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Fetoscopic, three-miniport technique for prenatal myelomeningocele repair: Technical evolution and outcomes in 60 consecutive cases

Amanda Kwasnicki[1], Smruti K. Patel[1, 2], Mounira Habli[3, 5], Foong-Yen Lim[3, 4], Jose Peiro[3, 4], Charles B. Stevenson[1, 3]

[1]Division of Pediatric Neurosurgery, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, United States, [2]Department of Neurosurgery, University of Cincinnati College of Medicine, Cincinnati, Ohio, United States, [3]Fetal Care Center of Cincinnati, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, United States, [4]Division of Pediatric Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, United States, [5]Department of Surgery, University of Cincinnati College of Medicine, Cincinnati, Ohio, United States

Background: The Management of Myelomeningocele Study (MOMS) demonstrated that open fetal closure of myelomeningocele (MMC) improved neurologic outcomes compared to postnatal closure. Fetoscopic MMC repair is a promising alternative to open fetal closure. We sought to describe the technical aspects, evolution, and clinical outcomes of a fully endoscopic technique providing a multi-layered, closure for prenatal fetoscopic MMC repair utilized at our center over the last 6 years.
Methods: IRB-approved study of the technique used for the first 60 consecutive fetoscopic MMC repairs performed at our center from 11/2015 to 07/2022. We review the technical details of the procedure, changes implemented over time, and clinical outcomes.
Results: Midline maternal limited laparotomy is performed under a combination of general and epidural anesthesia. The uterus is exposed and three fetoscopic ports (6/10/10 Fr) are placed under US-guidance and direct visualization via Seldinger technique. The neural placode is released utilizing bimanual technique with laparoscopic scissors and microdissectors. Duraplasty is performed by overlaying two umbilical-cord-matrix allograft patches sequentially and securing them with absorbable suture. The skin is then closed primarily with running suture; an acellular dermal allograft patch was utilized in 15 cases (25%). No procedures required conversion to open hysterotomy. There were no significant intraoperative complications. A watertight seal was achieved in all cases, with no evidence of CSF leak at birth. Average estimated gestational age at delivery was 33.4 weeks. Cesarean-sections were performed in 32 patients (53%) and vaginal delivery in 28 patients (47%). No infants demonstrated significant brainstem dysfunction. Fourteen (23%) required CSF diversion.
Conclusions: A laparotomy-assisted, three-miniport fetoscopic approach using dural patches offers excellent access and magnification for watertight closure in prenatal myelomeningocele repair. Initial results demonstrate clinical outcomes comparable to those of open fetal repair reported in MOMS, while obviating the need for delivery via cesarean section in every case.

Risk factors for postnatal cerebrospinal fluid diversion after percutaneous fetoscopic open spina bifida repair

Alexander Van Speybroeck[1, 2], Ramen Chmait[2, 3], Jason Chu[1, 4], Huyen Pham[2, 5], Arlyn Llanes[2, 3], Lisa Korst[6], Eftichia Kontopoulos[6, 7], Ruben Quintero[7]

[1]Pediatrics, Children's Hospital Los Angeles, Los Angeles, California, United States, [2]USC Keck School of Medicine, Los Angeles, California, United States, [3]Division of Maternal Fetal Medicine, Los Angeles, California, United States, [4]Division of Neurosurgery, Los Angeles, California, United States, [5]Department of Obstetrics and Gynecology, Los Angeles, California, United States, [6]Childbirth Research Associates, North Hollywood, California, United States, [7]The USFetus Research Consortium, Miami, Florida, United States

Background: The aim of this study was to assess the rate and risk factors for (CSF) diversion at 1 year of life after percutaneous fetoscopic repair of open spina bifida (OSB).
Methods: This is a prospective study of patients who underwent percutaneous fetoscopic OSB repair from February 2019 to July 2022. The decision to offer CSF diversion was based on postnatal magnetic resonance imaging and clinical findings. Preoperative, operative, and neonatal characteristics were

examined as predictors for a CSF diversion procedure (ventriculoperitoneal shunt or endoscopic third ventriculostomy) within the first 12 months of life, using bivariate analysis and multiple logistic regression models. Continuous variables are presented as mean ± standard deviation.

Results: 57 patients were enrolled; of these, 17 were <12 months old and 1 had an incomplete surgery, leaving 39 study patients. Any degree of hindbrain herniation (HBH) reversal was noted in 35 (89.7%), and complete HBH reversal in 19 (48.7%). CSF diversion by 12 months of age was required in 13 cases (33.3%). Patient characteristics are compared in the Table. In bivariate analysis ($p<0.05$), either fetal ventricular measurement >= 15 mm prior to surgery and lack of complete hindbrain HBH reversal at neonatal assessment were associated with the need for a CSF diversion procedure. Logistic regression showed that cases with a lateral ventricle >=15 mm were nearly 11 times more likely to have CSF diversion (OR 10.93, 95%CI 1.71-69.94, $p = 0.0116$), and cases lacking complete HBH reversal were > 6 times more likely than cases of complete reversal to have CSF diversion (OR 6.61, 95%CI 1.11-39.23, $p=0.0378$).

