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A post-natal myelomeningocele repair completed on a 26-week premature neonate

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ABSTRACT

Myelomeningocele (MMC) is a common neural tube defect that leads to lifelong physical and neurological disabilities. Fetal surgery for MMC repair, first studied in the Management of Myelomeningocele Study (MOMS) trial, showed improved neurologic outcomes when performed between prenatal estimated gestational age (eGA) of 19 0/7 to 25 6/7 weeks[1]. Limited data exists on postnatal MMC repairs at less than 30 weeks eGA and extremely low birth weight due to high mortality rates. No literature exists regarding MMC repair at the bedside.

Our case describes an MMC repair performed postnatally on a 26 6/7 weeks eGA neonate weighing 650 grams on day of life 3. Due to significant hemodynamic instability requiring vasoactive medications and high-frequency oscillator ventilator (HFOV) support, the repair was performed at the bedside. The neonate did well with no postoperative infectious concerns or need for cerebrospinal fluid (CSF) diversion to date. Serial brain MRIs were performed and showed stable ventriculomegaly with minimal hindbrain herniation.

This case report describes a successful bedside postnatal MMC repair on an extremely premature, low birth weight neonate on HFOV and inotropic medications. Per the MOMS trial, fetal MMC repair is associated with a 50% reduction in CSF diversion procedures, thus lowering the risk of life-long risk of malfunction and negative neurologic outcomes[1]. This case illustrates that postnatal repair in extremely preterm neonates may also be associated with decreased risk of CSF diversion. Long-term follow-up and larger case series are needed to determine the impact of postnatal repair in premature neonates.

1. INTRODUCTION

Myelomeningocele (MMC) is a birth defect due to a failure of neural tube closure. Its incidence in the United States is about 0.2 to 0.4 per 1000 live births[2]. MMC results in an exposed neural placode and resultant neurological deficits such as hindbrain herniation and brainstem anomalies. These hindbrain anomalies can result in CSF flow obstruction and hydrocephalus[3].

Outcomes of fetal surgery for MMC repair were first analyzed by a randomized clinical trial in the Management of Myelomeningocele Study (MOMS) trial[1]. This study showed improved neurologic outcomes with fetal repair performed between estimated gestational age (eGA) of 19 0/7 to 25 6/7 weeks[1].

We present a case of a female neonate born at 26 6/7 weeks eGA with MMC that was repaired at the bedside in the NICU while the patient was on an HFOV. There is essentially no data on MMC repair being successfully completed on extremely premature infants (EPI) (<30 weeks eGA) or babies of extremely low birth weight (ELBW) (<1.0 kg). There is also no data on the safety of performing such procedures at the bedside. Our case report will show that post-natal repair surgery can be successfully done in this age group and that there may be some long-term benefits of repair, such as decreased need for CSF diversion.

2. PATIENT CASE PRESENTATION:

A preterm female neonate was born at an outlying hospital at 26 6/7 weeks eGA by spontaneous vaginal delivery to a 22-year-old G1P0 mother due to preterm labor and premature rupture of membranes. Their birth weight was 810 grams. The patient was intubated and placed on mechanical ventilation. She was started on Ampicillin and Gentamicin due to preterm labor.

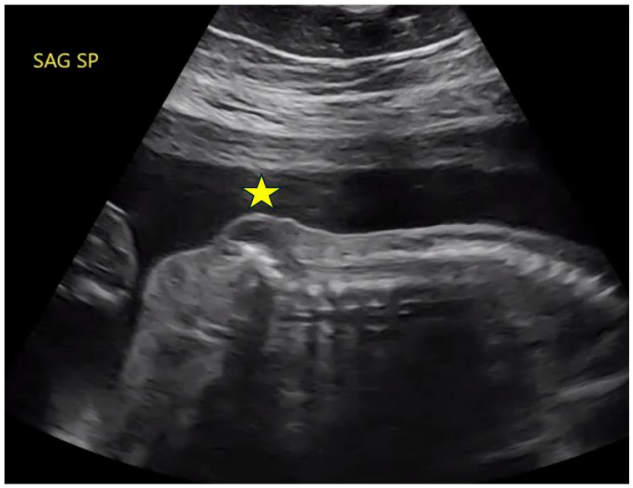

Figure 1: Transabdominal ultrasound at 24 3/7 weeks EGA showing sagittal view of thoracic and lumbar spine with an MMC noted at the caudal aspect of the image (yellow star).

