Where is the pulmonary valve and what does it do?

The pulmonary valve is situated at the junction of the right ventricle and the pulmonary artery. When the heart pumps, de-oxygenated (blue) blood is pushed from the right ventricle through the pulmonary valve into the pulmonary artery. From here, it travels to the lungs where the blood picks up oxygen again. The pulmonary valve stops blood flowing backwards into the right ventricle (regurgitation). If a small amount of blood flows back into the right ventricle with each pump of the heart, in time, the ventricle can become stretched and weak.

If the problem is not corrected, it can have a ‘knock on’ effect on the tricuspid valve, which is between the right atrium and right ventricle. These problems can have damaging effects, leading to heart failure, arrhythmias (abnormal heart rhythms) and rarely, sudden unexpected death, if not treated.

Why might the pulmonary valve need replacing?

The most common cause in children and young adults is related to treatment of congenital heart disease, such as surgery involving the creation of a new connection between the heart and lung artery (tetralogy of Fallot, pulmonary atresia, truncus arteriosus, Rastelli operation for transposition of great arteries) or balloon dilatation of the pulmonary valve or after surgery for left...
Treatment of these conditions requires the use of tissue valves that can become narrow over time or start leaking. In the past, the only treatment for such conditions was open heart surgery. With this technique, surgery can be postponed or completely avoided in some cases.

Pulmonary valve replacement is usually suggested when a child has symptoms of heart failure, such as tiredness on exercising. Research has shown that the results of replacement are better when it is carried out sooner rather than later so that irreversible injury to the right ventricle is avoided.

**What is pulmonary valve replacement?**

At GOSH, pulmonary valve replacements are carried out without needing open-heart surgery in cases where this technique can be performed with less invasive procedures using cardiac catheterization. A technique has been developed where the pulmonary valve is replaced using cardiac catheterisation (percutaneously or through the skin). This procedure is also called percutaneous pulmonary valve implantation. If this technique is used, it avoids the need to operate through the open chest and use of heart lung bypass. Instead, a catheter (thin, plastic tube) is passed through the vein in the groin up to the heart, through which the valve is inserted. X-rays are used to visualise the catheters moving around the heart. The advantage of this approach is that your child will have less pain and a shorter recovery period and no chest scar from the procedure.

**Preparing for the procedure**

Your child may need to come to the hospital before the procedure for a preadmission check. For more information, please see our Pre-admission clinic for cardiology information. The doctors will explain the procedure in more detail, discuss any worries you may have and ask you to give your permission for the procedure by signing a consent form.

Another doctor will visit you on the ward to explain about the anaesthetic. Further information about anaesthetics is available in our Cardiac anaesthesia information sheet. The doctors may also ask for some blood samples to check that your child is well before the procedure and sometimes, some tests may be repeated like an echocardiogram (Echo).

If your child has any medical problems, like allergies, please tell the doctors about these. If your child is taking warfarin, this will need to be stopped three days before the procedure, to reduce the risk of serious bleeding during or afterwards. We also advise that your child does not visit the dentist in the weeks before the procedure – this will reduce the risk of developing a dental infection, which could spread elsewhere in the body.
What happens before the procedure?
On the day of the procedure, your child will need to arrive on the ward at the time stated in your admission letter. The nurses will make sure that you and your child understand the procedure and get your child ready. They will then take your child to the cardiac catheterisation room.

What does the procedure involve?
Once your child is under general anaesthetic, a small area of their groin will be cleaned with antiseptic solution. The vein and artery in the groin that connect to the heart are used to insert various small tubes and wires. These help in measuring and outlining the parts of the heart where the valve would be inserted. Pressure inside the vessels and heart can be measured and dye injected to produce pictures and take measurements (angiography).

After assessing the need for and suitability of this technique, the narrow or leaky area where the valve will be implanted is prepared by either stretching it with a balloon or by inserting a slotted metal tube called a stent that provides a platform to implant the valve. The valve is loaded onto a specially made delivery catheter. After careful positioning and checks, the valve is implanted by inflating a set of two balloons on the delivery catheter. Repeat measurements and angiography are performed to confirm a successful result, after which the catheter is removed.

