Basic Fetal Cardiac Evaluation

Mert Ozan Bahtiyar, MD

Director, Fetal Care Center
Division of Maternal Fetal Medicine
Department of Obstetrics, Gynecology and Reproductive Sciences
Background

- CHD is a leading cause of infant mortality
- Prenatal detection may improve outcomes
  - TGA, HLHS, coarctation
- Society guidelines
- Screening exam vs. echocardiogram
Fetal Echocardiography

Guideline developed in conjunction with the American College of Obstetricians and Gynecologists (ACOG), the Society for Maternal-Fetal Medicine (SMFM), and the American Society of Echocardiography (ASE), and endorsed by the American College of Radiology (ACR).

Consensus Report on the Detailed Fetal Anatomic Ultrasound Examination

Indications, Components, and Qualifications

76811 Task Force

Ultrasound Obstet Gynecol 2013; 41: 348–359
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ISUOG Practice Guidelines (updated): sonographic screening examination of the fetal heart
Examination “Levels”

• **Basic Ultrasound (76805)**
  - 4 chamber view
  - RVOT
  - LVOT

• **Detailed Ultrasound (76811)**
  - Basic +
  - Aortic arch
  - SVC/IVC
  - 3VV
  - 3V&T

• **Fetal echocardiogram**
Fetal Echo - Some Fetal Indications

- Abnormal cardiac screening exam
- First degree relative of fetus with CHD
- Abnormal heart rate or rhythm
- Fetal chromosomal anomaly
- Extracardiac anomaly
- Hydrops
- Increased NT
- Monochorionic twins
### Congenital Heart Defects in Monochorionic Twin Gestation

#### Review:
- Congenital Heart Disease in Monochorionic Twin Gestations: A Systematic Review

#### Comparison:
- 02 CHD Prevalence in Monochorionic Twin Gestation

#### Outcome:
- 01 CHD Prevalence in MC/DA Twin Gestations

<table>
<thead>
<tr>
<th>Study or sub-category</th>
<th>RR (random) 95% CI</th>
<th>RR (random) 95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>01 No TTTS</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kratza</td>
<td>4.04 [1.53, 10.66]</td>
<td></td>
</tr>
<tr>
<td>Manning</td>
<td>7.93 [4.74, 13.25]</td>
<td></td>
</tr>
<tr>
<td>Lopriore</td>
<td>4.11 [1.04, 16.20]</td>
<td></td>
</tr>
<tr>
<td>Subtotal (95% CI)</td>
<td></td>
<td>6.51 [4.23, 10.01]</td>
</tr>
</tbody>
</table>

- Total events: 20 (MC Gestation), 8013 (Singleton Gestation)
- Test for heterogeneity: $\chi^2 = 1.98$, df = 2 ($P = 0.37$), $I^2 = 0\%$
- Test for overall effect: $Z = 8.52$ ($P < 0.00001$)

<table>
<thead>
<tr>
<th><strong>02 TTTS</strong></th>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Kratza</td>
<td>12.34</td>
<td>[5.70, 26.73]</td>
</tr>
<tr>
<td>Herberg</td>
<td>13.48</td>
<td>[7.63, 23.83]</td>
</tr>
<tr>
<td>Lopriore</td>
<td>9.67</td>
<td>[3.73, 25.11]</td>
</tr>
<tr>
<td>Subtotal (95% CI)</td>
<td></td>
<td>12.35 [8.17, 18.67]</td>
</tr>
</tbody>
</table>

- Total events: 21 (MC Gestation), 8013 (Singleton Gestation)
- Test for heterogeneity: $\chi^2 = 0.35$, df = 2 ($P = 0.84$), $I^2 = 0\%$
- Test for overall effect: $Z = 11.92$ ($P < 0.00001$)

- Total (95% CI)
- Total events: 41 (MC Gestation), 16026 (Singleton Gestation)
- Test for heterogeneity: $\chi^2 = 7.08$, df = 5 ($P = 0.22$), $I^2 = 29.3\%$
- Test for overall effect: $Z = 11.40$ ($P < 0.00001$)

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*Bahtiyar et al. JUM 26(11):1491-98. 2007*
Fetal Echo - Some Maternal Indications

