

## Ebstein's Anomaly

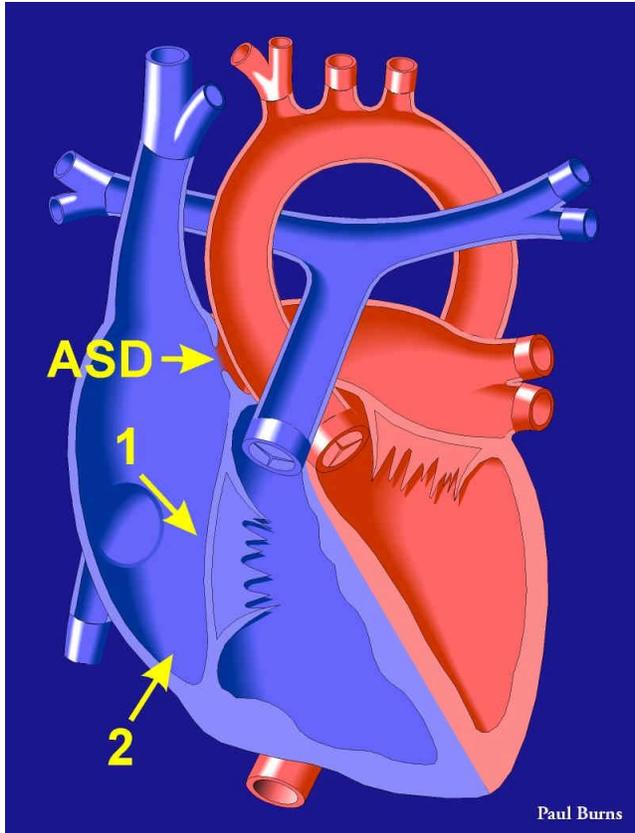


### What Is It?

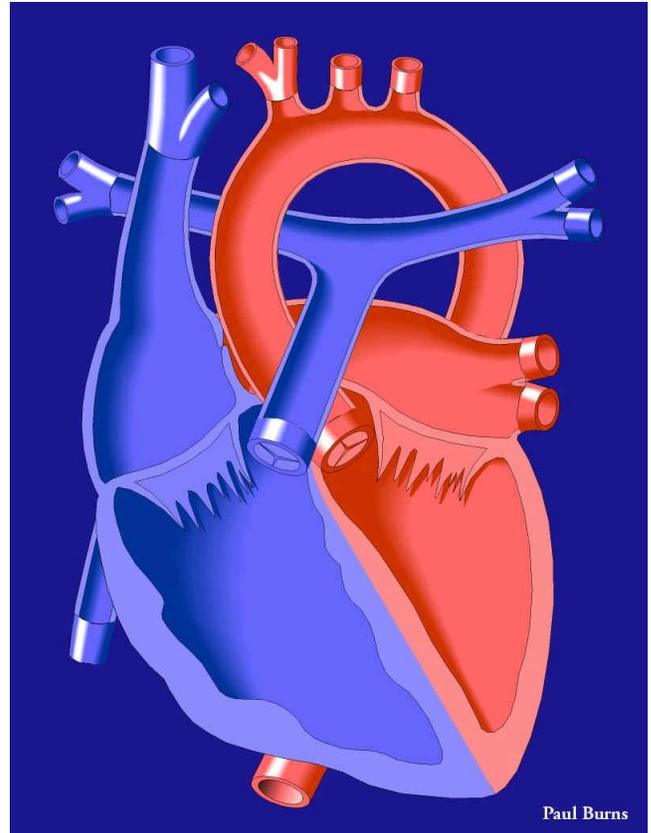
This rare defect involves an abnormality in the Tricuspid Valve, which connects the right atrium with the right ventricle. In Ebstein's Anomaly of the Tricuspid Valve, the valve forms abnormally and is lower than usual in the heart (number 1 in illustration).

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.



