FETAL ECHOCARDIOGRAPHY

Contents:
Scanning methods
View
Congenital heart disease
Fetal arrhythmia
Management and outcome of CHD

Study target
Scanning methods of fetal echocardiography
To know cardinal views
Sonographic characteristics of congenital heart diseases

I. SCANNING TECHNIQUE

A. Preparation:
   1. Use 5-7 MHz, color, Doppler, M mode
   2. Increase frequency
   3. Increase frame rate
   4. Increase contrast
   5. Attach the examining hand to the abdominal surface, then move the probe gently.

B. Sequential segmental analysis (Anderson, 1993)
   1. Divide heart into three parts (atria, ventricles, arteries)
   2. Determine atrial situs with structure
   3. Confirm left and right sides of each parts
   4. Confirm type of connections
   5. Then describe structural abnormalities.

C. Confirm Situs:
   1. Confirm fetal location first, stomach next
   2. Atrial Situs
      - With location of IVC and Aorta in transverse upper abdominal view
2. With Pulmonary vein? drainage

3. Ventricular Situs
   - RV-valve attached to more apex region
     - moderator band

4. Great artery Situs
   1. Aortic arch, Ductal arch
   2. Confirm each branch(PA-2, Ao-3)

D. Description

1. Determination of atrial situs
   - Solitus
   - Inversus
   - Right isomerism
   - Left isomerism

2. Analysis of Atrioventricular(AV) junction
   Connection:
     - Concordant
     - Discordant
     - Anomalous
     - Double Inlet
     - Absent connection
   Mode of Connection:
     - Two valves
     - Common valve
     - Straddling valve
     - Imperforate valve

Ventricular morphology
   - Biventricular heart
   - Univentricular heart

Ventricular Relationships

3. Analyse Ventriculo-Arterial junction
   Connection:
     - Concordant
     - Discordant
Double outlet
Single outlet

Relation:

4. Describe Morphology

II. VIEW

A by Benacerff

- four chamber view
- both outflow tract view

B. Basic echocardiographic views

1. the transverse view of the fetal upper abdomen
2. the 4-chamber view (apical / basal / subcostal)
3. the 3-vessel view (PV/Aorta/SVC)
4. the right / left outflow tract view
5. the aortic arch view (By Yoo) / Ductal arch view (candy cane / hockey stick)
6. the short axis view
7. Vena cava inflow view

C. Ghil View

Four chamber view
Three vessel view

D. Lesions missed on four chamber view

TGA
TOF
VSD
CoA
VSD
IVC interruption
AS, PS
DORV

E. Pitfalls

-thin membranous part; “drop out” in apical 4 chamber view
- difficult small VSD and may close in fetal life
- difficult mild valve stenosis
- impossible secondum ASD
- Ductus arteriosus normally open in fetal life
- mild isthmic narrowing present in fetal life

III. CONGENITAL HEART DISEASE

Incidence; 8 to 9 / 1000 birth

1. Etiology of CHD
   - Chromosomal abnormality 10%
   - Mendelian disorder 5%
   - Multifactorial 90%

2. Specific cardiac defects with chromosomal abnormality
   - Trisomy 21; CHD (40-50%), endocardial cushion defects (m/c)
   - Trisomy 18; CHD (>90%), double outlet of RV, VSD
   - Trisomy 13; CDH (>80%), dextrocardia, VSD, ASD
   - Turner syndrome: coarctation of the aorta

3. Recurrence risk (isolated heart disease)
   One sibling; 3-4%
   Two siblings; 7-10%
   Affected mother : Father
   - AS 13-18% : 3%
   - VSD 6-10% : 1%
   - ASD 4-15% : 1.5%
   - TCF 2.5% : 1.5%
   - CoA 4% : 2%

4. Maternal conditions directly affect fetal cardiac disease
DM
Pheochromocytoma
Hyperthyroidism
SLE
Smoking
Rubella, CMV, Toxo, Syphilis, Listeriosis

