

Pulmonary Stenosis

What is Pulmonary Stenosis?

Pulmonary means 'of the lungs'.
Stenosis means narrowing.
Pulmonary Stenosis is a narrowing of the pulmonary valve. This means that the right ventricle has to work harder to pump blue (deoxygenated) blood through the valve, into the pulmonary artery and so to the lungs to pick up oxygen.

Fig 1 – Pulmonary Stenosis

Pulmonary Stenosis

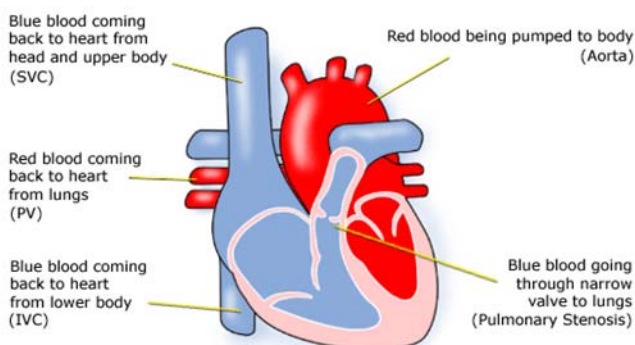
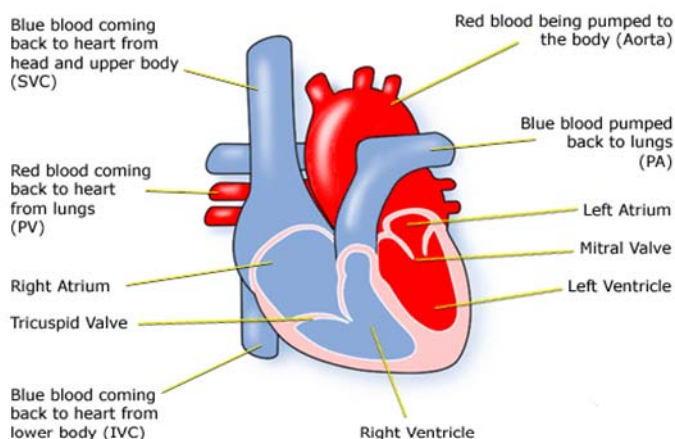


Fig 2 – Normal Heart

Normal Heart



Diagnosis

Your child's Pulmonary Stenosis may have been seen on a scan during pregnancy.

After birth the sound of blood moving through the narrow valve can be heard as a heart murmur.

Other heart disorders are sometimes found with Pulmonary Stenosis.

When a heart problem is suspected the tests used can be:

- pulse, blood pressure, temperature, and number of breaths a baby takes a minute
- listening with a stethoscope for changes in the heart sounds
- an oxygen saturation monitor to see how much oxygen is getting into the blood
- a chest x-ray to see the size and position of the heart
- an ECG (electrocardiogram) to check the electrical activity
- an ultrasound scan (echocardiogram) to see how the blood moves through the heart
- checks for chemical balance in blood and urine
- a catheter or Magnetic resonance Imaging test may be needed.

At home

After your child is diagnosed.

- You, your GP and Health Visitor should have details of your baby's condition from the heart doctor (paediatric cardiologist). If not, call the hospital at which your child was seen, ask for the name of the paediatric cardiologist and their telephone number. Call and explain that you need the information to pass on to, for example, your local casualty

Department should he or she have a sudden illness.

- You should have the number of a cardiac liaison nurse or outreach nurse to call should you have questions or any fears about your child's heart problem.
- You should have the number of a parent support group.

Treatment

Treatment depends on how narrow the valve is.

In many cases it will not cause the child to be unwell, but a cardiologist will need to keep an eye on it as sometimes the problem gets worse. Any such change occurs slowly, so expect appointments to be infrequent – every two years or so.

Severe narrowing will give the right ventricle too much work to do.

The aim of the procedures is to make the entrance to the pulmonary artery bigger, allowing the blood to be pumped to the lungs at lower pressure.

