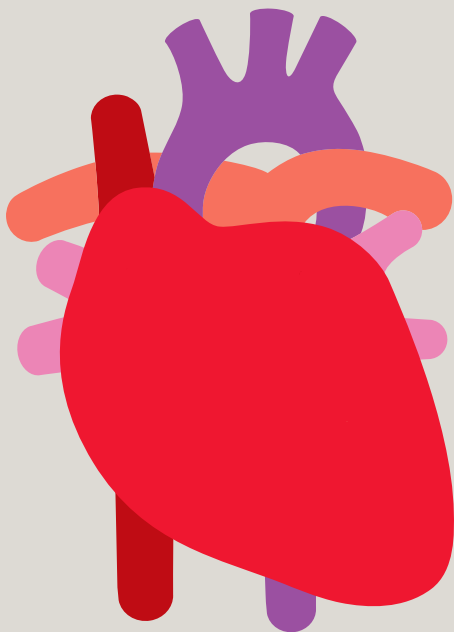


Understanding your child's heart

Pulmonary stenosis

4



**FIGHT
FOR EVERY
HEARTBEAT**

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About this book

If you're reading this book, you've probably just had some very upsetting news, and have lots of questions running through your mind.

We've written this book to help answer some of those questions. We'll go through:

- what pulmonary stenosis is and how it is diagnosed
- how pulmonary stenosis is treated
- the benefits and risks of treatments
- what happens as your child grows up
- where to go for more support.

Please be aware that this booklet shouldn't replace the advice your doctors or nurses may give you. But it should help make what they tell you that little bit clearer.

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What is a congenital heart condition?

It's an abnormality of the heart that developed in the womb. Sometimes, a **congenital** heart condition is diagnosed when the baby is still developing in the womb, but most times it's not discovered until after the baby is born. There are lots of different types of **congenital** heart conditions.

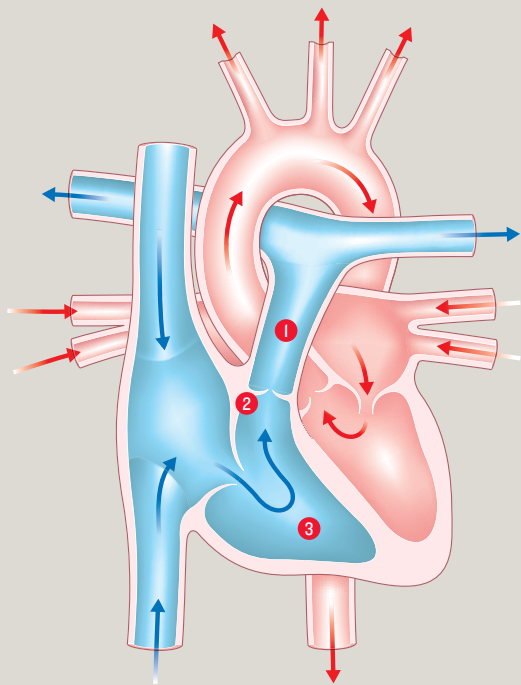
Each day 12 babies are diagnosed with a **congenital** heart defect in the UK. We continue to support research to improve the understanding, diagnosis and treatment of **congenital** heart conditions. For more information into our pioneering research visit **bhf.org.uk/research**

What causes a congenital heart condition?

In most cases, it's caused by something going wrong during the very early stages of the pregnancy.

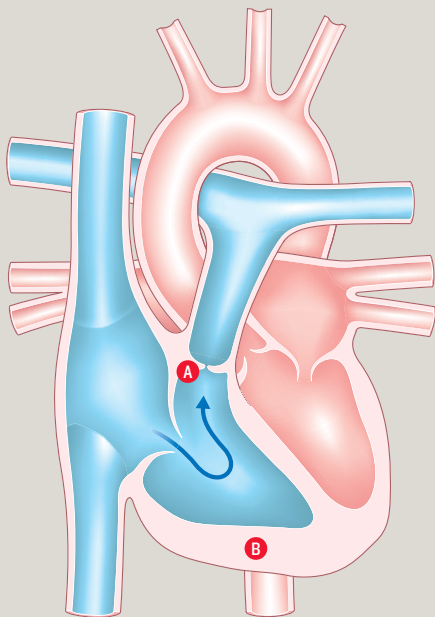
At the moment we don't fully understand why a baby's heart doesn't develop normally. But sometimes a **congenital** heart condition can be part of a syndrome which the baby is born with. *(A syndrome is a group of symptoms that commonly appear together as part of a condition).*

Normal heart



- 1 Pulmonary artery
- 2 Pulmonary valve
- 3 Right ventricle

Pulmonary stenosis



- A** Narrowed pulmonary valve
- B** Thickened muscle

What is pulmonary stenosis?