The pregnancy was uncomplicated aside from inadequately treated maternal syphilis. Prenatal ultrasound at 22 weeks eGA first showed the sacral MMC measuring approximately 2.0 x 1.4 x 1.4 cm in the sacral region (Figure 1). Concern for an absent corpus callosum was also noted (not shown). The head circumference was less than 2-3 standard deviations below the mean for gestational age. Genetic testing via amniocentesis showed a 46XX karyotype with a 1Q21 microdeletion. The patient was referred to Children's Hospital of Philadelphia for possible fetal MMC repair but was unable to make the appointment due to transportation and financial issues.

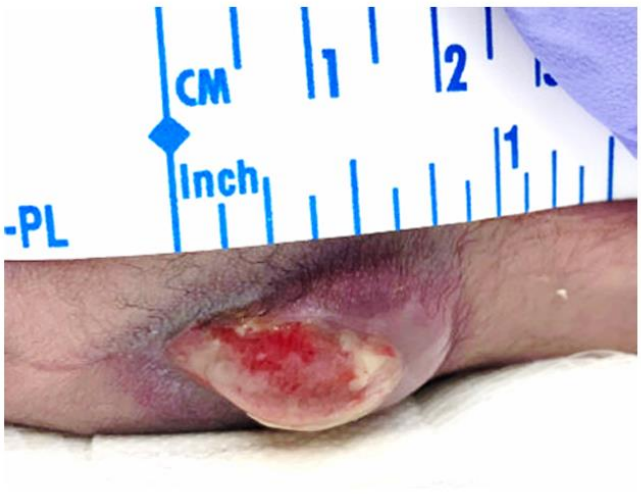

Figure 2: Photograph of open lumbosacral myelomeningocele defect measuring 2 x 1 cm present at birth. Exposed neural placode in the center with surrounding dysplastic skin and increased hair surrounding defect.

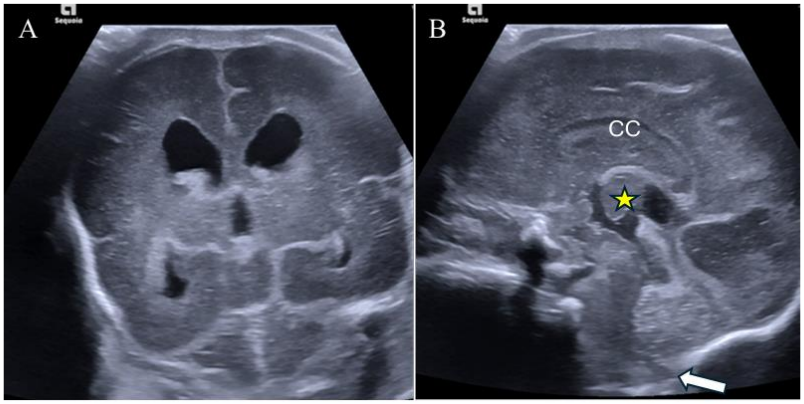

Figure 3: Admission head ultrasound A) coronal view showing mild-moderate ventriculomegaly and B) midline sagittal view showing the presence of corpus callosum (CC), interthalamic adhesion (yellow star), and small posterior fossa with hindbrain herniation consistent with Chiari II malformation (white arrow).

After transfer to our Neonatal Intensive Care Unit (NICU), the patient was transitioned to a HFOV. The spinal MMC defect measuring approximately 2 cm x 1 cm was noted on the low lumbar/upper sacral region (Figure 2). Initial neurological exam showed absent bilateral plantar flexion, consistent with an L5/S1 functional level. The patient had a head ultrasound (HUS) on admission that showed mild-to-moderate supratentorial ventriculomegaly (Figure 3A), a present corpus callosum (Figure 3B), and a Chiari II malformation (Figure 3B).

