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MAGNETIC RESONANCE-GUIDED FOCUSED ULTRASOUND FOR ABLATION OF MESIAL TEMPORAL EPILEPSY CIRCUITS: MODELING AND THEORETICAL FEASIBILITY OF A NOVEL NON-INVASIVE APPROACH

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Objective: We tested the feasibility of magnetic resonance-guided focused ultrasound (MRgFUS) ablation of mesial temporal lobe epilepsy (MTLE) seizure circuits. Up to one third of mesial temporal sclerosis (MTS) patients suffer medically refractory epilepsy requiring surgery. Because current treatment options of open surgical resection, laser ablation, and gamma knife radiosurgery pose potential risks such as infection, hemorrhage, and ionizing radiation, and often produce visual or neuropsychological deficits, we aim to develop a non-invasive MRgFUS ablation strategy for mesial temporal disconnection to mitigate these risks.

Methods: 3 Tesla MRI scans with diffusion tensor imaging (DTI) were retrospectively reviewed. The study group included 10 patients with essential tremor (ET) who underwent pre-treatment CT and MRI prior to MRgFUS, and 2 patients with MTS who underwent MRI. Fiber tracking of the fornix-fimbriae and inferior optic radiations was performed, ablation sites mimicking targets of open posterior hippocampal disconnection were modeled, and theoretical MRgFUS surgical plans were devised. Distances between the targets and optic radiations were measured, helmet angulations were prescribed, and the numbers of available MRgFUS array elements were calculated.

Results: Tractograms of fornix-fimbriae and optic radiations were generated in all ET and MTS patients successfully. Of the 10 patients with both the CT and MRI necessary for the analysis, 8 patients had adequate elements available to target the ablation site. A margin (mean 8.5 mm, range 6.5 – 9.8 mm) of separation was maintained between target lesion and optic radiations.

Conclusions: MRgFUS offers a non-invasive option for seizure tract disruption. DTI identifies fornix-fimbriae and optic radiations to localize optimal ablation targets and critical surrounding structures, minimizing risk of postoperative visual field deficits. This theoretical modeling study provides the necessary groundwork for future clinical trials applying this novel neurosurgical technique to patients with refractory MTLE and surgical contraindications, multiple prior surgeries, or other factors favoring non-invasive treatment.

Key Words: focused ultrasound; mesial temporal sclerosis; disconnection surgery; fornix-fimbriae; tractography; diffusion-tensor imaging
USE OF LAPAROSCOPY FOR PERITONEAL SHUNT CATHETER PLACEMENT

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Introduction: Ventriculoperitoneal (VP) shunt placement is one of the most common treatments for hydrocephalus in children and has a failure rate of up to 40% within the first year. Reoperation is frequently required to revise the intraperitoneal portion of the shunt. There has been a trend toward laparoscopic-assisted placement of the intraperitoneal portion of the shunt but there is little data regarding its potential benefit. We examine the outcomes of laparoscopic assisted versus open placement of VP shunt.

Methods: We performed a retrospective chart review of all pediatric (<18 years old) neurosurgical cases from July 2012 through June 2017 performed at a single institution. Surgery was performed by either the neurosurgeon alone (open peritoneal catheter placement) or with a pediatric surgeon (laparoscopic-assisted peritoneal placement). Data obtained include patient age, surgery performed, surgical control time, length of stay, and short term follow up of any medical complications.

Results: 230 patients underwent new VP shunt placement during the study period. Patient demographic data between the two groups was similar. There was a significant decrease in surgical time in the laparoscopic-assisted shunt placement group, with the average surgical control time of 32 minutes versus 58 minutes in the open group. There was no significant difference in complications or readmissions within 90 days. There was a trend toward decreased length of stay in the laparoscopic-assisted surgery group.

Conclusion: For patients undergoing new VP shunt placement, there is a decrease in surgical time with laparoscopic-assisted placement of the peritoneal catheter as opposed to open placement. There was also a trend toward decreased length of stay in this group. Outcomes and safety are consistent between the two groups.
INCIDENCE AND MORTALITY IN POST-HEMORRHAGIC HYDROCEPHALUS (PHH) REQUIRING NEUROSURGICAL INTERVENTION AMONG NEONATES WITH SEVERE INTRAVENTRICULAR HEMORRHAGE (IVH) IN CALIFORNIA

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Keywords: Intraventricular hemorrhage (IVH), Post-hemorrhagic hydrocephalus (PHH)

**Objective:** Recent evidence demonstrates decreased incidence of severe (grade III or IV) intraventricular hemorrhage (IVH) in infants born prior to 32 weeks gestation in California - from 9.7% in 2005 to 5.9% in 2015 - following changes in standardized obstetrics and delivery room practices for premature births (increased antenatal steroid administration and decreased delivery room intubations). The aim of this study is to determine if decreased rates of IVH correlated with decreased rates of post-hemorrhagic hydrocephalus (PHH) treatment in the same cohort.

**Methods:** This observational cohort study utilized clinical admissions and discharge data collected prospectively in the California Perinatal Quality Care Collaborative (CPQCC). Infants born at 22⁹/⁷⁻31⁶/⁷ weeks gestation with severe IVH were included. The primary outcome measure was PHH requiring temporizing or permanent neurosurgical intervention.

**Results:** From 2008 to 2016, 7% of infants with severe IVH (238 of 3380) were treated with a temporizing neurosurgical intervention (ventricular access device, ventriculosubgaleal shunt, or external ventricular drain). 11.8% of infants with severe IVH required permanent CSF diversion (shunt) (398 of 3380). Of those who underwent a temporizing neurosurgical intervention, 78% later underwent permanent CSF diversion. Of those requiring permanent CSF diversion with a shunt, 54% underwent shunt placement as the primary surgical procedure, with no prior temporizing surgical procedure. No decrease in rates of temporizing or permanent CSF diversion were observed from 2008 to 2016. In-hospital mortality was observed in 37% of infants with severe IVH, with lower mortality observed in infants treated for PHH: 12.7% for temporizing intervention and 7.6% for permanent CSF diversion (shunt) (p<0.001).

**Conclusion:** Despite decreasing rates of severe IVH in premature infants, no corresponding decreases in rates of PHH treatment are observed in California. Further investigation is needed to determine hospital level and regional variations in practice that may account for this.
THE IMPACT OF MULTI-DISCIPLINARY ENGAGEMENT IN BLOOD CONSERVATION PROTOCOLS FOR CRANIOSYNOSTOSIS

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Background: Patients undergoing open cranial vault remodeling for craniosynostosis frequently experience substantial blood loss requiring blood transfusion. Multiple reports in the literature have evaluated the impact of individual blood conservation techniques in blood transfusion rates during craniosynostosis surgery. We engaged a multi-disciplinary team and were able to assess the impact of multiple stakeholder input into the evolution of a comprehensive protocol.

Methods: Over a 4-year period from 2012-2016, thirty-nine non-syndromic patients were operated on by a single craniofacial plastic surgeon. In 2014, a new pediatric neurosurgeon joined the craniofacial team, and additional stakeholders in anesthesiology, transfusion medicine, critical care, and hematology were brought together to evaluate opportunities for development of a blood conservation protocol. Evolution of the comprehensive protocol involved initial standardized administration of intraoperative aminocaproic acid (ACA) followed by the addition of preoperative erythropoieten (EPO) and usage of the cellsaver device. In addition to this, resuscitation and transfusion guidelines were more clearly defined. The primary outcomes of estimated blood loss, transfusion rate and intraoperative transfusion volume were analyzed. The secondary impact of the multidisciplinary stakeholder input was inferred by the trends in the data with the implementation of the partial and full protocols.

Results: Administration of EPO significantly increased starting hemoglobin. The group of patients receiving ACA had lower intraoperative EBL than those not receiving ACA, and trends of transfusion volume in the final cohort which received both preoperative EPO and intraoperative ACA demonstrated decreasing transfusion volumes though it did not reach statistical significance. The final protocol patient cohort had a 66% transfusion-free rate at the time of discharge.

Conclusion: Patients undergoing open calvarial vault remodeling procedures benefit from the input of a multi-disciplinary stakeholder group. Further research into comprehensive protocols for blood conservation may benefit from input from the full surgical team (plastic surgery, neurosurgery, anesthesiology) as well as additional pediatric subspecialty stakeholders including transfusion medicine, critical care, and hematology.
IS HOUSEHOLD ESTIMATED INCOME ASSOCIATED WITH PROPER SEATBELT USE IN PEDIATRIC PATIENTS INVOLVED IN A MOTOR VEHICLE CRASH?

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Introduction: Injury prevention relies on identifying associated risk factors associated for patterns of injury. While low income families are known to be an at risk population for proper child restraints, Survey data in 2013 suggested that high income families were more likely to make exceptions about properly restraining their children than low and middle income families. However no study has confirmed the survey results in actual trauma patients. We used the pediatric trauma database at our institution, to examine the effect of income on seatbelt use in motor vehicle crashes (MVC).

Methods: We examined a prospective pediatric trauma database at a single academic institution for children <16 years old involved in an MVC. Patients with unknown seatbelt or proper restraint use were excluded. Age, admission and discharge information, mortality, presence of head or spine injury, injury severity score, and zip code were all examined to identify factors influencing trauma outcomes. Zip code was referenced in the Proximal One database to estimate household income, and classify patients as low, middle, and high income. Fisher’s exact test was used to compare the groups (p<0.05)

Results: 495 patients met inclusion criteria, 48 were excluded due to unknown restraint use. And 30 excluded for unknown income. Of the remaining 417 patients, 206 (49%) were female. Average age was found to be 8.7 years in those properly restrained,9.4 in those not. Average household income was $45,915 in those properly restrained, $45,063 in those not. There was a significant difference the observed and expected use of proper restraints and income.

Discussion: We found a higher incidence in pediatric patients in the middle income group, however no statistically significant difference among income groups and proper restraints. This supports prior survey data, and may provide an important area of focus for education to prevent injuries.

References
WORKING TOWARDS TARGETED THERAPY IN DIPG TUMORS: AURORA KINASE INHIBITORS AND THE ROLE OF H3SER28 PHOSPHORYLATION

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DIPG is one of the most aggressive pediatric brain tumors with no effective therapies. Majority of the midline high-grade pediatric gliomas carry somatic mutations of histone H3 resulting in the replacement of lysine 27 by methionine (K27M) and global loss of methylation on H3K27. In an effort to identify drugs for rapid clinical translation, we previously performed rigorous high-throughput drug and shRNA knockdown screens to identify potential molecular targets. As a class of compounds, the aurora kinase inhibitors (AKI) were one of the strongest hits, as these drugs potently reduced cell viability and proliferation of H3K27M tumors and also restored H3K27 trimethylation.

We confirmed distinct expression of Aurora Kinase-related genes in DIPG patients compared to normal brain and patients with WT high grade gliomas—these results extended to our H3K27M xenografts. Alisertib is a brain penetrant AKI that is orally available and already found to be safe in children which will allow for rapid translation to the clinic. Importantly, gene expression of twelve AK related genes that we identified in our H3K27M cell lines correlated directly with sensitivity of that cell line to alisertib.

We validated the therapeutic effects of alisertib in patient derived orthotopic xenografts and observed a decrease in tumors size and increase in survival. To understand the epigenetic effects of AK inhibition on H3 chromatin, we evaluated the phosphorylation status of important H3Ser residues with AKI treatment. Importantly we observed a decrease in H3Ser28 phosphorylation with treatment of AKIs and concurrent increase in H3K27 trimethylation which is a similar pattern found in stem cell programming and differentiation. The summation of these results supports the hypothesis that Aurora Kinases are critical for epigenetic programming in H3K27M tumor cells and represents a targeted approach for treating tumors with this mutation.
Brains grow rapidly in early childhood. They achieve their most substantial growth during the first 2 years of life, which intersects with the neurosurgical management of infant hydrocephalus. Although we drain and divert fluid to treat hydrocephalus, our ultimate goal should be to grow brains. Over the past 7 years, we have explored a wide-ranging exploration of brain growth. We demonstrated the feasibility of normative brain volume growth in mice, contrasted against different patterns of hydrocephalus. We also demonstrated that brain volume is correlated with neurocognitive function in human hydrocephalus, and that there is growth arrest and catch-up growth with successful treatment of hydrocephalus in infants. Most recently, we have demonstrated the construction of normal infant male and female clinical brain growth curves. Synthesizing these results, I discuss the potential for the use of brain growth curves in the management of childhood hydrocephalus.
TWO-YEAR COGNITIVE OUTCOMES IN PREMATURE INFANTS WITH POST-HEMORRHAGIC HYDROCEPHALUS CORRELATES WITH TOTAL CSF VOLUME TAPPED

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Background: Intraventricular hemorrhage (IVH) in premature infants of very low birth weight (<1500g) is a major cause of morbidity. The incidence of IVH in this cohort of patients has decreased from 50% to about 20% as of 2005, however the incidence of posthemorrhagic hydrocephalus (PHH) continues to be present in 25-50% of patients with IVH and is associated with long-term neurodevelopmental impairment and shown to increase cognitive and psychomotor delay. There is no consensus regarding timing or type of surgical intervention for treatment; with shunt or ventriculostubgaleal shunt (VSGS). We hypothesize that CSF removal with frequent ventricular reservoir taps results in improved two-year neuro-cognitive outcome compared to infrequent taps in premature infants with VSGS for PHH.

