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Intra-uterine closure of myelomeningocele defects with primary linear repair versusbipedicle fasciocutaneous flaps: A post-MOMS cohort study with long term follow up

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Background: The objective of this study was to determine the effects of in-utero bipedicle flaps on maternal-fetal morbidity/mortality, the need for cerebrospinal fluid diversion, and long-term functional outcomes.
Methods: 86 patients from a single institution who underwent fetal myelomeningocele repair from 2011-2021 were prospectively enrolled and retrospectively reviewed. 22 patients underwent bipedicle flap closure and 64 underwent primary closure. Questionnaires were derived for parents with 18 questions patient outcomes. Primary outcomes included intra-uterine fetal demise, postnatal death, postnatal myelomeningocele repair dehiscence, and cerebrospinal fluid diversion. Secondary outcomes included ambulation status and bowel and bladder function.
Results: The cohort was not statistically different with regards to race, ethnicity, maternal age at fetal surgery, body mass index, gravidity, parity, gestational age, estimated weight, or lesion level. There were no significant differences in rates of intra-uterine fetal demise, postnatal mortality, midline repair site dehiscence or the need for cerebrospinal fluid diversion by final follow-up. Gestational age at delivery was lower in the bipedicle flap cohort (232 vs. 241 days, $p<0.01$). Long-term functional outcomes and ambulation skill level was not different between cohorts. Similar rates of gastrointestinal surgery/enema dependence, genitourinary surgery/CIC dependence were also seen.
Conclusions: Analysis of the total cohort affirms the long-term benefits of fetal myelomeningocele repair. In-utero bipedicle flaps are safe and can be used for high-tension lesions without increasing perioperative risks to the mother or fetus. In-utero flaps preserve the long-term benefits seen with primary linear repair, and may expand inclusion criteria for fetal repair, providing life-changing care for more patients.

PELVIS syndrome: A rare form of spina bifida occulta

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Background: Introduction: PELVIS syndrome is a spectrum of disorders consisting of perineal hemangiomas, external genitalia anomalies, lipomyelomeningocele, vesicorenal abnormalities, imperforate anus, and skin tags. Within the literature, there is no established diagnostic criteria with minimal long term outcomes reported for this rare syndrome.
Objective: To identify the most common presenting signs/symptoms, describe medical and/or surgical treatments, and present long-term outcomes in children with PELVIS syndrome.
Methods: Retrospective chart review on all patients diagnosed with PELVIS syndrome at Ann and Robert H. Lurie Children's Hospital over the last 15 years.
Results: Eight patients (89% females) have been diagnosed with PELVIS syndrome and follow with our spina bifida team (avg length of follow up: 6.7 years). All patients had perineal hemangiomas on intial presentation. Hemangiomas were widely distributed involving the back, perineurm and/or lower extremity regions. Five patients (63%) have abormal external genitalia with 4 (50%) requiring anorectal surgery. Six (75%) were treated with propranolol while 4 also required pulse dye laser to reduce the hemangioma size and ulceration. All patients underwent a spine MRI with findings of: 6 (75%) lipomyelomeningocele, and 2 (25%) thickened, fatty filum. Seven (88%) patients underwent a laminectomy

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with tethered cord release (avg age: 7.71 months); 2 with partial untetherings due to transitional lipomas. Surgical findings, postoperative wound issues and long-term functional outcomes will be reviewed.
Conclusions: PELVIS syndrome is a rare compilation of symptoms often noticed at birth that requires multidisciplinary care. Children often have associated spina bifida occulta with tethering lesions. Optimum outcome is achieved with aggressive, prospective management.