The patient's MMC repair was performed on day of life (DOL) 3 at the bedside in the NICU. The OR equipment was brought to the patient's bedside. The operating microscope was not used due to space constraints. The surgery was completed with a standard MMC closure technique. The placode was imbricated with a simple interrupted 7-0 Prolene (polypropylene blue monofilament, Ethicon) along the pial edges to reform the neural tube. The dura was identified, bluntly dissected off the surrounding tissues, but insufficient in length to meet in the midline. A Durepair patch (suturable dural collagen substitute, Medtronic) was sutured to the dural leaflets with 6-0 Prolene (polypropylene blue monofilament, Ethicon), to entirely enclose the placode by the dural patch. The skin was then closed primarily over the defect in a multilayer fashion. The estimated blood loss for the surgery was 5 ml out of an estimated 78 ml total blood volume for this patient; approximately 6% of the total blood volume[4]. The total operative time was 104 minutes and the patient remained hemodynamically stable throughout.

Following MMC repair, a repeat HUS on DOL 7 showed slightly increased ventriculomegaly compared to prior. Serial weekly HUS revealed essentially stable ventriculomegaly. Daily head circumference measurements tracked along the third percentile line using the Fenton Premature Girls head circumference chart[5]. She did not show any other clinical signs of elevated intracranial pressure, evidence of a CSF leak from the repair site, or worsened neurological lower extremity motor function during her hospital stay.

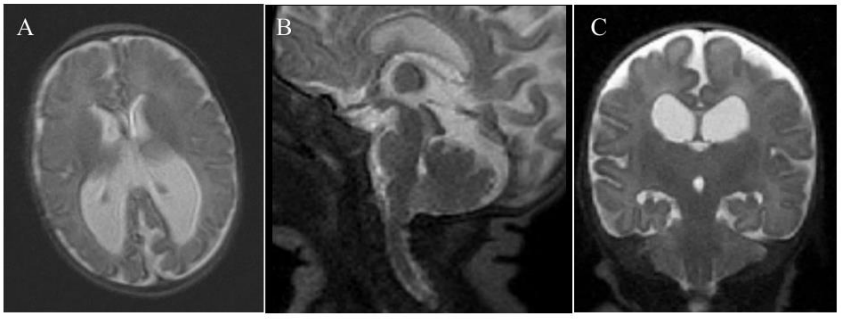

Figure 4: MRI at term-corrected 41 weeks A) axial T2 showing presence of colpocephaly in the occipital horns bilaterally B) mid-sagittal T2 showing tectal beaking, minimal hindbrain herniation C) coronal T2 showing ventriculomegaly stable compared to prior head ultrasounds, preserved extra-axial spaces and minimal cerebellar tonsillar descent.

A term-corrected (40-week) brain MRI without contrast was performed prior to discharge and showed ventriculomegaly

stable compared to prior HUS with minimal hindbrain herniation (Figure 4). We were unable to compare the degree of hindbrain herniation directly as she was unable to get an MRI prior to the operation due to respiratory status. She was successfully discharged at DOL 99 (41 weeks corrected) on room air and full oral feedings.

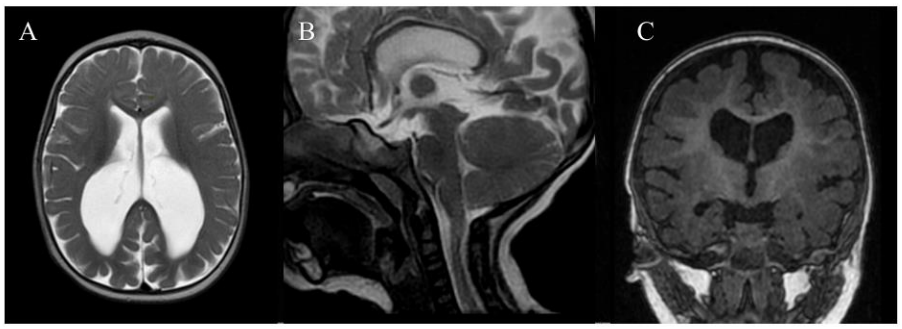

Figure 5: MRI imaging at 12 months of life showing A) Axial T2 showing colpocephaly of occipital horns of lateral ventricles without evidence of hydrocephalus B) mid-sagittal T2 showing improved hindbrain herniation with minimal Chiari malformation C) coronal T1 showing ventriculomegaly stable compared to prior MRI, with preserved extra-axial spaces.

