Tissue Oxygen Monitoring in Pediatric Severe Traumatic Brain Injury

Shih-Shan Lang MD, Nankee Kumar, Chao Zhao MS, Alexander M. Tucker MD, Phillip B. Storm MD, Greg G. Heuer MD, Matthew Kirschen MD PhD, Alexis Topjian MD MSCE, Chong Tae Kim MD, Ian Yuan MD, Todd J. Kilbaugh MD, Jimmy W. Huh MD

Introduction: Severe traumatic brain injury (TBI) is a leading cause of disability and mortality in pediatrics. Monitoring brain tissue oxygenation may help limit secondary brain injury and improve outcomes. We aimed to analyze 1) the association between intracranial pressure (ICP) and invasively acquired brain tissue oxygenation (PbtO2) on functional outcomes, and 2) the association between non-invasively acquired cerebral oxygenation using near-infrared spectroscopy (NIRS) and PbtO2.

Methods: Patients ≤ 18 with severe TBI who received ICP +/- PbtO2 +/- NIRS monitoring were retrospectively reviewed between 1998-2021. We evaluated differences between patients with ICP monitoring only versus ICP + PbtO2 monitoring on hospital and pediatric intensive care unit (PICU) length of stay (LOS), length of intubation, Pediatric Intensity Level of Therapy (PILOT) score, and functional outcome using Glasgow Outcome Score – Extended (GOS-E) scale at 6 months post-injury.

Results: Forty-nine patients (30 with ICP only and 19 with ICP + PbtO2) were analyzed. Lower overall PbtO2 and an increased number of hypoxic events (<20 mmHg) were associated with longer hospital (ρ = 0.55) and PICU LOS (ρ = 0.63) and more favorable GOS-E score (ρ = - 0.55). Patients with ICP + PbtO2 monitoring experienced longer PICU LOS (p = 0.018) compared to patients with ICP only monitoring with no GOS-E score difference between groups (p=0.733). There was a weak negative correlation between ICP and PbtO2 (r = -0.03, β = -0.22). Lower average ICP values predicted favorable GOS-E scores (r = -0.61). In seven patients with NIRS monitoring, NIRS values increased as PbtO2 values increased.

Conclusion: Lower overall PbtO2 and increased number of hypoxic episodes resulted in longer hospital admissions with less favorable functional outcomes. Lower ICP correlated with more favorable functional outcomes. There was a poor correlation between ICP and PbtO2 supporting the importance of multimodal monitoring.

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Abstract #39: Fetoscopic Closure of Myelomeningocele: Surgical Technique and Case Series

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[1] Midwest Fetal Care Center, [2] Children's Minnesota, [3] Pediatric Surgical Associates, [4] Mayo Clinic

Introduction: Open maternal-fetal myelomeningocele/myeloschisis (MMC) closures are superior to standard postnatal repairs, demonstrating improvement in hindbrain herniation, motor function, and shunt rates; however, they necessitate Cesarean delivery and increase risk for preterm birth. Since 2016, we have successfully performed 42 open closures using a myofascial flap technique. We recently adopted a fetoscopic technique aimed at preserving fetal and improving obstetrical outcomes, including vaginal delivery, gestational age at delivery, and risk of uterine and placental implantation complications in subsequent pregnancies.

Methods: Eligible patients underwent fetoscopic MMC repair involving a multi-layer myofascial flap and skin closure, distinguishing this technique from those previously reported. We will describe our fetoscopic protocol, including anesthetic, surgical, and intraoperative fetal cardiovascular monitoring techniques, along with outcomes data for the program's first year (October 2020-October 2021).

Results: Fourteen fetoscopic closures were successfully performed at a mean (SD) of 25.0(0.7) weeks gestation. Eight of 12 infants were delivered vaginally, with an overall mean gestational age of 35.1(2.4) weeks. Membrane complications occurred in 8/12 patients. Favorable outcomes at last follow-up (mean=66(58) days) included significant hindbrain herniation improvement in 10/11. One infant required shunting in the neonatal period. Gross motor functional improvement of ≥1 level was seen in 6/9 (mean=2.4(2.6)). One intrauterine fetal demise of unknown cause occurred 4 weeks post-surgery, and 8/12 required neonatal skin wound revisions with intact myofascial flap, including 2 with uncomplicated CSF leaks. Comparisons with MOMS Trial outcomes data will be reviewed.

Conclusions: We have demonstrated the feasibility of the fetoscopic myofascial flap and skin closure technique. Continued refinements of our surgical technique are needed to reduce rates of membrane complications and fetal skin edge dehiscence. Early findings regarding hindbrain herniation reversal, functional motor levels, and shunt rates are encouraging. Longer-term outcomes, including those concerning mobility, bladder and bowel function, and neurodevelopment, need to be evaluated.

