

# 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

## **.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.

-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

Ambiguous

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

.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 secundum ASD
- Ductus arteriosus normally open in fetal life
- mild isthmic narrowing present in fetal life

## . CONGENITAL HEART DISEASE

Incidence ; 8 to 9 / 1000 birth

Etiology of CHD

- Chromosomal abnormality 10%
- Mendelian disorder 5%
- Multifactorial 90%

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

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%
- TOF 2.5% :1.5%
- CoA 4% :2%

Maternal conditions directly affection 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%)

	No	% of survival
VSD	52	40
AV canal defect	154	37
CoA	104	51
TOF	27	35
TOF/PA	11	11
TGA	20	92
HPLHS	100	4

#VSD only;  
TOP—48%  
Chromosomal anomaly-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 dysarrhythmia

Embryonic heart beat  
;Starts embryonic heart beat at 21 to 22 days after conception  
(5weeks LMP)  
threshold 5mm CRL( Goldstein,1992)

Change of EHR:

Early in 5<sup>th</sup> LMP---85 BPM

Accelerates 3.3 BPM per day to 25 mm CRL(LMP 9 wks)

Peak at 175 BPM at 9 weeks(+/- 20BPM)

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

Conduction 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

Fetal dysarrhythmias

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(>180bpm) 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

Anomalous pulmonary venous connection

-pulmonary vein drain to anomalous sites:

intracardiac(to coronary sinus)

supracardiac(to SVC, innominate vein)

inreacardiac(to hepatic vein, IVC)

- Sonofindings
  - RA, RV dominant
  - small LA
  - no pulmonary vein draining to LA

### C. Conduction abnormalities at the atrioventricular junction

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

Tricuspid atresia

- no connection between the RA and RV

-sonofindings:

dilated RA, small or no RV

muscular VSD

TGV (20%)

Mitral atresia

- small LV

-asso. With

atrial atresia in HPLHS

VSD with patent aorta

VSD with DORV

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

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

Pulmonary atresia with intact ventricular septum

- hypertrophied RV
  - tricuspid regurgitation at high velocity
- Aortic atresia(hypoplastic left heart syndrome)
- small, or no discernible LV
  - hypoplastic Aao. Aortic arch
  - ass. with chromosomal anomalies

Simple transposition of the great arteries

- no cross over
  - wide sweeping aortic arch
- TGA with VSD
- can be mistaken for DORV

Corrected transposition

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

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

Common arterial trunk

- single great artery from the heart

-usually normal 4 chamber view

#### E. other anomalies

##### VSD

- most common congenital heart disease
- outlet, perimembranous defect in long axis view

##### Tumor

- rhabdomyoma, only intracardiac tumor in fetus
- usually multiple, can be single
- regress spontaneously
- strongly associated with tuberous sclerosis, particularly multiple pericardial effusion
- >2mm: pathologic
- karyotyping should be done.

#### .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 3<sup>rd</sup> & 4<sup>th</sup> 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%  
b. Large defects tends to become smaller with age  
c. Infundibular stenosis may develop  
d. Pulmonary vascular obstructive disease (PVOD)  
e. Prolapsing aortic regurgitation (AR) in subarterial (ventricular) 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 1-2mo after birth

b. Recurrent pneumonia common

c. Majority die in 2-3yrs without surgery

d. Survivors develop PVOD and die in late childhood or as young adults

#### . Partial AVSD

Management:

- Surgical repair: elective at 2-4yr, 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 hypertention  
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 encephaloapthy 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 aritical AS

Outcome:

- Mortality: 15-20%  
Higher mortality in sick neonates (at 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 hypoxemic 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

#### M. 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

#### N. Hypoplastic Left Heart Syndrome:

##### Management:

- Norwood procedure (3stage op) with extremely high mortality and morbidity

- Heart transplantation

##### Outcome:

- Progressive CHF, hypoxemia and acidosis,  
resulting in death, usually in the 1<sup>st</sup> month of life