Conclusions: Percutaneous fetoscopic repair of OSB was associated with a 33% risk of postnatal CSF diversion at 1 year of life. Risk factors for CSF diversion included preoperative ventriculomegaly >= 15 mm and lack of complete HBH reversal at birth.

Effect of allograft patch closure upon incidence of spinal inclusion cyst formation following open fetal myelomeningocele repair

Smruti K. Patel[1, 2], Sara Hartnett[1], Mridula Bethi[1], Mounira Habli[3], David N. McKinney[3], Foong-Yen Lim[3, 4], Jose Peiro[4, 3], Charles B. Stevenson[1, 3]

[1]Division of Pediatric Neurosurgery, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, United States, [2]Department of Neurosurgery, University of Cincinnati College of Medicine, Cincinnati, Ohio, United States, [3]Fetal Care Center of Cincinnati, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, United States, [4]Division of Pediatric Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, United States

Background: Studies reporting the relationship of prenatal myelomeningocele repair and the incidence of spinal inclusion cyst (sIC) formation and related spinal cord retethering are relatively scarce and primarily predate the Management of Myelomeningocele Study (MOMS) trial. The aim of this study was to evaluate the incidence of sIC formation following open fetal myelomeningocele (fMMC) repair and the effect of dural patch closure.

Methods: The authors conducted a retrospective review of patients who underwent open fMMC repair at our institution between March 2011 and June 2020. All patients met criteria for intervention defined by the Management of Myelomeningocele Study (MOMS). Primary outcomes included: need for CSF diversion, extent of reversal of hindbrain herniation, motor function/ambulatory status, and sIC formation.

Results: Of the 56 patients who underwent open fMMC repair, 52 had adequate spinal imaging for review. Twelve of these patients (23%) were found to develop sIC. Six patients experienced symptoms and required surgical detethering with sIC resection. Six additional patients had evidence of sIC on surveillance MRI but remained asymptomatic. We found a statistically significant relationship between the use of a dural allograft patch and sIC formation ($p=0.003$). When compared to patients without sIC development, there was no statistically significant difference in primary closure vs. the use of allograft at the level of the fascia ($p=0.47$), or skin ($p=0.51$). The rate of hydrocephalus requiring CSF diversion was 52%. Interestingly, 98% of patients had improvement in the extent of hindbrain herniation. Dural patch closure did not have any effect on the rate of progressive hydrocephalus ($p=0.07$) or degree of reversal of hindbrain herniation ($p=1.0$).

Conclusions: Our study suggests that children with prenatally repaired MMC are at higher risk for development of sIC and associated symptoms than following postnatal repair. The presentation of symptoms was also earlier in these patients than previously reported following postnatal repair. The use of a dural allograft patch appears to have a positive correlation with sIC formation. Future investigations evaluating the incidence of sIC after fetoscopic MMC repair, in which primary dural closure typically cannot be achieved and a dural patch is most often required, will be helpful in facilitating prenatal counseling for patients considering fetal intervention.

Tethered cord release in patients after open fetal myelomeningocele closure: Intraoperative neuromonitoring data and patient outcomes

Tracy Flanders[1], Alier Franco[2], Kristen Lincul[1], Samuel Pierce[1], Katie M. Schmidt[1], Julie Moldenhauer[1], N. Scott Adzick[1], Gregory G. Heuer[1]

[1]Division of Neurosurgery, Children's Hospital of Philadelphia, Bryn Mawr, Pennsylvania, United States, [2]Specialty Care, Nashville, Tennessee, United States

Background: To better understand the clinical course and impact of tethered cord release surgery on patients who have previously undergone open spinal dysraphism closure *in utero*.
Methods: This is a single-center retrospective observational study on patients undergoing tethered cord release after having previously had open fetal myelomeningocele (MMC) closure. All patients underwent tethered cord release surgery with a single neurosurgeon. A detailed analysis of the patients' preoperative presentation, intraoperative neuromonitoring (IONM) data, and postoperative course was performed.
Results: From 2009 to 2021, 51 patients who had previously undergone fetal MMC closure had tethered cord release surgery performed. On both preoperative and postoperative manual motor testing, patients were found to have on average 2 levels better than would be expected from the determined anatomic level from fetal imaging. The electrophysiologic functional level was found on average to be 2.5 levels better than the anatomical fetal level. Postoperative motor levels when tested on average at 4 months were largely unchanged when compared to preoperative levels. Unlike the motor signals, 46 (90%) of patients had unreliable or undetectable lower extremity somatosensory evoked potentials (SSEPs).
Conclusions: Tethered cord surgery can be safely performed in patients after open fetal MMC closure without clinical decline in manual motor testing. Patients often have functional nerve roots below the anatomic level. Sensory function appears to be more severely affected in patients leading to a consistent motor-sensory imbalance.