Adult patients with occult spinal dysraphism experience chronic but not acute pain at high rate

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Background: Chronic pain is an important co-morbidity in patients with spinal dysraphism. Patients with spinal dysraphism harbor low back, buttock, groin, and proximal lower extremity pain from sources including tethered spinal cord. This project surveys the longitudinal burden of chronic pain in a cohort of adult patients with open and closed forms of dysraphism.
Methods: An observational, retrospective single institution cohort study was performed on 222 patients followed in adult Spina Bifida clinic. Pain scores were assessed acutely by self-report of a pain index score. Chronic pain was detected by surveying clinic notes for primary complaint of pain. Descriptive statistics summarized the cohort and univariate, and multi-variate logistic regression analysis was performed to survey the relationships between variables. In addition, a multidisciplinary survey filled out by patients of the spina bifida clinic. Survey participants were asked directly about daily pain burden.
Results: Pain was mentioned as one of the primary concerns for the most recent clinic visit in 92 of 222 patients (41.4%). The median pain score on the 0-10 scale was 0 (IQR 0-3). Univariable logistic regression analysis showed significantly more chronic pain in patients with closed dysraphism than open MMC (OR 7.88, 95% CI 3.55-17.53, p<0.001). Logistic regression analysis of association showed older patient age and female sex show significant correlation with pain as a primary concern (Age: OR 1.03, 95% CI 1.01-1.06, p=0.011, Sex: OR 2.42, 95% CI 1.36-4.33, p=0.003). In multivariable analysis, sex, and closed v. open dysraphism remain statistically significant. Age and functional lesion level were not independently significant. In addition, the multidisciplinary survey showed 39/105 patients (37.1%) currently live in chronic pain.
Conclusions: Patients with closed but not open dysraphism develop significant chronic pain over the long term. Strategies to mitigate and treat this problem are likely to contribute to quality of life in these patients.

Evaluation of multidisciplinary high-risk pregnancy clinic

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Background: Prior to the COVID-19 pandemic, parents expecting a newborn with myelomeningocele at the authors' institution were routinely seen by a neurosurgeon for prenatal consultation and education to prepare for future challenges as part of a multi-disciplinary clinic. The purpose of this study is to evaluate the perception of usefulness and effectiveness of the prenatal, multidisciplinary, high-risk pregnancy clinic (HRPC) and retrospectively determine what information families wish they had learned.
Methods: Mothers of children with spina bifida completed a demographic survey about their HRPC experience. Using a five-point Likert scale, mothers evaluated their overall preparedness, spina bifida education, delivery plans, surgical expectations, and expectations in terms of quality of life and development. Comments were collected regarding which portion of the educational process was most useful, what was unhelpful/anxiety-causing, and what they wish would have been included in the process. Descriptive statistics are reported.
Results: Seventy-one mothers completed the survey (of 72 approached, 98.6%). Thirty-eight (53.5%) received no prenatal education, 9 (12.7%) received

prenatal education at other institutions, and 24 (33.8%) attended HRPC at the authors' institution.
Mothers who attended HRPC felt more informed and prepared throughout their pregnancy ($M = 4.28$) compared to mothers who did not ($M = 3.47$). Mothers who received prenatal counseling from the authors' HRPC environment or other clinics perceived it as beneficial, answering "agreed" or "strongly agreed" with the statement "I benefited from the discussion in prenatal clinic" (79.2% and 78%, respectively).
When scoring how well their experience met their expectations, based on what they knew before childbirth, the average was 3.66 among parents who attended HRPC clinic versus 3.26 for those who did not. The main perceived benefits were connecting parents to social support groups and meeting the coordinator to help navigate the process.
Conclusions: Prenatal counseling and the HRPC provide perceived utility to families and mothers.

Perinatal features of myelomeningocele patients after total folate fortification in Costa Rica: A 25-year experience

