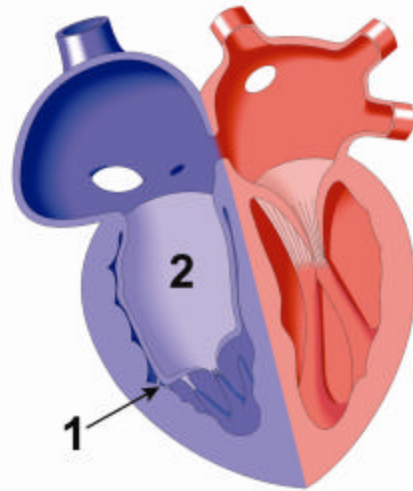


Ebstein's Anomaly

This rare defect involves an abnormality in the Tricuspid Valve, which connects the right atrium with the right ventricle. In Ebstein's Anomaly of the Tricuspid Valve, the valve forms abnormally and is lower than usual in the heart (number 1 in illustration).

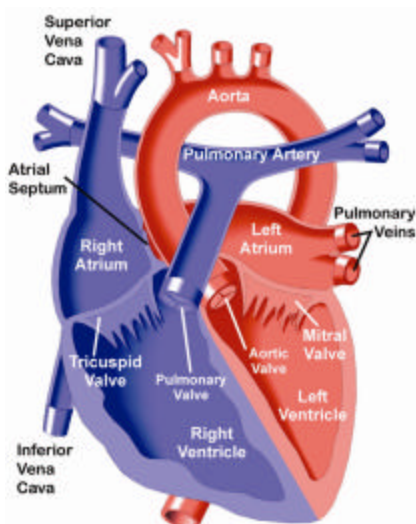
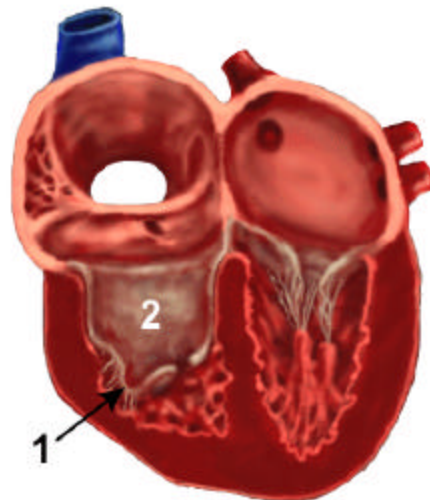
This displacement of the tricuspid valve results in insufficiency (leakiness) of the valve, which causes the right atrium, or collecting chamber, to be larger than normal. In addition, the "leaflets" or flaps of the tricuspid valve are usually abnormal in form. This stretched enlargement of the right atrium can predispose children to abnormal heart rhythms. Also, the abnormal position of the tricuspid valve causes part of the right ventricle to become functionally part of the right atrium. This is known as atrialization of the right ventricle (number 2 in illustration).



Frequently associated with this defect is a hole in the muscle wall that separates the atria, or upper chambers of the heart, known as an Atrial Septal Defect (ASD in diagram). Because of the increased pressure in the right atrium as a result of the leaky tricuspid valve, blue (oxygen-depleted) blood in the right atrium will often cross to the left atrium through the ASD. This results in de-oxygenated blood in the left side of the heart and decreased oxygen levels in the body.

The effects of Ebstein's Anomaly depend on the position and functioning of the tricuspid valve. In mild cases, no symptoms may be present and there is no need for treatment. In more severe cases, the baby may become "blue," or cyanotic, because there is a significant amount of de-oxygenated blood crossing the atrial septal defect (ASD in diagram) to the left heart.

The "blueness" is caused by a blockage of the opening into the pulmonary artery (PA) and the leakiness of the tricuspid valve. The blockage is a result of the abnormal position of the tricuspid valve. The greater the blockage, the leakier the tricuspid valve and the more deoxygenated blood crosses the atrial septal defect into the left heart.



25% of patients with Ebstein's Anomaly will have pre-excitation on their ECG and are prone to supraventricular tachycardia, or other atrial tachycardias.

Top and Above:

1. Ventricular displacement of the tricuspid valve.
2. Atrialized right ventricle.

Left: Normal Heart