Fetal Myelomeningocele Repair

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Lily now has a better chance to walk thanks to a new treatment being offered in the Midwest for the treatment of Myelomeningocele (MMC). Lily was diagnosed at 19 weeks gestation with MMC during her normal anatomy screen. At 25 weeks gestation, her mother underwent open fetal surgery to repair her MMC. She is now 4 months old, a thriving baby, kicking her legs and shunt free.

In February, 2011, the future of unborn babies diagnosed with MMC was changed forever when the results of the MOMS (Management of Myelomeningocele Study) were released. Just two months after the release of these results, the St. Louis Fetal Care Institute team from Cardinal Glennon Children’s Medical Center performed the first fetal MMC repair in the Midwest at SSM St. Mary’s Health Center.

MMC, a severe form of spina bifida, is an open neural tube defect where the fetus’ spinal cord fails to close during development. This normally happens between 20 and 28 days of gestation, but in MMC the meninges and vertebral arches do not close around the spinal cord, leaving the delicate neural tissue exposed to injury (Liptak, G. & Dosa, N., 2010).

The effects of MMC impact the nervous system, muscles and bones, bowel, and bladder, leading to lifelong disability. Because nerve damage at the MMC prevents function below that level, the higher the spina bifida level, the greater the impact on normal development and function. It is thought that in the early stages of MMC, the spinal cord remains normal. However, as the pregnancy progresses, prolonged intrauterine exposure of the open spinal cord tissue to amniotic fluid and trauma leads to progressive damage (Danzer, E. & Adzick, S., 2011). There is also loss of the cerebral spinal fluid. In cases of MMC above the sacral level, another defect, the Arnold Chiari II malformation, is usually associated: the brainstem and part of the cerebellum are displaced downward through the foramen magnum into the upper cervical spinal canal. When this occurs, the normal flow of cerebrospinal fluid out of the brain is obstructed, causing hydrocephalus, an excess of fluid within the brain.

Hydrocephalus occurs in 60% to 95% of children with MMC (Liptak, G. & Dosa, N., 2010). After birth, 80% or more children with hydrocephalus need to have the extra fluid shunted out of the brain into the abdomen via a ventriculoperitoneal shunt. Myelomeningocele affects about 3.4 per 10,000 live births in the United States, and it ranges in severity. Infants born with MMC have a death rate of approximately 10%. Some children, with mild cases very low on the spinal cord, can function nearly normally. In more severe cases, damage to the spinal cord and peripheral nerves is irreversible, even with surgical closure after birth (Adzick, S., et al, 2011).

However, with the results of the MOMS trial, it is now known that repairing the MMC prenatally between 19 weeks and 26 weeks can improve the baby’s outcome as compared to repair after birth. The results of the trial found prenatal treatment helps reduce, or even eliminate, the major complications of Myelomeningocele—the hydrocephalus, the Chiari malformation, and the lack of movement in the lower extremities. The MOMS trial showed that children with prenatal MMC repair are half as likely to need a VP shunt, often have reversal of the Arnold-Chiari malformation, and are more likely to walk, at least until 30 months. Long term follow up data of children treated with prenatal surgery is still being collected, so the benefit beyond 30 months is not fully known (Adzick, S., et al, 2011).

Prenatal MMC repair requires open fetal surgery. Because this is considered to be major surgery to a pregnant mother, maternal safety is of utmost concern. A mother must meet strict inclusion and exclusion criteria to be considered a candidate for open fetal surgery. Inclusion criteria include:

- Maternal age greater or equal to 18 years of age
- Gestational age between 19 weeks and 25 weeks 6 days
- Normal fetal karyotype
- MMC between T1 and S1
- Confirmed Arnold-Chiari II malformation on prenatal US and MRI

Exclusion criteria include:

- Multiple gestation pregnancy
- A fetal anomaly unrelated to MMC

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At 25 weeks gestation, Lily underwent open fetal surgery to repair the myelomeningocele.

