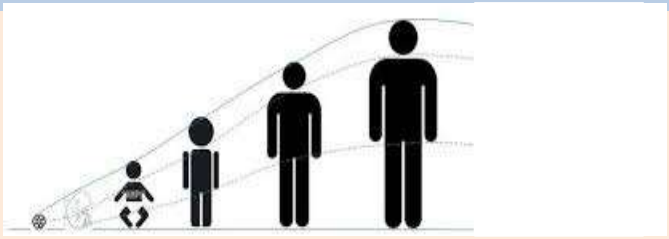
Ebstein Anomaly from fetus to adult



By

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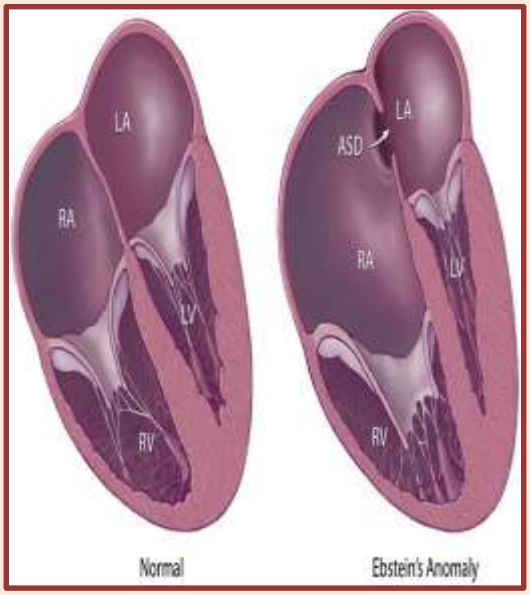
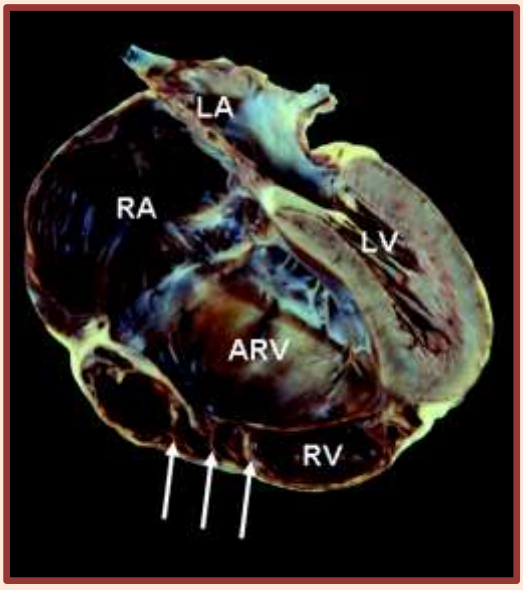
Ebstein ' s Anomaly

- It is a rare entity (incidence 0.3-0.5 % of congenital cardiac malformations

Ebstein's anomaly is the anterior-inferior displacement of the septal & posterior leaflets of the TV into the inlet portion of RV.





Ebstein ' s Anomaly
has a wide range of
clinical presentations

fetal

neonatal

childhood

Adult

So .....

Ebstein ' s Anomaly
has a wide range of
patients' outcome

fetal

neonatal

childhood

Adult

Extremely variable….!