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Abstract #40: Intra-uterine Closure of Myelomeningocele Defects with Primary versus Bipedicle \ Fasciocutaneous Flaps: A Single Center Post-MOMS Retrospective Cohort Study

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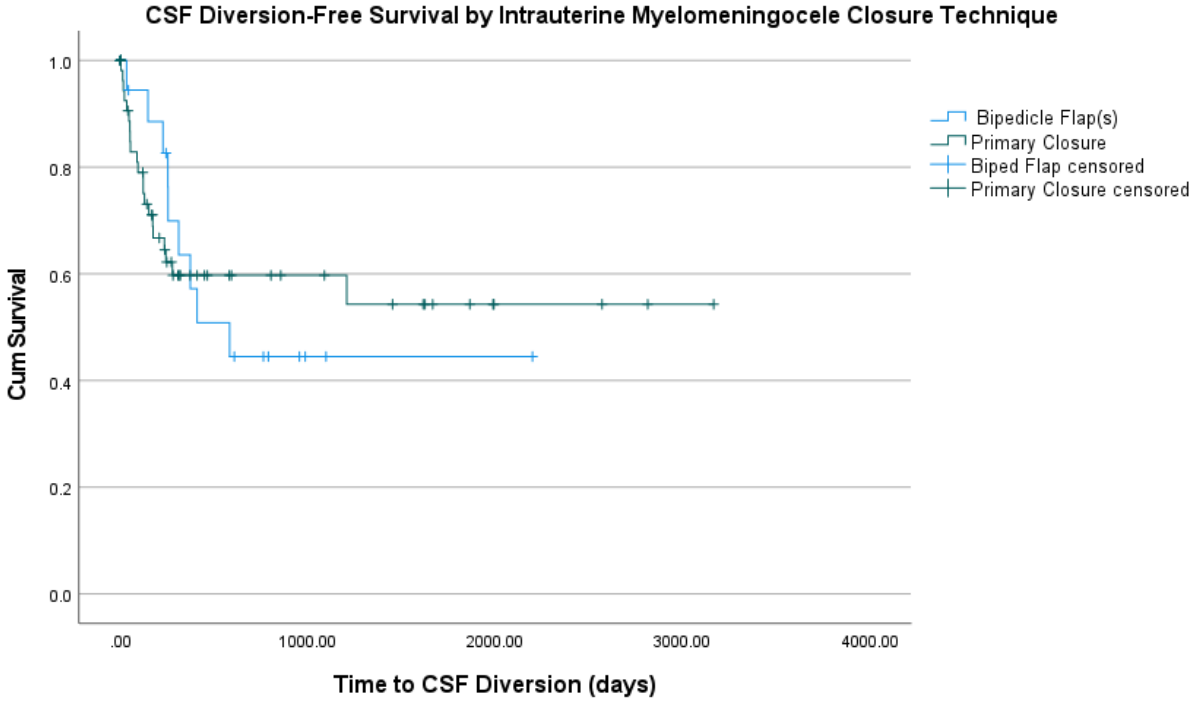
Introduction: The Management of Myelomeningocele Study concluded that prenatal myelomeningocele (MMC) repair improves postnatal outcomes. Lesions too large for primary closure present a unique challenge. This study compares the use of intrauterine bipedicle fasciocutaneous flaps for closure of MMCs not amenable to primary repair. We hypothesized that obstetric, wound, early CSF diversion, and functional outcomes would be similar between groups.

Methods: A retrospective review was conducted of all patients who underwent in-utero MMC repair from 2011-2021 at a single institution post-MOMS. Univariate analysis was conducted using Chi-squared, Fisher's exact, independent samples t and Log-Rank tests.

Results: Twenty-two patients underwent flap closure, while 64 underwent primary closure. Median follow-up time was 1.6 years and ranged from 0-9.6 years. Maternal cohorts did not differ significantly with respect to maternal or gestational age at time of fetal surgery, parity, BMI, or fetal lesion level. Operative times were longer in the flap cohort (32.5±9.93 vs. 18.7±4.88 minutes, $p<0.01$). There were no significant differences in gestational age at delivery (33.09±4.65 vs. 34.31±2.58 weeks, $p=0.13$) or repair site dehiscence (5% vs. 10.17%, $p=0.67$). There were no significant differences in major intra-uterine or postnatal complications. Rates of CSF diversion were similar at one year (33.3% vs. 38.5%, $p=0.70$) and overall at the most recent follow-up (50% vs 41.5%,$p=0.53$). There was no difference in CSF diversion-free survival between groups ($p=0.88$). Of those CSF-diverted by 1 year, 44% overall were by ETV/CPC. Of those with follow-up >2 year, a similar proportion in each group were ambulating independently or with orthotic devices (90.0% vs 79.3%,$p=0.64$).

Conclusion: Larger fetal MMC lesions present a challenge to adequate primary closure. This study suggests that bipedicle flaps can achieve adequate closure without substantially impacting obstetric, wound, early CSF diversion and functional outcomes.



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Abstract #41: Superior Long-Term Appearance of Strip Craniectomy Compared to Cranial Vault Reconstruction in Metopic Craniosynostosis

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Disclosures: The authors have no conflicts of interest to disclose.

Purpose: Strip craniectomy with orthotic helmet therapy (SCOT) is an increasingly supported treatment for metopic craniosynostosis, though its long-term efficacy remains poorly defined. We sought to compare the long-term outcomes of SCOT versus open cranial vault reconstruction (OCVR).