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Background: Following recommendations derived from several studies showing a preventive role of folate on the incidence of spina bifida, in 1997 a congress decrete introduced folate fortification of wheat in Costa Rica, this was followed by additional laws mandating fortification of maize, rice and dairy products over the next 5 years. There was a dramatic reduction of blood folate deficit that declined from a 25% overall deficit to a 3% in women in childbearing age before and after these policy was introduced.
Methods: We conducted a retrospective analysis of data obtained from medical records from 412 patients from the Spina Bifida Clinic at the Hospital Nacional de Ninos, the sole provider for specialized care of these children in the whole country. We conducted direct data recopilation from mothers of children with Spina bifida in order to know if the children had been conceived within the territorial boundaries of Costa Rica, the number of prenatal appointments, Number and timing of ultrasound studies, the detection rate of these ultrasounds according to trimester, public or private setting of US testing, number of weeks at delivery, method of delivery, time to closure of Spina bifida and defect size, functional level, the presence of Kyphosis and symptomatic Chiari malformation, Timing of shunt insertion or ETV procedures.
Results: 98.6% of the children born with spina bifida were conceived in Costa Rica after folate fortification had been implemented. Those children conceived outside Costa Rica had more severe defects with higher functional levels and bigger spinal defects and the timing for their repair was longer whenever they were treated in other countries. 98% of mothers has prenatal care and had an average of 3.5 US studies during their pregnancy with a detection rate of 65% for SB and 77% for congenital hydrocephalus. The average delivery age was 38.4 weeks and all cases detected prenatally were delivered by C section. All children were born in a hospital with specialized medical care and were sent to the National Childrens Hospital for their neurosurgical care. 84% of spina bifida patients were closed 12-24 h after delivery, 12% 24-48 h, 2% in less than 8 h and 2% later than 48 hrs. There was no acute perinatal mortality related to surgical procedures. The children born with SB have a functional level: paraplegic: 13%, L2 10%, L3 47%, L4 9.8%, L5 12% and sacral 6% which corresponds to a gross total of 74% children who are able to walk assisted with orthesis.
Conclusions: Folate fortification not only decreases the incidence of spina bifida in Costa Rica to a 4.8 per 10,0000 live births but also decreases the severity of the defect allowing almost 74% of children to deambulate with the use of orthesis which enables them to carry out more daily functional activities such as attending school and leading future productive lives.

Fetoscopic spina bifida repair: Feasible, safe, preferred

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Background: Introduction: Open fetal spina bifida back closure has been shown to decrease the need for hydrocephalus treatment and improve hindbrain

herniation and lower extremity function. This prenatal surgical procedure increases maternal and fetal perinatal risks.
Objective: Fetoscopic surgical intervention is a less invasive maternal surgical approach.
Methods: We discuss development of a fetoscopic spina bifida surgical training model and implementation of this surgical technique. Outcome data is presented for all spina bifida newborns who underwent back closure at our institution - 23 fetoscopic; 25 open fetal; 24 postnatal - since 2017.
Results: Our fetoscopic technique was utilized on 23 patients via an exposed uterine, 3 (3.5 mm) port approach. All patients underwent 3-layer back closure; however, placode imbrication with primary dural and primary skin closure was statistically more likely in the postnatal cohort. Operative length was significantly increased for fetoscopic closures in comparison to other closure procedures. No prenatal closure patient had a perinatal CSF leak. One fetoscopic and one postnatal patient required a subsequent wound revision. A majority of fetoscopic and postnatal patients were delivered vaginally, whereas all open fetal patients underwent Cesarean section. Mean gestational age at delivery for the fetoscopic group was improved in comparison to the open fetal cohort (31.8 vs. 34.3 weeks) with 1/3 of the cohort born at 37+ weeks. Primary outcome (hydrocephalus treatment by 12 months of age or death) was 50% (10/20) for postnatal closure, 40% (10/25) for open fetal, and 29% (2/7) for fetoscopic. A majority of the fetal cohort experienced improvement in hindbrain herniation with expected or improved lower extremity function.
Conclusions: Fetoscopic closure of an open spina bifida is technically feasible and provides improved maternal risks with equivalent fetal outcomes. A large, multi-institutional study utilizing standardized surgical technique is needed.

Prenatal myelomeningocele closure is associated with higher risk of early detethering procedure for tethered cord: An NSBPR study

