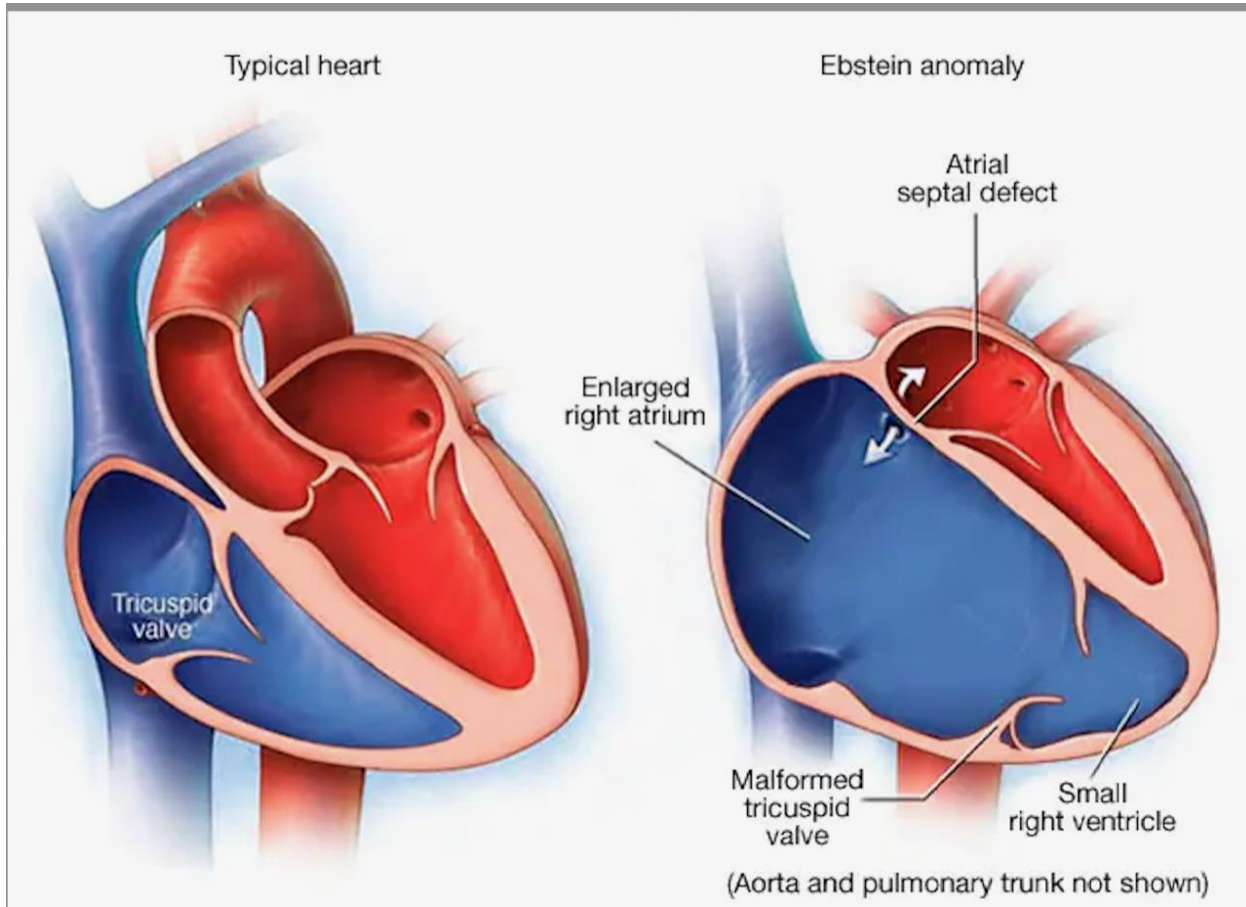


Ebstein's Anomaly

Patient Information Series – What you should know, what you should ask.

What is an Ebstein's anomaly?



Ebstein's anomaly is a defect of the tricuspid valve. The tricuspid valve separates the right atrium (the chamber that receives blood from the body) from the right ventricle (the chamber that pumps blood to the lungs).