Outcomes of prenatally diagnosed CHD
N=1006, Allen et al., JACC, 1994
Chromosomal abnormalities (17%)
TOP (55%)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Nb</th>
<th>% of survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>VSD</td>
<td>52</td>
<td>40</td>
</tr>
<tr>
<td>AV canal defect</td>
<td>154</td>
<td>37</td>
</tr>
<tr>
<td>CoA</td>
<td>104</td>
<td>51</td>
</tr>
<tr>
<td>TOF</td>
<td>27</td>
<td>35</td>
</tr>
<tr>
<td>TOF/PA</td>
<td>11</td>
<td>11</td>
</tr>
<tr>
<td>TGA</td>
<td>20</td>
<td>92</td>
</tr>
<tr>
<td>HPLHS</td>
<td>100</td>
<td>4</td>
</tr>
</tbody>
</table>

#VSD only;
TOP—48%
Chromosomal abnormality—48%
IUD—16%
Neonatal death—15%
Survival—40% in ongoing pregnancy

AV canal defect—die under age of 3 (75%), including Op. Cases

A Fetal dysrhythmia

Embryonic heart beat
Starts embryonic heart beat at 21 to 22 days after conception (5 weeks LMP)
Threshold 5 mm CRL (Goldstein, 1992)
Change of EHR:

Early in 5th LMP --- 85 BPM
Accelerates 3.3 BPM per day to 25 mmCRL (LMP 9 wks)
Peak at 175 BPM at 9 weeks (+ 20 BPM)
Rapid descent until 15 to 18 weeks
Slow or flat until term

*all EHR dropped 24 BPM or more under mean - demise
all EHR upper 95 percentile normal at delivery

2 Conductive System

Contraction 5-6 weeks
A-V node connects to His bundle by 8 weeks
S-A node by 10 weeks
Conduction system mature by 16 weeks

3 Fetal dysrhythmias

Frequency: 1-3%

(n=344, Korea)  (n=1235, Yale experience)

Premature Atrial contraction 70.3%  1095
Premature Ventricular Contraction 9.0%
Complete AV block 7.2%  31
Supraventricular tachycardia (>=180 bpm) 6.7%  60
Sinus bradycardia (<100 bpm) 4.1%  4
Atrial flutter 1.5%  19
Atrial fibrillation 0.6%  2
Ventricular tachycardia 0.6%  3

B. Connection abnormalities at the veno-atrial junction

4 Anomalous pulmonary venous connection
- Pulmonary vein drain to anomalous sites:
  Intracardiac (to coronary sinus)
  Supracardiac (to SVC, innominate vein)
  Infracardiac (to hepatic vein, IVC)
- Sonofindings
  RA, RV dominant
  small LA
  no pulmonary vein draining to LA

C. Connention abnormalities at the atrioventricular junction

1. ASD (complete)
   - defect in the lower part of the atrial septum
   - inlet part of the ventricular septum
   - crux of the heart
   - Sonofindings:
     loss of the normal differential insertion of the two A-V valve

2. Tricuspid atresia
   - no connection between the RA and RV
   - Sonofindings:
     dilated RA, small or no RV
     muscular VSD
     TGV (20%)

3. Mitral atresia
   - small LV
   - asso. With
     atritic atresia in HPHS
     VSD with patent aorta
     VSD with DORV

4. Double inlet ventricle
   - both atrioventricular valves drain to one ventricle
   - 4 chamber view
   - no ventricular septum between the two atrioventricular valves
   - great arteries frequently transposed

5. Ebstein’s malformation
   - attachment of the tricuspid valve (septal, posterior leaflets)
     is displaced into the RV
   - RV outflow tract obstruction common
   - Sonofindings
variable degrees of cardiomegaly with RA, AV dilatation
tricuspid regurgitation
apical displacement of tricuspid valve
usually small pulmonary artery

