



Understanding your child's heart

Aortic stenosis



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About this booklet

This booklet is written for the parents of babies and children with aortic stenosis, and for their relatives and friends. It explains:

- what aortic stenosis is and how it is diagnosed
- how aortic stenosis is treated
- the benefits and risks of treatments
- how to cope as a parent of a child with aortic stenosis
- where to go for more support.

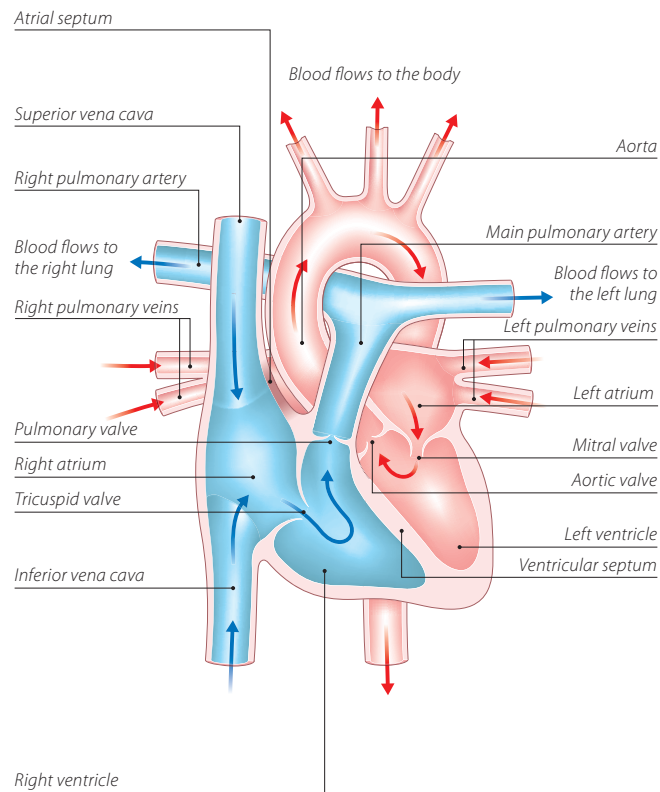
This booklet does not replace the advice that doctors or nurses may give you, but it should help you to understand what they tell you.

The normal heart

The heart is a muscular pump which pumps blood through the body and lungs. There are four chambers in the heart. The two upper ones are called the right atrium and left atrium. These are separated by a wall called the atrial septum. The two lower chambers are called the right and left ventricles, and are separated by a wall called the ventricular septum. See the illustration opposite.

On each side of the heart, blood passes from the atrium, through a heart valve – the tricuspid valve on the right, and the mitral valve on the left – into the ventricle. The ventricles are the main pumping chambers of the heart. Each ventricle pumps blood out into an artery. The right ventricle pumps blood – blue in the illustration – into the pulmonary artery (the blood vessel that takes blood to the lungs). The left ventricle pumps blood – red in the illustration – into the aorta (the blood vessel that takes blood to the rest of the body). Blood flows from the right side of the heart, through the pulmonary valve into the pulmonary artery, and then to the lungs where it picks up oxygen. The oxygen-rich blood flows back into the left side of the heart through the pulmonary veins. The left ventricle then pumps the oxygen-rich blood out of the heart through the aortic valve and into the aorta, and all around the body. The blood then returns to the right side of the heart through two main veins – one from the upper body (superior vena cava), and the other from the lower body (inferior vena cava).

The normal heart



What is congenital heart disease?

Congenital heart disease is an abnormality of the heart that developed in the womb. In some cases, congenital heart disease is diagnosed when the baby is still developing in the womb, but in most cases the problem is not discovered until after the baby is born. There are lots of different types of congenital heart disease. **Aortic stenosis** is one type.

What causes congenital heart disease?