- Autoimmune antibodies (SSA/Ro, SSB/La)
- Familial inherited disorders (e.g. 22q11.2 del)
- Metabolic disease (e.g. DM, PKU)
- Teratogen exposure (e.g. retinoids, lithium)
- IVF
<table>
<thead>
<tr>
<th>Order</th>
<th>n/N</th>
<th>Frequency</th>
<th>OR Against Historical Data</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>Fyler\textsuperscript{14}</td>
</tr>
<tr>
<td>Total</td>
<td>8/749</td>
<td>10.7 (3.3–18.0)</td>
<td>7.3 (3.6–14.7)\textsuperscript{c}</td>
</tr>
<tr>
<td>Per pregnancy</td>
<td>8/1001</td>
<td>8.0 (2.4–13.5)</td>
<td>5.5 (2.7–11.0)\textsuperscript{c}</td>
</tr>
<tr>
<td>Per fetus</td>
<td>4/512</td>
<td>7.8 (0–15.4)</td>
<td>5.3 (2.0–14.3)\textsuperscript{c}</td>
</tr>
<tr>
<td>Twins</td>
<td>4/222</td>
<td>18.0 (0.5–35.5)</td>
<td>12.5 (4.6–33.5)\textsuperscript{c}</td>
</tr>
<tr>
<td>Per pregnancy</td>
<td>4/444</td>
<td>9.0 (0.2–17.8)</td>
<td>6.2 (2.3–16.5)\textsuperscript{c}</td>
</tr>
<tr>
<td>Per fetus</td>
<td>0/15</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>Triplets</td>
<td>0/45</td>
<td>ND</td>
<td>ND</td>
</tr>
</tbody>
</table>

\textsuperscript{n} indicates number of patients with CHDs; N, total number of patients in the study; and ND, not determined.

\textsuperscript{a}Per 1000 pregnancies or fetuses as appropriate.

\textsuperscript{b}Odds ratio with 95% CI compared with historical data.

\textsuperscript{c}P < .05.

Bahtiyar MO. J Ultrasound Med 2010; 917-922
Timing/Technique

- Usually 18-22 weeks
- Technical limitations (obesity, position, late gestation)
- Optimization of equipment (zoom, frequency, harmonics, narrow field, high frame rate, etc)

AIUM: *Because the heart is a dynamic structure, a complete evaluation can only be made if real-time imaging with acquisition of analog recordings or digital video clips is used a standard part of every fetal echocardiogram.*

- Clips of (at least): 4 chamber, LVOT, RVOT, 3VTV, sag AA/DA with and without Color
Parameters

- Visceral/abdominal situs
- Atria
- Ventricles
- Great arteries
- Atrioventricular junction
- Ventriculoarterial junction
- Heart rate/rhythm
- Cardiac biometry (optional)
- Cardiac function assessment (optional)
Specific Views

- Grayscale
  - 4 chamber view
  - LVOT
  - RVOT
  - 3 vessel and trachea view
  - Short-axis – low for ventricles, high for outflow
  - Long-axis view
  - Aortic arch view
  - Ductal arch view
  - SVC/IVC
1. Four Chamber View
2. Left Ventricular Outflow Tract
3. Right Ventricular Outflow Tract
4. Three Vessels Trachea View
Specific Views

- **Color**
  - Systemic veins – SVC/IVC, DV
  - Pulmonary veins
  - Foramen ovale
  - AV valves
  - Atrial and ventricular septa
  - Semilunar valves
  - Ductal arch
  - Aortic arch
  - Umb vein/artery (optional)
Specific Views

- Pulsed Doppler
  - AV valves
  - Semilunar valves
  - DV
  - Umb vein/artery (optional)
  - Cardiac rhythm disturbance
  - Any structure in which an abnormality on Color Doppler is detected
Upper Abdomen

- Stomach
- Aorta on left
- IVC on the right and more ventral
- Umbilical vein to the left portal sinus
Abdominal Situs Inversus
Four Chamber View

- Heart area ~1/3 of chest area
- Hypoechogenic rim
- Long axis to the left, 45° ± 20°
  - Abnormal axis a/w CHD, esp outflow tract anomalies
  - Abnormal axis a/w chromosomal anomaly
  - Left deviation with gastroschisis/omphalocele

