Focused patient history and workup

- Ex-32w1d F neonate delivered via C-Section due to reversal of umbilical artery end-diastolic flow (REDF)
- Cyanosis: peripheral > central
- Moderate chest retractions
- Holosystolic blowing murmur obscuring S1 and S2
- CPAP +5 was applied for respiratory support given retractions
- Transferred to NCCC for further management with plan to intubate depending on saturations and work of breathing
DDx

**Cardiovascular**
- Pulmonary atresia with intact ventricular septum (PAIVS)
- Ebstein’s anomaly
- Truncus arteriosus
- Transposition of great vessels
- Tricuspid atresia
- Tetralogy of Fallot
- TAPVR
- Pericardial effusion

**Respiratory**
- Neonatal respiratory distress syndrome

**Infectious**
- Infectious myocarditis
- Congenital rubella
List of imaging studies

- XR Chest Portable
- Echocardiogram
XR Chest Portable

AIRWAYS
- Midline trachea
- Visible carina
- (R) w/o atelectasis
- (L) grossly obscured

BONES
- No acute osseous abnormalities
EFFUSIONS
- No effusions or pneumothoraces

DIAPHRAGM
- Mild lung hyperinflation
- Gastric bubble present

CARDIAC SILHOUETTE AND MEDIASTINUM
- Cardiomegaly
- “Wall-to-Wall” heart
LUNG FIELDS
- No focal opacities
- Sparse vasculature

Endotracheal tube

Umbilical Venous Catheter

EKG leads

Enteric tube with tip overlying the gastric fundus
IMPRESSSION:

1. Massive cardiomegaly

2. Endotracheal tube terminating approximately 0.5 cm above the carina, consider slight retraction
PATIENT FINDINGS

1. Severe dilation of right atrium
2. Severe apical displacement of the tricuspid valve leaflets into RV
3. Moderate to severe tricuspid valve regurgitation
4. “Atrialized,” hypoplastic RV
5. Patent foramen ovale with a moderate-size right-to-left shunt
6. Tortuous patent ductus arteriosus, left to right shunt, large and unrestrictive

IMAGE CAPTION: Apical 4 chamber, 2 -dimensional echocardiogram shows Ebstein's anomaly of the tricuspid valve. There is displacement of the tricuspid valve toward the apex of the right ventricle (RV) and a patent foramen ovale (PFO). LA: left atrium; LV: left ventricle; RA: right atrium.
Patient treatment and outcome

- Patient was intubated and remained in NCCC
- Developed bradycardia with poor perfusion and pulseless electrical activity
- CXR maintained baseline enlarged cardiac silhouette without pulmonary effusions or pneumothoraces
- Patient expired
- Primary Cause of Death: Ebstein’s anomaly
- Secondary Cause of Death: Cardiac arrhythmia
Discussion: Ebstein’s anomaly

- Congenital malformation
- Displacement of malformed tricuspid valve leaflets into the RV
- Enlarged RA
- “Atrialized” hypoplastic RV
- Moderate to severe tricuspid regurgitation
Discussion: Ebstein’s anomaly

- Unknown etiology
- Estimated risk in the general population is 1 in 20,000 live births with no gender predilection
- Associated with maternal lithium use during the first trimester of pregnancy
- Clinical presentation varies widely, ranging from critically ill neonates to asymptomatic adults
Discussion: Ebstein’s anomaly

- Routine Imaging of Asymptomatic Children and Adults
  - ECG, Doppler transthoracic Echo, Cardiac MRI

- Medical Management of Symptomatic Children
  - Alprostadil (prostaglandin E1) to keep the ductus arteriosus open and improve oxygenation
  - Nitric oxide to decrease PVR and improve antegrade blood flow through the pulmonary artery
  - Milrinone for its inotropic and pulmonary vasodilator effects

- Surgical Intervention
  - Tricuspid valvuloplasty (widening/ballooning) or replacement, selective plication of the atrialized right ventricle (folding it back to expand the LV), closure of intracardiac shunts (i.e. ASD), right reduction atrioplasty, and any indicated arrhythmia procedures
Arrhythmias are amongst the most feared complications for patients with Ebstein’s anomaly.

In these patients, congenital and acquired substrates for arrhythmias coexist and can manifest at different disease stages.

Many arrhythmias are based on accessory pathways located along the anomalous atrioventricular valve (TV).

Associated arrhythmias may include atrial ectopic tachycardia (AET), atrial flutter, atrial reentry tachycardia (ART), atrial fibrillation, WPW and ventricular tachyarrhythmia.
1. Ebstein’s anomaly is a congenital malformation characterized by malformed and displaced tricuspid valve leaflets and a “wall-to-wall” heart on chest radiograph

2. Although many patients with Ebstein’s anomaly are asymptomatic, surgical and/or medical intervention are critical if symptoms develop

3. Cardiac arrhythmias are amongst the most feared complications for patients with Ebstein’s anomaly


