Case Report

Fetoscopic Repair of Meningomyelocele

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BACKGROUND: Currently, maternal-fetal surgery for repair of myelomeningocele requires an upper-segment hysterotomy, which likely increases maternal postsurgical risks. If fetoscopic repair of myelomeningocele achieves similar or better fetal outcomes while decreasing maternal risks, it would be a better option.

CASE: A patient with a fetus with a L3–S1 meningomyelocele underwent a laparotomy and fetoscopic repair using a two-port, in-CO_2 approach at 23 2/7 weeks of gestation. The neonate was delivered at 30 6/7 weeks of gestation by lower segment cesarean delivery and required no further surgery, has not needed a shunt (5 months), and has normal, age-appropriate neurologic function.

CONCLUSION: This innovative fetoscopic approach may offer an alternative to open fetal surgery and may prevent the need for hysterotomy and cesarean delivery in index and subsequent pregnancies.

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Teaching Points

1. Fetoscopy in a CO_2 environment allows improved visualization, access to tissue, and a wider field of view compared with standard in-amniotic fluid fetoscopy.
2. A simplified fetoscopic neurosurgical closure for meningomyelocele can be performed using a two-port approach and a self-locking suture.

Open fetal surgery for meningomyelocele has become a commonly offered option to postnatal repair since the publication of the Management of Myelomeningocele Study.[1] The Management of Myelomeningocele Study primarily showed a reduction in the need for shunt placement in survivors during the first year after fetal surgery compared with postnatal repair. The maternal risks of hysterotomy, which include uterine rupture and dehiscence postfetal repair[1] as well as uterine rupture (20%)[2] and placenta accreta[3] in future pregnancies, must be considered. We report a case of in utero meningomyelocele repair using a novel fetoscopic approach. The infant has survived and thrived, did not require postnatal surgical revision of the repair, and has not required treatment for hydrocephalus in the first 5 months of life.

CASE

A 32-year-old woman, gravida 2 para 1, was referred to the Texas Children's Fetal Center carrying a fetus with a meningomyelocele (L3–S1) at 22 weeks of gestation. She satisfied all Management of Myelomeningocele Study criteria for open fetal surgical repair.[1] The upper border of the lesion itself was well within the T1–S1 range, and the fetus had evidence of hindbrain herniation on ultrasonography and magnetic resonance imaging (MRI).[1] Lateral ventricular diameter was 14 mm. After first requesting and then agreeing to open surgery, the patient was offered the option of an experimental fetoscopic repair under a Baylor College of Medicine institutional review board–approved protocol. She gave written informed consent.

At 23 2/7 weeks of gestation, the patient underwent laparotomy under general anesthesia, and, because of an anterior placenta, the uterus was exteriorized to allow access to an area of the uterine wall more than 5 cm from the placental edge. The fetus was positioned for surgery and then given an injection of analgesia (fentanyl [5 micrograms/kg]) and a paralytic (vecuronium [400 micrograms/kg]). After this, the first of two 12-French (4 mm) vascular cannulae was inserted into the uterus (Seldinger technique under ultrasound guidance). A 4/0 Monocryl full-thickness stay suture on either side of the port fixed the membranes to the uterine wall. A pediatric cystourethroscope (3.2 mm

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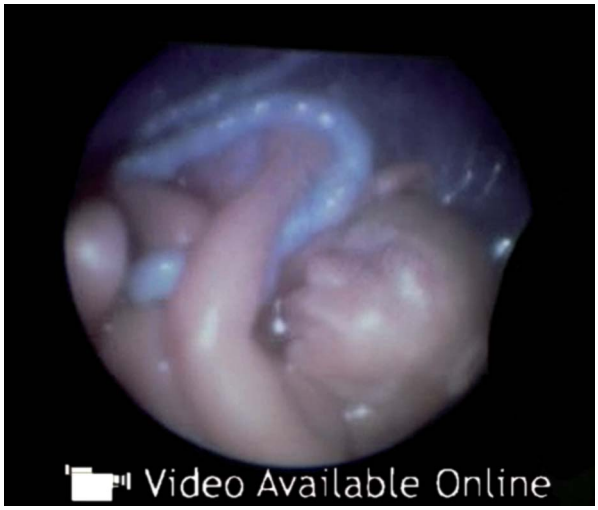