Pulmonary stenosis means that the **pulmonary** valve – the valve that lets blood flow from the heart to the lungs via the **pulmonary** artery – is narrowed. When the **pulmonary** valve is too narrow, the right ventricle has to work harder. This means that the muscle will get bigger, just like any other muscle in the body does when it works hard. If the **pulmonary** valve is severely narrowed, this can limit how much exercise your child can manage.

Most children have only mild **pulmonary stenosis** and don't need any treatment even when they are older, but the valve can become more narrowed as time goes by. You will need to take your child for regular check-ups at the outpatients department, even if they appear to be well and doesn't have any symptoms.

What are the symptoms of pulmonary stenosis?

Most children with **pulmonary stenosis** won't have any symptoms at all, and will appear perfectly well. Usually the only sign of **pulmonary stenosis** is a heart **murmur**, (*an extra sound from the heart*), which may be picked up during a routine medical check.

If your child has significant **pulmonary stenosis**, they may feel tired when playing or doing physical activity. In some rare cases where it is very severe, the child may have fainting episodes. If this happens, you should let your paediatrician or the **paediatric** liaison nurse know immediately. Talk to your doctor or nurse about who to contact in these circumstances.

Babies who are diagnosed before birth usually have a severe form of **pulmonary stenosis** and will usually need treatment shortly after birth.

What other conditions are associated with pulmonary stenosis?

Pulmonary stenosis mostly occurs on its own, without other abnormalities. However, sometimes children with **pulmonary stenosis** may also have a genetic abnormality such as Noonan syndrome. For more information on Noonan syndrome, visit www.nhs.uk/Conditions/noonan-syndrome.

Sometimes children with **pulmonary stenosis** also have other heart abnormalities. Your child's **cardiologist** will tell you if this is the case for your child.

How is pulmonary stenosis diagnosed?

In most cases, **pulmonary stenosis** is not diagnosed until after the baby is born, but some severe cases may be detected before birth using an **echocardiogram**. This is an ultrasound scan of the heart and it won't hurt your baby at all.

How is pulmonary stenosis treated?

If the **pulmonary valve** is severely narrowed, your child will need treatment shortly after diagnosis. It is very rare for a child with **pulmonary stenosis** to need open-heart surgery, most cases can be treated using a procedure called balloon **valvuloplasty**.



For more information and support about growing up with a heart condition, visit [bhf.org.uk/heart-health/children-and-young-people](https://www.bhf.org.uk/heart-health/children-and-young-people)

Balloon valvuloplasty

This is usually done under a general anaesthetic. A **catheter** (*a thin, hollow tube*) with a small collapsed balloon at its tip is inserted into a vein at the top of your child's leg. This is guided up the vein into the right side of the heart and across the narrowed valve. Using X-ray, the balloon is positioned in the narrow valve, and is then inflated, stretching the valve open. The balloon is then deflated and removed.

What are the risks of valvuloplasty?

Valvuloplasty is a very effective form of treatment and the fatality risks are very low. However, do be aware that in new-born babies with severe **pulmonary stenosis**, the risk will be greater. Your child's **cardiologist** will be able to talk through the details with you.

In a small proportion of children, the thickened heart muscle doesn't return to normal and the muscle itself can obstruct the normal flow of blood. If this causes significant narrowing inside the heart, your child will need to have surgery to remove some of the muscle.

What happens after a balloon valvuloplasty?

Your child may have to stay in hospital for a few days afterwards. If the narrowing has been only partly relieved by the balloon **valvuloplasty**, it's advisable the procedure be repeated at a later date. Because the thickened heart muscle itself can cause some narrowing, it is not always possible to tell if the procedure has been successful straight away. It can take several weeks for the thickened muscle to return to normal.