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Background: Although benefits resulting from prenatal closure of myelomeningocele (MMC) have been demonstrated, its impact on future spinal cord tethering is not well known. We compared the risk for spinal cord tethering between children with prenatal and those with postnatal MMC repair in a large national registry.
Methods: We included children born at or after 1990 who had prenatal MMC closure and followed in the National Spinal Bifida Patient Registry (NSBPR). We included children from the same period with MMC closed postnatally. The main outcome was time to a secondary detethering procedure. Covariates included year of birth, gender, functional level of neurologic lesion, and reasons for surgery. A multivariable Cox proportional hazard model was fitted. Registry sites were included as random effects in the model to adjust for clustering.
Results: This study identified 474 prenatal and 6,406 postnatal MMC closures in children with spina bifida. 50% were males. 90 (19%) and 1,281 (20%) of those whose MMC was closed prenatally and postnatally, respectively, underwent secondary spinal surgery. The median age for secondary detethering surgery was 2.9 (IQR 1.3-6.7) years in the prenatal closure group and 6 (IQR 3-10.3) years in the postnatal closure group ($p<0.001$). Prenatal MMC closure was associated with early need for secondary spinal cord surgery ($p<0.001$). After adjusting for year of birth, sex, functional level of neurologic lesion, NSBPR sites, the hazard of detethering surgery for an individual with prenatal MMC closure was twice the hazard for an individual with postnatal MMC closure (HR 2.0, 95%CI 1.4-2.7), $p<0.001$). Sensitivity analysis including individuals born before 1990 showed similar results.
Conclusions: Compared with postnatal closure, *in utero* MMC closure was associated with a higher hazard for early spinal cord tethering. This insight demonstrated the need for periodic and comprehensive monitoring of these children. Aggressive intervention may be needed to ensure preservation of functions from tethered spinal cord.

Treatment of sleep disordered breathing in children with myelomeningocele

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Background: Studies have shown a high prevalence of sleep-disordered breathing (SDB) in children with myelomeningocele, but there are minimal published data on the longitudinal care of these patients. The objective of this study is to determine the effectiveness of standard treatments for SDB in children with myelomeningocele.

Methods: We analyzed records from two multidisciplinary spina bifida clinics (2005-2020) to identify all patients with both myelomeningocele and SDB, diagnosed by polysomnography (PSG). The primary outcome of this study was a change in Apnea-Hypoxia Index (AHI, the number of apneic or hypoxic events per hour of sleep) before and after various clinically recommended SDB treatments. Clinical and demographic variables were recorded and evaluated for possible association with improvement of AHI. Analysis included change in AHI (a continuous variable) and whether SDB improved (defined as AHI<2.5 or decrease of AHI by >50% from baseline).

Results: Fifty-nine eligible patients (age 1 mo to 21 yrs, 50% male) had initial AHI > 2.5 and had follow up PSG after treatment for SBD. Mean AHI decreased from 19.5 at baseline to 12.7 after treatment (p=0.01). Children treated with supplemental oxygen and with continuous positive airway pressure (CPAP) had improvement on PSG (13 of 19 and 8 of 12, respectively). Children treated with airway surgery were less likely to improve (7 of 19). Fifteen patients improved from baseline abnormal AHI (AHI > 2.5) to normal (AHI<2.5) after treatment.

Conclusions: SDB is common among children with myelomeningocele. This is the first study to explore their response to treatment. Airway surgery (tonsillectomy/adenoidectomy) may be less effective in treating SDB in this population, compared to other treatments.

First-time tethered cord release among adults with myelomeningocele: An analysis of people in the National Spina Bifida Patient Registry

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Background: We sought to determine the ongoing longitudinal risk, and the roles of lesion level, ambulation status, and prior treatment for Chiari and hydrocephalus, for *first-time* tethered cord release (TCR) during adulthood (≥ 21 years) with myelomeningocele in the NSBPR.

Methods: Adults in the NSBPR registry who had not previously undergone a TCR during childhood were studied. Kaplan Meier curves, using time to first TCR, were constructed independently for males and females. Cox proportional hazards analyses sought correlations between lesion level, ambulation status, sex, and previous treatment for hydrocephalus and Chiari malformation.

Results: 47 of 967 adults (4.9%) in the NSBPR underwent their *first* TCR during adulthood. There were 35 females (74.5%) and 12 males (25.5%); this sex distribution was significantly different from the entire adult cohort (p = 0.015). Kaplan-Meier curves for first TCR for females and males were significantly different (log-rank test, p = 0.01). TCR rates were correlated with sex (males at lesser risk; OR = 0.31 (CI 0.16-0.62), p < 0.001); prior treatment for hydrocephalus (prior treatment at lesser risk; OR = 0.21 (CI 0.20-0.42), p < 0.001) and prior treatment for Chiari malformation (prior treatment at greater risk; OR = 3.84 (CI 1.50-9.88), p = 0.005).