- Severe fetal kyphosis > 30 degrees
- History of maternal alloimmunization problem or fetal Rh isomunization
- History of Kell sensitization
- History of neonatal alloimmune thrombocytopenia
- Risk of preterm birth (history of spontaneous preterm birth, cervix <20mm, presence of cerclage)
- History of a placental problem (history of abruption or placenta previa)
- Body-Mass Index of 35 or more
- Maternal contraindication to surgery (Insulin-dependent pregestational diabetes, hypertension or preeclampsia, HIV, hepatitis-B, or hepatitis-C positive, or other medical condition of risk to mother)
- Uterine problem (previous hysterotomy in the active uterine segment, severe fibroids, malformation such as bicornuate uterus)
- Psychosocial problem (No support person for periprocedural period, inadequate support at home for pregnancy, inadequate understanding of risks and benefits of fetal surgery, inability to comply with medical restrictions or follow up after fetal surgery)

Fetal evaluation and care coordination for a mother interested in open fetal repair of MMC requires the collaboration of many different specialties, including maternal fetal medicine, pediatric surgery, pediatric neurosurgery, neonatology, anesthesia, social services, cardiology, radiology, genetics, and nursing. Fetal evaluation includes Level II ultrasound, fetal echocardiography, fetal MRI, and amniocentesis for chromosome analysis. Extensive counseling and psychosocial evaluation are necessary to assure that a mother understands the risks versus benefits to open fetal surgery. At the St. Louis Fetal Care Institute, this collaboration is a partnership of St. Louis University and SSM St. Mary's Health Center, and the Co-Director of the St. Louis Fetal Care Institute.

The operation for open fetal surgery is done under general anesthesia. Uterine relaxation achieved through general anesthesia is essential to allow for exposure of the appropriate fetal part, to prevent placental separation, and to avoid preterm labor (Danzer, E., & Adzick, S., 2011). At the St. Louis Fetal Care Institute, mothers are given antibiotics intravenously and intrauterine to prevent infection. Indomethacin is given prophylactically to decrease the risk of preterm labor.

At the start of an open fetal MMC repair, the pregnant uterus is typically exposed through a low transverse incision. A hysterotomy site is chosen using sterile ultrasound, which must be at least 6 cm away from the placental edge. Stay sutures are placed through the uterine wall and an initial incision is made using electrocautery. An absorbable uterine stapling device is then inserted into the uterus, which opens the uterus while sealing the membranes to the myometrium on either side of the incision. The stapler creates a 6 to 7 cm uterine incision large enough to expose the fetal MMC. Warmed saline is infused into the uterine cavity, which helps prevent placental separation, fetal expulsion, and uterine contractions.

During open fetal MMC repair, continuous fetal ultrasonography is performed to monitor fetal well being. The fetus is also given intramuscular fentanyl, vecuronium, and atropine prior to repair. The fetal repair of the MMC is done by the pediatric neurosurgeon. Once the repair is completed, the hysterotomy site is closed by the fetal surgeons.

At the St. Louis Fetal Care Institute, mothers are given an epidural to help with pain control after surgery. They are also started on tocolytics to prevent preterm labor. Oral tocolytics are continued for the remainder of the pregnancy. Post operative hospitalization typically lasts four days. Mothers are required to stay locally in the St. Louis area for about two weeks to be monitored for post-operative complications. After this, the mothers can return home to the care of their primary prenatal care provider. Weekly ultrasound follow up is essential to monitor for preterm labor, premature rupture of membranes, and fetal well-being. Close follow up is essential by the care providers. Because of the weakness caused by the surgery on the uterus, a cesarean section is required for the birth of the baby and all future babies. Cesarean section is performed at 37 weeks gestation.

As with any prenatal surgery, there are risks to both the mother and the baby. The potential benefits must be balanced against the risks of open fetal surgery, and this decision is unique to every mother and her family. For Lily and her family, fetal surgery was the right decision.