Methods: Patients who underwent OCVR or SCOT for isolated metopic synostosis, with preoperative imaging and over 3 years of follow-up, were identified. Interfrontal angle and interzygomaticofrontal distance were used to stratify patients by baseline severity into two groups, mild-moderate and moderate-severe synostosis. At latest follow up, anthropometric measurements were made from postoperative 3D photos and all patients' parents completed satisfaction surveys. Independent adolescent and craniofacial surgeon raters, blinded to each patient's treatment, rated the appearance of postoperative 3D photos.

Results: Thirty-five patients were included (SCOT n=15 vs. OCVR n=20) with similar follow-up time between groups (SCOT 7.9±3.2 years vs. OCVR 9.2±4.1 years, p=0.33). Postoperatively, the glabellar angle, frontal width, and intercanthal width were equivalent between SCOT and OCVR after stratification by baseline severity (p>0.05, all comparisons). Among patients with moderate-severe synostosis, craniofacial surgeons assigned Whitaker class I to a greater proportion of SCOT patients (72.2%±5.6%) compared to OCVR patients (33.3%±9.2%, p=0.01), though there was no difference among patients with mild-moderate synostosis (SCOT 77.8%±11.1% vs. OCVR 61.9%±15.3%, p=0.34). Adolescents rated the overall appearance and forehead appearance of SCOT patients as "normal" significantly more frequently than OCVR patients, across both severity groups (p<0.05, all comparisons). Parents of patients who underwent SCOT and OCVR reported equivalent levels of satisfaction with overall results of the surgery (100% vs. 95%, p>0.99) and rates of appearance-related bullying (7% vs. 15%, p=0.82).

Conclusions: In metopic craniosynostosis, strip craniectomy with orthotic helmet therapy was associated with superior long-term appearance compared to open cranial vault reconstruction, with equivalent anthropometric measurements and patient satisfaction.

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Abstract #42: Addition by Reduction: The Addition of Wedge Osteotomies in Sagittal Strip Craniectomies May Reduce Helmeting Times and Improve Outcomes

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Introduction & objectives: Strip craniectomy via sagittal craniectomy with post-operative helmeting is recognized as a viable intervention for non-syndromic sagittal craniosynostosis. Previous studies have indicated wide ranges in postoperative helmet therapy (PHT), but largely suggest PHT times around 7-9 months. It has been noted in several studies, however, that helmeting beyond peak correction of craniometrics does not improve long term outcomes. Some previous literature has not supported the addition of non-reduction barrel stave osteotomies to the craniectomy to improve overall outcomes. The aim of our study was to demonstrate that the addition of reduction wedge osteotomies in our patients, when compared to published data, not only significantly reduces the length of optimal PHT (improving cost-effectiveness) but also results in better long-term outcomes.

Material & Methods: An IRB-approved retrospective review of patients treated with sagittal strip craniectomy and reduction wedge osteotomies by a single primary surgeon between January 2013 and July 2021. Age at time of surgery, duration of helmeting, mean duration of follow up, and craniometric data were analyzed. Inclusion criteria included only cases of isolated, non-syndromic sagittal synostosis. The craniometric data tracked were traditional and normative cephalic indices before & after surgery, and throughout helmeting and post-helmeting follow up.
Bilateral wedge osteotomies were performed either anteriorly & posteriorly or anteriorly / posteriorly only. Osteotomies were made posterior to the coronal suture and/or anterior to the lambdoid suture, with care taken not to cross or disrupt the natural sutures, and extending down to or past the squamosal suture.

Results: Eighty-seven patients were identified in this time period who met inclusion criteria. The median age at time of initial surgery was 89 [48-168] days. Initial mean pre-treatment cranial indices were 71.3 [SD +/- 3.98] for traditional CI and 62.9 [SD +/- 5.33] for normative CI. Mean helmeting time was found to be 129 [SD +/- 54.1] days or approximately 3-5 months. Mean traditional and normative CI's at time of helmet discontinuation were 84.2 [SD +/- 3.56] and 82.4 [SD+/- 3.97], respectively. Median follow up to was 22.4 months [range 3-76 mo]. Mean post-helmeting regressions were noted to be -4.2 [SD +/- 2.9] and -3.0 [+/-3.7] for traditional and normative CI's. Subgroup analysis of anterior & posterior vs posterior only osteotomies did not demonstrate significant differences. Duration of helmeting time did not correlate with degree of treatment correction. Further helmeting beyond peak correction did not impact craniometric outcomes or alter the degree of regression over time. No patients have required re-operation to date. Comparisons of our data against several peer reviewed publications are uniformly comparable if not favorable in craniometric outcomes, and superior in regard to helmeting times. All patients were treated with 1-2 helmets, 59% with only one. Usual and customary price per helmet is $3995.00, not including economic costs of travel and time off for orthosis appointments, with variable rates of insurance coverage per family.

