Fetal Surgery for Spina Bifida

Spina bifida (myelomeningocele) is an opening in the spinal column that results in impairment of the nervous system. While most children with spina bifida survive, many are left with major disabilities, including paralysis, bowel and bladder control difficulties, brain abnormalities (i.e., hydrocephalus and brain herniation, also known as Chiari II malformation) and learning disabilities. Spina bifida can be detected with routine testing and confirmed with ultrasound.

Traditionally, infants with spina bifida have surgery shortly after birth to repair the spinal cord defect. Any additional treatments are focused on the symptoms associated with spina bifida. For example, if an infant develops hydrocephalus, a ventriculoperitoneal (VP) shunt may be inserted to drain the fluid. Other symptoms, such as paralysis, require the care of medical specialists. Today, fetal surgery for spina bifida is considered an option for treatment of the symptoms in eligible candidates, but not a cure for the disease.

Results of the MOMS Trail
The Management of Myelomeningocele Study (MOMS) was a National Institutes of Health-sponsored multicenter clinical trial that evaluated the best treatment for spina bifida: fetal surgeries or surgical repair after birth. The study randomized 183 pregnancies (91 fetal surgeries and 92 postnatal surgery). The NIH stopped the trial at the fourth interim analysis (78 fetal surgeries and 80 postnatal surgeries) because there were significantly better outcomes in fetal surgery patients.

The primary outcome studied by the trial was perinatal death or need for a VP shunt at 12 months of age. Secondary outcomes included the Bayley Mental Developmental Index and difference between functional and anatomical level of lesion at 30 months of age.

Beneficial Results of the Trial

<table>
<thead>
<tr>
<th>PARAMETER</th>
<th>FETAL SURGERY</th>
<th>POSTNATAL REPAIR</th>
<th>P VALUE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary outcome</td>
<td>68%</td>
<td>98%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Placement of VP shunt</td>
<td>40%</td>
<td>82%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Any hindbrain herniation</td>
<td>64%</td>
<td>96%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Bayley Mental Index</td>
<td>89.7 ± 14.0</td>
<td>87.3 ± 18.4</td>
<td>0.53</td>
</tr>
<tr>
<td>Motor and anatomical levels</td>
<td>0.58 ± 1.94</td>
<td>-0.69 ± 1.99</td>
<td>0.001</td>
</tr>
<tr>
<td>Walking independently</td>
<td>42%</td>
<td>21%</td>
<td>0.01</td>
</tr>
</tbody>
</table>
Maternal and Neonatal Risks of the Trial

<table>
<thead>
<tr>
<th>PARAMETER</th>
<th>FETAL SURGERY</th>
<th>POSTNATAL REPAIR</th>
<th>P VALUE</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>MATERNAL</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chorion-amnion separation</td>
<td>26%</td>
<td>0%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Pulmonary edema</td>
<td>6%</td>
<td>0%</td>
<td>0.03</td>
</tr>
<tr>
<td>Oligohydramnios</td>
<td>21%</td>
<td>4%</td>
<td>0.001</td>
</tr>
<tr>
<td>Abruption</td>
<td>6%</td>
<td>0%</td>
<td>0.03</td>
</tr>
<tr>
<td>Spontaneous PROM</td>
<td>46%</td>
<td>8%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Status of hysterotomy incision</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intact</td>
<td>64%</td>
<td></td>
<td>–</td>
</tr>
<tr>
<td>Very thin</td>
<td>25%</td>
<td></td>
<td>–</td>
</tr>
<tr>
<td>Area of dehiscence</td>
<td>9%</td>
<td></td>
<td>–</td>
</tr>
<tr>
<td>Complete dehiscence</td>
<td>1%</td>
<td></td>
<td>–</td>
</tr>
<tr>
<td><strong>NEONATAL</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gestational age at delivery</td>
<td>34.1 ± 3.1</td>
<td>37.3 ± 1.1</td>
<td>0.003</td>
</tr>
<tr>
<td>Term delivery (&gt; 37 weeks)</td>
<td>21%</td>
<td>85%</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Birth weight</td>
<td>2383 ± 688</td>
<td>3039 ± 469</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>RDS</td>
<td>21%</td>
<td>6%</td>
<td>0.008</td>
</tr>
<tr>
<td>Neonatal/infant death</td>
<td>2*</td>
<td>2#</td>
<td></td>
</tr>
</tbody>
</table>

* - One fetal demise at 26 weeks EGA and one neonatal demise at 23 weeks EGA; both 5 days after fetal repair
# - Both deaths in first year of life due to Chiari malformation complications (both with VP shunt)

The valuable information from the MOMS study has enabled physicians to properly counsel patients about the risks and benefits of fetal surgery. A follow-up study is being conducted to evaluate the long-term results of fetal surgery for spina bifida.