Methods: The surgery database containing all records of patients who underwent VSGS at a single institution between 2006-2014 was reviewed, and their birth weight, sex, IVH grade, age at VSGS placement, ventricle size at VSGS placement, maximum/minimum ventricle size, ventricle size at 2 years, number of VSGS taps, volume tapped at each event, cognitive scores at 2 years (Bayley scores) and history of shunt infection/malfunction/seizures were recorded in a secured database.

Results: There were 32 patients, 15 females (47%), with an average birth weight of 808 grams. Eighteen (56.3%) had grade IV, 9 had grade III, 4 had grade II and one had grade I IVH. 13 patients had their VSGS tapped at least one time. In a general linear model, total volume tapped correlated with Bayley scores at 2 years (p=0.025). Average ventricular volume had no correlation with Bayley scores at 2 years (p= 0.489). Total volume tapped ranged from 2-105.5 cc over the lifetime of the VSGS.

Conclusions: Total volume tapped during the lifetime of a VSGS leads to improved Bayley scores at two years of age. Ventricular size did not correlate with outcomes.
SHUNT FAILURE – THE FIRST 30 DAYS

Paul Klimo

Background: Shunt malfunction continues to occur at an unacceptable rate. Incontrovertible predictors of malfunction remain elusive.

Objective: To determine predictors of shunt failure within the first 30-days of index surgery.

Methods: This was a single-center retrospective cohort study. We used a research database of all ventricular shunt operations from January 2010 through November 2016. For each shunt operation, demographic, clinical and procedural variables were procured. An “index surgery” was defined as implantation of a new shunt or revision or augmentation of an existing shunt system. The primary outcome was shunt failure of any kind within the first 30-days of index surgery. Bivariate models were first created followed by a final multivariable model using a backward-forward selection procedure.

Results: Our data set contained 655 unique patients with a total of 1206 operations. The median age for the cohort at the time of first shunt surgery was 5 years (range, 0–35.7) with 56% males. The 30-day failure rates were 12.4% (when analyzing the first index operation only, 81/655) and 15.7% (when analyzing all index operations, 189/1206). Small or slit ventricles at time of index surgery and prior ventricular shunt operations were found to be significant covariates in both the “first index” (p=0.0026 and p=0.050) and “all index” (p=0.021 and <0.001) multivariable models. Intraventricular hemorrhage at time of index surgery was an additional predictor in the all index model (p=0.007).

Conclusions: This study demonstrates that only 3 variables are predictive of 30-day shunt failure, 2 of them being potentially under direct control of the surgeon. Our findings suggest that drivers of shunt failure may be a function of time from index surgery.
RESULTS OF AN ELECTIVE “SHUNT REMOVAL PROTOCOL”

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Introduction: Complications related to cerebrospinal fluid (CSF) shunt dependence carry significant morbidity, mortality, inconvenience, and expense. The availability of endoscopic options to treat hydrocephalus has obviated the need for shunt dependence in many patients. We report the results of a 2 or more-stage protocol whose goal was removal of dependence on CSF shunting.

Methods: We reviewed the clinical, radiological, and surgical data of patients who were admitted to the hospital for attempted elective shunt removal. Patients were identified as candidates in clinic if they had symptoms such as positional headaches. Patients were electively admitted and underwent shunt externalization and closure. If they proved to be shunt independent the shunt was removed or tied off in a second stage. If the patient proved to be dependent on the shunt we attempted to induce ventriculomegaly, perform an ETV, place a frontal Omaya tapping reservoir and remove the shunt. Patients with ETV were monitored/weaned with a temporary EVD placed at the time of shunt removal. In patients who could not be weaned, developed csf leak, or developed delayed symptomatic ventriculomegaly, the shunt was replaced.

Results: From 2012 -2018, 34 consecutive patients (20 female:14 male) were studied. Average age was 16.6 years (range 0.6-60.7 years). 25 patients were less than 22 years. Patients had been shunt-dependent for a mean of 7.7 years. Eleven (32.4%) patients demonstrated no need for cerebrospinal fluid diversion and tolerated removal of their shunt systems without further treatment. The 23 csf diversion-dependent patients underwent an ETV after induction of ventriculomegaly. Thirteen (56.5%) of 23 patients were shunt independent at last follow-up (mean 14.4 months). Overall, 24 patients (70.6%) were shunt independent at last follow up (mean 13.3 months). Two patients suffered temporary complications (csf infection, transient diplopia). During follow-up, 100% of patients/parents, regardless of success, stated that they would repeat the process again if offered the same options.

Conclusions: A significant number of patients previously believed to be shunt-dependent can safely have their shunts removed or be managed with ETV using this stepwise “shunt-removal protocol.” Shunt removal should be considered the ultimate goal for patients harboring CSF shunts.
DIRECT VISUALIZATION OF FENESTRATION WITH PRECISE CATHETER PLACEMENT IN LOCULATED HYDROCEPHALUS USING SELDINGER TECHNIQUE: A REPORT OF THREE CASES

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Topic: Technical Pearls

Introduction: Hydrocephalus from neonatal intraventricular hemorrhage and meningitis remains a significant source of morbidity. Loculations due to scarring create complex situations. Surgical treatment presents challenges with short and long-term complications. We present a surgical technique for endoscopic placement of ventricular catheters through directly visualized fenestrations.

Methods: The first two patients, five and fourteen months old, both with history of prematurity and intraventricular hemorrhage with ventriculoperitoneal shunts presented with entrapment of the fourth ventricle. Using a frontal entry point with image guidance and an endoscope, the fourth ventricle was fenestrated under direct visualization through the atrium and quadrigeminal cistern. With concerns that the fenestrations would close, a novel technique placing a catheter was completed. A 0.27mm pediatric central line guidewire was placed through the endoscope into the desired location in the fourth ventricle. The endoscope was carefully removed and a shunt catheter passed over the guidewire after cutting a small slit in the tip. The guidewire was removed and replaced with the image guidance probe which confirmed the location of the tip of the catheter in the fourth ventricle. The catheters were then connected to the patients’ existing shunt. A third patient that had meningitis and developed significant scarring and loculations had fenestration into a trapped third ventricle through thickened tissue. The same technique was used to precisely place the catheter through the already created hole into the third ventricle.

Results: The first two patients recovered well and were discharged on post-operative day one. Follow up imaging showed decompression of the fourth ventricle and good placement of the fourth ventricular catheter. The third patient’s immediate post-op scan showed excellent placement of the catheter. No one had complications or new deficits from catheter placement.

Conclusion: An endoscope can be used with Seldinger technique to precisely place shunt catheters in complex ventricular systems and challenging locations with directly visualized fenestrations avoiding potential complications.

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INTRATHORACIC MIGRATION OF DISTAL VENTRICULOPERITONEAL SHUNT CATHETER VIA A MORGAGNI HERNIAL: CASE REPORT AND LITERATURE REVIEW

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Case summary: A former 23-week gestational age infant developed post-hemorrhagic hydrocephalus and was initially managed with a ventriculo-subgaleal shunt. At 3 months of age, this was converted to a left ventriculoperitoneal shunt in an unremarkable surgery. At 5 months of age, the child returned with signs and symptoms of shunt malfunction and imaging demonstrated that the distal catheter was in the left pleural cavity, with significant pleural fluid. A distal shunt revision was done by the initial surgeon.

At 7 months of age he returned with clinical evidence of shunt malfunction and the shunt was explored. No clear obstruction was found, but subsequent work-up of respiratory distress revealed pleural fluid and evidence of a left anteromedial diaphragmatic hernia, with migration of small bowel into the left pleural space. Laparoscopic repair of the Morgagni hernia was done without incident. At two months follow-up, the child was doing well.

Discussion: Congenital diaphragmatic hernias (CDH) most often occur in the posterolateral region of the diaphragm. Commonly known as Bochdalek’s hernia, these account for 95% of all CDH. Less than 2% occur anteriorly, usually near the xiphoid, and are referred to as retrosternal, or Morgagni, hernias. Fewer than 10 cases of pleural migration of a peritoneal shunt into the pleural cavity have been identified, and this case represents only the second confirmed migration via a Morgagni hernia, which was diagnosed in a delayed fashion after shunt revision. The literature and case management will be discussed.
IMMUNOPARALYSIS AFTER TRAUMATIC BRAIN INJURY WITH HEMORRHAGIC SHOCK: A JUVENILE RAT MODEL OF POLYTRAUMA

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**Introduction:** Traumatic brain injury in children is known to cause immune suppression. When severe, this immune suppression has been termed immunoparalysis. Polytrauma involving a traumatic brain injury and an extra-cranial injury may increase this degree of immunoparalysis. Suppression of the immune response can lead to an increased risk of developing nosocomial infections, potentially causing secondary brain injury and worsening patient outcomes. Despite the prevalence of polytrauma with traumatic brain injury in children, mechanisms of immune suppression following such injuries remain poorly understood.

**Methods:** We developed an animal model of traumatic brain injury and hemorrhagic shock to assess immune function after injury. Pre-pubescent rats were injured using a prefrontal controlled cortical impact method and a controlled hemorrhage by femoral arteriotomy. The degree of brain injury was sufficient to produce significant deficits in spatial memory testing. Immune function was measured by examining plasma cytokine concentrations, measuring the percentage of monocytes by flow cytometry, and by whole blood ex-vivo TNFα production capacity following incubation with lipopolysaccharide.

**Results:** Both isolated hemorrhage and traumatic brain injury with hemorrhage (combined injury) significantly reduced plasma concentrations of inflammatory cytokines. Only combined injury (traumatic brain injury and hemorrhage) correlated with reduced concentration of monocytes and reduced TNFα production capacity at post-injury day 1.

**Conclusion:** These results demonstrate that a juvenile animal model of traumatic brain injury with polytrauma can be used to study post-injury immune suppression. Our results indicate that traumatic brain injury with polytrauma leads to reduced plasma inflammatory cytokines and whole blood TNFα production capacity. Traumatic brain injury with polytrauma is associated with a reduction in circulating monocytes and reduced MHCII expression on circulating monocytes. These effects may be responsible for the depressed overall immune response seen following the combined injury.
THE VALUE OF COMPUTED TOMOGRAPHY AND MAGNETIC RESONANCE IMAGING IN DIAGNOSING PEDIATRIC THORACOLUMBAR COMPRESSION FRACTURES

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**Purpose:** To examine the sensitivity of CT and MRI in diagnosing pediatric thoracolumbar compression fractures.

**Methods:** Patients <18 years at a single Level 1 pediatric trauma center (1/2013-12/2016) with a mild thoracolumbar compression fractures (< 30% loss of height) were retrospectively identified. Patients who underwent both CT and MRI were included. Patients with complex spinal injuries (burst and fracture-dislocations) were excluded. MRI was used as the standard for presence or absence of injury.

**Results:** 52 patients (21 male/31 female, mean 10.6 yrs) fit inclusion criteria. There were 191 individual fractures identified (3.7 per patient). Ten patients (19%) had a single level injury. Of those with multiple levels, 81% (34/42) were contiguous and 19% (8/42) had noncontiguous. Fracture distribution was bimodal, with the most common locations in the mid-thoracic spine (T3-T6) and thoracolumbar junction (T12-L1). Complete imaging agreement in fracture number and distribution was noted in 23 patients (44%). MRI identified additional levels in 15 patients (29%, mean 3.3), whereas 14 had fewer fractures on MRI than CT (27%, mean 2.0). Two patients had injuries on CT who subsequently had a normal MRI (false positive). Only one patient had fractures on MRI after a normal CT (false negative). Patients 11-14 years had 82% correlation (9/11) between CT and MRI, compared to 34% (14/41) outside of this range. Sedation for MRI was required in 29% (15/52; mean 8 yrs) and 60% (9/15) were < 10 years of age. All patients were managed non-operatively.

**Conclusion:** In children with thoracolumbar compression fracture(s), CT demonstrates high sensitivity in determining the presence or absence of a fracture. While variability exists in the number of levels, the addition of MRI did not change treatment or outcome. The information obtained from MRI must be weighed against time and cost, as well as risk associated with sedation when necessary.
A PRELIMINARY STUDY ON HOW A NEW MULTI-MODAL PRE-SURGICAL EVALUATION STRATEGY HAS IMPACTED POST-SURGICAL OUTCOMES FOR POORLY-DEFINED FOCAL EPILEPSY IN CHILDREN

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Introduction: Starting in November 2014, we modified our pre-surgical evaluation strategy for patients referred to the Montreal Children’s Hospital with poorly defined focal epilepsy. Instead of simply relying on gross concordance between electroencephalography (EEG) and 3T magnetic resonance imaging (MRI), and performing PET, SPECT, and subdural grid intracranial recording on a case by case basis, we systematically performed EEG, 3T-MRI (including perfusion sequences), positron emission tomography (PET), single photon emission computed tomography (SPECT), and magnetoencephalography (MEG) on all patients. If these studies did not provide a strong hypothesis regarding the epileptogenic zone (EZ) and its borders, EEG-fMRI and advanced MRI post-processing with voxel-based methods were performed. Finally, if these combined methods did not provide a good definition of the EZ, intracranial recordings, preferably with stereoelectroencephalography (SEEG), was used. The goal of the current study is to examine preliminary data on how our new pre-surgical evaluation strategy has impacted post-surgical outcomes for poorly defined cases (PDCs) of pediatric focal epilepsy.

