Ebstein's Anomaly

What is it?

- An abnormally formed tricuspid valve with apically displaced leaflets.
- Is a relatively rare disease.
- Can be associated with lesions such as ASD, PFO, VSD, PS, pulmonary atresia, ToF or Coarctation.
- More common if the mother has used lithium or benzodiazepines during pregnancy.

How is it diagnosed?

- Can present with trivial symptoms or profound cyanotic heart defect.
- Symptoms are arrhythmias, dyspnoea, fatigue, poor exercise tolerance, chest pain and/or cyanosis.
- Echocardiogram.
- Cardiac MRI
- Chest X-ray
- Auscultation and physical exam
- ECG may show right atrial hypertrophy, prolonged PR interval, low voltage and supraventricular and ventricular arrhythmias

How does it affect the heart?

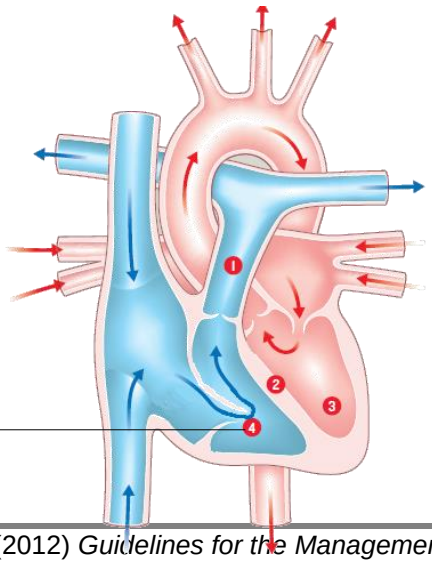
- Abnormally formed tricuspid valve.
- Tricuspid valve often regurgitant.
- Frequently associated anomalies include a shunt at the atrial level, ASD or PFO
- Can result in chronically low systemic cardiac output.

What is the long term management?

- Oral anticoagulation therapy for patients with history of paradoxical embolism or AF.
- Symptomatic rhythm disorder treated with EP intervention.
- Tricuspid valve repair or replacement is possible.
- Normally lead active lives except for extensive static sport to competition level.
- Asymptomatic females may tolerate pregnancy well. Higher risk in presence of cyanosis, serious arrhythmia and right heart failure.

What is the follow up?

- Annual cardiac follow up.
- Look for persisting or new tricuspid regurgitation, usual valve replacement complications, failure of RV or LV, residual atrial shunts, arrhythmias and higher grade heart blocks.



Displaced tricuspid valve

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Reference: The European Society of Cardiology (2012) *Guidelines for the Management of Grown-Up Congenital Heart Disease.* Oxford University Press: Oxford.