Moving Forward with Prenatal Congenital Heart Disease

3rd Annual Fetal Cardiology Symposium: What Not to Miss
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Epidemiology

- Most common birth defect 1% of births per year (~40,000 neonates/yr)
- 57% of infantile mortality as a result of CHD


Categories of CHD

**Mild CHD**
- Muscular VSD
- Small PDA
- ASD
- Mild pulmonary stenosis
- Bicuspid aortic valve

**Moderate CHD**
- Mild aortic stenosis (AS)
- Moderate pulmonary stenosis (PS)
- Non-critical coarctation of the aorta
- Membranous VSD
Categories of CHD

**Severe CHD**

d-Transposition of the Great Arteries
Tricuspid and Pulmonary Atresia
Hypoplastic Left Heart Syndrome (HLHS)
Single Ventricle Anatomy
Double Outlet Right Ventricle (DORV)
Categories of CHD

Severe CHD

Truncus Arteriosus

Total Anomalous Pulmonary Venous Return

Critical Pulmonary or Aortic Valve Stenosis

Aortic Arch Abnormalities (IAA, COA)
Optimal Timing of Diagnosis
Sub-Optimal Timing of Diagnosis
Unrecognized CCHD

Morbidity & mortality:

- Metabolic acidosis
- Intracranial hemorrhage
- Hypoxic-ischemic encephalopathy
- Necrotizing enterocolitis
- Cardiac arrest
- Death

Impact on Surgery

HLHS postnatal diagnosis = less stable prior to surgery

Prenatal dx = better surgical survival

Neurocognitive effects

Pre vs. postnatal diagnosis of dTGA

- Remedial school services 45% pre vs 69% post
- Postnatal show poorer executive function
- Deficits more prevalent & severe with postnatal dx
  - Cognitive flexibility
  - Social cognition

Delivery Location

- Neonates with HLHS:
  - Birth <10 minutes from tertiary center = 21% mortality
  - Birth >90 minutes away = 40% mortality

Delivery Location

- Delivery in cardiac OR
- Immediate transfer to cath lab
- Emergent cardiac stent
Parental Benefits

Family preparation

• Emotional
• Financial
• Social
Multi-Disciplinary Support

Integrated Consult for Heart Center Providers and other Sub-Specialists

Tour of FHC, NICU, and PICU

RMH and RM Room

Chapel
Multidisciplinary Teams

Cardiology

Child Life/Chaplains

Social Work/PACT

MFM/OB

Neonatology

Genetics

CV surgery
Parental Education

“Core curriculum” for families

58 point checklist

Individualized counseling

Fetal Cardiology Program Counseling Checklist

Cardiac Anatomy and Physiology
- Normal cardiac anatomy and physiology
- Specific fetal cardiac diagnosis with anatomy/physiology
- Physical limitations
- Cardiac disease etiology
- General considerations

Immediate Postnatal Management
- Immediate postnatal stabilization and management
- Umbilical lines and possible intubation
- Prostaglandin
- Expected immediate postnatal cath/surgery interventions

Neonatal Management
- Feeding issues (NPO, breastfeeding)
- NICU parent visitor policies

Surgical Planning
- Details of surgery/cath intervention expected
- Timing of surgery/intervention
- Possible complications of surgery/cath
- Ventilator Support
- Inotropic needs
- Drains
- Open Chest
- Possible Blood Transfusions
- ECMO

Post-Surgical Management
- Length of hospital stay
- Other possible medical issues (need for NGT, g-tube, malrotation)
- Likely discharge medications
- Other anticipated discharge, home nursing, home monitoring

Long-term Issues
- Subsequent surgeries/interventions expected
- Possible transplant
- Projected life expectancy

- Neurocognitive outcomes
- Physical limitations
- Long-term medications
- Long-term cardiology follow-up needs

Extracardiac Issues
- Other diagnosed extracardiac fetal abnormalities interaction with CHD
- Underdiagnosed known associated extracardiac anomalies
- Known genetic associations

Pregnancy Counseling
- Location of delivery
- Timing and mode of delivery
- Expectations for the rest of gestation
- Risk for hydrops
- Possible intrauterine demise
- Outcome of delivering preterm or Hybrid

Family Issues
- Acute and long-term housing needs
- Educational effect of child with significant medical needs
- Effect of special-needs child on family
- Genetic implications for future family planning
- Introduction of community family support groups
- Connections of family to other family with similar CHD

Consults
- PACCT
- Genetics
- CV Surgeon
- Social Work
- Surgery

Other System Issues
- ENT: vocal cord dysfunction
- Hematology: clot bleeding
- Neurodevelopmental
Parental Stress

+ time + support + education =
National Prenatal Detection

(Donofrio 2014; Levy et al 2013)
Prenatal Detection Process

- Fetal Cardiology
- Abnormal OB Screening Ultrasound
- MFM/Perinatal
- Fetal Cardiology

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Early detection

- Nuchal Translucency
- Ductus Venosus Flow
- Tricuspid Regurgitation

Fetal Anomaly Screening

Normal

Not Normal
Fetal Anomaly Screening

CHD detection rates vary widely

- 55-65% with four-chamber view alone
- 80-84% with outflow tract view in addition
Kansas City Region
Prenatal Detection by Disease

Prenatal Detection of Disease Specific
Neonatal Critical Cardiac Lesions (2010-2014)
Barriers to Prenatal Detection

Retrospective cohort study in Utah

- Utah Birth Defect Registry, 1997-2007
- Mandatory prenatal screening
- Objectives:
  - Determine rate of prenatal detection of CHD
  - Identify maternal & encounter risk factors for failed prenatal detection

Barriers to Prenatal Detection

Results:

- 97% of mothers had ≥ 1 prenatal US
- 77% between 16-24 wks
- 39% of CHD was detected prenatally

Barriers to Prenatal Detection

- Abnormal ultrasound:
  - 89% seen by MFM
  - ...but 42% never had fetal echo
- 35% of mothers with FamHx did not receive fetal echo
- Of those with fetal echo, 3% had a missed CHD Dx

Barriers to Prenatal Detection

- Missed cases prenatally
  - 42% expected abnormal four-chamber view
  - 64% expected abnormal outflow-tract view
  - 30% both views expected to be abnormal

- CHD detection rates did not improve over 10 years

KC Region Prenatal Detection of Critical CHD
National Prenatal Detection

Percent Detection

- Bull 1999 (n=4799)
- Game 2001 (n=2454)
- Jaeggi 2001 (n=659)
- Tegnander 2006
- Acherman 2007
- Pinto 2007
- Khoo 2008 (n=200)
- Acharya 2008
- Friedberg 2009 (n=39)
- Marek 2011 (n=1604)
- Levy 2013 (n=93)

74%
Standardized Fetal Echo Screening

- Educational program for sonographers
- Rotate sonographers with Pediatric Cardiologists
- Include 4 chamber view and both outflow tracts on routine screening U/S
- Video Clips (not still-frames)

(Donofrio 2014; Levy et al (2013))
Reasons for Referral

• Concern for CHD
• Maternal Risk Factors
  • Maternal disease
  • IVF
  • Family History
• Fetal Risk Factors
  • Genetic Abnormality
  • Extracardiac anomaly
  • Multiple gestations
How do we identify neonatal CHD?
Newborn CCHD Screening Progress

Click on a state for additional details.
Summary

• 1% of neonates have CHD
• Prenatal diagnosis can save lives
• Many other benefits to prenatal diagnosis
Summary

• Prenatal diagnosis is challenging
• Team effort
• Currently 50% of our regional children receive prenatal dx...

…but we can do better!
Thank you!
References


References


