Ebstein's Anomaly

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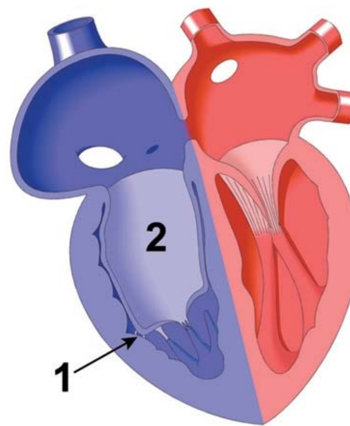
I. Embryology
- A. Formation of the normal tricuspid valve.
  1. Three leaflets (anterior, septal, and posterior) are derived from the right ventricular wall.
- B. Abnormal process in Ebstein's anomaly
  1. The septal and posterior leaflets remain tethered to the right ventricular muscle.
  2. These valve leaflets are displaced apically.

II. Anatomy
- A. Tethering of leaflet tips due to non-delamination from the RV muscle.
  1. Apical displacement of the septal and posterior leaflets of the tricuspid valve (Number 1 in Illustration)
  2. Divides the atrium into two parts
     - a. Normal right atrium
     - b. 'Atrialized right ventricle' - the area above the displaced tricuspid leaflets (Number 2 in Illustration)
       - (1) Anatomically right ventricle (RV)
       - (2) Functionally right atrium (RA)



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- B. Atrial septal defect (ASD)/Patent foramen ovale (PFO)
  1. PFO does not close in the presence of high right atrial pressure.
  2. ASD present in approximately 30% of cases
- C. Tricuspid regurgitation (TR)
- D. Abnormal left ventricular muscular wall (Non-compaction) seen in 18% of patients

III. Physiology

A. High Right Atrial (RA) pressures

1. Results from tricuspid regurgitation increasing the volume in the right atrium
2. Prevents closure of the PFO
3. Excessive RA pressure leads to right to left shunting at the atrial level across the PFO or ASD. May result in:
   a. Hypoxia
   b. Cyanosis with exercise or at rest
   c. Clubbing of digits
   d. Increased risk of paradoxical embolus.

B. Right Ventricular dilation

1. Secondary to volume overload from TR
2. Results in right bundle branch block

C. Electrical Conduction Disturbances

1. Wolff Parkinson White Syndrome seen in 20% of patients

   WPW syndrome is a rare congenital heart disease due to an accessory pathway between the atrium and the ventricle. Its ECG pattern is characterized by a short PR interval, a delta wave, a wide QRS complex and an abnormal ventricular repolarisation. Patients are usually asymptomatic, or have frequent paroxysmal episodes of tachycardia. These tachycardias are supraventricular tachycardias related to either reentry between the AV node and the accessory pathway, or atrial tachyarythmias descending through this bypass tract, or both. In the latter clinical situation, sudden cardiac death may occur in case of short refractory period within the accessory pathway. (Milliez P, Slama)

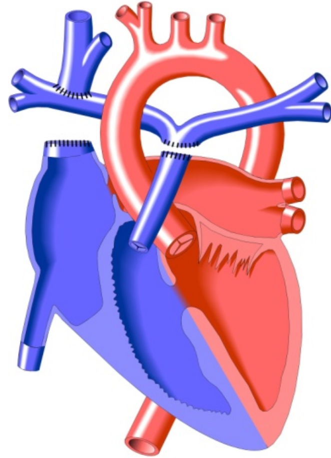
2. Atrial arrhythmias
   a. Occur in 30-40% of patients
   b. Atrial Flutter/Fibrillation from atrial dilation
   c. Supraventricular tachycardia
   b. Tend to be recurrent and resistant to drug treatment
3. May result in sudden death

IV. Surgical Repairs

A. Tricuspid Valve Repair/Replacement

B. Additional procedures

1. ASD closure
2. Anastomosis of superior vena cava (SVC) to right pulmonary artery (RPA) creating a Glenn Shunt (As Illustrated)



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3. Right Atrial reduction
4. Right sided 'Maze' procedure: Incisions made in the right atria to interrupt possible reentrant pathways that result in atrial arrhythmias including atrial fibrillation/flutter

V. Late Complications (See Problem List included with these documents)

A. Atrial arrhythmias
   1. Atrial fibrillation
   2. Atrial flutter
   3. Supraventricular Tachycardia (SVT)

B. Congestive Heart Failure

C. Cyanosis

D. Paradoxical Embolus

E. Endocarditis

F. Sudden Death

References:

Attenhofer Jost, et al: Left heart lesions in patients with Ebstein's Anomaly, *Mayo Clin Proc* 80:361-368, 2005.

Milliez P, Slama R: Wolff-Parkinson-White syndrome [Article in French], *Rev Prat.*54(16):1747-53, 2004.

Oechslin E, Buchholz S, Jenni R: Ebstein's anomaly in adults: Doppler-echocardiographic evaluation, *Thorac Cardiovasc Surg.* 48(4):209-13, 2000.

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