



Ebstein's Anomaly

Definition:

Ebstein's Anomaly, is a heart defect in which the tricuspid valve is abnormally formed. The tricuspid valve is the valve separating the right upper chamber (atrium) from the right lower chamber (ventricle). Normally, the valve has 3 leaflets or parts. In Ebstein's anomaly, 1 or 2 of the leaflets are abnormally attached to the wall of the heart and will not move normally. This condition is usually associated with a hole between the two upper chambers (atria). The abnormal valve will leak, causing some of the blood to return back to the right atrium and thus the right side of the heart will be affected.

What causes it?

Cause is usually unknown, however there is a definite known association between mothers taking lithium (prescribed by psychiatrists) in their first trimester and babies born with the anomaly.

How will it affect the heart?

In addition to the affection of the right side of the heart, problems with heart rhythm are known to occur. Children may have a rapid heart rate called supraventricular tachycardia (SVT). An episode of SVT will cause palpitations (the child will feel his heart beating rapidly), and sometimes fainting, dizziness, lightheadedness or chest discomfort can occur. If your child has any of these symptoms, they must immediately consult their doctor and go to the hospital if the symptoms persist. If the SVT is recurrent, medical treatment will be mandatory and sometimes they may need a management option known as radiofrequency ablation can be done by a catheter to remove the source of the abnormal heart rhythm.

Management Options:

If mild, then usually, surgery is not required. If the valve is severely affected and causes enough of a leak to cause heart failure then surgery might be required.

Repair of the tricuspid valve with closure of the ASD (if present) will be done. If the valve cannot be repaired, then it can be replaced with an artificial valve.

What activities can your child do?

If the surgery is successful and there's no associated heart rate problems, then the child should have a normal life with no restriction. However, if there is an associated with heart rate problems, the cardiologist will need to be consulted and, most likely, restriction of severe sports will be necessary.

Follow up in the future:

There will be continued following up with the pediatric cardiologist, especially due to the heart rate problems that may occur. Your doctor might require doing non invasive tests such as ECGs, Holter monitoring and echocardiograms.

Children may need to receive antibiotics before any surgical or dental procedures to prevent infective endocarditis.