Fetus with poor outcome

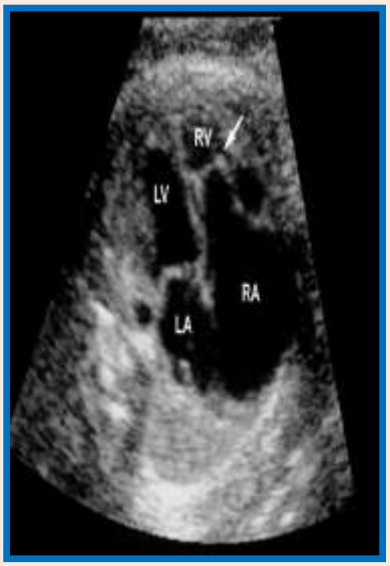
Asymptomatic adult

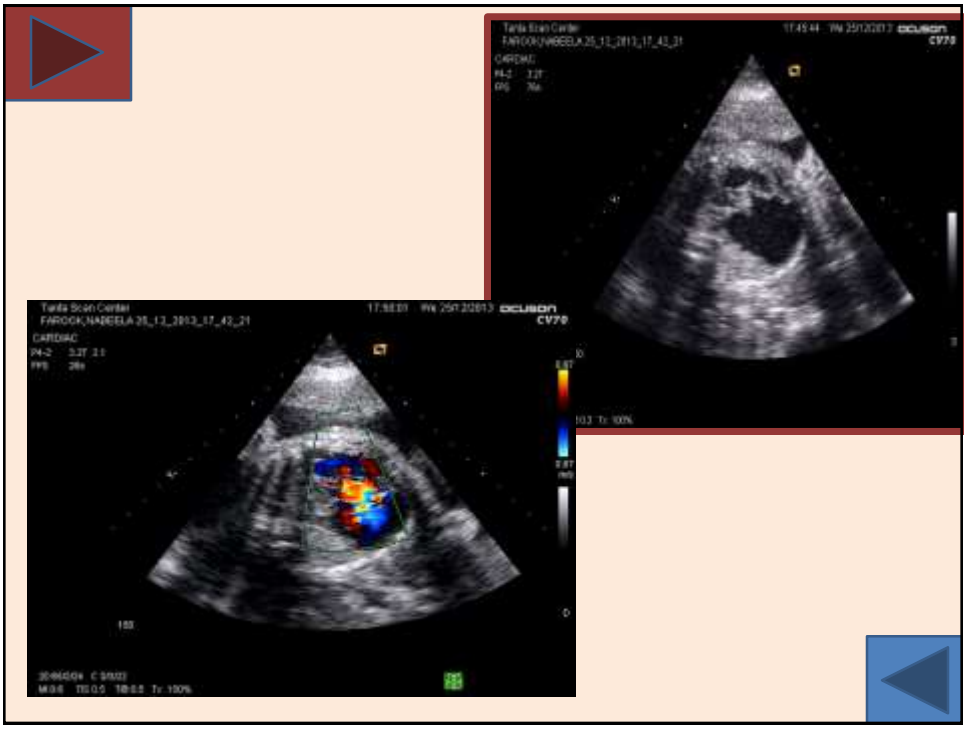
Fetal presentations

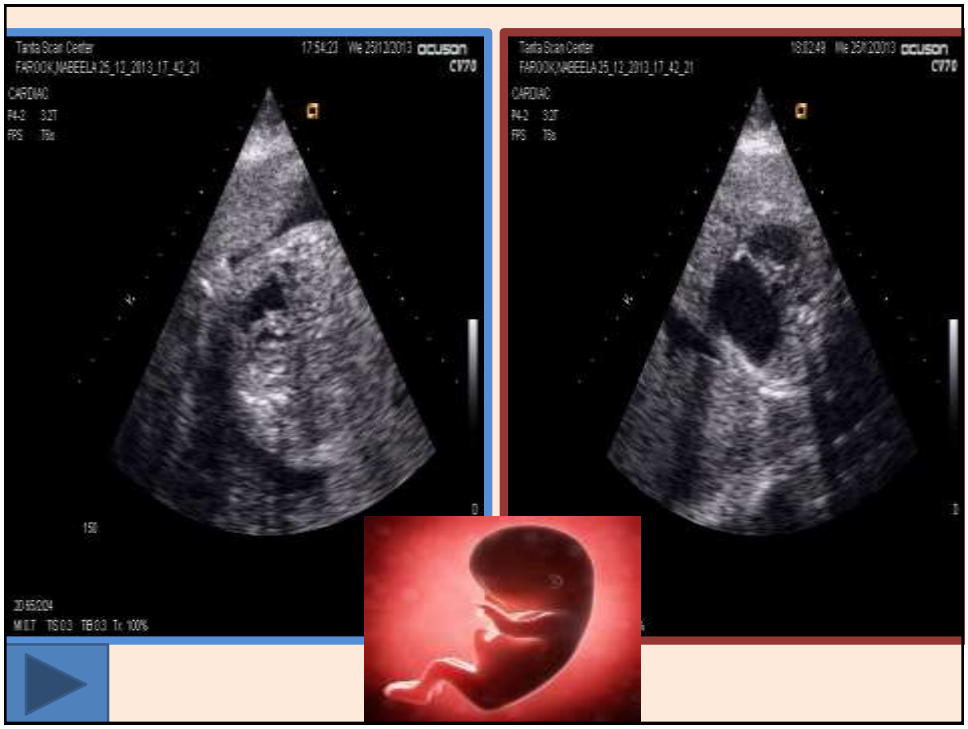
EA can be diagnosed in mid to late pregnancy