Methods: Chart and imaging review was performed for PDCs of focal epilepsy who followed the new pre-surgical evaluation strategy and underwent surgical resection since 2014. We compared this group of patients to the group of patients operated on before the new strategy was implemented. STATA 15 was used for statistical analysis.

Results: Fifteen patients operated on following the new pre-surgical evaluation protocol were compared to 10 patients using the previous protocol. Overall, the average age was 110.9 months at the time of surgery, patients were using an average of 3.0 antiepileptic drugs (AEDs) pre-operatively, and 40% of the lesions were temporal versus 60 % extra-temporal. These factors were not significantly different between the new protocol patients and previous protocol patients (p> 0.05). The patients all had very frequent seizures, most having several seizures a day in the period preceding surgery. The patients worked up with the new protocol had more tests performed than the previous protocol patients with 5.4 tests performed on average versus 2.6 tests (p < 0.0001, Odds Ratio 2.8 CI95 1.6-3.9). In the new protocol group, 5 patients underwent invasive neuromonitoring (2 SEEGs, 1 grids case, 2 combined grids and depth electrodes). In the previous protocol group, three patients had invasive monitoring (3 grids). In the new protocol group, 13 of the 15 patients (86.7%) were seizures-free at the time of last follow-up, and for patients followed at least 1 year, 7 of 8 patients (87.5%) had decreased their number of AEDs used. For patients treated with the previous protocol, there were 5 of 10 patients (50%) who were seizure free at last follow up, and 5 of 10 (50%) had a decrease in number of AEDs prescribed. The average follow-up for the new protocol patients was 12.2 months, and the average follow up was 59.2 months for the old protocol patients. On univariate analysis, there was no significant difference in seizure-freedom between patients investigated with the new
protocol versus the previous protocol (Odds Ratio 6.5 CI 95% 0.94 – 45.10) for this preliminary data.

**Conclusion:** Since the introduction of a new protocol for the work up of patients with PDCs of focal epilepsy, there has been an increase in the percentage of patients who have become seizure-free at last follow-up, from (5/10) 50% to (13/15) 86.7%. However, this difference was not found to be statistically significant on univariate analysis, perhaps due to the small numbers and statistically underpowered nature of this preliminary study. Future research will aim to better quantify the effect of using systematic advanced neuroimaging and intracerebral recording (SEEG and depth electrodes) for the pre-surgical evaluation of PDCs of focal epilepsy in children.
COUPLING OF CALCIUM CHANNEL ACTIVITY, OPTICAL INTRINSIC SIGNALS, AND LOCAL BLOOD OXYGENATION DURING CORTICAL SPREADING DEPOLARIZATIONS

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**Introduction:** Optical intrinsic signal imaging (OISI) is an established tool for assessing cortical spreading depolarizations (CSD) and offers potential for clinical application. OISI analysis can include information about local blood oxygenation (BOLD) as well as neuronal activation. To better understand the relationship between neuronal polarization states and the OIS in the intact *in vivo* cortex, we examined simultaneous OISI data and calcium channel activation in transgenic GCaMP6f mice during stimulus induced (SI-CSDs) as well as terminal spreading depolarizations (TSD).

**Methods:** Thy1-GCaMP6f (n=8) heterozygous C57Bl6 mice underwent surgery with isoflurane anesthesia to install a cranial window while fixed in a stereotaxic frame. Imaging took place on a Nikon Ti-E SpectraX system with ~10 µs fast gating acquisition of 488 nm fluorescence and reflected light from 555 and 640 nm channels. A capillary tube pulled to a sharp tip was inserted into the cortex through the cranial window to induce a CSD. Inhaled isoflurane was increased to 5% to investigate TSDs.

**Results:** A cortical pinprick or isoflurane overdose consistently initiated CSDs. However, GCaMP activation traveled faster following the pinprick (4.25 mm/min) compared to ischemic events (2.63 mm/min). In SI-CSD, compared to the calcium changes, the OIS peak increase was delayed by 18 s. However, the OIS peak was delayed by 35 s in TSD events. Spatial coupling was excellent with consistent delays across selected regions of interest. Simultaneous use of 655nm light allowed for isolation of the neuronal component of the complex OIS signal.

**Discussion:** Fast gated multi-channel imaging allowed the characterization of neuronal activation and provided additional information about local tissue oxygenation levels from the simultaneous OISI. CI-CSDs initiated a subsequent increase of oxygenated blood in the region; isoflurane was associated with an anticipated ischemic decrease in oxygenation before TSD, followed by an increase just prior to neuronal activation. Both the calcium channel signal propagation through the cortex and OIS data match previously reported velocity and alterations in blood oxygenation. The OIS changes in *in vivo* cortex appear to couple to calcium changes differently in SI-CSDs vs. ischemic TSDs temporally but not spatially. These changes may not be entirely explained by BOLD differences.
OUTPATIENT ASC PEDIATRIC NEUROSURGERY

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Introduction: Previous reports have provided evidence that appropriately selected patients and procedures can be performed efficiently and safely in ambulatory surgery centers. Initially this occurred with spinal and peripheral nerve interventions. More recently, appropriate adult intracranial procedures have been completed at ASC, including biopsy and endoscopy. We demonstrate that for appropriate pediatric patients, several neurosurgical procedures can be performed safely and routinely at ASC, and present our initial experience with pediatric neurosurgical procedures performed at an ASC.

Methods: We performed a retrospective chart review of all pediatric neurosurgical cases from April 2012 through August 2018 performed at a single ASC jointly owned/managed by a large neurosurgical practice and large healthcare system. Data obtained include patient age, surgery performed, length of stay at the surgery center, and short term follow up of any medical complications.

Results: Seventy-two pediatric neurosurgical procedures were performed during the study period at the ASC. All cases were performed under general anesthesia or laryngeal mask airway anesthesia. Patient median age was 16 years and age range was 2 to 18 years. 18 of the patients were 12 years of age or less. Median length of stay from start of surgery to discharge was 5 hours with a range of 2 to 9 hours. There were no surgical or anesthetic complications. Outcomes were in line with patients whose procedures were done as inpatients during the same period. No patient required admission to the hospital for complications related to surgery within the first 30 days post-operatively. One patient presented to the emergency department post-operative day 2 for psychogenic non-epileptic seizure (PNES).

Conclusion: For appropriately selected pediatric patients and indications, neurosurgical procedures are performed with safety at an ASC and discharged rapidly. Outcomes and safety are consistent with inpatient procedures.
ONCOLYTIC ADENOVIRUS FOR RECURRENT MALIGNANT BRAIN TUMORS IN CHILDREN

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High-grade pediatric brain tumors encompass a number of different diseases including high-grade glioma (pHGG); atypical teratoid rhabdoid tumor (AT/RTs), primitive neuroectodermal tumor (PNETs) or, ependymoma. Despite the improvement of diagnostic and therapeutic approaches, children with malignant brain tumors have a very poor prognosis representing the leading cause of cancer death in children. It is clear that a therapeutic paradigm change is needed to improve the survival and quality of life of children with these tumors. Delta-24-RGD (DNX-2401 in the clinic) is a replication-competent adenovirus that has already shown efficacy in animal models of adult gliomas. Of clinical significance, previous clinical trials in adults indicate that. DNX-2401 has a favourable safety profile, strong tumor-killing potential and can trigger an antitumor immune response. We developed preclinical data regarding the efficacy and safety of this virus in relevant immunosuppressed and immunocompetent models of pHGG, DIPGs, medulloblastoma, AT/RTs and PNETs.

Therefore, with the background of the clinical studies in adults and the promising preclinical data in pediatric models we designed a phase I dose escalation trial for recurrent high grade tumors in children. Patients receive an stereotactic intratumoral injection of DNX-2401. For those who presented with a potential resectable tumor and measurable disease 2-3 weeks after the initial injection, resection of the tumor would follow 21 days after the first surgery to obtain post-treatment samples. The aims of the study are to determine the safety, tolerability and toxicity of DNX-2401 and to assess the efficacy of the treatment as the objective response rate, the 6 months progression free survival (PFS-6) and 12 months overall survival (OS-12). At the meeting we will present the last update of the trial. We will present the current status of the trial and the preclinical data at the meeting.
COMPLICATION IN PATIENT MANAGEMENT:
METASTATIC RENAL CELL CARCINOMA IN VON HIPPEL LINDAU DISEASE WITHOUT
APPROPRIATE SCREENING

Robert P Naftel

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A 17yo male with von Hippel Lindau Disease (VHL) was under the care of neurosurgery for management/surveillance of multiple CNS hemangioblastomas. During this care, he underwent a cervical laminoplasty for resection of a symptomatic, enlarging cervical spinal cord hemangioblastoma. He was seen by neurosurgery every 3 months with surveillance MRI’s because of multiple enlarging posterior fossa and spinal cord tumors. He had been lost to follow up in the oncology clinic for other screening surveillance. On a spine MRI he was noted to have multiple metastatic tumors. Systemic workup and biopsy diagnosed metastatic renal cell carcinoma. He had not been receiving appropriate VHL screening examinations that may have detected this disease process prior to metastasis.

Although it is not necessarily the responsibility of the neurosurgeon to manage such surveillance, the neurosurgeon should be aware of this surveillance and ensure that proper surveillance is occurring. In this case the neurosurgeon did not ensure that the patient was receiving other appropriate care and remained only focused on the neurosurgical disease process. This presentation will discuss this specific case, appropriate screening in VHL, and correlates to other more common phakomatoses.
Tumors of the choroid plexus account for less than 1% of central nervous system tumors in the population as a whole but 12% of intracranial tumors in children less than two years of age. Surgical resection is the mainstay of treatment be it carcinomas or papilloma’s. The vascularity of these tumors makes such a resection a daunting undertaking, accomplished only 30% of the time at the initial surgery. Blood loss is the main limiting factor. Average blood replacement is total body all blood products. Operate mortality associated with bleeding has been reported to be as high as 12%. Therefore, devascularization of these tumors is mandatory. Endovascular techniques have tremendously evolved over the last 10 years. In children under two years of age, the inherent size of the cerebral vasculature and anatomy can make this is a technically difficult endeavor. The very volumes of contrast required to complete the study requires aggressive IV crystalloid to maintain a delicate balance between renal and pulmonary function.

Two patients with Choroid Plexus Papilloma’s and their tumor devascularization strategies are the topic of this presentation. Both patients required only a single blood transfusion (10cc/Kg PRBC) with gross total resection at definitive surgery. Standard endovascular approach was undertaken in the first. A novel endoscopic approach was undertaken in the second.

A 20-month-old male underwent CT imaging for routine of evaluation of blunt head trauma. A large intraventricular tumor was discovered and subsequently proved on follow up MRI imaging to be a Choroid Plexus Tumor. Choroidal arterial feeders were noted on the MRI. The patient underwent endovascular embolization of both the anterior and posterior choroidal arteries, however this required two separate settings. He underwent an uncomplicated occipital trans sulcal craniotomy and gross total resection of the tumor was accomplished. He required a single postop blood transfusion, sustained no neurological deficits, and was discharged home on the second postoperative day. A discussion of the vascular anatomy of choroid plexus tumors, embolization nuances, followed by surgical videos of resection will be presented.