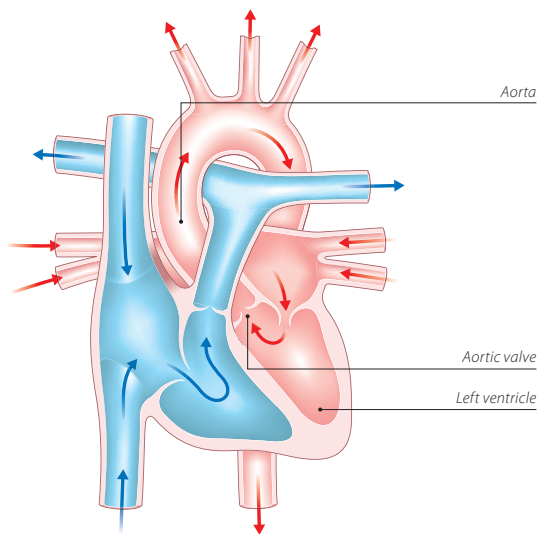
We know that, in most cases of congenital heart disease, something has gone wrong in the early development of the fetus, at the very early stages of the pregnancy. In most cases, we don't understand why the baby's heart did not develop normally. In some cases, congenital heart disease can be part of a syndrome that the baby is born with. (A syndrome means a group of symptoms that appear together.)

Why me? Why my child?

It is not unusual for parents of children with congenital heart disease to blame themselves or to be angry. Anger, disappointment, fear and guilt are all normal feelings to have when you are told that there is something wrong with your child. At first it may be difficult to cope with and it can take a while for the news to sink in. Many pregnant women or mothers, and their partners, ask themselves what they did during their pregnancy that could have caused their baby's heart to develop with heart disease. But the reality is that it can happen to anyone. In fact congenital heart disease happens in about 7 in every 1,000 pregnancies.¹ For more than half of these children, the heart disease is only a minor problem which either doesn't need any treatment, or which can be successfully corrected with surgery.

What is aortic stenosis?

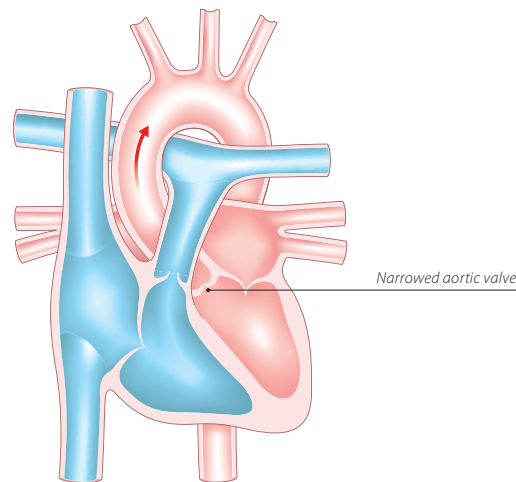
The normal heart



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 the illustrations above.

When the aortic valve is narrow, the muscle of the left pumping chamber – the left ventricle – has to work harder

Aortic stenosis narrowed aortic valve



than normal. This can lead to thickening of the muscle of the left ventricle. The thicker the muscle becomes, the less efficient it is at pumping blood in the long term. If the narrowing is very severe, the heart cannot pump normally and this can limit the amount of exercise or play your child can do. If this is left untreated, serious complications such as **heart failure** can occur, or in some cases the child may even die.

What are the symptoms of aortic stenosis?

The symptoms vary depending on how narrow the aortic valve is. Most children will not have any symptoms, but the symptoms that can occur include a lack of energy or being tired, or breathlessness when exercising or playing. In some rare cases, if the aortic stenosis is severe, fainting can occur. If your child does faint, you should seek specialist advice.

What other conditions are associated with aortic stenosis?

Some children with aortic stenosis may have other heart abnormalities as well – for example, affecting the aorta or the mitral valve. Your cardiologist will tell you if that is the case for your child.

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 is very similar to the scans that are carried out during pregnancy. It doesn't hurt your child at all.

How is aortic stenosis treated?

Most children with aortic stenosis will need **surgery to replace the narrowed aortic valve** at some stage in their lives. The age at which they will need the surgery varies from one child to another. Some children will need this surgery in early childhood, while others can wait until they are young adults or in middle age. The timing of the surgery depends on how narrow the valve is at birth, how much further it narrows as the child grows (it almost always gets narrower with age), and how the heart is coping with the extra work. The 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. We explain more about the surgery and the various types of valves on page 15. First, we describe two procedures – **balloon valvoplasty** and **valvotomy** – that can be used to stabilise your child's condition in the years before valve replacement.

Balloon valvoplasty and valvotomy

If the valve is very narrow in early childhood, a procedure called a **balloon valvoplasty** can be carried out to stretch open the narrowed valve. Your child will be given a general anaesthetic. A catheter (a fine, hollow tube) with a small collapsed balloon at its tip, is inserted into an artery in the groin. It is guided up into the heart and across the narrowed valve. Using X-rays for guidance, the balloon is positioned in the aortic valve, and is then gently inflated, stretching the valve open. The balloon is then deflated and removed.

