**EBSTEIN’S ANOMALY**


**Take home points:**
1. Ebstein’s anomaly results in an abnormally placed tricuspid valve and “atrialization” of the RV with decreased RV outflow
2. 80% of patients with Ebstein’s anomaly have R→L shunt(s) so cyanosis usually occurs
3. In adults with Ebstein’s anomaly, Eisenmenger’s syndrome and WPW can be complications

**Anatomy of Ebstein's anomaly: a problem with the tricuspid valve apparatus**
- The septal leaflet (and often posterior leaflet) of the tricuspid valve are displaced into the RV
- The anterior leaflet is abnormal, large, and adherent to the RV free wall
- A large portion of the RV is “atrialized” so that the functional RV is very small
- Tricuspid valve usually regurgitant (but may be stenotic)
- 80% of patients have associated ASD and/or PFO through which R→L shunting may occur

**Clinical manifestations:**
- Presents with cyanosis very early in life due to R→L shunt
- Differential diagnosis of cyanotic congenital heart disease: remember the T’s…
  - Tetralogy of Fallot, Transposition of the great vessels, Total anomalous pulmonary venous return, Tricuspid problems (Ebstein’s anomaly, tricuspid atresia)
- Severity of hemodynamic derangements depends on how much of the RV is left functional; if the anomaly is minor, patients may present later in life (adulthood) with Eisenmenger’s syndrome
- Predictors of outcome in adults with Ebstein’s anomaly:
  - NYHA functional class
  - Heart size
  - Presence or absence of cyanosis
  - Presence or absence of paroxysmal atrial tachycardias
- Physical findings: TR murmur, hepatomegaly, widely split S2
- ECG findings: tall, broad P waves, RBBB, 1°AVB; 20% of patients have WPW so look for delta wave
- CXR findings: cardiomegaly (due to huge RA), decreased pulmonary vascular markings (R→L shunt)

**Management:**
- In adults, definitive therapy involves repairing the tricuspid valve
- Medical management includes prophylaxis for endocarditis, diuretics, and digoxin
- Treat the atrial arrhythmias (catheter ablation with EP study)