The second patient was diagnosed on prenatal sonography to have an intraventricular tumor that was confirmed on MR imaging at birth to be a choroid plexus tumor. No intervention was undertaken at that time. The patient was referred at seven months of age. MR imaging revealed the child had developed ventriculomegally. His young age was concerning to consider endovascular intervention. On evaluation of the anatomy it became apparent that an endoscopic approach could be undertaken. He was taken to surgery where endoscopic microvascular clipping with a Yasirgil micro clip was accomplished and the remaining choroid plexus arterial feeders were endoscopically cauterized and transected with micro scissors. That technical approach was uncomplicated, however the endoscopic sheath simply touching the tumor would lead to bleeding that required irrigation to clear. The patient required a single blood transfusion based upon that. Two days later he was taken to surgery where an uncomplicated trans occipital sulcal approach removed a devascularized free-floating tumor in 60 minutes surgical time. The child was discharged on the second postoperative day and remains neurologically normal.
The contrast and management of these two patients suggest this novel endoscopic approach for devascularization should be considered if feasible in patients with these tumors and is associated with shorter preoperative management, less potential complications and ultimately easier definitive surgical resection.
ACQUIRED TEMPORAL ENCEPHALOCELE CAUSING MEMORY LOSS AND EPILEPSY IN A 17 YO FEMALE WITH GORHAM’S LYMPHANGIOMATOSIS: COMPLICATIONS IN MANAGEMENT

Sean Lew, MD

Gorham’s lymphangiomatosis, aka “disappearing bone disease”, is a rare condition characterized by lymphatic proliferation leading to resorption of bone. A 17 yo female presented with Gorham’s lymphangiomatosis and a left middle fossa encephalocele secondary to resorption of the middle fossa floor. Over time she developed both memory decline and epilepsy prompting an attempt at surgical correction. This case presentation will review the management and complications that ensued.
COMPLICATION: INTRACRANIAL HEMORRHAGE AFTER PLACEMENT OF A VENTRICULOPERITONEAL SHUNT IN A NEONATE WITH LARGE VENTRICLES

Corbett Wilkinson, MD

A baby girl with prenatally-diagnosed hydrocephalus was born at 38 5/7 weeks by planned Caesarian section to a 21-year-old mother with a history of smoking and methamphetamine use. I had met the family prenatally by videoconference and reviewed a fetal MRI and maternal ultrasounds. At birth, the patient had an extremely large head with an orbitofrontal circumference of 51.5 cm, splayed sutures, and a bulging fontanelle. Ultrasound (A) showed extremely large ventricles with an interhemispheric cyst (not shown).

At 2 days of life the patient underwent placement of a right frontal ventriculoperitoneal shunt with an Orbis-Sigma valve. She needed an extra-large shoulder roll due to the size of her head. The dura was opened with a #11 scalpel. Cerebrospinal fluid was sent for routine studies. She became hypothermic during surgery, and afterwards was kept in the operating room 15 extra minutes for warming.

Postoperatively, a CT scan (B) obtained for possible seizures showed numerous subdural and intraparenchymal hematomas. The patient developed disseminated intravascular coagulation, and repeat CT several days later (C) showed increased number and size of the intraparenchymal hematomas. She received multiple red blood cell and fractionated free plasma transfusions.

At 12 years of age, she remains shunted and is nonverbal, g-tube fed, cortically blind, and wheelchair-bound with quadriplegic spasticity and seizures.

MRI scan at 12 years of age (D) shows extreme cortical atrophy.

Lessons learned by the surgeon include the importance of losing as little CSF as possible when placing cerebrospinal fluid shunts in neonates with large ventricles and the importance of preventing intraoperative hypothermia.
SPONTANEOUS INTRACRANIAL HYPOTENSION MIMICKING SURGICAL LESIONS:
REPORT OF TWO CASES

Naina L. Gross, MD  Lacey M. Carter, MD

Intro: Spontaneous intracranial hypotension (SIH) is a rare occurrence, especially in the pediatric population. One case of Chiari malformation in a child with connective tissue disease and no cases of ventriculomegaly due to SIH have been discussed in the literature.

Methods: We present two patients who appeared to have surgical lesions needing intervention found to be due to SIH that resolved spontaneously.

Results: Four-year-old healthy female presented with headaches and vomiting that started after repeatedly riding on a child-sized rollercoaster. MRI showed a Chiari Type 1 malformation. Review of the MRI showed bilateral cerebellar hygromas and clumping of the cauda equina on spine MRI, which was suspicious for SIH. The patient’s symptoms resolved prior to clinic visit. Follow-up imaging revealed spontaneous improvement in tonsillar descent from 9mm to 4mm, resolution of cervicomedullary kink, and separation of the lumbar nerve roots.

Separately, a two-year-old healthy female presented with headaches and vomiting. Imaging was concerning for hydrocephalus with fourth ventricular outlet obstruction, but showed bilateral cerebellar hygromas along with clumping of the cauda equina. The patient’s symptoms resolved after a brief period of observation. Four months later, imaging revealed spontaneous resolution of the ventriculomegaly and hygromas as well as improvement of the clumping of the cauda equine. Neither patient received or required intervention, and they experienced no return of symptoms.

We hypothesize that these patients had spinal subarachnoid blebs which ruptured causing intracranial hypotension that resulted in their presentations.

Conclusion: We show two cases of SIH presenting as two different neurosurgical pathologies with spontaneous resolution. These cases are the first case of Chiari in a healthy child and the first case of ventriculomegaly due to SIH presented in the literature.

Authors have no disclosures.
SEVERE TONGUE SWELLING FOLLOWING POSTERIOR FOSSA SURGERY
CASE PRESENTATION AND REVIEW OF THE LITERATURE

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The patient was a 6-year-old boy, who presented with headaches and morning vomiting. An MRI showed a left superior anterior cerebellar brain tumor abutting the midbrain. He was taken to the operating room for a posterior fossa craniotomy for resection of the tumor with neuromonitoring. He was positioned prone with his head in a Mayfield headholder. A bite block was placed by anesthesia. The surgery was uneventful, and postoperatively he was extubated and taken to the PICU. After surgery, his tongue looked normal, and he was saying words with appropriate neurological exam.

The following day, he began to have significant swelling of the tongue, and his tongue began to protrude outside of his mouth. Postoperative MRI showed near-total resection of the tumor, but there was edema in the anterior tongue. He was seen by ENT and oral surgery, and he was treated with supportive care. He was given bite blocks to make sure he did not bite his tongue and to allow room for his swollen tongue. In addition, his tongue was wrapped in wet gauze (changed every few hours) to keep it moist. The tongue edema started to improve after a few days, and it completely resolved to normal after about 10-14 days, after which he was able to talk appropriately.

Severe macroglossia after posterior fossa surgery has been described in a few case reports in the literature. There are two main theories regarding the cause. One theory is that the tongue was bitten during or after surgery. A second theory is that the swelling is due to venous congestion in the tongue. These theories will be discussed.
HYDROCEPHALUS CLINICAL RESEARCH NETWORK REGISTRY REVIEW OF ETV AFTER SHUNT FAILURE

Curtis Rozzelle, MD, Anastasia Arynchyna, MPH, Jessica Alvey, MSc, Ron Reeder, PhD, Abhaya Kulkarni, MD, PhD, & John Kestle, MD for the Hydrocephalus Clinical Research Network

**Introduction:** Treatment of hydrocephalus with Endoscopic Third Ventriculostomy is well described in the neurosurgical literature, reporting a wide range of success and complication rates. Prior reports on safety and efficacy of ETV included previously shunted patients as a subgroup. Multi-center investigation of outcomes in this "post-shunt ETV" (PSETV) patient population would help surgeons provide more accurate pre-operative counseling about procedural risks and shunt freedom potential.

**Methods:** Prospectively collected data in the Hydrocephalus Clinical Research Network (HCRN) Registry database was reviewed for inclusion criteria. ETV procedures following shunt procedures from 2008-2016 (inclusive) comprised the final cohort. Descriptive statistics and univariable analyses were conducted using SAS software. Kaplan-Meier survival curves for ETV failure and shunt free survival were compared with a mathematical estimate of the cohort’s ETV Success Score (ETVSS).

**Results:** Initial registry review identified 205 PSETV procedures. After excluding patients >18 years and ETVs done with simultaneous shunt revision, the final PSETV cohort comprised 149 procedures. Average age at PSETV was 4.2 (1.3-10.2) years, 56% male, 73% Caucasian; etiologies were 27% IVH of prematurity, 20% aqueductal stenosis, 13% myelomeningocele, and 40% other. PSETV was performed concurrent with shunt infection treatment in 24% of cases and supplemented with an EVD in 59%. New neurological deficits occurred in 3.3%, diabetes insipidus - 0%, CSF leak - 9.8%, and wound infection - 2.0%.

Shunt-free survival of PSETV procedures was 40% by 6 months and stable thereafter, compared with 60% predicted by ETVSS. Univariable analysis of potential failure predictors showed no significant pre- or intra-operative factors except that severe bleeding predicted earlier failure, O.R. 4.82 (95% C.I.: 1.15, 20.12).

**Conclusions:** In this registry review of PSETV complication rates were slightly greater than in our registry analysis of first-time ETV, and failure rate was higher than predicted by ETVSS. Identifying factors that may account for this discrepancy will require further study.
AUTOMATICALLY MEASURING THE VENTRICULAR VOLUME FROM PICTURE ARCHIVING AND COMMUNICATIONS SYSTEM (PACS) CLINICAL IMAGES

F. Yepes-Calderon, MD Nelson and J. Gordon McComb

The picture archiving and communications system (PACS) is currently the standard platform to manage medical images but lacks analytical capabilities. Staying within PACS, the authors have developed an automatic method to retrieve the medical data and access it at a voxel level, decrypted and uncompressed that allows analytical capabilities while not perturbing the system's daily operation. Additionally, the strategy is secure and vendor independent.

Cerebral ventricular volume is important for the diagnosis and treatment of many neurological disorders. A significant change in ventricular volume is readily recognized, but subtle changes, especially over longer periods of time, may be difficult to discern. Clinical imaging protocols and parameters are often varied making it difficult to use a general solution with standard segmentation techniques. Presented is a segmentation strategy based on an algorithm that uses four features extracted from the medical images to create a statistical estimator capable of determining ventricular volume.

When compared with manual segmentations, the correlation was 94\% and holds promise for even better accuracy by incorporating the unlimited data available. The volume of any segmentable structure can be accurately determined utilizing the machine learning strategy presented and runs fully automatically within the PACS.
THE QUALITY OF YOUTUBE VIDEOS ON ETV AND ETV+CPC PROCEDURES AVAILABLE TO PEDIATRIC HYDROCEPHALUS FAMILIES

Sader N, Kulkarni A, Ahmed S, Eagles M, Riva-Cambrin J

Introduction: YouTube has become a vital information source for modern pediatric neurosurgical patients and families. However, no published data exists examining the relationship between the informative quality of these videos and established metrics of online popularity.

Methods: This cross-sectional study used comprehensive search terms to identify videos pertaining to endoscopic third ventriculostomy (ETV) and ETV+CPC (choroid plexus cauterization) on YouTube. Two pediatric neurosurgeons, one neurosurgery resident and two patient families independently reviewed and scored the selected videos. Videos were scored for informational quality using a validated 5-point Global Quality Scale (GQS) and compared to metrics of popularity such as views, likes, likes/views ratio, comments/views ratio and likes/dislikes ratio. Weighted kappa scores were used to measure agreement between reviewers.

Results: A total of 58 videos (47 ETV, 7 ETV+CPC, 4 both) met the inclusion criteria. In terms of GQS, substantial agreement was seen between surgeons (Kappa 0.67 [0.55, 0.80]) and excellent agreement was found between each surgeon and the neurosurgical resident 0.77 [0.66, 0.88], 0.89 [0.82, 0.97]. Only fair to moderate agreement was seen between professionals and patient families ranging from 0.27 to 0.55. There were significant associations between better GQS scores and more likes (p=0.01), views (p=0.02) and likes/dislikes ratio (p=0.016). Video styles included technical (62%), lecture (24%), patient testimonial (4%) and other (10%). Video style was associated with likes (p=0.02), views (p=0.03) and likes/dislikes ratio (p=0.015). Specifically, a lecture style was more likely rated good or excellent on GQS (86% vs. 0%, p <0.001).

Conclusion: Academic lecture type videos were more highly rated by both surgeons and patients and were associated with increased metrics of online popularity. Neurosurgeons seeking to increase their online footprint via YouTube would be well advised to focus more on the academic lecture format rather than technical videos.
RATES OF HINDBRAIN HERNIATION REVERSAL AFTER PRENATAL REPAIR OF MYELOMENINGOCELE

Edward S. Ahn, MD; Kendall Snyder, MD; David J. Daniels, MD, PhD

Purpose: To describe the rate and timing of reversal of hindbrain herniation after in utero repair of myelomeningocele at a single fetal surgery center.

Methods: This retrospective study was approved by our Institutional Review Board. We screened our surgical database for all repairs of myelomeningocele from 2010 until present. We evaluated the following features: presence of hindbrain herniation preoperatively, presence of hindbrain herniation postoperatively, gestational age at delivery, surgical management of hydrocephalus, and perioperative morbidity and mortality.

Results: Twelve consecutive patients (2 males, 10 females; gestational age at myelomeningocele repair ranging 23 weeks 3 days to 26 weeks 1 day) were identified. Of 9 patients with available pre- and post-op imaging, 7 (78%) of patients definitively exhibited Chiari II malformation with hindbrain herniation preoperatively. 6 (86%) of these patients showed resolution on MRI taken a mean of 3.3 months after delivery. Three patients (33%) required surgical treatment for hydrocephalus. Two patients had a third ventriculostomy with choroid plexus cauterization and one patient required a ventriculoperitoneal shunt. Early in the series, there were two cases of in utero fetal demise at the time of myelomeningocele repair and one patient died 4 days after delivery secondary to complications of a perforated bowel.