Conclusion: Patients who undergo strip craniectomy with reduction wedge osteotomies continue to experience excellent long term primary outcomes, comparable to published results for longer helmeting times, as well as significant reduction of optimal PHT duration by the simple addition of reduction wedge osteotomies and monitoring plateauing of craniometrics. Reduction of PHT duration improves

the cost-effectiveness of treatment. Both regarding degree of correction and degree of regression, our current review suggests comparable if not improved outcomes compared to prior studies that did not utilize wedge osteotomies.

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Abstract #43: Pediatric High-Grade Gliomas

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Pediatric high-grade gliomas including diffuse midline glioma (DMG) harboring the H3K27M mutation are uniformly lethal tumors for which no therapy has found to be effective. Children diagnosed with DMG (previously referred to as DIPG) typically succumb to their disease within 12 months of diagnosis and the 5-year survival rate is less than 1%. In 2012, groundbreaking studies revealed that the majority of these tumors have a somatic mutation of the *H3F3A* gene resulting in and substitution of a methionine at lysine 27 (K27M) on histone H3. This mutation resulting in K27M among H3.3 histones drives the global loss of tri-methylation of K27 wild-type histone proteins. While this increased understanding of DMG genetic and epigenetic biology has unsheathed new potential therapeutic vulnerabilities, nearly 100 DMG clinical trials have failed to show any therapeutic benefit beyond palliative radiation. Currently, this tumor remains a death sentence.

Our lab and others have identified numerous drugs of high interest for treating this devasting disease, however, every trial has failed to show benefit. Is our failure to translate these laboratory findings due to the fact we have not found the right drugs or treatment regimens yet? Or is this a failure to get the drugs to the tumor? Or is this simply a very bad disease to treat? The answer is likely multifactorial. Our lab, in collaboration with others, have strong data to show that simply getting drugs to the tumor will not result in a positive benefit. Most drugs are rapidly cleared from their target space. Utilizing techniques that increase the drug-tumor resonance time is necessary to translate our basic science findings to our patients. Picking the right drug and the delivery mechanism is not enough for a positive effect. Here we will discuss what is necessary to finally deliver effective therapy to these patients and offer hope to the families that need it most.

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Abstract #44: Use of Bipolar Release of the Sternocleidomastoid to Treat Refractory Torticollis in Patients with Normal Skeletal Anatomy and Patients with Congenital Vertebral Body Anomalies

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A pediatric neurosurgeon may be the first provider to recognize or evaluate torticollis in an infant. Torticollis may be primarily realized by referring providers, or it may be the cause of positional plagiocephaly, which could be the primary referring diagnosis to a pediatric neurosurgeon. Conservative treatment such as stretching, physical therapy, and potentially botulinum toxin are mainstays of treatment. For refractory torticollis, Surgical release of a tight or foreshortened sternocleidomastoid may be indicated. The unipolar and bipolar release of the sternocleidomastoid have been used to treat torticollis by plastic and orthopedic surgeons. Pediatric neurosurgeons are technically equipped to perform this operation, and they likely see most of the surgical candidates through plagiocephaly and cervical spine clinics. In addition, bipolar release of the SCM may be indicated for adequate correction of cervical scoliotic deformity. The author will describe the bipolar release of the SCM and illustrate its utility in treating severe torticollis in a patient with normal skeletal anatomy and in a patient with C7 hemivertebra requiring SCM release in conjunction with hemivertebra resection. Technique and outcomes will be described.

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Abstract #48: CDC Head Circumference Charts Not Representative of Racial-Ethnic Groups: A Study of New Mexican Native American Children

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Background: The existing head circumference charts provided by the United States Centers for Disease Control (CDC) lack sufficient data from each racial/ethnic group and patients under the age of 2 years. As a consequence, non-white children, including Native Americans with normal but larger head circumferences than established charts delineate, can inaccurately be diagnosed with macrocephaly and hydrocephalus and undergo unnecessary diagnostic workups.