Video 1. This video shows images of a 2 port, in-CO2 fetoscopic repair performed on a human fetus with a L3 to S1 meningomyelocele. Video courtesy of Mr. Wally Crow, Texas Children's Hospital. Used with permission.

diameter, 2-mm operating channel) was placed through one port to visualize the lesion, which appeared closable (see the Video online at http://links.lww.com/AOG/A633). Approximately 60 cc of amniotic fluid was removed and replaced with CO_2 gas (0.5 L/min, maximum pressure 12 mmHg). A 1-mm Karl Storz grasper (1151OC) placed through the cystourethroscope was used to stabilize the meninges, and 3-mm Storz scissors (30210MSS), placed through a second 12-French port, were used to dissect the placode (open spinal cord forming a flattened plate with the spinal nerves exiting from the lower surface; Fig. 1A) free of the meninges without undermining the skin. The arachnoid was completely divided between the placode and junctional zone. Once the placode had retracted into the defect, a Storz laparoscopic needle driver (302200FNS) and Storz grasper were used to oppose the junctional zone edges (including skin and dural layers) with a V-Loc 4-0 barbed suture (Fig. 1) in a running stitch placed 4–5 mm from the skin edge. Two interrupted 5-0 Monocryl sutures were placed to buttress the repair. The fetus was monitored throughout the procedure by a pediatric cardiologist, and heart rate and cardiac function remained normal. There were no signs of maternal hypercarbia (no elevated end-tidal CO_2) and no evidence of fetal acidosis (no heart rate decelerations or bradycardia). At the completion of the surgery, the gas was slowly removed and replaced with warmed saline, the ports were removed, the punctures were closed with 2-0 Vicryl sutures, the uterus was returned to the abdomen, and the patient's abdomen was closed. Total surgery time was approximately 4 hours. The patient's postoperative recovery was uneventful, and, according to our institutional review board's protocol, because we are monitoring patients undergoing fetoscopic repair as we do patients undergoing open repair, she was discharged on postoperative day 5 to the Ronald MacDonald House (a housing facility in the Texas Medical Center) for outpatient follow-up.

Weekly ultrasonograms revealed decreased hindbrain herniation within 4 weeks. At 29 2/7 weeks of gestation (6 weeks postoperatively), a fetal MRI (Fig. 1) confirmed reversal of the hindbrain herniation and ultrasonography and MRI showed retained functional neurologic level (S1 level as inferred from active plantar flexion of the feet) compared with the preoperative level.

At 30 weeks of gestation, the patient presented with preterm premature rupture of the membranes and was admitted for in-house management (steroids and magnesium sulfate for neuroprotection). At 30 6/7 weeks of gestation, she developed persistent fetal tachycardia (170 beats per minute) for 1 hour. Despite a lack of vaginal bleeding, occult abruption was considered likely given a normal maternal temperature and white count. The decision was made to deliver, and, because the fetus was in the breech presentation, a cesarean delivery was performed.

The neonate had Apgar scores of 4 and 7 at 1 and 5 minutes of life, and his cord blood gases were normal. There was no sign of abruption, and the cause of the fetal tachycardia was never clear. He had an uneventful neonatal intensive care unit course from the standpoint of his prematurity. A Steri-Strip was placed over a suspected pinhole defect with cerebrospinal fluid leak, and there was no further leakage and good skin apposition (Fig. 2). The neonate was discharged after a 4-week neonatal intensive care unit stay. He has a normal neurologic motor examination and an intact anal wink reflex (S2–4). He has mild ventriculomegaly (which was present preoperatively), but serial ultrasonograms show stable ventricular size. The head is growing at a normal rate, and the fontanelle remains sunken with apposed cranial sutures. At 5 months of age, the infant is being monitored by the Texas Children's Hospital Spina Bifida Clinic team (including neurology, neurosurgery, urology, orthopedic surgery, and physical therapy) and he has not required treatment for hydrocephalus nor has he needed revision of the surgical scar. He has age-appropriate respiration, swallowing, eye movements, and arm strength.