Conclusions: Adults with myelomeningocele who escape childhood without a TCR have lesser risk for spinal cord tethering, clearly not due to spinal column growth and reflecting other factors such as dy-

namic changes in spinal cord health. Among people with MMC who underwent their first TCR as adults, females seem to be over-represented. Similar to our prior childhood study, people with a previous Chiari decompression seem to be over-represented whereas those with a previous treatment for hydrocephalus seem to be under-represented. These novel findings deserve further study.

Inclusion cysts after open prenatal spina bifida aperta repair: Data from the Zurich Cohort

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Background: Open prenatal spina bifida aperta repair (OPSBAR) has been shown to produce better outcomes than postnatal repair. However, the benefits gained with OPSBAR may be challenged postnatally by the occurrence of inclusion cysts (IC). Here we present incidence, presentation, diagnostic work-up, management and outcomes of IC in our cohort from the Zurich Center for Spina Bifida.
Methods: We analyzed data of 110 children born after OPSBAR at the Zurich Center for Fetal Diagnosis and Therapy with particular focus on the above variables. Indications for IC surgery included symptoms (back pain, change in motor, bladder, and bowel function) and/or IC size progression and/or worsening hydro- or syringomyelia on spinal MRI within 6-12 months after initial diagnosis. Perioperative assessment included neurologic function, cystometry, anorectal manometry, and MRI.
Results: In twenty-five children (23%), IC were detected on MRI within the first 2 years of life. Fourteen children (13%) underwent IC resection in different centers. After IC resection, all but one child (93%) developed a neurogenic bladder and bowel dysfunction. Motor loss of one functional level was found in four patients (29%). Re-tethering was common (100%).
Conclusions: There is a high incidence of IC in patients with former OPSBAR (23%) at our center. The incidence seems to be higher than after postnatal spina bifida repair. It remains unclear if IC after OPSBAR are related to surgical technique (incomplete resection of the zona epithelioserosa). Inclusion cysts endanger the benefits of OPSBAR, thus an active surveillance for the occurrence of IC is mandatory. The decision to treat must be considered diligently, ideally in a multidisciplinary team. In order to improve techniques and long-term outcome, we encourage all centers to report their IC data openly.

Long-term outcomes among people with lipomyelomeningocele (LMM): An analysis from the National Spina Bifida Patient Registry (NSBPR)

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Background: Previous studies of people with LMM have provided conflicting results regarding outcomes, particularly retethering rates (ranging from 2-47%). The NSBPR, a multi-institutional registry presently with more than 10,000 registrants, provides an opportunity to study long-term outcomes, and compare in particular the long-term retethering rates after early initial versus later repair among a large cohort.
Methods: Registrants with LMM enrolled between 2009-2019 were studied. Two groups were contrasted – those undergoing initial repair ≤ 6 months of age as a proxy for prophylactic repair (Group I; n = 547), and those undergoing repair > 6 months of age (Group II; n = 623). Clinical outcomes and rates of

subsequent tethered cord release (TCR) were contrasted. Acute clinical deterioration following initial repair and subsequent TCR were studied.

Results: Of 1170 identified registrants, at the last visit 68.7% had sacral level function, 80.1% were community ambulators, 66.4% had bladder dysfunction, 55.8% were on a bowel regimen, and 51.8% had bowel continence. Subsequent TCR were performed in 24.9%with a greater rate among Group I (27.8%) compared with Group II (22.3%) ($p = 0.036$). Kaplan-Meier analysis confirmed that TCR also occurred earlier after initial repair in Group I ($p < 0.001$). Mean length of follow-up was significantly greater ($p < 0.001$) in Group II (16.1 yrs) than in Group I (12.24 yrs). However, there were no significant clinical differences between Groups I and II with respect to functional level, ambulation, use of braces or wheelchairs, or bladder/bowel function at last follow-up. Although there were no significant changes in neurological or urological function after initial repair, there was a significant post-operative decline in functional motor level after subsequent TCR when compared with pre-operative status ($p < 0.001$).

Conclusions: In this, the largest long-term study of people with LMM, long-term clinical outcomes are significantly worse than previously reported. Those who underwent early repair (≤ 6 months) had higher retethering rates than those who had delayed repair (>6 months) with no significant differences in functional outcomes. TCR itself was associated with significant post-op deterioration in functional level.