Fetal presentations

- Abnormal 4 CH view
- Fetal arrhythmias
- Hydrops fetalis.



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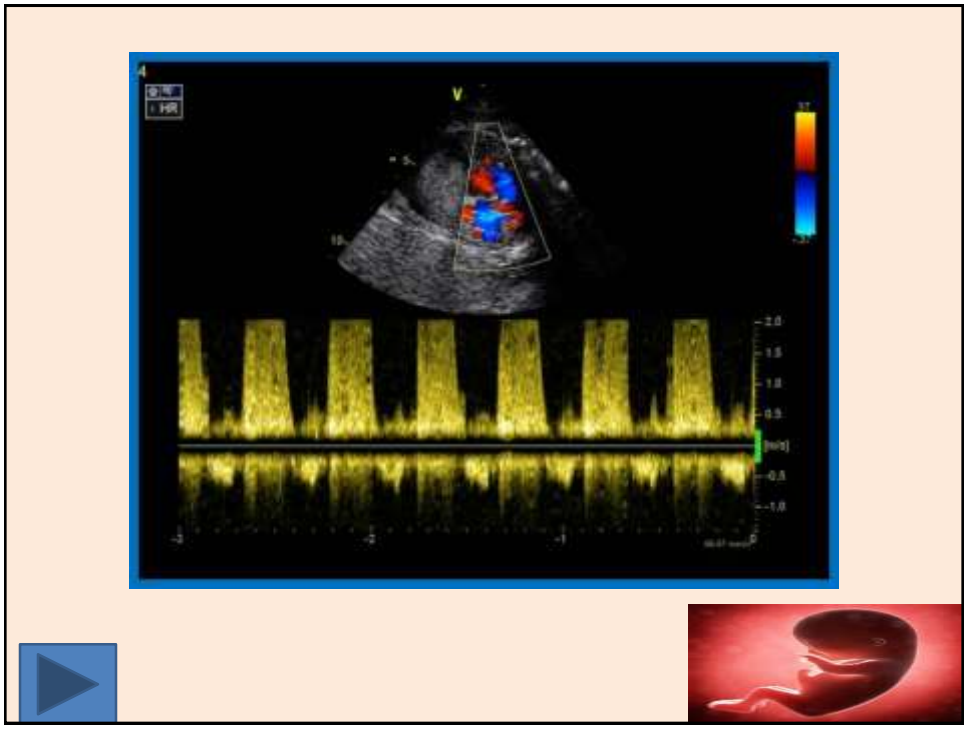

Essentials of fetal diagnosis of EA

- Apical displacement of tricuspid valve septal and mural leaflets
- Long anterior tricuspid leaflet
- Variable degrees of TR (PWD).
- Variable degrees of RA enlargement
- Cardiomegaly (often massive)
- Pulmonary stenosis ,hypoplasia or atresia.

The degree of displacement.....

- In the first trimester: it is not possible to detect any offset between tricuspid &mitral valves in normal fetus.
- with progression of gestation, the distance gradually increases in normal hearts
- Abnormal displacement is variable from between (5-20 mm) as published in different fetal series