Your child may have to stay in hospital for a few days after the procedure. You will need to take your child to the outpatients department again to see the paediatric cardiologist, for a check-up.

Balloon valvoplasty does not make the valve normal and it does not always work, but in many cases it can help to reduce the severity of the narrowing, helping to delay surgery. The procedure can be carried out more than once if necessary and if your child is suitable for it.

If a balloon valvoplasty is not successful, or if your child is unsuitable for it, he or she may need to have a **valvotomy**. A valvotomy 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. You will need to take your child to the outpatients department again to see the paediatric cardiologist, for a check-up.

Cutting the valve open will almost always make the valve leak. (This is called **aortic incompetence** or **regurgitation**.) It is impossible to predict how significant the leak will be, but if it is very severe, your child may need to have surgery to replace the valve in childhood after all.

What are the risks associated with balloon valvoplasty and valvotomy?

The risks of both these procedures depend very much on the age at which treatment is needed. Babies with very severe aortic stenosis may need treatment very early in life. When treatment is needed very early in life there is a significant risk that the baby might die. Your cardiologist will discuss the risks with you, taking into account the specific problems of your child. In older children, the chances of survival are much higher, with about 95 out of 100 children being alive a year after treatment.

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. There are several different types of surgical treatment for aortic stenosis. We describe two options briefly below. The doctors will discuss the treatments with you in more detail.

One option is to replace your child's natural aortic valve with an artificial metal 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. This can prove problematic for girls, as it will make the planning and management of pregnancy complicated in their adult life. For more information on warfarin, see our website bhf.org.uk

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 a 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 in the future – which means further open-heart surgery.

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 around 98 in every 100 patients survive overall.² The risk varies depending on the exact nature of the problem in the individual patient, as well as on the exact type of surgery used. Your surgeon will discuss this with you in detail.

What happens after surgery?

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 again, 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**. He or she 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

The specialist centre for congenital heart disease

Your child will continue to have check-ups at a specialist centre for congenital heart disease throughout their life. Up to the age of about 16, he or she will go to a centre which may be called a 'specialist centre for children with congenital heart disease', or a 'specialist paediatric centre'. (Paediatric means to do with children.)

If the specialist centre is quite a distance from your home, it may be possible to make arrangements for your child to be looked after under a 'shared care' system. This means that your child will be looked after locally, but will go to the specialist centre for specialised treatment. (See page 22 for information about claiming travel expenses for visits to the specialist centre.)

The specialist team

At the specialist centre, a large multidisciplinary team of people will be involved in caring for your child and your family's needs. (Multidisciplinary means that it includes several different types of health professionals.) The team usually consists of:

- a consultant paediatric cardiologist
- a specialist registrar (a doctor who is specifically training in children's heart problems)
- a cardiac nurse specialist or cardiac liaison nurse
- a consultant paediatric surgeon or paediatric cardiothoracic surgeon
- cardiac technicians

- a paediatric physiotherapist
- a paediatric dietitian
- ward-based paediatric nurses
- a social worker
- an occupational therapist
- a speech and language therapist, and
- a play specialist.

All of these people are involved in planning the care for each patient. You probably won't need to see all of them, but it is important to know who is there and available to help you with any problems you may come across. For example, the social worker can be a very useful source of information on what benefits you might be entitled to claim, and whether you can claim your travel expenses for visiting your child in hospital, or for visits to the specialist centre. (For more on this, see page 22.)

The specialist centre will also have access to psychology services which you can use to help your child or your family if you need help at difficult times.

Specialist adult congenital heart disease centres

When your child grows into adulthood, it is important that he or she carries on going to a specialist centre for check-ups. There are several specialist centres in the UK for adults with congenital heart disease. Your child's care will be transferred to an adult specialist centre usually at around the age of 16. These centres usually have a multidisciplinary team with the same mix of professionals as in the children's specialist centre (see

page 18). When your child is nearing adulthood, your specialist paediatric centre will start preparing you and your child for the move to the adult specialist centre, to make the transition as smooth as possible.

Coping with everyday life

Having a child with a heart condition in hospital can be very difficult for a number of reasons. You are having to care for your child in hospital, and may be anxious about the treatment your child has to have or how well he or she is recovering. You might also be worrying about being away from home, or about your other children and who's looking after them, or about your work or your finances.