Prevalence of ADHD in a pediatric hydrocephalus prospective sample

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Background: Little is known about the prevalence of attention-deficit/hyperactivity disorder (ADHD) in children with hydrocephalus. Furthermore, the relationship between ADHD and intellectual disability is unknown in this population. The primary aim of this study is to provide an estimate of the prevalence of ADHD in children with hydrocephalus.

Methods: We conducted a cross-sectional study of parents of children with hydrocephalus ages 6-12 using phone surveys. The Child and Adolescent Intellectual Disability Screening Questionnaire (CAIDS-Q) was used to screen for intellectual disability. The National Institute for Children's Health Quality (NICHQ) Vanderbilt Assessment Scale was used to screen for ADHD. Variables tested for association included gender, race, age, involvement of Individualized Educational Program (IEP), family income level, family educational level, etiology of hydrocephalus, and primary treatment (Shunt versus endoscopic third ventriculostomy (ETV)).

Results: Seventy-five children (51%) had CAIDS-Q scores indicating no significant intellectual disability. Of these, 25 (33%) were positive for the NICHQ assessment scale, indicating likely diagnosis of ADHD. The 95% confidence interval for this estimate of 33% prevalence was 22.9% to 45.2% and does not include the estimated prevalence of ADHD in Alabama (12.5%). Therefore, children with hydrocephalus may be at higher risk for ADHD than the general population. Among the 147 patients screened for intellectual disability, etiology of hydrocephalus, lower family income, and VP shunt as primary treatment were associated with presence of intellectual disability on univariate analysis. Multivariate analysis showed only family income category to be independently associated with presence of intellectual disability.

Conclusions: The prevalence of ADHD among children with hydrocephalus is significantly higher than the general population. Intellectual disability is also common (49%). Routine screening for ADHD and intellectual disability in children with hydrocephalus may be indicated to assure adequate resources are provided to optimize educational outcomes.

Scope of care in the first four years of life for individuals born with myelomeningocele: A single institution experience

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Background: Children with myelomeningocele are known to be consumers of substantial healthcare resources, with many early hospital encounters. The purpose of this study was to survey the extent of

medical and surgical care that patients with myelomeningocele receive during the first four years of life.

Methods: Clinical and demographic data were collected on newborn infants with open myelomeningocele from the Children's of Alabama Spina Bifida Web Tracker, a prospective, comprehensive spina bifida database. Additional data pertaining to all hospital admissions, surgical procedures, and clinic visits were collected from the medical record.

Results: One hundred and fourteen subjects with a primary diagnosis of myelomeningocele between 2004 and 2015 were included. Males slightly predominated (55%), 72% were Caucasian, 11% Hispanic/Latino; 28% had a mid-lumbar functional lesion level.

Over the first four years of life, 688 total surgical procedures were performed (an average of 86 per child): 438 in year 1, 100 in year 2, 84 in year 3, and 66 in year 4. The mean number of hospital visits was 40.5. Total average drive time per patient over 4 years being 103.8 hours. Average number of nights spent in the hospital was 51.

Conclusions: Children with myelomeningocele need multiple hospitalizations, surgeries, and medical encounters in the first 4 years of life. These data will be valuable when counselling new parents and prospective parents of children with this condition.

Trends in the early care of infants with myelomeningocele in the United States 2012-2018

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Background: The early care of children with spina bifida has changed with increasing availability of fetal surgery and evidence that fetal repair improves the long-term outcomes of children with myelomeningocele. We sought to determine current trends in the prevalence and early care of children with myelomeningocele using a national administrative database.

Methods: Retrospective, cross-sectional cohort study of infants with spina bifida admitted within the first 28 days of life using the 2012 -2018 Healthcare Cost and Utilization Project National Inpatient Database. Patients with spina bifida were identified by ICD code and stratified into a cohort with a coded neonatal repair of the defect and those without a coded repair. This database had no identifier specific for fetal surgery, but it is likely that a substantial number of infants without a coded repair had fetal surgery.