**Ebstein's Anomaly**



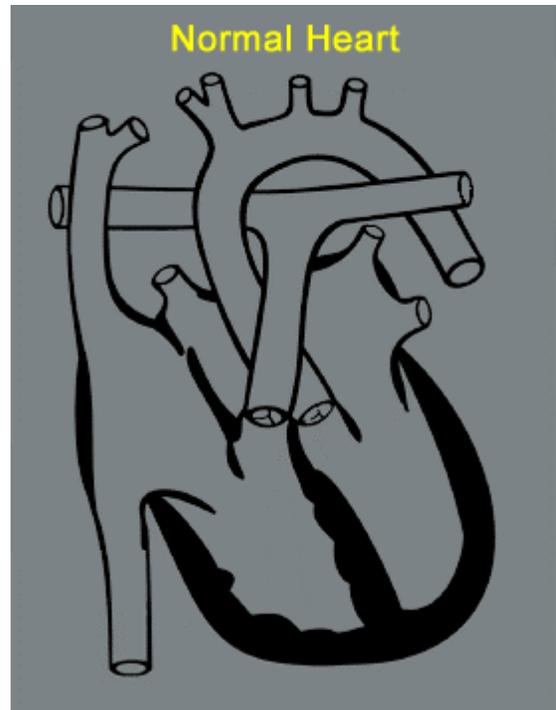
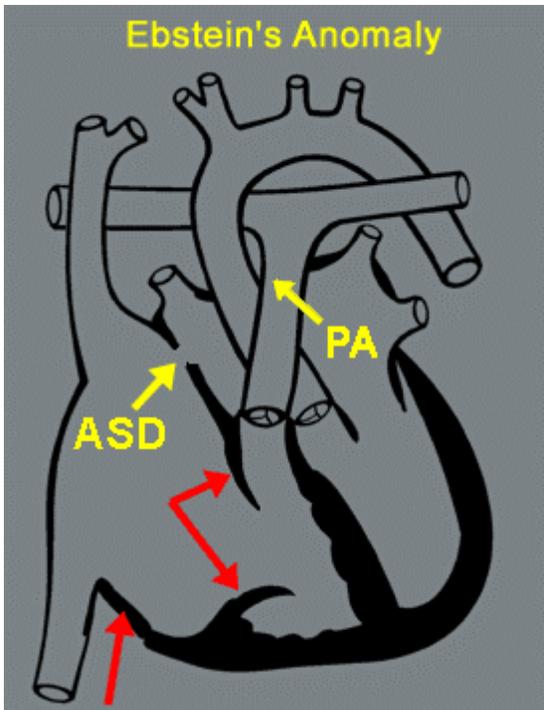
**Normal Heart**

### **What Are Its Effects?**

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 oxygenated blood crosses the atrial septal defect into the left heart.

In some cases, an abnormal heartbeat may occur with Ebstein's Anomaly that requires medicine.