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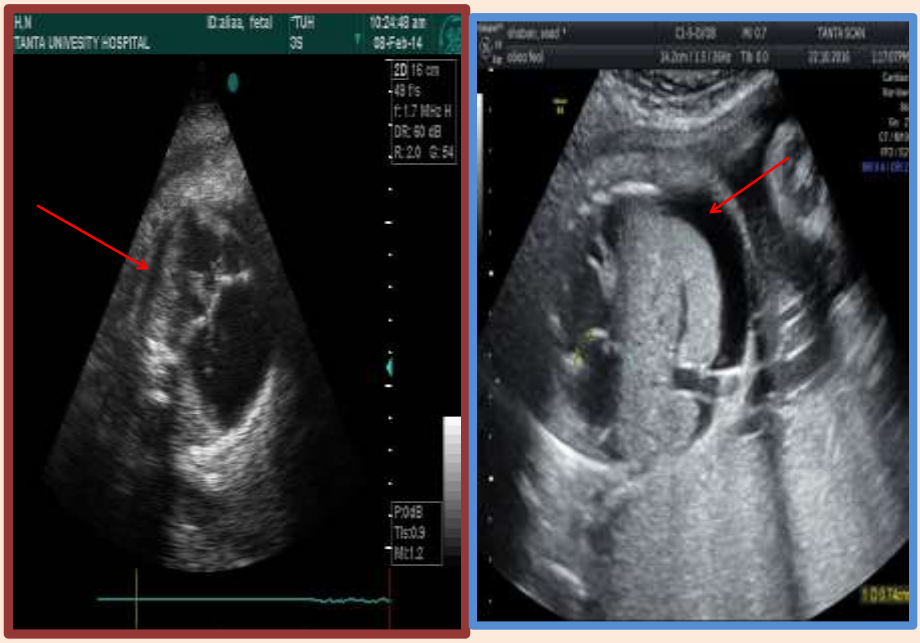
Fetal diagnosis of complications

- Hydrops fetalis
- Fetal arrhythmias (premature beats, SVT)
- Functional pulmonary atresia. (indicates poor RV function)



- Even in mild forms of the disease, serial monitoring is mandatory for the development of arrhythmias and effusions.

Associated Anomalies with EA

Cardiac

- VSD
- TOF
- AVSD
- Ao Coarc
- TGA

Extra cardiac

- craniofacial
- Digital

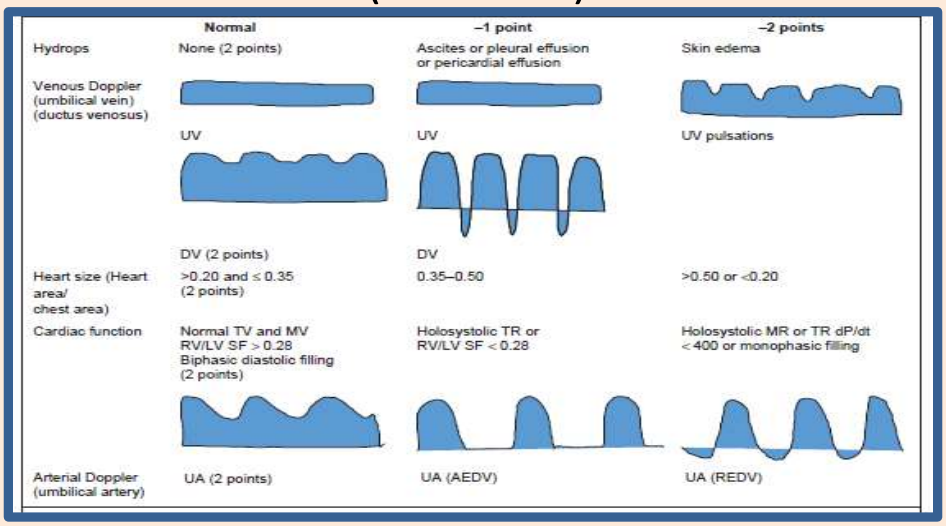
Prognosis

Fetal echo features of poor prognosis in EA

- Tethered distal attachment of the anterior leaflet (three or more accessory attachment between the valve and the RV wall)
- Dysplasia of the arterialized portion of the RV (decreases wall thickness &RV dyskinesia)
- Marked RA dilatation (RA > Functional RV+LV+LA)
- LV compression with narrowing of the LVOT.