Significant brainstem dysfunction in neonates with myelomeningoceles: A comparison of prenatal versus postnatal closure

Paige A. Lundy[1], Paul Grabb[2], Emmanuel Vlastos[2], Michael Partington[2]

[1]Pediatric Neurosurgery, University of Alabama-Birmingham, Birmingham, Alabama, United States, [2]Childrens Mercy, Kansas City, Missouri, United States

Background: The purpose of this study was to compare the incidence of significant brainstem dysfunction (SBD) in neonates with myelomeningocele who have been treated with prenatal versus postnatal closure at a single institution.
Methods: The records and imaging of all children undergoing either prenatal (n = 27) or postnatal (n = 60) closure of myelomeningocele at the authors' institution from December 2014 through May 2021 were reviewed. SBD, fetal ventricular size, gestational age at fetal imaging and delivery, postnatal ventricular size, need for and type of hydrocephalus treatment, spinal neurological level at birth, anatomical Chiari severity, death, and prenatal or postnatal repair were factors recorded. SBD was defined by need for airway surgery or gastrostomy tube, or endotracheal intubation because of apnea, aspiration, or airway control problems. Comparisons between prenatal and postnatal cohorts and between the cohorts with and without SBD were performed.
Results: SBD occurred in 25% and 0% of neonates who underwent postnatal and prenatal closure, respectively. There were no differences in fetal ventricular size or spinal neurological level between the prenatal and postnatal cohorts or between those with or without SBD. Anatomical severity of the Chiari malformation after birth was worse in the postnatal cohort. Hydrocephalus treatment was required in 70% and 33% of infants who underwent postnatal and prenatal closure, respectively. All three deaths were in the postnatal group from SBD.
Conclusions: Prenatal closure of myelomeningocele is associated with a significant reduction in SBD.

The Zurich spina bifida experience: A model for comprehensive pre-and postnatal care and research

Beth Padden[1], Nicole Ochsenbein[2], Nele Struebig[2], Barbara Casanova[1], Ueli Möhrlen[1], Martin Meuli[1], Brigitte Seliner[1], Alexandra Wattinger[1], Spina Bifida Study Group Zurich[1, 2], Luca Mazzone[1]

[1]Rehabilitation, University Children's Hospital Zurich, Zurich, Switzerland, [2]University Hospital Zurich, Zurich, Switzerland

Background: In accordance with the position statement on fetal myelomeningocele repair, the Zurich Center for Fetal Diagnosis and Therapy (ZCFDT), the Zurich Center for Spina Bifida (ZCSB) and the Spina Bifida Academy (SBA) are committed to upholding the 6 key areas for teams performing prenatal spina bifida aperta repair (PSBAR).
Methods: Open PSBAR began in 2010 at the ZCFDT. In order to meet these requirements and treat the growing number of spina bifida patients, the ZCSB was founded in 2018. The center has a matrix organization and includes core disciplines neurorehabilitation, neuroorthopedics, neurourology, neuroproctology, neurosurgery and nursing. Developmental medicine, dietary consultation, social work, psychology, radiology, gynecology, endocrinology, physical therapy, pneumology and wound specialists complete the multidisciplinary team. Full day multidisciplinary clinic at 3, 6, 12, 18, 24 months and annually thereafter are planned. Care coordination by advanced practice nurses and administrative coordination are essential in our care-system. The team meets for a morning briefing and post-clinic debriefing on clinic days and for a conference monthly. Data is collected prospectively in the REDCap™ database and reporting occurs regularly. The SBA study group oversee research.
Results: To date 192 open PSBAR were performed; 25 on average in the past 5 years. The Covid pandemic had a negative impact. Patients come from 11 countries: Switzerland 38, Germany 86, Austria 33, Russia 9, Slovakia 8, France and Italy 7 and 1 each from Sweden, UK, Romania and Croatia. Until 2022 follow-up appointments were provided regardless of insurance status; only 8 families had declined care at ZCSB. Since then follow-up requires insurance coverage and 31 families from abroad have declined initial care or discontinued care at ZCSB.
Conclusions: The ZCSB offers comprehensive, structured multidisciplinary care from infancy to adulthood including a transition clinic. Our outcomes results are on par with international reporting following open PSBAR.