Surgery

Sometimes the **pulmonary** valve cannot be stretched open using the balloon **valvuloplasty** procedure, and open-heart surgery is needed to carry out a **valvotomy**. This involves having a general anaesthetic. During the operation, the heart is stopped and the function of the heart is taken over by a 'heart-lung machine', which makes sure that blood is still pumped around your child's body. The surgeon will then open the heart and make a small cut in the narrowed valve, to allow more blood to flow through. The heart is then restarted. After the operation, your child will have a scar down the centre of their chest, along the breast bone.

What are the risks of surgery?

The good news is, surgery for this condition carries a very low fatality risk. The risk of major complications such as brain damage is also very small. Other uncommon complications – such as a small amount of fluid collecting around the heart or lungs – can also occur after the operation, but these are rarely serious.

What happens after surgery?

Your child will need to stay in hospital for a few days after the surgery. You will have to bring your child back to the outpatients department to see the **paediatric cardiologist**.

What happens as my child grows up?

Those who have not needed any treatment by the time they are fully grown usually don't ever need treatment for their **pulmonary stenosis**. However, in some rare cases the valve can become narrower in later adult life. Balloon **valvuloplasty** is usually effective in adult life just as it is in children.

After any kind of treatment for **pulmonary stenosis**, the **pulmonary** valve never works completely normally, and will leak to some extent. This leak is normally nothing to worry about, but there is a very small chance that some children might need surgery to replace or repair their valve in later life.

What is the risk of having another child with a congenital heart condition?

If you have one child with a **congenital** heart condition, there is around a 1 in 40 chance that if you have another child, they will have a heart condition too.¹ However, this risk may be higher (*or lower*) depending on the type of **congenital** heart condition your child has. Because your risk of having another child with congenital heart condition is higher than it is for other people, your doctor may offer you a special scan at an early stage in future pregnancies, to look at the baby's heart.

Do ask your midwife or GP for more information on having a scan earlier than usual. Do be aware that if you have more than one child with **congenital** heart condition, the specific condition may not always be the same.

Coping with everyday life

For information on the topics below, please visit **bhf.org.uk/congenital**

- Financial issues
- Low-income benefits
- Disability benefits
- Carer's Allowance
- Fares for visiting your child in hospital

The medical terms and what they mean

Cardiologist A consultant specialising in heart disease.

Catheter A fine, hollow tube.

Chromosomes Found in the nucleus of every cell in the body, chromosomes contain the genes, or hereditary elements, which establish the characteristics of an individual.

Congenital From birth.

ECG See electrocardiogram.

Echocardiogram An ultrasound scan used to produce pictures of the heart and blood vessels.

Electrocardiogram A recording of the electrical activity of the heart. Also called an ECG.

Genetic To do with the information that is passed from parents to children through genes in sperm and eggs.

Murmur An extra sound that is sometimes heard when listening to the heart through a stethoscope.

Paediatric To do with paediatrics – the study of children's diseases.

Pulmonary To do with the lungs.

Pulmonary stenosis When the pulmonary valve is very narrow.

Stenosis See pulmonary stenosis.

Valvuloplasty A procedure to stretch open a narrowed valve.

Valvotomy A surgical procedure to open a narrowed valve.

Ventricle One of the two lower chambers of the heart.

References

1. Gill HR, Splitt M, Sharland GK, Simpson JM. 2003. *Patterns of recurrence of congenital heart disease: An analysis of 6,640 consecutive pregnancies evaluated by detailed fetal echocardiography.* Journal of the American College of Cardiology; 42: 923-9.

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For more information and support about children and young people with a heart condition, visit [**bhf.org.uk/heart-health/children-and-young-people**](https://bhf.org.uk/heart-health/children-and-young-people)



**British Heart
Foundation**

For over 50 years we've pioneered research that's transformed the lives of millions of people living with cardiovascular disease. Our work has been central to the discoveries of vital treatments that are changing the fight against heart disease.

But cardiovascular disease still kills around one in four people in the UK, stealing them away from their families and loved ones.

From babies born with life threatening heart problems, to the many mums, dads and grandparents who survive a heart attack and endure the daily battles of heart failure.

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