Prognosis

Fetal Cardiovascular profile score (CVP& EA)



	Normal	–1 point	–2 points
Hydrops	None (2 points)	Ascites or pleural effusion or pericardial effusion	Skin edema
Venous Doppler (umbilical vein) (ductus venosus)	UV DV (2 points)	UV DV	UV pulsations
Heart size (Heart area/ chest area)	>0.20 and ≤ 0.35 (2 points)	0.35–0.50	>0.50 or <0.20
Cardiac function	Normal TV and MV RV/LV SF > 0.28 Biphasic diastolic filling (2 points)	Holosystolic TR or RV/LV SF < 0.28	Holosystolic MR or TR dP/dt < 400 or monophasic filling
Arterial Doppler (umbilical artery)	UA (2 points)	UA (AEDV)	UA (REDV)

Prognosis

Fetal Cardiovascular profile score (CVP& EA)

- It is an echo bases approach for assessment of fetal hemodynamics.
- It has been reported that **CVP< 7** is associated with poor outcome in fetus with EA.

Prognosis

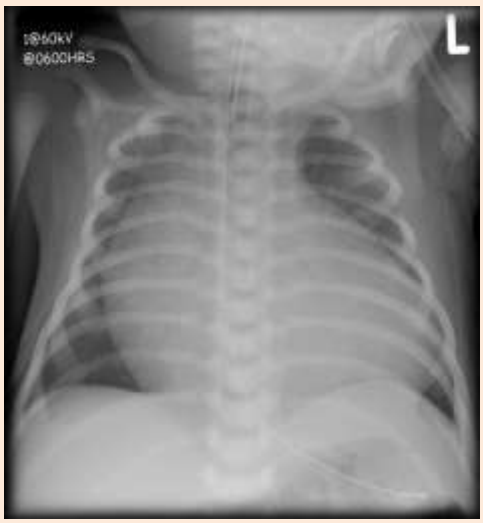
- fetal survival is linked to the ability of the fetal heart to increase the left ventricular volume flow.
- Therefore, the size of the fossa ovalis allowing the required increase of right-to-left shunt and a sufficient left ventricular diastolic and systolic function are mandatory in fetuses with EA.

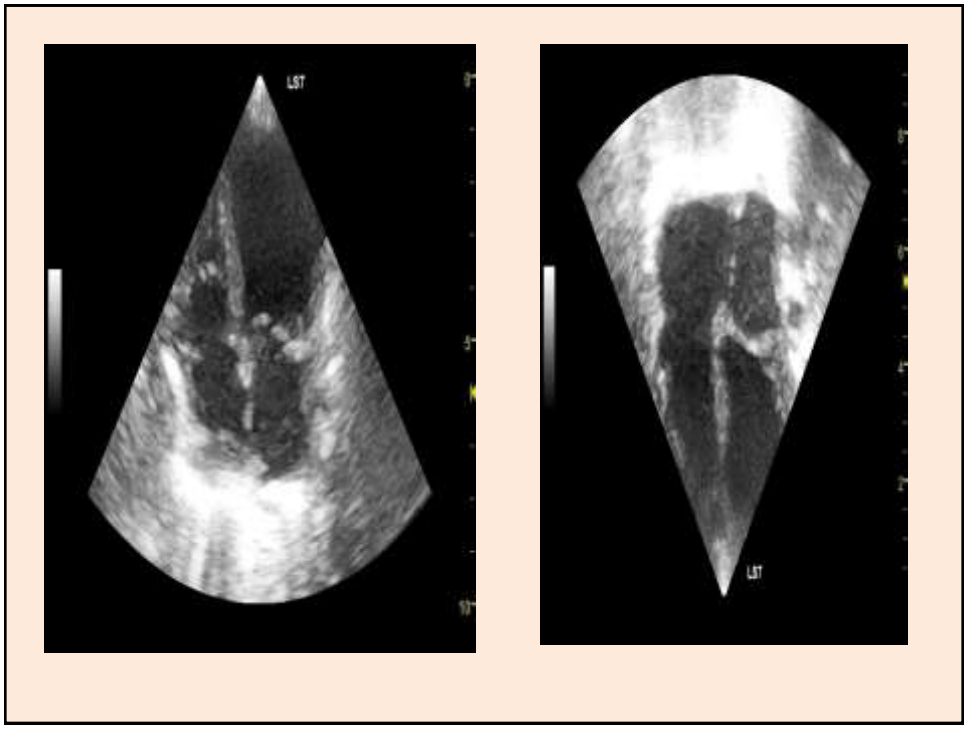
Parent counseling
(Addressing the bad news)

