Fetal Surgery

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Our Early Beginnings

The Concept:
fix simple anatomic defects which lead to disastrous physiologic consequences
UCSF Fetal Treatment Center

- Founded in 1981
- First Fetal Treatment Center in the United States
- Multidisciplinary Team Approach
- University Appointed Oversight

First Animal Studies - 1981

Hysterotomy for fetal surgery & maternal safety demonstrated in a monkey model.

Fetal Rx: Judging Risks vs. Benefits

- Risks to Mother
- Benefits/Risks to Fetus
- Future Benefits to Fetal Patient

Maternal Safety
Fetal Surgery: Open Hysterotomy

Minimally Invasive Techniques

Fetal Conditions Treated

- Tumors/Lung Lesions
- Congenital Heart Disease
- Congenital Diaphragmatic Hernia
- Spina Bifida
- Urinary Obstructions
- Complications of Twin Pregnancies
- Fetal Anemias

Urinary Obstruction

- Clinical Findings
  - Obstruction Can Lead to Renal Impairment
  - Decreased Amniotic Fluid = Lung Hypoplasia
Urinary Obstruction

- Obstruction Can Lead to Renal Impairment
- Decreased Amniotic Fluid = Lung Hypoplasia
- Open Vesicostomy
- Bladder Catheter
- Fetal Cystoscopy

Criteria for Intervention

- Isolated anomaly
- Appearance of Kidneys
- Urine analysis
  - Sodium < 100
  - Chloride < 90
  - Osmolality < 200
  - Beta 2 < 6

Tumors

- CCAM
- SCT
- Cervical Teratomas
- Pericardial Teratomas

Tumor Treatment

- Prenatal or Postnatal Therapy
- Size of the Mass
- Amount of Blood Flow/Vascularity
- High Output Failure/Hydrops

References:
Sacrococcygeal Teratomas

Neck Masses/EXIT Procedure

Congenital Diaphragmatic Hernia
- Incomplete Formation of the Diaphragm
- Resultant Pulmonary Hypoplasia
CDH: Natural History

- Hidden Mortality
- Hidden Morbidity
- Spectrum of Severity

First CDH Clinical Trial

- First Fetal Surgery Clinical Trial
- Completed in 1994
- Studied “Liver down” patients
- Operation successful but not necessary


CHAOS

- Accident of nature
- Laryngeal or Tracheal Stenosis/Atresia
- Prevents egress of lung fluid
- Result: Overexpanded Lungs

Promoting Lung Growth

- Increases Lung Mass
- Improves Gas Exchange
- Reduces Viscera
Balloon Tracheal Occlusion

- Minimally Invasion Procedure
- Performed Under Ultrasound Guidance
- Inhibits Egress of Fetal Lung Fluid

NIH-sponsored Clinical Trial

- Randomized Clinical Trial
- March 2003
- Prenatal Surgery versus Postnatal Surgery
- 200 women to be enrolled
- Compare safety and efficacy of prenatal vs postnatal surgery
- Inclusion Criteria:
  - S1-T1
  - Normal karyotype
  - US resident
  - Presence of Chiari II
  - 19 to 25 weeks’ gestation

Primary Hypotheses

- Midtrimester repair of fetal myelomeningocele compared with standard postnatal repair:
  - Reduces the risk of death or ventricular decompression shunting
  - Results in an improvement in neurologic and neuromotor function
Myelomeningocele

- Hydrocephalus
- Arnold-Chiari II Malformation
- Impairment of Bowel and Bladder Control
- Motor and Sensory Losses

First Primary Outcome (12 months)

<table>
<thead>
<tr>
<th></th>
<th>Prenatal N=78</th>
<th>Postnatal N=80</th>
<th>RR (95% CI)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary outcome</td>
<td>53 (68)</td>
<td>78 (98)</td>
<td>0.70 (0.58–0.84)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Components of primary outcome</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Death before shunt placement</td>
<td>2 (3)</td>
<td>0</td>
<td></td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Shunt criteria met</td>
<td>51 (65)</td>
<td>74 (92)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Shunt placed not met criteria</td>
<td>0</td>
<td>4 (5)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Placement of shunt</td>
<td>31 (40)</td>
<td>66 (82)</td>
<td>0.48 (0.36–0.64)</td>
<td>&lt;0.001</td>
</tr>
</tbody>
</table>

Two perinatal deaths in each group:
Prenatal: IUFD at 26wks, NND at 23wks
Postnatal: NND with severe symptoms of Chiari II

Secondary Outcome: Hindbrain Herniation (12 months)