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References


Children under the age of 2 months have the greatest risk of hospitalization and death related to pertussis infection, due to the fact that they are too young to be vaccinated. Since 2005, the Advisory Committee on Immunization Practices (ACIP) has recommended Tdap booster vaccines to unvaccinated postpartum patients and other family members of newborn infants to protect the infants from pertussis, a strategy called “cocooning.” Over this time, these programs have been difficult to implement widely among family members and mothers. In June 2011, the ACIP made recommendations for Tdap use in pregnant patients.

The safety information for the vaccines was established through pregnancy registries that concluded that the vaccine did not cause any increase in frequency of adverse events or teratogenicity. The ACIP concluded that administration after 20 weeks is preferred to minimize any potential risk for harmful effects to the fetus. The administration of the vaccination during pregnancy provides the infant with transplacental antibodies that may provide protection for the infant in early life prior to their first immunization series. To provide the best protection for the baby, the mom should receive Tdap in the third trimester. By receiving the vaccine, the pregnant mom will be less likely to transmit pertussis to her newborn infant.

There has been fear that the maternal antibodies may interfere with the infant’s immune response to their first DTaP vaccination. This response is referred to as blunting and it is believed to be short-lived as the maternal antibodies decline rapidly. There are currently clinical trials looking at the immune response of infants at 2, 4, and 6 months of age in moms who received the Tdap vaccination. The interim data suggest the potential benefit of protection from the maternal antibodies in newborn infants outweighs the potential risk of shifting the disease burden to later in infancy.

There are two Tdap vaccines available in the United States, Adacel, licensed for use in patients 11-64 years and Boostrix, for use in patients greater than 10 years.

<table>
<thead>
<tr>
<th>Vaccine</th>
<th>Before pregnancy</th>
<th>During pregnancy</th>
<th>After pregnancy</th>
<th>Type of Vaccine</th>
<th>Route</th>
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</thead>
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<tr>
<td>Hepatitis A</td>
<td>Yes, if at risk</td>
<td>Yes, if at risk</td>
<td>Yes, if at risk</td>
<td>Inactivated</td>
<td>IM</td>
</tr>
<tr>
<td>Hepatitis B</td>
<td>Yes, if at risk</td>
<td>Yes, if at risk</td>
<td>Yes, if at risk</td>
<td>Inactivated</td>
<td>IM</td>
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<tr>
<td>Human Papillomavirus (HPV)</td>
<td>Yes, if 9 through 26 years of age</td>
<td>No, under study</td>
<td>Yes, if 9 through 26 years of age</td>
<td>Inactivated</td>
<td>IM</td>
</tr>
<tr>
<td>Influenza TIV</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Inactivated</td>
<td>IM, ID (18-64 years)</td>
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<tr>
<td>Influenza LAIV</td>
<td>Yes, if less than 50 years of age and healthy; avoid conception for 4 weeks</td>
<td>No</td>
<td>Yes, if less than 50 years of age and healthy; avoid conception for 4 weeks</td>
<td>Live</td>
<td>Nasal spray</td>
</tr>
<tr>
<td>MMR</td>
<td>Yes, avoid conception for 4 weeks</td>
<td>No</td>
<td>Yes, give immediately postpartum if susceptible to rubella</td>
<td>Live</td>
<td>SC</td>
</tr>
<tr>
<td>Meningococcal: polysaccharide conjugate</td>
<td>If indicated</td>
<td>If indicated</td>
<td>If indicated</td>
<td>Inactivated</td>
<td>SC IM</td>
</tr>
<tr>
<td>Pneumococcal Polysaccharide</td>
<td>If indicated</td>
<td>If indicated</td>
<td>If indicated</td>
<td>Inactivated</td>
<td>IM or SC</td>
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<td>Tetanus/Diphtheria Td</td>
<td>Yes, Tdap preferred</td>
<td>Yes, Tdap preferred if 20 weeks gestational age or more</td>
<td>Yes, Tdap preferred</td>
<td>Toxoid</td>
<td>IM</td>
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<td>Tdap, one dose only</td>
<td>Yes, preferred</td>
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<td>Yes, preferred</td>
<td>Toxoid/ inactivated</td>
<td>IM</td>
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<tr>
<td>Varicella</td>
<td>Yes, avoid conception for 4 weeks</td>
<td>No</td>
<td>Yes, give immediately postpartum if susceptible to varicella</td>
<td>Live</td>
<td>SC</td>
</tr>
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</table>