Scan this image to view Video 1 on your smartphone.

DISCUSSION

There have been three prior case series of fetoscopic meningomyelocele repair reported in the United

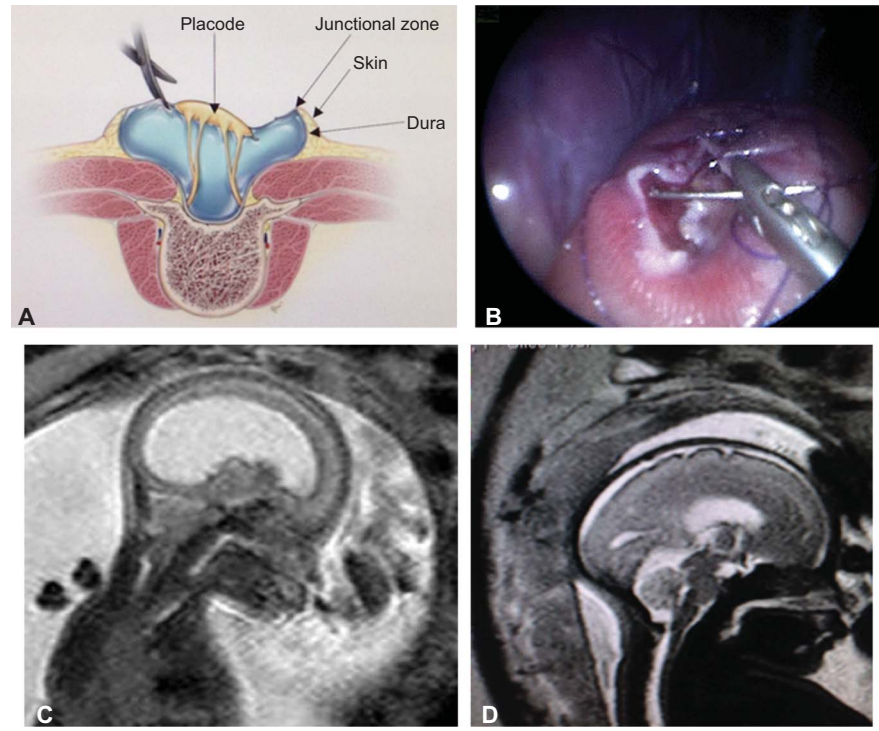

Fig. 1. This composite figure shows a drawing depicting the anatomy of a myelomeningocele (**A**), a photograph showing the dissected lesion during the repair (**B**), and two magnetic resonance images showing the preoperative Arnold-Chiari malformation (**C**) and reversal of the Arnold-Chiari malformation at 29 2/7 weeks of gestation (**D**). In the surgical image, the placode is seen beneath the midshaft of the straightened needle. Illustration by Kathy Relyea, MS, CMI. Printed with permission from Baylor College of Medicine.

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States: four cases by the same group, including two cases reported both in 1997[4] and in 1999[5] and three cases reported in 2003.[6] The results were not encouraging. Four of the seven neonates died and all three surviving neonates needed both revision of the meningomyelocele repair and ventriculoperitoneal shunting within the first 6 weeks of life. After these publications, fetoscopic repair was abandoned in the United States. Two other groups (Pedreira et al[7] in Brazil and Kohl et al[8] in Germany) have recently published results on fetoscopic repairs using percutaneous CO_2 gas insufflation with different closure techniques (significant dissection, cellulose, collagen and Teflon patches, and sutures). These groups use a three- or four-port percutaneous technique with various sized (11–16 French or 5 mm) ports. Although preterm premature rupture of membranes occurred within 2–7 weeks of the surgery in most cases, almost all had delayed delivery until beyond 30 weeks of gestation, and in the case of Kohl et al,[8] 49% of their 51 neonates delivered at or beyond 34 weeks of gestation. Shunt frequency in Kohl et al's patients[8] is not currently known, whereas two of the four patients (50%) of Pedreira et al[7] required ventriculoperitoneal shunting for hydrocephalus. These results have raised concerns about clinical use.[9]