After discharge, Neurosurgery follow-up showed improving leg strength and fine motor skills with her feet. A brain MRI without contrast (Figure 5) was performed at 12 months of age (9 months corrected eGA) and showed stable ventricular size with no evidence of hydrocephalus compared to imaging at 3 months of age. Her head circumference was tracking along the 3rd percentile growth curve (when corrected for prematurity) on the CDC girls head circumference[6].

Follow up at NICU developmental clinic at 6 months corrected eGA showed an INFANIB score[7] of 73, denoting age-appropriate development in all domains tested. At 18 months corrected eGA, a Bayley Scales of Infant and Toddler Development (IV edition) test showed fine and gross motor, cognitive, socio-emotional, and communication delays, indicating an overall developmental delay of 50.9% when compared to other children of the same adjusted age[8].

3. DISCUSSION:

We presented a case report of a 26 6/7 weeks eGA neonate who underwent post-natal repair of a MMC. The MMC defect was repaired at the bedside in the NICU while the patient was on an oscillator ventilator. To our knowledge, no data exists on post-natal MMC repair being successfully completed on EPI (<30 weeks eGA) or ELBW (<1.0 kg) neonates or on performing such procedures at the bedside in the NICU.

Initial studies looking into the feasibility of prenatal / fetal repair of MMC used postnatal repair as the control group for comparison of outcomes[9,10]. However, none of these postnatal repair patients were ever born prior to 30 weeks eGA. The MOMS trial postnatal cohort did not include any patients at <30 weeks eGA or <2000g[1]. Our patient was born at 26 6/7 weeks and has still seen a benefit of repair despite an eGA later than what is considered inclusion criteria for fetal surgery.

Although there are some neurosurgical procedures commonly done at the bedside in adults, this is often limited to emergency situations, such as placement of external ventricular drains or ICP monitors in an acute setting. There is also limited literature describing bedside temporary CSF diversion procedures, such as ventricular reservoirs[11], ventriculosubgaleal shunts[12] and external ventricular drains[13] in pediatrics. There is no literature that describes more complex pediatric neurosurgical operations completed at the bedside in the NICU.

Our case report aims to show that surgery can be successfully done in this age group. Fetal or EPI surgery is complicated by the fragility and high water content of tissues[14]. In our patient, the placode was imbricated with 7-0 prolene and the dura was closed with 6-0 prolene successfully without tears. The surgery was completed in 104 minutes with only 5 mL of estimated blood loss, maintaining hemodynamic stability throughout the surgery.

Although fetal surgery for myelomeningocele is an effective established treatment that shows improved functional outcomes and less risk of requiring ventricular shunt placement, socioeconomic and racial disparities exist in families who receive care[15]. Our patient's mother was affected based on socioeconomic status (SES). She was unable to financially afford to take time off work, as well as obtain travel to a fetal surgery center within the appropriate gestational age window for MMC repair. Extending surgical inclusion criteria to up to one week later (such as seen in our patient) may not only allow a wider number of patients to benefit from intervention, but also help alleviate the disproportionate burden of low SES.

There are several limitations to note. This paper analyzes a single case report; the results may not be generalizable to a broader population. Although the initial data from this case is promising, there needs to be long-term data to assess neurologic and developmental progress. In the future, additional cohort studies/trials should examine expanding the inclusion criteria

for fetal MMC repair determined by the MOMS trial.

4. CONCLUSIONS

We present a case of a female neonate born at 26 6/7 eGA with MMC. Due to socioeconomic factors, our patient was unable to access a fetal center capable of prenatal MMC repair. Thus, surgery was successfully completed postnatally at the bedside in the NICU. Our patient is currently over 22 months old, and to date has not required CSF diversion. Our case indicates that post-natal MMC repair, specifically <30 eGA, can be successfully done in EPI and ELBW neonates. It also shows that there may be some long-term benefits of repair in this age group, despite being a post-natal repair, such as decreased need for CSF diversion.

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