If your child has other heart defects, the kind of surgery needed will depend on how the heart can best be modified to cope with all the problems he or she has.

For most children the procedures are low risk, but it can depend on how well your child is otherwise. The doctors will discuss risks with you in detail before asking you to consent to the operation.

Balloon dilation: A tube is inserted through a vein in the groin and into the heart. It is then passed through the narrow valve into the pulmonary artery. A balloon on the end of the tube is inflated, so stretching the pulmonary valve. This does not leave any scar and recovery time is usually overnight, so your child only spends one or two days in hospital.

Corrective surgery: in very severe cases open heart surgery may be needed – the heart will need to be stopped and opened to repair it. This means that a machine will have to take over the job that the heart normally does – the heart bypass machine. The aim of the operation is enlargement of the area around the pulmonary stenosis – sometimes needing a valve replacement.

The length of time in hospital after surgery will usually be only 5-7 days, of which one or two will be spent in the intensive care and high dependency unit. Of course this depends on how well your child is before and after the surgery, and whether any complications arise.

The valve used to replace the child's narrow valve is usually a homograft – from another person. These valves need replacing after a few years.

How the child is affected

Most children are completely well, active, and gaining weight a few days after a balloon catheter or surgery. After surgery he or she will have a scar down the middle of the chest, and there may be small scars where drain tubes were used. These fade very rapidly in most children, but they will not go altogether. Smaller scars on the hands and neck usually fade away to nothing.

Some of these problems can occur after procedures or later in life:

- It is common for the pulmonary valve to leak and it may need further repair or replacement.
- Hearts that are not normal are more likely to have an infection called endocarditis. Although rare this is a difficult disease to treat.

People born with pulmonary stenosis will need to take antibiotics if there is a chance that a large number of bacteria

will get into the blood stream. The most common way for this to happen is during ear-piercing or tattooing, or surgery or a dental procedure such as de-scaling of teeth or an extraction.

These problems may not become serious until the teen years or adulthood.

Parents' stories

Charlie

Now she is thirteen, we have had a good long talk to Charlie about her heart. The treatment she had on her valve was when she was only just over a year old, so she doesn't remember it, and her problem really has been far more to do with asthma – that's what has kept her off games and needing to keep going to the doctors.

Last year at her heart check up, the doctor started to explain that the valve was getting narrower, and she would need more treatment in a year or so.

That seems a long time to Charlie but by then she will need to take over talking to the doctor, and getting her appointments sorted out herself. She just doesn't seem interested.

It can't help that when I start to talk about it I get upset, so I think she blots it out – all she says is 'Have you finished?' and 'Don't get so stressed about it.'

I'm going to tell her that her next appointment she will go to see the doctor by herself, and I'll wait outside. Next time she has treatment, she will sign the consent form, and if there is a choice to be made between an operation and a catheter, it will be Charlie's choice.

Liam

It was our GP who spotted that Liam has a heart condition. He was four months and such a big, healthy boy, that I really couldn't believe it – I mean, heart

problems is something you expect your old uncle to have after a lifetime of chips and booze.

Then we had to wait to see a cardiologist for three weeks. During that time Mark and I didn't talk about it. I really felt I had let him down having a baby with that sort of problem. When it got near to the appointment date I asked him if he would come, but he said he didn't see the need.

Liam had an ultrasound, an echo, like the ones I had when I was expecting and they told me straightaway that part of his heart was narrow, and that was why the GP heard the heart murmur. We just need to keep an eye on it, they said, and it may need treatment when he is older.

I forgot to ask all those things I knew Mum, and Mark and my friend Carrie would want to know. I didn't ask if he would grow up, if he would be able to walk, go to school, play football. For two weeks I just cried whenever I looked at him, and kept myself to myself.

It was my GP who came to the rescue, got me information about Pulmonary Stenosis, and brought Mark in for a talk as well.

Three years on it seems a lot of fuss – you wouldn't think Liam had anything wrong at all, and with another baby on the way, I've got other things to think about.

Please contact CHF if you have suggested amendments or changes as we like to keep our information sheets relevant and up-to-date.

