Objectives

- Understand how fetal diagnosis, prenatal counseling and fetal interventions are relevant to pediatrics now and in the future
- Review what is currently available for fetal evaluation and care at CMH and what might be feasible in the near future
NMR by Level of Delivery Hospital: Meta-analysis

COMMITTEE ON FETUS AND NEWBORN Pediatrics 2012;130:587-597
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- Full range of pediatric medical subspecialists, surgical subspecialists, pediatric anesthesiologists
- Capable of surgical repair of complex congenital or acquired conditions
- “concentrating care at designated Level IV centers will allow these centers to develop expertise...”

Incidence of Birth Anomalies

<table>
<thead>
<tr>
<th>Sample conditions</th>
<th>Estimated Incidence1</th>
<th>Percent</th>
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<tbody>
<tr>
<td>Cardiovascular</td>
<td></td>
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<tr>
<td>Hypoplastic left heart syndrome</td>
<td>0.02</td>
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<td>Tetralogy of Fallot</td>
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<td>Transportation of the great arteries</td>
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<tr>
<td>Neurology</td>
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<tr>
<td>Cerebral dysplasia</td>
<td>0.12 - 0.22</td>
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<tr>
<td>Spinal dysraphism</td>
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<tr>
<td>GI</td>
<td></td>
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<tr>
<td>Esophageal, duodenal, intestinal atresia</td>
<td>0.03 - 0.13</td>
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<tr>
<td>Anorectal malformation</td>
<td></td>
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<tr>
<td>Other</td>
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<tr>
<td>Abdominal wall malformation (gastrochisis,</td>
<td>0.28 - 0.47</td>
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<tr>
<td>omphalocele)</td>
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<tr>
<td>Congenital hydrocephrosis</td>
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<tr>
<td>Cystic hygroma/cervical teratoma</td>
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<tr>
<td>Cystic adenomatoid malformation of lung</td>
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<tr>
<td>Diaphragmatic hernia</td>
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<td></td>
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<tr>
<td>Sacrococcygeal teratoma</td>
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<td></td>
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<tr>
<td>Total</td>
<td>1.28 - 1.62</td>
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</table>

1 Per all pregnancies in the United States.

Prenatal diagnostic testing

Invasive:
- Amniocentesis, CVS
- PUBS
Non invasive:
- ultrasound testing, echocardiogram, MRI
- cell-free DNA in maternal blood
Fetal Ultrasound: Detection Rate for fetal anomalies

- Fadda (2009): 55% (23 yr experience)- varied by dx (cardiac, 33%, CNS 81%)
- Wong (2004): 73% (6 yr Australian cohort)
- Nicolaides (2011): “combined testing” for T21 – 93-96%

Why does prenatal diagnosis and treatment matter?

Structural anomalies are the leading cause of neonatal mortality in US

Continuum of Care
Fetus is a patient when a diagnosis is made
Pediatric subspecialty input
“Prenatal Pediatrics”
Interdisciplinary care for family – integrated counseling
† Fetal treatment

Influence of prenatal diagnosis on HLHS outcomes

- Higher Apgar scores
- Less preoperative acidosis
- Less inotropes or ventricular dysfunction
- Less end organ damage
- Less pre-op ventilation
- Delivery at higher level center
- Earlier surgery

Levey, 2010; Kips 2011; Sivarajan, 2009
Elizabeth J. Ferrell
Fetal Health Center

- Integrated prenatal services to include pediatric subspecialty and surgical input focused on the fetus with a birth defect
- Multidisciplinary, collaborative approach to assist with diagnosis, comprehensive family counseling, fetal interventions as available, delivery plan, and postnatal care
- Working cooperatively with area obstetricians, maternal-fetal specialists

FHC Integrated Consults (n=312)

- OB/MFM referrals for consultation/testing
- Fetal echocardiogram
- Fetal MRI
- Integrated prenatal consultation
- FHC delivery
- Following consultation: written f/u with OB, including postpartum recommendations when appropriate
  - If not delivering at CMH, primary pediatrician/neonatologist contacted with summary of findings and recommendations
  - FHC social service and nursing f/u with family
  - Delivery at CMH: transfer of care to FHC perinatology

Fetal Diagnosis for referrals to FHC

- Normal 25%
- Cardiac 17%
- GU 12%
- GI 11%
- Craniofacial 7%
- Pulmonary 7%
- Skeletal 5%
- Complex 1%

Prenatal testing is stressful

- Related to severity of lesion but even testing creates anxiety
- Decision making after abnormal testing results is stressful and individualized support is needed
- Prenatal dx of congenital malformation may increase psychological distress
- Parents want realistic information, specific for their infant, provided empathetically

Miquel-Verdes et al, Pediatrics, 2009
Parent Satisfaction
FHC survey 2011

- Questions answered 21/23
- Data useful for decision making 20/23
- Consult useful 24/24

Comments:
“we were pretending nothing was wrong” – provided “reality”
“Useful to have everyone together”
“helped with our understanding”

Fetal Health Center

- Delivery of selected infants on-site at CMH
- potentially unstable birth defects
- assist with transition to neonatal care
- immediate, life-saving interventions
- allow mother to be with her infant during first critical hours of evaluation and care
**FHC Deliveries**
- 139 Live births
- 1 Stillbirth
- 16 (12%) neonatal deaths
- 94 (68%) neonatal surgery and/or cardiac cath
- High intensity (urgent interventions):
  - Complex CHD 12
  - CDH 7
  - Pulmonary hypoplasia 3
  - Airway obstruction 2