D. Connection abnormalities at the ventriculo-arterial junction

1. Pulmonary atresia with intact ventricular septum
   - hypertrophied RV
   - tricuspid regurgitation at high velocity

2. Aortic atresia (hypoplastic left heart syndrome)
   - small, or no discernible LV
   - hypoplastic Aao. Aortic arch
   - ass. with chromosomal anomalies

3. Simple transposition of the great arteries
   - no cross over
   - wide sweeping aortic arch

4. TGA with VSD
   - can be mistaken for DORV

5. Corrected transposition
   - RA to LV to PA, LA to RV to Ao
   - reversed A-V valve location in 4 chamber view
   - long, uncomplicated life

6. Tetralogy of Fallot
   - anterior deviation of the aorta
   - VSD
   - infundibular pulmonary stenosis
   - RV hypertrophy
   - normal 4 chamber view
   - abnormal 3 vessel view
     - anterior displaced large aorta
     - posterior displaced small pulmonary artery

7. Common arterial trunk
   - single great artery from the heart
- usually normal 4 chamber view

**E. Other anomalies**

1. **VSD**
   - most common congenital heart disease
   - outlet, perimembranous defect in long axis view

2. **Tumor**
   - rhabdomyoma, only intracardiac tumor in fetus
   - usually multiple, can be single
   - regress spontaneously
   - strongly associated with tuberous sclerosis, particularly multiple

3. **Pericardial effusion**
   - >2mm pathologic
   - karyotyping should be done.

### IV. MANAGEMENT AND OUTCOME

**A. Surgical Management Policy:**
- Biventricular correction vs Univentricular correction (Fontan-type operation)
- Anatomic correction than Functional correction
- Early corrective surgery
- Palliative surgery for Nonreparative lesions
- Preserve Native valves (Valvuloplasty) vs Artificial valve (Valve replacement)
- Heart/Heart-Lung Transplantation

**Management & Outcome of Individual Lesions:**

**B. Patent Ductus Arteriosus:**

**Management:**
- Surgical ligation/division: 1-2yr, anytime in older, neonatal
- Device closure

**Outcome:**
- Surgical mortality: less than 1%
- Complications are rare

**C. Atrial Septal Defect** (Ostium secundum defect):

**Management:**
- Surgical repair in significant L-R shunt: 2-5yr, anytime in older
- Device closure in small defect

**Outcome:**
- May symptomatic after 3rd & 4th decades
- Surgical mortality: less than 1%
- Depends greatly on the age of operation
- Preoperative arrhythmias (atrial flutter/fibrillation, atrial tachycardia, nodal rhythm) frequently persist, even develop postoperatively

**D. Ventricular Septal Defect:**

**Management:**
- Surgical closure: elective at 6mo-2yr, anytime in older
  - Any age under 6mo in baby with medically unresponsive CHF/poor weight gain

**Outcome:**
- Surgical mortality: less than 1-3%
- Natural history: a. Spontaneous closure in 30-50%
  - Large defects tends to become smaller with age
  - Infundibular stenosis may develop
  - Pulmonary vascular obstructive disease (PVOD)
  - Prolapsing aortic regurgitation (AR) in subarterial (oriental) defect

**E. Atrioventricular Septal Defects:**

- Complete AVSD (30% occur in Down's syndrome)

**Management:**
- Corrective surgery at 6-12yr

**Outcome:**
- Surgical mortality: 5-10%
  Complications: Persistence or worsening of MR (10%)
  Complete heart block & supraventricular arrhythmia

- Natural History & Complications:
  a. Heart failure occurs 1-2 mo after birth
  b. Recurrent pneumonia common
  c. Majority die in 2-3 yrs without surgery
  d. Survivors develop PVOD and die in late childhood or as young adults

2. Partial ASD
   Management:
   - Surgical repair: elective at 2-4 yr, anytime in older
     Earlier in infants with medically unresponsive CHF or MR

