Ebstein Anomaly from fetus to adult

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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

So ......

Ebstein’s Anomaly has a wide range of patients’ outcome
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.
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.
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)

- It is an echo bases approach for assessment of fetal hemodynamics.
- It has been reported that $\text{CVP} < 7$ 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 prenatally 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 severe 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)
Neonatal Echo signs of poor prognosis

- Marked cardiomegaly
  - 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