### How Is It Treated?

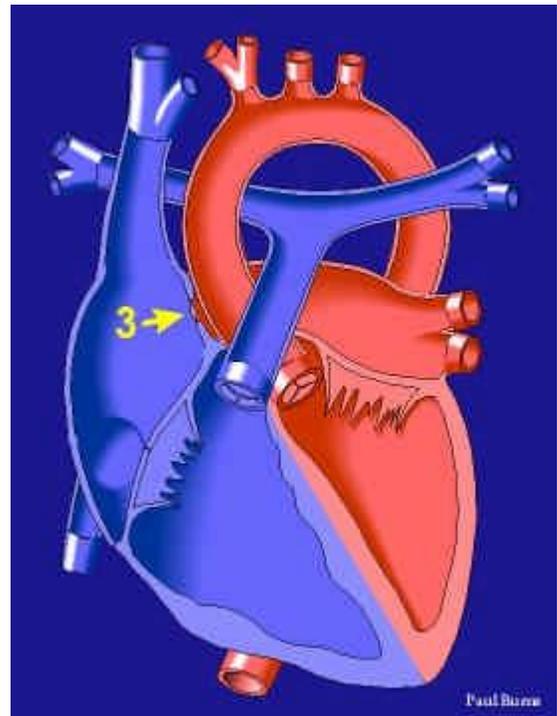
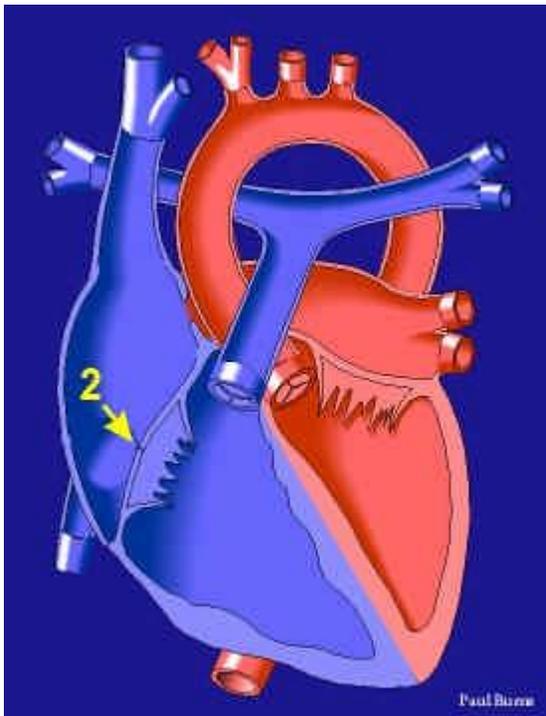
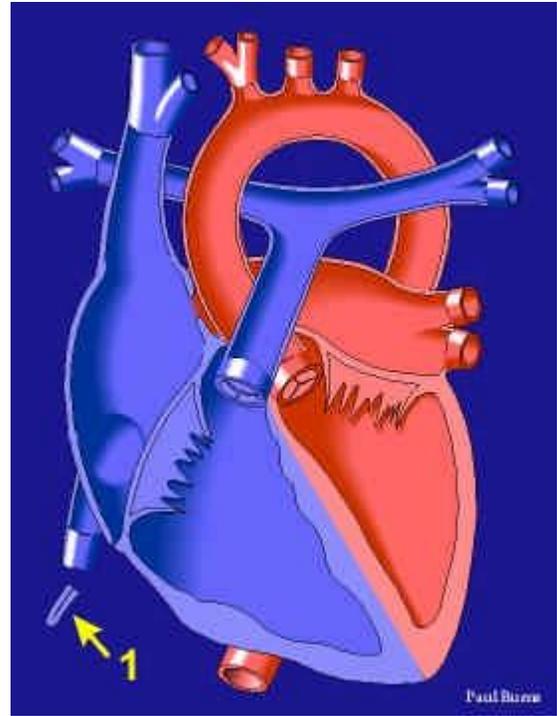
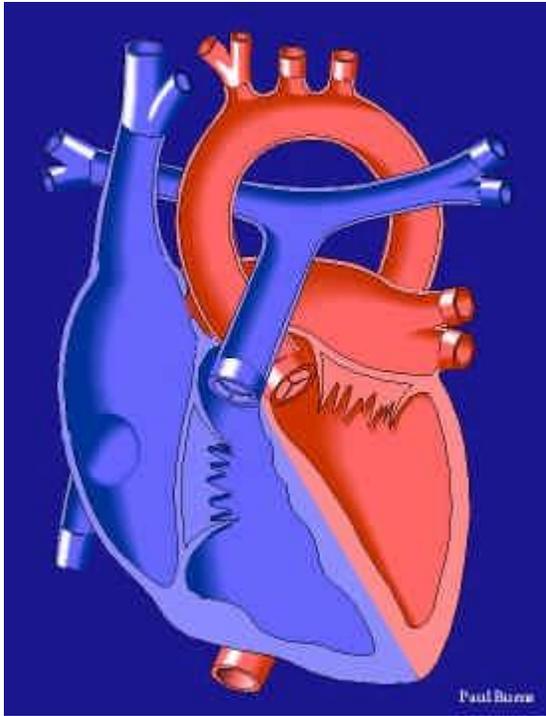
In mild cases of Ebstein's Anomaly, no treatment is necessary. However, if negative symptoms occur, such as cyanosis (blue baby syndrome), surgical intervention may be required.

Initial treatment shortly after birth may be necessary to treat a baby's "blueness." This may involve medications to keep the Ductus Arteriosus open, or Patent (see PDA), as well as a Modified Blalock-Taussig Shunt, involving the insertion of a tube between the pulmonary artery and a branch of the aorta. These are temporary measures to ensure that enough blood is carried to the lungs to receive oxygen.

To effect a permanent repair of Ebstein's Anomaly, the valve may be effectively repositioned by shortening the wall of the right atrium (1 in the illustration) and the tricuspid valve may be strengthened by modifying the shape of the valve leaflets (2 in the illustration).

Frequently, Ebstein's Anomaly is associated with an Atrial Septal Defect (ASD). The ASD may be closed with a patch made of pericardium (part of the membrane that covers the heart) or of a synthetic material. This patch (3 in the illustration) is shown as a pink oval.

In extreme cases, an artificial tricuspid valve may be introduced to correct the problem. The hospital stay after repair of Ebstein's Anomaly may be from two to three weeks.



**Surgical Repair of Ebstein's Anomaly**