

## **Ebstein's Anomaly**

### **What is it?**

Ebstein's anomaly, also called Ebstein's malformation, is a heart defect in which the tricuspid valve is abnormally formed and placed lower than normal in the right ventricle. The tricuspid valve normally has three "flaps" or leaflets. In Ebstein's anomaly, one or two of the three leaflets are stuck to the wall of the heart and don't move normally. The valve is lower than normal in the right ventricle. Often there's also a hole (atrial septal defect) in the wall between the heart's two upper chambers. S

### **What causes it?**

Most cases don't have an identifiable cause. Some studies have found an association between mothers on lithium (a mood stabilizing medication) and their children having Ebstein's anomaly. More recent studies suggest this isn't the case. Currently, pregnant woman on lithium are usually referred for fetal echocardiography to assess for Ebstein's anomaly in the fetus.

### **How does it affect my heart?**

Because the tricuspid valve is malformed in Ebstein's anomaly, it often doesn't work properly and may leak. If the valve leaks, some of the blood pumped by the right ventricle goes backwards through the valve with each heartbeat. This may result in significant enlargement of the right atrium. In more extreme cases the size of the right ventricle is too small to allow for enough blood to go to the lungs.

### **How does it affect me?**

If the leakage of the tricuspid valve is moderate or severe, symptoms including exercise intolerance and swelling of the abdomen and legs may develop. Heart rhythm problems may also occur. In extreme cases when the right ventricle is underformed, babies may be very blue. In these cases, patients may have required surgeries similar to patients with single ventricles.

### **Surgery in childhood and adulthood**

Ebstein's anomaly is mild in most adults who have it, so they don't need surgery. But sometimes the tricuspid valve leaks severely enough to result in heart failure or cyanosis. Then surgery may be required. Several different operations have been used in patients with Ebstein's anomaly. The most common involves a repair of the tricuspid valve. The valve can't be made normal, but often surgery significantly reduces the amount of leaking. In some cases the tricuspid valve can't be adequately repaired. Then it's replaced with an artificial valve. If there's an ASD, it's usually closed at the same time. In some patients, the atrial septal defect is the main problem and can be closed either with a device or with surgery.

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### **Problems You May Have**

#### **Heart Rhythm Disturbances**

People with Ebstein's anomaly may have a rapid heart rhythm called supraventricular tachycardia (SVT). An episode of SVT may cause palpitations. (You feel your heart racing.) Sometimes this is associated with fainting, dizziness, lightheadedness or chest discomfort. If you have these symptoms, contact your doctor. If your symptoms persist, seek immediate attention. Recurrent SVT may be prevented with medicines. In many cases, the source of the abnormal heart rhythm may be treated by a catheter procedure called radiofrequency ablation.

#### **Other Problems**

If the valve abnormality is especially severe, you may have decreased stamina, fatigue, cyanosis and sometimes fluid retention. These problems usually develop because the valve has become leakier. If you have these symptoms, contact your cardiologist. The symptoms may respond to medicines such as diuretics, which cause you to lose excess fluid. In some instances surgery (described above) may be recommended.

### **Ongoing Care**

#### **What will I need in the future?**

People with Ebstein's anomaly should receive continued care from a cardiologist with expertise in congenital heart defects. You should also consult a cardiologist with expertise in care of adult congenital heart disease if you're undergoing any type of non-heart surgery or invasive procedure.

#### **Medical**

Besides getting information from routine exams, the cardiologist may use tests such as electrocardiograms, Holter monitor and echocardiograms.

#### **Activity Restrictions**

Being physically active is good for your cardiovascular system, so stay active. If valve leakage is mild and tests show no abnormal heart rhythms, you can usually participate in most sports. Your cardiologist may recommend avoiding certain intense competitive sports. Ask your cardiologist which activities are right for you.

#### **Endocarditis Prevention**

You may need antibiotics before certain dental or surgical procedures to prevent endocarditis if you are cyanotic (blue) or have a prosthetic valve.

#### **Pregnancy**

Pregnancy puts many increased demands on the heart. In most cases women with mild Ebstein's anomaly will be able to safely deliver a baby. Still, each woman should be evaluated individually. Consult with your cardiologist about the safety of pregnancy.

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### **Will you need more surgery?**

Patients with mild tricuspid valve leakage are unlikely to require surgery at any point. Patients with more moderate or severe leakage may need initial or subsequent surgery. Patients who have undergone pacemaker placement need close follow-up and eventually may need their pacemaker replaced.