- Position
  - Displacement with CDH, space-occupying lesions (CPAM, etc), lung hypoplasia/ageneis

- RV ≈ LV, RA ≈ LA
- Visualize the crux of the heart
- Examine the interventricular septum
Four Chamber View
Interventricular Septum
Interventricular septum
Differentiating the Ventricles

Right
- Shape – “square”
- Trabeculated
- Moderator band apical
- Papillary muscles attach to interventricular septum
- Tricuspid valve belongs to RV

Left
- Shape – “oval”
- Smooth
- LV forms apex of heart
- Papillary muscles attach to free wall
- Mitral valve belongs to LV
Four Chamber View

Comstock CH. Obstet Gynecol 1987
Ventricles

• Single ventricle
  – HLHS
  – Pulmonary atresia
  – Double inlet
• AV valve attachment
• AV discordance
• Dextrocardia
• Heterotaxy
AV valves
AV valve attachment
Differentiating the Atria

Right
• Anteriorly located
• Receives IVC, SVC, coronary sinus
• Appendage is pyramidal in shape with broad base
• *Posterior portion is smooth, anterior portion is trabeculated*

Left
• Posteriorly located, over the spine
• Receives 4 pulmonary veins
• Appendage is narrow, fingerlike with coarse walls
• Foramen ovale flap into LA
• *Anterior and posterior portions are smooth*
Left atrium
Left atrium
Pulmonary Veins
Right atrium
AV discordance

McEwing & Chaoui, UOG 2004
4 chamber view

- Normal
  - TOF
  - DORV
  - dTGA
  - Truncus arteriosus
  - VSD (malalign, outlet)
  - AV, PV stenosis
  - AV, PV atresia
  - Hypoplastic or interrup AA

- Abnormal
  - Single ventricle variants
  - Complete AVCD
  - ccTGA
  - HLHS
  - VSD (membr)
  - TV, MV atresia
  - Ebsteins
  - RV disproportion (TAPVR, coarct)
Outflow tract views

- RVOT \approx LVOT
- Cross at right angles
- Connection to appropriate vessels
- Opening of valves
- Relationship of great arteries
  - dTGA: Ao ant/rt of PA
  - ccTGA: Ao ant/lt of PA
  - DORV: side by side (or other)
LVOT

- Vessel arising from the LV → Aorta
- Continuity of ventricular septum and aorta
- Post wall of AAo contiguous with ant cusp of MV
- Valve moves freely, not thickened
- 3 head vessels
- Outlet VSDs, conotruncal anomalies
- MV/AV share fibrous continuity
- LV is bullet shaped
- PV/AV not seen in same plane
RVOT

- Vessel arising from the RV → Pulmonary artery
- PA is slightly larger than Ao
- Crosses ascending Ao at ~right angle just above origin
- Branches into RPA (1st), then LPA
Valve integrity
Overriding aorta

1. Narrowing of the pulmonary valve
2. Thickening of wall of right ventricle
3. Displacement of aorta over ventricular septal defect
4. Ventricular septal defect - opening between the left and right ventricles

Tetralogy of Fallot
Four abnormalities that results in insufficiently oxygenated blood pumped to the body
## DDx VSD with great vessel override

<table>
<thead>
<tr>
<th>Condition</th>
<th>Diagnostic clue</th>
<th>Additional signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>TOF</td>
<td>Patent, narrow PA</td>
<td>Antegrade or retrograde flow in DA</td>
</tr>
<tr>
<td></td>
<td>Antegrade flow in PA</td>
<td></td>
</tr>
<tr>
<td>Pulm atresia w VSD</td>
<td>Very narrow PA</td>
<td>DA tortuous with retrograde flow</td>
</tr>
<tr>
<td></td>
<td>No antegrade flow in PA</td>
<td></td>
</tr>
<tr>
<td>Absent pulm valve</td>
<td>Very large PA</td>
<td>No DA generally</td>
</tr>
<tr>
<td></td>
<td>To-and-fro blood flow in PA</td>
<td>Aortic root is more narrow than PA</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>PA arises from the overriding aorta</td>
<td>Valve of the overriding vessel may show regurg</td>
</tr>
<tr>
<td>DORV</td>
<td>PA is overriding and aorta courses in parallel</td>
<td>Mimics TGA with VSD</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Aorta or PA may be of normal size or narrow</td>
</tr>
</tbody>
</table>