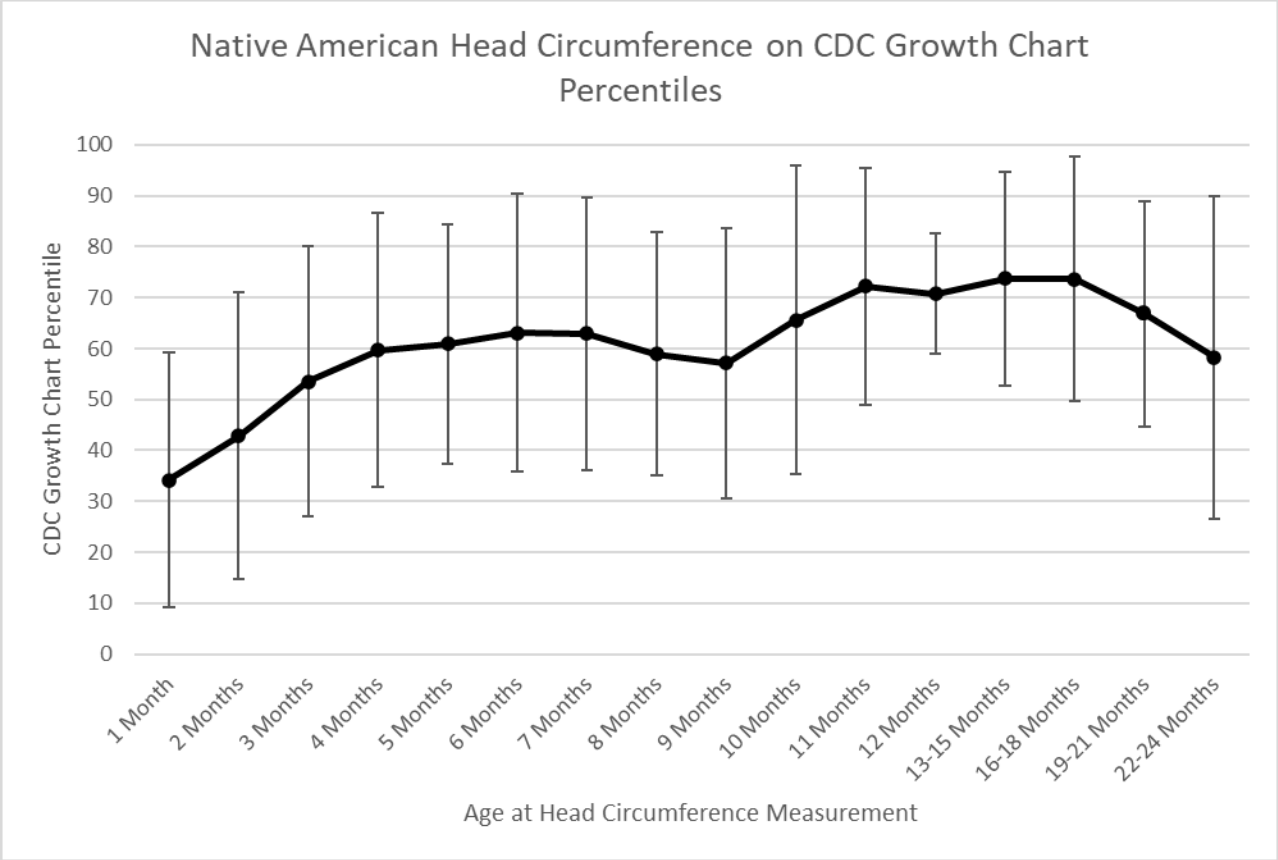
Methods: In this retrospective cross-sectional study conducted at the University of New Mexico, the electronic medical records (EMR) of 100 random New Mexican Native American well-child visits were reviewed. Children with any intracranial pathology, including but not limited to hydrocephalus, tumors, abnormal neuro-imaging, genetic abnormalities affecting the growth of the head, fetal alcohol syndrome, prenatal drug abuse, and preterm birth, were excluded. Head circumferences were entered based on the CDC percentile by gender. The data were then grouped according to age. For the first 12 months, the data were grouped per month of life. For ages 12 to 24 months, the data were grouped per 3-month intervals. A normal curve was created for each group based on CDC growth chart percentiles and the mean and standard deviations were plotted on a graph.

Results: This pilot study included 100 Native American children in New Mexico with a mean age of 9.7 months. Within the first 3 months of life, Native American children had head circumferences that distributed normally around the 50th percentile of the CDC head circumference growth charts. Between 3 to 12 months, the head circumferences for Native American children distributed higher at the 65th percentile of the CDC head circumference growth charts. Between 12 and 24 months, the Native American circumferences peaked at the 73rd percentile at 16-18 months and then decreased to the 58th percentile at 22-24 months.

(See Figure 1. Native American Head Circumference by CDC Percentile)

Conclusion: On average, New Mexican Native American children have larger head circumferences after three months of life than predicted by CDC head growth charts. This discrepancy in head circumference warrants updating the reference charts, as larger head circumferences can lead to inaccurate diagnoses, unnecessary scans and increased anxiety for families.

Figure 1. Native American Head Circumference by CDC Percentile



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Abstract #49: A Standardized, Evidence Based Institutional Approach to Simple Tethered Cord Management

Moriarty, TM, Gump, WC, Mutchnick, IS, Comingore, J, Peppas, D, Rosenberg, E, White, J, Sizemore, E, Moeller, K

Disclosures: none for all authors

Introduction: There is significant evidence in the literature and decades of practice experience that indicate filum sectioning can improve the symptoms of simple tethered cord (TC). The diagnosis of TC and indications for TC surgery vary widely throughout the USA and Canada. The incidence of TC surgery is increasing across USA. We sought to create and evaluate an evidence-based algorithm for the management of tethered cord.

Methods: An institutional standard for TC management was established, based on an EBM analysis of 93 papers from the literature, ranked as per Rutka, J., Neurosurg 126:1747–1748, 2017. A prospective, IRB approved study of all patients treated with these strictly delimited indications in the year 2019 was undertaken. Demographic, clinical, diagnostic, consultative and outcomes data were collected for all patients. Operated patients were seen at 6 weeks and at 6 months for follow-up. Post-operative outcomes were recorded on a four-point ordinal scale (worse, same, better, resolved) for analysis.