In Ebstein's anomaly, two leaflets of the tricuspid valve are not in the right place. The third leaflet is longer than normal. It may be tied to the wall of the ventricle. Rarely, the valve is so deformed that it will not allow blood to flow forward in the normal direction (right atrium to right ventricle). These defects cause the tricuspid valve to leak blood back into the right atrium when the right ventricle contracts. As a result, the right atrium becomes larger than normal. If the tricuspid regurgitation is severe enough, congestive heart failure can result.

Ebstein's anomaly may occur with other heart defects, such as pulmonary valve stenosis or atresia, atrial septal defect or ventricular septal defect. Many patients with Ebstein's anomaly have an extra electrical conduction pathway in the heart. This can lead to times of abnormal fast heart rate called supraventricular tachycardia.

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How does an Ebstein's anomaly happen?

Ebstein's anomaly occurs as a baby develops in the womb. There may be a genetic component in Ebstein's anomaly, however in most cases no specific cause is determined. The use of certain drugs (such as lithium or benzodiazepines) during pregnancy may play a role. The condition is rare. It is more common in white people. Ebstein's anomaly may be associated with many different genetic problems and complex heart defects. In Ebstein's anomaly, the leaflets are placed deeper into the right ventricle instead of the normal position. The leaflets are often larger than normal. The defect most often causes the valve to work poorly, and blood may go the wrong way. Instead of flowing out to the lungs, the blood flows back into the right atrium. The backup of blood flow can lead to heart enlargement and fluid buildup in the body. There may also be narrowing of the valve (pulmonary valve) that leads to the lungs.

Should I have more tests done?

Your caregiver may refer you for genetic counseling and genetic testing. This can provide essential information regarding your individual case. In addition, your caregiver may refer you to specialists in fetal heart problems, such as a maternal-fetal medicine specialist and/or a pediatric cardiologist. They can monitor your baby's progress with specialized ultrasound scans focused on the fetal heart and the blood vessels around it. Those specialized ultrasounds are called fetal echocardiography. The tests offered by your caregiver will depend on many associated factors.

What are the things to watch for during the pregnancy?

Your caregiver may order serial ultrasound scans to assess whether the Ebstein's anomaly is affecting your baby's growth and well-being. Your caregivers will advise you as the pregnancy progresses.

What does it mean for my baby after it is born?

Ebstein's anomaly is a spectrum disorder with a very wide range of immediate, medium-term and longer-term problems. Ebstein's anomaly can be very mild to very severe. Many patients with milder forms of Ebstein's anomaly do not have symptoms. When symptoms are not present until the patient ages, a diagnosis is made when a heart murmur is heard. Some babies and children have blue color to their skin (cyanosis). This happens because of the flow of blood from the right atrium to the left atrium. Children may complain that their heart races, skips a beat, or just "beats funny." They may tire more easily than other children or become short of breath. Severely affected babies are often critically ill at birth. They may have low oxygen saturations (cyanosis) and heart failure needing immediate intensive care.

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Each baby with Ebstein's anomaly is unique. Your caregivers will evaluate your baby carefully to determine whether any intervention is necessary, and, if so, what will suit your baby best. Children with Ebstein's anomaly should receive continued care from a pediatric cardiologist. Besides getting information from routine exams, the cardiologist may use tests such as electrocardiograms, Holter monitor and echocardiograms.

Will it happen again?

This depends whether or not a genetic cause was found to explain the Ebstein's anomaly, and what other problems were present, if any. All these factors affect the likelihood of having another baby with Ebstein's anomaly. Your caregiver will likely order early targeted fetal scanning to rule out fetal heart defects in future pregnancies.

What other questions should I ask?

- Does this look like a severe case of Ebstein's anomaly?
- When is my next follow-up appointment?
- Should I have genetic counseling? What genetic testing should we consider?
- Where should I deliver?
- Where will the baby receive the best care after it is born?
- Can I meet the team of doctors that will be assisting my baby when it is born in advance of my delivery?

Last updated December 2022