**Evaluation for Surgery**

Physicians select candidates for surgery carefully based on, but not limited to, the following:

- Comprehensive ultrasound for other fetal anomalies
- Fetal echocardiography for heart structure and function
- Fetal brain and spine MRI for presence and severity of Chiari II malformation and other anomalies
- Extensive consultations with the multidisciplinary team

Candidates for surgery must meet the following criteria:

- The spina bifida defect must start between T1-S1.
- The fetus must have normal fetal chromosomes and no other anomalies.
- Gestational age at referral must be between 19 and 25 weeks.
Exclusion Criteria

FETAL
- Fetal anomaly not related to myelomeningocele
- Kyphosis in the fetus greater than 30 degrees
- Multigestational pregnancy

MATERNAL
- Maternal contraindications to surgery or anesthesia
- Morbid obesity (BMI of ≥ 35)
- Current or planned cervical cerclage or documented history of incompetent cervix
- A short cervix (less than 20 mm by vaginal ultrasound)
- Preterm labor in the current pregnancy
- History of spontaneous preterm delivery
- Placenta previa or placental abruption
- Maternal red cell or platelet alloimmunization
- Maternal pre-gestational insulin-dependent diabetes
- Uterine anomaly (multiple or large leiomyomata or müllerian duct abnormality)
- Infection with HIV or hepatitis B or C
- Inability to comply with travel or follow-up requirements

Consultation
In addition to comprehensive counseling regarding fetal surgery, patients are educated on having a child with spina bifida. All patients, regardless of whether they qualify for fetal surgery, undergo extensive two-day counseling with the multidisciplinary experts in fetal surgery and spina bifida.

These include:
- Maternal-fetal medicine specialists
- Pediatric surgeons
- Pediatric neurosurgeons
- Long-term spina bifida outcomes specialists
- Neonatologists
- Anesthesiologists
- Social work
- Child Life specialists

Surgery
Fetal repair of spina bifida requires a multidisciplinary team of experts. Although the operation is much like a cesarean section, the fetus is not removed from the uterus. An incision is made in the mother’s abdomen and the uterus, just large enough for the spinal defect to be operated on. The spina bifida defect is surgically repaired by the pediatric neurosurgeon, much as it would be after the baby is born if the fetal surgery were not undertaken. After the procedure, the incisions in the mother are closed and the pregnancy is allowed to continue.

The Fetal Spina Bifida Surgery Program
The Texas Fetal Center is dedicated to providing exceptional care by practicing evidence-based medicine and providing an experienced team of fetal myelomeningocele specialists.

The Center is dedicated to the education of its patients.
- All patients and their families are counseled by every specialist involved in the care of mom and baby. Online educational materials have been developed to provide the full scope of spina bifida and fetal surgery.

The Center believes in an experienced fetal surgery team.
- The spina bifida team has visited all three centers that participated in the MOMS Trial, including observation of patient counseling and the actual surgical technique.
- The experienced surgeons have performed fetal myelomeningocele repair prior to the MOMS Trial.
- The experienced MFM specialist participated in the MOMS Trial.
- The fetal surgical team has participated in the UCSF Fetal Surgery for Spina Bifida Training Course, and includes neurosurgeons, MFM specialists and obstetrical anesthesiologists.

The Center believes that the care of the unborn patient with spina bifida does not end with fetal surgery. The Texas Fetal Center has established a collaboration with the long-term spina bifida program at the world-renowned Shriner’s Hospital of Houston.
Meet the Team
The fetal surgery team at the Texas Fetal Center consists of fetal surgeons with extensive previous fetal myelomeningocele repair experience (10 operations prior to the MOMS Trial) and fellowship training at the University of California, San Francisco (lead institution in the MOMS Trial), as well as a maternal-fetal medicine specialist who has counseled over 100 patients during the MOMS Trial at the Children’s Hospital of Philadelphia. In addition, specialists at the Shriner’s Hospital for Children in Houston provide the most comprehensive and state-of-the-art long-term care for all children with myelomeningocele.

KuoJen Tsao, M.D.  Co-director and associate professor of pediatric surgery, completed his fellowship in fetal therapy with the Fetal Treatment Center at the University of California, San Francisco, during the inception of the MOMS Trial and the early fetal surgeries for spina bifida.

Kenneth J. Moise Jr., M.D.  Co-director and professor of obstetrics, gynecology and reproductive sciences, headed the fetal surgery program for spina bifida at the University of North Carolina and had performed 10 cases prior to the MOMS Trial.

Lynette J. Mazur, M.D., M.P.H.  Medical director of the Shriner’s Hospital Spina Bifida Clinic and professor of pediatrics, leads the multidisciplinary team that provides long-term care for all children with spina bifida, including one of the most advanced gait labs in the United States.

Michael W. Bebbington, M.D., M.H.Sc.  Director of prenatal diagnosis and fetal imaging and professor of obstetrics, gynecology and reproductive sciences, joined the Texas Fetal Center from the Children’s Hospital of Philadelphia where he was one of the lead MFM specialists for the MOMS Trial. While there, he counseled over 100 patients and participated in the 30 fetal operations for spina bifida.

Anthony Johnson, D.O.  Co-director and professor of obstetrics, gynecology and reproductive sciences, has over 25 years of fetal intervention including open fetal surgeries for fetal lung masses, sacrococcygeal teratomas and EXIT procedures.

Referrals
The Texas Fetal Center is a collaboration between Children’s Memorial Hermann Hospital and The University of Texas Health Science Center at Houston (UTHealth) Medical School. To refer a patient, please call 832.325.7288 or toll free 1.888.818.4818.

For more information, visit texasfetalcenter.org/spina-bifida.