British Heart Foundation

Understanding your child's heart

Aortic stenosis

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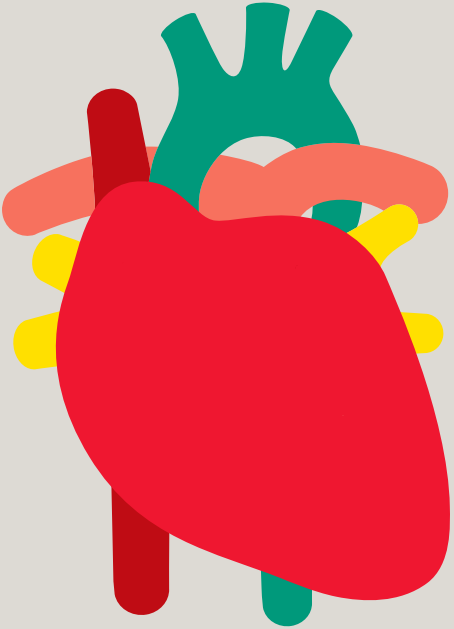


FIGHT FOR EVERY HEARTBEAT

bhf.org.uk

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 aortic stenosis is and how it is diagnosed
- how aortic 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.

Contents

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.

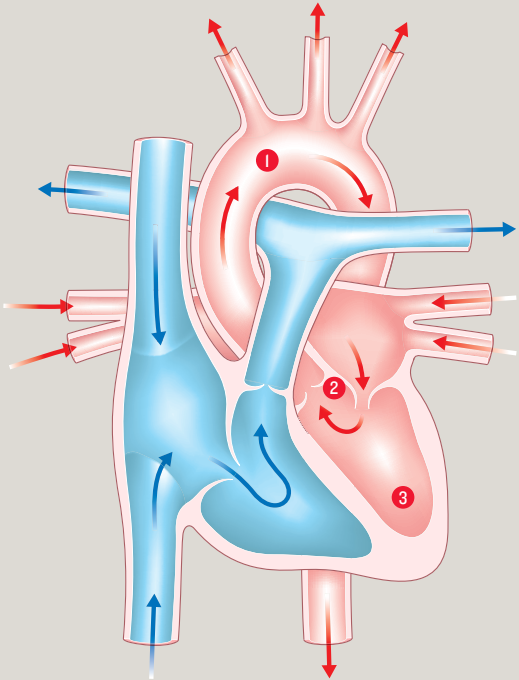
Each day 12 babies are diagnosed with a **congenital** heart defect in the UK. We continue to support research to improve the understanding, diagnosis and treatment of **congenital** heart conditions. For more information into our pioneering research visit **bhf.org.uk/research**

What causes a congenital heart condition?

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

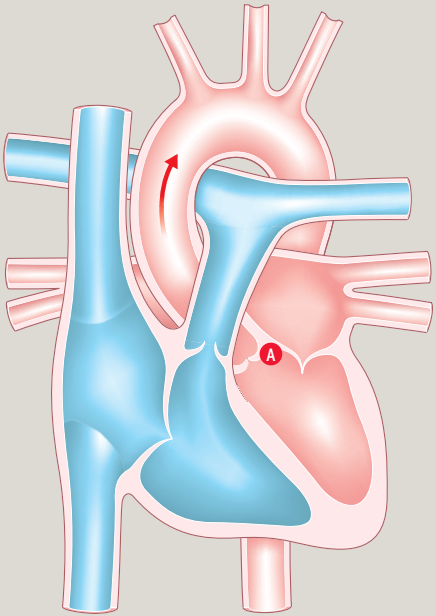
At the moment we don't fully understand why a baby's heart doesn't develop normally. But sometimes a **congenital** heart condition can be part of a syndrome which the baby is born with. *(A syndrome is a group of symptoms that commonly appear together as part of a condition).*

Normal heart



1. Aorta
2. Aortic valve
3. Left ventricle

Aortic stenosis



A Narrowed aortic valve

What is aortic stenosis?

Aortic stenosis means that the aortic valve cannot open fully. This is the valve which allows blood to flow from the heart to the main artery supplying the body – the **aorta**. *(See illustration on page 5)*.

When the aortic valve is narrow, the muscle of the left pumping chamber – the left **ventricle** – has to work harder than normal. This can lead to thickening of the muscle. The thicker the muscle becomes, the less efficient it is at pumping blood. If the narrowing is very severe, this can limit the amount of exercise or play your child can can manage.

It's important to understand that if left untreated, serious complications such as **heart failure** can occur, or in some rare cases sadly, could be fatal.

What are the symptoms of aortic stenosis?

Most children will not have any symptoms, the symptoms can include a lack of energy or being tired, or breathlessness when exercising or playing. Do be aware that in some rare cases, if the aortic stenosis is severe, episodes of fainting can occur.

For more information and support about growing up with a heart condition, visit **bhf.org.uk/heart-health/children-and-young-people**

What other conditions are associated with aortic stenosis?

Some children with aortic stenosis may have other heart abnormalities as well – for example, conditions affecting the **aorta** or the mitral valve. Your child's **cardiologist** will tell you if that is the case for your child, and will be able to give you more information about what this means.