Our approach was developed using a planned stepwise program involving animal[10] and simulation training. Despite initial enthusiasm for a "patch and glue" repair in fetal sheep,[10] we elected to perform a full surgical repair in our human trial. We developed a unified single-layer surgical closure using

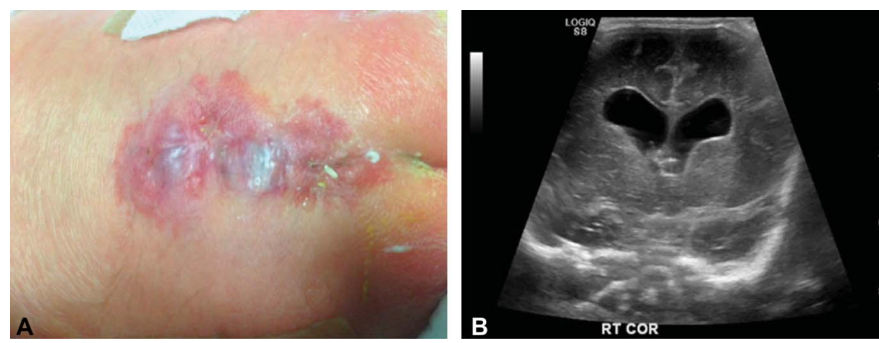

Fig. 2. This composite figure shows the healed scar on the neonate's back a few days after birth (**A**) and an ultrasonogram of his brain at his 3-month visit (**B**).

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a self-anchoring suture and performed it successfully on three open cases with excellent closure and no need for postnatal revision (oral presentation by Dr. W. Whitehead at the American Association of Neurological Surgeons meeting, Amelia Island, December 5, 2014). We developed a simulator and practiced our technique extensively (to be published separately). After we successfully performed our technique in the fetal sheep model, we obtained institutional review board approval for this human study, and this patient is the first enrollee. Information on the trial can be found at http://clinicaltrials.gov/ct2/show/NCT02230072.

This report suggests that fetoscopic meningomyelocele repair without fetal or neonatal adverse outcome (except preterm delivery), or the need for revision or shunting in the first weeks of life, is possible. The long-term neurologic outcome remains to be seen, but at 5 months postdelivery, our patient has normal bowel, bladder, and motor–sensory neurologic function. The issue of cord tethering, higher in fetal than in postnatally repaired cases,[11] will require long-term follow-up. Our technique, by reducing tissue trauma and by apposing dural tissue over the placode, may potentially decrease tethering. Although the gestational age at delivery in this case was less than that seen with open fetal surgery,[1] the outcome of the neonate is currently comparable with that seen after open surgery, and the mother has the option of a vaginal delivery in her next pregnancy. Further research on membrane preservation is required to reduce preterm premature rupture of membranes, which appears to occur within 6–7 weeks of most fetoscopic procedures.

Our case differs from reported fetoscopic cases in that we use a two-port technique in CO_2 with an exteriorized uterus and use a neurosurgical closure that does not require dissection, multiple-layer closure, or the use of patches or glues. We appose the skin edges using a barbed suture that does not require knots but which prevents cerebrospinal fluid leak, exposure of the neural elements to amniotic fluid, and protects the placode from trauma. If needed, the procedure can be converted to an open technique by opening the uterus between the two port sites. Although data are few, CO_2 in the uterus has not appeared to cause measurable harm to the fetus in animal[10,12] or human studies.[8] In our case, there was no evidence of either maternal or fetal acidosis based on maternal end-tidal CO_2 and fetal cardiac monitoring. Although our patient delivered by cesarean (preterm breech presentation), she is capable of having a vaginal delivery in her next pregnancy. If a neonate in our trial is in a cephalic presentation at term, and imaging suggests a successful closure, a vaginal delivery is an option. We feel that if there is a way to safely and appropriately treat fetal meningomyelocele without subjecting the mother to surgery that permanently and significantly scars her uterus, as a discipline, we should consider further exploring this line of research.

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