<table>
<thead>
<tr>
<th></th>
<th>Prenatal N=70</th>
<th>Postnatal N=69</th>
<th>RR (95% CI)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Any hindbrain herniation</td>
<td>45 (64)</td>
<td>66 (96)</td>
<td>0.67 (0.56–0.81)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Degree of hindbrain herniation</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>25 (36)</td>
<td>3 (4)</td>
<td></td>
<td></td>
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<tr>
<td>Mild</td>
<td>28 (40)</td>
<td>20 (29)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>13 (19)</td>
<td>31 (45)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>4 (6)</td>
<td>15 (22)</td>
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</table>

Secondary Outcome: Ambulation

<table>
<thead>
<tr>
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<th>Prenatal N=62</th>
<th>Postnatal N=67</th>
<th>RR (95% CI)</th>
<th>P</th>
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<tbody>
<tr>
<td>Walking independently on exam</td>
<td>26 (42)</td>
<td>14 (21)</td>
<td>2.01 (1.16–3.48)</td>
<td>0.01</td>
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<tr>
<td>Walking status</td>
<td></td>
<td></td>
<td></td>
<td>0.03</td>
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<tr>
<td>No</td>
<td>18 (29)</td>
<td>29 (43)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Walking with orthotics or devices</td>
<td>18 (29)</td>
<td>24 (36)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Walking without orthotics</td>
<td>26 (42)</td>
<td>14 (21)</td>
<td></td>
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Fetal and Neonatal Outcomes

<table>
<thead>
<tr>
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<th>Prenatal N=78</th>
<th>Postnatal N=80</th>
<th>RR (95% CI)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bradycardia at repair</td>
<td>8 (10.3)</td>
<td>0</td>
<td>0.003</td>
<td></td>
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<tr>
<td>Perinatal death</td>
<td>2 (2.6)</td>
<td>2 (2.5)</td>
<td>1.03 (0.14-7.10)</td>
<td>1.00</td>
</tr>
<tr>
<td>GA at birth</td>
<td>34.1±3.1</td>
<td>37.3±1.1</td>
<td>&lt;0.001</td>
<td></td>
</tr>
<tr>
<td>&lt; 30 wks</td>
<td>10 (12.8)</td>
<td>0 (0.0)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>30-34 weeks</td>
<td>26 (33.3)</td>
<td>4 (5.0)</td>
<td>15%</td>
<td></td>
</tr>
<tr>
<td>35-36 weeks</td>
<td>26 (33.3)</td>
<td>8 (10.0)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;=37 weeks</td>
<td>16 (20.5)</td>
<td>68 (85.0)</td>
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</tr>
</tbody>
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Summary

- Prenatal surgery associated with maternal and fetal risks
  - Preterm birth: 80% vs 15%
  - RDS in 21% vs 6%
  - Bradycardia
  - Oligohydramnios
  - Placental abruption
  - Transfusion at delivery
  - Uterine dehiscence at surgical site (35%)
Twin-Twin Transfusion Syndrome

- Disease of the Placenta
- Up to 50% Mortality
- Treatment Options:
  - Amnioreduction
  - Laser of Placental Vessels
  - Septostomy

Placental Vascular Anatomy

- A placenta is designed to support one fetus
- When one placenta supports two fetuses, there is no set plan or roadmap for how it will do this
- The angioarchitecture is always different; each MC placenta is like a snowflake
  - how the two circulations interact is always unique

Polyhydramnios/Oligohydramnios

Oligohydramnios/Polyhydramnios
Quintero Staging for TTTS

- Stage I (abnormal amniotic fluid levels)
- Stage II (no donor bladder)
- Stage III (abnormal doppler flow in either twin)
- Stage IV (hydrops in either twin)
- Stage V (demise of either twin)

Arterio-Venous Anastomosis

Normal Artery & Vein
Laser ablation

- Laser is minimally invasive, single port, 3mm device
  - Does carry up to 10-15% risk of PPROM/PTL/PTD
- Eurofetus trial (2004) had shown single/dual survivorship of 76% and 36%, respectively
- UCSF data shows single/dual survivorship of 84% and 33% respectively; mean GA 32wks
  - Well selected pts
  - Operator and experience dependent

Laser Ablation for TTTS

Advances in Imaging: Ultrasound vs MRI

Conditions We Treat

- Congenital diaphragmatic hernia
- Esophageal atresia
- Hirschsprung's disease
- Imperforate anus
- Congenital cystic adenomatoid malformation
- Pulmonary sequestration
- Gastrochisis / Omphalocele
- Hepatic and biliary anomalies
- Sacrococcygeal Teratoma