Most often, the skin entry in the groin is closed with stitches which will need to be removed later.

Are there any risks?
The team looking after your child are very experienced. However, as with all procedures, there are risks, which although unlikely, you should understand. The procedure is performed under general anaesthetic, and although every anaesthetic carries a risk, this is extremely small. There is only a small risk of infection because no surgical incisions are necessary. The implantation procedure may rarely be complicated by events needing emergency rescue surgery. In this situation, the surgeon would insert a new valve in the same procedure.

Your child may bleed from the area where the catheter is inserted, but this can be minimised by applying pressure for a few minutes after the procedure. He or she may develop a bruise where the catheter was inserted, and feel some discomfort in this region, but pain relief like paracetamol is usually enough to deal with any discomfort. Your child may develop a high temperature after the procedure, although this usually reduces over the following day or two. Sometimes, blood samples are taken to rule out infection.

The valve inserted by catheter may also wear out in time, but another valve can be inserted using catheterisation as before.
What happens afterwards?
Your child will return to the ward after he or she has recovered from the anaesthetic. After a general anaesthetic, some children feel sick and may vomit. Your child may have a headache or a sore throat or experience some dizziness, but these side effects are usually short-lived and not severe.

Your child will need to stay in bed for a few hours after the procedure to make sure the catheter site has started to heal. The nurses will check them regularly during this time. Most children stay one night on the ward to make sure that the valve is working properly, and that they have fully recovered from the anaesthetic. The sutures from the groin will be removed. Once the doctors are happy that your child is well enough, you and your child will be able to go home. Your child will be discharged with a prescription of aspirin that has to be taken on a daily basis to prevent any clots forming.

When you get home
You should call the hospital if:

- your child starts bleeding from where the catheter was inserted. Apply firm pressure to the area for 5 to 10 minutes. If the bleeding does not stop, call 999 for an ambulance
- your child is in a lot of pain and pain relief does not seem to help
- the area where the catheter was inserted looks red, swollen and feels hotter than the surrounding skin
- your child is not drinking any fluids after the first day back home
- your child develops a high temperature that does not reduce with paracetamol or remains high for more than a day or two

Precautions

- A direct hit to the chest may affect the stent (part of the valve which holds it open), breaking it or making it misshapen, which will weaken it. Your child should avoid contact sports, such as rugby and football until allowed by your doctor.
- If your child starts to develop symptoms such as breathlessness or tiredness on exercising, please contact your doctor for an appointment. A chest x-ray and Echo will be needed to check that the valve is not damaged.
- Infection of the valve can cause symptoms such as a high temperature, poor appetite, weight loss and generally being unwell. It is important to suspect valve infection if your child develops these symptoms.
- Since March 2008, guidelines from the National Institute for Health and Clinical Excellence (NICE) advise that children and adults with congenital heart defects need not have antibiotic prophylaxis for dental treatments. However, maintenance of good dental hygiene is of paramount importance to reduce the risk of infection.
Follow up
We will arrange appointments to see your child at one month, six months and one year after the procedure and more tests may be requested by your doctors (such as Echo, ECG, 24 hour Holter, MRI, Exercise test).

What is the outlook for children who have had pulmonary valve replacement?
The results of the new way of doing pulmonary valve replacement are good, with few lasting side effects. Depending on any other heart problems, your child should be able to lead a near-normal life after the procedure. As with all valve replacements, the valve itself might need replacing in time, but this is not usually needed for many years.

Is there a support group?
The following organisations can offer advice and support to parents of children with any heart problem – visit their websites for further information.
- HeartLine Association: www.heartline.org.uk
- Children’s Heart Federation: www.chfed.org.uk
Further information about the valve and its development is available from the manufacturers at www.medtronic.com/intl/melody

If you have any questions, please telephone Walrus Ward (Cardiac Day Care) on 020 7813 8347. Out of hours, please telephone Bear Ward on 020 7829 8829.

Notes

Compiled by the Cardiac Surgery department in collaboration with the Child and Family Information Group

Great Ormond Street Hospital for Children NHS Foundation Trust, Great Ormond Street, London WC1N 3JH
www.gosh.nhs.uk