Conclusions: As previously demonstrated, prenatal closure of myelomeningocele results in early reversal of hindbrain herniation as demonstrated on perinatal imaging. A comparison between timing of reversal in prenatal versus postnatal closure is warranted.
PARENTAL REFUSAL TO CONSENT TO LIFE-SAVING THERAPY IN PEDIATRIC NEUROSURGERY-
CASE REVIEW AND FRAMEWORK FOR DECISION MAKING

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Introduction: Parental refusal of life-saving intervention is an uncommon, but ethically challenging scenario. We present the case of a prenatally diagnosed choroid plexus tumor where parents refused consent for their child to receive perioperative blood products for faith-based reasons, discuss our management of the case and review the range of how similar refusals are managed in pediatric neurosurgery centers in Canada.

Methods: We contacted the legal/risk management offices of the 12 pediatric health care centers with pediatric neurosurgery programs in Canada to request hospital policies regarding parental/care-giver refusal of potentially life-saving use of blood products.

Results: Canadian pediatric health care centers varied in their guidelines, policies and approaches to parental refusal of blood products. All centers indicated that their approach depended on the age and developmental status of the patient and the nature of the patient’s illness and proposed intervention. Approaches varied from no specific policy (cases dealt with on an ad hoc basis) to apprehension of the child for the purposes of authorizing transfusion, to the use of a “Letter of Understanding”, a document signed by parents acknowledging their understanding that their child will receive blood products if deemed to be life-saving and/or in the child’s best interests, while also acknowledging that they have not consented to receipt. In our case, parents signed our institutional Letter of Understanding and the infant was not apprehended by our Child Protection Service for the purposes of authorizing a blood transfusion.

Conclusion: Parental refusal of potentially life-saving blood products presents a rare but challenging ethical dilemma for pediatric neurosurgeons. Policies and approaches vary across Canadian pediatric health care centers. Our patient ultimately received a blood transfusion post-operatively but because parents had signed a Letter of Understanding prior to surgery, the child did not require apprehension for the purposes of authorizing the transfusion.

Disclosure: The authors have no relevant disclosures
DEVELOPMENT AND IMPLEMENTATION OF THE INTERSURGEON ONLINE MATCHING PLATFORM FOR GLOBAL NEUROSURGERY COLLABORATION

James M. Johnston, Matthew Davis, Michael Dewan, Brandon Rocque, William Harkness

Introduction: Over the past decade, there has been a revolution in handheld technology and social media, leading to an increasingly interconnected global community. Surgery has more recently emerged as a major global health priority, declared by the WHO, World Bank and Lancet Commission on Global Surgery. Encouragement and coordination of institutional collaborations will play a crucial role within the larger context of national surgical capacity building. Traditionally, global neurosurgical collaboration has been mediated by various non-governmental and neurosurgical organizations, with attendant inefficiencies of access and coordination. We describe the development and implementation of an online surgical matching application (InterSurgeon.org) to more effectively connect the global neurosurgical community.

Methods: After completion of a preliminary needs assessment, an application interface, database, and matching algorithm were developed in collaboration with Novagram (London, UK). Funding was secured from the International Society of Pediatric Neurosurgeons, the University of Alabama at Birmingham, and private donors. Beta testing was performed by an international group of pediatric neurosurgeons prior to formal launch of InterSurgeon.

Results: Since April 2018, InterSurgeon has enrolled more than 200 pediatric neurosurgical centers in 54 countries, with 39 training and research collaborations facilitated to date. The application was introduced at the WHO Emergency and Essential Surgery Programme during the World Health Assembly in May 2018, generating pledges for collaboration from global representatives in adult neurosurgery, pediatric surgery, and urology. The second phase of development is underway, including design and implementation of an adult neurosurgery module.

Conclusion: Rapid advances in handheld technology and social media are enabling a revolution in the reach and training capacity of global neurosurgery. InterSurgeon has demonstrated preliminary success in pediatric neurosurgery, and efforts continue to expand its membership and ensure a user-friendly, efficient and scalable Internet-based facilitator of the global surgery effort.
LONG-TERM OUTCOMES OF SPRING-ASSISTED SURGERY FOR SAGITTAL CRANIOSYNOSTOSIS

Daniel Couture

Background: Early intervention for sagittal craniosynostosis using spring-assisted surgery (SAS) has gained popularity due to decreased blood loss and transfusion requirement, and operative/recovery time, compared with traditional open approaches. Long-term stability of correction and complications have not been previously reported due to the more recent implementation of this technique.

Methods: To examine long-term outcomes of SAS for correction of sagittal craniosynostosis, we performed a retrospective examination of our consecutive series from 2000-2016.

Results: Of 174 patients receiving SAS, the mean age at spring-placement was 4.6 months, with surgery reserved for only those younger than 6 months of gestationally-corrected age. The mean age of follow-up at the time of the study was 5.0 years. Mean lengths of spring placement and removal operations were 45.8 and 26.7 minutes, respectively. No patients required blood transfusions for either operation. There were no serious complications, including CSF leak, cerebrovascular accidents, or deaths. Ten patients required unplanned surgery for complications (5.7%), including spring malposition (6, 3.4%), and infection (2, 1.1%). One patient experienced incomplete correction of frontal bossing, and another developed secondary coronal suture synostosis; both underwent secondary calvarial vault remodeling (CVR). Other long-term complications included a suture abscess (1, 0.3%), referral to neurology for headaches (2, 1.2%) or for seizures (1, 0.3%). Mean cephalic index (CI) prior to SAS was 71.0. This improved to 74.8 post-op (p<0.001). There were no significant differences between immediate post-op CI and those measured at post-operative years 1 (73.8), 6 (74.8) and 12 (73.9).

Conclusions: Correction of sagittal craniosynostosis using SAS is associated with an excellent complication and morbidity profile compared with open CVR. Maintenance of phenotype improvement with SAS is durable over time. SAS should be considered amongst the standard of care for treatment of sagittal craniosynostosis in infants less than 6 months of age.
PROPOSED MECHANISM OF CORPUS CALLOSUM TRAUMA IN SPORTS CONCUSSIONS

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The corpus callosum is involved in the pathophysiology of a concussion following mild traumatic brain injury. However, the mechanism by which the biomechanical forces and tissue deformations penetrate the deep structures like the corpus callosum and how these relate to a clinical concussion is unknown. Our laboratory has shown that coronal and horizontal but not sagittal rotation forces displace the falx cerebri, which in turn, strains the corpus callosum. We analyzed head accelerations of 115 sports impacts in collegiate football players measured with instrumented mouthguards to test the hypothesis that coronal and horizontal rotation produces motion in the falx cerebri, which in turn strains the corpus callosum. Coronal rotational accelerations distinguished impacts with diagnosed mTBI (8592 avg.) from those without (1412 rrrrr/s^2). Using 3D finite element simulations, coronal rotational acceleration strongly correlated with deep lateral motion of the falx center (r = 0.85) while horizontal rotational acceleration correlated with deep lateral motion of the falx periphery (r > 0.78).

The relationship between corpus callosum strain and the falx was unique: removing the falx from the finite element model halved peak strains in the corpus callosum from 35% to 17%. Among all brain structures, tract oriented strain x strain rate in the corpus callosum was the strongest predictor of a mild TBI providing evidence of clinical relevance. Furthermore, we also found evidence of corpus callosum trauma on diffusion tensor imaging (DTI) in the same injured players. Our results suggest that the corpus callosum is sensitive to coronal and horizontal rotations because they drive lateral motion of a relatively stiff membrane, the falx, in the direction of commissural fibers below. We named this proposed injury mechanism “corpus callosum concussion”.
HOW A PEDIATRIC NEUROSURGERY TELEMEDICINE CLINIC CREATES SOCIO-ECONOMIC BENEFITS FOR PARENTS/CAREGIVERS

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**Introduction:** We present the socio-economic benefits of a pediatric neurosurgery telemedicine clinic.

**Methods:** A pediatric neurosurgery telemedicine clinic was organized by the University of Florida College of Medicine Jacksonville/Wolfson Children's Hospital/Baptist Health of Northeast Florida in collaboration with Georgia Children’s Medical Services (CMS) to service the Southeast Georgia Health District (Area 9-2). Monthly telehealth sessions are held with the CMS nursing personnel at the remote location. Pertinent clinical history and findings are presented audiovisually to the pediatric neurosurgeon in the presence of the patient and parents. From the clinic database, the authors review the composition of the clinic and report on the factors affecting family visits. MapPoint 2013 (Microsoft Corporation) was used to calculate the distance, time and cost savings.

**Results:** This report addresses the clinic activities from August 2011 to January 2017. Fifty-five patients were seen in a total of 268 appointments (initial and follow-up). The average distance of travel for a family from home to the UF Pediatric Neurosurgery Center in Jacksonville versus the CMS remote location was 93 miles versus 28 miles. The families saved, on average, 2.4 hours of travel time and 132 miles per visit. Transportation cost savings for all visits was $194 average per family, and $10,853 for all families. The cost savings for lost work time for all visits was an average of $42 per family, and $2239 for all families; which led to a cost savings for all visits of an average per family of $236 and $13,232 for all families.

**Conclusions:** Managing pediatric neurosurgery patient/families via telemedicine is feasible and saves families substantial travel time, travel cost, and time away from work.

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INFUSION OF 5- AZACYTIDINE (5- AZA) INTO THE FOURTH VENTRICLE OR RESECTION CAVITY IN CHILDREN WITH RECURRENT POSTERIOR FOSSA EPENDYMOMA: A PILOT STUDY

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Background: DNA methylation inhibitors are logical therapeutic candidates for ependymomas originating in the posterior fossa of the brain. Our objective was to test the safety of infusing 5-Azacytidine (5-AZA), a DNA methylation inhibitor, directly into cerebrospinal fluid (CSF) spaces of the fourth ventricle or tumor resection cavity in children with recurrent ependymoma originating in the posterior fossa.

Materials and Methods: In patients with recurrent ependymoma whose disease originated in the posterior fossa, a maximal safe subtotal tumor resection was performed. At the conclusion of the tumor resection, a catheter was surgically placed into the fourth ventricle or tumor resection cavity and attached to a ventricular access device. Cerebrospinal fluid (CSF) flow from the posterior fossa to the sacrum was confirmed by CINE phase contrast magnetic resonance imaging (MRI) postoperatively. 12 consecutive weekly 10 milligram (mg) infusions of 5-Azacytidine (AZA) were planned. Disease response was monitored with MRI scans and CSF cytology.

Results: Six patients were enrolled. One patient was withdrawn prior to planned 5-AZA infusions due to surgical complications after tumor resection. The remaining 5 patients received 8, 12, 12, 12, and 12 infusions, respectively. There were no serious adverse events or new neurological deficits attributed to 5-AZA infusions. All 5 patients with ependymoma who received 5-AZA infusions had progressive disease. Two of the five patients, however, were noted to have decrease in the size of at least one intraventricular lesion.

Conclusions: 5-AZA can be infused into the fourth ventricle or posterior fossa tumor resection cavity without causing neurological toxicity. Future studies with higher doses and/or increased dosing frequency are warranted.
ITGA2 AS A THERAPEUTIC TARGET FOR Glioblastoma

Peng Guo, Alexander Moses-Gardner, Jing Huang, Marsha A. Moses and Edward R. Smith

To date, there is no effective targeted therapeutic to treat glioblastoma (GBM) in the clinic. The recent rapid development of nanomaterials has created a promising opportunity to engineer “virus-like” nanovehicles (termed nanomedicines) to circulate in the body and selectively deliver various therapeutic, diagnostic, and theranostic agents to the diseased sites (e.g., tumor and metastatic lesions) while sparing healthy organs and tissues.

We reasoned that first-generation nanomedicine development may have been limited by delivery of their payloads in a non-specific, non-targeted manner and that drug availability to the GBM may be severely hindered by blood-brain tumor barrier (BBTB) and tumor heterogeneity. To resolve these issues, we hypothesized that functionalizing non-specific nanomedicines with antibodies against GBM-specific antigens can guide them to selectively recognize and ablate GBM tumors in a more precise and efficient manner.

In this study we have identified and characterized the cell surface antigen ITGA2 as a novel molecular target for GBM that is robustly expressed in multiple representative GBM cell lines while being absent in normal glial cells. We found that ITGA2 is significantly upregulated in human GBM tumor tissues and high ITGA2 expression has a negative impact on GBM patient survival, suggesting ITGA2 as a promising therapeutic target for GBM. We have developed an ITGA2 antibody-directed, doxorubicin encapsulating liposome (ITGA2-Dox-LP) as a novel GBM-targeted nanomedicine that selectively binds to and kills GBM cells in vitro. We also demonstrated that these GBM-targeted ITGA2-Dox-LP could effectively breach an in vitro BBTB via GBM induced angiogenesis, but not a normal intact in vitro BBB. These findings may have significant clinical potential for GBM therapy and diagnosis and support further research into the use of ITGA2 as a therapeutic target for GBM.
ENDOSCOPIC ENDONASAL SURGERY FOR RESECTION OF PEDIATRIC CRANIOPHARYNGIOMA: COMPARISON TO OPEN RESECTION WITH A FOCUS ON CEREBROVASCULAR COMPLICATIONS

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Introduction: Craniopharyngiomas are benign tumors but attempts at gross total resection (GTR) can lead to serious complications, especially those related to cerebrovascular insult. Endoscopic endonasal surgery (EES) aimed at GTR has the potential to generate fewer general and cerebrovascular complications, but there has been limited comparison to open surgery. We performed a review of open surgical and EES resections of pediatric craniopharyngioma within our institution to elucidate any potential benefits and assess differences in complications.