Supporting patients and families with a fetal spina bifida advanced practice nurse program coordinator

Katie M. Schmidt, John Chuo, John Flibotte, Lori Howell

Children's Hospital of Philadelphia, Philadelphia, Pennsylvania, United States

Background: Patients affected by spina bifida or a neural tube defect have morbidities and mortalities as well as quality of life issues. Options after prenatal diagnosis include fetal surgery to close the myelomeningocele (MMC) or myeloschisis or traditional postnatal closure. Fetal surgery to close the MMC proved to be safe and effective in reducing the need for a shunt and improved motor outcomes at 30 months of age for those mothers and fetuses that qualify and parents willing to proceed with fetal surgery. Families faced with this diagnosis encounter several transition periods in a short time from the prenatal period, delivery, care in the neonatal intensive care unit (NICU), then discharge home and transition of care to the pediatrician and outpatient specialty providers.
Methods: The Fetal Spina Bifida APN Program Coordinator, counsels in the prenatal period to provide anticipatory guidance, works closely with the patient, medical team, and parents while in the NICU, and coordinates discharge and follow-up, including telemedicine. The APN develops programmatic systems to support this patient population.
Results: The Fetal Spina Bifida APN Program Coordinator meets parents during the initial prenatal diagnosis then again later in pregnancy to provide anticipatory guidance about delivery and NICU stay. During the inpatient period, the multidisciplinary team, including various providers from neurosurgery and neonatology, round weekly, in-person in the NICU for all spina bifida patients. The use of telemedicine and close follow-up with surgical subspecialists and the spina bifida clinic takes place after discharge. To date over the past 21 months, 51 telemedicine visits with the patient and parents or tele-signout visits with the pediatrician, patient, and parents took place. The telemedicine/tele-signout visit unveiled safety issues in and around the dis-

charge process and education. Among the 51visits, 23 (45%) have been evaluated using an impact tool. Of these evaluated visits, 17 (74%) led to identification of one or more discharge issues requiring intervention. Examples of these included incorrect formula sent to the patient leading to an inpatient hospitalization, inaccurate mixing of formula by parents preventing an emergency department visit, need for outpatient specialty follow-up appointments, identification of the need for a sooner pediatrician appointment, and notification to the apnea team that a patient was sent home on caffeine and a monitor. Education provided during these telemedicine visits also prevented parents from needing to call a provider. The Fetal Spina Bifida APN Program Coordinator also ensures all patients have routine neurodevelopmental evaluations to assess development and attainment of milestones.

Conclusions: The Fetal Spina Bifida APN Program Coordinator improves the course of care for patients with spina bifida and their families from prenatal diagnosis through childhood by providing consistency of care in critical transition periods. This role could be adapted for various complex diagnoses from the prenatal period through childhood.

Neurodevelopmental outcomes of 5-year-old patients after open prenatal spina bifida aperta repair

Beth Padden, Theres Möhrlen, Luca Mazzone, Ueli Möhrlen, Spina Bifida Study Group Zurich, Beatrice Latal

Rehabilitation, University Children's Hospital Zurich, Zurich, Switzerland

Background: The Zurich Center for Spina Bifida (ZCSB) provides comprehensive follow-up after open prenatal Spina Bifida Aperta repair (OPSBAR) at the Zurich Center for Fetal Diagnosis and Therapy. Comparison studies of pre- vs postnatal repair have shown improvement in motor function outcome but unchanged cognitive outcomes. We thus examined neurodevelopmental and functional outcomes at 5 years.

Methods: We prospectively follow-up all children with OPSBAR at our center with a standardized program, data obtained is entered in a REDCap™ database. Assessments include motor function in daily life, therapies, need for cerebral spinal fluid diversion (CSFD) and IQ (Wechsler Primary Preschool Intelligence Scale (3rd and 4th version) and fine motor function (Zurich Neuromotor Assessment).

Results: 58 patients turned 5 years by July 31, 2022; 39 had complete follow-up data. Mean age at follow-up was 5.2 years (SD 0.36). Nine (23%) children were able to walk independently, 25 (64%) with orthotics and/or assistive devices and 5 (13%) were unable to walk. 74% were community walkers (Hoffer FMS). Functional level (all between L2 and S1) correlated with ambulation status (p=0.015). All with a functional level L5 and S1 were community walkers and 19/21 (90%) with a functional level L4 were ambulatory. 38% required a manual wheelchair and 84% wore daytime orthotics. Mean IQ of 36 children with valid results was 88.6 (SD 15.4) and fine motor function score was -2.4 (SD 2.1), both significantly lower than the norm (100 ± 15 SD, $p<0.001$, and 0 ± 1 SD, $p<0.001$, respectively). Lower IQ was not associated with the need for CFSD (n = 19, 49%, p=0.50). Ninety-five percent of children received physical therapy, 39% special needs therapy and 31% occupational therapy.

Conclusions: The results from the ZCSB demonstrate that benefits of prenatal repair persist until school-age. However, cognitive and fine motor impairments occur in this population, which may impact school-performance.