Results: 729 unique patients were evaluated for possible TC. 151 operations were done. Two minor complications were seen (wound problem/ UE DVT). All operated patients were clinically symptomatic, 76% of whom had 3 or more symptoms of TC syndrome. 65% of patients were seen by 3 or more subspecialties before surgery; no patient had less than two subspecialties involved in diagnosis. Post op results recorded by neurosurgery were strongly concordant with results observed by consultants. Significant improvement of symptomatic TC was seen across all symptoms (bladder/bowel/back pain/leg pain/gait/HA/tone) in majority of patients.

Conclusion: A low morbidity, high impact intervention guided by a multidisciplinary, evidence-based algorithm can be beneficial to many children.

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Abstract #50: High Social Vulnerability Affects Survival of Pediatric Patients with Abusive Head Trauma: Comparison During COVID-19

Donoho DA; Singer T; Cain C; Risen S; Weiner HL

Background: Abusive Head Trauma (AHT) is a syndrome of life-threatening intracranial injuries. The COVID-19 pandemic imposed new stresses upon socially vulnerable populations, but the relationships between social vulnerability, COVID-19 and AHT outcomes are not known. We investigated whether patient or social factors predicted survival after AHT and whether these factors and outcomes were modified during COVID-19.

Methods: A single-institution database was queried for all admissions of children with a confirmed diagnosis of AHT from 2018-2021. Clinical information, radiographs and clinic follow-up data were reviewed. Social vulnerability index (SVI) was calculated based on published methods (atsdr.cdc.gov). Univariate and multivariate analyses were performed.

Results: One hundred and three cases of AHT were reviewed. Median age at presentation was 4 months (IQR 2-10) in the overall cohort, males outnumbered females overall (76 males, 27 females). 18 patients died (17.5%), higher than previously reported rates. Nonsurvivors had higher social vulnerability index (.867 vs .719, p=0.004); 71% had high social vulnerability compared to 39% of survivors.

There was no difference in fatality rate before (19%) or during (15%) COVID-19.

All nonsurvivors were intubated on admission, compared to 36% of survivors (p<0.001) and all nonsurvivors were comatose compared to 29% of survivors (p<0.001); 61% of nonsurvivors had cardiac arrest on admission compared to 3% of survivors (p<0.001). The injury severity score of nonsurvivors was higher than that of survivors (27 vs 17, p=0.02 in univariate analysis). Nonsurvivors were less likely to have multiple fractures (11% vs. 43%, p=0.01). Nonsurvivors were more likely to have bilateral hypoxic ischemic injury (HII, 89% vs 29%, p< 0.001, Crude OR for survival 0.33, p<0.001, p=0.017 in multivariate analysis). There was no difference in rates or types of neurosurgical intervention, intracranial hemorrhage location, or presence of spinal hemorrhage between nonsurvivors and survivors.

Discussion: Mortality from AHT in our series was higher than previously reported: more than one out of six children in our series did not survive. Although nonsurvivors were more likely to live in highly vulnerable social settings, COVID-19 did not change survival rate. Nonsurvivors are more likely to present in coma requiring intubation and in cardiac arrest. Subdural hematomas are seen in survivors and non-survivors but surgical mass lesions are rare and surgery does not improve survival. We identify a strong association between completed bilateral HII on admission and fatality in AHT. The high mortality of AHT in association with HII, and the low efficacy of intervention after completed HII supports a public health effort towards treatment and prevention focusing on socially vulnerable communities.

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Abstract #51: The Use of In-patient Concussion Consultation in a Cohort of Pediatric Patients to Improve Symptom Score and Associated Health Outcomes: A Proof-of-concept Study

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Objective: To improve patient care and clinical outcomes of mild traumatic brain injury (TBI) patients treated at our institution and validate the effectiveness of in-patient concussion consultation.

Introduction: It is estimated that 10% of all pediatric emergency room visits are due to concussions. Although most children recover without complications, problems including physical, emotional, somatic symptoms and sleep disturbances may occur. We evaluated if early in-patient concussion consultation for mild TBI decreases recovery time, increases symptom reduction, and improves recovery to baseline neurocognitive functioning.

Methods: This is a single institution retrospective study. The control cohort includes patients treated in the emergency department for head trauma (4/12018-9/2019) and the in-patient experimental group (10/2019-4/2021) admitted for head trauma and having received an in-patient concussion consult. Primary clinical endpoints of concussion symptom score at initial clinic follow-up and time to follow-up were assessed and compared between the groups. Secondary analysis was performed to assess symptom score reduction in the experimental group who received both in-patient and outpatient symptom score evaluation along with evaluation if certain covariates and comorbidities impacted symptom score and patient retention.

Results: The Mann-Whitney U Test was used to evaluate the difference in symptom score at initial clinic follow-up between the groups yielding a significant difference of p=1.13E-7. Time to follow-up after controlling for outliers was compared using a two-independent sample student's t-test which yielded a non-significant reduction for the experimental group.

