Ebstein's Anomaly

Note: before reading the specific defect information and the image(s) that are associated with them, it will be helpful to review normal heart function.

What is it?

Ebstein's anomaly, also called Ebstein's malformation, is a heart defect in which the tricuspid valve is abnormally formed. 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. Often there's also a hole in the wall between the atria, the heart's two upper chambers. This hole is called an atrial septal defect or ASD. (See the section on Atrial Septal Defect.) 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. In some children, the right ventricle downstream from the tricuspid valve is smaller than normal and doesn't work properly.

Can it be repaired?

Ebstein's anomaly is mild in many children that they don't need surgery. But sometimes the tricuspid valve leaks severely enough to result in heart failure or cyanosis (see the Glossary). 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. If there's an ASD, it's usually closed at the same time. In some cases the tricuspid valve can't be adequately repaired. Then it's replaced with an artificial valve.

What ongoing care will my child need?

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.

What activities will my child be able to do?

If valve leakage is mild and tests show no abnormal heart rhythms, your child can usually participate in most sports. Your cardiologist may recommend avoiding certain intense competitive sports. Ask your child's cardiologist which activities are appropriate.

What problems might my child have?

Children with Ebstein's anomaly may have a rapid heart rhythm called supraventricular tachycardia (SVT) often as a result of a condition called Wolf-Parkinson-White syndrome (WPW). An episode of SVT may cause palpitations (older children may feel your heart racing.) Sometimes this is associated with fainting, dizziness, lightheadedness or chest discomfort. Infants may be unusually fussy or have other symptoms that can't easily be connected with rapid heart rhythm. If you child has had these symptoms, contact your doctor. If your symptoms persist, seek immediate attention. Recurrent SVT may be

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prevented with medicines. In many cases, the source of the abnormal heart rhythm may be removed by a catheter procedure called radiofrequency ablation.

If the valve abnormality is especially severe, you may have decreased stamina, fatigue, cyanosis, and sometimes fluid retention. Infants may not feed or grow normally. The symptoms may respond to medicines such as diuretics. In some instances surgery (described above) may be recommended.

What about preventing endocarditis?

People with unrepaired or incompletely repaired Ebstein's anomaly are at risk for endocarditis. Ask your pediatric cardiologist about your child's need to take antibiotics before certain dental procedures to help prevent endocarditis. See the section on endocarditis for more information.