Methods: A retrospective chart review was performed of pediatric patients undergoing resection of primary or recurrent craniopharyngioma at our institution either from craniotomy 2001-2010 and endonasal surgery 2010-2017. Volumetric analysis of imaging assessed tumor volumes and extent of postoperative ischemic injury between the 2 treatment options. Imaging review also determined the incidence of pseudoaneurysm formation.

Results: 43 patients were identified for analysis with an average age of 8.2 years. Open surgery was the initial intervention in 15 patients and EES in 28. EES was performed in patients aged 3 to 17 years. This revealed no difference in residual tumor volume ($p = 0.28$), but volume of postoperative ischemia was significantly larger in the open group ($p = 0.004$). GTR was more often achieved in the EES group. Pseudoaneurysms were only observed in the open surgical group. Panhypopituitarism developed almost uniformly across both groups. Postoperative weight-gain was significantly more common in the open surgical group, and body mass index correlated with volume of ischemic injury in regression analysis ($p = 0.05$).

Conclusions: EES resection was associated with similar, if not better, extent of resection and significantly less ischemic injury than open surgery. Pseudoaneurysm formation was only seen in the open surgical group. Weight-gain was also decreased in the EES cohort.
STEREOTACTIC BIOPSIES IN THE MANAGEMENT OF DIFFUSE INTRINSIC PONTINE GLIOMA

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Introduction: Biopsies had been previously avoided in the setting of DIPG, given high morbidity and lack of therapeutic options. In recent years, stereotactic biopsies have become more common, often done as part of clinical trials. We present our series of stereotactic biopsies of DIPG’s.

Methods: This was a retrospective review of patients at Children’s National Health System from 2015 to 2018 who underwent stereotactic biopsy when DIPG was suspected. All surgeries were done by 2 pediatric neurosurgeons. Charts were reviewed for demographic and clinical information and pathology.

Results: There were 16 patients who underwent stereotactic brainstem biopsies (10 male, 6 female). All surgeries were done via a trans-middle cerebellar peduncle approach, using frameless navigation. Mean age at surgery was 7.96 years (range 3.24 years to 25.95 years). 7 were done on the left side and 9 were done on the right. The most common presenting symptoms were gait instability (13/16), visual symptoms such as double vision (10/16), facial weakness (6/16), and extremity weakness (6/16). All biopsies except one were done after diagnosis and before radiation treatment (mean time from diagnosis to biopsy was 9.26 days). One biopsy was done after radiation treatment. Pathology showed all to be DIPG’s with the following WHO grades: Grade II: 1, Grade III: 1, Grade IV: 14. 14/16 had an H3-K27M mutation. There was one postoperative morbidity of increased facial weakness. There were no surgical mortalities.

Conclusions: Stereotactic biopsies of DIPG’s can be done safely and effectively. In our series, there was 1 (out of 16) morbidity, and no mortalities. Accuracy of frameless navigation registration can be a challenge for lesions in the posterior fossa, and we will discuss modifications to our technique to improve registration accuracy.
DOSIMETRY-BASED CONVECTION-ENHANCED DELIVERY (CED) FOR BRAIN TUMORS

Mark Souweidane, MD, FACS, FAAP

Introduction: Convection-enhanced delivery (CED) has recently been explored as a therapeutic modality for brain tumors by optimizing local drug concentrations while avoiding systemic exposure. Seemingly, drug distribution and dose should affect therapeutic response. Strategies for reliably assessing drug distribution and more importantly concentration have not been validated but are needed to better gauge CED as a therapeutic platform for brain tumors.

Methods: Children with diffuse intrinsic pontine glioma (DIPG) were treated on a Phase I clinical trial using CED of a theranostic agent (\(^{124}\)I-8H9). MRI was used for estimating tumor volume (TV). Volume of infusion (Vi) was defined by prescribed dose level. Volume of distribution (Vd) was measured using T2 signal change relative to base line (\(\Delta T2\)) and positron emission tomography (PET). Absorbed dose (whole body, brain, brain stem) was obtained using PET. These measurements were then used to calculate percent tumor coverage (Vd/TV x 100), and tentatively defined intralesional therapeutic index (Vd/TV × brain stem absorbed dose).

Results: Mean tumor volumes was (17.58 ± 10.89 cm\(^3\), DL 7: 21.35 ± 14.71 cm\(^3\)). Vi ranged from 250 to 4,000 µl. Vd measured by T2 and PET was volume-dependent. Vd measured by T2 ranged from DL 3 to DL 7: 1.84, 2.76, 5.99, 17.44, 15.81 cm\(^3\), respectively, and measured by PET ranged from DL 1 to DL 7: 1.72, 7.67, 2.30, 2.86, 6.07, 9.50, 6.69 cm\(^3\), respectively. Mean Vd/Vi ratio based on T2 was 3.40 (DL 3 to DL 7: 2.53, 2.74, 2.32, 5.00, 3.68, respectively). Percentage of tumor overlap ranged from 4.4% to 95.6% (DL3 average 31.1%, DL4 28.0%, DL7 59.3%). On DL 7, percentage of tumor overlap ranged from 9.2% to 95.6% (average 59.3%) (n=6). Intralesional absorbed dose ranged from 0.6 to 89.1 Gy (average from DL1 to DL7: 1.22, 4.09, 9.27, 12.17, 23.88, 60.35, 51.90 Gy, respectively).

Conclusions: Accurate and reproducible dosimetry data can be obtained when positron-emitting agents are delivered via CED followed by PET imaging. Early estimations provide confidence in predicting infusion volumes that are required to match intended tumor volumes. Tumor overlap is inconsistent and can benefit from some evolving surgical planning utilizing algorithms predicting infusedate distribution incorporating anatomical structures and infusion parameters. Intralesional drug dose can be monitored and should be integrated into clinical trials designed to test therapeutic response.
THE PRO-INFLAMMATORY EFFECT OF CYST FLUID IN PEDIATRIC ADAMANTINOMATOUS CRANIOPHARYNGIOMA MAY BE DRIVEN BY INTERLEUKIN-6

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Introduction: Pediatric Adamantinomatous Craniopharyngioma (ACP) is a clinically aggressive albeit, histologically benign tumor well known for its association with poor quality of life in young patients. Understanding the immunopathology of the disease is critical for developing targeted drug therapies, as none are currently available. Prior studies have shown that the ACP cyst fluid (CF) is immunologically enriched with cytokines, of which IL-6 is among the most prominent. We seek to understand the role of cyst fluid IL-6 in the pathology of ACP.

Methods: Cyst fluid samples were collected intraoperatively from pediatric ACP patients. Normal human astrocytes were treated in vitro with media containing ACP CF (at 1:10 dilution) in the presence or absence of an IL-6 neutralizing antibody, mimicking the activity of the FDA approved chimeric monoclonal antibody, Siltuximab. After 48 hours, drug-enriched media was replaced with fresh media which was subsequently collected after 24 hours and was assayed for IL-6 levels by ELISA.

Results: Astrocytes treated with ACP CF demonstrated slightly increased IL-6 secretion compared to astrocytes treated with control media (185.23 +/- 24.6 pg/mL vs 143.5 +/- 19.5 pg/mL; p = 0.06). Astrocytes treated with ACP CF in the presence of the IL-6 neutralizing antibody had decreased secretion of IL-6 compared to astrocytes treated with ACP cyst fluid alone (111.5 +/- 7.8 pg/mL vs 185.23 +/- 24.6 pg/mL; p=0.05).

Conclusion: This study attempts to elucidate the role of IL-6 in pediatric ACP. We show that under the influence of ACP CF, normal human astrocytes may be driven towards a pro-inflammatory state. This effect of the cyst fluid is mediated, likely among other things, by IL-6 signaling. With the availability of FDA-approved drugs targeting IL-6, further studies to advance this line of research and additional work to explore IL-6 as a potential therapeutic target for pediatric ACP are underway.
NOVEL MOUSE MODELING OF ATYPICAL TERATOID RHABDOID TUMOUR: PROMISE OF NEW THERAPEUTIC TARGETS

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Introduction: Atypical Teratoid Rhabdoid Tumour (ATRT) is a rare, devastating and largely incurable pediatric brain tumor diagnosed early in childhood. Use of intensive, high-dose chemotherapy and/or craniospinal irradiation has improved short term survival for some patients; however, long term survival rates remain dismal with 70-75% of children succumbing to their disease. Poor survival outcomes and treatment toxicity necessitate targeted and more efficacious therapeutics for children suffering from this disease. Genetic characterization of primary tumors has revealed that in addition to SNF5 loss, ATRTs comprise three molecular subtypes which differ at the level of global gene expression and DNA methylation and are found in distinct anatomical compartments.

Methods: We describe the generation of a genetically engineered mouse model that recapitulates the spatial pattern of human ATRT upon targeted loss of Snf5 to putative cells of origin using the Cre lox recombination strategy. We have also developed a robust drug screening strategy that employs a rigorous dose assessment methodology for high throughput analysis of a library of 477 kinase inhibitors against a panel of subtyped ATRT cell lines.

Results: Using this methodology, we identified Multiple Kinase Inhibitors which demonstrate therapeutic benefit in Group 2a/b ATRT. Interestingly, numerous kinase targets showed subtype specific expression. Among these, ERBB2 is a promising target which is exclusively expressed in Group 2a and Group 2b ATRTs and is exquisitely sensitive to dual EGFR/ERBB2 Inhibitors. More broadly, we found that ATRT cell lines are highly sensitive to PI3K and MAPK signaling blockade and that combinatorial inhibition of these pathways acts synergistically to reduce cell viability. Lastly, we combined our drug screening strategy with radiation treatment and found that dasatinib and MTOR inhibitors are capable of sensitizing Group 2a/b cells to radiation.

Conclusions: This work has advanced ATRT research on two important fronts: 1) the much needed development of better preclinical models, and; 2) the identification of novel, subtype-specific therapeutics.
THERMAL DYNAMICS, VOLUMETRICS AND SEIZURE OUTCOMES FOLLOWING MAGNETIC RESONANCE-GUIDED LASER INTERSTITIAL THERMAL THERAPY (MRgLITT) IN PEDIATRIC LESIONAL EPILEPSY

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Introduction: Magnetic resonance-guided laser interstitial thermal therapy (MRgLITT) has emerged as a safe and effective minimally invasive ablation technique for a variety of intracranial pathologies. Commercially available systems allow for pre-operative trajectory planning and real-time intra-operative monitoring based on thermal damage estimates. However, the effects of variable thermal energy on tissue ablation dynamics are poorly understood, particularly in the pediatric population.

Methods: A single-center retrospective review of patients undergoing MRgLITT for lesional epilepsy was performed. Volumes of ablation were measured independently by two raters in Osirix viewer, using post-ablation T1-weighted MR images. Inter-rater variability was analyzed via one-way ANOVA. Seizure outcomes were assessed using the Engel scale. Linear Regression analysis was performed to determine the relationship of energy to volume ablated. Fisher’s exact test was used to assess seizure outcomes in relation to pathologic substrate.

Results: Thirteen patients with an average age of 11.6 years had twenty total lesions ablated. Two patients had tuberous sclerosis, two had focal cortical dysplasia, four had hypothalamic hamartomas, three had periventricular nodular hyperplasia and one had mesial temporal sclerosis. Outcomes were documented to a mean follow-up of 10 months. Postoperative seizure control outcome was Engel I in nine patients (69.2%), Engel II in two patients (15.4%) and Engel III in two patients (15.4%). A linear regression was calculated to predict the effect of energy on volume of ablation ($p=6.47E-6$, $R=.841$). Volume ablated per joule of energy was not significantly different between pathologic substrates by ANOVA ($p=.47$). Linear correlation between energy and volume was found to be strongest in tuberous sclerosis ($R=.88$) and weakest in cortical dysplasia ($R=.057$). Inter-rater differences were statistically insignificant ($p=.73$). There was no significant relationship between pathological substrate and seizure control by Fisher’s Exact Test ($p=.67$).

Conclusions: We were able to characterize a linear relationship between the in vivo application of thermal energy and the volume of pediatric brain ablated. The strength of this linear correlation was variable across different pathologic substrates, but pathologic substrate did not predict seizure outcome. This study highlights the need for further, preferably prospective studies using larger sample sizes to better characterize these relationships.
INTRACRANIAL EEG AND INSULAR LASER ABLATION IN NON-LESIONAL EPILEPSY – CASE SERIES

Jim Baumgartner, MD

Introduction: Insular epilepsy has long been recognized but poorly characterized. With the introduction of stereo EEG monitoring, the electrographic patterns of insular epilepsy have been better described. We have explored the use laser ablation as a strategy for the management of sEEG localized insular seizures. We present our sEEG methodology, seizure semiology and laser surgical experience with a series of patients with non-lesional insular epilepsy.