Sub-group analysis for select comorbidities and covariate impact on the experimental group outcomes of patient follow-up and reduction in symptom score respectively were detailed. The comorbidities of ADD/ADHD, and anxiety had significant impacts on both while depression, migraines and other headaches had significant impact on one or the other secondary endpoints. In-patient consultation also resulted in a significant reduction between initial symptom score and symptom score evaluated at initial clinic follow-up of 8.289, p<0.001.

Conclusion: This study highlights the importance of in-patient concussion consultation as an early intervention tool for improving mild TBI outcomes in children. We revealed that it improves outcomes at initial follow-up.

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Abstract #53: Management of Brain Tumor Patients with Medically Refractory Epilepsy

James Baumgartner, MD

Objective: To describe a single institution experience managing pediatric brain tumor patients with medically refractory epilepsy (MRE).

Methods: A retrospective observational study was performed in patients less than 18 years of age who underwent combined brain tumor and epilepsy management at Advent Health for Children between 2017 and 2021.

Results: Eight patients presented to our comprehensive epilepsy center with both brain tumors and medically refractory epilepsy. All subjects underwent a standard phase 1 multidisciplinary epilepsy evaluations. Based upon the results of the phase 1 evaluation, 8 patients underwent a phase 2 evaluation which included video-EEG monitoring with intracranial depth and/or grid electrodes. The scalp and intracranial EEG recordings seen in MRE patients were starkly different than those obtained from MRE patients without brain tumors. Intracranial EEG recordings revealed seizure onset zones both adjacent to and at a distance from the tumor, particularly with DNETs. Once iEEG data were analyzed, a single stage procedures were planned that addressed both the tumor and the cortex identified as epileptogenic. Surgical pathology included ganglioglioma and DNET. With the exception of one patient who experienced a propofol related infusion reaction at the time of electrode placement, gross total tumor resection was accomplished in all patients, and additional cortical resection or ablation was included in six cases. All surviving subjects achieved seizure freedom.

Conclusions: Phase 2 evaluation including placement of intracranial electrodes can be safely achieved in this patient population. An epilepsy evaluation, particularly in cases of DNET, often identifies foci of epileptogenic cortex away from the primary tumor site. Seizure freedom and gross total tumor removal can be achieved in this patient population.

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Abstract #54: Investigating Outcomes following Endoscopic Third Ventriculostomy (ETV) with or without Choroid Plexus Cauterization (CPC) in Patients with Aqueductal Stenosis (AS) and AS plus other Genetic Disorders and Cerebral Malformations (AS+)

Jonathan Pindrik, MD and Jeffrey Leonard, MD

Objective: Endoscopic third ventriculostomy (ETV) with/without choroid plexus cauterization (CPC) represents a popular therapy for obstructive hydrocephalus due to aqueductal stenosis (AS). Unreported in published literature, patients with AS plus genetic and/or neurologic syndromes with cerebral malformations (AS+) may respond poorly to ETV/ETV-CPC, a hypothesis investigated in this study.

Method: This single institution retrospective review investigated electronic medical records and prospectively collected data within the Hydrocephalus Clinical Research Network (HCRN) for pediatric patients with AS or AS+ undergoing ETV or ETV-CPC from 12/2016 through 06/2021. Demographic and clinical data, new onset post-operative seizures, and frequency of ETV/ETV-CPC failure requiring shunt insertion were summarized using descriptive statistics and compared to published outcomes. Treatment outcomes were compared to ETV success scores (ETVSS), a validated percent prediction of ETV success.

Results: Twenty-one subjects total (AS, 12; AS+, 9) underwent ETV/ETV-CPC. Among 12 subjects with isolated AS (median and mean age at surgery, 142.7 months and 122.8 months +/- 78.8 months, respectively), none (0%) experienced ETV/ETV-CPC failure with mean follow-up 22.2 months +/- 17.9 months, comparing favorably with related published hydrocephalus outcomes. Among 9 subjects with AS+ (median and mean age at surgery, 2.3 months and 27.3 months +/- 51.0 months, respectively), 3 (33%) experienced new onset seizures and 7 (78%) experienced ETV/ETV-CPC failure with mean follow-up 19.0 months +/- 13.6 months, exceeding relevant published frequencies for post-operative seizure (1-5%) and treatment failure (18-51%) in AS. Treatment success rates for subjects with AS (100%) exceeded the sample mean ETVSS (82% +/- 12%) while success rates for subjects with AS+ (22%) fell below the sample mean ETVSS (53% +/- 19%).

Conclusion: Patients with AS+ responded poorly to ETV/ETV-CPC regarding seizure and hydrocephalus outcomes. Larger retrospective and prospective studies through the HCRN are needed to verify these preliminary results and justify alternative therapy with shunt insertion.

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Abstract #56: QuickBrain MRI in a Tertiary Pediatric Emergency Department: A Retrospective Study of Use Patterns and Accuracy

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Background: QuickBrain MR imaging (qbMRI) is most commonly used in the pediatric emergency department (PED) setting for the evaluation of patients with surgically treated hydrocephalus. This study describes the use of qbMRI at a single tertiary referral PED for all other indications.