Hospital staff recognise the stress that you and your family might be under. Ask the nurse who is looking after your child about the support services available within the hospital – such as psychology services, welfare rights advisers and social workers. Below we give a brief guide to the benefits and help you may be able to get.

Financial issues

Financial problems may arise because you need to stay in hospital with your child. The hospital may be a long way from home, and you may not know how long you will need to stay there for. This can affect your finances, as you may have to take time off work and will have extra costs such as travel expenses and buying meals while in hospital. This can be an extra anxiety, and difficult to cope with.

If you are worried about your finances, it is important to discuss your situation with a hospital social worker or cardiac liaison nurse, or with the Citizens Advice Bureau. They may be able to advise you on the benefits you can claim. Also, an organisation called Working Families can give you advice on the phone – on 0800 013 0313 – about your rights as an employee if you need to take time off work to be with your child.

Low-income benefits

Benefits for people on a low income include Income Support, housing and council tax benefits, and Tax Credits. If your income goes below a certain amount, you may be able to claim benefits. However, you have to meet certain criteria in order to get these benefits. The criteria vary from one benefit to another, so you should get specialist advice from a hospital social worker, Citizens Advice Bureau or Jobcentre Plus.

Disability benefits

Some children with congenital heart disease will qualify for a **Disability Living Allowance** (DLA), but most will not. Ask the specialist nurse or social worker for advice. If your child needs extra care because of his or her condition, you may be able to apply for this benefit. There is a mobility and a care component to the benefit. It can be difficult to get Disability Living Allowance for a child. You will need to show that your child needs more attention or supervision than other children of the same age. To get a claim form for Disability Living Allowance, call 0800 88 22 00. Or you can get a form from your GP or your local Jobcentre Plus office, or claim online at www.direct.gov.uk

Carer's Allowance

You may be entitled to *Carer's Allowance* if your child is awarded the middle or higher care component of Disability Living Allowance. However, you will only get Carer's Allowance if your earnings are below a certain limit. To get a claim form for the Carer's Allowance, call 01253 856 123. Or you can get a claim form from your local Jobcentre Plus, or claim online at www.direct.gov.uk

Fares to hospital

If you get Income Support, or have a Tax Credit exemption card, or are assessed as being on a low income, you are entitled to get reimbursed for your fares to hospital appointments and inpatient treatment. If you think you might qualify because of your income, complete a form HC1. To get one, call 0845 850 1166. The forms are usually also available from Jobcentre Plus offices and NHS hospitals.

Fares for visiting your child in hospital

If you are getting Income Support, income-based Jobseeker's Allowance, or Pension Credit, you may be able to get help towards the cost of fares for hospital visiting, from the Social Fund. You should apply for a Community Care Grant on form SF300, which is available from your local Jobcentre Plus office or from the website of the Department for Work and Pensions at www.dwp.gov.uk. However, these payments are not given in all cases.

The benefits system is very complex, so it is important to get specialist advice on what you may be entitled to, from the hospital social worker or Citizens Advice Bureau.

General advice for the future

Endocarditis

Everyone who has aortic stenosis is at risk of getting *infective endocarditis*, both before and after surgery or treatment. Infective endocarditis is a rare condition where the inner lining of the heart, most commonly one of the heart valves, becomes infected.

Infective endocarditis is a serious condition which can be life-threatening if it's not treated quickly. Nowadays, if it is diagnosed early, most people with it will recover well with antibiotic treatment, although some damage may occur to the heart valves as a result of the infection.

Endocarditis is caused by a bacterium, or (rarely) another type of infective organism that is in the bloodstream, which settles onto the abnormal structure or defect in the heart. Although it is not possible to prevent all bacteria from getting into the bloodstream, there are some things your child can do, as he or she grows up, to reduce the risk of getting endocarditis:

- Maintain good oral hygiene and have regular check-ups with a dentist
- Avoid body piercing and tattooing
- Never inject recreational drugs.

If your child develops flu-like symptoms with a temperature **which persist for over a week**, you should visit your GP as your child may need a blood test. Make sure that the GP knows that your child is at increased risk of getting endocarditis. You can

do this by showing the GP an *Endocarditis warning card*. You can get this card from the British Heart Foundation (BHF) by calling either the Heart HelpLine on **0300 330 3311** (local rate number) or the BHF Orderline on **0870 600 6566**.

