Before we are Born: Fetal Diagnosis of Congenital Heart Disease

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First Pediatric Cardiology Symposium of Kidsheart

Welcome to KidsHeart
Disclosure

- No thing to disclose except:
  - I am the Co-founder of Kidsheart: American Fetal and Children heart center.
Objectives:

- Prenatal detection of CHD
- Indication for fetal echo.
- Program for fetal echocardiography in UAE.
Prenatal Ultrasound

1979 “of academic interest only. It should not be used to influence clinical judgment.”

2013:
- diagnosis of CCVM as early as 12 weeks of gestation
- assess physiology of heart failure
- diagnosis of cardiac arrhythmias and guide treatment
- gain insight into the natural history of congenital and acquired cardiac defects
Why screen for CHD

- CHD are the most common lethal congenital malformations and

- The most common congenital defect (8/1000 live births)

- The majority of CHD are from low risk pregnancies
An Ultrasound is an Ultrasound?

Level I:  Fetal size and Gestational age  
cardiac motion  
**OB/Technician (do and read)**

Level II:  Fetal Structural Survey  
four chamber view  
**Perinatologist/Radiologist**

Level III:  Fetal Echocardiogram (Pediatric Cardiologist/some prenatologist)
Four Chamber View (Level II)
Detection of CHD: How Good is a Screening Ultrasound?

• Accurate 4 chamber view detects 4 – 50% of “serious” CHD.

• **DOES NOT** detect CHD with outflow tract anomalies that can be ductal dependent lesions.
Abnormalities Detectable by 4 Chamber Screening Examination

- “Single” Ventricle
- Complete AV septal defect
Cardiac Surgery on “Single Ventriles” at UOC (July 1999 – April 2008)

Diagnosed before birth

- SV: 11%
- HLH: 37%
- HRJ: 18%
Hypoplastic left heart Syndrome
Cardiac Surgery on “Complete AV Septal Defect” at UOC (July 1999 – April 2008)

Diagnosed before birth

8%
Tricuspid valve dysplasia

Tricuspid atresia
Detecting Fetal CHD: 
Non-Selected Population at 18-24 Weeks

- 5347 fetuses evaluated with 4 chamber view PLUS follow-up scan if imaging suboptimal in first scan

- Scan time (US and 4-C heart) 30 minutes

- Detected 48% of CHD

- Incorporating great vessel views increased detection to 78%

Abnormalities Detectable by 4 Chamber Plus Outflow Tract Exams

- Transposition of the great vessels
- Tetralogy of Fallot
- Persistent truncus arteriosus
- Interrupted aortic arch
Improving the detection rate

Adding the LVOT to the scan

Adding the RVOT to the scan
Transposition of Great Arteries
TOF

DORV
Rhabdomyoma
Pulsed Doppler
Color Flow Mapping

[Images of ultrasound scans with annotations]

RV

RA
Detecting Fetal CHD:

- Unpublished data from Tawam hospital database

- Detection rate is 92%. Initially was 87%, then improved to 96% in the last few years
Prenatal Detection of CHD

1. CHD almost never manifest before birth except valve regurgitation.
2. Screening ultrasound for all patients at 20 – 22 weeks.
3. We cannot screen everyone with fetal echo (time and money).
4. Screen high risk, and suspicious screening ultrasound.
Risk factors for CHD

Maternal Risk

1. Diabetes
2. Anti-SSA/SSB (Ro/La) antibodies
3. Family hx CHD or cardiac syndrome
4. Teratogen exposure
5. Abnormal serum screening
Family History of CHD

• If either parent: risk is 2.5 – 4%
• If sibling: risk is 1 – 4%
• If more than one sibling affected risk is 3 – 10%
• Highest recurrence risk is for left outflow obstruction
  • (AS, COA, HLH variants): 10 – 15%
Family History of Syndromes with Cardiac Involvement

- Williams syndrome
- Marfan syndrome
- Tuberous sclerosis
- 22 q 11 Deletion syndrome
- Hypertrophic cardiomyopathy
- Noonan syndrome
Risk factors for CHD

Fetal Risk

1. Non-cardiac anomaly
2. Suspected cardiac defect (Very important)
3. Chromosome abnormalities.
4. Increased nuchal translucency
5. Hydrops fetalis
6. Arrhythmia
Chromosome Abnormalities and CHD

• Trisomy 21: 40%
  • CAVCD, VSD, TOF

• Trisomy 18: 90%
  • VSD, DORV, Polyvalver disease

• Trisomy 13: 80%
  • VSD
Miscellaneous

- Hydrops or isolated effusion
- Suspected twin: twin transfusion
- IUGR
- IVF
Abnormal Rate or Rhythm

- Bradycardia
  - HR < 100 bpm

- Tachycardia
  - HR > 180 bpm

- Irregular rhythm
M – Mode and arrhythmia

---1---
Dist = 0.41 cm
ΔT = 0.133 s
ΔT → = 450 bpm
Slope = 3.10 cm/s

---2---
Dist = 0.30 cm
ΔT = 0.261 s
ΔT → = 230 bpm
Slope = 1.13 cm/s
Advantages of Prenatal Diagnosis

- Parental reassurance
- Multidisciplinary counseling
- Therapeutic intervention
  (arrhythmias – not structural defects)
- IMPROVED OUTCOME
- No surprises after birth
Pediatric Fetal Cardiology program

Multidisciplinary team:

- Pediatric Cardiologist
- Prenatal OBGYN

NICU team

Pediatric Fetal Cardiology program

TEAM WORK
Why we are doing all of this?

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<th>Year</th>
<th>Children With CHD</th>
<th>Adults with CHD</th>
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Thank you
Where is this Park located?