   Outcome:
   - Surgical mortality: 1-2%
     Complications: Persistence or worsening of MR (10%)
     Atrial or nodal arrhythmia

F. Pulmonary Stenosis:
   Management:
   - Balloon valvuloplasty at any age
   - Urgent surgical valvotomy in some cases with critical PS or Pulmonary atresia

   Outcome:
   - Mortality: 0-1% in older children, Higher in critically ill infants
   - Pulmonary Atresia with intact ventricular septum
     Hypoplastic RV (may need Fontan-type op).
     Severe and progressive cyanosis since birth
     Exceedingly poor prognosis; 50% die in 1 mo, 85% by 6 mo without Tx.

G. Coarctation of the Aorta:
Management:
- Surgical repair: Asymptomatic children – elective at age 3-4yrs
  Symptomatic infants – need intensive anticongestive measures and PGE1, then urgent op at diagnosis
- Balloon angioplasty, esp in Recoarctation

Outcome:
- Surgical mortality: Less than 1% in older, less than 5% in infants
- Complications: Recoarctation
  Persistence of hypertension
  Postop renal failure, postcoarctectomy syndrome - rare

- Natural History & Complications:
  Asymptomatic Children:
  a. LV failure may develop in adult life
  b. Bicuspid aortic valve: may have ASR
  c. Infective endocarditis
  d. Intracranial bleeding, hypertensive encephalopathy or cardiovascular disease

Symptomatic Infants:
  a. Heart failure occurs in over 80% by 3 mo of age
  b. Early death from CHF and renal shutdown

H. Valvar Aortic Stenosis:
Management:
- Surgical valvotomy, valve replacement, Ross operation
- Balloon valvuloplasty
  At any age in infant with CHF from critical AS

Outcome:
- Mortality: 15-20%
  Higher mortality in sick neonates (as high as 50%)
  In older children: 1-2%
- Complications: Significant AR, Residual AS, Infective endocarditis, CHF
  Only about 25% of the patients survive event free for 20 years
I. Tetralogy of Fallot:
Management:
- Total surgical correction: Elective in 6-24 mo of age
  Shunt op. In hypoxenic infants with small pulmonary artery
Outcome:
- Surgical mortality: under 5%
- Complications: Pulmonic valve regurgitation
  Ventricular arrhythmia & sudden death (2-5%)

J. Complete Transposition of the Great Arteries:
Management:
- Arterial switch operation: within 2 weeks of age
  - Balloon septostomy
Outcome:
- Death in 90% under 6 mo, without intervention
- Surgical mortality 0-15% with reasonable long term prognosis

K. Total Anomalous Pulmonary Venous Connection:
Management:
- Urgent or emergency surgery at any age,
  esp. in cases with pulmonary venous obstruction
Outcome:
- Mortality: high (10-25%), esp. in infracardiac type with PV obstruction
- Reasonably good long term prognosis in postop. Survivor without PV obstruction
- Most patients die by 2 mo of age without surgery

L. Ebstein’s Anomaly:
Management:
- Tricuspid annuloplasty, TV replacement, Fontan procedure
  in cases with severe limitation, cyanosis and CHF
Outcome:
- Extremely variable natural history, depends on TV dysfunction and RV hypoplasia
- 30% die before the age of 10yr.
- Attacks of PAT are common

**Single Ventricle / Tricuspid Atresia**

Management:
- Fontan-type procedure
- Shunt op or PA banding as palliative procedure

Outcome:
- 50% of patients die by 1 yr of age
- CHF, PVOD or cyanosis; depends on the associated problems

**Hypoplastic Left Heart Syndrome**

Management:
- Norwood procedure (3 stage op) with extremely high mortality and morbidity
- Heart transplantation

Outcome:
- Progressive CHF, hypoxemia and acidosis,
  resulting in death, usually in the 1st month of life