**Recommendations:**

- **Maternal vaccination:** During the third trimester (or at least 20 weeks gestation) or postpartum.
- **Cocooning:** Those adults or adolescents (siblings, caregivers, etc.) who will have close contact with a newborn should get Tdap at least 2 weeks before having close contact with the infant.
- **Pregnant women due for tetanus booster:** If greater than 10 years have passed since the pregnant woman’s Td booster, then she should receive Tdap during the third trimester.
- **Wound management:** If 5 years or more have elapsed since the last Td, the pregnant woman should get Tdap (unless she received it previously).
- **Pregnant women with incomplete or never receiving tetanus vaccination:** The pregnant woman should receive the normal 3 vaccinations of Td, preferably starting in the 2nd trimester, unless high risk. The first two doses should be 4 weeks apart, with the third dose 6-12 months after the first dose. Tdap should replace one of these doses after 20 weeks gestation.

10/2011 CDC
The Monitor Corner
A CASE STUDY
Sharon Rector, RNC, MSN

Presentation
Ms. T is a 32 year old, G2P0101 who presented to labor and delivery at 37 weeks gestation for induction of labor due to poorly controlled Type 1 Diabetes Mellitus. Her HbA1c test two months prior was 8.3%. Attempts were made to control her blood sugar through nutrition counseling and insulin management. Her first pregnancy was complicated by preterm labor at 32 weeks gestation that resulted in a cesarean section for breech presentation.

Background
Ms. T’s cervical exam on admission was 1 cm, 50% effaced, -3 station, moderate consistency and midposition resulting in a Bishop score of 4. The low Bishop score suggested a need for cervical ripening. Laminaria were inserted, and approximately 9 hours later they were removed and an oxytocin infusion was started. The oxytocin infused for approximately 12 hours with a maximum dose of 7 mu/min. The oxytocin was discontinued once due to fetal heart rate changes. It was restarted one hour later and increased as indicated. The patient received an epidural for pain management. After approximately 11 hours with oxytocin infusing the fetal heart rate tracing revealed a baseline rate of 125 bpm with moderate variability, occasional accelerations and no decelerations. Thirty minutes later the following tracing was observed. How would you interpret the tracing? What interventions are indicated? What do you think happened?

Outcome
At the time of this tracing Ms. T was comfortable with her epidural. Her vital signs included BP 100/60, HR 68, R 16, oxygen saturation 97%. As a result of the tracing, the oxytocin was discontinued and a vaginal exam was performed noting the cervix to be 4 cm dilated, 75% effaced and -3 station with a small amount of vaginal bleeding. The patient was repositioned on her side, oxygen was administered at 10L/min per non-rebreather face mask, and a bolus of intravenous fluids was initiated. The physician arrived at the bedside, and when the FHR dropped below 60 bpm the decision was made to perform an emergency cesarean section. During surgery, the infant’s head was observed in the patient’s abdomen as a result of a uterine rupture where the previous low transverse cesarean section had been performed. The baby boy was delivered and handed off to the pediatric team. Apgars were 2 and 7, and he weighed 2815 grams or 6 pounds 3 ounces. Venous cord blood gases included a pH of 6.85, CO2 148, and BE -11.8. Arterial cord blood gases included a pH of 6.81, CO2 152, and BE -14.3. The infant was observed in the neonatal intensive care unit for one day. He received oxygen for several hours and was easily weaned to room air. Both mom and baby had an otherwise normal postpartum course and were discharged on day three.