How is aortic stenosis diagnosed?

In most cases, aortic stenosis is not diagnosed until after the baby is born, although some severe cases may be diagnosed before birth. The diagnosis is usually made because a heart **murmur** *(an extra sound from the heart)* is detected at birth or during a routine examination when the child is older.

Usually, the only test that is needed to make the diagnosis is an **echocardiogram**. This is an ultrasound scan of the heart. It won't hurt your baby at all.

How is aortic stenosis treated?

Most children with aortic stenosis will need surgery to replace the narrowed aortic valve. The age at which they will need the surgery varies from one child to another. The timing of surgery will depend on how narrow the valve is at birth, how much further it narrows as your child grows *(it almost always gets narrower with age)*, and how their heart is coping with the extra work.

Surgery is usually delayed as long as possible, because an artificial valve will not grow as your child grows and will need to be replaced again.

First, we describe two procedures – balloon **valvuloplasty** and **valvotomy** – which can stabilise your child's condition in the years before valve replacement.

Balloon valvuloplasty and valvotomy

If the valve is very narrow in early childhood, a procedure called a balloon valvuloplasty can be used to stretch open the narrowed valve. Your child will be given a general anaesthetic. A **catheter** *(a thin, hollow tube)* with a small collapsed balloon at its tip will be inserted into an artery in their groin. The **catheter** will be guided up into the heart and across the narrowed valve. Using X-ray, the balloon will be positioned in the aortic valve, and will then be inflated, stretching the valve open. The balloon will then be deflated and removed. Your child may have to stay in hospital for a few days after the procedure.

It's important to know that Balloon **valvuloplasty** doesn't make the valve normal and isn't always completely successful, but more often than not it helps to reduce the severity of the narrowing, helping to delay surgery. The procedure can be carried out more than once if necessary.

If a balloon **valvuloplasty** is not successful, or if your child is unsuitable for it, they may need to have a **valvotomy**.

This is a surgical procedure to open the narrowed valve. Your child will be given a general anaesthetic. The operation involves using a heart-lung machine to pump blood around the body while the heart is stopped briefly and the valve is cut open. Your child will need to stay in hospital for a few days after the operation.

Cutting the valve open will almost always make the valve leak. *(This is called aortic regurgitation, and can also happen with balloon valvuloplasty).* Unfortunately, it's impossible to predict how significant the leak will be, but if it's very severe, your child may need to have surgery to replace the valve.

What are the risks associated with balloon valvuloplasty and valvotomy?

Babies with very severe aortic stenosis may need treatment very early in life, which sadly does increase the fatality risk. Your child's **cardiologist** will discuss the risks with you, taking into account the specific condition of your child. In older children, the level of risk is much lower.

Surgery to replace the aortic valve

This open-heart operation involves using a heart-lung machine to pump blood around the body, while the heart is stopped briefly and the valve is replaced.

One option is to replace your child's aortic valve with an artificial mechanical one. This means that your child will need to take a drug called warfarin to reduce the risk of a blood clot forming across the metal valve. Be aware that this can prove problematic for girls, as it will make the planning and management of pregnancy complicated in their adult life.

The second option is to remove the natural valve in your child's pulmonary artery (the pulmonary valve) and use it to replace the narrowed aortic valve. A 'tissue valve' (usually from a pig or cow, or donated human valve) is then used to replace the pulmonary valve. This is known as a Ross procedure. However, the tissue valve in the pulmonary artery will not last

forever and will need to be replaced approximately every 10-15 years - which means further open-heart surgery. Some adults may be suitable for valve replacement by key hole catheter technique.

Deciding on the type of surgery is complicated and will vary from patient to patient. Your surgeon will discuss this in detail with you.

What are the risks associated with surgery to replace the aortic valve?

Replacing the aortic valve is a major operation, but the good news is the risk of fatality is very low. The risk varies depending on the exact nature of the problem, as well as the exact type of surgery used. Your child's surgeon will talk this through with you in detail.

What happens after surgery or Balloon valvuloplasty and valvotomy?

Most children can go home within a few days after surgery, although it may be longer than this if there are any complications.

You will need to take your child to the outpatients department within a few weeks to see the **cardiologist**, for a check-up.

What happens as my child grows up?

Most children and adolescents lead normal, active lives after surgery. However, replacement valves can develop problems over the years, so regular check-ups with a **cardiologist** are very important – not just in childhood, but throughout adult life too.

If a metal valve is used, your child will need to take warfarin. They will need to have regular blood tests *(weekly to monthly)* to make sure they are getting the correct dose of the drug – a dose that is safe yet effective enough to reduce the risk of blood clots forming.

For more information on **warfarin**, see our website **bhf.org.uk**

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

Aorta The main artery of the heart. It supplies oxygen-rich blood to the body.

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.

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.

Heart failure When the heart becomes less efficient at pumping blood around the body.

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.

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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Notes

Notes

For more information and support about children and young people with a heart condition, visit **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.

Join our fight for every heartbeat in the UK. Every pound raised, minute of your time and donation to our shops will help make a difference to people's lives.

Text FIGHT to 70080 to donate £3

FIGHT FOR EVERY HEARTBEAT

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