Methods: We report a series of (n=8) patients with insular epilepsy who underwent stereo EEG evaluation followed by laser ablation. The patients ages ranged from 7-19 years at the time of laser ablation and included 3 males and 5 female patients. All patients underwent initial video EEG evaluation in our epilepsy monitoring unit where typical clinical seizures were captured. In addition, they underwent MRI, SPECT, PET, MEG and Neuropsychological evaluation. The cases were discussed at epilepsy case conference and a treatment plan developed by consensus. Patients underwent intracranial monitoring with stereo EEG with or without subdural electrode arrays. The epileptogenic zone was within insular regions based on intracranial EEG monitoring.

Results: At least two depth electrodes with 5-10 contacts were placed into each insular region. Insular laser ablation was planned based on the electrode contacts showing epileptic activity. 6 patients underwent left Insular laser ablation and two had right insular ablation. Two cases required repeat insular laser ablation following continued seizure activity after the initial ablation. One patient experienced an intracerebral hemorrhage during laser ablation, requiring additional surgical treatment. Six patients had ILAE class I outcome. One patient had class 3 outcome and one class 4 outcome.

Conclusions: Our experience suggest that intracranial monitoring can be used as an effective strategy for localizing non-lesional epileptogenic zones in case of insular epilepsy. Also, LASER ablation can be an effective surgical management tool for treating these patients. The details of the intra-LITT hemorrhage and its management will be discussed.
THE ROLE OF MINIMALLY INVASIVE SURGERY FOR CHILDREN WITH TUBEROUS SCLEROSIS COMPLEX (TSC)

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Introduction: Significant advances have been made in the treatment of children with medically refractory epilepsy, in general, and for patients with TSC, specifically, due to philosophic and technologic progress in pediatric epilepsy surgery. Increasing experience at many centers has revealed that children with TSC may benefit from surgery despite not being seizure free. Given the natural history of TSC, and the evolving concept of palliative surgery, we developed a minimally invasive option and report our initial experience with this approach.

Methods: Over 29 months, we have treated 34 children with TSC and medically refractory epilepsy. All patients underwent a Phase I evaluation, consisting of VEEG, MRI, CT, MEG, PET, and group discussion at conference, and were referred for either resective surgery with subdural electrodes (SDE) or a minimally invasive approach, consisting of stereo EEG (SEEG) and stereotactic laser ablation (SLA). Outcome was considered improved if at least a 50% reduction of the targeted seizure type was achieved.

Results: 23 children underwent an initial minimally invasive approach (22 SEEG and 1 SLA only) and 11 had resective surgery (10 with SDE and 1 resection only). 19 SEEG patients had SLA, and 3 had resection. At early follow up, 13 of the 19 SLA and 6 of 10 resection patients were improved. 3 SEEG patients had resection and all improved, as did the two who had either SLA or resection only. Complications included one abscess and one hemorrhage seen on MRI, not requiring treatment. Several children had prior surgery, with improvement, but underwent additional interventions in an iterative fashion.

Conclusion: Children with TSC can improve after surgery, despite not being completely seizure free by definition. A new, less invasive neurosurgery approach is possible for select children with TSC and medically refractory epilepsy. TSC has facilitated a new iterative epilepsy surgery paradigm.
COMPARATIVE STUDY OF OPEN SURGICAL CALLOSOTOMY AND LASER INTERSTITIAL THERMAL THERAPY (LITT) IN PEDIATRICS

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Background: Corpus callosotomy has been efficacious for patients with medically refractory epilepsy, especially those with drop attacks (atonic or tonic) and secondarily generalizable seizures. Given the morbidity of the open procedure, there has been an increasing trend to seek alternative treatment options. MR-guided Laser Interstitial Thermal Therapy is a minimally invasive surgical option for ablation of epileptogenic foci and it is increasingly being used for disconnection procedures. To date, there is no comparative study; therefore authors report their results from open corpus callosotomy (OCC) and Laser Interstitial Thermal Therapy (LITT).

Methods: All patients operated with either, and in some cases both, open surgical callosotomy and LITT from January 2005 to June 2018 were reviewed at a single center. Patient demographics, presurgical seizure burden, operative variables, postoperative outcome, complications, revision operation, need for steroid/duration of steroid, length of hospital stay (LOS), disposition to rehab facility or home, and overall recovery from seizure burden were recorded.

Results: 20 patients male to female ratio of 4:1 with mean age of 10 years {OCC versus LITT, (9.2 vs. 11.6)} were operated for medically refractory epilepsy. 6 (30%) patients underwent LITT procedure. When compared with OCC cohort, statistically significant decrease in EBL (p<0.003), and decreasing trend towards LOS (p<0.149) was observed in LITT cohort {mean±SEM, (50.71±8.40 vs. 7±3.41), and (6.07±0.44 vs. 4.50±1.18)}.

Reported follow-up (in months) for OCC cohort was 87.64, and for LITT was 6.91. In OCC cohort 9 (64%) patients were transferred to inpatient rehab facility while, all LITT patients were discharged home postoperatively. Interestingly, 10 (71%) patients in OCC, and 5 (83%) in LITT cohort documented recovery from seizure burden.

Conclusions: The concept of treatment of various epileptogenic foci in children using LITT has been relatively recent. And it is considered a safe and effective therapeutic option. Corpus callosotomy using LITT is a novel alternative procedure to conventional open surgical approach with significant decrease in EBL, and LOS. When compared to OCC, LITT offers quicker recovery, almost complete cure from drop seizures, and final disposition to home.
PROGRESSIVE EXPERIENCE DIFFERENTIALLY AFFECTS IMRI USE FOR RESECTION IN PEDIATRIC EPILEPSY AND TUMOR PROCEDURES

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Introduction: Intra-operative magnetic resonance imaging (iMRI) reveals the extent of even subtle parenchymal brain lesions, including gliomas and malformations of cortical development in children. Little is known about the adoption curve for iMRI technology in pediatric neurosurgery and its differential impact on surgery for tumor and medically refractory epilepsy.

Methods: With IRB approval, prospective data for the author’s first 60 pediatric 3.0 Tesla iMRI procedures from March 2016 to July 2018 were analyzed, including surgical indications, principal findings, number of scans, and the occurrence of additional resection. Other data were collected retrospectively. Comparison was made between the first and second epochs within the cohort and between procedures for tumor and medically refractory epilepsy.

Results: iMRI was used in 29 epilepsy and 28 tumor resections plus single AVM and cavernous malformation resections and a craniocervical decompression for Chiari I malformation. Maximal safe lesion resection was achieved in all cases. We obtained an average 1.34 scans per epilepsy case (range 1-3) and 1.46 scans per tumor case (range 1-2; p = 0.062). We obtained an average of 1.53 scans per early epoch case and 1.23 scans per late epoch case (p=0.042). iMRI data were more likely to alter resection in early than late epoch cases (54% versus 31%) and in tumor than epilepsy cases (67% versus 38%), although a logistic regression model utilizing these factors was non-significant. In the remaining three vascular and Chiari procedures, a single scan did not prompt changes in surgical management.

Conclusions: In this single surgeon series, there was a propensity to obtain more MR scans during early than late cohort cases and during tumor than epilepsy cases. iMRI was somewhat more likely to lead to additional resection early in the process of technology adoption and during tumor rather than epilepsy procedures.

Dr. Selden has no relevant conflicts of interest for the conduct or reporting of this work.
GENERATING IN VIVO SOMATIC MOUSE MOSAICS WITH LOCUS-SPECIFIC, STABLY-INTEGRATED TRANSGENIC ELEMENTS FOR STUDYING PEDIATRIC BRAIN TUMORS

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In situ transgenesis methods such as virus and electroporation can create in vivo somatic transgenic mice quickly, but they lack the exquisite control over copy number, zygosity, and locus specificity. Here, we establish mosaic analysis by dual recombinase-mediated cassette exchange (MADR), which permits stable labeling of mutant cells expressing transgenic elements from precisely-defined chromosomal loci. We provide a toolkit of MADR elements for combinatorial labeling, inducible/reversible transgene manipulation, VCre recombinase expression, and genetic manipulation of human cells. Further, we demonstrate the versatility of MADR by creating novel glioma models with mixed, reporter-identified zygosity or with “personalized” driver mutations from pediatric glioma or ependymoma. For example, introducing H3f3a mutation variants with MADR regulates the spatiotemporal profile of pediatric glioma, and single-cell RNA sequencing analysis demonstrates a recapitulation of developmental hierarchy seen in K27M-mutant human glioma. MADR is extensible to thousands of existing mouse lines, providing a high-throughput, flexible platform for mechanistic discovery.
TUMORS OF THE SUPERIOR MEDULLARY VELUM IN CHILDHOOD

Tadanori Tomita, MD

The superior medullary velum (SMV) consists of a thin lamina of white matter between the superior cerebellar peduncles horizontally and between the tectum and the cerebellum vertically. It forms an anterior roof of the fourth ventricle. The average thickness and length of the SMV is 0.3mm and 4.25mm respectively. In spite of its small size, it becomes an origin of certain tumors. The tumors arising in the SMV extend to the quadrigeminal cistern dorsally and into the fourth ventricle ventrally. Thus the tectum is displaced anterosuperiorly, and the superior vermis and the fastigium are displaced posteriorly. The pineal gland is not affected.

The author had 9 cases of SMV tumors; 8 were ATRT which all occurred during infancy and another one, JPA in a 5-year old boy. All patients presented with symptoms and signs of raised intracranial pressure, due to obstructive hydrocephalus. Tumors were removed effectively through the occipital transtentorial approach. This approach allows access to the fourth ventricle through the sectioned tentorial opening and the quadrigeminal cistern without sectioning neural structures. At this presentation, neuroimaging characters and surgical techniques are demonstrated.

Disclosures: The author declares no conflict of interest concerning the materials or methods used in this study or findings specified in this presentation.
DESMOPLASTIC INFANTILE GANGLIOGLIOMA: THE GREAT NEUROSURGICAL MASQUERADER

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Intro: Desmoplastic infantile ganglioglioma (DIG) is a rare, distinctive, supratentorial embryonal tumor with a generally favorable prognosis, first described by VandenBerg in 1987. Clinical, radiographic and pathologic features of this unusual neoplasm can mimic those of a malignant tumor and other serious intracranial disorders. Here the author describes 3 cases of DIG, each of which initially masqueraded as a different neurological disease with a very different prognosis.

Methods: Retrospective analysis of 3 personal cases.

Results: The author has cared for 3 patients with a DIG, each of whom was initially diagnosed with a vastly different neurological disorder. Case 1 was an 8-month-old boy referred for evaluation of a left posterior temporal hemorrhagic infarction at birth. Case 2 was a 2-month-old girl referred for a second opinion after pre- and postnatal MR imaging studies were interpreted as showing a large left holohemispheric malignant neoplasm. Case 3 was a 9-month-old girl referred with a large enhancing right frontoparietal parasagittal extra-axial tumor with massive edema felt to be a sarcoma. In each case, a gross total resection of the lesion was achieved, and each lesion proved to be a DIG, WHO Grade 1. Each child has had a benign postoperative course with a follow-up duration of 15 years for Case 1, 4 years for Case 2 and 5 months for Case 3.

Discussion: DIG is a great masquerader, and may be mistaken for other neurosurgical conditions including malignant neoplasms, stroke and infection. It is essential to consider this rare, low grade resectable tumor in the differential diagnosis of atypical intracranial masses of infancy, as the impact on prognosis can be profound. The author discusses management strategies for DIG, including the impact of molecular markers in selected cases.
RECENT TRENDS IN NORTH AMERICAN PEDIATRIC NEUROSURGICAL FELLOWSHIP TRAINING

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Objective: The goal of this study was to evaluate trends in the pediatric neurosurgical workforce in the United States.

Methods: From a database maintained by the Accreditation Council for Pediatric Neurosurgery Fellowships (ACPNF), all graduates of ACPNF-accredited pediatric neurosurgery fellowships were identified and an internet search was conducted to determine gender, undergraduate and graduate degrees, location and dates of residency and fellowship training, current practice/employment environment, American Board of Neurological Surgery (ABNS) or Fellowship of the Royal College of Surgeons certification status, American Board of Pediatric Neurological Surgery (ABPNS) certification status, and extent of current pediatric-focused practice. The graduates were further studied to determine whether they had completed a neurosurgical residency at a program with an affiliated, ACPNF-accredited pediatric neurosurgery fellowship program, and their residency training programs were further classified by whether the program ranked in the top 50 by NIH funding awards. The fellowship graduates’ current practice was also ranked in a similar fashion.