**FHC Case TC**
- 27 year old, G3, P2
- Referred 32 weeks GA, care transferred
- Anhydramnios since 29 weeks
- Multicystic dysplastic left, poorly functioning right kidney
- 46 XY
- Integrated prenatal visit:
  - Peritoneal dialysis, renal transplant
  - Neurodevelopmental risks
  - Pulmonary hypoplasia, ECMO
  - Ethical considerations

**FHC Care TC**
- Delivery at 36 weeks GA, 2330g BW
- Hospital course
  - Immediate respiratory distress
  - Ph 6.8, PaCO₂ 128
  - Pneumothorax, HFOV, iNO
  - ECMO 15 days, hemodialysis
  - Nephrectomy, PD at 26 days
  - Hospital discharge, 6 months
  - 17 months: continues outpatient dialysis

**FHC Case AY**
- 32 year old G5 P4, referred for CHD 23 weeks GA
- Echocardiogram: HLHS, intact septum
- Integrated prenatal visit
  - Reviewed high mortality
  - Need for urgent catheterization, possible ECMO
  - Delivery plans, relocate to KC
  - Surgical plans: hybrid procedure
  - Assisted with family, social stress
  - Delivery planning-multidisciplinary meetings, simulation training
FHC Care AY

- Elective Cesarean Section
  - Neonatology, Cardiology, Anesthesia, CV Surgery, ECMO team
- Hypoxic (saturation 35%), resuscitation: intubation, UVC, Echocardiogram
- Prepped for ECMO? but stabilized – moved to cath lab < 1 hour
  - atrial septectomy on bypass
- 12 days: hybrid procedure (PDA stent, pulmonary bands)
- Remains in PICU

What’s next: Fetal Interventions

- Shunting
- Fetoscopy
- Exit Procedures
- Open procedures
Endoscopic fetoscopy

- Urinary tract obstruction
- Twin-Twin Transfusion
- Acardia (TRAP)
- CDH

Twin-to-Twin Transfusion Syndrome

- Monochorionic, diamniotic
  - Single placenta
  - Two amnions
  - 1 in 360 pregnancies
- Vascular connections
  - 15-20% unbalanced: donor & recipient (oligo, polyhydramnios)

Main Outcomes of Eurofetus trial compare laser photocoagulation versus serial amnioreduction for stage 1 to 4 TTTS

<table>
<thead>
<tr>
<th>Survival of at least one twin</th>
<th>Laser group 76% (55/72)</th>
<th>Amnioreduction group 51% (36/70)</th>
<th>P-value 0.002</th>
</tr>
</thead>
<tbody>
<tr>
<td>Periventricular leukomalacia</td>
<td>6% (8/144)</td>
<td>14% (8/140)</td>
<td>0.02</td>
</tr>
<tr>
<td>Free from neurological complication</td>
<td>52% (75/144)</td>
<td>31% (44/140)</td>
<td>0.003</td>
</tr>
</tbody>
</table>

FETO for CDH

- Fetal Tracheal Occlusion
- Compensatory lung growth
  - In-utero repair of CDH = lung growth
- Patient selection
  - Intrathoracic liver
  - LHR < 1.4
- Clip vs balloon

Fetoscopy to place balloon

CDH Response to TO

Fetal Outcome DOL #1 CXR

CDH

CDH + TO
**EXIT**

- EX utero Intrapartum Treatment
- NOT Cesarean Section
  - controlled uterine hypotonia
  - preserved uteroplacental circulation

**EXIT: indications**

- Reversal of tracheal occlusion
- EXIT to airway: neck mass, high airway obstruction, micrognathia
- EXIT to ECMO: CDH, CHD
- EXIT to resection: congenital lung lesion, SCT

**Open Fetal Procedures**
Sacrococcygeal teratoma

Posterior Urethral Valves

Myelomeningocele
- Incomplete neural tube closure
- 1 in 2,900 LBs
- 2 hit theory
  - Developmental defect
  - Neural elements damaged from exposure

MOMS Trial (Management of Meningomyelocele)
- Study concluded early (2011)
- Powered for 200
- 183 enrolled
- Primary outcome: Death or need for VPS
  - 68% vs 98% (p< 0.001)
- Decreased hindbrain herniation
  - 64% vs 96% at 12 months (p< 0.001)
- Walk independently 30 mo
  - 42% vs 21% (p = .01)

MOMS Trial
- Other significant morbidity
  - Increased preterm labor
    - 34.1 weeks versus 37.3 weeks
    - Lower birth weight in prenatal group
  - Higher placental complications
  - Uterine dehiscence
    - Partial 9%
    - Complete 1%
  - Maternal blood transfusion (9 vs 1%)
  - Maternal pulmonary edema (6 vs 0%)

Myelomeningocele
- Fetoscopy?
- Renewed interest in patches
Conclusions

- Fetal assessments are becoming more refined – clearly the fetus is a “pediatric patient”
- Fetal diagnosis begins the process of care, with comprehensive planning for in-utero, intrapartum and neonatal management decisions in collaboration with informed parents
- Fetal interventions will be less invasive; surgical and non-surgical approaches have potential to provide improved survival with less morbidity for infants with congenital malformations

References