Methods and Findings: We reviewed the medical records and imaging studies of 865 pediatric patients (mean age 6.45 years) evaluated with qbMRI at a single tertiary PED between January 2010 and June 2017 for indications other than previously diagnosed hydrocephalus. Standard MR and/or CT neuroimaging (sMR-CT) completed during the same encounter or at a follow-up encounter within 3 months for the same indication was available for 179 of these patients. Presenting complaints and final diagnoses included: headache, seizure, vomiting, traumatic brain/head injury, and other. Radiologists interpreted 54% of qbMRIs as normal and 12% as showing stable, previously known findings. Of those with comparison sMR-CT available, radiologists interpreted qbMRI as normal in 28%, showing intracranial hemorrhage in 23%, and demonstrating stable, previously known findings in 9%. QbMRI demonstrated a sensitivity of 91% and specificity of 97% for findings evident on sMR-CT. No clinical intervention was undertaken as the result of positive sMR-CT findings in 9 of the 10 patients with discordant qbMRI results. The 10th patient received pharmacological therapy on the basis of ongoing seizures which led to additional imaging without a delay in treatment.

Conclusions: In the pediatric ED setting, qbMRI obtained for indications other than hydrocephalus demonstrated very high specificity when compared to sMR-CT, but was less effective in excluding the presence of minor, generally clinically insignificant findings. No patient suffered delayed clinical care or an altered outcome due to the use of qbMRI as the initial imaging modality. Further multi-center investigation to validate use in various patient care settings is indicated.

Key Words: Neuroimaging; trauma; headache; seizure; hydrocephalus; MRI

Disclosure: Neither author has any conflict of interest with regards to this abstract. This research was supported in part by an Oregon Health & Science University Medical Research Foundation (MRF) grant (Sheridan: PI).

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Abstract #57: Intracerebral Hemorrhage Occurring Years After Revascularization Surgery in Pediatric Moyamoya Patients

R. Michael Scott and Edward R. Smith

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Eight patients in a series of 460 operated for pediatric moyamoya have presented 6 to 27 years after surgery with intracerebral hemorrhage. Seven of these hemorrhages were fatal, and the one patient who survived suffered permanent disability. Although all patients had been taking daily low-dose aspirin, in none had there been any history of bleeding or easy bruising, and two had stable MRI and MRA imaging within 6 months of the fatal bleed. All but one patient's initial pediatric presentation was ischemic. Three of the patients had undergone cranial radiation therapy prior to the onset of their moyamoya syndrome. Pre-morbid CT scans revealed intraventricular hemorrhage with or without basal ganglion involvement. A post-mortem examination in one patient failed to demonstrate an etiology for the hemorrhage. We suggest that pediatric patients operated for moyamoya disease or syndrome undergo lifelong follow-up, with periodic imaging and general physical examination to detect the late development of comorbidities that might increase hemorrhage risk such as hypertension, obesity, hyperlipidemia, etc. -- although it is not clear how any of the deaths in our series could have been prevented. A national database with extended follow-up of operated pediatric patients with moyamoya disease might shed further light on the etiology of these hemorrhages, and provide information to help prevent late morbidity and mortality in these patients.

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Abstract #58: The Neurosurgical Management of Severe Hemophilia A and Moyamoya (SHAM): Challenges and Strategies

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Moyamoya arteriopathy is a rare disorder characterized by progressive stenosis of the terminal branches of one or both intracranial carotid arteries. It is associated with development of abnormal compensatory collateral vessels and can lead to ischemic or hemorrhagic stroke.

Hemophilia is a rare bleeding disorder in which blood does not clot normally. Hemophilia A is an X-linked recessive genetic disorder characterized by absence or deficiency of clotting factor VIII (F8). Severe Hemophilia A occurs in individuals with less than 1% of normal F8 activity and is associated with episodes of spontaneous bleeding.

Recently, a novel association has been identified between Severe Hemophilia A and Moyamoya arteriopathy (SHAM).

The gene for F8 is located on the proximal portion of chromosome Xq28 and contains 26 exons and spans 186 kilobases.

An X-linked recessive form of moyamoya arteriopathy has been identified in association with deletion of the BRCC3 gene, which encodes the deubiquitinating protein, BRCC3, which is involved in angiogenesis. The BRCC3 gene is also located on chromosome Xq28. It contains 11 exons and spans 55 kilobases.

A deletion within chromosome Xq28 that involves both the F8 gene and the BRCC3 gene causes SHAM. These two genes are located close to one another, separated by only 50 kilobases.

The author recently cared for a 5 year old boy with SHAM. Whole genome chromosomal microarray analysis demonstrated an interstitial deletion of 338 kilobases within Xq28 that involved 11 genes, including F8 and BRCC3. The patient had a history of prior spontaneous systemic bleeding, including a severe gastrointestinal bleed. He also had symptomatic moyamoya with multiple bilateral ischemic infarcts.

The patient underwent staged bilateral surgical revascularization with non-anastomotic pial synangiosis bypasses. Strategies for managing the perioperative risk of hemorrhage are described. The importance of recognizing the association between Hemophilia A and moyamoya arteriopathy in SHAM syndrome is emphasized.

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