Results: We identified 5,090 patients with a coded repair and 5,715 without a coded repair. Overall prevalence of spina bifida was 3.94 per 10,000 live births. The percentage of patients without neonatal repair increased during the study period compared to those with repair (p=0.0002). The cohort without neonatal repair had a higher risk of death (p<0.001), prematurity (p<0.001) and low birth weight (p<0.001). More shunts were placed in patients who underwent neonatal repair (p <0.001). Patients without neonatal repair were less likely to have public insurance (p=0.0052) and more likely to reside in zip codes within the highest income quartile (p=0.0002).

Conclusions: The prevalence of spina bifida from 2012-2018 was 3.94 per 10,000 live births, with an increasing number of patients without neonatal repair of the defect, suggesting increased utilization of fetal surgery. Patients without neonatal repair had a higher risk of death, prematurity and low birth weight, but were more likely to have commercial insurance and reside in high-income zip codes.

Time to shunt failure in children with myelomeningocele: An analysis of the NSBPR

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Background: Hydrocephalus is common among children with myelomeningocele, often treated with

a shunt. Although much is known about factors related to first shunt failure, less data are available about shunt failures after the first one. The purpose of this study was to use a large data set to explore time from VPS placement to first shunt failure in children with myelomeningocele and to explore factors related to multiple shunt failures.

Methods: Data were obtained from the NSBPR. Children with myelomeningocele enrolled within the first 5 years of life and with all lifetime shunt operations recorded in the registry were included. Kaplan-Meier survival curves were constructed to evaluate time from shunt placement to shunt failure. The total number of children who experienced at least 2 shunt failures was calculated. A proportional means model was performed to calculate adjusted hazard ratios (HR) for shunt failure based on sex, race/ethnicity, lesion level, and insurance status.

Results: A total of 1691 children met the inclusion criteria. Fifty-five percent (938 of 1691) experienced at least 1 shunt failure. Median length of follow-up was 5.0 years. The estimated median time from initial shunt placement to first failure was 2.34 years (95% CI 1.91–3.08 years), 26% of patients had at least 2 shunt failures, and 14% of patients had at least 3. Male children had higher likelihood of shunt revision (HR 1.25, 95% CI 1.09–1.44). Some children from racial and ethnic minority groups had a lower likelihood of shunt revision (non-Hispanic Black children HR 0.74, 95% CI 0.55–0.98; Hispanic children HR 0.74, 95% CI 0.62–0.88).

Conclusions: Among the children with myelomeningocele, the median time to shunt failure was 2.34 years. The observed higher likelihood of shunt revisions among males and lower likelihood among some children from racial and ethnic minority groups illustrate a possible disparity in hydrocephalus care.

Utilizing a clinical pathway for coordination of care of infants with myelomeningocele

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Background: An increasing number of infants are born with myelomeningocele and are transferred to our Neonatal Intensive Care Unit (NICU) for management after birth. As of 2013, approximately 1.61 infants out of 10,000 live births were born in Massachusetts with spina bifida. Due to the many medical complexities of infants with Spina Bifida, a standardized guideline to direct the management of care was needed. The complexities of care include management of the lesion, bladder management, bowel management, infection control, coordination of consultations and routine health maintenance.

Methods: A review of the literature and existing protocols and pathways at Boston Childrens Hospital was performed prior to creation of the clinical pathway

Collaboration of the NICU director and Advanced Practice Providers as well as neurosurgery director of Neonatal and Congenital Anomaly and Advanced Practice Providers to determine the needs of infants born with myelomeningocele immediately after birth and during the course of their admission and follow-up at discharge Review of the clinical pathway by the QI team and clinical pathway committee Publication and distribution of clinical pathway and order set.

Results: Physicians, Nurses, Advanced Practice Providers, residents, and therapists for infants with myelomeningocele are using the pathway to standardize care. NICU and neurosurgery teams collaborate by using the pathway and order set. Consultations are requested as part of the order set and therefore routine health maintenance and consultations required for infants with myelomeningocele (specific imaging, urology, orthopedics, physical therapy) needs are addressed early in course of treatment.

Use of antibiotics and preop dressing to cover the lesion to prevent meningitis have been standardized. The use of the clinical pathway offers clinical guidance and standardization for specialty clinicians based on evidence, supporting continuity in care for a complex population. Supports a seamless transition from the inpatient team to providers within the multidisciplinary clinic, including opportunity for families to meet ambulatory care providers.

