In-Utero Repair of Meningomyelocele

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Meningomyelocele
THE CLINICAL SPECTRUM

- Paralysis of muscles with sensory and motor deficit. Dependant on clinical exam level not the skin lesion.
- Neurogenic bladder and bowel.
- CSF circulation problems with resultant Hydrocephalus and Syringomyelia.
- Abnormalities of cerebral migration.
- Brainstem abnormalities with clinical sequelae.
- Progressive scoliosis secondary to Syringomyelia, osteologic abnormalities and muscle imbalance.
History

John Cleland-1883

Hans Chiari- 1891
Fetal Open Neural Tube Defect
MRI
MOMS Trial
Management of Meningomyelocele Study

- Multi-Institutional Study: UCSF, Vanderbilt, CHOP
- Funded by NIH
- Patient exclusion: High BMI, fetal MRI qualifiers not met, psychological well being of mother
- Fetus must have open neural tube defect, hydrocephalus, and tonsillar herniation
- Operative timeline about 23-24 weeks gestation
- Variability of repair techniques
- Risks of fetal death, premature labor and birth, uterine injury and maternal mortality/morbidity, c-section obligation for future pregnancy
Benefits                                   Fetal Morbidities
• Decrease in need for VP shunt by 30%
• Improvement in hindbrain herniation by 30%
• Improvement in neurological level
• Difficulty with skin closure
• Surgical Blood loss
• Operative time
• Need for skin graft
• Risk of future tethered cord (may be even worse if graft used)
Why Does this Work

• Stop spinal leak and low pressure herniation which causes Chiari II
• Allows ‘normalization ‘ of CSF flow up and down neuraxis
• Tonsils float back up into the posterior fossa
• CSF pathways from ventricles are re-established: hydrocephalus better
• What evidence?
FIRST FETAL LUMBAR OPEN NEURAL TUBE DEFECT
FIRST FETAL REPAIR
HYDROCEPHALUS

IN UTERO HYDROCEPHALUS

POST DELIVERY VENTRICLES
FIRST FETAL CHIARI II MALFORMATION

IN UTERO TONSILLAR HERNIATION           POST DELIVERY NORMAL POSITION
Current Research and Trends – UT/Children’s

vs

Current Trends coupled with Research

1. The move to endoscopic approaches for repair. Less uterine trauma. Longer surgical times and higher morbidity and mortality

2. The desire to find the best ‘patch’ or bioadhesive that can be used in-utero

3. UT Fetal Research Program:
   1. Laser activated bioadhesive (Sandcastle Snail secretion) capable of adherence underwater. Partner University of Utah
   2. Development of flexible, endoscopes for in utero repair. Partner Mischer Neuroscience Institute
   3. Immune responses of amniotic fluid and CSF to various foreign materials. Partner Univ. of Mississippi
   4. Radiological assessment of Fetal Surgery for Meningomyelocele Repair. Partner Univ. of Texas Neuroradiology
Rat/Ovine model, sandcastle snail adhesive, human umbilical cord patch
Normal Open defect Repair with HUC

Ovine Meningomyelocele Model
First FDA approved use of Human Umbilical Cord for Fetal Meningomyelocele Repair
The Fetal Center at Children’s Memorial Hermann Hospital
The University of Texas Medical School

First In-Utero Repair of a meningomyelocele in the State of Texas
First ever use of cryopreserved Human Umbilical Cord for Cutaneous Patch for in-utero repair of a patient with lumbar myeloschisis
Ongoing research program involving specialists in the field of immunology, fetal medicine, pediatric neurosurgery, neuroradiology, ophthalmology, and veterinary medicine.
Partnership with researchers at the University of Mississippi, University of Utah, Ocular Surface Center in Miami, and private industry.
The University of Texas Fetal Center and Children’s Memorial Hermann Approach to Fetal Surgery for Open Neural Tube Defects

• Multidisciplinary approach that includes neuroradiology, fetal surgeons, pediatric surgery, neonatology, general pediatrics, and social work
• Comprehensive assessment of risks and psychological makeup of the mother and father
• An analysis of available care in the region where they live, especially neurosurgical care
• A thorough discussion and assessment of the management of the hydrocephalus. Shunting was an end measure of the MOMS trial
• We like other centers consider endoscopy as a reasonable alternative to management of hydrocephalus if the anatomy is favorable
• Thorough discussion of the long term needs of the spina bifida patient in general from a pediatric, urological, orthopedic and therapy standpoint
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