Pregnancy

If you have a daughter with congenital heart disease, you need to be aware that pregnancy could carry risks to both the mother and the baby. So, when your daughter gets older, it's particularly important that she avoids having an unplanned pregnancy. You will need to discuss this with your daughter in whichever way you, as a parent, think is appropriate for her. If your daughter wants to have a baby, it's best that she speaks to her cardiologist about it first, so that the pregnancy can be planned for when your daughter's heart condition is most stable.

People who have congenital heart disease themselves have an increased risk of having a child with a heart problem. This applies to both males and females. You can discuss this with your cardiologist. Early scans in pregnancy can be arranged to look for heart disease in the baby.

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

If you have one child with congenital heart disease, there is about a 1 in 50 chance of having another child with congenital heart disease.³ However, this risk may be higher (or lower) depending on the type of congenital heart disease your child

has. Because your risk of having another child with congenital heart disease is higher than it is for other people, you may be offered a special scan at an early stage in future pregnancies, to look at the baby's heart. Ask your midwife or GP for more information on having a scan earlier than usual.

If you have had two children with congenital heart disease, the risk of having another child with heart disease rises to about a 1 in 10 chance. This may sound like a high risk, but you still have a much better chance of the baby's heart being normal than abnormal. If there is a recurrence, the heart disease may not always be of the same type.³

Support groups

The following support groups may be able to offer you further information, advice and support:

Action for Sick Children

36 Jacksons Edge Road
Disley, Stockport SK12 2JL
Phone: 0800 074 4519
Website: www.actionforsickchildren.org

Children's Heart Federation

Level One, 2-4 Great Eastern Street
London EC2A 3NW
Phone: 0808 808 5000
Website: www.childrens-heart-fed.org.uk

About the British Heart Foundation

The British Heart Foundation is the nation's heart charity, saving lives through pioneering research, patient care and vital information.

What you can do for us

We rely on donations to continue our vital work. If you would like to make a donation to the BHF, please ring our Supporter Services team on **0844 847 2787** or contact us through our website at **bhf.org.uk/donate** or send it to us at the address on the back cover.

For more information

British Heart Foundation website

bhf.org.uk

For up-to-date information on heart disease, the BHF and its services.

Heart HelpLine

0300 330 3311 (local rate number)

For information and support on anything heart-related.

Booklets

To order any of our booklets

- call the BHF Orderline on **0870 600 6566**
- email **orderline@bhf.org.uk** or
- visit **bhf.org.uk/publications**

You can also download many of our publications from our website.

For information on other BHF booklets, and on DVDs and videos, ask for a copy of the *Heart health catalogue*.

Understanding your child's heart series

This booklet is one of the booklets in the *Understanding your child's heart* series. For a full list of the booklets available in this series, see our website **bhf.org.uk** or call the Heart HelpLine on **0300 330 3311** (local rate number).

Operation Fix-it

A short story book about eight-year-old Tom's experience in hospital for a heart operation. Prepares children for their hospital visit in an interesting and sometimes humorous way.

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3. 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.

Contacts

Use this page to keep contact details of the health professionals who are caring for your child.

Paediatric cardiologist

Paediatric nurse

Paediatrician

Social worker

Surgeon

Others

Hospital visits

Use this page to write down the dates of your hospital visits.

The medical terms and what they mean

aorta	The main artery of the heart. It supplies oxygen-rich blood to the body.
atrial	To do with the atrium. (See below.)
atrium	One of the two upper chambers of the heart.
cardiac	To do with the heart.
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 <i>electrocardiogram</i> .
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.
endocarditis	Infection of the lining of the heart or its valves.

genetic	To do with the information that is passed from parents to children through genes in sperm and eggs.
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.
septum	The wall that keeps the right and left sides of the heart separate.
valvoplasty	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.
ventricular	To do with the ventricle or ventricles. (See above.)

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Have your say

We would welcome your comments to help us produce the best information for you. Why not let us know what you think? Contact us through our website at [bhf.org.uk/contact](https://www.bhf.org.uk/contact) Or, write to us at the address on the back cover.

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- Dr John Gibbs, Consultant Paediatric Cardiologist, Leeds General Infirmary.

HEART HELPLINE

For information and support on anything heart-related



0300 330 3311

local rate number



bhf.org.uk

Phone lines open 9am to 6pm Monday to Friday



British Heart Foundation

Greater London House
180 Hampstead Road
London NW1 7AW
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Fax: 020 7554 0100
Website: bhf.org.uk