- Most cases of EA that have been diagnosed pre natally are associated with poor outcome.
- There is significant risk of developing intrauterine heart failure causing spontaneous IUFD.
- Survivors to delivery also have high neonatal mortality.
- Cases with mild cardiomegaly ,mild TR may progress to a more sever form thought gestation

Neonatal presentation of EA

- Pulmonary vascular resistance is high immediately after birth.
- Severe TR.
- Right to left shunt across ASD
- Severe cyanosis
- Dysfunctional RV.
- Survival will depend on adequacy pulmonary blood flow.(PGE)

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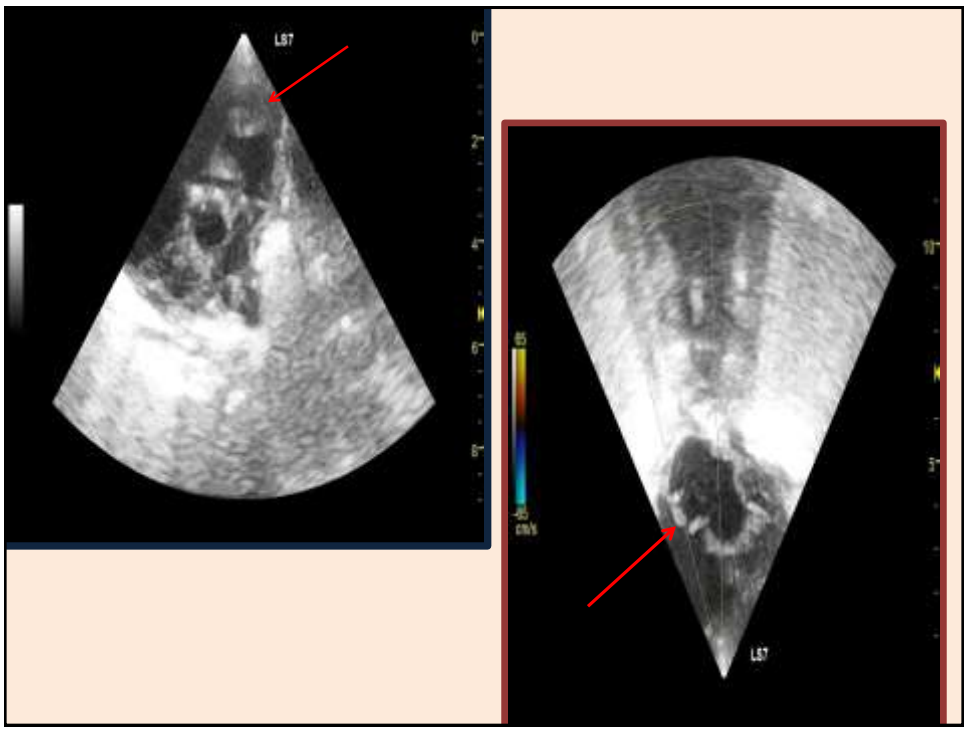
LST
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Neonatal Echo signs of poor prognosis

- Marked cardiomegly
  - Impact on Lung Mechanics
  - Celermajer Index (apical 4CH):

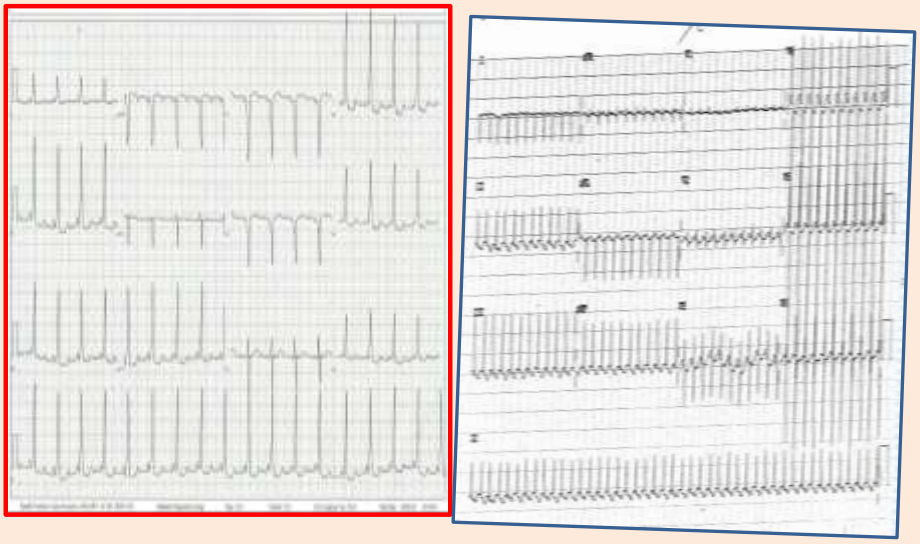
Ratio of areas (RA+aRV to RV+LA+LV) Value > 1 suggests a poor prognosis in the newborn.