Strip 1

Strip 2

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Uterine Rupture

Uterine rupture is a rare but very serious obstetric complication. Factors associated with uterine rupture include grand multiparity, use of prostaglandin preparations or high dosages of oxytocin, uterine tachysystole, abdominal trauma and obstetric manipulations such as forceps delivery or external and internal version (Bowers et al., 2008). However, the greatest risk for uterine rupture occurs in women with a history of previous uterine surgery. Intrapartum rupture of a low transverse uterine scar occurs in 0.5% to 1% of women who undertake a trial of labor after a cesarean birth (Thorp, 2009). Women with a vertical, T or J shaped scar are at greater risk for uterine rupture.

The terms uterine rupture and uterine dehiscence are sometimes used interchangeably and are usually not differentiated in the research. Uterine dehiscence refers to a separation of a previous uterine scar, usually partial, with intact membranes and with the fetus remaining inside the uterus. Uterine rupture refers to the actual separation of the uterine myometrium or previous uterine scar, with rupture of the membranes and possible protrusion of the fetus or fetal parts into the peritoneal cavity (Bowers et al., 2008).

According to Bowers et al. (2008) the clinical presentation of uterine rupture depends on the specific type of rupture and may develop slowly over several hours or very quickly within several minutes. Impending rupture may be preceded by increasing uterine tone or uterine tachysystole. Changes in the fetal heart rate may be the most common early sign of uterine rupture. The fetal heart rate pattern may include decreased variability, recurrent variable, late or prolonged decelerations or fetal bradycardia. The woman may experience the sudden onset of sharp abdominal pain and tenderness, nausea and vomiting, syncope, vaginal bleeding, tachycardia or pallor. Bleeding can quickly lead to maternal hypotension, shock and cardiovascular collapse. When performing a vaginal exam the practitioner may be unable to reach the presenting part and the fetus may be palpated through the abdominal wall. Complete or partial placental abruption often occurs with uterine rupture. Dehiscence of a prior uterine scar is usually asymptomatic at first. The woman may continue to have contractions without cervical change. If the dehiscence extends past the scar tissue, the woman may begin to complain of unrelieved pain in the lower abdomen.

Treatment of uterine rupture includes hemodynamic stabilization and immediate cesarean birth. With prompt intervention maternal death is a rare complication. There is significant risk of brain damage and death for infants who were extruded into the maternal abdomen during the rupture. The key to a healthy outcome for mom and baby is prompt diagnosis and intervention.

References


Changes in the fetal heart rate may be the most common early sign of uterine rupture.
The Numbers Do Not Lie!
STATISTICS WOMEN CONSIDERING CESAREAN SECTION NEED TO KNOW
Suwan Mehra, M.D., and Gilad Gross, M.D.

The number of cesarean sections (CS) performed continues to increase in the United States. Most recent figures show that nearly 1 in 3 babies are now delivered surgically. CS delivery on demand remains uncommon in the U.S., but it too is on the rise. Repeat CS accounts for 40% of all cesareans done in the US. A history of multiple prior CS need not exclude a patient from the option of trial of labor. There is growing concern regarding an association between CS and long-term maternal morbidity as the rate of CS rises. Clinicians and patients need to be aware of the long-term risks associated with CS so that they can be considered when determining the best method of delivery for initial and subsequent births.

Evidence exists to suggest that the risk of morbidity increases as the number of CS increases. The dominant maternal risk is placenta accreta in which the placenta grows into the muscular or deeper layers of the uterus. This complication is highly associated with hemorrhage and the need for hysterectomy. Having multiple CS is the major risk factor for this severe complication. The risk of placenta accreta increases dramatically from 0.57% after three CS to 2.13% after four CS. The incidence of delivery-related hysterectomies is more than five times higher for women undergoing their fourth CS than for women having their second CS. Recent data shows that the chance of a woman requiring a hysterectomy is 0.4% after the second CS, compared with 2.4% following a fourth CS, and 9% after six or more CS. Women delivering by CS for the sixth time or more require blood transfusions 10 times as often when compared to women having their second CS. Subsequent pregnancies are also at an increased risk for other types of abnormal placental conditions, reduced fetal growth, preterm birth, and possibly stillbirth. Chronic maternal side effects associated with CS include pelvic pain and adhesions. Adverse reproductive effects may include decreased fertility and increased risk of spontaneous abortion and ectopic pregnancy.

