Tricuspid valve abnormalities (including Ebstein’s anomaly)

This information sheet from Great Ormond Street Hospital explains the causes, symptoms and treatment of tricuspid valve abnormalities (including Ebstein’s anomaly) and where to get help.

Tricuspid valve abnormalities (including Ebstein’s anomaly) are congenital heart defects – that is, they were present when a child is born. Around eight in every 1,000 babies born have a congenital heart defect, though Ebstein’s anomaly is rarer than that.

The normal heart

The heart consists of four chambers – two upper filling chambers (atria) and two lower pumping chambers (ventricles). In between each atrium and ventricle is a valve that stops blood flowing backwards. There is also a valve where the pulmonary artery and aorta join the heart.

The right and left sides of the heart are divided by a thick wall of heart muscle called the septum.

The function of the heart is to pump blood around the body. Blood comes into the right atrium from the body and through the tricuspid valve into the right ventricle. From here, it is pumped up the pulmonary artery to the lungs to pick up oxygen. Oxygen-rich blood comes back to the heart through the pulmonary veins into the left atrium. It flows through the mitral valve into the left ventricle. This pumps the blood into the aorta and from there around the body.

What is Ebstein’s anomaly?

Ebstein’s anomaly is the name given to the defect where the tricuspid valve is abnormal. The tricuspid valve lies between the upper right filling chamber (atrium) and the lower right pumping chamber (ventricle). As the valve is abnormal, oxygen-poor blood can flow back into the right atrium instead of being pumped to the lungs to pick up oxygen.

Most children also have an atrial septal defect, which is a hole between the two upper filling chambers (atria). This is
another route for oxygen-poor blood to travel around the body instead of being pumped to the lungs.

In many children, the pulmonary valve between the right ventricle and the pulmonary artery is narrower than usual, so the heart has to work harder to pump blood to the lungs.

**What causes Ebstein's anomaly?**

The heart is formed very early in pregnancy. Doctors do not fully understand why some children's hearts do not develop properly. However, they know that the chance increases a little if one or both parents had a congenital heart defect.

Occasionally some conditions such as diabetes or medicines taken during pregnancy can also increase the risk.

Congenital heart defects can also be more common in children with other congenital conditions.

**How is Ebstein's anomaly normally diagnosed?**

The child's defect may have been diagnosed antenatally by our foetal team and will be confirmed on admission. Doctors will use chest X-rays, electrocardiograms (ECG) and echocardiograms (Echo) to diagnose Ebstein's anomaly. An ECG measures the electric current passing through the heart. An Echo is an ultrasound of the heart and shows not only the structure of the heart but the blood flow through it.

**How is Ebstein's anomaly treated?**

Not all patients need surgery.

Before any surgery is planned, the doctors may suggest medicines to help the heart work better if it is already coping poorly. Surgery if it is necessary is aimed at making the tricuspid valve less leaky and normalising the flows in the heart.

**What happens next?**

All children with congenital heart defects (even when corrected) will need regular check ups in hospital for a long time, usually continuing into adulthood.

These will usually involve repeat Echo and ECG scans and sometimes cardiac magnetic resonance imaging (MRI) scans. The aim of these check ups is to monitor your child's heart function so that any future heart problems are diagnosed and treated quickly.
Further information and support

Our Cardiorespiratory Unit regularly refer to information published by the British Heart Foundation (BHF) and the Children's Heart Federation when explaining tricuspid valve abnormalities (including Ebstein's anomaly) to our patients and their families.

- Visit the BHF website to download their tricuspid atresia factsheet
- Read about Ebstein's anomaly on the Children's Heart Federation website

The following support organisations may also help:

**British Heart Foundation**
Tel (Heart Help Line): 0300 330 3311 (calls charged at local rate)
Website: www.bhf.org.uk

**Heartline**
Tel: 03300 224 466 (local rate number)
Website: www.heartline.org.uk

Notes

Compiled by the Web team in collaboration with the Child and Family Information Group

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