Conclusions: Due to the many medical complexities of infants with Spina Bifida, a standardized guideline to direct the management of care was needed. The complexities of care include management of the lesion, bladder management, bowel management, infection control, coordination of consultations and routine health maintenance. Creating this streamlined approach did improve care for these patients by ensuring that the require consultations

happened automatically, without long wait times and encouraged continuous collaboration through the specialties. Future work would include completing a QI project to determine if this has impacted LOS, infection rate, and to determine fidelity to the pathway. Using a workgroup and QI projects to improve discharge process for infants with myelomeningocele. Other future work could look at the impact of pathway on family experience during this admission.

Post-traumatic stress symptoms in caregivers of children with hydrocephalus

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Background: Hydrocephalus is most often treated with the installation of shunt catheters to direct excess cerebrospinal fluid outside of the brain. While effective, shunts have an unpredictable pattern of failure. The omnipresent threat of shunt failure along with the potential need for invasive investigations can be stressful for patients and caregivers and may lead to post-traumatic stress symptoms (PTSS) in this population.
Methods: A cross-sectional analysis of children with hydrocephalus and their caregivers was conducted. Caregivers completed a measure of their own PTSS (the Post-Traumatic Stress Disorders Checklist for the DSM-V (PCL-V)) and resilience (the Connor Davidson Resilience Scale (CD-RISC)). Pediatric patients rated their own PTSS and resilience using the Acute Stress Checklist for Kids (ASC-Kids) and CD-RISC.
Results: Ninety-one caregivers completed the PCL-V. Mean score was 17.0 (SD 15.7; median 13.0). 14% scored above 33, the threshold suggestive of a preliminary diagnosis of post-traumatic stress disorder. There was a statistically significant association between caregiver post-traumatic stress and marital status, child's race, and caregiver education. 52% of caregivers reported their child's hydrocephalus as the most significant source of their PTSS. Children did not have markedly elevated levels of PTSS. 41% of caregivers and 60% of children scored in the lowest resilience quartile compared to the general population.
Conclusions: Results from this study suggest that post-traumatic stress affects caregivers with hydrocephalus with low levels of resiliency being a potential catalyst. In order to address this, we have begun to implement a program created to teach skills to caregivers in order to increase their ability to handle stressful situations.

Neurosurgical management of myelomeningocele in severely premature infants

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Background: Myelomeningocele (MMC) is the most common neural tube defect, but rarely seen in premature infants. Given the rarity of this population, we aimed to share our institutional experience and outcomes with severely premature infants with MMC.
Methods: We performed a retrospective review of severely premature infants (< 32 weeks gestational age) identified through our multidisciplinary spina bifida clinic (1995-2021). Descriptive statistics were compiled, including timing of MMC closure and incidence of adverse events such as sepsis, CSF diversion, and meningitis.
Results: Twelve patients were identified with MMC and born at or before 32 weeks gestational age. Mean gestational age was 27.4 weeks (SD 2.9). Median time to MMC closure was 1 day (IQR 0.5 - 84.5). Five patients were closed within the 48hr recommended window, 2 experienced delayed closure (107 and 139 days), and one patient's defect epithelized without surgical intervention. 8/12 (66%) patients required permanent cerebrospinal fluid (CSF) diversion; mean of 3 years after birth, ranging from 1 day to 16 years. Only 2 patients necessitated additional CSF diversion. Two developed sepsis (2/4 SIRS criteria), and 2 patients experienced intraventricular hemorrhages. Of the 7 patients with known MMC closure dates, none developed meningitis (positive CSF cultures before MMC closure).

Median follow up duration was 9.7 years. 3 patients passed away; 2 before 2 years of age and of causes unrelated to surgical intervention, and one within 24 hours of closure due to severe IVH.

Conclusions: While some patients underwent delayed closure, the overall rate of meningitis, sepsis, and mortality for preterm children with MMC was analogous with those born at term. Nearly all patients required permanent CSF diversion but rates of revision were not different between term and preterm patients. Overall, outcomes for premature infants with MMC appear to be comparable to term infants with MMC, without significant consequences for delayed closure.