Abuhamad & Chaoui, 2010
3 Vessel View

- Number of vessels = 3

- Vessel arrangement (relative position)
  - Left $\rightarrow$ Right = PA, Ao, SVC

- Vessel size
  - PA $>$ Ao $>$ SVC

- Vessel alignment
  - Anterior $\rightarrow$ Posterior = PA, Ao, SVC
3 Vessel Trachea View

- Ductal & aortic arches:
  - are to the LEFT of the trachea
  - form a V as they join the descending aorta

- Nl 4 chamber/Abnl 3V
  - cTGA
  - TOF
  - Pulmonary atresia w VSD

- Abnormal 3VT
  - Coarctation
  - Right aortic arch
  - Double aortic arch
  - Vascular rings
Azygous vein
Persistent LSVC with interr IVC
d-TGA
1. High Short Axis View - Great Arteries

Fetal Heart - Coronal View
Fetal Heart - Sagittal View

2. Low Short Axis View - Ventricles

RV
PA
RA
Ao
Papillary muscle
LV
Short axis view
The 4 second echo
<table>
<thead>
<tr>
<th>Box 1</th>
<th>Cardiac examination check list</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Situs</td>
</tr>
<tr>
<td></td>
<td>Size</td>
</tr>
<tr>
<td></td>
<td>Location</td>
</tr>
<tr>
<td></td>
<td>Axis</td>
</tr>
<tr>
<td></td>
<td>Heart rate and rhythm</td>
</tr>
<tr>
<td>Four chamber</td>
<td></td>
</tr>
<tr>
<td></td>
<td>LA = RA</td>
</tr>
<tr>
<td></td>
<td>LV = RV</td>
</tr>
<tr>
<td></td>
<td>RV has moderator band, anterior</td>
</tr>
<tr>
<td>AV valves</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Two distinct AV valves</td>
</tr>
<tr>
<td></td>
<td>Tricuspid slightly apically displaced</td>
</tr>
<tr>
<td>Interventricular septum intact</td>
<td></td>
</tr>
<tr>
<td>Foramen ovale present</td>
<td></td>
</tr>
<tr>
<td>Foraminal flap opens in to LA</td>
<td></td>
</tr>
<tr>
<td>LVOT</td>
<td></td>
</tr>
<tr>
<td>RVOT</td>
<td></td>
</tr>
<tr>
<td>3-VV/3-VTV</td>
<td></td>
</tr>
</tbody>
</table>

**Abbreviations:** AV, atrioventricular; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract; RA, right atrium; RV, right ventricle; RVOT, right ventricular outflow tract; 3-VTV, 3-vessel trachea view; 3-VV, 3-vessel view.
Box 2
Anomalies can be identified due to concerns during initial fetal heart assessment

1. Situs inversus
2. Abnormal location (shift of the heart with or without axis change)
   a. CDH
   b. Agenesis of fetal lung
   c. Congenital lung abnormalities
3. Axis deviation (extreme left or right axis deviation)
   a. CDH
   b. Outflow abnormalities
4. Cardiomegaly
5. Rate and rhythm
   a. Bradycardia
   b. Tachycardia
   c. Irregular rhythm

Box 3
Anomalies can be identified through basic cardiac examination

Four-chamber view
1. AVSD
2. Hypoplastic left heart syndrome
3. Mitral stenosis
4. Tricuspid atresia
5. Epstein anomaly
6. Hypertrophy
7. Pericardial effusion
8. Intracardiac masses

LVOT
1. Overriding aorta
2. Double outlet RV
3. Aortic valve stenosis
4. Transposition of great arteries
5. Truncus
6. Ventricular septal defect

RVOT
1. Pulmonary valve stenosis
2. Transposition of great arteries
3. Truncus arteriosus

3-VV
1. Vascular ring
2. Right-sided aortic arch
Conclusions

- Levels of examination
- Systematic approach
- Color Doppler
- Referral as indicated
• A systematic approach to fetal heart examination, regular feedback, and implementation of training programs could improve detection rates and in turn neonatal outcome.

• In utero detection of congenital heart disease (CHD) allows possible prenatal interventions.

• In utero detection of CHD improves postnatal outcome.
Thank you