Results: There were 391 graduates of ACPNF-accredited pediatric neurosurgery fellowship programs from 1993 to 2018. The number of graduates per year has grown steadily over time, as has the percentage of women, now over 40% compared with no women in the first 3 years of fellowship accreditation in the mid-1990s. Approximately 71% of graduating fellows have a pediatric-focused practice but only 63% went on to obtain ABPNS certification. Of all graduates practicing in the United States, 68% practice in academic settings. Ninety-five percent of graduating fellows who were ABNS-board eligible were ABNS-certified.

Conclusions: A study of the graduates of accredited pediatric neurosurgical fellowships from 1993 to 2018 has revealed a growth in the number of graduates from ACPNF-accredited fellowship programs over time. A substantial portion of graduates will practice at least some adult neurosurgery and not go on to obtain ABPNS board certification.
FAST MRI IS FEASIBLE AND ACCURATE FOR TBI IN YOUNG CHILDREN

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Background: Due to concern about the risks of ionizing radiation, efforts have been made to supplant CT with fast MRI in various clinical settings. Several centers have proposed fast MRI as an alternative to CT or conventional MRI when evaluating for abusive head trauma where missing a relatively asymptomatic injury could be devastating. Small studies suggest that gradient echo (GRE) sequences are the most sensitive for trauma, but they have also found limited sensitivity of fast MRI. No study has prospectively compared the feasibility and accuracy of fast MRI in acute TBI.

Objective: To assess the feasibility of fast MRI and to compare its accuracy to that of CT in children with acute TBI.

Methods: Inclusion criteria included age <6 years and having had a head CT for TBI. All participants had fast MRI performed within 24 hours of CT. MRI sequences included fast version of T2, T1, FLAIR, GRE, and diffusion weighted images. All scans were interpreted independently by 2 pediatric neuroradiologists and results of the two imaging types were compared.

Results: Fast MRI was completed in 223 of the 225 subjects in whom it was attempted. Median imaging time was 6 minutes, compared to 58 seconds for CT. Radiographic evidence of TBI was present in 50% of CTs and unclear in 3%. Using CT as the criterion standard, fast MR had a sensitivity of 92.8% and specificity of 96.2%. Six isolated skull fractures and two isolated subarachnoid hemorrhages were identified on CT but missed on MRI. In 5 cases, small intra-cranial hemorrhages or contusions were identified by fast MRI but not by CT. These were felt to be real findings.

Conclusion: Fast MRI with GRE is a reasonable alternative to CT in stable children with acute TBI.
Guidelines for the diagnosis of brain death in children were first published in 1987 and last revised in 2011 by Nakagawa et al. The working group for the last revision included one pediatric neurosurgeon, however the guidelines were not endorsed by the AANS or CNS because of conflicting viewpoints of guidelines methodology. Recent national news stories such as the Jahai McMath case in Oakland, CA, prompted the initiation of a new working group by the American Academy of Neurology to create an updated, unified brain death guideline applicable to both children and adults. I will present a brief history of the development of pediatric brain death guidelines, a brief update of current controversies, and a brief account of my current experience as a member of a working group to revise these guidelines into a unified adult and pediatric version. During the discussion, I hope to receive feedback that I can bring back to the brain death guidelines working group.
CHRONIC INTRATHECAL ACCESS IN PATIENTS WITH SPINAL MUSCULAR ATROPHY AND COMPLEX SPINES

Michael G Muhonen, MD; Sam Rosenfeld, MD

Background: Spinal Muscular Atrophy (SMA) is a rare autosomal recessive progressive neuromuscular disorder. Patients with SMA often present with complex spinal pathology, including extensive spinal fusions, that complicate access to the intrathecal space. Nusinersin is an antisense oligonucleotide approved for the treatment of SMA requiring periodic intrathecal bolus injections for duration of life. To overcome major spinal access challenges in patients requiring repeated spinal injections, a novel infusion system, with a cervical subarachnoid catheter connected to suboccipital access port was implanted.

Methods: Retrospective review was undertaken to evaluate general device and procedure safety and tolerability in 7 SMA patients (age 11-48, 6 SMA Type II, 1 SMA Type III); that underwent a cervical laminotomy with implantation of a Medtronic intrathecal catheter connected to an Angiodynamics vortex flow smart port for periodic intrathecal injection of Nusinersin. 28 injections were administered in the 7 patients.

Results: The implant procedure was tolerated well. All 7 patients were stable in postoperative setting despite cases of severe pulmonary impairment commonly observed in SMA. CSF leak was witnessed intraoperatively around the catheter in three, but none developed a post-op CSF leak. One patient developed a perioperative CSF collection around a flipped port, necessitating re-operation to anchor the port. The port/catheter patency was evaluated by ease of CSF aspiration from port through the 2 month loading injections, followed by chronic injections every 4 months. 24 injections were done with the patients sitting in their wheelchairs. None of the ports or catheters have failed. All patients reported stabilization of motor function with some reporting gains during first 16 months of the study.

Conclusion: Preliminary observations reveal this cervical port/catheter procedure and device to be relatively safe and well-tolerated. This port/catheter construct vastly simplifies the Nusinersin intrathecal injection procedure in patients with complex spinal anatomy.

Disclosures: Dr Muhonen is a member of the Medtronic medical advisory board. Drs. Muhonen and Rosenfeld have attended paid advisory board meetings for Biogen.
INVESTIGATING THE NECESSITY FOR SPINAL CORD UNTETHERING IN PATIENTS WITH SPINAL DYSRAPHISMS PRIOR TO SCOLIOSIS CORRECTION

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Purpose and Background: Tethered spinal cord (TC) due to spinal dysraphism (SD) including myelomeningocele (MMC), split cord malformations (SCM), lipomyelomeningocele (LMMC) and re-tethering after the initial treatment of these pathologies may progress to neurologic, musculoskeletal, and other systemic deterioration. Even though patients may be clinically asymptomatic otherwise, tethered cord is thought to be one of many factors causing particularly scoliosis in these patients.

Initial treatment is releasing the cord which offers the opportunity to stabilize or improve the scoliosis. Scoliosis related to re-tethering of the spinal cord associated with repair of SD, specifically LMCC and MMC have poorer outcomes. Nonetheless, tethered cord release (TCR) is common in this population prior to spinal deformity correction with the goal of either halting the progression of scoliosis or mostly preventing any post-operative neurological decline related to manipulation of tethered spinal cord. No standard recommendations were established for performing TCR prior to deformity correction. The neurological outcomes have not been reported in a large patient sample. Our primary purpose is to assess the need for TCR in these patients.

Material and Methods: We retrospectively reviewed patients treated at Arkansas Children’s Hospital between 2006 and 2017 who underwent surgery for spinal deformity and have follow-up for at least one year. Data including demographics, preoperative diagnosis, surgical indications, neurological findings, surgical timing between TCR and scoliosis, operations, complications and outcomes were collected. Surgical and neurological outcomes were analyzed.

Results: Forty patients were identified. All of these patients had MMC. Average age at spinal deformity surgery was 10.66 years, and 7.81 years at TCR. In twenty patients, TCR was performed before spinal deformity surgery, and in three TCR was performed after. Overall, 69.23% (27/39) patients had complications, 14.81% (4/27) being shunt related. TCR before
spinal corrective surgery was not significantly correlated to outcomes or to occurrence of postoperative neurologic complications.

**Conclusions:** Tethered cord release prior to spinal deformity correction surgery did not significantly improve outcomes in our study group. Therefore, it may not be indicated in all patients prior to their spinal deformity corrective surgery in the absence of related symptoms. However, given the limited data and patient population we expanded our study as an international multicenter project. A multicenter study to identify a larger number of patients would provide a stronger scientific basis for making a management decision.

**Keywords:** spinal dysraphism, tethered cord, surgery, scoliosis

Conflict of interest statement: Authors do not have conflict of interest for this study
MINIMALLY INVASIVE TRANSFORAMINAL LUMBAR INTERBODY FUSION (MI-TLIF) IN CHILDREN

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Introduction: Minimally invasive transforaminal lumbar interbody fusion (MI-TLIF) has been found to have significant advantages as compared to traditional open spinal fusions in the adult population, but this technique has not previously been reported in children.

Methods: Medical records and radiologic studies were reviewed and reported.

Results: Two consecutive patients with lumbar spondylolistheses and failure of conservative management were identified. The first patient was a 13-year-old male with a Wiltse classification IIa, Meyerding grade II, mobile L4-5 spondylolisthesis. The second patient was a 17-year-old female with a Wiltse classification IIa, Meyerding grade II, mobile L5-S1 spondylolisthesis. Both patients had persistent severe lower back pain despite physical therapy, medical pain management, activity medication, and external bracing. The patients underwent MI-TLIF with interbody cage placement and bilateral percutaneous pedicle screw instrumentation at the level of spondylolisthesis. Operative time, blood loss, and length of stay were decreased compared to review of institutional historical controls. Postoperative imaging demonstrated appropriate reduction of the spondylolistheses and maintenance of baseline lumbar lordosis. Both patients had immediate postoperative resolution of their previously noted axial back pain and there were no perioperative complications. At one-year follow-up, bony fusion was confirmed radiographically, and both patients continued to be asymptomatic.

Conclusion: Early results following MI-TLIF in children are encouraging. Given the demonstrated relative benefits in the adult population, this technique should be considered as an option in pediatric patients with mobile spondylolisthesis and failure of conservative management.

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INFRATENTORIAL INTRACRANIAL PRESSURE IN PATIENTS WITH CHIARI I MALFORMATION

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Objective: To assess the infratentorial compartment intracranial pressure with patients with Chiari I malformation undergoing decompressive craniectomy procedures

Methods: 71 patients (26 males, 45 females) presenting with Chiari I malformation were treated with suboccipital craniectomy and duraplasty. The intracranial pressure was monitored using a Codman ICP sensor prior to removing the bone flap, prior to dural opening and after dural opening. ICP measurements were taken before and after Valsalva maneuvers and in three different positions: flat, Trendelenburg, and reverse Trendelenburg. The patient’s pCO2 was monitored and maintained in the same narrow range for all patients.

Results: The average pre-craniotomy ICP (mmHg) in neutral was 24 (5-49), Valsalva 29 (11-61). The average post-crani/pre-dural opening ICP was 22 (12-41), Valsalva 28 (4-47). The average post craniotomy/post dural opening ICP was 2 (1-8) and Valsalva 3 (2-11). The mean percent decrease in ICP from pre-crani to post-crani was 18% and between pre-crani and post-dural opening was 89%. The decreased was more pronounced in patients undergoing Valsalva maneuvers with pre-crani to post-dural decrease of 91%.

Conclusion: Patients with Chiari I malformation were found to have significant elevation in posterior fossa ICP prior to and after craniectomy. Dural opening significantly decreased the patient’s ICP from baseline. These results indicate that craniectomy alone as the primary mode of treatment may not be sufficient to treat the patient’s symptoms and elevated intracranial pressures.
AXIAL MR ANGIOGRAPHY IN EVALUATING REVASCULARIZATION AFTER INDIRECT BYPASS SURGERY FOR MOYAMOYA

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Introduction: Moyamoya disease and syndrome are often treated with encephaloduroarteriosynangiosis (EDAS). Traditionally, post-EDAS revascularization is evaluated using catheter angiograms. However, angiograms involve radiation exposure and a risk of stroke. Our center has transitioned from routine postoperative catheter angiograms to routine magnetic resonance angiograms (MRAs). Revascularization appears on MRA as new cerebral surface collaterals at the surgical site. It may also appear as increased signal intensity and diameter of the graft vessel versus the corresponding contralateral vessel (Figure 1). In this study, we evaluate these relative increases as evidence of revascularization.

Methods: We reviewed all subjects who underwent EDAS from 2005 through 2017. For each graft vessel and corresponding contralateral vessel we a) evaluated the change in relative signal intensity (0=no change, 1=increase, 2=exceptional increase) and b) calculated the change in ratio of their diameters between a preoperative and a 9-to-24-month-postoperative axial MRA. We also correlated these changes with the extent of new surgical site surface collaterals (none, fair, robust). We are currently correlating these changes with the angiographic degree of revascularization in subjects who also had postoperative catheter angiograms.

Results: Sixty-five moyamoya cases were screened in 45 subjects. 32 cases were included. Reasons for exclusion included bilateral surgery, non-EDAS surgery, and ineligible timing of postop MRA. Of 29 subjects in whom the change in relative signal intensity has been assessed, 13 (45%) were graded 0 and 13 were 1; 3 (10%) were 2. The average preoperative-postoperative change in diameter ratio was (+) 0.26. There was a trend for extent of this change to vary with the extent of new surface collaterals.

Conclusions: Revascularization of the brain after EDAS can be qualitatively and quantitatively evaluated on axial MRA. Increased signal intensity and diameter of the donor vessel versus the corresponding contralateral vessel on the postoperative versus the preoperative MRA strongly suggests successful revascularization.
Figure 1. Axial MRA scans. A. Pre-EDAS  B. 12 months post-EDAS. Large arrows point to the vessel used for EDAS, the parietal branch of the superficial temporal artery (STA). Small arrows point to the contralateral STA parietal branch.