- High volume of TR.

Childhood presentations of EA

- Murmur of tricuspid regurgitation.
- Palpitations, chest pain or syncope due to tachyarrhythmias (WPW)

- Treatment: medically treat or ablate WPW pathway .
- Follow conservatively with echo
- Surgery :



Current surgical indications of EA

- 1. Symptoms or reduction in exercise tolerance
- 2. Cyanosis if ASD or FOP are present
- 3. Arrhythmias: New onset or worsening.
- 4. Progressive RV dilation or dysfunction (CMR)

Surgical repair be performed between 2 and 5 years of age.

Surgical options in EA

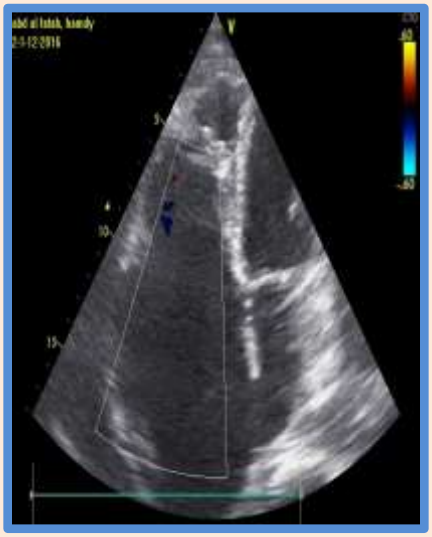
- Tricuspid valve repair.
- Tricuspid valve replacement
- Atrial septal defect (ASD) closure.
- Bidirectional Glenn procedure ("1.5 repair")
- Heart transplant.

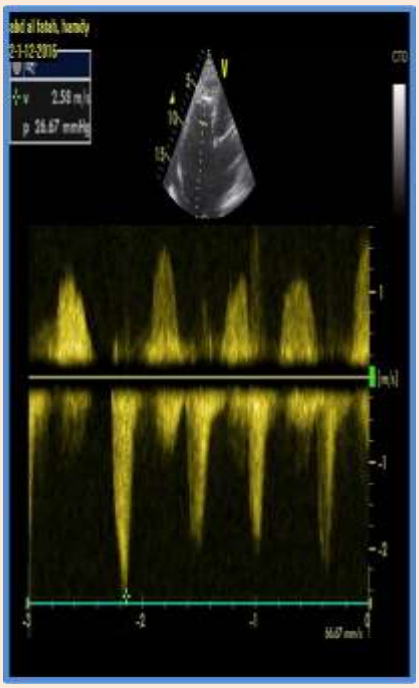
Adult presentation of EA

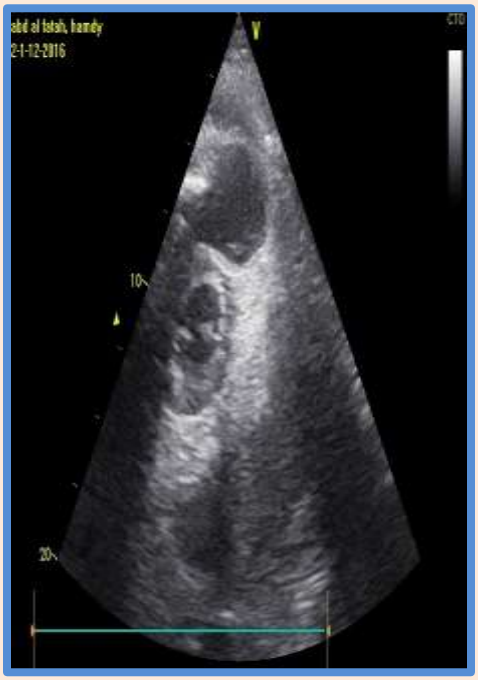
Similar to childhood presentation.