In 2006, the vaginal birth after cesarean (VBAC) rate was 8.5% in the United States. As the number of prior CS a patient has varies, so does the rate of attempted VBAC. Those with one prior CS have a rate of 10.1%, compared with 3.2% for women with two prior CS, and 2.8% for women with three or more prior CS. The success rate of VBAC is 71% with two prior CS, 63% with three prior CS and 55% with four. Indications for CS that are considered non-recurring such as malpresentation and non-reassuring fetal status have higher success rates (60-87%). This rate is similar to primigravidas when compared to a CS done for labor related problems such as cephalopelvic disproportion and failure to make progress (50-60%). Additional factors associated with lower VBAC success rates include labor induction after 40 weeks and larger babies. An induced labor for VBAC is successful 68% of the time compared to women who VBAC spontaneously (78%). Mothers delivering newborns with birth weight greater than 4000 grams have a success rate of 58% compared to those greater than 4500 grams (43%). In addition maternal obesity is also associated with lower rates of success. Certain factors do not seem to influence the success of VBAC attempt, namely twin gestation and grand-multiparity (more than six or more prior deliveries).

The most serious complication for women undergoing a trial of labor after a prior CS remains uterine rupture. While rates of uterine rupture vary, the absolute risk ranges between 0.5 and 4 percent. Patient-specific factors cannot be used to accurately predict the relatively small proportion of women who

Most recent figures show that nearly 1 in 3 babies are now delivered surgically.

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will experience a uterine rupture during an attempted VBAC. There are several contra-indications to attempting a VBAC. These include prior classical, inverted-“T” or “J” shaped uterine incisions that involve the more muscular lining of the uterus. There is a high risk of uterine rupture associated with types of incisions. There are, however, patients in whom an unknown prior incision exists. Past studies show that success and complication rates are similar to patients whose past incision was amenable to VBAC, that being the low transverse incision. Unless clinical factors linked to the prior unknown scar could suggest a classical, “T”, or “J” incision, it is reasonable to offer VBAC. A prior uterine rupture of the lower uterine segment has a recurrence rate of 9% and of the contractile portion of the uterus 52%. More recent data suggests that, after accounting for labor duration, induction is not associated with an increased risk of uterine rupture in women attempting VBAC. Most consider a previous uterine rupture a contraindication to an attempted VBAC and encourage delivery by elective cesarean after confirming fetal lung maturity after 37 weeks. Also well accepted are the risks of uterine rupture with labor inducing agents containing prostaglandins. When attempting VBAC, these agents should be avoided.

VBAC is a safe and appropriate choice for most women with one prior cesarean and for some women with two prior cesareans according to the American Congress of Obstetricians and Gynecologists. Data regarding the risk for women undergoing VBAC with more than two previous cesarean deliveries are limited. Previous vaginal delivery and prior successful VBAC delivery are favorable factors in predicting the success of VBAC and are associated with low rates of uterine rupture. Factors that work against the safety and success of VBAC include multiple prior CS, short inter-pregnancy intervals, prior single layer uterine closures, prior preterm CS, labor induction and augmentation with prostaglandins. Unfortunately there is no accurate prediction tool for uterine rupture as well as VBAC success. The best approach remains a careful consideration of both historical and current pregnancy related factors. Each patient and her pregnancy require individual assessment of risk factors, plus an evaluation of risks versus benefits when choosing VBAC or CS.

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References
Additional references available upon request.
Letters
The Perinatal Times welcomes comments on any of its articles and will consider such letters for publication. Suggestions for future topics of interest or announcements are encouraged. Correspondence should be addressed to:

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