- Fatigue with exercise.
- Mild cyanosis due to ASD shunt (R→L)
- Murmur of tricuspid regurgitation .
- Tachyarrhythmias (WPW).
- Right sided heart failure.

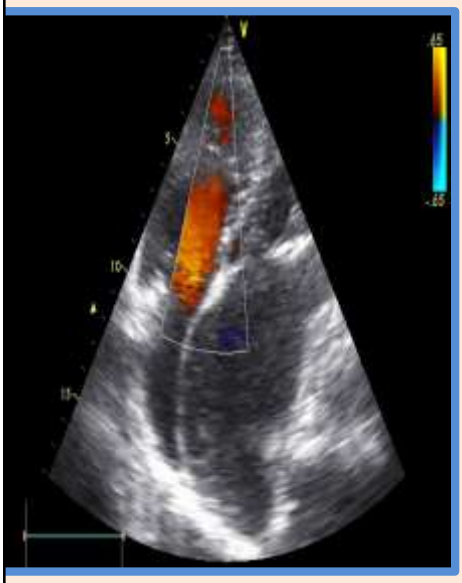
Patient 1

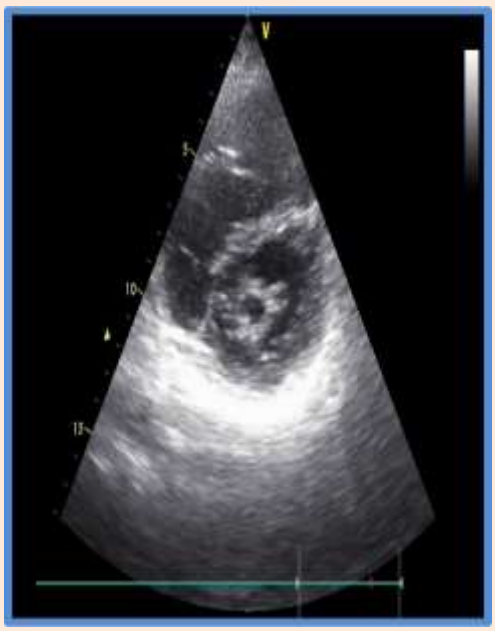


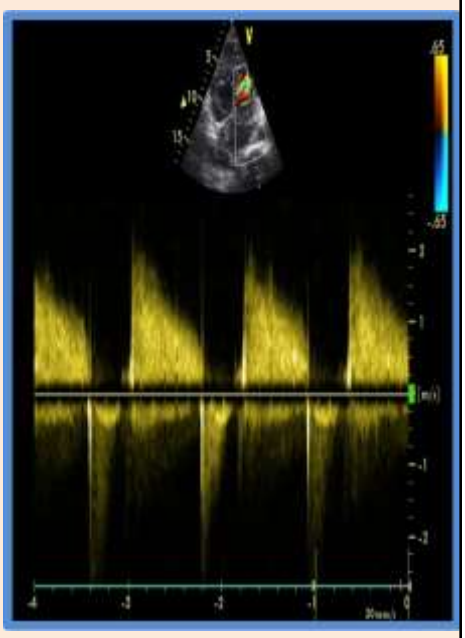




Patient 2







Take Home Message

- Ebstein's anomaly of the TV is rare complex anomaly with broad spectrum of clinical presentations
- Management of EA should be **individualized**
- The management of Ebstein's anomaly depends on the age at presentation, anatomic severity of the lesion, and clinical features

Take Home Message

- Fetuses diagnosed with EA have grim prognosis.
- Neonates with severe EA require early surgical care with higher rates of re-operation.
- Asymptomatic children/adults can be monitored and expect normal life expectancies and low-normal exercise ability.


Thank
You